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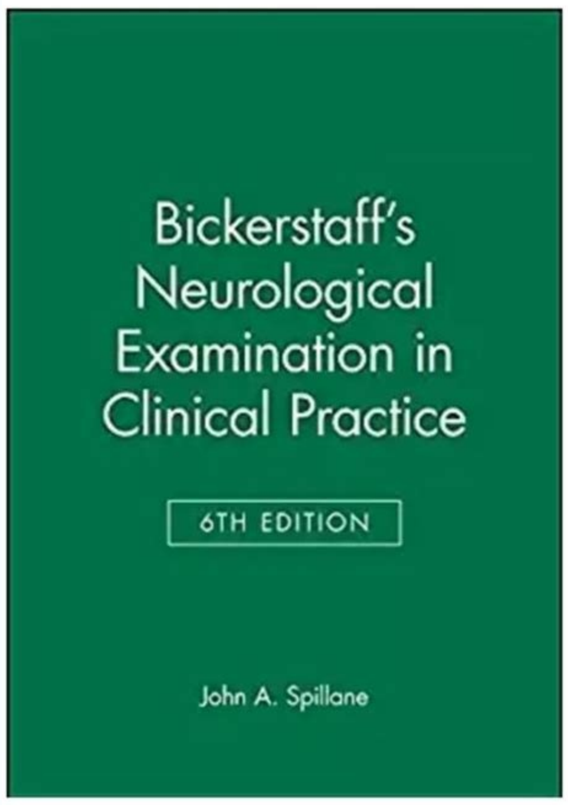
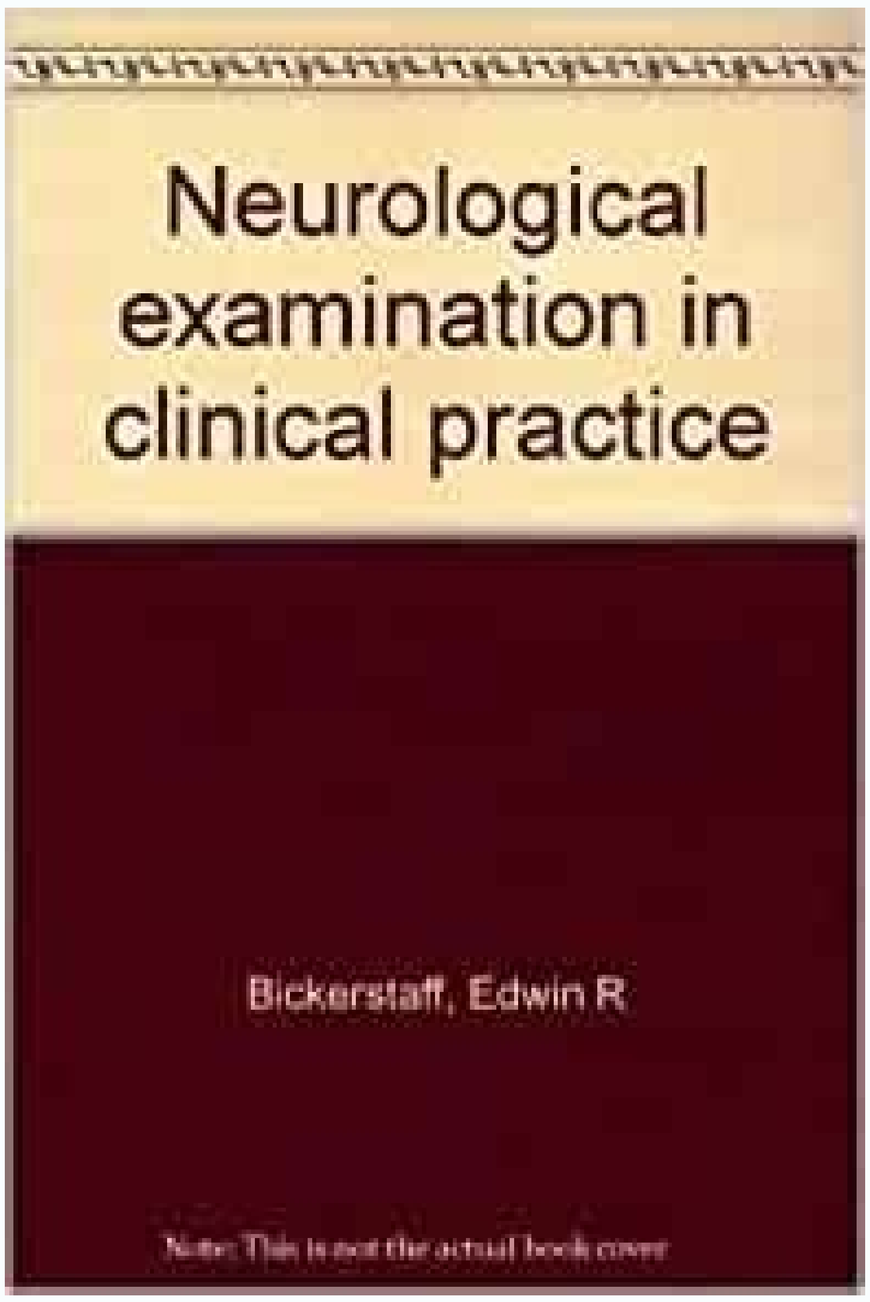

Bickerstaff's
Neurological
Examination in
Clinical Practice
6TH EDITION
John A. Spillane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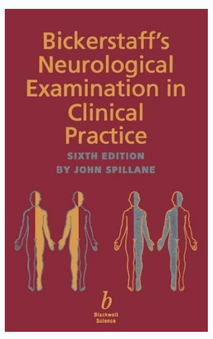
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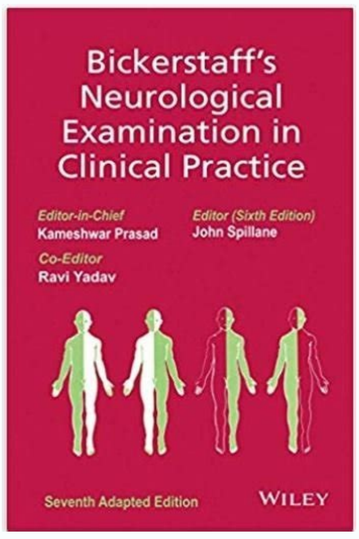
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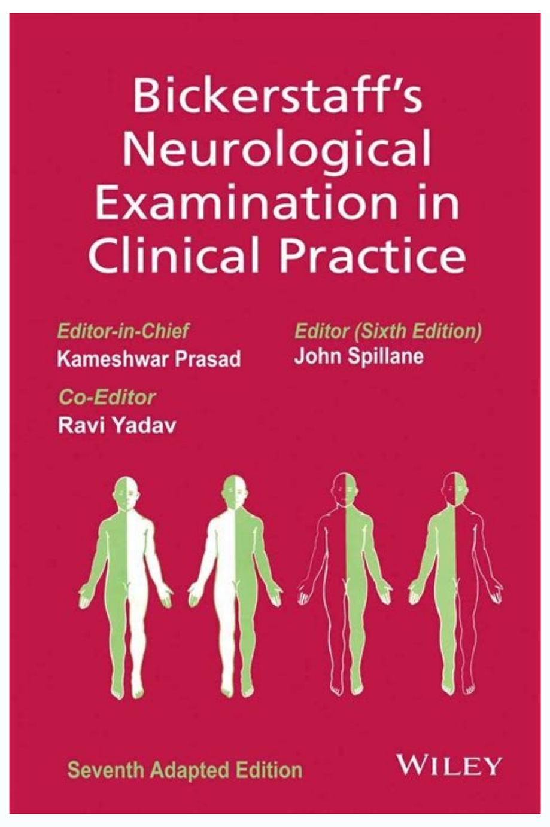

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Ltd. www.wileyindia.com Printed at: SDR Printers Contents Preface, ix Preface to the Sixth Edition, xi Part 1: The Introductory Stages, 1 Chapter 1 Approaching a neurological problem, 3 Chapter 2 Equipment, 6 Chapter 3 The history, 8 Chapter 4 First impressions, 13 Chapter 5 The general physical and mental examination, 20 Part 2: The Cranial Nerves, 41 Chapter 6 The first cranial nerve: the olfactory nerve, 43 Chapter 7 The second cranial nerve: the optic nerve, 46 Chapter 8 The third, fourth and sixth cranial nerves: the oculomotor, trochlear and abducent nerves, 62 Chapter 9 The fifth cranial nerve: the trigeminal nerve, 84 Chapter 10 The seventh cranial nerve: the facial nerve, 91 Chapter 11 The eighth cranial nerve: the auditory nerve, 99 Chapter 12 The ninth and tenth cranial nerves: the glossopharyngeal and vagus nerves, 109 Chapter 13 The eleventh cranial nerve: the accessory nerve, 115 Chapter 14 The twelfth cranial nerve: the hypoglossal nerve, 119 vi Contents Part 3: The Motor System, 123 Chapter 15 Muscle bulk, 125 Chapter 16 Muscle tone, 134 Chapter 17 Muscle power, 140 Chapter 18 Posture, stance, spinal movement and gait, 161 Chapter 19 Involuntary movements, 169 Part 4: The Sensory System, 185 Chapter 20 Basic principles for examination of sensation, 187 Chapter 21 Pain, touch and temperature, 194 Chapter 22 The proprioceptive sensations, 198 Chapter 23 Stereognosis, discriminative sense and graphaesthesia, 202 Chapter 24 Common patterns of abnormal sensation, 205 Part 5: The Motor-Sensory Links, 213 Chapter 25 The reflexes, 215 Chapter 26 Coordination, 234 Part 6: Examinations of Particular Difficulty, 243 Chapter 27 Tire unconscious patient, 245 Chapter 28 Disorders of speech, 263 Chapter 29 Apraxia, 274 Chapter 30 Agnosia and disorders of the body image, 277 Chapter 31 The autonomic nervous system, 283 Contents vii Part 7: The Investigation of Neurological Problems (Excluding Neuroradiology and Imaging), 291 Chapter 32 Towards a balanced attitude (some introductory observations), 293 Chapter 33 General medical investigations, 297 Chapter 34 Tire cerebrospinal fluid, 300 Chapter 35 Tine clinical value of electroencephalography, 311 Chapter 36 Peripheral electrophysiology, 319 Chapter 37 Biopsy, 326 Part 8: Localization of Lesions, 331 Chapter 38 Localization of lesions affecting various parts of the nervous system, 333 Part 9: Appendices, 349 Appendix A Checklist for history taking and neurological examination for undergraduates, 351 Appendix B Recording the neurological examination, 364 Appendix C Screening neurological examination, 367 Appendix D A suggested scheme for the examination of higher cerebral function, 370 Index Preface Thousands of students of medicine and neurology around the world have read Bickerstaff's Neurological Examination in Clinical Practice. As students of medicine and neurology, we considered this very popular, and much praised book, as a bible for the techniques of neurological examination. So, when we received an invitation to edit the seventh edition of the book, we considered it a great hon our indeed. We were also conscious of our responsibility to retain the inherent character and simple language of the book. Neurological examination often presents a challenge to students, residents or practitioners. Medical students often fear coming across a neurology case in their examinations and general practi tioners and physician specialists promptly refer a neurology case to a neurologist if the presentation demands neurological exam ination. This is because there is a common perception that neu rological examination is lengthy, cumbersome, time-consuming and difficult. This book is intended to dispel this perception. It describes techniques and examinations in an easy, clear way with free-flowing language. The main target audience of the book remains trainees in neu rology and general medicine, but this edition is also intended for undergraduate medical students for whom we have added an appendix containing a brief checklist of the essential items under graduate students will need for history taking and neurological examination. Continuing with the thoughtful omission of some chapters such as Paediatric Neurology in the fifth edition and Indi cation for Full Investigation in sixth edition, we have, for the sev enth edition, omitted the chapter on Neuroradiology and Imaging, since to adequately cover the advances in neuroradiology would have required expansion of the chapter far beyond the scope of the book. Instead, we have added a section on Tests of Lobar Functions in Chapter 5 and a new chapter on Localization - both very useful additions to the book. The section on lobar functions will be useful for trainees in neurology when dealing with patients of neurodegenerative diseases or diseases affecting higher mental functions. The chapter on localization will be helpful for all stu dents in determining the site of a lesion after they have gathered the clinical findings following the neurological examination. We have also added many new figures and changed the illustrations to make them more attractive. We have added a 'Causes List' in IX x Preface many chapters to help students generate diagnostic hypotheses in patients with different clinical findings. We hope students will also find the video clips (represented by video icon G] at relevant places in the book) in the free companion CD-ROM to illustrate various movement disorders useful. We hope these changes will enhance the usefulness and popular ity of an already much-praised book and expand the target student base. Grateful thanks to Dr. Vallika Devi for her hard work, patience, sincerity and flexibility. Our thanks are also due to the production team, Wiley India, for the excellent work and Dr. Bharathi for ensur ing good quality neuroanatomy figures. We are also thankful to the . Department of Mental Health Education, NIMHANS, Bengaluru for the help in the production of both photographs and videos. We enjoyed editing the book and hope that students, both under graduates and postgraduates, will find the book more useful and enjoyable to read than previous editions. Kameshwar Prasad Ravi Yadav April 2013 Preface to the sixth edition The date recorded inside my first copy of Neurological Examination in Clinical Practice reminds me of its purchase in 1973 when just embarking upon the initial nervous steps into neurology. Little did I anticipate the possibility let alone the reality, of succeeding Dr Bickerstaff at the Midland Centre nor the double honour of an invitation to join him for the fifth edition of his much praised book. A few years on and Edwin has graciously handed over the reins for this sixth edition. The task remains unchanged - to outline the techniques of neu rological examination, the principal methods of investigation and to suggest how the latter may best applied. The book was never intended to be a comprehensive text of neurology, nor of neurological diagnosis. The temptation, therefore, to expand this edition along those lines has been firmly resisted. Many older methods of investigation have been superseded, and are there fore omitted. To have properly updated the chapter on 'Indica tions for full investigation' in a way to adequately complement the advances in neuroradiology and imaging, alone, would have required an expansion in the text far beyond the above declared aims. So, rather than change the character of the book that chapter has been omitted. The wish has been to modernize the text and illustrations, as required, but to maintain the overall balance of the book, in particular so that it remains affordable for those to whom it has always been directed, trainees in neurology and gen eral medicine. Grateful thanks are due to Dr David Yates for providing the new CT and MRI scans, to the Oxford University Press for permission to reproduce four more illustrations originally published in The Atlas o f Clinical Neurology, additional to those already acknowledged in the preface to the fifth edition. This applies to Figs 14.1d, 15.1,15.2 and 15.3a. The collaboration of the Department of Medical Pho tography of Sandwell Hospital NHS Trust is gratefully acknowl edged for provision of the new illustrations of the limb reflexes. The obliging subject, our registrar, prefers to avoid formal identifi cation lest this should adversely affect his career! To Stuart Taylor, commissioning editor at Blackwell Science, grateful thanks for the help and encouragement in planning this new edition and thanks xii Preface to the sixth edition also to jane Andrew for guiding it through production. Filially, for sacrifice beyond the call of duty when typing the manuscript (bilat eral carpal tunnel syndrome) very many thanks to my secretary Mrs Jacqui Penk. Birmingham, 1995 J ohn spillane Part 1 The Introductory Stages 1 Approaching a neurological problem Solving a neurological problem can be the most fascinating exer cise in medical detection and logical deduction in the whole clinical field, yet there can be few branches of medicine, excluding, per haps, dermatology, in which practitioners from other disciplines feel more un-informed, ill-trained and even impotent. This is par ticularly disappointing because, at least in the UK, the majority of patients with neurological symptoms do not, in the first place, come into the hands of a neurologist or special unit. In India, how ever, the patient may come to a neurologist directly, although the majority sees general practitioners at the first instance. The general practitioner and general physician still see a vast amount of neuro logical material and will continue to do so until there is a significant expansion in the number of neurologists. Sadly, the undergraduate curriculum in many medical schools does not allow anything like sufficient time in the neurological unit. Neurological symptoms, after all, lead to at least 10% of consultations with general practi tioners and about 20% of all acute medical admissions. In order to compensate for these shortcomings, the non-neurologist and stu dent must develop an organized line of thought in approaching each problem so that the pieces of what, after all, is a diagnostic jigsaw can be specifically looked for, and, if found, fitted together to form a recognizable portrait of a disease. Those who feel they are doomed to fail diagnostically display a method of approach that soon rubs off on the patient, who loses confidence, and later may repeat a phrase so often heard in neurological consultations - 'the other doctors didn't seem to know where to begin'. Each case poses and demands an answer to four vital questions: 1 Is there a lesion of the nervous system present? This is deter mined by analysis of the history and physical examination. 2 Where does this lesion lie in the nervous system? Is it possible to locate it at one site, or must multiple sites be involved? This can be worked out only by relating the symptoms and signs to a basic knowledge of neuroanatomy - but there is no need to be scared by this - only rarely does this have to be very detailed. 3 What pathological conditions are known to be capable of caus ing lesions at this site (or sites)? 4 In this particular individual, first from careful analysis of the history and examination, and later by intelligent use of the 3 Part 1 The introductory stages ancillary services, which of these suspected conditions is most likely to be present? Each step taken in the study of the case, from the first inter view onwards, should aim at answering each of these questions in turn. Each examination or investigation should have behind it the planned purpose of including, or excluding, one specific member of the 'shortlist' of suspected conditions. It is always the failure to have such an organized plan of approach that makes neurological problems so artificially difficult. Routine steps must, of course, be followed, but blind routine and blunderbuss investigations, 'just so as not to miss anything', show that such a plan has not existed. The more experienced one becomes, the more one recognizes 'pat terns' of disease, and it may appear to younger doctors that diag noses are often arrived at by inspired guesswork in which many corners have been cut. In fact it is that the experienced clinician moves that much more quickly over these basic questions while they still remain the foundation of his decision. Diagnosis purely by comparison with previous cases is reserved for those who are very experienced, remember their cases truly accurately and fol low them carefully and critically, and they are rarities. The begin ner will come nearer to diagnostic accuracy by logically reasoning out each step along the lines suggested. There is, however, another psychological barrier to be overcome. The view still widely held is that the exact solution of a neurologi cal problem does not matter all that much, as it will be of academic interest only, there being no useful treatment. Nowadays, this is just arrant nonsense. It may be true that we know of no treatment for motor neuron disease; that we cannot cure the hereditary atax ias and that we have not yet found a reliable method of prevent ing relapses in multiple sclerosis, but, contrary to many people's belief, these occupy a relatively small part of the neurologist's time. Think for a moment of the transformation in the last 40 years in the treatment of epilepsy, meningitis, neurosyphilis and deficiency neuropathies; of the influence of immunosuppression in myasthe nia gravis, not to mention plasma exchange and immunoglobulin treatment in acute demyelinating neuropathies; of the help avail able in multiple sclerosis with the advent of interferon; of the con tinuing progress in the drug therapy of migraine, parkinsonism, many pain syndromes and dystonic disorders; of the enormous advances of neurosurgery in the fields of stereotaxis and 'radio surgery' in extra-pyramidal disease, some types of epilepsy and all forms of cerebral and spinal compression. This list includes only a few examples of the way in which neurological practice offers possibilities for therapy which compare very favourably with all 4 Chapter 1 Approaching a neurological problem other branches of medicine. Complete eradication of the pathologi cal process may not be achieved, but this unhappily also applies in medicine as a whole. If, however, an exact diagnosis is not reached, it is important that this be admitted, and that the patient does not become firmly labelled with one of the differential possibilities (such as, for instance, multiple sclerosis), for this may result in any and every future neurological event being accepted as 'just another symptom of the old trouble', when in fact these are new develop ments giving the essential clue to the diagnosis of a much more remediable condition. Finally, remember that the solution of a neurological problem takes time. It cannot be rushed, and examiners must never allow their approach to be influenced by exhortations from optimis tic colleagues to 'just glance at this case while passing', or to 'just run over the nervous system; it won't take five minutes'. It will. It always does, and so it should. No one can expect to go through their career and to be right all the time, but most errors arise from inadequate taking of history and inadequate physical examination - particularly the inadequate history. The physician who is careful with both will rarely have difficulty in deciding which cases come into the category where organic neurological disease is a serious possibility, and then go forwards to appropriate investigation. If the results of any investi gations do not alter the management of the problem, then they have not been appropriate. 5 2 Equipment The instruments required for bedside neurological examination are relatively simple and, with one exception, inexpensive (Fig. 2.1). There are, however, a few points of guidance which may help those planning their equipment. The ophthalm oscope - auroscope A simple instrument is quite adequate provided it gives a steady, even, white disc of bright light shining in the plane of the exam iner's visual axis when properly and comfortably held. One may improvise many things for neurological examination, but nothing will replace a good ophthalmoscope. The torch Pocket-size torches giving a fine bright beam are better than the diffuse light of the larger variety. The percussion hammer The handle should be long and flexible, the ring of thick resilient rubber, without a heavy centre. The pins Sharp mapping pins with red or white heads are also useful for testing visual fields. Fig. 2.1 Instruments required for neurological examination at the bedside. 6 Chapter 2 Equipment ti he two-point discriminator Dealers may call this an aesthesiometer. Its points must be blunt, and besides its primary purpose, it can be used for testing ocu lar movement and the superficial reflexes, while the prong can be inserted under a plaster case to test the plantar reflex - a not uncommon problem to be faced. The stethoscope Its end should be adapted to fit closely to the skull or orbit in order to hear intracranial bruits. Optional extras A portable, reduced-size Snellen's chart or Rosenbaum's chart fits neatly into a briefcase. Two or three small bottles for testing smell will obviate a time-wasting search on a general or orthopaedic ward. A striped tape to test for optokinetic nystagmus is a useful adjunct. The exa onin at ion couch This should be warm and securely covered so that the patient is comfortable and not afraid of slipping, for this maintains muscu lar tension. Its headpiece should be adjustable and its height such that one can reach easily over an obese patient's stomach and yet not have to kneel to examine his legs. It should not stand opposite bright light and it should be possible to reach both sides easily. In developing countries, the examination couch may not be avail able in which case a screening neurological examination may need to be conducted, with the patient sitting on a chair (for details, see Appendix C). The patient The required state of dress or undress will vary with the clinical sit uation. The modern businesswoman with migraine neither expects nor deserves a breast examination. Indeed, she may not allow it. But another, suntanned, woman may justifiably require a search for melanoma. 'Shirt sleeves and socks' may conceal diagnostic physi cal signs. Long pants in the elderly and tight jeans in the young should be removed, because if rolled above the knees, they are effi cient only as a tourniquet. 7 3 The history Aii accurate and detailed history is the supremely important part of the investigation of a neurological problem. By the time the his tory is complete, the physician should be three-quarters of the way towards the diagnosis, and, if not, then there is something wrong with the way in which it has been taken. Experience of consultations has shown that taking a history is almost invariably the weakest part in the presentation of a clinical problem. The modes of onset and progression of symptoms are illdefined, the terms used are vague and woolly, and items of unhelp ful information often predominate instead of the vital facts. The art of taking a good neurological history lies in paying particular attention to certain important points. The age of the patient Surprisingly enough, this is often omitted from verbal case reports, but it influences the management of the problem more than almost any other single factor. There is generally a fairly high degree of cor relation between age and the most probable diagnosis. For example, at the age of 20, a very rapidly developing brainstem lesion is most likely to be due to demyelinating disease, whereas in the 60s it is probably due to basilar artery occlusion. A cerebellar tumour below the age of 12 is probably a medulloblastoma; in the late teens or early 20s, often an astrocytoma or ependymoma; and between 35 and 50, usually a haemangioblastoma, while later on a metastasis becomes likely. Record the year of birth in the case notes, because records have a habit of perpetuating the age at which tine patient first attended. Clarifying the symptoms It must be made absolutely clear what a patient means by his description of his symptoms. By all means put it down in his own words first, but do not be content with that. The term 'giddiness' may mean, to some, rotational vertigo; to others, a sense of insta bility, ataxia of gait, disturbance of vision, loss of contact with sur roundings, nausea, or the term may be used as a socially acceptable description of a full-scale epileptic convulsion. 'Blackouts' may mean loss of consciousness, loss of vision, loss of memory or just loss of confidence. 'Numbness' can be a substitute for 'stiffness' 8 Chapter 3 The history or even weakness. A 'numb' face is often just tense from bruxism (teeth-clenching). By explaining to the patient the great importance of the doctor knowing as well as he does what he means by the terms he uses, and if necessary giving illustrations of the way in which different people mean different things, these points can be clarified by careful insistence. Having reached this stage, the patient must then be forced to be precise about the nature, position and duration of his symptoms. Do not be satisfied with generalizations and vague gestures. A note such as 'the patient complains of continuous pain in the right side of the face' is of very little practical value. 'Continuous' may mean truly continuous or frequently repeated. A pain in the right side of the face lasting continuously all day is not trigeminal neuralgia, whereas attacks of pain, each lasting a few seconds, but repeated every few minutes throughout the day, may very well be, yet it is likely that either will be described as 'continuous'. The patient should clarify the word 'pain'. Many use this word as the easiest in their vocabulary to describe some quite different sensation, such as tingling or even numbness. The character of the pain is impor tant, whether it is aching, shooting, burning, searing, etc., and the patient should be asked to give an honest assessment of its severity. Adjectives such as 'agonizing' slip off the tongue too easily. Finally, and still taking the same example, the patient must be made to indicate with his forefinger the exact part of the face affected. Often, by this method, he may clearly trace out a sensory dermatome or the distribution of a peripheral nerve. The mode of onset and progression of the symptoms , v Most patients, if made to realize how important it is, will be able to say whether a symptom developed abruptly or gradually. As vas cular accidents form so large a part of neurological work, this dis tinction is vital. Patients and young doctors alike overuse the word 'sudden', especially when describing headache. A rapidly evolving occipital pain may not be truly instantaneous. If the patient is persis tently vague in this respect, he should be asked to compare the onset with either a clap of the hands or a gradually rising movement of the hand. In the same way, the progression must be clarified, whether it has been steadily worsening, worsening in a series of steps, or relapsing and remitting. Here again, illustration by hand movement or drawing a graph helps the inarticulate or the vague. Examiners must, however, constantly check themselves from putting words into their patient's mouth, for this is so easy a thing to do. Part 1 The introductory stages 1 Onset sudden, peak within minutes: ° Traumatic • Vascular 2 Onset acute, evolving over minutes to hours: • Vascular ° Traumatic 0 Demyelinating • Sometimes infections/inflammatory 8 Metabolic 3 Onset rapid, evolving over weeks: ° Infections/inflammatory • Metabolic 8 Demyelinating • Sometimes compressive (includes neoplastic) 4 Onset gradual, evolving over weeks to months: ° Infections/inflammatory 8 Neoplastic • Degenerative (sometimes, may be in years) 8 Congenital (may be in years) The chronological sequence of events Sooner or later, someone else is going to read the notes; someone, perhaps, who will have to rely on them entirely to formulate an opinion. They should, therefore, be easily intelligible, and the order of events in time be clearly documented, keeping the main com plaints together and not interposing disorders in other systems unless of obvious relevance. It should be possible from rapidly glancing at a history for a newcomer to build up a clear mental pic ture of the development of the disease without having to go over each point again with the patient. This would be particularly useful in multiple sclerosis, epilepsy and other episodic illnesses. It would be worthwhile to make a summary of the events mentioning the date of event, main symptoms, treatment administered and inves tigations at the time of episode. The value of negative information From the symptoms, the examiner will have formed a good idea of the general group of disorders into which the patient's case falls. Direct enquiry as to the absence of certain symptoms is often as helpful in differentiation as positive information. Chapter 3 The history Excluding irrelevancies Every physician likes to form his own opinion; every patient likes to relate the opinions given to him by others. Patients delight in describing in detail the hospitals they have been to (with dates, usually accurate); the doctors they have seen (with names, usu ally inaccurate); the detailed conversations that they have had with them (with quotations, usually incredible) and the treatment ordered (with results, usually disastrous); together with a wealth of other information which contributes not one iota to the value of the history. These recitals must be checked and irrelevancies rigorously excluded, although an appendix giving previous hospital admis sions may sometimes be of value if one wishes to obtain the results of previous investigations. Drugs Direct questions should be asked concerning medication, whether taken regularly, or at whim, and regarding exposure to habit forming drugs, by mouth, by injection or by inhalation. Many physicians, including neurologists, shy away from this question, and so, on occasions, may miss a vital cause of symptoms. Ques tions regarding prescribed medication apply particularly to anti parkinsonian remedies, anticonvulsants and psychotropic drugs, taken singly or in combination, their type and their dosage. Special enquiry should be made as to the use of oral contraceptives, tact fully, but nevertheless at any age after puberty, and without regard to marital status or religion. Interviewing the relatives While it is always wise to get an external observer's view of the story, in certain circumstances it is absolutely obligatory. These circumstances are: 1 When the patient is a child. 2 When the patient suffers from episodes of impairment of con sciousness. 3 When there is obvious memory defect or mental change. 4 When details concerning other members of the family need to be checked. No case record should ever start with the words 'history unob tainable', even if the only history is what the policeman bringing in the patient can give. 11 Part 1 The introductory stages History is incomplete unless enriched by interviewing an eyewit ness or relatives. V ______ J In determining whether a patient is having epileptic attacks, and if so, whether these are focal or generalized, the need to interview a relative or witness would seem to be obvious. Yet, at every clinic, one sees a patient sent up entirely by himself, suffering from attacks of loss of consciousness of which he himself knows nothing until he comes round, with a request for a decision as to whether or not he has epilepsy, presumably in the pious, but misguided, belief that an electroencephalograph will answer the question. Interviewing relatives will often give valuable insight into per sonal relationships in the family or household which the patient himself may be too embarrassed or too loyal to mention, but which may influence diagnosis and ultimate disposal. Finally, the relatives maybe able to give full details of birth, infancy and education; they may describe a voluntary or enforced change of handedness and they may open up a whole new aspect to the case by confiding that one or other, or both, is or are not the true parents at all, so that a negative family history may be pure assumption. Some extra details Hospital residents should remember that the general practitioner is often able to clarify points confused by patients or relatives. A telephone call in time may save the patient numerous unreward ing investigations and the nation considerable expenditure. This applies particularly where the question of compensation following an injury may arise. T never had any trouble with my back before' may turn out to be the understatement of the year. In children, details of performance at school obtained from the school authorities will often show a recent deterioration not real ized or not admitted by the parents, but having direct bearing on the clinical problem. When the record of the history is completed, it should be stud ied critically. If an intelligent layman, reading it for the first time, would end with a clear picture of the development of the case, then that history will probably be a good one. Remember, however, that it must also be an accurate one, and beware of the fallacy that the neat typing of notes, so perfected in some departments, guarantees their accuracy, as well as their legibility. It does not! 12 9 4 First impressions To the experienced physician, first impressions are invaluable, but if they are allowed to develop into spot diagnosis, it will sooner or later lead to disaster. The purpose of this chapter is to empha size that intelligent analysis of clinical observations made from the moment of first contact with the patient can give a wealth of infor mation which may not be obtainable later under the somewhat arti ficial conditions of formal examination. The circumstances differ when the patient enters the physician's room and when the physician enters the patient's room, and they are dealt with separately. Patient to physician Before the patient enters Listen first to the patient's approach. Loud conversation outside the room usually indicates a patient well accustomed to attending doctors, one who is highly nervous and trying to prove to him self and others that he is not, one who has incoordination of the muscles of articulation such as in multiple sclerosis or one who has recently become deaf. From the sounds made while walking towards the door, it should be possible to distinguish unilateral dragging of the foot in hemi plegia, bilateral dragging in spastic paraplegia and the double 'ker-lump' soimd of the dropped foot. The parkinsonian shuffle is a short, variable, accelerating sound likely to come to a halt at the door itself. Anxious patients, particularly those with medico legal problems, frequently arrive three-quarters of an hour or more before they are due to be seen but those coming for the umpteenth medico-legal report often arrive late, 'having had difficulty in find ing the address'. * * Key point Some neurological diagnoses are clear from the way the patient enters the physician's room and from the sounds made while the patient is walking towards the door. 13 Part 1 The introductory stages Coming through the door The stooping, flexed, rigid attitude of parkinsonism gives the impression that such patients are trying to come through the exact centre of a very narrow space. Although they may smile, the facial immobility and absence of blinking often result in an appearance rather of startled surprise. They may appear to stick in the doorway, or come through the last few feet in a little run. Each turn is made by the body as a whole (en bloc). Patients with cerebellar ataxia may reel into the door frame, but usually successfully fend themselves off. Those who hit it before they realize it, and then turn their heads apparently unnecessarily far round to see what they have hit, probably have a hemianopia. Those who reel only when they are within sight may be doing it especially for the examiner's benefit, but they nearly always forget on the way out. The patient who reels explosively in on a broad base, stick thrust out forwards and sideways, an exaggerated, rather fatu ous smile on his face, looking in the examiner's direction, but not exactly at him, and who begins talking equally explosively from the doorway has almost always fairly advanced multiple sclerosis. Moving from door to chair The patient's chair should not be too near the entrance, hi the few movements needed for him to reach it, a great deal can be learnt. The size and shape of the patient Note dwarfism, excessive height, obesity, wasting and obvious skeletal deformities. Look for an abnormal size of head such as in hydrocephalus, acromegaly, achondroplasia and Paget's disease. Mildly acromegalic features are not uncommon in tall people. The mode of dress Note whether the dress is unkempt, as in the depressed and men tally deteriorated; overcared for; or too casual, often indicating a habitual patient, or merely a modern youth. The style and disposi tion of any jewellery, the type of spectacles worn, the presence of tattoos, may all tell a tale. 14 Chapter 4 First impressions The hair The bedraggled hairstyle of the depressed female is very striking; this is also seen in women showing premature baldness associated with organic disease and is very obvious in myotonic dystrophy. A very sharp demarcation line between the hair and scalp may suggest a wig, and a history of alopecia with all its causative or attendant psychological stresses, or a previous cranial operation. A tightly curled 'perm' in a middle-aged woman with pink-tinted spectacles is often associated with psychoneurosis. Gait and posture This is an ideal time to observe the gait. During the examination, it will be necessary to reassess this formally, but now the patient is moving without realizing that special observations are being made, and unnatural postures are less frequently adopted. The types of gait are dealt with in Chapter 18. Look for the presence of kyphosis, scoliosis, torticollis, the short retracted neck of basilar invagination and the forwards drooping neck of profound muscular weakness, such as in progressive mus cular atrophy, myasthenia gravis or carcinomatous neuropathy. Shake hands with the patient, and compare the grip with the muscular development of the patient. A hand like a wet fish is com mon iii psychoneurosis and in those with an 'inadequate' personal ity. hi those with non-organic states, the grip is often strong under the automatic stimulus of a handshake, but during formal exami nation for strength may be almost non-existent. On sitting down Patients who ask to remain standing, or who collapse into a chair puffing, blowing and fanning themselves, do not very often have organic disease. Similarly, those who sit on one buttock only should be viewed with caution. Involuntary movements, tics and habit spasms are now at their most obvious (see Chapter 19). A sudden distortion of one side of the face, to which the patient clasps his hand, probably holding a handkerchief, is almost pathognomonic of trigeminal neural gia. The tremor of extra-pyramidal disease appears as soon as the patient is settled, by which time, through force of habit, he may have attempted to conceal the affected limb. Some parkinsonian patients, unable to get comfortable, con stantly change their position; others, due to intense akinesia, 15 Part 1 The introductory stages remain totally immobile and may be unable to rise when asked to do so. It is important for therapeutic reasons to recognize this aki nesia. Severe dance-like involuntary movements (dyskinesia) may be seen in advanced Parkinson's disease. Hearing aids indicate bilateral marked deafness and therefore make it very unlikely that the deafness is due to a cerebellopontine angle tumour, which is a common query in the request for a neuro logical opinion. In children and adolescents, a sudden pause, a stare into space, a flicker of the lids, a few mumbled words and a little shake of the upper limbs, followed by an embarrassed smile, may illustrate an 'absence' attack before a word of history has been given. As the hands become visible, excessively long finger nails, partic ularly if painted unusual colours, often share with bizarre spectacle frames a method of drawing attention to an individual who might otherwise escape notice. Multiple burns suggest syringomyelia, but may be occupational. Note the presence of rheumatoid arthritis often associated with wasting of the small muscles. The fa ce Be on the alert to identify the plethoric, fat, hairy face of Cushing's syndrome; the round, smooth, hairless face with tine yellowish pallor of hypopituitarism or the sunken, wasted face with the brownish-yel low pallor of malignant cachexia. Note any scars. Exophthalmos and lid retraction occur in hyperthyroidism, gross exophthalmos occurs with chemosis and ocular paresis in dysthyroid eye disease. Watch tine eyelids carefully for the drooping which varies from moment to moment in myasthenia gravis, or tine fixed bilateral droop with wrin kled forehead in ocular myopathy or the fixed, unilateral droop of a third nerve palsy. A unilateral fixed ptosis may be part of Homer's syndrome, when there will be enophthalmos and a small pupil. The combination of baldness, ptosis, downwards-drawn face and scraggy neck are typical of myotonic dystrophy (Fig. 4.1). Watch the patient's blinking. Absence of blinking is common in parkinsonism. When tirere is a lower motor neuron facial paralysis, the eyes turn up without the lids closing. Note any tremor of facial muscles, clonic spasm of orbicu laris oculis arid oris, and the flickering around the eyes of myokimia - which may also be seen in other parts of the facial musculature. The voice Listen to the volume of sound, the clarity of the words and the con tent of the speech. Disorders of speech are dealt with in Chapter 28. 16 Chapter 4 First impressions Fig. 4.1 Myotonic dystrophy:baldness, ptosis, myopathic facies and absence of stemomastoids. See also Fig. 13.1(c). Patients with pseudobulbar palsy and occasionally with primary brainstem lesions often show 'incontinence' of emotion, laughing or crying involuntarily and irrelevantly. Correlating the symptoms with the patient's condition During the course of taking the history, the examiner must always ask himself, 'Would this patient look or behave in this manner if he had the symptoms of which he complains?' Failure of correla tion does not, of course, necessarily mean that the patient's con dition is not organic. It may be merely an indication that the exact nature of the patient's complaint must be clarified. A patient who says he has no use in the right leg and yet has just walked in probably means that he has a disturbance of sensation in the leg. On the other hand, a patient who complains of unremitting and intolerable pain in the face, driving him to the point of suicide, and present at that moment, but who appears to be in no discom fort of any sort, is unlikely to be describing an organic condition. These are only two examples of comparisons too numerous to mention. Physician to patient This is a rather different situation and arises when the physician approaches a bed in the ward or enters the patient's room in his home. It is in the latter instance that clinical acumen is given its stiffest test, because before the physician leaves that room he will be expected to have formulated a complete analysis of the case. 17 Part 1 The introductory stages The patient seated in a chair Does the patient rise on one's entry? If not, note first the posture. It is easy to recognize the forwards stooped immobility of the par kinsonian; the lateral slump of the hemiplegic; the poorly focused, grimacing welcoming of the patient with multiple sclerosis and the grossly swollen legs that usually mean that the patient has been confined to that chair for many months. Glance around at the rest of the room. Innumerable bottles and pillboxes suggest a demanding patient, and probably a hypochon driacal one, and the stick close at hand for thumping the floor demonstrates the command that the patient has over his family. An empty room suggests a recent illness. The patient in bed In the home, the state of the bedroom will show whether the patient is well accustomed to being there or is an unwilling invalid. In hos pital, the environment of the ward is too standardized, and, apart from family photographs, one may obtain few clues from it. But huge, flamboyant greeting cards and cuddly soft toys surround ing a teenage girl often denote a degree of immaturity. In a young * woman, or even more in middle age, there is a strong association with dissociative disorders. Note the condition of bedclothes and nightclothes for evidence of cleanliness, vomiting or incontinence. Next, assess the patient's conscious level, using the methods dealt with in Chapter 27. If the patient is alert, it can be seen by his drawn expression if he is in pain, or by his perfectly normal appearance that he evidently is not in pain however much he may complain. Intense photophobia occurs in meningeal irritation and migraine. However, do not confuse eye closure and turning of the head away from the light with blepharospasm, where the lids are forcibly, if tremulously, closed. The patient with vertigo lies still, not daring to turn. Patients with respiratory distress lean across a bed table; those surrounded by writing materials are obviously not very ill. The approach generally should be no different from that in the outpatient department. Accuracy of history is just as vital, and in the home one usually has the unique opportunity of talking pri vately and unobtrusively to the relatives, and full advantage of this must be taken. There is, however, that subtle difference produced by the fact that it is the physician who is the visitor. In modern life, a doctor's visit is too common an event necessarily to make a deep 18 Chapter 4 First impressions impression, and for him to restore the position of control he must show confidence and precision both in history taking and in exami nation. This is not difficult if he knows the facts and features he must elicit, but a visit to patients of great age, perceived grandeur or wealth can be an intimidating prospect for the young doctor. V A si f 19 5 The general physical and mental examination r On the ward, so many neurological disorders form a part of general systemic disease that a complete physical examination is absolutely obligatory in each case. To this must be added an assessment of the mental state. The remainder of this chapter lists relationships between general systemic diseases and nervous disease which may be forgotten. A. GENERAL PHYSICAL EXAMINATION Co-operation Uncooperativeness may be due to disturbance of conscious level, to confusion, suspicion, delusion, hallucination, aggressiveness or violence. It must be realized, however, that an apparent lack of cooperation may be due to dysphasia (see Chapter 28), and many unfortunate patients have found themselves admitted to psychiat ric units for what is a wholly organic disturbance of speech. The head and neck Note the size, shape and position of the head; palpate, percuss and auscultate the surface when appropriate. Inspection Gross degrees of hydrocephalus are obvious. To detect lesser degrees, note that the hydrocephalic head and face resemble an inverted trian gle, the forehead being large, bossed and bulging forwards over the orbits, and the eyes being displaced slightly forwards and downwards. In microcephaly, the head appears as a triangle the right way up, the forehead sloping backwards, the occiput forwards and the cra nium coming to a rounded point. In craniosynostosis, the head is deformed in a manner that var ies according to which sutures are prematurely fused. The fused sutures may be felt as hard ridges. The fontanelles are closed, the orbits are flattened and the eyes protrude, sometimes to an extreme degree. In these children, there is often associated finger webbing, and also bronchiectasis. 20 Chapter 5 The general physical and mental examination In acromegaly, the size of the head is increased by elongation, with enlargement of the jaw, and of the ears and nose, while the teeth are separated, the skin coarsened and excessively folded around the eyes and, of course, the hands and feet are also enlarged, the digits blunt-ended and spade-like. The head in advanced Paget's disease is enlarged and appears unnaturally rounded, the scalp being red, warm and covered with dilated vessels. In basilar invagination, the head appears to lie in extension on a shortened neck, the hairline is low and movement is limited. It gives the appearance of being thrust downwards and backwards upon the cervical spine (Fig. 5.1). Palpation Feel the surface of the skull for bony irregularities or deficien cies. The latter are sometimes congenital, sometimes traumatic, often postoperative, but may represent erosive lesions such as eosinophilic granulomata or xanthomata. Skull defects, like fontanelles, are normally concave and pulsating, but become convex and tense in raised intracranial pressure in very young children. Localized bony lumps may lie over an exostosing meningi oma, may be evidence of a sarcoma or may be developmental abnormalities of no significance. Fig. 5.1 Basilar impression. Note the short, retracted neck and low hairline. Part 1 The introductory stages In infants, the anterior fontanelle must be felt and measured. It is usually closed by 18 months, but this varies greatly. It is enlarged in hydrocephalus from any cause, but is tense and bulging in states of continued high intracranial pressure. It is prematurely closed in craniosynostosis. Percussion In children with hydrocephalus and separation of the sutures, tap ping the skull with the fingertip produces a tympanitic, impure and rather high-pitched note, the so-called cracked-pot sound. Auscultation The stethoscope is placed on both frontal bones, on the lateral occipital regions and then on each closed eyelid in turn, hi the last case, if the other eye is opened, the noise due to eyelid flicker is reduced. Listen for a systolic bruit, which when present is usually faint and distant, and may be heard only if the patient holds his breath. Bruits are heard: 1 In young children, too frequently to be of value in diagnosis. 2 Arteriovenous communications. This includes angiomata and caroticocavernous fistulae, tumours of the glomus jugulare (best heard over the mastoid and jugular vein) and in advanced Paget's disease. It cannot be too strongly emphasized that intra cranial bruits are very uncommon in berry aneurysms. 3 Overenlarged external vessels supplying a vascular meningioma. 4 When conducted upwards from the neck, such as in carotid stenosis or aortic stenosis. Neck movement and meningeal irritation Place both hands under the occipital region and, by flexing the wrists, gently raise the head forwards until the chin rests on the chest. Neck rigidity occurs in meningeal irritation of any cause; in inflammatory and destructive disease of the cervical spine, and in cervical fusion; to some degree in cervical spondylosis and in parkinsonism; and in high intracranial pressure, it is a danger sign of tonsillar herniation; posterior fossa tumours may well invoke cervical pain and stiffness without other evidence for raised intra cranial pressure. Extreme tenderness at the side of the neck over the course of the jugular vein is seen after extension of a lateral sinus thrombosis. Tenderness of scalp muscles is extremely rare in organic disease, 22 Chapter 5 The general physical and mental examination but common in psychoneurosis, tension headache and simulated head pains, hi the same way, persistent tenderness in neck muscles rarely has organic significance, yet is complained of for years after a potentially compensatable neck injury. Now flex each hip in turn and then try to extend the knee. This is greatly limited in meningeal irritation (Kernig's sign), and not affected in the other conditions mentioned above. Straight leg raising In suspected lumbar disc prolapse, the ability to raise the extended leg is limited on the side of the lesion (Lasegue's sign), and raising the normal leg may produce root pain on the affected side. In the rare high lumbar disc lesions, root pain may be reproduced by hyperex tending the hip. Though similar to Kernig's sign, there is, of course, no neck rigidity. If the knee has been painlessly fully flexed, and then the hip flexed, and root pain is still claimed to be produced, one should be wary of the organic nature of the case. The spine Spinal examination, normally carried out late in the examination, is described together with tests of stance and gait in Chapter 18. The peripheral nerves When relevant, palpate the ulnar nerve at the elbow, and the lat eral popliteal nerve below the knee as it courses round the head of the fibula. Note if they are very superficial, for they are then prone to trauma, particularly the ulnar nerve if there is also cubitus val gus. Sometimes, especially after old injury, the ulnar nerve may be shrouded in thickened tissues at, or just below, the elbow. Thicken ing of nerves occurs in some forms of hereditary neuropathy and in leprosy. In the latter, the greater auricular nerve is almost always involved and patches of sensory loss and trophic changes are evi dent. Neurofibromata may be palpable along the course of a nerve. Following nerve injury, and during regeneration, tapping the point to which the nerve has regrown may produce paraesthesiae in its distribution ('Uriel's sign). Pressure at the wrist over the ulnar (and sometimes the median) nerve may reproduce the patient's symp toms, especially if the nerve is compressed by a deep ganglion at this site. In patients with median nerve comparison at the wrist (carpal tunnel syndrome), Phalen's test and reverse Phalen's test may be done to reproduce the symptoms and clinical confirmation. 23 Part 1 The introductory stages General exam ination The ears Most deafness is not due to eighth nerve disease. Otitis media must be most carefully looked for in any case of meningitis, meningism, facial paralysis, or if there are any other features suggestive of intracranial infection, such as rapidly spreading local fits, or a pleo cytosis in the cerebrospinal fluid. Auroscopy should be routine in a case of vertigo. In cases of unilateral lower cranial nerve palsies, a polyp in the middle ear seen behind the drum or extending into the external meatus may be the vital clue to the presence of a tumour of the glo mus jugulare. Under any circumstances, do not interfere with such a polyp; torrential haemorrhage may result. The skin Watch the skin carefully throughout the examination, for many fea tures vary from moment to moment. Look in particular for: 1 Signs o f vasomotor instability and peripheral vascular deficiency, often associated with migraine, syncope and anxiety. 2 Allergic lesions and dermatographia. 3 Scleroderma. Systemic sclerosis may present as muscle weakness and wasting, dysphagia and dysarthria, and resemble motor neuron disease. 4 Cutaneous von Recklinghausen's phenomena. These include cafe au lait patches, mollusca fibrosa, subcutaneous and plexiform neurofibromata. These may be associated with spinal and intra cranial neurofibromata and meningiomata and may be familial. 5 Adenoma sebaceum. In the early stages, this consists of pink, globular discrete spots on the cheeks, nose and chin, at first fad ing on pressure, but later darkening to deep red and coalescing (Fig. 5.2). Tire spots almost spare the forehead and upper lip. This is associated with tuberous sclerosis. 6 Cutaneous angiomata. Consider if they are distributed over a seg mental dermatome. Facial naevi occur in Sturge-Weber disease, and spinal segmental naevi occur with disorders of the neu ral crest, syringomyelia and spinal astrocytoma. Midline pink naevi centrally placed high in the neck are not uncommon in normal individuals, but may be very prominent in congenital spinal or craniospinal abnormalities. Spider naevi occur with hepatic cirrhosis and pregnancy, and in some normal people. In familial haemorrhagic telangiectasia, telangiectases may occur on the spinal arachnoid as well as on the skin. 24 Chapter 5 The general physical and mental examination f 'g- 3-2 Adenoma sebaceum. 7 Heiyes zoster. Apart from the disease itself, this type of eruption may occur at the segmental level of spinal lesions due to sec ondary deposits, multiple sclerosis and even trauma. 8 Herpes simplex. Note this carefully, especially in children, for herpes encephalitis is now recognizable and treatable. 9 The common exanthemata may be complicated by an encepha litis. 10 A variety of rashes occur in systemic vasculitis, glandular fever and drug toxicity. 11 Angular cheilitis and pellagra-like skin disorders may be associated with deficiency neuropathies, and Hartnup disease (q.v.). 12 Skin malignancy, in particular melanomata, may have multiple metastases in the nervous system and the primary may have been removed some years before. 13 Light-sensitive eruptions occur in porphyria (polyneuropathy and mental changes with colicky abdominal pain). 14 Erythema ab igne is common in the lonely and depressed. 15 Bed sores are particularly prone to develop in anaesthetic areas. 16 Septic skin areas often precede epidural spinal abscess formation. 17 Scars, burns, destruction o f terminal phalanges and all degrees of peripheral mutilation can occur in syringomyelia, leprosy and hereditary sensory neuropathy. 18 lllcers on the external genitalia, in the mouth and eyes occur in Beh cet's syndrome, which may be accompanied by diffuse cerebral and spinal cord disease not unlike multiple sclerosis. 19 Tufts o f hair often overlie developmental spinal abnormalities, particularly a 'split cord'. The heart There are several ways in which cardiac abnormalities can be correlated with disease of the nervous system. Part 1 The introductory stages Pulse rate and regularity 1 A very slow rate occurs in increased intracranial pressure, vaso vagal seizures and in complete heart block with Stokes-Adams attacks. 2 A very fast rate is found in nervousness, systemic infections, thyrotoxicosis, paroxysmal tachycardia and severe haemor rhage. 3 A fibrillating heart may produce cerebral emboli. 4 Changes in heart rate or rhythm, including rims of extra-systoles (so often considered unimportant), may cause attacks of faintness or vertigo particularly if atheroma of the carotid, vertebral or subclavian arteries is present. 5 Cerebral emboli may follow any form of open heart surgery. 6 Absent pulses are seen in Takayasu's arteritis in relation to eval uation of a young patient with stroke. 7 Carotid bruit. Heart sounds 1 Cardiac murmurs may be significant in patients with chorea. 2 Mitral and aortic disease may be associated with cerebral emboli, especially if there is any suggestion of bacterial endocarditis. 3 Aortic stenosis causes episodes of fainting. 4 Aortic regurgitation may be associated with neurosyphilis. 5 Congenital cardiac lesions predispose to cerebral abscess forma tion. 6 Subclavian stenosis may result in a steal of blood from the ver tebrobasilar system, especially on exercise. A subclavian bruit may be accompanied by a delayed radial pulse and a lower blood pressure in the affected arm. The blood pressure This may be very important, and in particular circumstances should be taken both lying and standing, and in both arms. Hyper tension may be of the benign type in so far as it is not producing renal damage or retinal disease, but in the presence of a sudden loss of function of some part of the body, it may very well be of great significance. Transient postural hypotension is a common cause of attacks of loss of consciousness. Persistent postural hypotension may signify autonomic failure (see Chapter 31, p. 283). 'Subclavian steal' syndrome will be accompanied by a significant difference between the two arms. Chapter 5 The general physical and mental examination The lyrags Attention during examination should be paid particularly to any suggestion of: 2 Bronchial carcinoma, because in the male this is the common est primary site for intracranial or multiple neurological metastases. 2 Bronchiectasis, a common origin for metastatic abscesses. 3 Tuberculosis, when there are signs of meningitis. 4 Severe bronchitis and emphysema, which can cause headaches, papilloedema, fluctuating consciousness and involuntary movements (asterixis). 5 A Pancoast tumour. This usually presents as inexorable pain and marked sensory impairment in the distribution of the lowest cord of the brachial plexus. 6 Small oat-cell carcinomata (and mesotheliomata) may be associ ated with many of the paraneoplastic syndromes, particularly Lambert-Eaton-type myasthenia. 7 Interstitial lung disease may be associated with dermatomyositis in mixed connective tissue disorder. 8 Diaphragmatic palsy may be seen in association with phrenic nerve palsy and motor neuron disease. The abdomen Carcinoma o f the stomach, colon and hypernephromata may first declare themselves as metastases in the nervous system. Metastases from these tumours may manifest themselves years after removal of the primary growth. Neuroblastoma in children may give rise to deposits in the orbit and frontal bones (Hutchin son's syndrome). Hepatosplenomegaly is associated with cerebral disease in the lipoidoses of children, in sarcoidosis, reticulosis and leukaemia in adults, and in the older age groups may be further evidence of met astatic malignancies, or may accompany the neurological features of chronic alcoholism, such as polyneuropathy, Wernicke's enceph alopathy and Korsakov's psychosis. The pelvis Malignancy of pelvic organs may cause lesions in the nervous sys tem, most frequently by metastasis to the spine (e.g. in prostatic carcinoma) or by direct invasion of peripheral nerves (e.g. in carci noma of the cervix). Pregnancy maybe associated with psychological 27 Part 1 The introductory stages disorders, chorea, vascular accidents and enlargement of pre-ex isting unsuspected cerebral tumours. As induced abortion is now commonplace, and not necessarily always carried out by the most expert hands, it must not be forgotten that fits, hemiparesis and coma may follow, due to air emboli, or even amniotic emboli, usu ally in unskilled hands. A pituitary tumour can cause amenorrhoea and it is unwise to diagnose primary amenorrhoea or a very early menopause without X-ray or computerized tomography scan of the sella turcica. The breasts The breast is the second most common site of the primary tumour in cerebral or spinal metastases. Careful examination is essential in all cases of spinal or cerebral disease, both in men and women. Evidence of previous mastectomy, even if very radical and with no signs of local recurrence, may be highly significant. Galactorrhoea is a symptom of prolactinoma. Premature breast development can occur in hypothalamic lesions. The thyroid Pay special attention to the thyroid, noting its size and consistency, the presence of any signs of thyrotoxicosis or the presence of a recent thyroidectomy scar. These may be significant in the presence of: 1 Exophthalmos with ophthalmoplegia. 2 Muscle wasting and weakness; thyrotoxic myopathy is a rare possibility. A periodic paralysis can accompany thyro toxicosis. 3 Multiple lesions in the nervous system; the thyroid is a site of primary tumours. 4 Tendon reflexes that are prolonged and relax slowly. These are a feature of myxoedema (see p. 223). 5 Thyroid enlargement, or a past

thyroidectomy, may have caused recurrent laryngeal nerve paralysis. The glands Glandular enlargement, an accompanying disease of the nervous sys tem, directs attention particularly to the following possibilities: 1 Metastatic carcinoma or possible Hodgkin's disease. 2 Granulomata such as sarcoid, which may cause cerebral deposits and polyneuropathy and many other neurological syndromes. 28 Chapter 5 The general physical and mental examination 3 Lymphatic leukaemia or reticulosis, in which, terminally, there may be severe cerebral demyelination, or an encephalitis due to invasion by torula, or other organisms, especially if immunosuppressive therapy has been given. Remember human immunodeficiency virus disease. 4 Glandular fever. A Guillain-Barre syndrome is a rare complica tion, encephalitis may occur and local pressure palsies are not unknown. The teeth Unsuspected dental abscesses are a possible source of cerebral abscesses, especially if there is congenital heart disease. Defective teeth, defective closure and temporomandibular joint dis ease can all cause facial neuralgic pain. Look for bruxism. Blue line along the gum is seen in chronic lead poisoning. Gum hypertrophy indicates heavy phenytoin dosage. The tongue For convenience, abnormalities are classified together in Chapter 12. The nails Nail-biting commonly indicates tension and anxiety. Koilonychia should make one suspect severe iron-deficiency anaemia. Clubbing may accompany congenital heart disease, chronic chest disease, bronchial malignancy and chronic hepatic disease. Mee's lines in the nails are suggestive of arsenic intoxication. The Feet Examine the dorsal as well as plantar surface of the feet for any ulcer, amputation of toe, or any deformity. A common deformity seen in neurological practice is pescavus, which consists of exag gerated long arch of foot with clawing of toes. Clawing involves hyperextension of metacorpophalyngeal joints and flexion of interphalangeal joints. In early cases of pescavus, only claw ing of toes and callosities on the ball of foot is seen. In definite cases, light can be seen through and through the arch of foot on standing and a foot point (place a wet or oily foot on flat sur face) will show interruption of isthmus connecting the ball of foot and heel. 29 Part 1 The introductory stages © A. Familial (usually bilateral or symmetric) 1 . Hereditary neurological disorders • Friedreich'sataxia • Charcot-marie-tooth disease • Hereditary spastic paraplegia ° Roussy-Levy syndrome • Refsum's disease 8 Dystonia musculorum deformans 8 Rarely, muscular dystrophy (if feet involved) 2 . Isolated pescavus in family members of patients with hereditary neurological disorders • Friedreich'sataxia 8 Charcot-marie-tooth disease 3 . Idiopathic familial pescavus B. Sporadic (usually unilateral or asymmetric) 8 Traum a • 8 8 • 8 8 Poliomyelitis Congenital Cerebral palsy Spinal dysraphism (e.g. diastematomyelia) Syringomyelia Clubfoot This need not be very elaborate, and indeed full mental status exam ination is beyond the scope of this book. Begin with brief screening tools such as Folstein's mini-mental status examination (MMSE) (published in Folstein MF, Folstein SE, McHaugh PR. Mini-mental state: A practical method for grading the cognitive state of patients for the clinician. / Psychiatric Res 1975;12(3):189-98.) but whenever indicated, e.g. in patients with minimal cognitive impairment, more rigorous set of neuropsychological tests will be required. Items that are used in assessing mental status are provided in detail in Appen dix A. A suggested scheme for assessment of higher cerebral func tion is given in Appendix D. Interpretation and limitations of MMSE The normal range of score depends on education. Some experts sug gest a normal cut-off of 19 for uneducated people, 23 for graduates 30 Chapter 5 The general physical and mental examination of elementary or junior high school, 27 for high school graduates and 29 for college graduates. Usually, a score of < 23 for educated people is taken to indicate cognitive impairment. Limitations of the M M S E The test is weighted towards orientation and language (these ele ments form 19 of the 30 score points). It can be normal in patients with right hemisphere and frontal lobe damage. An abnormal score does not distinguish between a focal and a diffuse disease of the brain. Orientation MMSE tests only orientation to time and place. Determine whether the patient is orientated to person - whether he knows his name, age, address; the number and names of his children and who the doctors and nurses are. Disorientation may occur in any or all of the time, place and person in organic dementias. Usually, disori entation occurs first to time, then to place and last to person (see Appendix A). Attention Serial 100 minus 7 and digit span are the commonly used tests for attention. Serial 100 minus 7 is a part of MMSE. For digit span test, read out to the patient clearly and slowly a series of numbers for him to repeat. Start with three numbers and increase by one each time until he makes consistent errors. It is wise to have written out the groups of numbers beforehand. Avoid consecutive num bers; avoid emphasizing any one number. Then carry out the test with different numbers, asking him to repeat them in reverse order. Many people can repeat seven or eight fonvards and four or five backwards correctly. Memory Memory testing usually involves tests for immediate memory, short-term memory and long-term memory. Immediate memory Dive the patient three unrelated words (e.g. carrot, ankle, honr'tv). Ask him to repeat. Keep saying them until he can repeat all three. 2 1 Part 1 The introductory stages Short-term memory Ask the patient to recall the three words you previously asked him to remember. Test the patient's ability to remember the events of that day. Ask what actions he has recently carried out, what he has eaten for breakfast, who have met him today, how he came to the pres ent building, where this building is and where he is going after he leaves. Find out if he has read the newspapers or watched television, and ask what have been the main news stories of recent days. If interested in sports, see if he can recall items of topical sports news. Long-term (remote) memory Ask the patient to recall personal and historic events. Ask where was he born, where did he go to school or college, where has he worked, what is his wife's or children's names and how old are they. For historical events, ask him to name four people who have been presidents or prime ministers during Iris lifetime, which years did world wars or 9/11 happen. Personal events must be verified by a reliable source other than the patient, and performance on the recall of historic events must be interpreted in the view of the patient's level of intelligence, edu cation and social experience. Many patients may fail these tests simply due to poor powers of concentration, but loss of recent memory and ability to retain and recall are common in all organic dementias, distant memory often being preserved. In Korsakov's psychosis, recent memory and orientation in time and place are so disturbed that the patient makes up stories, often interesting ones, to fill in the gaps. This is termed as confabula tion, and may be so detailed as to convince an unwary examiner. Though classically associated with alcoholism, anterior hypotha lamic lesions due perhaps to anterior communicating aneurysms or their surgical treatment can temporarily produce an identical picture. Emotional state Note if the patient is anxious, excited, depressed, frightened, apa thetic or euphoric. Euphoria is apparently common in patients with multiple sclerosis, but often only when visiting the doctor. Some patients with frontal lobe disease repeatedly make fatuous remarks and treat all parts of the examination as something of a joke, a condition termed 'witzelsucht'. A somewhat aggressive euphoria commonly follows severe head injuries. 32 Chapter 5 The general physical and mental examination Incontinence of emotion, laughing or crying suddenly and often inappropriately, occurs in pseudobulbar palsy, usually due to bilateral cerebrovascular disease or amyotropic lateral sclerosis. Delusions and hallucinations Listen to the flow of speech; is there evidence of delusions, and, if so, are they systematized or grandiose; does he appear to have auditory, visual or olfactory hallucinations, and can he describe them? Temporal lobe lesions produce well-organized and formed hallucinations, sometimes terrifying; occipital lobe visual halluci nations are ill-formed or unformed. Patients with non-dominant hemisphere lesions are usually able to describe hallucinations more clearly and in greater detail. Great assistance can be given by a qualified psychologist in psychometric examination, in assessing intelligence, educational state and capacity, and finding evidence of recent deterioration in organic cerebral disease. In children, it may be difficult clinically to distinguish between mental deterioration and a pre-existing men tal defect which has merely become more obvious as they grow older. Here, serial recordings are important. Tests for organic dementia are more than of academic interest these days for in the so-called low-pressure hydrocephalus, beneficial therapy may be available for the appropriate few. Tests for speech, apraxia, agnosia, disorders of body scheme and lobar functions are dealt with in the appropriate sections of Chapters 29 and 30. Higher mental function assessment can be done by focussing on the tests of four lobes of the cerebral hemispheres (Fig. 5.3). Although various standardized batteries are available for the assessment, the following tests may reveal involvement of various Abes and its extent. This part of the chapter presents approach to test the higher mental functions as lobar functions, though many auctions involve association areas in other lobes via different networks. Co-operation from the patient and the caregiver is very import er : for carrying out these tests. To seek the cooperation, give ade: - ; : e instructions to reassure the patient and alleviate the anxiety r ; : may arise due to failure to perform the tests. Part 1 The introductory stages Fig. 5.3 Four lobes of the brain. Frontal lobe It is the largest lobe of the brain. The most anterior part of frontal lobe is the prefrontal cortex which is the seat of the executive func tions (dorsolateral prefrontal cortex) such as planning, program ming, judgement, motor sequencing, abstract thinking and set shifting. The mediofrontal and orbitofrontal cortex are the centres for motor and social inhibition and personality traits. Executive functions test (prefrontal cortex of frontal lobe) The aim is to check the fund of information, motor sequencing, set shifting, problem solving, judgement and abstract thinking. 1 Ask and note how the patient had planned the visit to the doc tor and his eagerness to reach on time. 2 Ask and note whether the patient is able to give the history properly and has preserved insight about his problems. 3 Forward and backward digit span. Ask the patient to repeat the number sequences in the increasing order, for example 4 then 46, then 469, then 4691 then, 46915 and check whether the patient is able to repeat it correctly. Alternatively, days of the week or months of the year backwards can be used. 4 Ask the patient to produce as many words as possible, in one minute, starting with any letter like F, then A and then S. 34 Chapter 5 The general physical and mental examination 5 Ask the patient to name animals, fruits or vegetables as many as he can in one minute (more than 10 is normal; abnormal is less than eight). Look for perseveration. 6 Motor Luria test (fist-edge-palm test). First explain the patient by doing it yourself. Make a fist and tap on flat surface with your thumb upwards; then straighten out your fingers and tap with the ulnar border of hand and then place your flat palm on the surface. Repeat this again. Once the patient has understood, ask the patient to repeat it. The hand motion may be reinforced by counting from 1 to 3 along with each segment. 7 Proverb interpretation test. Give five proverbs in the patient's own language in the increasing order of difficulty keeping in mind social and educational background of the patient. Ask him to explain what it means. Note the degree of abstract think ing shown by the patient to explain the interpretation of the proverb. 8 Luria graphic test. Patient is asked to draw alternating sequence like a triangle and a square after being shown by the examiner. An example is shown below. 9 Set shifting (a) Trail making test A — X © S ' © ( ) W ® @ © *© © @ 22 e © © © © © © © ©© © ® © © © © 35 Part 1 The introductory stages (b) Trail making test B Orbital and basal areas of frontal lobe The aim is to examine the patient's ability to inhibit his responses, both social and motor. 1 Check for the inappropriate dressing sense and behaviour that suggests lack of concern with the feelings of others or without concern to accepted social customs. 2 Go/No-go test. Ask the patient to make a response to one signal (the go signal) and not to respond to another signal (the no-go signal). • Direction. Patient is asked to tap once when examiner taps once and stop when examiner tap twice • Score (a) No error: 3 (b) 1-2 errors: 2 (c) > 2 errors: 1 (d) Patient taps like the examiner at least four consecutive times: 0 • Significance. Difficulty in withholding a response - lesion in ventral part of frontal lobes. 36 Chapter 5 The general physical and mental examination 3 The stroop test. This test examines the patient's ability to inhibit his responses. Patients are asked to state the colour in which words are printed rather than the words themselves. Example: Ask the patient to tell the colour of the letters in the following: Red, Blue, Black, White, Orange, Violet Parietal lobe Ideational apraxia Patient is unable to perform a task which involves a series of motor activity or steps, for example, 'Fold an envelope, apply a stamp and seal it'. The patient can be asked to open a toothpaste, take the toothbrush and place the paste on the brush or to place the candle on table, light a matchstick, light a candle and blow out a match. Right-left orientation Test can be done in four steps of increasing difficulty: 1 Identification on self (e.g. show your left hand) 2 Cross command on self (e.g. touch your left ear with right hand) 3 Identification on examiner (e.g. show my right hand) 4 Cross command on examiner (e.g. touch my left hand with your right hand) Nine percent of normal males and 17% of normal females are reported to demonstrate difficulty in doing the above in the Western literature. Finger agnosia Test can be done in three steps of increasing difficulty: 1 Nonverbal finger recognition (touch the patient's finger in a closed-eye state and have eye opened and ask the patient to point the same finger on the examiner's hand). 2 Identification of the named finger of the examiner. 3 Verbal identification (naming) of fingers of self and the exam iner, for example, What is the name of this finger? Both right-left disorientation and finger agnosia are part of Gerstmann syndrome due to a lesion of dominant parietal lobe. Visuo-spatial functions Show the patient a drawn map of his or her house as told by the caregiver. Now ask the patient to navigate the way in the house. 37 Part 1 The introductory stages Cortical sensations Look for sensory extinction (tactile), astereognosis, graphaesthesia (refer to Chapter 23 and 30). Simple and complex calculations Test both verbal and written in increasing order of difficulty - first verbal simple, then verbal complex and then written complex. Give verbal simple calculations with single digit addition, subtraction, multiplication and division. Next give verbal complex calculations (e.g. 12 + 19,42 - 36, etc.), and then written complex calculation like 106 + 59; 405 - 75, etc. Adjust depending on the educational status of the patient. Geographical orientation Ask the patient to mark the important cities and towns on the map of the country. Look for the correctness in terms of near the ocean, direction and accuracy. Constructional ability tests in tw o steps 1 Reproduction of two- and three-dimensional diagrams (at least two of each type) Scoring - 0 ,1 ,2 ,3 for poor, fair, good and excellent, respectively. This tests constructive praxis/visuo-constructive ability involv ing integration among parietal, occipital and frontal lobe. 2 Drawing on command: Clock face drawing Ask the patient to draw a wall clock face by drawing a large cir cle, putting numbers from 1 to 12 and two hands showing time ll:10.If patient is unable to initiate drawing a circle, then ask again to do it. Scoring - give 3 for putting no. 12 in position, 1 for 12 numbers, 1 for hands and 1 for correct time. Score of less than 4 suggests definite impairment. This tests constructional apraxia, visuo-spatial orientation, hemineglect, right-left orientation, planning and sequencing. Temporal lobe The tests for temporal lobe functions involve testing of the memory and language. Chapter 5 gives tests for memory and Chapter 28 for language. 38 Chapter 5 The general physical and mental examination O ccipital lobe Prosopagnosia (inability to recognize familiar faces) The photographs of family members, popular figures like sports men, political, historical or religious figures (Fig. 5.4) can be shown and patient can be asked to identify the person in picture. Presence of any hemianopia with macular sparing also supports the involve ment of occipital lobe. Visual m em ory (five hidden objects) 0 Direction - Name each item as it is hidden, provide interfering stimuli for 5 minutes, ask the patient to name and indicate the loca tion, If the patient cannot locate, ask the name of the hidden object. 0 Score - less than 4 is abnormal. M iscellaneous Some of the tests of higher mental functions involve the functional circuits across multiple lobes and can provide us the clue to know the abnormality. For example, clock face drawing test described above. Similarly, language functions involve predominantly Frontal and temporal lobe (details can be seen in Chapters 28-30). A sum mary of lobar functions and their assessment tests and effects of lesions is presented in Table 5.1. t ig. 5.4 Examples of historical and political figures. 39 Table 5.1 Functions of the four lobes of the brain and the recommended tests for their assessment Frontal Parietal (dominant) Functions 1 Control of emo tions 2 Personality 3 Social behaviour 4 Contralateral motor control 5 Language 6 Micturition Language calculation Lesion effects 1 Disinhibition 2 Inertia 3 Antisocial behaviour 4 Memory impair ment 5 Motor aphasia 6 Incontinence 1 2 3 4 5 Examine 1 Executive functions (planning, programming, judgement, motor sequencing, abstract thinking and set shifting) 2 Working memory 3 G o/N o-go test 4 Si mop lest 1 Language (spontaneous speech, naming, both read ing and verbal comprehen sion, reading, writing) 2 Praxis tests (for ideo-motor and ideational apraxia) 3 Calculation (simple and complex) 4 Right-left disorientation Aphasia Acalculia Alexia Apraxia Agnosia Parietal (non-dominant) Temporal (dominant) Temporal (non-dominant) Occipital 1 Spatial orien tation 2 Construc tional skills 1 Auditory perception 2 Language and verbal memory 3 Smell 4 Balance 1 Auditory perception 2 Nonverbal memory 3 Smell 4 Balance Visual 1 Contralateral hemineglect 2 Spatial disori entation 3 Construc tional apraxia 4 Dressing apraxia 1 Sensory aphasia 2 Alexia 3 Verbal memory impairment 1 Nonverbal memory impairment 2 Amusia Visual inattention Visual loss Visual agnosia 1 Clock drawing test 2 Visuo-spatial tests 3 Construction and dressing 1 Language 2 Memory 1 Language 2 Memory IT % 1 Visual memory 2 Prosapognosia 3 Hemianopia with macular sparing Part 1 The introductory stages 4* O Part 2 The Cranial Nerves Olfactory nerve Optic nerve Oculomotor nerve Trochlear nerve Abducent nerve Trigeminal nerve Vestibulocochlear nerve Facial nerve Glossopharyngeal nerve Vagus nerve Hypoglossal nerve Accessory nerve Brainstem with cranial nerves © The first cranial nerve: the olfactory nerve It is unfortunate that this is the first nerve because tests of its function form one of the least satisfactory parts of the whole exami nation. At times, however, anosmia may give the vital clue to local ization of an intracranial lesion. A n a to m y The olfactory nerve begins in the form of about 20 branches on each side of upper nasal cavity from the bipolar cells located in the olfac tory epithelium (the first-order neurons). These branches pierce the cribriform plate of the ethmoid bone and synapse with the mitral and tuft cells. The axons of these cells (mainly mitral cells) continue in the olfactory bulb and tract and lie in the olfactory groove in the basifrontal region. The olfactory tract divides into medial and lat eral olfactory striae. Tire primary olfactory cortex is located in the medial temporal lobe (hippocampus). Fyoctnoo To carry sensations of smell from the nasal mucosa to the olfactory bulb. The stimuli then pass through the olfactory tract and roots, especially the lateral root, running under the temporal lobe to the periamygdaloid and prepiriform areas of the cortex, the uncus and hippocampal gyrus. Pyrpose o f th e te s t To determine whether any impairment of the sense of smell is uni lateral or bilateral, and whether it is due to local nasal disease or to a neural lesion. M ethod o f testin g Small bottles containing essences of very familiar odours are required. These must indeed be familiar. Asafoetida and even musk are sometimes advised, but are useless. Coffee, almonds, chocolate, oil of lemon and peppermint are amongst the many that are suitable. 43 Part 2 The cranial nerves O • Local acute or chronic inflammatory nasal disease; heavy smoking. • Head injury*- usually thought due to tearing of olfactory fila ments. The trauma may be surprisingly slight and often occipital. • Intracranial tumours, most commonly inferior frontal, malig nant or benign, compressing the olfactory bulb or tract. • Atrophy* of the olfactory bulbs. • Chronic meningeal inflammation (e.g. syphilis) or infiltration (e.g. sarcoidosis). • Parkinson's disease. May be accompanied by cerebrospinal fluid rhinorrhea. The patient must compress each nostril in turn and, by sniffing through the other, show that the airway is clear. The test odour is then placed under one nostril while the other is compressed, and the patient is told to take two good, but not overexuberant, sniffs. He is then asked (i) if he can smell anything and (ii) if he can iden tify the odour. The test is then repeated using the other nostril and he is asked (iii) if the odour is the same in each nostril. After an interval to allow that odour to disperse, the test is then repeated with two further odours and, in addition to the above, he is asked (iv) if he can distinguish the different odours. Interpretation of results Patients will fall into several categories: 1 Those who recognize and name the odours quickly (usually women). 2 Those who recognize but cannot name them (usually men). 3 Those who can detect a smell, and easily distinguish differ ences, but can neither recognize nor name them. 1, 2 and 3 should be accepted as normal. 4 Those for whom each odour smells the same but is distorted and unpleasant - parosmia. 5 Those who can smell nothing in one or both nostrils, or whose sense is much reduced on one side compared with the other. This represents true anosmia, but careful examination of the nasal passages is necessary to distinguish neural from local dis ease. 6 Those whose responses are very vague and variable. This is probably going to be a long and tiring neurological examination, 44 Chapter 6 The first cranial nerve: the olfactory nerve and to avoid impatience at this early stage, it is wise to return to the problem later. Parosmia is not uncommon in incomplete olfactory recovery following head injuries. It may also occur in depressive or schizophrenic states and in hysterical conversion syndromes. Difficulties and fallacies This is a purely subjective test. The examiner has to take the patient at his word and unfortunately, anosmia as a potential sequel to head injury is now a well-known symptom - well known to litigants as well as to doctors. If an attempt is made to produce objective results by using substances such as ammonia or ether, something other than the olfactory nerve is being stimulated as well. It is widely believed that an olfactory groove meningioma is a common cause of anosmia. It is certainly an important cause, but it is also a rare tumour, and if a tumour is present, an inferior frontal glioma is, in practice, more likely. 45 7 The second cranial nerve: the optic nerve Although the optic nerve is the most important cranial nerve, it is not a peripheral nerve in the true sense of the word, but a forward extension of a part of the brain. As it can reflect conditions existing inside the cranial cavity, no amount of trouble is too great to ensure a satisfactory examination. It is customary at this stage to include tests designed to detect lesions throughout the whole extent of the visual pathways. Anatom y The optic nerve consists of axons of the retinal ganglion cells. Optic nerve fibres exit the globe at scleral canal. The fibres then continue till optic chiasma where there is decussation of nasal fibres, result ing in grouping of all the fibres of each hemifield in the opposite optic tract. Most of the axons in the optic tract terminate at the lat eral geniculate body (LGB). From LGB, the fibres continue as optic radiation which terminates in the visual cortex of the correspond ing occipital lobe (Fig. 7.1). Functions To carry visual impulses from the retina to the optic chiasma and on in the optic tract to the LGB; to act as the afferent pathway for the pupillary light reflex by means of fibres travelling to the supe rior colliculus of the midbrain. Purpose of th e te s ts 1 To measure the acuity of vision and to determine if any defect is due to local ocular disease. 2 To chart the visual fields. 3 To take the unique opportunity of inspecting directly a part of the nerve itself. 46 Chapter 7 The second cranial nerve: the optic nerve Visual acuity M ethods of testin g The standard Snellen's Type charts are used for testing distant vision and the Jaeger Type cards for near vision. The Snellen's Type chart is placed, evenly illuminated, 6 m from the patient, who covers one eye and is asked to read the smallest line he can see accurately. He should not close an eye, as this may make him partially close the other and give misleading results, especially in myopia. Acuity is recorded as a fraction (e.g. 6 /2 4 or 20/80). The numer ator indicates the distance at which the patient has to be from the chart in order to read the same type that the normal person could read at a distance indicated by the denominator. 6 / 5 - 6 / 6 is within the average normal range. The Jaeger Type card must be held 30 cm from the patient's eye and a similar test is then carried out. The different types are labelled as, for instance, J.4, according to their size. Average acuity lies between J.l and J.4. There are of course a number of other test types, but the general principles remain the same. If visual acuity is severely depressed, hold up varying numbers of fingers to be counted with each eye. If unable to do this, see if hand movements can be detected. Always equate the degree of visual deficit claimed with what you observe the patient to be able to do. Loss of visual acuity The commonest causes of visual failure lie in lesions of the eye itself. These are too numerous to list, but include all refractive errors, cataracts, vitreous opacities etc. In all cases, and especially if routine inspection of the eye reveals no such lesion, the next stages are the examination of the visual fields and of the optic nerve head and the retina. The visual fields Charting the visual fields is the most important method of locating a lesion in the visual pathways or of interpreting certain fundus appearances, yet in clinical notes it is often regrettably difficult to find evidence that any attempt has been made to examine them at all. 47 Part 2 The cranial nerves o Sudden unilateral vision Sudden bilateral vision loss loss 4 ° Central retinal artery occlusion • Anterior ischaemic optic neuropathy - arteritic or nonarteritic 0 Branch or central retinal vein occlusion ° Traumatic • Central serous choroidopathy 0 Retinal detachment 0 Vitreous haemorrhage 0 Psychogenic Transient monocular blindness 0 Embolism of 0 Occipital lobe central retinal infarctions artery 0 Pituitary apoplexy 0 Migraine (vasospasm) 0 Functional (psychogenic) 0 Hypoperfusion (hypotension, vision loss hyperviscosity) ° Traumatic 0 Vasculitis 0 Uhthoff's phenomenon 0 Psychogenic Bilateral transient vision loss 0 Migraine ° Occipital lobe hypoperfusion 0 Epilepsy • Papilloedema (transient visual obscurations) Purposes o f th e te s ts To chart the periphery of the visual fields; to detect the position, size and shape of the blind spot and any abnormal scotomata; to compare any defects shown with those abnormalities known to be reproduced by lesions at specific points in the visual pathways. Methods of testing Do not underestimate the value of testing by confrontation. A sur prising degree of accuracy is possible with a cooperative patient. Confrontation The patient and examiner face each other. Tire examiner covers (say) the right eye and the patient covers the left, either with a shield or with his fingers, taking care to avoid obscuring the nasal field of the other eye. He must fix carefully on the examiner's pupil while tire examiner moves test objects of varying size (the whole hand, a mov ing finger, a white or red pin) inwards in each of the four quadrants from just outside the limits of his own field. The patient must say the moment he sees the object and whether it is of equal clarity in each quadrant, for 'greying' of vision often precedes a measurable defect. 48 Chapter 7 The second cranial nerve: the optic nerve The central area of vision can be tested in order to map out the blind spot and any scotomata by using a disc of white paper about 5 mm in diameter attached to a rod or long pin, or a white or red hat pin (see below). The examiner should move the test object from blind areas to areas of vision, because it is easier to detect appearance than disappearance of objects. When this is completed, both eyes are uncovered, the examiner holds both hands in the outer part of the fields and, moving one or other hand, or both together, asks the patient to point to the hand moved. This will detect the phenomenon of visual inattention com mon in parietal disorders where, although the fields tested in each eye separately are normal, only one object is appreciated when both are moved simultaneously (see Chapter 30). If a homonymous hemianopia is present, this can also very quickly he picked up by this method. Visual field defects and their significance. 43s* . s * * S & \ & So many and varied are the possible defects that it is not possible to describe them all, but the principles by which lesions are located are best illustrated by comparing a diagram of the visual pathways (Fig. 7.1) with the principal field defects (Fig. 7.2(a)-(n)). Difficulties and fallacies Curious bilateral defects of the upper fields or nasal fields can be produced by mephistophelian eyebrows or an overgenerous nose. These can be avoided simply by tilting or turning the patient's head appropriately. A patient with a central scotoma cannot fix on a central object. Two objects, placed just at the edge of the scotoma on the screen, can be held in their relative positions by a cooperative patient. Always be suspicious of the accuracy of the small, concentri cally constricted field if visual acuity is adequate. The real shape of the fields will be determined by using successively larger objects, unless the defect is of hysterical origin. Poor cooperation in fixation can often be overcome simply by enlarging the object on which the patient is to fix. Modern perimeters, however, allow the perimetrists to view the patient's fixation directly while each point is tested, thus increasing accuracy, if not solving the problem. 49 Part 2 The cranial nerves Fig. 7.1 Diagrammatic representation of the visual pathways. The common sites of lesions are lettered and the characteristic field defects so caused are illustrated in Personal equations vary. Move the object slowly. It may take time for its appearance to register with the patient and for the patient to react. Examining the fundus The fundus must always be carefully examined no matter what the patient's complaints may be. Only by inspecting the maximum possible number of fundi can that experience of the wide variation (a) Total unilateral loss of vision. A lesion of the optic nerve (Fig. 7.1(a)). Common causes: Injuries, optic neuritis, optic nerve compression. Involvement of nasal crossing fibres from the other eye will cause a contralateral upper tem poral field defect (shaded area), usually with an ipsilateral scotoma rather than total blindness. (b) Altitudinous hemianopia. A partial lesion of the blood supply to the optic nerve (Fig. 7.1(b)). Common causes: Trauma, vascular accidents. Fig. 7.2 (a)-(n) Diagrammatic representation of common field defects to be studied in conjunction with Fig. 7.1. Chapter 7 The second cranial nerve: the optic nerve (c) H o rn * o n y m o u s hem ianopia. The commondefect cau sed by a lesion anywhere est m a jo c tract t o occipital cortex. In the tract, it from c>p t com plete, incongruous, without mac is usurill ing (Fig. 7.1(C,)). In the radiations, it is ular s p a usually * jm com plete, congruous, with macular ig. 7.1 (CO)- hi the calcarine cortex, it is sparing C mplete, congruous and with macular usually Maut m ay show associated scotomata sparing, ,)). C ongru ity and macular sparing (Fig- 7 A le. are varU C om m o tumou (d) U p p T em p o ral ations "W horn o i com m on ally in p a C o m in o accident t s . causes: Vascular accidents, cerebral 'vascular anomalies, injuries. quadratic homonymous defect. lobe lesions involving the optic radicre they sweep round the temporal e lateral ventricle (Fig. 7.1(d)). Less in low er calcarine lesions, occasiontial tract lesions. causes: Cerebral tumours, vascular cerebral abscesses, injuries. (e) L o w e r quadratic homonymous defect. Lesions o J C the upper radiations or calcarine area (F i g . > ^ .l (e)) . C om n rort causes: Vascular accidents, injuries, tu m o u rs. (f) B i t e » n j ^ « o r a l hemianopia. Lesions at the optic c h i a s* m a (Fig. 7.1(f)). C om m on causes: Pituitary tumours, craniop h a ry n g ^ io n n a ta , suprasellar meningiomata, midline a r m o u r y sms, hypothalamic neoplasms, ^ross th i r d t ventricular dilatation, optic chias mal g lio » r» ^ » *a. Fig. 7.2 (continued) 51 Part 2 The cranial nerves (g) Bitemporal upper quadratic defect. The early stages of chiasmal compression from below (Fig. 7.1(f)). Common causes: Pituitary tumours. (h) Bitemporal lower quadratic defect. The early stages of chiasmal compression from above (Fig. 7.1(f)). Common causes: Intrinsic tumours of the hypothalamus, suprasellar cysts or meningiomata. This is not a very common defect. (i) Enlarged blind spot: Enlargement of the optic nerve head (Fig. 7.1(g)). Common causes: Papilloedema from increased intracranial pressure. (j) Central and centrocaecal scotomata. Intrin sic lesions of the optic nerve between the chiasma and the nerve head (Fig. 7.1(h)). Common causes: Demyelinating lesions, optic nerve gliomata. (k) Fibre bundle (arcuate) defects. Lesions of the optic nerve between the chiasma and the retina (Fig. 7.1 (i)). Common causes: Vascular lesions, toxins, optic nerve gliomata, demyelinating lesions. (l) Bitemporal scotomata. Chiasmal lesions. The peripheral field may be affected later (Fig. 7.1(f)). Common causes: The same as for bitemporal hemianopia, but with special reference to glio mata of the optic chiasma if in children. Fig. 7.2 (continued) 52 Chapter 7 The second cranial nerve: the optic nerve (m) Homonymous scotomata. Unilateral lesions of the tip of the calcarine cortex (Fig- 7.l(j)). Common causes: Injuries, tumours. (n) Purely nasal defects. Although theoret ically due to lateral chiasmal lesions, this is rarely the case. Common causes: Glaucoma, and consecutive optic atrophy following papilloedema. In the latter, the lower nasal field is first affected, then the upper nasal and lower temporal, so that finally only an island of vision is left in the upper temporal fields. Fig. 7.2 (continued) of normal be gained which makes it possible to detect early abnor malities, and quite unexpected abnormalities may be found which alter the whole aspect of the problem. Presently, the visual field is tested by automated perimetry. An example of output of automated perimetry is shown in Fig. 7.3. Methods of examination To obtain a clear view of the fundus, there are certain requisites: (i) a good ophthalmoscope; (ii) a large pupil and (iii) a still field. In order to overcome the light reflex, diminish, if necessary, the illumination in the room; to overcome accommodation for near vision, instruct the patient to look at a distant point, which has been clearly defined, thus keeping the eye still. The temptation to look at the light of the ophthalmoscope is very great, and the patient must be warned not to do so. Use your right eye to examine the patient's right eye holding the ophthalmoscope in your right hand and then walk around the bed to examine the left eye with the left eye holding the ophthalmoscope in your left hand. Leaning over the patient is uncomfortable and prevents him from fixing on the distant object easily, while trying to use the wrong eye results in nasal collision. Part 2 The cranial nerves • i 9 w a 1 * , * «c it ft • « 1 • X >• • % -k • a * * a a a • i« •» •i t XI * "« * 4 •* ft :i X a a • t 2* X ry 17 • » •- * X r ti •i i If these measures fail, and especially when careful inspection of the periphery of the retina is important, it is perfectly justifiable to introduce a few drops of a quick-acting mydriatic. If homatropine is used, it must be remembered that (i) the pupillary reflexes must be tested first, (ii) it takes some time to achieve maximal dilation and (iii) it is followed by the use of eserine; otherwise, the results of the examination will be unpopular, in the elderly dangerous and in children the cause of a dilated, non-reacting pupil, perhaps for several days. Quick-acting mydriatics are readily available which avoid most of these problems. Features to be examined First find the optic disc. If the patient is carrying out instructions, this will immediately be seen; if not, it will be the vessels first seen, and by following them to their point of convergence the disc will come into view. Note its colour, the clarity of its edges and the size of the depression in its centre - the optic cup. Next, look at the vessels as they leave and enter the disc, the veins being the wider and darker; note the degree of curvature as they pass over the edge of the disc. 54 Fig. 7.3 Upper quadrant homonymous field defect, (a) Left eye. (b) Right eye. Chapter 7 The second cranial nerve: the optic nerve See if the vessels are continuously visible or if they appear and dis appear; if their curves are gradual or acute and tortuous. Now look at the points of crossing of the arteries and veins; note if one vessel appears to obliterate the other. Look next for haemorrhages, their size, shape and relationship to the vessels; and for exudates, their size, colour and texture. Finally, inspect the retina itself, noting any deviations from the uniformity of colour. While doing this, ask the patient to look directly at the light, when the vessel-free macula area can be inspected. Mydriatics may be required for this. A Sometimes the patient gets confused how to look at a distant point when the examiner is obstructing the field of vision of one eye. To avoid this, the patient may be instructed to use the free eye to focus at a distant point and imagine that they are looking at a distant object. Th e disc Pallor Disc colour varies greatly, probably even in the same individual at different times, and only long experience makes it possible to assess minor degrees of pallor. There are four main varieties of marked pallor: 1 Primary optic atrophy. The whole disc is chalky white, with well-defined sharp margins standing out dramatically like a full moon against a dark red sky. The causes include some neurodegenerative diseases (e.g. Freidriech's ataxia), toxic neurop athy or without any discoverable cause. The word 'primary' is purely a visual description. 2 Secondary optic atrophy. This follows papillitis or papilloedema and may also be called postneuritic or postpapilloedemic optic atro phy. The disc margins are blurred. Hie disc is grey or dirty green. Peripapillary sheathing and tortuous veins may also be present. 3 Consecutive optic atrophy. This follows the extensive disease of retina as in retinitis pigmentosa. The disc is yellowish waxy pale or white with disc margin being normal. The vessels are markedly contracted. 4 The disc is strikingly pale in a quadric or crescentic maimer on the temporal half, the so-called temporal pallor, due to lesions principally of the papillomacular bundle. This is often seen in multiple sclerosis and nutritional optic neuropathy, but is neither constant in that disease nor pathognomonic of it. 55 Part 2 The cranial nerves Difficulties and fallacies The temporal side of the disc is usually paler than the nasal side and, in the absence of other signs, this alone should not be considered pathological. Optic atrophy is a description of what the examiner thinks he is seeing and is not a diagnosis. One might just as well say that a patient is suffering from an extensor plantar response. There is always an underlying cause for optic atrophy and failure to appreciate this has often led to patients being allowed to go blind from remediable optic nerve compression. The myopic disc normally looks very pale, appears greatly enlarged and often has also a crescent of pallor immediately around it. Examine it through the patient's own spectacles to get the most accurate view. The optic cup is sometimes so marked as to give an impression of optic atrophy. As this is especially the case in glaucomatous cupping, when seen, the ocular tensions must be tested. Even here there is usually a rim of normal disc. Swelling Normally, the nasal edge of the disc is somewhat blurred. As swell ing develops, this blurring extends to the upper and lower margins. The changes in disc include: ® First, the optic cup becomes less evident and the disc of more uniform colour with the rest of the retina. The temporal edges are the last to disappear. ° The area covered by the disc then enlarges, the margins can not be defined and irregular radial streaks appear which give the disc an 'angry' appearance. The veins become swollen and engorged and the hump of the vessels entering and leaving the disc becomes very pronounced. The vessels appear and disap pear as they course near the disc. • Finally, small haemorrhages or large irregular ecchymoses appear, usually near a vein. In addition, white streaks and patches of exudate appear near the disc margins and spread outwards towards the macula to produce the macular fan. This is the appearance of papilloedema due to increased intracra nial pressure from any cause. Despite the degree of swelling, vision may be well preserved, the visual fields showing only enlargement of the blind spot (Fig. 7.2(i)). If the swelling is due to local lesions in the nerve (e.g. anterior-positioned optic neuritis, the so-called papillitis), there are important differences. The degree of swelling, although variable, is usually slight and is usually unilateral. The veins are not engorged, the humping is only slight and the disc area is not greatly enlarged. 56 Chapter 7 The second cranial nerve: the optic nerve There may be peripapillary haemorrhages and vascular 'sheath ing'. Vision is grossly disturbed due to large central or centrocaecal scotomata (Fig. 7.2(j)). On rare occasions, especially in childhood and adolescence, pap illitis may look more alarming, accompanied by a macular 'star' of exudates. The profound visual loss distinguishes this so-called neuroretinitis from papilloedema due to increased intracranial pressure. The Foster-Kennedy syndrome A tumour in the posterior inferior frontal region can cause optic atrophy on one side by compressing the optic nerve, and papilloedema on the other by obstructing venous return or causing raised intracranial pressure. Difficulties and fallacies • The hypermetropic disc is pink and its edges appear blurred, but it is also a small disc and the vessels and fields are normal. • Swelling occurs not only in intracranial disease, but also in generalized vascular disease, severe emphysema, profound anaemia and leukaemia. This emphasizes the importance of a general physical examination. • Central venous obstruction causes an appearance similar to that of increased intracranial pressure, but it is usually unilateral, is even more dramatic and angry-looking, the onset is abrupt and the visual failure marked. Haemorrhages are larger and more extensive, vary from day to day and may occur in the media. • Pseudopapilloedema is a disc appearance very similar to mild swelling, but without vessel engorgement, and symptomless throughout the patient's life. This term must not be confused with "pseudotumour", or benign intracranial hypertension, which is a syndrome where there is true papilloedema but not due to intracranial tumour. • Obliquity of the optic nerves, Drusen bodies (small colloid bod ies) near the nerve head, juxtapapillary choroiditis and haziness of the vitreous may all give the appearance of disc swelling. Some of these can be distinguished by the use of fluorescein (see p. 60). Th e vessels • Arteries are lighter and narrower than veins and often have a central reflecting line, so that a 'silver wire' appearance can be quite normal. • A very variable lumen, widening and narrowing throughout the course, is characteristic of arterial hypertension. Part 2 The cranial nerves 0 The arteries are thin and scanty following consecutive optic atrophy, and very thin after ophthalmic artery thrombosis when there is corresponding retinal pallor. • Minute emboli may be seen obstructing a retinal arteriole, while fragments of atheroma may be seen as refractile bodies blocking small vessels. These often indicate carotid artery disease. If the arteries are thickened (arteriosclerosis) in association with high pressure (hypertension), they compress the veins at points of crossing, obliterating them for a short distance, particularly on the side nearer the disc. Veins become engorged in states of high intracranial pressure and in ophthalmic venous obstruction, such as in thrombosis of the central vein. Tortuosity accompanies severe engorgement and veins may appear and disappear rather like a sea serpent. It is important to correlate veins with the appearances with quite normal veins is at all or papilloedema that edges. the degree of engorgement of the of the disc. Dubious papilloedema probably either not papilloedema has subsided leaving blurred disc Both arteries and veins show gross tortuosity in angiomatosis of the optic nerve and retina, producing bizarre appearances. An unusually tortuous artery should be traced to its periphery because it may lead to a retinal angioma, and retinal angiomata may be associated with cerebral angiomata, hamartomata or cerebellar haemangioblastomata. Th e retina The number of possible abnormalities in the retina and choroid is so large that descriptions will best be sought in textbooks of oph thalmology. It is important to have a working knowledge of these appearances, as they may be the cause of visual failure or field defects thought to be of more central origin. The important neuro logical features are described below. Haemorrhages These occur as: 1 Small streaks, near the vessels, long and linear, or flame-shaped. 2 Much larger ecchymoses capable of obscuring local vessels and retina. Types 1 and 2 occur in raised intracranial pressure and Chapter 7 The second cranial nerve: the optic nerve in venous engorgement of any cause: in hypertension, systemic vasculitides and haemorrhagic disorders. In the latter, they extend to the retinal periphery. 3 Small rounded pin-head areas. These are microaneurysms of the retinal vessels and occur in diabetes. 4 Subhyaloid haemorrhages. Seen in subarachnoid haemorrhage, they appear as large effusions of blood related to and often below the disc, with crescentic inner and clear-cut outer borders extending forwards towards the lens. V 4^ txudafes • d P!®0 Hard exudates are the result of extravasation of fluid which is rich in fibrin and proteins. These exudates are deposited as masses. They accumulate and are seen as a cluster around leaking microan eurysms. In the macular region, sometimes exudates may form the shape of a star (macular star as in malignant hypertension). Cotton-zvool retinal patches, so-called because of their fluffy appear ance, are seen in papilloedema, diseases such as renal failure asso ciated with hypertension and in arteriole disease associated with systemic vasculitis such as polyarteritis nodosa or systemic lupus erythematosus. They can also occur in retinal embolism and severe anaemias. The cotton-wool spot is essentially a focal ischaemic reac tion (microinfarct) of injured axons. Exudates are not themselves diagnostic, and must be correlated with the appearance of the disc and vessels, and indeed with the rest of the clinical findings. Retinal 'vasculitis' This phrase denotes the combination of inflammatory changes in relationship to retinal vessels. There may be focal or extensive sheathing of vessels, or actual occlusion, retinal haemorrhages, with cells in the vitreous and abnormal leakage on fluorescein angiography (see below). Some causes are infectious, namely cyto megalovirus in association with acquired immune deficiency syn drome, tuberculosis or syphilis, others inflammatory in association with multiple sclerosis or systemic vasculitides. Tubercles Choroidal tubercles appear, under ophthalmoscopic examina tion, much larger than tubercles as seen at post-mortem. They are rounded, about half the size of the disc, yellowish at the centre, but with ill-defined pink edges, slightly raised and, if present for any length of time, surrounded by pigmentation. Part 2 The cranial nerves Phakoma A phakoma is a collection of neuroglial cells appearing in the ret ina as a large white, or bluish-white, plaque with almost translu cent edges, about one-half to two-thirds the size of the optic disc. Although on occasion seen in neurofibromatosis, they are most characteristically a feature of tuberous sclerosis. Pigmentary abnormalities Many choroidoretinal pigmentary lesions are not directly of neu rological importance. Reduction of pigment in fair-haired people may allow choroidal vessels to be seen so clearly as to mimic an angiomatosis. Gross choroidoretinitis sometimes accompanies neurosyphilis. hi cerebromacular degeneration, there may be a cherry-red spot at the macula, surrounded by darker spots of pigment, sometimes a granular peppery appearance over a wide area, with retinal atrophy. In toxoplasmosis, large areas of pigmentary abnormality, irreg ularly oval in shape with clearly defined edges, are seen near the macula. At the periphery, the most important change is the spidery black 'bone corpuscles' pigmentation spreading towards the centre that is seen in retinitis pigmentosa, and to a lesser degree in several other heredofamilial disorders. Opaque nerve fibres If myelinated nerve fibres persist in the retina, a vivid white fan spreads from the disc, curving in the course of the nerve fibres. This is often mistaken for an exudate, but the vivid whiteness and direction are quite different. The edges of such a patch are usu ally curvilinear and clear-cut, while the peripheral edge gradually streaks off into the retina. Fluorescein and fundus photography Retinal photography is a standard procedure used by ophthalmological and neurological departments. The preliminary injection, intravenously, of a carefully sterilized solution of 10% fluorescein adds greatly to the interpretation of certain appearances. The dye does not escape from normal arterioles, but in hypertensive reti nopathy, areas of arteriolar necrosis are clearly seen; in retinal arte rial occlusion, the failure of filling is seen, and in central venous occlusion gross exudation will occur. This is also seen in true Chapter 7 The second cranial nerve: the optic nerve papilloedema, but is absent in pseudopapilloedema. In true optic atrophy, as opposed to doubtful disc pallor, the disc remains dark throughout serial photographs. The microaneurysms of diabetic retinopathy are seen earlier, and shown to be more extensive than ordinary ophthalmoscopy can detect. But interpretation may be equivocal; taken in isolation, the result should not of itself trigger extensive further investigation. 61 8 The third, fourth and sixth cranial nerves: the oculomotor, trochlear and abducent nerves The actions of these nerves are so closely linked that they are con sidered together, and it is at this stage that abnormalities on inspec tion of the eyes are usually detected. Some of these relevant to neu rological disease are mentioned in this section. Anatomy Three cranial nerves control eye movements: oculomotor, trochlear and abducent (Fig. 8.1). O culom otor nerve and nucleus (lllrd nerve) The nucleus is made up of both paired and unpaired nuclei and is located in periaqueductal grey matter of upper midbrain. This complex has four paired subnuclei giving innervations to the medial, superior, inferior recti and inferior oblique muscles. Levator Oculomotor nerve (lllrd nerve) Superior obliqi (IVth nerve) Superior rectus (lllrd nerve) Lateral rectus (Vlth nerve) (IVth nerve) Inferior rectus (lllrd nerve) 62 Inferior oblique (lllrd nerve) Medial rectus (lllrd nerve) Fig. 8.1 Course of the lllrd, IVth and Vlth nerves. Chapter 8 The third, fourth and sixth cranial nerves palpebrae superioris is innervated by a single caudal midline nucleus, which when involved results in bilateral ptosis. The innervations to the superior rectus are contralateral and the motor fibres decussate inside the Illrd nerve nucleus and join the fascicle of the opposite oculomotor nerve. Therefore, a lesion on one side of the Illrd nerve nucleus can involve both superior recti due to involvement of ipsilateral subnucleus and contralateral decussating fibres. Tire fascicles combine to form a nerve at the exit from midbrain. Trochlear nerve (IVth nerve) Tire trochlear nerve arises from the nucleus located in the midbrain at the level of inferior colliculus and exits from the dorsum of mid brain. It has the longest course and supplies superior oblique muscle. Abducent nerve (Vlth nerve) The abducent nerve arises from a nucleus located in the mid to lower pons in the floor of the fourth ventricle. It supplies the lateral rectus muscle. Functions Control of all the external ocular muscles and the elevators of the lids. Autonomic fibres running in relation both to these nerves and the Vth nerve regulate pupillary muscles. Purposes of the tests 1 To inspect the pupils and determine if any abnormalities dis covered are due to local disease, a peripheral autonomic lesion or nuclear involvement in the brainstem. 2 To examine eye movement and to determine if any defect is of local muscular origin, or due to lesions of the oculomotor nerves peripherally, their nuclei in the brainstem or the path ways of supranuclear control. 3 It is also convenient at this stage to detect and analyse nystag mus, though this is closely related to vestibular dysfunction. M ethods of examination Preliminary inspection While talking to the patient, note the presence or absence of ptosis and squint, whether it is unilateral or bilateral, constant or variable. 63 Part 2 The cranial nerves Compare the size of the palpebral fissures and note if the eyeballs are protruded (exophthalmos) or recessed (enophthalmos). Exophthalmos Assess protrusion by looking down on the eyes from above and behind the patient's head. Ocular prominence may be normal, but true exophthalmos, with lid retraction, occurs in thyrotoxicosis, and becomes severe, with injection and chemosis of the conjunc tiva, oedema of the lids and oculomotor paralysis in dysthyroid eye disease (exophthalmic ophthalmoplegia). Protrusion can become severe in craniosynostosis and may be associated with downwards displacement of the globes in hydrocephalus. Unilateral exophthalmos, still most commonly due to thyroid dysfunction, may also be caused by orbital or retro-orbital neo plasms and granulomata, and when the globe cannot be replaced by pressure, all eye movements may be limited, but the lid is nor mal. The exophthalmos, ptosis and oculomotor palsy become gross iit tumours or thrombosis of the cavernous sinus, and if the globe pulsates and there is a bruit audible to patient and examiner, this is characteristic of a caroticocavernous fistula. Enophthalmos Although most commonly part of Horner's syndrome (q.v.), this may be due to a maldeveloped eye. Do not be caught out by a well fitting but recessed prosthesis, or by the true abnormality being prominence of eye or orbit on the other side. Hypertelorism Though not uncommon in normal individuals, this should alert one to the possibility of intracranial congenital abnormalities. Look next at the conjunctiva, cornea and iris while the patient moves the eyeball in all directions. Note colours, pigmentary abnormalities, surgical or developmental defects. The conjunctiva 1 Subconjunctival haemorrhage is common following cranial trauma and rare in spontaneous subarachnoid haemorrhage. 2 Telangiectases may be associated with skin telangiectases and cerebellar ataxia in the syndrome 'ataxia telangiectasia' (LouisBar syndrome). 3 Retro-orbital tumours may grow forwards as a red or grey felting visible on extreme deviation of the eyeball. 4 A yellow tint may be the only sign of liver disease and raise the possibility of metastatic or diffuse systemic disease. 64 Chapter 8 The third, fourth and sixth cranial nerves Fig. 8.2 KF rings as crescent most marked at the upper margin. 5 An intense inflammatory reaction occurs in leptospira canicola infection, which also causes an acute meningitis, and dining the migration of filaria (loa loa), which also may cause an encephalitis. 6 Conjunctival ulcers form a part of the triad of Behcet's syndrome (q.v.). The cornea and iris Look particularly for different colouring of the two eyes; the pale atrophied iris of the tabetic and the Kayser-Fleischer (KF) ring. For merly held to indicate premature vascular disease, the arcus senilis, or translucent ring overlying the iris, is a non-specific sign common in the elderly. The KF ring This is a very characteristic golden-brown ring, lying just inside the limbus of the cornea, sometimes forming a complete circle, but at other times merely a crescent at the upper and lower margins (Fig. 8.2). There is a clear zone between the KF ring and the limbus, which distinguishes the ring from arcus senilis or juvenilis. It is seen most easily in light-coloured eyes and is diagnostic of hepa tolenticular degeneration. If it is suspected, its presence should be confirmed by slit-lamp examination. After treatment with penicilla mine, the vivid colour of the ring changes to a dull mottled brown. The eyelids Note the position of the lids in relation to the iris, and the width of the palpebral fissure. Then ask the patient to open the eyes widely and notice both the lid movement and the degree of movement of the frontalis muscle. Ask the patient to follow an object upwards and maintain forwards gaze for at least 30-45 seconds without blinking. 65 Part 2 The cranial nerves O Congenital Acquired Neurogenic • Horner's syndrome • Illrd nerve palsy Myogenic 0 Mitochondrial myopathies • Oculopharyngeal muscular dystrophy • Myotonic dystrophy Neuromuscular junction • Myasthenia gravis Ptosis is always partial. Ptosis If the pupil is partly covered by the upper lid, there is a slight degree of ptosis, which may be due to paralysis of the levator palpebrae superioris as a result of a Ulrd nerve lesion, or to weakness of the tarsal mus cles due to a sympathetic lesion. In the latter, tire lid can still be raised voluntarily, but in the former, the frontal muscles contract to overcome the drooping and there may be a permanent wrinkling of tire forehead. In myasthenia gravis, the degree of drooping of the lid varies from moment to moment, and may change sides. The lid will droop progressively on prolonged upwards fixation, but a blink restores its position to normal. In ocular myopathy (mitochondrial or dystrophic), the ptosis is fixed and the head is often held extended in an attempt to see under the drooping lids. Lid retraction The lid is buried under the brow and the sclera is clearly visible above the iris in hyperthyroidism, after large doses of anticholin esterase, and in some normal patients. The pupils Look at the size, shape, equality and regularity of the pupils. Corre late their size with the surrounding illumination, remembering that the pupil nearer a bright window is often smaller. It is normal for 66 Chapter 8 The third, fourth and sixth cranial nerves the pupils to be very small in early infancy, old age, during sleep and in bright light, and to be large in poor light, myopia and fright ened children. The reaction to light It must be possible to see this reaction. If, therefore, the pupils are small, first darken the room, and give the patient the same instructions as for examining the fundi. A bright beam of light is then shone suddenly from slightly to one side of the eye (shining from directly in front may cause the patient to converge the eyes, when the pupils will contract anyway). The pupil should constrict briskly. Repeat the test for the other eye and compare the two reac tions. Finally, shield one eye, shine the light in the other, and watch for the consensual reaction, which is the constriction of the shielded pupil as well. The reaction to convergence and accommodation for near vision The patient should still be fixing on a distant object. Explain that he is about to be asked to look at a near object, and then place a pencil suddenly about 22 cm in front of the bridge of the nose. This position prevents lowering of the lids which obscures the normal pupillary constriction, and the sudden movement emphasizes it. Return the eyes then to the distant object, for the subsequent dilata tion may be even easier to see. Pupillary abnormalities The constricted pupil (miosis) This indicates a lesion at some point in the very circuitous path way taken by the sympathetic supply to the pupillary dilator muscle. Thus, the lesion may be in the hypothalamus, brainstem, lateral aspect (the spinal cord as far down as the upper thoracic segments), the sympathetic chain, the cervical sympathetic gan glia, the pericarotid plexus or in the sympathetic fibres which run to the orbit accompanying the ophthalmic division of the Vth cranial nerve. Pontine tumours or haemorrhages, primary or sec ondary tumours involving the cervical sympathetic chain, and vascular lesions of the carotid artery or its sheath, are the most common causes, but there are many others. Bilateral spontaneous sympathetic palsy almost invariably means an upper brainstem lesion. Remember, however, that a sleeping patient, even if his 67 Part 2 The cranial nerves eyes are not closed, may have very small pupils which enlarge rapidly when roused. Horner's syndrome In its complete state, this consists of miosis, ptosis, enophthalmos and dryness and warmth of that half of the face (see Fig. 8.4(c)); cocaine will not dilate the pupil as it normally does, and adrenaline dilates it more than normally. This syndrome in its purest form is seen in lesions of the superior cervical ganglion in the neck, and of the fibres surrounding the carotid artery, but it can be seen in other lesions of the sympathetic supply, though usually with other physi cal signs to help localization. It used to be common after carotid arteriography carried out by direct puncture. It may occasionally still be seen after carotid catheterization. It occurs after many bouts of periodic migrainous neuralgia or cluster headache. The dilated pupil (mydriasis) In practice, this results from paralysis of the parasympathetic fibres, either at their origin from the pretectal nuclei and the EdingerWestphal nucleus in the midbrain, during their course with the Illrd nerve, or at the ciliary ganglion in the orbit. The possible sites of a lesion extend over a much smaller distance than in the case of the sympathetic division. Most commonly, such lesions are due to vas cular accidents in the midbrain, tentorial herniation (due to cerebral space-occupying lesions) or aneurysms of the carotid artery. Never forget, however, that a very common cause of a dilated pupil is that someone has put in a mydriatic. When atropine was commonly used, it could be seen in children for several days afterwards. An eye that is almost completely blind may have a dilated pupil. The pupil that does not react to light This is due to a break in the pathways for the light reflex, and the lesion may lie in the afferent loop, i.e. the retina, optic nerve or chiasma; or in the efferent loop, i.e. the parasympathetic supply from the midbrain running with the Illrd nerve. In the former, as no stimulus can be received there will also be no consensual reaction, but if the lesion is unilateral (e.g. optic nerve compression or neu ritis) when the normal side is stimulated, both pupils will react. If the efferent loop is involved, however (e.g. causes as listed above), the affected pupil is unable to react no matter which side is stimu lated. Bilateral failure to react, with intact vision, usually means a midbrain lesion. Bilateral blindness with non-reacting pupils must be due to a lesion between the retina and the first part of the optic 68 Chapter 3 The third, fourth and sixth cranii Chapter 8 The third, fourth and sixth cranial nerves tract, because after that the pupilloconstrictor fibres have left the visual fibres. All these remarks assume that someone has not recently instilled a mydriatic to examine the fundi and omitted to record the fact. The pupil that fails to accommodate for near vision This is most commonly due to a failure in the technique of the examination. If, however, it is genuine, it usually results from fail ure to converge owing to upper brainstem lesions, such as tumours or encephalitis, or in parkinsonism. The Argyll Robertson pupil Often incorrectly hyphenated, this name is given to a small irreg ular pupil with impaired or absent reaction to light, reacting to accommodation, but responding poorly or not at all to atropine, physostigmine and methacholine. The pupil is, however, not always small and the two sides may differ. It usually is caused by syphilis, but may occasionally be seen in other midbrain lesions. The site of tire lesion has never been fully determined. Some favour the tectum of the midbrain, others the ciliary ganglion, but bearing in mind the nature of neurosyphilitic lesions, multiple sites camrot be excluded. The myotonic pupil (Holmes-Adie syndrome) Correctly hyphenated, when this presents it is often in young women and is first seen as unilateral dilatation of one pupil with failure to react. However, if the patient is kept in a dark room for a quarter of an hour and the pupil then exposed to a diffuse light, a slight slow reaction may be seen. The most important feature, and one which is much less time-consuming to elicit, is the very slow constriction that occurs on maintaining convergence for 45 seconds or more. If the patient then looks at a distant object, the dilatation of the myotonic pupil is similarly slow, so that there may be a stage at which the once larger pupil is now smaller than its fellow. As reaction to light is almost absent, the myotonic response to conver gence can often be better seen by illuminating the pupil at the same time with not too bright a torch. Minute doses of methacholine 2.5%, insufficient to affect a normal pupil, cause brisk contraction. Bilateral myotonic pupils may rarely occur, hi the full syndrome, the knee and ankle jerks are also absent and occasionally there is complete areflexia. The site of the lesion causing the pupillary abnormality is probably in the ciliary ganglion. 69 Part 2 The cranial nerves Examining ocular movement The patient must look at a clear and definite point, such as the point of a pen or a fine point of light. Hie examiner then moves this point precisely and deliberately to right and left horizontally, upwards and downwards in the midline and vertically when the eyes are deviated to one side. Do not attempt to make the eyes deviate beyond the point of comfort, and hold each deviation for at least 5 seconds. The aim is to: 1 Observe lagging of one or the other eye. 2 Analyse any diplopia the patient may describe (see below). 3 Detect nystagmus. The action of the ocular muscles Complex though these are, one cannot understand the tests of ocu lar movement without learning a scheme of muscle action. This is a simplified form of considerable practical value. I A. The lateral rectus (Vlth nerve) - moves the eye horizontally outwards. B. The medial rectus (Bird nerve) - moves the eye horizontally inwards. C. The superior rectus (Illrd nerve) - elevates the eye when it is turned outwards. D. The inferior oblique (Illrd nerve) - elevates the eye when it is turned inwards. E. The inferior rectus (Illrd nerve) - depresses the eye when it is turned outwards. F. The superior oblique (IVth nerve) - depresses the eye when it is turned inwards. Tire muscles paired together above act together. The vertical recti and obliques have other important actions, e.g. the vertical recti do, of course, turn the eye upwards or downwards, but it is only when the eye is in the positions mentioned that these muscles have the pure elevating or depressing action that is easiest to analyse. A secondary action of the superior oblique is to rotate the vertical meridian (see importance following p. 74). If the action of one of the obliques is remembered, the others can easily be deduced (Fig. 8.3). Ocular muscle paralysis From this scheme, the following general rules can be deduced. 1 If an eye fails to move outwards, there is either a Vlth nerve lesion or a local lesion of the external rectus muscle (see (A) above). Chapter 8 The third, fourth and sixth cranial nerves Inferior oblique Tem poral sid e Lateral rectus Fig. 8.3 The direction of muscle action in right eye. Superior oblique © Site Common causes Nerves involved • Brainstem • Stroke (ischaemic/haemorrhagic) • Demyelination • Intraxial neoplasm • Illrd - midbrain • Vlth-pontomedullary junction • Meningeal • • • • Meningitis Raised intracranial pressure Aneurysms Cerebellopontine angle tumour • Trauma • inrd, IVth and Vlth • Vlth; Illrd (uncal herniation) • Illrd (posterior communicating artery aneurysm) • Vlth • Illrd, IVth and Vlth • Cavernous sinus • Infection/Thrombosis • Aneurysm • Caroticocavernous fistula • Illrd, IVth and Vlth • Superior orbital • Granuloma, tumour fissure • Illrd, IVth and Vlth • Orbit • Illrd, IVth and Vlth • Ischaemic (diabetes, vasculitis) • Infection • Tumour • Trauma 71 Part 2 The cranial nerves 2 If the eye, when deviated inwards, will not then move down wards, there is either a IVth nerve lesion or a local lesion of the superior oblique muscle (see (F) above). 3 All other defects in movement are due to Illrd nerve lesions, a local lesion of the muscles, myasthenia gravis or an internuclear ophthalmoplegia. In addition, however, when there is ocular muscle paralysis, the unopposed pull of the normal antagonist will displace the eye ball. In paralysis o f the Illrd neroe, therefore, the eye is displaced outwards and will not move inwards or vertically. There is accom panying ptosis and pupillary dilatation (Fig. 8.4(a)), though the latter may be absent if the lesion is recovering (Fig. 8.4(b)) or is incomplete. In Vlth neroe paralysis, the eye is deviated inwards and will not move outwards beyond the midline, but will move vertically when deviated inwards (Fig. 8.4(d) and (e)). Unfortunately, incomplete paralysis may show little visible abnormality and it is then that the diplopia produced must be investigated. The analysis of diplopia Diplopia can indicate ocular muscle weakness before it is evident to the examiner. The light rays fail to fall on exactly corresponding parts of the two retinae, and a false image is formed which is usu ally paler and less distinct. The rules governing the relationship of these two images are as follows: Rule 1 Displacement of the false image may be horizontal, vertical or both. Rule 2 Separation of the images is greatest in the direction in which the weak muscle has its purest action. Rule 3 The false image is displaced furthest in the direction in which the weak muscle should move the eye. M ethod of examination Cover one of the patient's eyes with a transparent red shield and, using a point of light, move the object as described above (p. 70). In each position, ask the patient: 1 Whether he sees one object or two. 2 If double, do the two images lie side by side or one above the other? 3 In which position are they furthest apart? 4 Which is the red image? 72 Chapter 8 The third, fourth and sixth cranial nerves Fig. S.4 Partial left Illrd nerve palsy; carcinoma of nasopharynx. (a) Ptosis and lateral deviation of left eye. (b) Upwards gaze; slight elevation of left lid reveals dilated pupil and paralysis of elevation of left eyeball, (c) Horner's syndrome; ptosis, raising of lower lid, miosis, enophthalmos, but no overaction of frontalis, (d) and (e) Vlth nerve paralysis. The right eye is deviated inwards and will not move outwards across the midline. 73 Part 2 The cranial nerves Interpretation of results If the images are exactly side by side, it will be only the external or internal recti that are involved. If they are one above the other, either of the obliques, or the superior and inferior recti, may be defective. The muscle pair involved It is the position in which there is maximum displacement that indicates which pair of muscles is involved (see Rule 2, p. 72), e.g. if maximal displacement occurs when the eyes are deviated to the right and upwards, this is the movement carried out by the right superior rectus and the left inferior oblique (see (C) and (D), p. 70). The individual muscle responsible To determine which of the two muscles in each pair is at fault, the patient must say which of the two images is displaced furthest, the red or the white. Using the same example, if the red glass were over the right

eye, arid on looking to the right and upwards the red image is furthest displaced, then it must be the muscle mov ing the eye in that direction (see Rule 3, p. 72), namely the superior rectus, that is at fault. If it were the white image that was furthest displaced, the fault would be with the left inferior oblique. It is a good exercise to work out the
situation in each direction for each possible muscle fault, and the more frequently this is done, the easier it becomes. In practice, of course, multiple faults occur, and, particularly when the Illrd nerve is involved, it is at times easier to work out which muscles are acting normally. But in the presence of a Illrd nerve palsy, it is particularly difficult to
detect paraly sis of the superior oblique muscle. The eye then is in the abducted position and as a result the superior oblique cannot produce a downwards movement of the eye; its contraction results only in torsion, a movement that is very readily overlooked. Various charts exist for representing diplopia diagrammatically, of which the Hess chart and
those used in orthoptic analysis are the most useful. They do not, however, offer better evidence than is afforded by carefully following the scheme of combination just described. Conjugate ocular movement Normally, of course, the eyes do not move independently. For instance, each external rectus muscle contracts in conjunction with the opposite
internal rectus, producing a movement so organized 74 Chapter 8 The third, fourth and sixth cranial nerves that the visual axes remain in the same relationship throughout and diplopia does not occur. The centre for the control of conjugate movement to, say, the right is situated in the posterior part of the left frontal lobe and there is probably a
further centre in the occipital region. The final common pathway for controlling this movement lies, however, in the brain stem and depends upon connections, running through the median longitudinal bundle, between the innervation of the left internal rectus and the right external rectus. A lesion of the frontal lobe causes contralateral paralysis of
con jugate gaze; a lesion of the brainstem causes ipsilateral paralysis of conjugate gaze. Paralysis of the normal control of conjugate deviation to one side usually results in deviation of the eyes to the opposite side, which may be slight and transient, or marked and permanent. Taking an example, it will be seen from these facts that deviation of the
head and eyes to the right can result from several different types and positions of lesion. 1 Destructive lesions (vascular or neoplastic), involving the supranuclear pathways between the right frontal lobes and the oculomotor nuclei, paralyzing normal conjugate deviation to the left. 2 Destructive lesions of the left side of the brainstem (vascular
accidents or pontine neoplasms), also paralyzing conjugate deviation to the left. 3 Irritative lesions (focal fits) of the left frontal lobe, stimulating deviation to the right. Not theoretical - it occurs. 4 Irritative lesions of the right side of the brainstem (encephalitis), also stimulating deviation to the right. Failure of upwards deviation follows an upper
midbrain lesion due either to a vascular accident, compression by a tumour in the neighbourhood of the Illrd ventricle or tentorial herniation. Fixed upwards deviation is rare, but prolonged upwards devia tion occurs in the oculogyric spasms following administration of certain drugs (benzodiazepines or metoclopramide) either in heavy dosage or to a
hypersensitive individual. As different sites of lesion can produce similar abnormalities of conjugate deviation, in order to distinguish them it is necessary to bring other neurological manifestations into the picture, and to test ocular movement on reflex fixation. In the supranuclear destructive lesions, the face and limbs are paralyzed on the side
opposite to the ocular deviation; in brainstem lesions, on the same side; but in the latter, if the face is paralyzed, it will be a lower motor neuron lesion on the side opposite to the 75 Part 2 The cranial nerves hemiplegia. The situation is reversed for irritative lesions, but the deviation is spasmodic. If the patient is able to cooperate and there is a failure
of con jugate movement in any direction, he should be told to fix his eyes on a particular object while the examiner turns or flexes the patient's head passively according to the defect. If the defect is of supranuclear origin, the eyes will move throughout their full range on reflex fixation, even though voluntarily they cannot do so. If the lesion is at the
level of the nucleus or peripheral to it, the eyes will still fail to move across the midline in the direction of the defect. Internuclear ophthalmoplegia When there is a lesion of the brainstem involving the median lon gitudinal bundle (and so disrupting connections between one Illrd nerve nucleus and the opposite Vlth nerve nucleus), there is disor-
ganization of the conjugate movement of the two eyes when look ing laterally (Fig. 8.5). Internal deviation of the adducting eye is defective and 'out of step' with the external deviation of the other eye. A very marked nystagmus may develop (see 'ataxic nystag mus', p. 81). Fig. 8.5 Internuclear ophthalmoplegia. (a) Ocular deviation to the left is normal.
(b) To the right, internal deviation of the left eye is defective and the eye turns downwards. External deviation of the right eye is dissociated, and accompanied by marked nystagmus. 76 Chapter 8 The third, fourth and sixth cranial nerves Detecting and analysing nystagmus The balance of tone between opposing ocular muscles is influenced by
impulses from the retina, the muscles themselves, the vestibular nuclei and their central and peripheral connections and by proprio ceptive impulses from neck muscles. It is normally so maintained that the eyes at rest remain in the midline. Any disturbance of this balance results in a drift of the eyes in one or other direction. If this drift is then
corrected by a quick movement back to the original position, and this cycle is repeated frequently, nystagmus results. It is intended here to describe nystagmus as it is actually seen by the examiner and to indicate the possible significance o f the d iffer ent varieties. Purpose o f th e te s ts 1 To detect nystagmus 2 To determine its direction, rate and
amplitude ' '•hm ei 3 To deduce from its character whether it is due to peripheral labyrinthine or central vestibular disease, muscle weakness or visual defects M ethods of testin g Watch the patient's eyes while taking the history. Nystagmus on forwards gaze can be noted at once. Note whether it is constant or occurring in bursts. On direct
examination, the patient must be told once again to look at a definite point, such as the tip of a pencil. Hold this object first in the midline, and then move it clearly and deliberately to left, to right, upwards and downwards. During this last movement, the eyelids should be held up by the other hand. Lateral deviation should be maintained for at least 5
seconds to give time for the nys tagmus to occur and for it to be assessed. Note that the deviation should never be to the extreme of lateral movement. If the patient has to strain in order to see the object, there is likely to be a spurious nystagmus in quite normal people. Recording nystagm us In describing nystagmus, the terms used must be quite
clear. There is a great deal of confusion caused by different people meaning different things. The information required is: 1 The position of the eyes when nystagmus occurs. 77 Part 2 The cranial nerves 2 The deviation that produces its greatest amplitude, usually to the side towards which the quick phase beats. 3 The direction, in each oscillation, of
the fast movement (not which deviation produces the quickest oscillation). (a) Conventionally, the direction of nystagmus is described according to its fast movement (see caloric tests, p. 104). To avoid confusion, it is probably wise to be a little long-winded and describe what is seen, e.g. 'on deviation to the right, fine horizontal nystagmus, fast
component to the right'. (b) Alternatively, especially useful in recording a changing clin ical situation, the nystagmus can be described by 'degree'. First degree: when present only on deviation of the eyes Second degree: when also present looking straight forwards Third degree: when visible even on gaze in the direction opposite to the fast beat The
presenting types of nystagmus Pendular nystagm us A rapid horizontal oscillation to either side of the mid line, of equal amplitude, but variable speed, present and usually obvious on forwards gaze, increased by fixation, but often losing its pendu lar character on lateral deviation. It is difficult to believe that any object is stationary to these patients.
Causes. Visual defects from earlier years including macular abnormalities, choroidoretinitis and albinism; high infantile myo pia; opacities of the media. Congenital nystagmus is also pendular, and horizontal, but becomes 'jerky' on lateral gaze, and remains horizontal on upgaze and downgaze. It is important to avoid unnecessary investigation in these
patients. Jerk nystagm us Horizontal nystagmus The rhythm of this common type of nystagmus is that of a slow drift in one direction and a fast correcting movement in the other. It may be present at rest, or only on ocular deviation; so coarse as 78 Chapter 8 The third, fourth and sixth cranial nerves almost to lose its characteristics, or so fine as to be
visible only when using the ophthalmoscope. 'Jerk nystagmus' may also be vertical or rotary, the significance of which is described below, but note that 'horizontal' nystagmus is a to-and-fro movement in the horizontal plane, no matter in which direction the eyes may be deviated to demonstrate it. Causes. Jerk nystagmus is produced by disturbances
of the ves tibular system. These may occur peripherally in the labyrinth, centrally at the nuclei, in the connections between these two (the vestibular nerve) or between the nuclei and the ocular muscles (the median longitudinal bundle); it is seen in lesions of the cerebellum (though probably because the type of lesion responsible usually involves the
cerebellovestibular connections) and in lesions of the uppermost cervical region. In more peripheral lesions, the quick phase is away from the lesion, and the amplitude is greater in the direction of the quick phase. In cerebellar lesions, and disturbances of its brainstem connec tions, the quick phase and the greatest amplitude is towards the side of
the lesion. In cerebellopontine angle lesions, there are both central and peripheral effects, but the amplitude is greater towards the side of the lesion. Peripheral lesions usually have additional vertigo, tinnitus and deafness, such as in Meniere's disease, and acoustic neuromata, though the last produce central effects as well. The more central lesions
tend to be more chronic, may cause no tinnitus or deafness and vertigo is less constant. Examples include multiple sclerosis, vascular lesions, or tumours of the cerebellum, IVth ventricle and cerebellopontine angles (when Vth and Vllth nerves may also be involved). Vertical nystagmus This is not nystagmus on looking upwards, but nystagmus in
which the oscillation is in an up-and-down direction, no matter what the position of the eye. It is never labyrinthine in origin, and the quick phase is most often upwards (upbeat nystagmus). Causes. It is important to recognize this type of nystagmus because whereas horizontal nystagmus can arise from many sites in the vestibulo-ocular pathways,
vertical nystagmus is essentially due to intrinsic disturbance of the brainstem, such as in vascular accidents, encephalitis, multiple sclerosis, syringo bulbia, and secondary to compression from cerebellar tumours, basilar invagination with tonsillar descent and the Arnold-Chiari 79 Part 2 The cranial nerves syndrome. However, drugs remain the
commonest cause of vertical nystagmus which, when combined with a horizontal jerk variety, again always indicates a brainstem dysfunction. Benzodiazepines, barbiturates and phenytoin are often responsible. Rotary nystagmus This type of nystagmus also has fast and slow components, but the oscillations are of rotary character. Causes. It occurs
both in labyrinthine and brainstem disease. In peripheral disorders, it is usually at the acute stage, and is tran sient; if long-standing, it indicates disease of the vestibular nuclei, especially the inferior portion. Any of the disorders causing vertical nystagmus can be included here. © P endular nystagm us • Visual defects from earlier years including
macular abnormali ties, choroidoretinitis and albinism • High infantile myopia 8 Opacities of the media. Je rk nystagm us Horizontal nystagmus 8 Disturbances of tire vestibular system 8 Lesions of the cerebellum 8 Lesions of the uppermost cervical region Vertical nystagmus 8 Intrinsic disturbance of the brainstem such as in vascular ac cidents,
encephalitis, multiple sclerosis, syringobulbia 8 Secondary to compression from cerebellar tumours, 8 Basilar invagination with tonsillar descent 8 Arnold—Chiari syndrome 8 Drugs such as benzodiazepines, barbiturates and phenytoin Rotary nystagmus • Labyrinthine and brainstem disease • Any of the disorders causing vertical nystagmus N ysta g
m u s of dissociated rh yth m in the tw o eyes • Lesions of the median longitudinal fasciculus 8 Multiple sclerosis 80 Chapter 8 The third, fourth and sixth cranial nerves Nystagm us of dissociated rhythm in the tw o eyes This is the so-called ataxic nystagmus in internuclear ophthalmo plegia. There is defective inwards movement of the adducting eye
with a fine nystagmus, accompanied by a coarse, irregular, rather dramatic nystagmus of the abducting eye, of dissociated rate and rhythm. 'Ataxic' nystagmus may be mimicked by asymmetrical muscle weakness in myasthenia gravis. Causes. Lesions of the median longitudinal bundle. This most com monly, but by no means invariably, is caused by
multiple sclerosis. Rare forms of nystagmus See-saw nystagmus The name is self-explanatory. This is spontaneous nystagmus, one eye moving up while the other moves down, the movement being accompanied by a conjugate rotation. This rare phenomenon has most often been seen in lesions in the suprasellar region anterior to the Illrd ventricle.
Convergence-retraction nystagmus Attempted upgaze, usually defective, provokes a jerk nystagmus with the fast phase inwards, in a convergent manner. Lesions of the upper midbrain near the pineal are responsible. Downbeat nystagmus A vertical nystagmus, but here the fast phase is downwards and it is particularly provoked by lateral gaze. It is
characteristic of lesions at the foramen magnum, commonly Chiari malformations, and the accompanying symptom is oscillopsia - awareness of the apparent vertical movement of objects. Difficulties and fallacies 1 Misleading results come from failure to ensure that the patient understands what is required of him, from expecting the eyes to be able to
deviate as far as a chameleon's and by not allowing the nystagmus time to appear. 2 A patient with a hemianopia often has apparent difficulty in looking to that side. If told to look to the left, or towards the window, and to keep looking that way, an object on which he can fix can then be brought into Inis normal field and the results will be the same. 81
Part 2 The cranial nerves 3 Weakness of an ocular muscle, most often the external rectus, will give an impression of nystagmus, but is usually irregular, non-rhythmic and accompanied by diplopia. In myasthenia gravis, a very irregular nystagmus is often seen which will dis appear on appropriate treatment. Note that a patient either has nystagmus or
does not. 'A few nystag moid jerks' is a terminological escape hatch for uncertain examiners. Optokinetic nystagm us This is a normal phenomenon best observed when sitting opposite someone in a railway carriage. His eyes will follow a portion of passing scenery until they can follow no longer, when they will quickly move back to fix on a new object
and follow that. The cycle is repeated regularly, so producing nystagmus and, as the quick phase is back towards the primary position, this is the reverse of all other forms. • The same effect can be obtained either by rotating in front of the patient's eyes a drum marked with alternate black and white stripes or by passing before the eyes a tape
marked with black and white squares, first in one direction, then in the other. • The physiological purpose is to stabilize images of stationary objects on the retina during head movement. In deeply situated parietal lobe lesions, the optokinetic response is absent or much reduced when the drum is rotated towards the side of the lesion. The particular
value of the test is that in patients with a homony mous hemianopia it is often difficult by other means to be certain whether the lesion lies in the optic tract or the temporal, parietal or occipital part of the radiations. Optokinetic nystagmus may, how ever, also be reduced in brainstem lesions, but there are usually signs of brainstem disease, and no
hemianopia. The test can be employed on patients whose degree of coopera tion is poor, or who are unable to understand commands, and being an involuntary movement depending on some degree of fixation, it is also of value in detecting the presence of vision in an infant It is sometimes stated that optokinetic nystagmus is absent 'to the side
opposite to the lesion'. This arises because nystagmus is described as being to the side of its fast component, which is the side opposite the lesion. It is, however, easier to remember which way the drum is being rotated. 82 Chapter 8 The third, fourth and sixth cranial nerves thought to be blind, or in an adult thought to b e feigning blindness. Its
absence cannot of course be a proof of blindness. IMon-nystagmus ocular oscillations Following ocular oscillations are rare but have certain characteris tics that help in localization: 1 Ocular flutter. It is a horizontal conjugate repetitive saccadic movement that occurs spontaneously in periodic bursts and increased on efforts to fixate the gaze. It may
be precipitated by a change in posture. It is caused by lesions of paramedian pon tine reticular formation or cerebellar neurons. It is associated with ocular dysmetria and may progress to opsoclonus. 2 Opsoclonus. It is a spontaneous, chaotic, multi-directional sac cadic eye movement disorder in which the abnormal move ments are almost always
conjugate. It is worsened by attempts to fixate. The common causes include encephalitis, toxic, metabolic and paraneoplastic disorders. Deep cerebellar nuclei involvement is hypothesized as the cause of this condition. 3 Ocular bobbing. It is a fast downwards movement of both the eyes followed by a slow drift back to the primary position. It is seen in
comatose patients with significant central pontine destruction. 4 Ocular dipping (inverse bobbing). It is a slow downwards move ment followed by a rapid return to primary position. It is seen in anoxic, metabolic and toxic encephalopathies. 83 9 The fifth cranial nerve: the trigeminal nerve The trigeminal nerve has a large sensory portion with
extensive central and peripheral ramifications, and a small motor element which runs in close association with the mandibular division. Anatomy The Vth nerve (Fig. 9.1) has two divisions. The sensory division, which is lower, has three divisions ophthalmic (VI), maxillary (V2) and mandibular (V3), which carry sensation from the face. The main
sensory and motor nuclei are located in midpons, although the spinal tract (pain and temperature) extends from pons to the upper cervical cord. The smaller motor part innervates the muscles of mastication. The proprioceptive component of the trigeminal nerve originates from mesencephalic nucleus, which is also located m pons. Mesencephalic
nucleus of Vth cranial nerve Motor nucleus of Vth cranial nerve Principal sensory nucleus of Vth cranial nerve Pons V1 (Opthalmic division) V2 (Maxillary division) V3 (Mandibular division) Medulla Trigeminal ganglion Spinal Cord Spinal nucleus of Vth cranial nerve (a) Fig. 9.1 (a) Schematic diagram showing origin of Vth cranial nerve and its main
divisions, (b) Distribution of cutaneous supply of V I , V2 and V3. 84 Chapter 9 The fifth cranial nerve: the trigeminal nerve Functions The important functions are: 1 To carry all forms of sensation from the face, the anterior part of the scalp, the eye and the anterior two-thirds of the tongue. 2 To give motor power to the muscles of mastication. 3 To
carry sensation from the teeth, gums, mucous membranes of the cheeks, nasal passages, sinuses and much of the palate and nasopharynx. Purposes of the tests 1 To determine which, if any, of the modalities of sensation are impaired. 2 To decide from this whether the lesion lies in one of the periph eral branches, in the Gasserian ganglion or sensory
root, or in the brainstem. 3 To determine whether motor weakness is unilateral or bilateral, and of lower or upper motor neuron origin. M ethods of examination Superficial skin sensation See Chapters 20 and 21 for general instructions regarding the tests of sensation. Here, pain and light touch are the main modalities exam ined. Six areas on each
side are tested near, but not at, the midline, • The forehead and upper part of the side of the nose (ophthalmic division) • The malar region and upper lip (maxillary division) • The chin and anterior part of the tongue (mandibular division). The area and extent of any abnormality should be carefully charted and compared with the known distribution of
the three divisions (Fig. 20.1). In addition, remember that: 1 On the scalp, the areas supplied by the Vth nerve and the sec ond cervical segment meet a little posterior to the vertex, but the exact point varies. 2 The mandibular division supply to the pinna is variable. The tragus is always included, and sometimes a strip along the upper and anterior
margin of the pinna. 3 The skin over the angle of the jaws is supplied not by the Vth nerve, but by the upper cervical segments. 4 All forms of sensation must pass to the Gasserian ganglion, sen sory root and brainstem. Fibres sewing touch enter the principal 85 Part 2 The cranial nerves sensory nucleus in the pons, cross and ascend to the thalamus.
Pain and temperature fibres pass downwards to the second cervical segment, gradually entering the descending nucleus. Ophthalmic division fibres pass to the lowest level. From this nucleus, all fibres cross the midline and ascend again in the quintothalamic tract. 5 'Numbness' of the entire exterior cheek sparing the mucosa, gums or tongue is
questionable. Abnorm alities of sensation 1 Total loss o f sensation over the whole distribution o f the nerve. This indicates a lesion of the ganglion or sensory root, or an exten sive lesion anterior to the ganglion, when the motor root is usu ally involved as well. Tumours eroding the base of tire skull, large neurofibromata of the Vth and Vlllth nerves,
epidermoids, chronic meningeal lesions such as sarcoid or syphilis and basal injuries are the commonest causes. There is an 'idiopathic' form of trigeminal sensory neuropathy, with pathological changes rather like a lupus or granulomatous syndrome in the Gasserian gan glion found in a very few. If this sensory abnormality is merely part of a total
loss down the whole of that side of the body, the lesion is in the neighbourhood of the opposite thalamus. 2 Total sensonj loss over one or more o f the main divisions. This can also be found in partial lesions of the ganglion (e.g. herpes zoster) or of the root (acoustic neuroma). More peripherally, the ophthalmic division is involved in the cavernous
sinus by carotid aneurysms, and in the orbital fissure by tumours. The maxillary division is rarely involved alone except as a result of local trauma, but it is also affected in tire cavernous sinus, while basal tumours involve the mandibular division, usually affecting the motor root as well. 3 Touch only is lost. This is a pontine lesion affecting the principal
sensory nucleus, and is usually due to vascular disease, pontine tumours or brainstem displacement by large tumours. 4 Pain and temperature are lost, but touch is preserved. Dissociated anaesthesia results from a lesion of the descending root and occurs in syringobulbia, foramen magnum tumours or anoma lies, and bulbar vascular accidents.
Traditionally, a thrombosis of the posterior inferior cerebellar artery causes ipsilateral loss of pain and temperature sensation on the face and contralateral loss on the rest of the body. In practice, however, this syndrome is usually caused by a vertebral artery deficiency, particularly if there is anomalous development of the two posterior inferior
cerebellar arteries in relation to each other. High cervical lesions can cause loss in the ophthalmic division but this is quite rare. 86 Chapter 9 The fifth cranial nerve: the trigeminal nerve 5 Hyperaesthesia over nil or port o f the distribution o f the nave. Apart from indicating an irritative rather than a destructive lesion, it gives little localizing value,
but is most common in vascular lesions and herpes and least common in syringomyelia. This is rather dif ferent from the trigger zones of trigeminal neuralgia. Here, a light touch on certain points - often the corner of the upper lip, the ala nasae, just in front of the jaw joint or just below the lower lip - will produce an intense spasm of pain in the
related division of the Vth nerve. In men, these areas may be left unshaven. 6 'Onion skin' type o f sensory loss. In nuclear lesions of Vth (sensory) nerve, sensory loss can be in concentric pattern, starting from the perioral region in a Balaclava helmet shape. This is because the fibres from the upper lip, mouth and tip of the nose (cen tral face) synapse
most rostrally in the nucleus of spinal tract of Vth nerve and those from the outer part of face synapse more caudally next to the sensory input from C2 and C3. This onion skin pattern is useful to understand the patterns of sensory loss in intrinsic brainstem and spinal cord lesions like syrinx and vertebrobasilar strokes. Multiple sclerosis is a common
cause of abnormal sensation or pain in a trigeminal distribution. A ^ The corneal reflex n f S Ji r _ This test must first be explained to the patient, who will otherwise undoubtedly flinch if some pointed object is suddenly thrust towards his eye. It must be the cornea, and not the lids, lashes or even the conjunctiva, that is stimulated. In order to widen
the palpebral fis sure as much as possible, the patient is told to look upwards as far as possible, and a piece of cotton wool teased to a point is touched just lateral to or below the pupil on either side. A little puff of breath on one cornea, while shielding the other, is a quick alternative method of value. Normally there is a bilateral blink whichever side is
tested, the facial nerve forming the efferent loop of the reflex arc. Reduction of the corneal reflex If there is no response on one side, this may be due to a breach of the sensory or motor sides of the reflex arc, i.e. the ophthalmic divi sion of the trigeminal nerve or the facial nerve. In Vth nerve lesions, there will be no response from either lid when the
abnormal side is stimulated, and a normal response from both lids when the normal side is stimulated. 87 Part 2 The cranial nerves In Vllth nerve lesions, there will be no response from the side of the facial paralysis no matter which side is stimulated, but, provid ing the Vth nerve is intact, there will be a blink on the normal side even when the
abnormal side is touched and both eyeballs can be seen to turn upwards. Loss of this reflex may be the first and only sign of a Vth nerve lesion and is of great value in early cerebellopontine angle tumours, and in aneurysms and tumours in relation to the cavernous sinus and orbital fissure. Difficulties and fallacies All the difficulties that surround
sensory testing are, of course, encountered here (see Chapter 21). Some people have very insensi tive corneae, especially if there is some degree of exophthalmos. If no response is obtained on either side, ask the patient if he can feel the touch and if it is equal on both sides. The cervical segmental supply to the angle of the jaw is very vari able and
may sometimes extend quite far up on to the cheek. This emphasizes the importance of testing near the midline. In the presence of a simple facial paralysis, the skin sometimes seems less sensitive and may give a false impression of a concomi tant trigeminal lesion. Sensory loss can be simulated, but the curious features of ana tomical distribution are
not reproduced. In particular, the area sup plied by the cervical segments is too unexpected, not unnaturally, to be included in an alleged Vth nerve loss. Remember that loss of facial sensation may be part of a total hemianaesthesia, which, if genuine, indicates a lesion high in the stem or in the region of the opposite thalamus. Th e m otor function of
the V th nerve The temporal muscles, masseters and pterygoids are tested. Note the symmetry of the temporal fossae, and the angles of the jaw, and then palpate the muscles while the patient clenches his jaws. Tire muscles can be compared as they stand out as hard lumps. Next, place the hand under the jaw and instruct the patient to open Iris
mouth. Motor abnormalities Wasting of temporal muscles and masseters due to a lower motor neuron or local muscular lesion produces hollowing of the temple and flattening of the angle of the jaw. This may be caused by nuclear 88 Chapter 9 The fifth cranial nerve: the trigeminal nerve lesions, as in motor neuron disease (bilateral), a peripheral
nerve lesion, as in compression of the motor root (unilateral), or muscular dystrophy. Pterygoid weakness causes the jaw to deviate towards the para lyzed side on opening as a result of the action of the normal muscle. If the jaw keeps falling open, but closes satisfactorily after rest, this indicates myasthenia, but not necessarily myasthenia gravis, for
this symptom is common in the myasthenia associated with a carcinoma. The jaw jerk This is a very important reflex, which sadly is often ignored. To obtain it, the patient is told to let his jaw sag open slightly, but not to push it open and not to open it wide. The examiner then places a forefinger or little finger below the lower lip and taps it in a down-
ward direction with the percussion hammer (Fig. 9.2). Sometimes the jaw jerk is not obtainable in the recumbent position, the reflex can then be elicited in the sitting position. There may be a slight palpable upward jerk immediately after the purely percussive effect is over, but in many normal people no response is obtained. Abnormalities An absent
jaw jerk is rarely helpful, but in lesions of the upper motor neuron above the level of the pons, great exaggeration, even amounting to jaw clonus, may be found. This is commonly the case in pseudobulbar palsy, motor neuron disease and quite often in multiple sclerosis. When a patient has pathological exaggeration of Fig. 9.2 The jaw jerk. A gentle
tap is essential, or the mechanical effect of the percussion will confuse the responses. 89 Part 2 The cranial nerves all tendon reflexes in arms and legs, an exaggerated jaw jerk shows that the lesion must be higher than the cervical spine. Be on guard, however, not to be misled by emotional hyperreflexia, which will be generalized, and without any
other signs of corticospinal tract abnormality. Trophic changes Erosion of the ala nasae and surrounding skin can follow severe sensory loss; this is occasionally seen after trigeminal ganglion injection for tic duoloureux, and in the idiopathic trigeminal neu ropathy. Corneal ulceration, infection and panophthalmitis may fol low profound ophthalmic
sensory loss but it would suggest that the Vllth nerve was involved as well so that the cornea was inadequately protected. 90 10 The seventh cranial nerve: the facial nerve Owing to the close anatomical relationship between the facial nerve and the nervus intermedius, they are considered together, though their functions differ greatly. Anatomy The
facial nerve nucleus is located in pons bilaterally. Each nucleus has two parts. The upper part (upper face) receives scant input from contralateral corticobulbar fibres, while the lower part (lower part of face) is dominantly innervated from only contralateral corticobulbar fibres. The nerve also receives inputs from nucleus ambiguous (swal lowing) and
nucleus tractus solitaries (salivation). The facial nerve has two parts: motor and sensory. These two parts emerge at the lower border of the pons. The motor part innervates muscles of the face, scalp and auricle, the buccinator and platysma, the stapedius, the stylohyoid and posterior belly of the digastric. The submaxillary and sublingual glands are
supplied via sympathetic fibres. The sensory part carries fibres for taste for the anterior two-thirds of the tongue and a sensation from the middle-ear region. A schematic dia gram of facial nerve and its branches is shown in Fig. 10.1. Functions For the purpose of neurological examination, the important func tions are the motor innervation of the
muscles of expression and facial movement, including platysma, and of the stapedius. The intermediate nerve carries secretory fibres to the lachrymal glands through the greater superficial petrosal nerve and to the salivary glands through the chorda tympani, which also carries the sensation of taste from the anterior two-thirds of the tongue.
Purposes of the tests To determine whether any weakness of the facial muscles detected is unilateral or bilateral, and of upper or lower motor neuron ori gin. If of peripheral origin, to determine by association with other abnormalities the site of the lesion along the course of the nerve. To detect impairment of taste. 91 Part 2 The cranial nerves Facial
nucleus Nucleus tract solitarius Superior salivatory and lacrimal nucleus Internal acoustic meatus Vestibular branches of Vllth cranial nerve Greater superficial petrosal nerve To lacrimal glands Meatal part Labyrinthine part To palate and nasal glands Horizontal part Pterygopalatine ganglion Mastoid part Geniculate ganglion Nerve to stapedius
Lingual nerve Chorda tympani To anterior two-thirds of tongue To sublingual gland To submandibular gland Posterior auricular nerve Cervicofacial division Temporal Stylohyoid Digastric facial division nerve nerve M ethods of examination Inspection Everything in this part of the examination is a matter of symmetry and asymmetry as seen while first
talking to the patient. Observe: 1 The face as a whole. 2 The wrinkles of the forehead and the nasolabial folds. 3 Blinking, and whether the eyeballs can be seen to turn up with each blink. (This of course happens normally, but is hidden by efficient closure of the lids.) 4 Movements of the mouth while talking, smiling etc. Note also the presence of
twitching, tremors or other iiivoluntary move ments in the facial muscles (see Chapter 19). Motor functions It is common practice to say to the patient 'show me your teeth'. This often results in a tedious explanation that they are false, or the more eager patients may actually take them out and show them. These undesirable experiences may be
avoided if the examiner bares his teeth himself and asks the patient to copy him. This also helps edentulous patients to overcome a curious difficulty in mak ing this particular movement. Note the symmetry of the movement 92 Fig. 10.1 Schematic diagram of facial nerve and its branches. Chapter 10 The seventh cranial nerve: the facial nerve and of
the nasolabial folds (zygomaticus, levator anguli oris, leva tor labii superioris). Next ask the patient to open his mouth and compare the nasolabial folds. Be on guard, however, for deviation of the jaw due to motor Vth nerve weakness. The upper facial muscles are tested by telling the patient to close his eyes (noting whether he can do so), then to
screw them up tightly and to resist attempts to open them. The orbicularis oculis is normally powerful enough to overcome this even in a puny child. He is asked to frown (corrugator supercilii), wrinkle the forehead (frontalis), and raisetbeeyeh Next ask the patient to bare his teeth and open his mouth at the same time; this enables the platysma to
stand out in the young and not too well covered. Other movements such as blowing out the cheeks(orbicularis oris) and pursing the lips tightly (orbicularis oris, buccinator) against resistance can be used as confirmation of strength or weak ness, and will distinguish proper weakness from mere asymmetry. Not all people can whistle (or even elevate
their eyebrows), but an abortive attempt often prompts a spontaneous smile. Examination of taste This shares with the tests of the sense of smell, the most unsatisfac tory part of the examination. There are only four primary tastes: sweet, salt, sour and bitter; all others are flavours, their appreciation depending upon an intact sense of smell. The tests
are carried out with sugar, salt, vinegar and quinine in that order. The patient must protrude the tongue to one side, keep it out throughout the test and not talk. The four possible tastes are written on a card. The tip of the tongue is held gently with a piece of gauze and the side of the tongue is moistened about 2 cm from the tip with a little of the test
substance. The patient should indicate the taste by pointing to the card. In between each test, he must swill out his mouth with water. Secretory functions The actual amount of tear production may be shown by hanging a strip of filter or litmus paper from the lower eyelids and measur ing the length of moistening on each side. More than 10 mm of
moistening is normal. This is Schirmer's test. The flow of saliva is compared by placing a highly spiced sub stance on the tongue and asking the patient to raise the tip so that the examiner may witness the submaxillary salivary flow. These tests related to secretory functions are not often required and the patient will usually be able to describe any
defects spontaneously. 93 Part 2 The cranial nerves Th e types of facial weakness These are dealt with in the order in which they are usually appreciated. Unilateral facial paralysis 1 A deviation of the mouth on smiling, which disappears on voluntary movement, constitutes the so-called emotional facial weakness. This occurs in deep-seated lesions of
the opposite thal amus, or its connections with the frontal lobe. O U nilateral em otional paralysis Neoplasms and vascular accidents affecting the thalamus and thalamofrontal connections. Bilateral em otional paralysis Parkinsonism, pseudobulbar palsy. Bilateral upper m o to r neuron paralysis Cerebrovascular accidents, early or late, neoplasms,
demyelinating lesions. Diffuse cerebrovascular disease producing a pseudobulbar palsy. Motor neuron disease. U nilateral lo w e r m o to r neuron paralysis 0 Bell's palsy ° Pontine neoplasms and vascular accidents ® Cerebellopontine angle tumours ° Petrous epidermoids (gradual onset), tumours, infections and operations in the middle ear 0
Fractures of the petrous bone 0 Trauma to the jaw and parotid regions ° Parotid tumours or sarcoid Bilateral lo w e r m o to r neuron paralysis (facial diplegia) ° Muscle - facioscapulohumeral dystrophy, myotonic dystrophy, congenital myopathies. 0 Neuromuscular junction - myasthenia gravis, botulism, organophosphate poisoning. 0 Peripheral
nerves - Guillain barre syndrome, Heerfordt's syn drome, sarcoidosis, bilateral Bell's palsy, Tick paralysis. 94 Chapter 10 The seventh cranial nerve: the facial nerve 2 Upper motor neuron facial palsy. Deviation of the mouth and deepening of the nasolabial fold but with normal strength of the orbicularis occurs in an upper motor neuron facial
weakness (Fig. 10.2). This is due to a lesion at some point between the opposite cortex and the facial nucleus in the pons. The upper facial muscles on each side are controlled by both cerebral corti ces, so that if one supranuclear pathway is damaged the other is still capable of performing its function. An associated hemianopia will mean a
hemisphere lesion; any hemiplegia will be on the same side. In an occasional patient with upper motor neu ron facial palsy, upper as well as lower half of the face may be involved on the same side as the hemiplegia. 3 Lower motor neuron facial palsy. Both upper and lower parts of the face may be paralyzed. The eye cannot be closed or can easily be
opened by the examiner, the eyeball is seen to turn upwards on attempted closure, the patient does not blink on that side and the normal wrinkling on that side of the forehead is absent (Fig. 10.3). This is a loioer motor neuron iveakness, occurring when the final common pathway between the nucleus and the muscle is interrupted, cutting off all
stimuli to both upper and lower facial muscles. The lesion may lie, on the same side, at any point along the course of the facial nerve, and it is necessary to con sider associated abnormalities to decide on its exact site. Thus: (a) If the Vllth nerve is also involved, the lesion is in the pons. Any hemiplegia will be on the opposite side. (b) If the Vth and
VUIth nerves are also involved, the lesion is in the cerebellopontine angle. (c) If taste, salivation and tear production are affected, the lesion lies between the brainstem and the departure of the chorda tympani in the middle ear. Distortion of sound due to stapedius palsy is also present. Fig. 10.2 Upper motor neuron facial paresis, (a) The weakness of
the lower part of the face is very much greater than the upper part, (b) In this case, associated movements of the right lower facial muscles were also affected. 95 Part 2 The cranial nerves Fig. 10.3 Lower motor neuron facial paralysis. Both upper and lower parts of the face are equally involved. Note the absence of wrinkling of right forehead and
visible sclera on screwing up eyes. (d) If taste and salivation are involved, but the secretion of tears is normal, the lesion is in the middle ear after the departure of the superficial petrosal branch, but before the departure of the chorda tympani. (e) If nothing other than motor weakness is present, the lesion may, in fact, be anywhere along the route of
the nerve, but is usually either at the nucleus or in the facial canal periph eral to the departure of the chorda tympani. (f) If only some of the facial muscles are paralyzed, and par ticularly if only the upper part, the lesion is usually very peripheral and often in the parotid gland, or in the muscles themselves. Perineural spread of certain skin
malignancies or infratemporal fossa tumours characteristically produce a gradually extending lower motor neuron type weakness. Bilateral facial paralysis This often causes difficulty in detection, because symmetry may be maintained. In bilateral emotional facial palsy, there is a mask-like face, complete lack of the normal play of expression and
dimin ished blinking, yet when it occurs it is normal, and there is transfor mation when the patient smiles. In bilateral upper motor neuron palsy, the masking is not so marked, blinking is little affected, but the mouth cannot be moved on command, yet often appears to move quite well during ordinary conversation. In bilateral lower motor neuron
palsy, there is a flattening of all normal folds, the corners of the mouth sag, all attempts at voluntary movement fail and the whites of the eyes are seen when the patient attempts to close them or to blink (Fig. 10.4). The patient talks as if protecting a very sore mouth. Long after a facial palsy, there may be aberrant reinnervation. If this is bilateral, the
whole face wrinkles when any expression is attempted, and the result may be quite inappropriate for the 96 Chapter 10 The seventh cranial nerve: the facial nerve Fig. 10.4 Bilateral lower motor neuron facial paralysis. The patient is trying to close her eyes. The eyeballs move upwards, but are uncovered; the lower part of the face is flattened and
expressionless. emotion being expressed and as unexpected to the onlooker as it is embarrassing to the patient (Fig. 10.5). Spontaneous twitching is also common during reinnervation and often interpreted by the patient as a sign of potential recurrence. Primary muscular disorders * ft tried Inmyasthenic states, facial expressions are markedly
diminished, the muscles are weak, particularly in the upper part, and there is usually associated ptosis and oculomotor paresis. These features are returned to normal by injection of neostigmine or edrophonium chloride (Tensilon) (see Figs. 17.42 and 17.43). Not many myopathies affect the facial or ocular muscles, but when present they are
characterized by a sagging of the whole facial musculature, downward-drawn lines at the corners of the eyes and mouth, generalized weakness of the muscles and inability to raise the corners of the mouth on smiling. The whole face is given an abnormally long, thin appearance, looking sad and lifeless. This Fig. 10.5 Old bilateral lower m otor neuron
facial paralysis w ith aberrant reinnervation: (a) at rest and (b) on smiling. 97 Part 2 The cranial nerves is the so-called myopathic facies. Myotonic dystrophy, ocular and oculopharyngeal dystrophy, facioscapulohumeral dystrophy and mitochondrial myopathy are the main causes, together with certain forms of spinal muscular atrophy. In myotonic
dystrophy, when the patient closes the eyes and is told to open them quickly, they remain closed for a measurable interval. Similarly, an expression such as a smile will persist after that emotion is, in fact, over. This is rare in myotonia congenita. Loss of taste Unilateral loss of taste, without Vllth or lingual nerve abnormality, is very rare, but could
occur in middle-ear lesions of the chorda tympani. More peripherally, lesions in the lingual nerve cause loss of common sensation over the anterior two-thirds of the tongue as well. Bilateral 'loss of taste' usually is a loss of flavours, due to anosmia, the primary tastes remaining intact when tested. Occa sionally, true bilateral loss occurs in
demyelinating or vascular lesions of the brainstem involving the nucleus of the solitary tract, but this is usually temporary. Difficulties and fallacies Natural facial asymmetry can usually be overcome if the problem is explained to the patient with the aid of a mirror. If a patient does not smile, asking him to whistle will often result in a successful smile,
if an unsuccessful whistle. In long-standing lower motor neuron lesions, contracture occurs and may suggest a contralateral palsy. This error is corrected when the patient makes a voluntary movement. In very severe upper motor neuron lesions, the upper part of the face may be so weak as to mimic a lower motor neuron lesion. Even here, however,
there is a far greater paralysis of the lower facial muscles and almost invariably a hemiplegia on the same side, whereas it would be on the opposite side if the paralysis were truly of lower motor neuron origin, because the neighbouring pyramidal fibres, if involved, would shortly be decussating in the medulla. 98 11 The eighth cranial nerve: the
auditory nerve* Unilateral total loss of hearing may easily be overlooked, both by the patient, who is either not aware of it or has grown to accept it (always using the telephone on the other ear), and by the doc tor who has not carried out any appropriate examination. Distur bances of the vestibular function are however so dramatic in their effects
rarely to be ignored by either, although the bedside tests of cochlear function are much simpler of the two. Anatomy The vestibulocochlear nerve arises at the ponto-medullary junction, bound by facial nerve posteriorly and inferior cerebellar peduncle anteriorly. The vestibular nerve is formed by the fibres which are concerned with equilibrium while
the auditory or cochlear nerve by the fibres associated with hearing. The vestibular nerve ends in the vestibular nuclei and carries sen sations from maculae, utricle and saccule (for linear acceleration) and ampullae of semicircular canals (for angular acceleration). Tire vestibular nuclei have extensive connections with cerebellum, ocu lomotor nuclei,
cerebral cortex and other areas of brain. It influ ences the eye and head movements, trunk muscles and limbs to maintain equilibrium. The auditory nerve receives input from the cochlea: the firstorder neurons are in the organ of Corti, the second-order neurons iii the cochlear nulcei, the third-order neurons in the inferior col liculus, the fourth-order
neurons in the medial geniculate body and the fibres terminate in Broadman's area 41 and 42 located in the transverse temporal gyrus of Heschl's. Fume! doms 1 The cochlear nerve. This carries impulses of sound from the hair cells of the organ of Corti, through the spiral ganglion in the cochlea, to the cochlear nuclei in the pons. Most fibres cross,
run in the lateral leminiscus to the medial geniculate body and are relayed to the superior temporal gyrus. But there is some Synonyms: the acoustic nerve, the vestibulocochlear nerve 99 Part 2 The cranial nerves uncrossed upwards transmission, so that deafness from a uni lateral cerebral cortical lesion is virtually precluded. 2 The vestibular nerve.
Impulses arise in the labyrinth by dis placement of endolymph affecting the hair cells in the ampul lae of the semicircular canals, and the otoliths in the saccule and utricle. Fibres run to the vestibular ganglia and on in the main trunk of the nerve to the vestibular nuclei in the medulla. These nuclei have connections with the cerebellum, the ocu-
lomotor nuclei via the medial longitudinal bundle, the nuclei of the upper cervical nerves, the spinal cord and the temporal lobes. Purposes of th e te s ts To determine whether any deafness is bilateral or unilateral, and whether due to disease of the middle ear, or of the cochlear nerve. To determine whether disturbance of vestibular function
originates in the labyrinth, the vestibular nerve or the brainstem. Examination of hearing A deaf patient turns his head to bring the unaffected side nearer. Hearing aids may be cunningly concealed, but if present, effi cient or inefficient, silent or singing, they almost invariably mean marked bilateral deafness, which considerably reduces the chances
of it being due to intracranial disease - reduces, but by no means excludes. Simple bedside screening test of hearing is carried out as follows: 1 The examiner rubs his index finger and the thumb in front of the ears bilaterally and then once at a time. The patient is asked about first whether he is able to hear the sound of friction of digits and second to
report any hearing loss on any side. 2 Place a finger in the patient's external meatus on one side and move it constantly in order to produce a masking noise. Posi tion the head so that lip-reading is impossible, and ask the patient to repeat the words said to him. In order to produce a standard volume of sound, breathe out, and at the end of expi ration
whisper a few numbers, for example, 26 or 68, to test high tones, and 42 or 100 to test low tones. If nothing is heard, the force of the whisper should be gradually increased and the two sides compared. Do not be surprised if the patient whis pers back. If deafness is present, the auroscope must be used to exclude the presence of wax and any disease
of the middle ear and drum. 100 Chapter 11 The eighth cranial nerve: the auditory nerve Th e type of deafness Conduction of sound through the air to the drum and bony ossicles is the normal method of hearing and is about twice as efficient as conduction through bone. Rirrne's test Strike a tuning fork gently, hold it near one external meatus, mask
the other, and ask the patient if he can hear it. Place it then on the mastoid, ask if he can still hear it, and tell him to say 'now' the moment the sound ceases. When he does so, at once place the blades, of the. fork, near the meabaa a%aba. M/armally, the nete. la still audible. In middle-ear deafness, this will not be so. ha nerve deaf ness, both air and
bone conduction are reduced, but air conduction remains better, and the note will still be heard. Further information must now be obtained from Weber's test. Weber's test The fork is placed on the centre of the forehead. Ask the patient if he can hear the sound all over the head, or in both ears or in one ear predominantly. In nerve deafness, the
sound appears to be heard in the normal ear, but in chronic middle-ear disease, it is conducted to the abnormal ear. Notes on terminology 'Conduction deafness' means middle-ear deafness. 'Perception deafness' means nerve deafness. 'Rhine's positive' is the normal response, which also occurs with nerve deafness. 'Rinne's negative' indicates reduced
air conduction and middle-ear deafness. Causes of deafness Causes of conduction and perception deafness are given in the box below. Deciding the level of the lesion is by no means easy. If deaf ness occurs as a result of a brainstem lesion, it is usually gross and profound neurological disability is present. If it is due to cerebello pontine angle tumour,
the Vth and Vllth nerves are also involved, but other lesions of the nerve trunk may be difficult to separate from cochlear lesions. Various additional tests of greater or less sophistication have been devised, none of which is specific, but a combination of which can help to distinguish between cochlear and neural deafness. 101 Part 2 The cranial nerves
O C o n d u ctio n deafness • All diseases of the external meatus, middle ear, and Eustachian tubes • Middle ear infection in suspected intracranial infection • Certain middle ear tumours (e.g. tumours of the glomus jugulare) Perception deafness ® At cochlear level - Meniere's disease, advanced otosclerosis, deaf ness due to drugs, internal auditory
artery occlusions, prolonged exposure to loud noise • In the nerve trunk - Old age, post-inflammatory lesions, toxic le sions, meningitis, cerebellopontine angle tumours, trauma • In the brainstem - Severe pontine vascular lesions, severe demyelinating lesions, occasionally tumours Tests of auditory function Puretone audiom etry This is a quantitative
measurement of hearing particularly impor tant in detecting early nerve deafness and still the most useful test. Musical notes of varying pitch are produced by an electric oscillator, and the intensity of the sound is increased until the patient can hear it and then decreased until the sound disappears. One ear is exam ined at a time while the other is
masked. The minimum intensity of each tone audible to the patient is recorded. A range of frequencies between 100 and 8000 Hz is used, with 0 dB representing 'normal'. Naturally, if hearing is impaired, the intensity of a particular note will have to be increased more than is usual for it to be heard, and the degree of loss of hearing is plotted against
the frequency of the note on special charts. High-tone loss is characteristic of nerve deaf ness and low-tone loss of middle-ear deafness (Fig. 11.1). Speech discrimination audiom etry Speech is employed, instead of pure tones, and discrimination of speech is more affected than the pure tone audiogram in retrocochlear lesions such as an VITIth nerve
tumour. Loudness recruitment Normally, as the intensity of a sound of a fixed frequency is increased, it is heard equally well in either ear. Under certain 102 Chapter 11 The eighth cranial nerve: the auditory nerve Fig. 11.1 Audiogram. A: Normal curve; B: nerve deafness loss of high tones; C: middle-ear deafness - loss of low tones; D: gross loss of
hearing. conditions of unilateral deafness, appreciation of a sound of low intensity may be diminished in the affected ear, but when the same sound reaches a high intensity it may be heard equally in both ears. The deafness of the affected ear is reduced at higher intensities of the same frequency. This is termed 'loudness recruitment' and occurs when
there is a lesion of the hair cells in the organ of Corti. It is a characteristic of cochlear disturbance, therefore, and will be found in Meniere's disease and otosclerosis rather than retrocochlear causes of perceptive deafness. Tone decay With time, the intensity of a pure-tone signal has to be raised appre ciably to remain audible to a patient with an
VUIth nerve tumour. Decay of some degree occurs in cochlear lesions but the need to raise the intensity by over 20 dB in less than 3 minutes suggests a nerve lesion. Bekesy audiom etry This is a method of showing a mixture of hearing threshold, loud ness recruitment and decay. Either interrupted or continuous tones of different frequency are
automatically presented to the patient who controls their intensity to remain just audible, and the intensity is graphically recorded. A continuous tracing falling abruptly away from a pulsed tracing is highly suggestive of a nerve lesion. Even more sensitive tests include measuring the response of the stapedius muscle to sounds, both determining
threshold response 103 Part 2 The cranial nerves and decay to long-continued stimulation (acoustic reflex measure ments), and acoustic impedance tests reflect any abnormality in the middle ear and ossicular chain. Direct measurement of the electrical output of the cochlea (electrocochleography) can distinguish between different types of sen-
sorineural impairment. But these tests require great experience both in performance and in interpretation. No one test gives a certain localization, but taking the cochlear tests together with vestibular tests can offer a high degree of probability. Tests of vestibular function All vestibular tests make use of the fact that disturbances of the balance of
vestibular function result in vertigo, nystagmus, past pointing and falling. The varieties of spontaneous nystagmus are discussed under Chapter 8. By stimulation of the vestibular system, usually the labyrinth, these features can normally be produced at will, and defects may be detected which are helpful in localizing the lesion. The rotational test The
patient is seated in a chair that can be rotated, with his head well-supported and fixed in a headrest. To test the horizontal canals, the head is flexed to 30° so that the eye/external meatus plane is horizontal. To test the vertical canals, the head is flexed to 120°. The chair is then rotated 10 times in 20 seconds. Normally, when rotation to (say) the right
has stopped, the endolymph continues to flow in that direction. This results in nystagmus with its slow phase to the right, this always being in the direction of the current. There is also past-pointing and falling to the right, and vertigo with apparent movement of objects to the left. Unfortunately, this test stimulates the labyrinths on both sides and for
neurological purposes the caloric tests are of greater value. The caloric tests Many people find difficulty in understanding these invalu able tests. If the patient lies supine with the head flexed 30°, the horizontal canals lie in the vertical plane, with the ampullae at the highest point. In this position, warming or cooling the endo lymph will produce
currents upwards or downwards, respectively. This movement stimulates the ampullae of the canal, producing 104 Chapter 11 The eighth cranial nerve: the auditory nerve nystagmus. For example, if, with the head in this position, warm water is run into the right external ear, a current flows upwards in the right horizontal canal towards the ampulla.
The position of the head makes no difference, however, to the fact that this is really the horizontal canal, and if such a displacement of endolymph took place when the canal was in its normal plane, the flow would be in an arc curving forwards towards the left. The nystagmus, therefore, has its slow phase towards the left, this phase always being in
the direction of the flow. Tradition, however, makes it more compli cated by decreeing that nystagmus is always named after its quick phase, which in this case is to the right. Therefore, irrigating the right ear with warm water produces nystagmus to the right; with cold water, to the left. Similarly, irrigating the left ear with warm water produces
nystagmus to the left; with cold water, to the right. During the test, about 250 ml of water is irrigated through the external auditory meatus over a period of about 40 seconds, first using water at 30°C and later at 44°C. The patient fixes his eyes on a given point immediately above his head and, after ceasing the irrigation, the time in seconds is
measured during which nystag mus on forwards gaze persists. The test is repeated on the other ear and the results are either charted as shown (Fig. 11.2) or more often nowadays recorded using electronystagmography (ENG). Normally, all four durations are of approximately the same value. If equivocal results are produced at 30°C, a much lower
tempera ture may be used. If the drum is perforated, air at a controlled tem perature can be blown in, instead of water irrigation. Caloric abnormalities If there is no response, or a much diminished response, to both warm and cold water on one side, this is termed canal paresis. It is caused by lesions of one labyrinth (Meniere's disease), or vestibular
nerve (e.g. acoustic nerve tumour, or vestibular neuronitis, the former showing deafness, the latter no deafness) or lesions of the vestibular nuclei. If the response is always reduced for irrigations producing nys tagmus in the same direction (e.g. cold water in the right ear and warm water in the left), there is said to be directional preponderance,
which in this case would be to the right. This is a sign of imbalance between the two sides of the vestibular system, normally held in a state of equilibrium. This balance can be disturbed by lesions of the peripheral or central vestibular apparatus or of the cerebellum and corticofugal fibres deep in the temporal lobe. Combinations of these two types of
abnormality may at times be seen in lesions of the vestibular nerve or labyrinth. This emphasizes, 105 Part 2 The cranial nerves Direction of nystagmus (quick phase) Ear L — S\— 30°C r\ r\ To right rs V - _A R To left 1 0 L ru\ U A 44°C 3 (minute) \J i R Conclusion f A To left U To right (a) L , 30°C R ^ |V ^ A 44°C r\ r\ — C\— n ' .Tr\ To right \J To left •
l 1 0 L r\ —#------ —e— ~ —o— — V —#— — 3 (minute) To left To right R Conclusion (b) To right 30°C To left 2 3 (minute) To left To right Conclusion (c) therefore, that the caloric tests must be interpreted in conjunction with audiometry and, of course, the clinical picture. Electronystagmography As the eye is surrounded by an electric field, probably
originat ing from the pigment epithelium of the retina, changes in potential difference produced by movement of the eyeball can be recorded graphically between skin electrodes on opposite sides of the eye. 106 Fig. 11.2 Caloric responses. The broken line marks the time in minutes for which nystagmus on forwards gaze persists after the end of
irrigation. The temperature of the water is indicated on the left, the direction of the nystagmus on the right. (a) Normal response. (b) Right canal paresis in the case of acoustic neuroma. (c) Directional preponderance to left in the case of left posterior temporal lobe tumour. Chapter 11 The eighth cranial nerve: the auditory nerve 0 • At labyrinthine
level (a) Meniere's disease (b) Motion sickness (c) Drug toxicity (d) Probably migraine 0 In the vestibular nerve. As for perception deafness, but add also 'vestibular neuronitis' ° In the brainstem (a) Vascular deficiency, especially vertebrobasilar artery disease (b) Cerebellar and IVth ventricular tumours (c) Acute demyelinating disease, migraine • In
the temporal lobe. As an epileptic manifestation, especially in children, or as an ischaemic lesion in the elderly Electrodes lateral to each eye and just above the bridge of the nose will record horizontal movement, above and below will detect less common vertical nystagmus. This technique enables the nystagmus to be measured at leisure, is not so
dependent upon a subjective impression and though neither simple nor without drawbacks, is the most frequently used means of detecting and recording eye movement. Optokinetic, caloric, positional and rotational tests can all use ENG, in addition to analysis of gaze-evoked spontaneous nystagmus. Tests can be carried out in darkness, with the eyes
shut, without fixation, and this constitutes much of the importance of the examination because fixation diminishes spontaneous nystagmus of peripheral origin, but increases it when the lesion is supranu clear, so the method has localization value. These differing effects oi closure ?.Ti-d 'iTfiteitSb on spontaneous nystagmus allow rec ognition of
characteristic patterns emerging from lesions periph eral to the vestibular nuclei, at nucleus level, or above. Positional vertigo and nystagmus Seat the patient on a couch, grasp the head, turn it 30-40° to one side, and then lower the patient backwards until he is supine with the head over the end of the couch and 30° below the horizontal. A patient
with positional vertigo will, after a short latent period, develop both nystagmus and vertigo, the fast phase of the eye move ment being towards the lower ear. Adaptation rapidly occurs, and these features cannot usually be elicited again within 10-15 min utes. This is the benign paroxysmal type, thought to be due to a 107 Part 2 The cranial nerves
utricle lesion, and common after infective or vascular lesions, and following head injury. Such patients experience transient vertigo on movement of the head and neck. If nystagmus appears at once, adaptation does not occur, nys tagmus reappears immediately on reassuming the position, and its direction is altered by varying the head posture, then
this is the cen tral type and may indicate deeply situated posterior fossa lesions - though not what type. Such patients may also have vertigo on movement but it is likely to be more prolonged and prostrating. Unfortunately, in practice, this distinction between 'benign' and 'central' does not always hold true. The clinical and even ENG findings may
suggest a peripheral lesion, only for the computer ized tomography or magnetic resonance imaging scan to reveal a cerebellar infarct or tumour. « 108 12 The ninth and tenth cranial nerves: the glossopharyngeal and vagus nerves Clinically, and even anatomically, it is so difficult to separate the actions of these two nerves that it is customary to
consider them together. Anatomy The glossopharyngeal nerve (IXth) carries motor, sensory and parasympathetic fibres. It exits from the posterior lateral sulcus of the medulla in close relation with the vagus nerve and the spi nal accessory nerve. The motor fibres originate from the nucleus ambiguus and supply the stylopharyngeus muscle and (with
the vagus nerve) muscles of the pharynx. It also carries taste sensa tions from the posterior third of the tongue and the pharynx, and sensations from the posterior third of the tongue, tonsils, palatal arch, soft palate, nasopharynx and tragus of the ear. The parasym pathetic fibres carried in the glossopharyngeal nerve originate in the inferior
salivatory nucleus, located in the medulla and sup plies the parotid gland. The vagus nerve (Xth nerve) carries motor, sensory and parasym pathetic nerve fibres. The six to eight rootlets of the vagus nerve emerge from the posterior sulcus of the lateral in close association with the glossopharyngeal nerve. The motor fibres arise from the dorsal motor
nucleus of the vagus and the nucleus ambiguus. The dorsal motor nucleus of the vagus gives rise to preganglionic para sympathetic fibres that innervate the pharynx, respiratory and gas trointestinal system. Through vagus, the nucleus ambiguus sup plies all of the striated musculature of the soft palate, pharynx and larynx except the tensor veli
palatini (cranial nerve V) and stylo pharyngeus (cranial nerve IX) muscles. Functions From the very widespread functions of the vagus and glossopharyn geal nerves, the following are of most importance in neurological examination: 109 Part 2 The cranial nerves 1 To carry common sensation from the pharynx, tonsils, soft palate and posterior one-
third of the tongue. 2 To carry the sense of taste from the posterior one-third of the tongue (probably almost purely by the IXth nerve). 3 To give motor supply to the palatal and pharyngeal muscles. 4 To give motor supply to the vocal cords (purely the vagus). Purposes of the tests 1 To determine the integrity of the reflex arc for the gag reflex. 2 If
abnormal, to differentiate between a breach on the sensory side by testing sensation in the pharynx and palate, and on the motor side by testing the elevation of the palate and contraction of the pharynx. 3 To examine the movements of the vocal cords. f J M ethods of examination Preliminary observations Notice the pitch and quality of the patient's
voice, and of his cough, and whether there is any difficulty in swallowing his saliva. Ask if there has been any nasal regurgitation of fluids. A high-pitched, hoarse voice may mean vocal cord paralysis; a nasal tone that increases if the head is bent forwards means palatal paralysis, when lying back this can become almost normal. If the patient chokes
on his saliva while talking, there may be both pala tal and pharyngeal weakness, and if any of these features increase towards the end of each sentence, this may be due to myasthenia gravis. Motor functions Ask the patient to open his mouth wide. A few moments' wait, allowing the tongue to rest in the floor of the mouth, will usually make it possible
to see the palate without the use of the unpopular tongue depressor. The patient is then asked to say 'A h' while breathing out, followed by 'U gh' while breathing in. In each case, the palate should move symmetrically upwards and backwards, the uvula remaining in the midline, and the two sides of the pharynx should contract symmetrically. The
patient should then be asked to phonate several times in suc cession and the palate is watched to see if any defect worsens with repeated use. 110 Chapter 12 The ninth and tenth cranial nerves Sensory functions Common sensation It is normally only the sense of touch that is tested, relying on its ability to stimulate a reflex arc. A throat swab, with
the cotton wool safely attached, is passed to one side of the back of the throat, while the tongue is gently and slowly depressed. Touching any part of the palate, tonsil or the back of the tongue will normally result in contraction of the pharynx, elevation of the palate and retraction of the tongue. This is called the gag reflex, and varies in sensitivity
from individual to individual. Taste Testing taste on the posterior part of the tongue is so difficult by normal means that it is hardly worth spending much time over it. Using a galvanic current of 2-4 mA, and touching the tongue in this area with the anode on either side, will produce a metallic taste which the patient should be able to detect if not
define, and allows the two sides to be compared, but the simple devices described in p. 93 can be more easily applied to this area than any of the conven tional methods. Abnormalities On inspection Tire uvula lies to one side of the midline due to: 1 Simple asymmetry of the palate. In this case, movement on phonation is normal. 2 Swelling in the
tonsillar region. This will be visible. Take this opportunity to detect other nasopharyngeal swellings, so eas ily overlooked, and yet so vital in cases of lower cranial nerve palsies. 3 Unilateral muscle paralysis. A constant rhythmic vertical oscillation of the palate, sometimes also involving the pharynx, is called palatal nystagmus or palatal myoclonus,
due to a lesion in the central tegmental tract. A more extensive form has been referred to as palato-phanyngo-laryngooculodiapliragmatic myoclonus, a name that if alphabetically uneco nomical is, at least, self-explanatory. Palatal fasciculation can be seen in motor neuron disease. On phonation The palate moves up and over to one side when there is
paralysis of the opposite side, owing to the pulling movement of the unopposed 111 Part 2 The cranial nerves normal muscle. In pharyngeal paralysis, the muscles will also appear to move towards the normal side, so resembling a flat sheet being drawn across that it is called the 'curtain movement'. These features are caused by a lower motor neuron
lesion of the vagus. If there is no movement of the palate and pharynx, there should also be dysphagia, nasal regurgitation and nasal speech, and this usually indicates either a bilateral medullary nuclear lesion, or a bilateral upper motor neuron lesion as seen in pseudobulbar palsy. These two groups of symptoms may be combined in motor neuron
disease. Repeated phonation can demonstrate the fatiguability that occurs in myasthenia gravis. This is a time to note the high arched palate of patients with Marfan's syndrome, which may be associated with intracranial vascular disorders, and a cleft palate, which may be associated with other congenital malformations. On testing sensation
Unilateral absence of the gag reflex may be due to loss of sensation, or motor power or both. Phonation will have shown if one side is paralyzed. If due to loss of sensation alone, stimulation of the nor mal side will produce a normal symmetrical reflex. This rare event would be due to a glossopharyngeal lesion. If the defect, however, is due to combined
motor and sensory paralysis, stimulation of the normal side will cause the palate to be pulled towards that side. This more common finding indicates a combined lesion of glosso pharyngeal and vagus nerves. No reaction on either side, but normal movement on phonation, is practically never due to organic disease, though theoretically it might possibly
occur in syringomyelia or tabes dorsalis. A combination of bilateral anaesthesia and bilateral motor paral ysis indicates a severe medullary lesion and is usually associated with other lower cranial nerve palsies. Difficulties and fallacies Some people have intensely sensitive fauces and pharynx and are unable to tolerate any touch. Others appear to
have complete insensitivity. In these, watch spontaneous movements on phona tion and inspiration, and ask them to attempt to swallow with the mouth open. A great deal of information can be obtained in this way without touching at all. 112 Chapter 12 The ninth and tenth cranial nerves 0 Muscle • Myotonic dystrophy • Polymyositis and other
inflammatory myopathies • Oculopharyngeal muscular dystrophy/hyperthyroidism Neuromuscular junction • Myasthenia gravis 0 Botulism • Organophosphate poisoning Peripheral nerve ° 8 • 8 Guillain-Barre syndrome Diphtheria Tick paralysis Porphyria Anterior horn cell disease 8 Fazio-Londe disease • Progressive bulbar palsy 8 Poliomyelitis
Central nervous system 8 8 8 8 • Arnold-Chiari malformation Pseudobulbar palsy Syringobulbia Basal meningitis Brainstem tumours tr U nilateral 8 • 8 8 • Mediastinal tumours Primary or secondary aortic aneurysms Enlarged cervical glands Malignant disease of the thyroid Trauma including surgical operations on the neck Bilateral • Enlarged

cervical glands including the thyroid • Trauma 113 Part 2 The cranial nerves In children, these tests are best left to a later stage or one may never regain the 'rapport' required for the rest of the examination. Hysterical insensitivity of the pharynx is not uncommon and may be associated with hysterical hoarseness, but movements on sudden respiratory intake are normal, even if phonation is not attempted. If local anaesthetic has been used to enable good visualization of the vocal cords, it is important to know when this was carried out. Patients vary greatly in the length of time that local anaesthesia lasts, and apparent sensory loss may be misleading. 114 13 The eleventh cranial nerve: the accessory nerve It is the spinal root of the Xlth nerve that is examined here, the cranial root joining the vagus after leaving the brainstem and being considered part of that nerve. Anatomy The spinal root originates from the spinal nucleus located in the spinal grey column (accessory nucleus) and descends up to the C5 spinal segment. The cranial part arises from the caudal part of nucleus ambiguus. The fibres emerge between the upper cervical and dorsal roots and form a trunk which leaves the cord between dentate liga ment and dorsal spinal roots to enter the skull via the foramen magnum. The cranial and spinal roots join and exit through the jugular foramen. The cranial part joins the Xth (vagus) nerve and supplies larynx and pharynx. The spinal part supplies sternocleidomastoid and trapezius muscles. Functions To supply motor power to the upper part of the trapezii and to the sternomastoids, and so to influence the posture and movements of the head and shoulder girdles. Purpose of the tests To detect wasting and weakness, unilateral or bilateral, of these muscles; to decide if the lesion is nuclear, in the nerve trunk or its branches, or due to local muscular disease. Methods of examination When the patient is first seen, severe trapezius weakness may be suspected if the head falls forwards, and sternomastoid weakness if it falls backwards. 115 Part 2 The cranial nerves Stermomastoids Place one hand against the right side of the patient's face and ask him to turn (not bend) his head against it. The left sternomastoid will stand out clearly (Fig. 13.1(a)). Repeat this in the opposite direction and compare the two sides for bulk and strength. Then rest a hand on his forehead and ask him to bend his head forwards. Both sternomastoids will stand out together and are easily com pared (Fig. 13.1(b)). Now ask the patient to sit up. Normally, the head leaves the pillow first and the movement is easy. Trapezius Go behind the patient and compare the line and curve of the trapezii and the position of the scapulae, making certain that he is sitting symmetrically upright. Then ask him to raise his shoul ders towards his ears. (Asking patients to 'shrug their shoulders' often produces a most unnatural convulsive movement.) Now try to depress the shoulders forcibly. Even the most feeble patient is normally able to resist the manoeuvre. Fig. 13.1 The sternomastoids. (a) Turning the head against resistance brings the opposite sternomastoid into action, (b) Raising the head forwards against resistance brings both muse e into action, (c) Tyr. i result of bilateral sternomastoid weakness. On sitt - g up, the patient's head lags behind and overaction ot platysma draws the mouth downwar_ 116 Chapter 13 The eleventh cranial nerve: the accessory nerve Abnorm alities When the sternomastoids are wasted or absent, the neck appears elongated, thin, scraggy and poultry-like, the thyroid cartilage and gland standing out abnormally. 1 In bilateral sternomastoid weakness, when the patient sits up, the head seems to be left behind on the pillow and then is raised with difficulty (Fig. 13.1(c)). The platysma may stand out even to the extent of drawing the mouth downwards, the resultant grimace making the whole movement look most unpleasant. 2 In unilateral sternomastoid weakness, the patient will fail to turn his head against resistance to the opposite side. The muscle will not stand out clearly either then or when the head is flexed for wards against resistance. Do not overdo this latter test, for it may put the anterior neck muscles into painful cramp. 3 Trapezius -weakness results in the shoulder dropping on one side and the scapula being displaced downwards and laterally, giving a steeper gradient to the contour of the neck. Shrugging of that shoulder may be weaker, though not absent, because part of the trapezius is supplied by cervical nerves. 4 Fasciculation in these muscles means a nuclear lesion, as in motor neuron disease, but coarse twitching of the trapezius is seen in irritative and compressive lesions of the nerve trunk near its origin. 5 Upper motor neuron lesions of the accessory nerve rarely pro duce much demonstrable abnormality unless the paralysis is very profound, when the face and arm at least are likely to be grossly affected. O Sternomastoid and trapezius paralysis Bilateral • Nuclear Xlth nerve: Motor neuron disease, spinal muscular atro phy, poliomyelitis • Nerve: Polyneuropathy or mononeuropathy • Muscle: Polymyositis, dermatomyositis, myasthenia gravis, myo tonic dystrophy, oculopharyngeal muscular dystrophy Unilateral • Nucleus: Poliomyelitis, syringobulbia • Nerve: Tumours at jugular foramen level, bony abnormalities of base of skull, trauma to the neck or base of skull, compression to the nerve with enlarged cervical lymph nodes 117 Part 2 The cranial nerves Difficulties and fallacies These tests are very simple. The main problems arise from varia tions in muscular development. Muscles with very little bulk may, nevertheless, be very strong, and the symmetry of their size and strength is o f most importance.

118 14 The twelfth cranial nerve: the hypoglossal nerve The Xllth nerve contains only motor fibres and all tests applied to it are truly objective. Anatomy The fibres arise from the hypoglossal nucleus in the spinal anterior grey column in medulla beneath the hypoglossal triangle in the floor of the fourth ventricle. The fibres of hypoglossal nerve are arranged linearly as a series of 10-12 rootlets in the anterolateral sulcus between the inferior olive and the pyramid. The nerve passes through the hypoglossal canal and gives the following branches. The branches are menin geal, descending branch (upper root of ansa cervicalis) to supe rior belly of omohyoid, nerves to thyrohyoid and geniohyoid. It finally supplies all the intrinsic (longitudinal, transverse and vertical muscles) and extrinsic muscles (hyoglossus, styloglos sus, genioglossus and geniohyoid muscles) of the tongue except palatoglossus. Functions To control all movements of the tongue, and certain movements of the hyoid bone and larynx during and after deglutition. Purposes of the tests To inspect the surface of the tongue; to detect wasting, weakness and involuntary movement; to examine voluntary muscle control; to detect myotonia. M ethod of examination If the patient opens his mouth, the surface, size, shape and position of the tongue can be inspected before his attention is drawn to it. Then ask that it should be protruded in the midline. When the jovial comments that accompany this traditionally impolite gesture have been accepted with good humour and the movement achieved, repeat these observations and note any difficulty in performing the 119 Part 2 The cranial nerves movement, any deviation from the midline and any involuntary movement. Myotonia of the tongue can be tested by asking the patient to protrude the tongue and holding it by a gauge piece. Subse quently, a sharp tap by the edge of the tongue depressor can be given over the bulk of the tongue. This brings out myotonia in the tongue. Abnormalities On inspection and protrusion Many deviations from normal are irrelevant to neurological diagno sis. The tcngue may be enlarged in Down's syndrome and infantile hypothyroidism. It will show corrosion after ingestion of caustic fluids. In vitamin deficiencies, the papillae atrophy and the tongue appears shiny, smooth and translucent with shallow, irregular, red dened ulcers flanked by desquamating tissue. It looks small though there is no muscle loss. This is particularly important in pernicious anaemia, vitamin B]? deficiency and multiple deficiencies of the B group of vitamins. If there is unilateral muscle wasting, the longitudinal folds on that side are greatly exaggerated and the tip and median raphe curve round towards the affected side on protrusion, owing to the unop posed pushing action of the normal genioglossus (Fig. 14.1(a)). There may be fasciculation, but speech is little affected. This is the result of an ipsilateral lower motor neuron lesion. When such wasting is bilateral, the tip and median raphe remain central, the tongue is greatly reduced in size, there may be diffi culty in protrusion and speech is grossly disturbed. Fasciculation is marked if the lesion is nuclear (Fig. 14.1(b)) as in motor neuron disease. If a normal-looking, symmetrical tongue moves constantly to one side, this can be due to a contralateral upper motor neuron lesion, but would then usually be part of a profound hemiplegia. As an isolated feature, it is more commonly due to faulty performance of the test and can be corrected. A small, tight, compact-looking tongue, lying in the floor of the mouth like a nut in an open shell, its surface little altered, but almost incapable of protrusion and with gross disturbance of speech, is the result of bilateral upper neuron lesions. The jaw jerk will be exag gerated. The features of nuclear and upper motor neuron lesions coexist in varying proportions in motor neuron disease. The tongue may become immobile. 120 Chapter 14 The twelfth cranial nerve: the hypoglossal nerve (b) (c) Fig. 14.1 (a) Right hypoglossal nerve palsy. Note the reduction in the size of affected side, excessive ridging and wrinkling, and the curve of tip and median raphe towards the side of the lesion, (b) Bilateral wasting and spasticity in motor neuron disease. Note the restricted protrusion, surface indentations and reduction in size without deviation, (c) Myotonia. Note the characteristic prolonged dimpling after percussion of tongue. O L o w e r neuron m o to r lesions Unilateral • • • • • • • Syringomyelia Poliomyelitis Tumours at or near the jugular foramen Angiomas of the brainstem Trauma Tumours or glandular enlargement high in the neck Early motor neuron disease Bilateral • Progressive bulbar palsy ® Syringomyelia ® Foramen magnum anomalies (occasionally) U p p e r m o to r neuron lesions Unilateral • Profound hemiplegia (due to vascular accidents or deep-seated neoplasms) Bilateral • Bilateral vascular accidents producing a pseudobulbar palsy, amyotrophic 121 Part 2 The cranial nerves A coarse, trombone-like tremor on protrusion may be seen in neurosyphilis and some cases of parkinsonism. An up-and-down flapping movement with alternate protrusion and retraction is common in chorea. Dystonic movements of the tongue follow high and prolonged dosage of chlorpromazine derivatives. Other invol untary movements are described in Chapter 19. On percussion Normally, the dent formed will disappear immediately. In myoto nia, it persists and may enlarge for a few seconds in a linear man ner. Tapping the tongue on one side will produce a dent on both sides (Fig. 14.1(c)). See Video clip on tongue myotonia in the free companion CD-ROM of this book. Difficulties and fallacies Apraxia of the tongue may prevent it from being protruded, but it will probably move normally during automatic speech, licking of lips etc., and there is likely to be other evidence of apraxia. The patient may have failed to understand the directions due to recep tive dysphasia. A very short frenulum holds back the tip, which curves downwards so that the centre of the tongue appears to be somersaulting forwards on protrusion. If there is a facial paralysis, the tongue may appear to deviate to one side owing to the asymmetry of the mouth. This is overcome by drawing back the corner of the mouth into its normal position and comparing the position of the median raphe with the central incisors. Many tongues have corrugated edges and deep clefts. These are not necessarily significant. Little muscular movements frequently occur on the surface, which must not be confused with fasciculation, for they are inconstant and not associated with wasting. Most people, of both sexes, are unable to keep their tongues still for long, so that great care should be taken before deciding that involuntary movements are present.

122 Part 3 The Motor System 15 Muscle bulk Normal movement depends upon the correct functioning of nor mal muscles, their motor neurons and the bones and joints that they have to move, any of which may develop primary disease pro cesses. To help in distinguishing the neurological lesions, careful preliminary inspection is essential. Preliminary general inspection The aim is to compare the size and shape of the limbs and to detect deformities. Examine the patient lying, sitting and later standing, placing the limbs in symmetrical positions. Any apparent differences in size must be checked by careful measurement. If definite asymmetry is confirmed, try and decide if this is due to: 1 Congenital maldevelopment. This will include absence of muscles or parts of a limb, webbing of fingers and toes, polydactyly etc. The possibilities are too numerous to mention in detail. 2 Long-standing neurological lesions. Lesions of the lower motor neu ron in infancy, such as b radii a1plexus injury or poliomyelitis, cause marked retardation in limb growth with wasting and absence of reflexes. Following infantile cerebral lesions, such as birth trauma, acute infantile hemiplegias, and sometimes with vascular anoma lies, there is again retardation in growth, but of lesser degree, with little wasting, a hemiplegic posture, and exaggerated reflexes. 'Old polio' used to be much too casually diagnosed in sudi patients. Muscle • Polymyositis, dermatomyositis and other collagen vascular diseases Nerve 9 Acute brachial neuritis ° Guillain-Barre syndrome • Diabetic radiculoplexoneuropathy (amyotrophy) Others - referred pain, arthritis Psychiatric - depression and dissociative disorders' Parkinson's disease 125 Part 3 The motor system 3 Acquired lesions o f local structures. These include deformities of bones, joints and muscles, surgical operations, wounds, osteo arthritis or rheumatoid arthritis and Dupuytren's contracture. Examination of joint movement Each joint should be put gently through its full range of movement. In Charcot's joints, once mainly seen in tabes dorsalis, but now more commonly in other forms of sensory neuropathy, particularly syringomyelia, there is swelling, abnormal motility, often alarm ing crepitus, associated effusion, but usually practically no pain on movement. In the frozen shoulder, the arm cannot be abducted or rotated, the joint is tender and all movement is very painful. Local inspection of muscle Next inspect the muscles of the shoulder girdles, upper arms, fore arms, hands, hip girdles, thighs and calves, the aim being to detect wasting, hypertrophy and fasciculation. Measure the circumference of the limbs at clearly stated places, e.g. 10 cm above or below the olecranon; 18 cm above the patella; 10 cm below the tibial tuberosity. If wasting is observed, inspect the groups affected individually to decide which of the component muscles is involved. Compare O P hysiological • Exercise Pathological • M uscular dystrophy (a) D uchenne m uscular dystrophy (b) Becker m uscular dystrophy (c) Lim b-girdle m uscular dystrophy • M yotonia congenita • Spinal m uscular atrophy • Storage disorders 0 H ypothyroidism • C ysticercosis • Localized m uscle m ass: m uscle rupture, m uscle haem orrhage, m uscle tumour, m yositis ossificans, pyogenic abscess 126 Chapter 15 Muscle bulk the muscles with their fellows on the other side, with experience of the normal for that build of individual, and observe if related structures are uncovered, e.g. the prominence of the tibia when the anterior tibial muscle is wasted, or of the spine of the scapula with wasting of the supra- and infraspinati. In the hands, note if rheu matoid arthritis is obvious, because small muscle wasting is often associated. Hypertrophy may be physiological, when the muscles are merely big and powerful, their texture being normal. • In the pathological pseudohypertrophy of Duchenne and Becker muscular dystrophy, which most frequently involves the calves, the muscle is abnormally globular, has a tense rubbery feeling due to excessive deposition of fat and is weaker rather than stronger than normal. Rather more diffuse hypertrophy may develop in myotonia congenita, including thighs and shoul der girdle. Myotonia-induced sustained muscle activation is the presumed cause, and the muscles are not necessarily weak. Hypertrophy of the calf muscles also occurs in a proportion of Kugelberg-Welander-type spinal muscular atrophy (SMA). Dropping of the shoulders due to trapezius wasting may give a false impression of enlargement of the shoulder-girdle muscles. Fasciculation is discussed in Chapter 19. T y p es o f m u scle w a stin g Generalized wasting Though commonly the result of systemic diseases such as malig nancy, or thyrotoxicosis, wasting is also seen in the very advanced stages of many crippling neurological diseases, but particularly myopathies and motor neuron disease. In the latter, fasciculation together with spasticity, increased reflexes and extensor plantar responses will be distinctive of amyotrophic lateral sclerosis. Addi tional changes in the tongue and bulbar region also serve to distin guish it from progressive muscle disease. The wasted limb muscles in SMA may be felt on palpation but superficially obscured by subcutaneous fat. Conversely, loss of sub cutaneous tissue can be mistaken for muscle wasting. Proximal muscle wasting Wasting mostly follows the pattern of progressively weakening muscles. Together they are more likely proximal than distal in most forms of muscle disease, but unchecked become widespread. 127 I Part 3 The motor system © Generalized muscle wasting Proximal muscle wasting Distal muscle wasting hands (C7-T1) • • • • • • Muscular dystrophies - FSHD, limb-girdle syndromes, dystrophinopathies • Motor neuron disease • Neuromuscular junction disorder - myasthenia gravis • Inflammatory - neuralgic amyotrophy, old polio, inflammatory myopathies • Compressive cervical radiculopathy • Axillary neuropathy, brachial plexopathy • Anterior horn cell disease - polio, motor neuron disease, syringomyelia, cervical cord tumours • Anterior root - cervical spondylosis, cervical tumours • Brachial plexus lesions • Lesions of radial, median and ulnar nerves Malignancy Thyrotoxicosis Tuberculosis Advanced myopathies Motor neuron disease FSHD: facioscapulohumeral dystrophy Myopathies tend to be symmetrical compared to SMA, and selec tive patterns of individual muscle involvement suggest a dystro phy. This pattern, together with the age of onset, any family history and the subsequent clinical course will give the likely diagnosis. Facial, ocular and neck muscles are affected early in a select group of disorders (See Chapters 8 and 11). Muscular dystrophies It must be realized that the end stage of muscular dystrophies tends to look similar. It is the early signs that differentiate the types and so are invaluable in prognosis. Facioscapulohumeral dystrophy (FSHD) As already discussed, facial involvement is prominent and early. There is selective, usually symmetrical, shoulder-girdle weakness tending to spare deltoids. Hypertrophy is rare; asymmetry commonly denotes SMA. In the legs, tire combination of early anterior tibial and hip muscle involvement can be confusing. Indeed, a scapuloperoneal dystrophy or SMA combining foot drop and proximal shoulder-andarm wasting may be difficult to distinguish from FSHD (Fig. 15.1). 128 Chapter 15 Muscle bulk Fig. 15.1 Facioscapulo humeral dystrophy with (a) winging of scapula on attempting to push against the wall and (b) Tolyhill' sign due to the selective muscle fibre atrophy in deltoid and biceps associated with winging of scapula. Limb-girdle syndromes Both shoulder and hip girdles may be involved. There may be asymmetry, but the face is spared. Several different condi tions can be grouped under this heading, only rarely is there a true dystrophy. Chronic forms of SMA (Kugelberg-Welander) and polymyositis are more likely. There are other congenital or metabolic myopathies and there is a genuine scapulohumeral dystrophy. There is a paraneoplastic 'axonal motor neuropathy' causing proximal wasting and weakness, simulating myopathy. An inflammatory polymyositis can also be associated with car cinoma, and there is a rare necrotizing myopathy of rapidly pro gressive course. It can be seen that any limb-girdle syndrome represents only a descriptive diagnosis. Full investigation, including metabolic stud ies and muscle biopsy, is always required. X-linked muscular dystrophies (Duchenne and Becker) The pattern of muscle involvement is similar in both, but the Becker variety starts later and is more benign. Pseudohypertrophy of calves is characteristic (Fig. 15.2 (a)); both affect mainly boys; Duchenne is associated with cardiomyopathy and mental retar dation. Weakness originates around shoulder and pelvic girdles, ultimately spreading more widely. Both classic 'waddling' gait and Gower's sign (Fig. 15.2(b) and (c)) are common. Motor neuron disease Wasting and fasciculation may often be marked in the shoulder girdles when distal muscles appear normal. Never be content with examination of hands or forearms only. Tendon reflexes are usually, but not always, exaggerated. The striking and well-known manoeuvre used to risefrom the floor. Part 3 The motor system F ig . 1 5 .2 (a) C a l f h y p e r tr o p h y i n a p a t i e n t w i t h m u s c u l a r d y s tr o p h y , (b) a n d (c) G o w e r 's s i g n , th e asm (c) Syringomyelia Often starting in the shoulder girdles, there is dissociated sensory loss to pain and temperature, with relative preservation of touch, affecting several spinal segments, and often including the occipital region. Tire reflexes in the upper limbs are absent. Inflammatory lesions Neuralgic amyotrophy Usually, but not invariably unilateral, this starts as a painful con dition sometimes following an illness, injury or inoculation, progressing to marked wasting of the shoulder-girdle muscles and, if present, impairment of sensation is limited to the area supplied by tire circumflex nerve (Fig. 15.3). Wasting may not appear for 2-3 weeks and may progress for months, but recovers over years. The muscles most commonly affected are the serratus, supraspinatus, infraspinatus, deltoid and trapezius. Various combinations are found. Old poliomyelitis Muscles having the same segmental supply are involved. There is no sensory loss, and the history of the onset is typical. 130 c h a r a c t e r i s t i c m e t h o d o f r is in g f r o m th e flo o r. Chapter 15 Muscle bulk ■ ig -15.3 Circumflex nerve sensory loss and periscapular ■.-.'asting in neuralgic amyotrophy. Inflammatory myopathies Myositis may be focal (e.g. Bornholm disease, sarcoidosis) or diffuse (polymyositis), acute or chronic. In the neurology clinic, the type most frequently encountered is the chronic, diffuse polymyositis some times associated with skin disease (dermatomyositis) or a specific vasculitis or collagen vascular disorder. Muscles may be tender in acute cases, the weakness is proximal and usually symmetrical. The erythrocyte sedimentation rate is high in most, but enzyme studies, electromyography and biopsy are mandatory for proper diagnosis. Compressive lesions Spondylotic or neoplastic compression of the lower cervical roots, usually the fifth or sixth, is accompanied by segmental pain and sensory impairment over the outer aspect of the arm. Cauda equina lesions, due to neoplasms, massive disc prolapse or arachnoiditis, cause wasting of the buttocks, loss of sensation in the saddle area and loss of sphincter control. Distal muscle wasting Nearly all the conditions mentioned already, if severe enough, will affect the periphery of the limbs as well. In the early stages, O • Amyotrophic lateral sclerosis • Syringomyelia • Bilateral brachial plexopathy ° Bilateral upper cervical radiculopathy • Juvenile motor neuron disease 131 Part 3 The motor system however, certain diseases may be confined to the distal muscles. One must again stress the difference between diagnostic early features and common end results. The forearm and small muscles of the hands • Wasting results from a lower motor neuron lesion affecting principally the segmental distribution of C7-T1. This may occur at many levels: (i) the anterior horn cell (poliomyelitis, motor neuron disease, syringomyelia, cervical cord tumours); (ii) the anterior root (cervical spondylosis, cervical tumours); (iii) the brachial plexus (injuries, cervical ribs, cervical glandular enlarge ment. superior pulmonary sulcus tumour) and (iv) traumatic lesions of the radial, median and ulnar nerves. • In addition, the median nerve is often compressed in the carpal tunnel. This causes wasting of the thenar eminence and slight sensory loss. If the ulnar nerve is damaged at, or just below, the elbow, there is wasting of most of the small muscles except opponens pollicis and abductor pollicis brevis, and marked sen sory change (Fig. 15.4). Injury at the wrist, or compression by a ganglion, may spare the hypothenar eminence and sensation, though pressure at the point of the lesion can produce paraesthesia over the ulnar distribution. • Muscular causes include myotonic dystrophy and the very rare 'distal muscular dystrophy of Wellander'. Do not forget wasting due to advancing age and arthritis. Very occasionally, wasting occurs in contralateral parietal lobe lesions without necessarily marked paralysis. Fig. 15.4 (a) Wasting of small muscles of hand, (b) Due to ulnar nerve lesion at the elbow. 132 Chapter 15 Muscle bulk The lower leg Isolated peripheral wasting is much less common than in the arm. It occurs as part of an extensive cauda equina lesion, as part of a poly neuropathy and in the early stages of hereditary motor and sensory neuropathy (HMSN). It will follow poliomyelitis and peripheral nerve trauma, especially to the exposed lateral popliteal nerve, e.g. by an inadequately padded plaster, or by skin traction techniques. Peripheral wasting in both upper and lower limbs Peripheral wasting in all four extremities is rare except in two conditions: Peroneal muscular atrophy (Charcot-Marie-Tooth disease) 9 This is the classic clinical syndrome arising from certain types of HMSN and distinguishable from a spinal muscular atrophy only on neurophysiological testing. Characteristically, there is a trans verse demarcation between normal and wasted muscle, often at mid-thigh level, giving the limbs the typical inverted bottle appearance, with similar but less-striking appearances in the upper limbs. Other features, according to type, may include pescavus, areflexia and mild sensory changes such as absent vibration sense at the feet. Despite all these features, disability is remarkably slight. Other chronic polyneuropathies 0 The causes of acquired peripheral neuropathy are, of course, le sion. 0 The chronic conditions are typified by progressive distal weak ness and wasting, but the transverse demarcation is not so obvi ous, pescavus is unlikely, there is no family history, sensory loss is greater and so, usually, is the degree of disability. Individual muscle group and muscle wasting 1 A compressive or traumatic lesion o f the nerve root. Wasting is often slight, sensory changes are almost invariable, but correspond to a lesser area than the segmental supply of the root, due to overlap from neighbouring segments. 2 A traumatic or vascular lesion o f the peripheral nerve. Both wasting and sensory disturbances in the distribution of the nerve are marked, but overlap again occurs. 3 Peripheral nerve lesions in metabolic (e.g. diabetic) or toxic (e.g. lead) neuropath]/. As in 2. 4 Local trauma to muscle.

133 16 Muscle tone Tone, most simply defined as the degree of tension present in a muscle at rest, is not easy to assess because of the variations that take place from one examination to another. The patient must be warm, comfortable and have confidence in the examiner, and this is where kindness and precision prove to be so important in pro ducing relaxation. Using the local phrase for the relaxed state will be much more successful than peremptorily ordering the patient to 'relax', for this usually induces a state of board-like rigidity. Con tinuing with some aspect of the history or merely gossiping about any item of mutual or national interest will put the majority at ease. Testing tone in the upper limbs • First pick up the patient's hand and forearm as if to take the pulse, holding his fingers in the right hand. The fingers should now be submitted to an undulating flexion-extension move ment, passing proximally from the terminal phalanges to and including the wrist. Such movements can be carried out before the patient realizes that it is not simply an examination of the pulse, a gesture so much to be expected that there is no dan ger of muscular tension. Similar movements are then applied to the proximal muscles. Flexion-extension at the elbow is simple enough. • Supination-pronation of the forearm may be more revealing. A sudden catch in the otherwise smooth movement is seen in spasticity. All movements must be gentle. Vigorous and violent movements are quite useless. • Now raise each arm in turn and let it fall back on to the bed, comparing on the two sides the checking movement that usually breaks the fall. This is of particular value in stuporose and uncooperative patients. Testing tone in the lower limbs • First gently roll the limbs with the palms of the hands on the shins. This not only provides an initial assessment of tone in the hip-girdle muscles, but in some curious way it also encour ages relaxation. Look towards the ankles; a floppy side-to-side movement of the feet indicates normal or reduced tone. In 134 Chapter 16 Muscle tone hypertonic states, the ankle and foot move all 'in one piece' as if the joint were 'fixed'. • Then proceed to passive flexion and extension of the hip, knee and ankle. • Place both hands under the knee, briskly flex the joint with an upward movement of the hands. As the knee rises, the heel should slide up the couch. When spastic, the leg rises all-inone. • Then flex the hip, raise the lower leg until it forms more than a right angle at the knee and allow it to fall, noting once again normal checking movements. • Finally, with the flat of the fingers, flap the calf muscles on each side. Loss of tone The muscles are lax; they assume a pendulous shape when allowed to hang freely, offer diminished resistance to passive movement and so widen the range of movement at a joint; they have difficulty in maintaining the position of a limb, allow it to be displaced easily and do not check its sudden release. The tendon reflexes (q.v.) are decreased or absent. In complete relaxation, tone is at its minimum, a state attained so easily by children that their limbs often appear hypotonic. Muscles out of use for long periods also become atonic. In neurological lesions, however, hypotonicity is produced by 1 A breach in the reflex arc 2 Cerebellar disease 3 Cerebral or spinal 'shock', i.e. very soon after a vascular acci dent or trauma C o m m o n causes of hypotonia 1 Lesions o f the motor side o f the reflex arc. Anterior horn cell disease, neuropathies, peripheral nerve injuries. 2 Lesions o f the sensory side o f the reflex arc. Neuropathies, tabes dor salis, herpes zoster. 3 Combined motor and sensory lesions. Syringomyelia, cord or root compression, gross cord destruction. 4 Lesions o f the muscle itself. Myopathies, spinal muscular atro phies, myasthenia gravis. In periodic paralysis, during an attack, the muscles, though paralysed and areflexic, feel tense, swollen and entirely different from the flabby flaccid feeling ir other hypotonic states. Part 3 The motor system 5 States o f neurological 'shock'. The earliest stages of a severe cord lesion, or a profound hemiplegia. This is temporary unless destruction is extreme. 6 Cerebellar lesions. Ipsilateral hypotonia is common, but rarely very marked, and the reflexes are prolonged and pendular rather than lost. 7 Chorea. In Sydenham's chorea, the involuntary movements are associated with marked hyperextensibility of the joints, espe cially the wrists and fingers, but reflexes are often retained. Increase of tone There are three main types of hypertonicity: 1 In the 'spastic' type, tone and resistance are greater in one group of muscles (e.g. the quadriceps) than in the antagonists (e.g. the hamstrings). The resistance is usually most noticeable when the movement is first made, and then is suddenly overcome, pro ducing the so-called clasp knife effect, most easily demonstrated at the elbow and knee. Supination-pronation of the forearm will reveal the so-called supinator catch. Affected muscles are compact at rest, feel firm, do not flap on palpation and tend to form contractures. This is a sign of an upper motor neuron, pyramidal pathway, lesion; hypertonicity is more evident in the flexor muscles and pronators in the upper limb, and the extensors and adductors in the lower limb. 2 In the 'plastic' type, there is equal resistance in both agonists and antagonists at any point, with the result that the same degree of hypertonicity is felt throughout each movement - the so-called plastic or lead-pipe rigidity. When this becomes very marked, a state of true rigidity is reached in which the muscles, though not greatly altered in appearance, hold the limb in com plete immobility. Although this may be seen in extreme spasticity from upper motor neuron lesions, it is a characteristic feature of a lesion of the extrapyramidal system. 3 In the third type, the agonists and antagonists contract alter nately, rapidly and regularly during the movement, producing the so-called cog-wheel rigidity. This may be present only dur ing the first moments of testing, and perhaps only at the wrist, so that the examiner must be on the alert for it from the very outset. It is a valuable sign of extrapyramidal disease, and may be felt in the absence of any visible tremor, though it is much increased when tremor is present. 136 Chapter 16 Muscle tone Sometimes, rigidity is only brought out by voluntary move ments in the opposite limb (Froment's sign). Ask the patient to perform opening and closing of the fist, abduction-adduction or pronation and supination of the opposite hand. Simultane ously examine for the tone at the wrist and elbow. 4 A type of increased muscle tone known as paratonia (gegenhalten) is seen in diffuse frontal lobe (particularly medial and premotor cortex, Broadman area) disease. In this, when a patient is asked to relax a joint to allow movement (e.g. at wrist or elbow), the involved muscles involuntarily contract instead, and the patient appears to be actively opposing any movement of the joint by the examiner. The resistance increases in propor tion to the speed and strength with which the exam iner tries to move the joint. Clonus ^ J' V£ Sudden stretching of hypertonic muscles produces redex contrac tion. If the stretch is maintained during the subsequent relaxation, further reflex contraction occurs and this may continue almost indefinitely, unless the stretch stimulus is released. It is most easily demonstrated by dorsiflexing the foot (ankle clonus) or by sharply moving the patella downwards (patellar clonus), but it may be present at any joint. There is nothing diagnostic about clonus. It merely represents an increase in reflex excitability and may be pres ent in a very tense patient, one who has been straining his mus cles, or one who has had a fright (e.g. the ankle clonus that most car drivers have experienced in their right foot after an alarming experience). Under these circumstances, it rarely becomes well sus tained, but as a physical sign it is only of great significance together with other signs of an upper motor neuron lesion. However, even in their absence, the presence of clonus on one side only may give a hint of a pyramidal system lesion on that side. Lesions producing increased tone Here, the results of experimentation differ from tire traditions of clinical experience, but providing one is aware of the limitations of one's knowledge; for practical purposes, it is wise to retain the view that a lesion of the pyramidal system produces spasticity, increased tendon reflexes, absent abdominal reflexes and extensor plantars, because long experience has proven its value in locational diagnosis. Increase in tone is produced by any destructive lesion of the upper motor neuron. This includes cerebral thromboses, haemorrhages, 137 Part 3 The motor system tumours, degenerative diseases, inflammatory lesions and injuries; spinal cord tumours, compressions, injuries and degenerative dis eases. Note, however, that immediately after a gross lesion of the corticospinal tracts tone may be lost, only to become increased after several days. The plastic type of rigidity occurs in parkinsonism; in some rare instances of basal ganglia neoplasms; and in catatonia. Cog-wheel rigidity occurs in the varieties of parkinsonism, including with diffuse cerebrovascular disease, but it does also occur after high dosage of reserpine or chlorpromazine and its derivatives and after carbon monoxide poisoning. Extrapyramidal types of hypertonicity are not infrequent in 'nor mal' elderly patients. Difficulties • A few moments spent in a gentle, friendly, even light-hearted manner, in establishing rapport will save many struggling with that failure of relaxation which defeats the estimation of tone. Re-examination, when the patient knows it will not be unpleas ant, may completely alter one's impression. • It is very difficult to assess minor degrees of hypotonicity. Other features, such as diminution of reflexes, have to be found before pathological conclusions can be drawn from limbs that are merely limp. • Joint changes or muscle contractures may give false impressions of increased tone which emphasizes the importance of feeling the muscles themselves and examining the joints for evidence of local disease. • Another impression of increased tone arises when there is gross loss of postural sensibility. While the examiner is trying passively to move the limb, the patient appears to be fighting against him. This is an important sign, rarely described (see p. 180). Myotonia This is a state in which muscle contraction continues beyond the period of time required for a particular movement to be made. It is best seen in the face and hand muscles. Ask the patient to screw up his eyes, or show his teeth, and then to 'let go'. There is a delay in relaxation so that the patient appears to have not understood the instruction. Ask him to grip the fingers, and then suddenly release his hold. This will result in flexion of the wrist, adduction and opposition of 138 Chapter 16 Muscle tone the thumb and incomplete extension of the fingers. He has to drag his hand away. Repeating the movement several times may over come the myotonia, so look carefully the first time. Percussing the tongue has already been mentioned (p. 122), but percussion of the thenar eminence results in slow adduction of the thumb and dimpling of the muscle, while percussion of other muscles may produce dimpling at first, followed by a raised lump to the side of the dimple. Merely a lump on hard percussion may occur as a result of myoedema in normal individuals. Myotonic states The most common conditions are: • Myotonic dystrophy in which there is myopathic facies, wasting of stemomastoids and distal involvement in the limbs (which may outweigh the myotonia), baldness, cataracts, testicular atrophy and steady progression. • Myotonia congenita. The myotonia is present from early years, infancy in Thomsen's disease or from childhood in the recessive Becker type. There is generalized myotonia with muscle hyper trophy (p. 127) but without dystrophic features. Characteristi cally, as the patient moves away from a stationary position, the myotonia freezes him in his tracks for a moment after which he is able to move briskly and normally. • In paramyotonia congenital, exposure to cold causes marked myo tonia in the exposed muscles. There may also be intermittent muscle paralysis, and there is a closer relationship to periodic paralysis than to the other myotonic disorders.

139 17 Muscle power Because muscles work in combination with other muscles, and testing of movements may mean testing several muscles, it is easy to be vague, both in testing and describing the results of testing a patient's strength. This remark applies to neurologists as well as to other physicians. To avoid this, the examiner must discipline him self to ask the following questions at each stage: 1 What muscle, or muscle group, am I about to test? 2 Is the limb in the right position for that muscle alone to be tested? 3 What is the segmental nerve supply of that muscle? 4 Which peripheral nerve supplies it? Once the positioning is correct, the patient must be told clearly the movement he is to make, possibly illustrating it for him first. The test of power can then be carried out in three ways: 1 Iu a fu lly contracted muscle. The patient first completes the move ment and then tries to maintain the muscle in full contraction while the examiner tries to overcome it. This method puts the muscle at maximum advantage. 2 In a fu lly relaxed muscle. The patient is asked to contract a fully relaxed muscle (e.g. asking the patient to flex his fully outstretched elbow) and the examiner resists the movement throughout the whole of the patient's attempt to contract it. This method puts the muscle at maximum disadvantage and may detect mild degrees of weakness. It is also a method some patients fail to understand and a false impression of weakness, or lack of cooperation, may be gained. 3 In a mid-contracted muscle. The patient is asked to partially con tract the muscle (e.g. flexing his mid-flexed elbow) and the examiner resists the movement. This method puts the muscle neither at maximum advantage nor disadvantage. Most of the large muscles are usually tested using method 3, whereas small muscles are tested using method 1. Steady exertion is required by both the patient and the examiner. Sudden move ments serve only to confuse. Application of great force is unneces sary, and indeed, in hypertonic muscles undesirable, for very pain ful cramps may easily be produced. While these tests are being carried out, further questions must be asked: 1 Is this muscle as strong as might be expected, bearing in mind the build and age of both the patient and the examiner? 140 Chapter 17 Muscle power 2 Is it as strong as the same muscle on the other side? 3 What is the degree of weakness, if any? (See below.) 4 Is the weakness constant or variable? Does it improve on rest or on encouragement? 5 Is there any painful condition (e.g. injury), or mechanical defect (e.g. ankylosis of a joint, or contracture of an antagonist), which hinders the movement? 6 Are the actions the patient is known to be able to carry out compatible with any apparent weakness demonstrated? (For example, has the patient, whose hands on formal examination are apparently almost paralyzed, just undressed himself?) Quantitative assessment of weakness The assessment of power may differ strikingly in the record of one examiner from that of another. For this reason, many classi fications of degrees of weakness have been suggested. None has been ideal, but the use of the scheme supported by the Medi cal Research Council at least ensures some degree of uniformity. Power is recorded by numbers ranging from the normal of 5 to complete paralysis represented by 0. It is worth remembering that even a very poorly developed individual is usually able to resist an examiner's attempt to overcome the power of a fully contracted muscle. 5 = Normal power 4 = The muscle, though able to make its full normal movement, is overcome by resistance 3 = The muscle is able to make its normal movement against grav ity, but not against additional resistance 2 = The muscle can only make its normal movement when the limb is so positioned that gravity is eliminated 1 = There is a visible or palpable flicker of contraction, but no resul tant movement of limb or joint 0 = Total paralysis Routine tests of muscle groups It is customary to direct attention first to major groups of muscles. These are the flexors and extensors of the neck; the adductors, abductors and rotators of the shoulder; the flexors and extensors of the elbow, wrist and fingers; the grip; the abdominal muscles; the extensors of the spine; the flexors and extensors of hip and knee; the dorsiflexors and plantar flexors of the feet and the flexors and extensors of the toes, particularly the great toe. 141 Part 3 The motor system Any weakness discovered is then more carefully analysed by car rying out the appropriate tests for individual muscles concerned in making the defective movement. It is here that the positioning of the limb is of such great importance. Testing individual muscles Full details of the actions of individual muscles are given in the textbooks of anatomy. The following pages deal with those muscles that are commonly of help in neurological diagnosis. The illustra tions are intended to show the movement required to bring a mus cle into action rather than to demonstrate a particularly prominent muscle belly. Normal individuals, without outstanding muscular development, have been photographed. The movements recommended should be carefully followed, for many patients learn tricks to overcome disability, which may cause confusion if the purest action of a muscle is not tested. Seg mental supply is subject to individual variation, and indeed is not necessarily universally agreed; simplification has therefore been attempted by giving the segmental supply most frequently found to be relevant in clinical practice. Muscles of the head and neck The facial muscles, jaw muscles, sternomastoids and trapezii are dealt with under the appropriate cranial nerve. Muscles of the shoulder girdle and scapula (Figs. 17.1-17.7) Fig. 17.1 M uscle: Deltoid. M ain segmental nerve supply: C5. Peripheral nerve: Circumflex. Test: The patient holds his arm abducted to 60° against the exam iner's resistance. 142 Chapter 17 Muscle power Fig. 17.2 M uscle: Supraspinatus. Main segmental supply: C5. Peripheral nerve: Suprascapular. Test: The patient tries to initiate abduction of the arm from the side against resistance. Fig. 17.3 M uscle: Infraspinatus. Main segmental supply: C5. Peripheral neive: Suprascapular. Test: The patient flexes his elbow, holds the elbow to his side and then attempts to turn the forearm backwards against resistance. Fig. 17.4 M uscle: Rhomboids. Main segmental supply: C5. Peripheral nerve: Nerve to rhomboids. Test: Hand on hip, the patient tries to force his elbow backwards. 143 Part 3 The motor system Fig. 17.5 Muscle: Serratus anterior. Main segmental supply: C 5 ,6 ,7 . Peripheral nerve: Nerve to serratus anterior. Test: Tire patient pushes his arms forwards against firm obstruction. Fig. 17.6 M uscle: Pectoralis major. Main segmental supply: C6, 7 ,8 . Peripheral neroe: Lateral and medial pectoral nerves. Test: Placing the hand on the hip and pressing inwards, the sternocostal part of the muscle can be seen and felt to contract. Raising the arm forwards above 90° and attempting to adduct it against resistance brings the clavicular portion into action. Fig. 17.7 M uscle: Latissimus dorsi. Main segm ental supply: C 7. Peripheral neroe: Nerve to latissimus dorsi. Test: (i) While palpating the muscles, ask the patient to cough, (ii) Resist the patient's attempt to adduct the arm when abducted to above 90°. 144 Chapter 17 Muscle power Muscles of the elbow joint (Figs. 17.8-17.10) M uscle: B ic e p s . Main segmental supply: C 5 . Peripheral nerve: F ig . 1 7 .8 M u s c u lo c u ta n e o u s . Test: T h e p a t i e n t f le x e s h i s e lb o w a g a in s t r e s is t a n c e , th e f o r e a r m b e i n g s u p in a t e d . M uscle: B r a c h i o r a d ia lis . Main segmental supply: C 5 , 6 . Peripheral nerve: F ig . 1 7 .9 R a d ia l. T e s t: T h e p a t i e n t p r o n a t e s th e fo re a rm a n d d ra w s th e th u m b to w a rd s th e n o s e a g a in s t r e s is t a n c e . M uscle: T r ic e p s . Main segm ental supply: C7. Peripheral nerve: R a d ia l. Test: T h e F ig . 1 7 .1 0 p a t i e n t a t t e m p t s to e x t e n d t h e e lb o w a g a in s t r e s is t a n c e . 145 Part 3 The motor system Muscles of the forearm and wrist joint (Figs. 17.11-17.15) Fig. 17.11 M uscle Extensor carpi radialis longus Main segmental supply C6, 7 peripheral nerve Radial,; Test - The patient holds the fingers partially extended and dorsiflesxes the wrist towards the radial side against resistance. Fig. 17.12 Muscle: Extensor carpi ulnaris. Main segmental supply: C7. Peripheral nerve: Radial. Test: As in Fig. 17.11, but dorsiflexion must be towards the ulnar side. Fig. 17.13 M uscle: Extensor digitorum. M ain segmental supply: C 7 . Peripheral nerve: Radial. Test: The examiner attempts to flex the patient's extended fingers at the metacarpophalangeal joints. Fig. 17.14 M uscle: Flexor carpi radialis. Main segm ental supply: C6, 7. Peripheral nerve: Median. Test: The examiner resists the patient's attempts to flex the w rist towards the radial side. Palmaris longus is also shown. 146 Chapter 17 Muscle power I ig. 17.15 Muscle: Flexor carpi ulnaris. Main segm ental supply: C 8. Peripheral nerve: Ulnar. Test: This m uscle is best seen w hile testing the abductor digiti minimi, w here it fixes its point o f origin. Muscles of the thumb (Figs. 17.16-17.22) N ote. A b d u ctio n o f th e th u m b is the m o v e m e n t th at b rin g s the th u m b to a rig h t a n g le w ith the p alm . E x ten sio n o f the th u m b d raw s the th u m b a w a y in the sa m e p lan e as the p alm . Fig. 17.16 Muscle: Abductor pollicis longus. M ain segmental supply: C8. Peripheral nerve: Radial. Test: The patient attempts to maintain his thumb in abduction against the exam iner's resistance. Fig. 17.17 Muscle: Extensor pollicis brevis. M ain segmental supply: C8. Peripheral nerve: Radial. Test: T he patient attem pts to extend the thum b while the exam iner attempts to flex it at the metaca rpophalangeal joint. 147 Part 3 The motor system Fig. 17.18 M uscle: Extensor pollicis longus. Main segmental supply: C8. Peripheral nerve: Radial. Test: The patient attempts to extend the thumb while the examiner attempts to flex it at the interphalangeal joint. Fig. 17.19 M uscle: Opponens pollicis. Main segmental supply: T l. Peripheral nerve: Median. Test: The patient attempts to touch the little finger with the thumb; preserved in ulnar nerve lesions when the rest of the hand appears very wasted. Fig. 17.20 Muscle: Abductor pollicis brevis. Main segmental supply: T l. Peripheral nei-ve: Median. Test: First place some object between the thumb and the base of the forefinger to prevent full adduction; then the patient attempts to raise the edge of the thumb vertically above the starting point, against resistance. This is an important muscle, being the first to show weakness in the common carpal tunnel syndrome. 148 Chapter 17 Muscle power Fig. 17.21 Muscle: Flexor pollicis longus. Main segm ental supply: C8. Peripheral nerve: Median. Test: An attempt to extend the distal phalanx of the thumb against the patient's resistance. It is w ise to hold the proximal phalanx. Fig. 17.22 Muscle: Adductor pollicis. Main segm ental supply: Tl. Peripheral nerve: Ulnar. Test: The patient attempts to hold a piece of paper betw een the thumb and th e palmar aspect of the forefinger. Muscles of the hand and fingers (Figs. 17.23-17.27) Fig. 17.23 M uscles: Lumbricals and interossei. Main segm ental supply: C8, T l . Peripheral nerves: M edian (lumbricals I and II); ulnar (interossei, lumbricals III and IV). Test: (a) The patient tries to flex th e extended fingers at the metacarpophalangeal joints (lumbricals). 149 Part 3 The motor system (b) N e x t t h e p a t i e n t a t t e m p t s to k e e p th e f in g e r s a b d u c t e d a g a in s t r e s is t a n c e (in t e r o s s e i) . Fig. 17.24 M uscles: First dorsal interosseus and first palmar interosseus. Main segmental supply: T1. Peripheral nerve: Ulnar. Test: Place the hand flat on a table. The patient then tries to abduct (illustrated) and adduct the forefinger against resistance. This test can be applied to other fingers, but the muscles are not easily visible. Fig. 17.25 M uscle: Flexor digitorum sublimis. Main segmental supply: C 8 . Peripheral nerve: Median. Test: The patient flexes the fingers at the proximal interphalangeal joint against resistance from the exam iner's fingers placed on the middle phalanx. 150 Chapter 17 Muscle power Fig. 17.26 Muscle: Flexor digitorum profundus. Main segmental supply: C8. Peripheral nerves: Median (I and II), ulnar (III and IV). Test: The patient flexes the terminal phalanx of the fingers against resistance, the middle phalanx being supported. Fig. 17.27 Muscle: Abductor digiti m inim i. Main segmental supply: T l. Peripheral nerve: Ulnar. Test: The back of the hand is placed on the table and the little finger abducted against resistance (see also F ig. 17.15), often the o n ly sign of an ulnar lesion. M u scles of the trunk Illustrations of these muscles are not helpful. M u scles: Extensors of the spine M ain seg m en tal su p p l y : All segments P erip h eral n erv es: Posterior rami of spinal nerves Test: The patient lies on his face and then attempts to raise his shoulders off the bed. M u scles: Intercostals M ain seg m en tal su p p l y : T1-T12 P erip h eral n erves: Intercostal nerves Test: A difficult test. Observe the movements of the ribs on expi ration and inspiration and the movements of the muscles in the intercostal spaces. M u scles: Abdominal muscles 151 Part 3 The motor system M ain seg m en tal su p p l y : T 5 -L 1 P erip h eral n erves: Intercostal, ilioinguinal, iliohypogastric nerves Test: T h e p a tien t lie s on h is b a ck an d attem p ts to raise the head ag ain st lig h t resistan ce. W atch th e m o v e m e n t o f th e u m bilicu s. Muscles of the hip girdle (Figs. 17.28-17.31) Fig. 17.2S M uscle: Iliopsoas. Main segmental supply: L I, 2 ,3 . Peripheral nerve: Femoral. Test: The patient lies on his back and attempts to flex his thigh against resistance. Similarly, with the hip fully flexed, he resists attempts to extend it. Fig. 17.29 M uscle: Adductor femoris. Main segmental supply: L5, S I . Peripheral nerve: Obturator. Test: The patient attempts to adduct the leg against resistance. Fig. 17.30 M uscles: Gluteus medius and minimus. Main segmental supply: L2, 3. Peripheral nerve: Superior gluteal. Test: The patient, lying face down, flexes the knee and then forces the foot outwards against resistance. These muscles also abduct the extended leg. 152 Chapter 17 Muscle power Fig. 17.31 Muscle: Gluteus maximus. Main segmental supply: L5, S I . Peripheral nerve: Inferior gluteal. Test: T h e patient, still lying o n his stomach, should tighten the buttocks so that each can be palpated and compared; he must then try to raise the th ig h against resistance with the leg flexed at the knee. H aving the knee flexed minimizes any contribution from the ham strings, which is im portant in caudal equina and conus m edullans lesions. Muscles of the thigh and knees (Figs. 17.32 and 17.33) Fig. 1 7 .3 2 M uscles: H am strings (biceps, sem itendhosus, semimembranosus). Main segm en tal supply: L 4 ,5, S I , 2 Peripheral nerve: Sciatic. Test: The patient, Iy.ng on his sto m ach , cttempts to flex the tcree against resistan ce.T h e biceps is seen l a ttrally, the sem i teid in osu s m edially. Fig. 1 7 .3 3 Muscle: Q u a d r ic e p femoris. Main segm en tal supply: L 3 ,4. P e r ip e r a l nerve: Femoral. TsA The patient, ly in g on his back, n tem p ts to extend tie knee against re-sstance. 153 Part 3 The motor system M uscles of the low er leg and ankle (Figs. 17.34-17.37) Note. The sciatic nerve divides into the medial and lateral popliteal nerves. The lateral popliteal further divides into anterior tibial and musculocutaneous branches. Fig. 17.34 Muscle: Tibialis anticus. Main segmental supply: L4, 5. Peripheral nerve: Anterior tibial. Test: The patient dorsiflexes his foot against the resistance of the examiner's hand placed across the dorsum of the foot. Fig. 17.35 Muscle: Tibialis posticus. Main segmental supply: L4. Peripheral nerve: Medial popliteal. Test: The patient plantar-flexes the foot slightly and then tries to invert it against resistance. Fig. 17.36 Muscle: Peronei. Main segmental supply: L5, S I. Peripheral nerve: Musculocutaneous (principally). Test: The patient everts the foot against resistance. Isolated weakness may be the earliest sign of peroneal muscular atrophy. 154 Chapter 17 Muscle power Fig. 17.37 Muscle: Gastrocnemius. Main segmental supply: SI. Peripheral nerve: Media' popliteal. Test: The patient plantarflexes the foot against resistance. Muscles of the foot and great toe (Figs. 17.33-17.41) Fig. 17.38 M uscle: Extensor digitorum longus. Ma/h segmental supply: L5. Peripheral nerve: Anterior tibial. Test: The patient dorsiflexes the toes against resistance. Fig. 17.39 M uscle: Flexor digitorum longus. Main segmental supply: S I, 2. Peripheral nerve: Medical popliteal. Test: The patient ilexes the terminal phalanges against resistance. Fig. 17.40 M uscle: Extensor hallucis longus. Main segm ental supply: L5, S I. Peripheral nerve: Anterior tibial. Test: The patient attempts to dorsiflex the great toe against resistance. 155 Part 3 The motor system Fig. 17.41 M uscle: Extensor digitorum brevis. Main segmental supply: S I. Peripheral nerve: Anterior tibial. Test: The patient dorsiflexes the great toe against resistance (especially middle 3 toes). Types of muscular weakness To repeat the same statement for each group or muscle in turn would be pointless, but there are certain general principles that help to distinguish different types of weakness. Weakness due to pyramidal tract lesions This tends to be a weakness that is incomplete except in the acute stages, or in the presence of a grossly destructive lesion. It affects par ticular movements rather than particular muscles, and is most marked in the abductors and extensors of the upper limb, and the flexors of the lower limb. Normally it is associated with increase of tone and exag gerated reflexes. Distribution is more distal than proximal, particu larly in the upper limbs, where hand movements are affected earliest. Weakness due to extrapyramidal lesions This is more of a hindrance to movement due to equal resistance from agonists and antagonists, than to true loss of muscle power. It is generalized throughout the limb and associated with rigidity and often with resultant suppression of the reflexes. Weakness due to lower motor neuron lesions This is usually very marked, but, except in extensive polyneurop athies, is limited to the muscles having that segmental supply. If of any standing, it is associated with marked wasting and loss of those tendon reflexes in which the affected muscles play a part. A lesion at anterior horn or anterior root level picks out those muscles whose sole or maximal supply is from that segment, and these muscles may show fasciculation. At peripheral nerve level, it affects all the muscles supplied by that nerve. In a polyneuropathy, this type of weakness is often maximal peripherally in the arms and legs, and usually symmetrical. 156 Chapter 17 Muscle power © • Common peroneal neuropathy • Sciatic neuropathy • Low er lum bosacral plexopathy • L4-L5 radiculopathy • Distal myopathy Weakness due to muscular lesions This can range from weakness of one muscle, such as after a local injury, to weakness of every muscle, such as in some cases of polymyositis. This type of weakness is either very localized or very widespread but patchy. The muscles affected correspond either to the supply of a particular spinal segment or a particular peripheral nerve. There is often individual muscle wasting, pseudohypertro phy or tenderness. The related reflexes are lost £ Myasthenia * P itied ^ Though this word, in the strict sense, means merely muscular weakness, by custom it has come to mean that type of muscle weakness seen in myasthenia gravis, where the degree of weakness varies from hour to hour, increases as the muscle is repeatedly used, even to the extent of total paralysis, and yet recovers to its previous con dition after a very short period of rest. This phenomenon, though capable of affecting any muscle in the body, is most commonly seen in the eyelids, the external ocular muscles, the facial muscles, the muscles of the tongue, throat and larynx, the muscles of the back, the shoulder girdle and the hand. Any of these should be tested for myasthenia either by repetition of a given action, such as maintain ing upward deviation of the eyes for testing the eyelids, counting successively up to 100 for the bulbar muscles, or repeatedly sitting up and lying down for the back muscles. The diagnosis can be confirmed by the intravenous injection of 10 mg of edrophonium chloride (Tensilon) when power returns within 1 minute (Fig. 17.42), the effect usually lasting only about 5 minutes, though in some patients it may persist longer. Eye mus cle weakness responds less completely than limb muscles. An injec tion subcutaneously of 2.5 mg of neostigmine (Fig. 17.43 (a)-(c)) is another striking test, when almost maximum power may be restored, but taking up to 45 minutes to do so. The effect may last 4 hours or longer. To minimize bowel discomfort, 0.6 mg of atro pine should be included. After either of these injections, fasciculation may be seen in unaffected muscle, but if it occurs throughout Part 3 The motor system Fig. 17.42 Effects of edrophonium chloride (Tensilon) on myasthenia gravis: (a) before injection and (b) 60 seconds after injection. all muscles, myasthenia gravis becomes unlikely, but not impos sible, however, if only the eye muscles are weak. In myasthenic syndromes associated with carcinoma (Lambert-Eaton syndrome), muscle strength temporarily increases with repetition, and there is no dramatic response to Tensilon. In contrast to myasthenia gravis, weakness of the limbs, particularly lower, is commoner than ocular or bulbar presentation. Cholinergic crises An important word of caution: in patients known to have myas thenia gravis who are needing increasing dosage of anticholines terases, increasing weakness may be a warning sign of impending cholinergic crisis, rather than worsening myasthenia. Always be on the alert for pallor, sweating, constricted pupils, hypersalivation and bradycardia. A very cautious test dose of intravenous edropho nium, using only small amounts (e.g. 1 mg) at a time will improve the situation in myasthenia and worsen it in cholinergic crises. Simply increasing the dosage of neostigmine or similar drugs may produce respiratory failure. Hysterical weakness This varies considerably both in degree and distribution, but never corresponds to a set pattern of nerve supply, nor does it follow the proper 'pyramidal' distribution. Movements are affected rather 158 Chapter 17 Muscle power (b) (c) Fig. 17.43 Effects of neostigmine on myasthenia gravis: (a) before injection, (b) 10 minutes after injection and (c) 20 minutes after injection. These photographs are intended also as a reminder that this disease can affect the very young. than individual muscles, most commonly involving both flexion and extension around a particular joint, e.g. at the knee or shoul der. The object of the examination, therefore, must be to note the distribution of the paralysis, the muscles affected and to discover whether the patient can still use those affected muscles to perform movements that he does not realize entail their use. Furthermore, the antagonists to the muscles being tested are in action simultane ously, and this produces tremor. When assessing wrist extension, perhaps, it is possible to feel strong contraction of the flexor muscles as one supports the forearm being tested. The power exerted by the patient is proportional to that exerted by the examiner, so that all degrees of strength produce the same failure of movement, but that failure varies from moment to moment. The wrist may suddenly collapse, usually in a jerky fashion; dorsiflexion of the foot is sud denly 'let go'. Grimacing or protests of pain may accompany the examination, and a request to grasp the examiner's hand, e.g. is usually accompanied by a 'shunting of effort' so that the muscles of the upper limb, shoulder and face are brought into powerful play, but the fingers of the affected hand remain limp and useless. The grimacing, clenching of the teeth and holding of the breath can be quite characteristic. In an extreme case, when the patient is in bed a request to raise a leg is followed by a preliminary ritual in which the patient takes a breath, holds it, clutches the side of the mat tress with his hands and strains with effort before finally collapsing 159 Part 3 The motor system back, puffing and 'exhausted'. By watching the patient out of bed, he can be seen to be carrying out actions that would be impossible if the degree of weakness just shown on examination was genu inely present. Thus, a patient in bed who is apparently unable to either dorsiflex or plantar-flex the feet may be able to walk on his heels or toes. When a lower limb is paralyzed, a test devised by Babinski is often very useful. A patient, lying in bed, is asked to raise himself to a sitting position while holding Inis arms across his abdomen. Normally, to do so the heels are pressed into the bed. hi organic hemiplegia, there is involuntary elevation of the paretic limb, as the heel cannot be pressed downwards. In hysteria, the sound leg may be raised, the paralyzed leg pressing into the bed. If the examiner's hand is placed under the heel of a paralyzed leg in hysteria, there may be no response when the patient is requested to press upon the observer's hand; but when he is asked to raise his sound limb, pressure may be felt. It is axiomatic that a patient whose elbow flexion or extension can be prevented by pressure from the dorsal surface of the examiner's little finger should not be able to dress or undress if such weakness were real. Indeed, in hysterical paralysis the patient appears to be even more helpless than a patient with an organic hemiparesis, e.g. a hysterical patient will make little effort to overcome the disability and rely wholly on others to assist in undressing and dressing. The tendency to calm unconcern contrasts strongly with the distress of a patient with organic paresis.

160 18 Posture, stance, spinal movement and gait The best time to observe the stance and gait is when the patient does not realize that he is being examined, because on formal test ing the average person assumes a most unnatural mode of both standing and walking. Watch the patient's movements closely on entering the room, and even more closely as he leaves. Simulated limps and other defects are frequently forgotten at the time of departure. Observations through a window of gait when leaving the building may be revealing. In the ward, a casual visit may often be more informative in this respect than a set ward round. Posture and stance Formal testing is usually postponed to the end of the examination. The patient, if well enough, is asked to stand up and the position he naturally adopts is noted. Then ask him to bring both feet close together, the heels and toes touching. Watch the ease with which he does this and how well the position is maintained. Now ask him to close his eyes, assuring him that he will not be allowed to fall if he feels unsteady (and being ready to fulfil this promise). Remember that many people feel a slight sense of instability in these circumstances, and that this increases with age. Ask the patient to turn around and note whether this movement disturbs his equilibrium. The opportunity can then be taken to examine the back and spine. The back Note the presence of kyphosis, scoliosis or abnormal lordosis, and then ask the patient to bend forwards to touch his toes. Again, note the line of the spinous processes, the conversion of the lumbar lor dosis to a smooth curve and the ease with which each part of the spine flexes. He should then flex the spine laterally in both direc tions. Palpate and lightly percuss the spinous processes and the paraspinal muscles. If a muscular dystrophy or weakness of the back muscles is sus pected, the patient should be told to squat down on his haunches, and to stand up again. If he can do this, he should then be asked first to lie flat on his back and then to get up on to his feet again. Most 161 Part 3 The motor system normal people will flex their hips and knees, raise their shoulders, place their hands behind them and push themselves forwards on to their feet, and so regain the upright position, possibly turning on to one side to gain better leverage. Abnorm alities of posture, stance and spinal m ovem ent There are many conditions other than neurological disease that alter the patient's posture. These cannot be dealt with here and mention will merely be made of those abnormalities which fairly frequently bear relationship to nervous disease, with an indication of the disorders that may be suspected. Bradykinesia This physical sign, seen principally in parkinsonism, comprises poverty and slowness of movement in the absence of weakness. There is particularly intense slowing of the initiation of volun tary movements of limbs and trunk, and of the many natural little movements that characterize human behaviour. It results in immo bility of expression and of posture, great difficulty in changing pos ture, turning over in bed etc., and inability to make rapid changes in movement of any sort. It is best seen simply by observing the patient's usual behaviour rather than by set tests. A stooped position This is common with age, excessive height, poor muscular devel opment, some psychotic states and in patients who have for years been under the control of overbearing relatives. In severe parkinsonism, the stoop affects mainly the upper spine, the head and neck being held forwards and the arms flexed at the side or in front of the body. In some cases of motor neuron disease, myasthenia gravis or polymyositis, the neck muscles are so weak that the head falls forward, and the patient can only look ahead by turning his eyes upwards. Kyphoscoliosis A kyphotic spine does not necessarily give the impression of stooping, but in ankylosing spondylitis the stoop may be extreme. Scoliosis is common in Friedreich's ataxia, muscular dystrophies, syringomyelia and von Recklinghausen's disease. 162 Chapter 18 Posture, stance, spinal movement and gait Gross kyphoscoliosis is capable of so distorting the theca as to cause cord compression and paraplegia. In lumbar disc disease, a scoliosis tends to be convex to the side of the lesion, it may be maximal at the affected level, and is greatly increased by bending forwards. If a congenital hemivertebra is present, there is extreme scoliosis on forward flexion. This may be of no significance, but in the lumbar region it is sometimes accom panied by disc disease. Excessive lordosis This occurs in muscular dystrophies, in some cases of generalized myasthenia gravis and in congenital hip disease. It may also be a normal racial characteristic. A rigid spine On forward flexion, the lumbar spine remains quite straight when there is paravertebral muscle spasm resulting from lumbar spine or disc disease. Flexion towards the side of the disc lesion increases the pain. In ankylosing spondylitis, the whole spine moves as one and flexion occurs at the hip joints. Many patients who have been wearing spinal supports, cervical or lumbar, for a long time develop a state of rigidity of their spinal movements, which can be found even when there is very little wrong with the spine. A tender spine Local bony tumours and infections (such as that accompany extra dural spinal abscesses) cause localized tenderness on percussion. Some spinous processes, however, are always tender. Patients who constantly flinch when the paraspinal muscles are palpated, but are otherwise well, rarely have organic disease. Abnorm alities of equilibrium Complete inability to remain upright is seen with lesions around the cerebellar vermis and IVth ventricle, and may be out of all pro portion to any ataxia found while the patient was recumbent. It may also be a hysterical manifestation. In this case, the posture is bizarre and varies. It is at its worst when examined and yet, while dressing, the patient may be capable of standing and pulling his shirt over his head without falling. Other hysterical symptoms are usually present. 163 Part 3 The motor system Falling to one side is seen in vestibular and cerebellar hemisphere lesions. Falling backwards is seen in elderly patients and those with parkinsonism (e.g. progressive supranuclear palsy); in lesions involving the cerebellar vermis and in foramen magnum lesions associated with descent of the cerebellar tonsils-such as basilar invagination and the Arnold-Chiari deformity. Such patients may be able to remain standing until they look upwards; others may find their legs becoming suddenly useless on coughing or sneezing. Romberg sign Tire patient sways from the heels, slightly when the eyes are open, but very markedly when the eyes are closed, to tire extent that he will either fall or separate his legs to achieve a broader base. Most experts discount the sway at the hips, and insist on seeing the sway at the ankles for a positive test. However, expert opinion varies to some extent as to how the Romberg test is performed and inter preted. Some experts test with the patients holding the arm out stretched in front but it is not what Romberg had described. It is characteristic of proprioceptive deficiency and is found par ticularly in posterior cord compression at a high level, sensory polyneuropathies, subacute combined degeneration of the spinal cord and tabes dorsalis. Some degree of swaying is not uncommon with advancing age, and in psychoneurosis. In the latter, distract ing the patient's attention may stop it (e.g. by repetitive tapping over the vertex). Squatting and standing up Weakness of the proximal leg and paraspinal muscles makes it impossible to rise from squatting. This is seen particularly in mus cular dystrophies, but

also in some cases of myasthenia gravis, polymyositis and chronic polyneuropathies. On attempting to stand up from lying flat, such patients will first turn on their face, draw their legs forwards towards their arms, place their hands on the lower part of their legs and push them selves upright by placing the hands at successively higher levels on the legs. This is usually termed 'climbing up the legs' or Gower's sign and is most obviously seen in children (see Fig. 15.1). Gait First impressions, both visual and auditory, of a patient's gait are often of more help than formal testing (see Chapter 4). 164 Chapter 18 Posture, stance, spinal movement and gait At the end of the clinical examination, the patient, if well enough, should be made to walk in a straight line for at least 9 m, then turn and walk back to the starting point. Note the posture of the body while walking, the position and movement of the arms, the relative ease and smoothness of move ment of the legs, the distance between the feet both in forwards and lateral directions, the regularity of the movement, the ability to maintain a straight course, the ease of turning and, finally, of stopping. Abnorm alities of gait Dragging the feet The patient who drags one foot usually has an upper motor neuron lesion of that leg. If this is part of a marked hemiparesis, he will throw the whole leg outwards from the hip, producing the movement called circumduction, leaning towards the opposite side with the arm flexed across the body, but often also abducting and cir cumducting it. This is common following hemiplegia of any cause. In bilateral upper motor neuron lesions, both feet drag, the steps are slow and short, the gait is stiff-legged and the patient tends to lean forwards. Additional disability may arise when there is adductor spasm. The feet tend to cross in 'scissors' fashion, most commonly seen in the spastic diplegias of childhood. When accom panied by calf muscle contracture, there will be a tendency to walk on the toes. Tire more mobile a patient is despite this apparently crippling gait, the longer the lesion has been present. High-stepping gaits The patient raises the foot high to overcome a foot drop; the toe hits the ground first, but the patient is not ataxic. He has to flex the limb as a whole at hip and knee so that the foot will clear the ground. It is an exaggeration of the normal stepping process. This type of gait occurs unilaterally in lumbosacral root or peripheral nerve lesions causing anterior tibial muscle paralysis, and bilaterally in polyneu ropathies, cauda equina lesions and peroneal muscular atrophy. In the latter condition, this gait may be very marked but apparently disable the patient very little. A similar gait is found when, as a result of loss of position sense, the patient does not know where his foot is. The heel tends to strike the floor first, but the gait is irregular and ill-controlled, the legs move in all directions, with accompanying reeling from side to side on a broad base. The abnormality is increased in the dark and 165 Part 3 The motor system walking may be impossible if the patient doses his eyes ('sensory ataxia'). This is classically seen in tabes dorsalis, but as this is now an uncommon disease in many countries, it is more common in disorder of the posterior columns and in sensory neuropathies. A shuffling gait Movement in a series of small, flat-footed shuffles is best typified by extrapyramidal syndromes, particularly Parkinson's disease. The combined rigidity and bradykinesia causes a characteris tic posture stooped forwards with the hips and knees flexed, the steps becoming quicker as the movement progresses ('festination'). There may be difficulty in stopping, and a slight push may cause rapid forward movement ('propulsion'). Sudden changes of direc tion cannot be made. Turning is slow, moving en bloc. Sometimes the patient may become rooted to the spot, especially approaching a doorway ('threshold akinesia'). He may have to go round several times in a revolving door before being able to extricate himself. Another small-stepped shuffling gait occurs as a result of dif fuse cerebrovascular disease (typified by multi-infarct states), or alternatively as part of the syndrome of 'normal or low-pressure hydrocephalus' (NPH). The gait is irregular and hesitant, and this is the marche a petits pas. The patient may lean backwards rather than forwards, and sometimes he may make curious little dancing movements. The term 'gait apraxia' is also used to describe this type of difficulty in walking, particularly when in association with NPH syndrome. An ataxic gait The gait of cerebellar ataxia is of two types: 1 In one, the patient swings the legs unnecessarily and irregu larly, and tends to reel and sway. He looks, and often has been suspected of being, drunk (drunken gait). Although he sways to and fro, he may tend to veer to the side of the lesion, if unilateral. 2 The other is an ataxia of the trunk. The patient is grossly unsta ble, reels in any direction including backwards and may need the support of two people (staggering gait). There is little or no limb ataxia. This is seen in midline posterior fossa lesions, including tumours of the vermis and foramen magnum anoma lies with descent of the tonsils (cerebellar ectopia). Tire titubant ataxia of multiple sclerosis combines all these fea tures, together with vertical oscillation of the head, tnmk and arm, the arm oscillating on its stick as it moves it forwards and sideways 166 Chapter 18 Posture, stance, spinal movement and gait to give additional support. There may also be spasticity with both sensory and cerebellar ataxia. Tire total combination is hardly ever seen in any other condition. A waddling gait The pelvis is rotated through an abnormally large arc, accompanied by compensatory movements of the upper trunk and associated with marked lordosis. This accompanies congenital dislocation of the hips, but is also seen in myopathies, particularly muscular dys trophy. The rest of the physical examination will make differentia tion easy. Hysterical gaits These are usually, but not invariably, quite bizarre, correspond to none of the above features, vary from moment to moment, and from examination to examination, are minimized when the patient does not know he is being watched, do not cause injury, can be altered by suggestion and are of a degree that would be accompa nied by other signs of, for instance, cerebellar disease if the process had reached a stage advanced enough to produce this type of gait. The main danger is that o f thinking that the midline cerebellar disequilib rium is o f hysterical origin, and o f course the hysteria may be exaggerat ing some genuine disturbance. The gait in chorea In Sydenham's and other forms of chorea, one is not so impressed by the gait as by the movements of arm, neck and face that accom pany walking. In Huntington's chorea, however, the patient walks wide-based, lordotic, lurching from heel to heel, with variable steps, starting and stopping, and marked by associated grimacing and vigorous movements of fingers and wrist. This type of gait is often thought hysterical. In the dyskinesia of L-dopa overdosage, bizarre movements resembling those of Huntington's chorea may be seen in the limbs, trunk and neck while walking. In dystonias, the patients may walk on the outsides of their feet, though they may be able to evert them normally on the bed. M inor abnormalities In order to detect minor abnormalities of gait, particularly the atax ias, the patient should be made to walk heel-to-toe along a straight line. It should be noted whether he tends to reel off consistently in Part 3 The motor system any particular direction. He should then be made to walk round a chair first in one direction and then the other. The patient with a right cerebellar lesion will stagger to the right and when walking with a chair on his right will tend to bump into it. On reversing the procedure, he will deviate outwards from it. These tests can be made even harder by asking the patient to close his eyes. He should also attempt to hop on one foot. This is not possible on the side of a cerebellar lesion, but its performance will also be defective on the side of a pyramidal lesion, being slowed rather than unsteady. There are probably few parts of the examination in which it is more important to correlate the findings with the patient's age and to temper diagnostic enthusiasm with kindness and consideration. 168 19 Involuntary movements Analysis of involuntary movement requires intelligent use of the eyes, supplemented by a few instructions aimed at increasing, decreasing or altering the character of the abnormality. Many move ments such as shivering or startled jumping are not voluntarily pro duced, yet do not represent any disease process. At times, however, such phenomena may become so exaggerated as to pose problems in differential diagnosis, and for this reason alone, observations must be made at different times and under varying conditions. If any involuntary movement is detected at any stage in the examination, it is important to learn certain facts about it: 1 What parts of the body are affected? 2 Is it constant, or episodic in occurrence? 3 Is it present only at rest, only on movement or both? 4 Does voluntary movement increase or suppress it? 5 Is it altered by any particular position of the trunk or limbs? 6 Is it affected by environment, temperature or emotion? 7 Is it altered by eye closure? 8 Does it disappear in sleep? 9 If the patient is aware of it, can he describe its onset? 10 Is it present when the patient does not know he is being observed? Many of these points can be observed while talking to the patient. During the formal examination, spend some time in careful obser vation of the whole body and then give specific instructions to bring out the behaviour of the movements more clearly. Ask the patient first to hold his hands out in front of him, to retain them there with his eyes closed and then to hold them above his head, palms forwards. He should then grasp the examiner's hand, carry out the finger-nose test, pick up a small object such as a pin, do up or undo buttons and make other voluntary move ments requiring some degree of skill both with the affected and unaffected limbs, watching all the time the influence of the activi ties on the abnormality. He should then sit up, stand up and walk. Information should be added from the nursing staff or relatives as to the situation during sleep. The quality of peripheral tremor can be emphasized by placing a piece of paper on the outstretched fingers. As many similar movements can be differentiated only by cor relating them with other physical signs, a final decision regarding 169 Part 3 The motor system their nature must await the completion of the neurological examination. See Video clips on different kinds of involuntary movements in free companion CD-ROM. Video icon adjacent to the condition point to the video clip in the CD-ROM on the respective move ment. Epilepsy and convulsive movements Generalized tonic-clonic epilepsy A generalized attack, characteristically, is of sudden or rapid onset with loss of consciousness, rigidity of the limbs (maintained for a variable period), turning of the head and eyes and followed by clonic jerking of face, neck, arms or legs, with cyanosis, teeth clenching, tongue biting, frothing at the mouth and incontinence. The rigidity then relaxes, the jerking dies away with a few slower jerks and the patient goes into a postictal state, with stertorous breathing, its duration varying from a few moments to several hours. An attack may stop short at any stage. Not all attacks exhibit the full range of features described above. Neither tongue biting nor incontinence is invariable. Some attacks display rigidity only (tonic seizures); in others, there is no tonic stage but with immedi ate jerking (clonic seizures). Focal or partial epilepsy affects one side or one part of the body only, without necessarily any loss of consciousness. If the same part undergoes epileptic twitching for hours or days, the condition is called epilepsia partialis continua. In Salaam attacks or infantile spasms, the arms are thrust upwards and forwards, the trunk at first extended and then flexed, the move ment being followed by a cry. An electroencephalogram abnormal ity termed 'hypsarrhythmia' accompanies this form of infantile spasm (p. 315). Typical absence seizures Motor manifestations are not necessarily a feature of a petit mal attack. It may consist of no more than a few seconds of loss of attention. However, this loss of awareness may be accompanied by slight twitching of the eyelids, head nodding, a little jerking of the hands and maybe a few repetitive words. The important features of the untreated case are the great frequency of the attacks, their very 170 Chapter 19 Involuntary movements short duration and the ease with which they can be stimulated by overbreathing. Myoclonus Myoclonus is a sudden, brief, shocklike involuntary movement arising from the nervous system. The myoclonus due to contraction of muscles is known as positive myoclonus, whereas an interrup tion in a contracting muscle leading to brief loss of postural tone is known as negative myoclonus. It can also be classified as epilep tic or nonepileptic myoclonus on the basis of the association with epilepsy. The type of myoclonus is decided on the basis of the body part(s) affected or the cause that triggered the jerk. Generalized - the whole body is affected Focal or segmental - a particular body part or segment is affected Multifocal - different parts of the body are affected, but not at the same time Action - triggered by action Reflex - triggered by a stimulus such as touch, light or sound It may occur singly or twice or three times in rapid succession, usually, but not invariably, affecting the flexors of the upper limbs and the extensors of the lower limbs; they vary in degree from a contraction insufficient to move a joint to one so violent as to throw the patient to the ground. They may be provoked by touching or by sudden noise. The commonest form occurs just as a person falls asleep (hypnic jerk or physiological). The legs are usually affected, but the jerks may be more widespread and can interfere with sleep. These are, however, of quite benign significance. The commonest epileptic form usually starts in adolescents, affects the upper limbs and occurs just after waking in the morning or shortly after rising. They may herald a generalized seizure or remain as tire only epi leptic feature, but can be provoked by overbreathing, sometimes simply by eye closure, and often by photic stimulation. Myoclo nus may also be seen in a number of degenerative and infective diseases of the brain and even of the spinal cord - including the spongiform encephalopathies, particularly Creutzfeldt-Jakob dis ease, Alzheimer's disease and the very rare subacute sclerosing panencephalitis - and is not of localizing value. 171 Part 3 The motor system Asterixis or negative myoclonus This consists of a sudden loss of posture in arms held outwards with wrists dorsiflexed (muscle contraction is interrupted). The drop may be followed by a compensatory jerk, so that objects may be thrown around. It is a 'negative' myoclonus. It can be either cortical or subcortical in origin. This is seen in patients with advanced liver disease or metabolic/toxic encephalopathies. Its particular importance is that its appearance is often a warning of an impending hepatic coma. Opisthotonus Opisthotonus is a state of extreme hyperextension of the neck and spine, varying from arching of the spine to a state of rigidity so great that only the heels and vertex touch the bed. It may be perma nent or episodic, precipitated by noise or interference. Maintained opisthotonus is seen in extreme meningeal irrita tion, usually in small children, or in extreme dystonias and rigid ity, such as in the late stages of subacute encephalitis and dis orders of Neurodegeneration with Brain Iron Accumulation. If spasmodic, it occurs in tetanus, in brainstem compression from posterior fossa neoplasms, occasionally in pontine haemorrhage secondary to tentorial pressure coning and has been seen in brainstem encephalitis. It can be a hysterical manifestation, and is more likely to signify a 'pseudoseizure' than a genuine epilep tic event. Chorea F^| This condition varies both in degree and on different occasions from the severe form where the limbs are flung about in rapid movements - no two in succession being the same, often described as semipurposive, but really having no purpose at all, accompa nied by respiratory irregularity and rapid protrusion and retraction of the tongue, with 'flapping' of its tip - to minor degrees where the movements may be slight, but being brought out by voluntary movement and disorganizing it may be mistaken for a cerebel lar ataxia. The limbs are hypotonic, yet the reflexes are usually retained. The joints can be hyperextended, and bizarre postures of the fingers and wrists are adopted; if the arms are held above the head with palms forwards, this position cannot be maintained. The movements are minimal at rest and in quiet surroundings and maximal when the patient is frightened or embarrassed. Patients 172 Chapter 19 Involuntary movements T ? Common causes of chorea ^ ______________ ,______ • Sydenham's chorea: usually in children, often with cardiac abnormality and a positive antistreptolysin titre • Huntington's chorea: patients in middle life, without hypotonicity, no cardiac lesion, progressive dementia and a family history • Chorea of pregnancy or chorea gravidarum: usually in first pregnancy • Chorea in patients on oral contraceptives • Senile or arteriosclerotic choreas: in the later age groups, often of sudden onset, and with evidence of degenerative vascular disease • Chorea in primary polycythaemia • Hereditary chorea without dementia: now a recognized entity with dominant transmission, not always 'benign' V______________ J rarely injure themselves or anyone else, but may frequently drop or knock things over. It is essential to look particularly for: (i) the flapping of the tongue (Darting tongue sign); (ii) the respiratory irregularity; (iii) hypotonia and hyperextensibility; (iv) the inability to hold the hands above the head with palms extended; (v) milkmaid sign, characterized by the inability to maintain a voluntary contraction in hand grip; (vi) hung-up or pendular reflexes due to chorea interfering in the reflex contractions and, of course, (vi) any cardiac lesion (rheumatic chorea). Athetosis In contrast to chorea, this is a slow writhing movement, best seen at wrists, fingers and ankles. The fingers writhe, the wrists flex, the forearm and arm rotate inwards, abduct, and then rotate outwards in abduction. The foot is inverted. The impression is one almost of voluntary movement, and except in severe cases, most marked in a limb not at that moment being examined. The movements are absent during sleep, little altered by eye closure, increased by voluntary movement and interfering with sleep. Respiratory rhythm is normal; there is no tongue flap, but similar movements of face and tongue may occur, especially when ath etosis is bilateral. Choreoathetosis This is a combination of the above. The movements have both slow and quick elements, and either may predominate. It is rather 173 Part 3 The motor system characteristic that just as one is about to diagnose, say, athetosis, a movement typically choreic appears, and vice versa. The respira tory rhythm and tongue movements are abnormal. The movements are very apparent on voluntary effort, and straining with one limb will increase the movements greatly in the others. No cardiac lesion is present. Both athetosis and choreoathetosis commonly accompany syndromes causing mental retardation, whether resulting from perinatal trauma or anoxia and with physical neurological disability (cerebral palsy). The retardation may often be much less than the impression given by the bizarre movements. Dyskinesias Dyskinesia is a generic term to describe all the above involuntary movements, but its special importance is the frequency with which it is seen in patients receiving L-dopa treatment for parkinsonism. The choreic element is less rapid than in other choreas, and the athetoid element is of lesser amplitude. Indeed, the movements, though so obvious, are often preferred by the patient to the previ ous akinesia. The movements may be absent from a limb previously treated stereotactically. Lowering the L-dopa dosage will reduce or stop the movements at first, these tending to occur only at the time of peak action of the drug. Later, the dyskinesias increase in dura tion and can occur throughout the day. Orofacial movements are most prominent. Hemiballismus f^t This is the most dramatic of all involuntary movements. Usually affecting the proximal joints of one arm, there are wild, rapid, fling ing movements of wide radius, occurring constantly, or with short periods of freedom, sufficiently violent to injure the patient and others. They are not altered by eye closure, are absent during sleep, but prevent sleep because of their violence and may be accompa nied by increased tone and reflexes in the affected limb. This condi tion is often of sudden onset, is totally disabling and exhausting, but may lessen under observation. It is due to a lesion in the vicin ity of the subthalamic nucleus interrupting its immediate connec tions, usually vascular in origin, though occasionally due to meta static or primary neoplastic infiltration. It may sometimes follow stereotaxic operations for parkinsonism, and may require a larger lesion to cure it. 174 Chapter 19 Involuntary movements Dystonia Dystonia is currently defined as a syndrome of involuntary movements characterized by continuous and repetitive muscle contractions of agonist and antagonist muscles that are patterned and lead to twisting movements and abnormal postures. Dystonia can be accompanied by other abnormal movements like myoclonus (myoclonic dystonia) and tremors (dystonic tremor). The dystonia can be subclassified on the basis of the severity, clinical features, distribution, age of onset and aetiopathogenesis. The following classification based upon the distribution is useful to label the dystonia in a particular category on bedside testing. Focal dystonia This involves a single part of the body. Examples include blepha rospasm, oromandibular dystonia, spasmodic dysphonia and writ er's cramps. Segm ental dystonia It affects one or more contiguous body parts. Examples include idiopathic craniocervical dystonia which includes blepharospasm, oromandibular such as facial, lingual, pharyngeal and cervical dys tonia. Other types can be crural (single leg/both legs/trunk) and axial (trunk and neck). Multifocal dystonia It affects two or more noncontiguous body parts. Generalized dystonia It should have a segmental crural dystonia and dystonia in one other body part (idiopathic generalized torsion dystonia). Hemidystonia This is characterized by the involvement of half of the body, and is associated with contralateral structural lesion in basal ganglia. The severity of dystonia is affected by emotions, fatigue, motor activity and also in some cases by diurnal variation (in doparesponsive dystonia, it is absent in the morning and increased in the evening). Paroxysmal kinesogenic dystonia is another type of Part 3 The motor system fluctuating dystonia where there is sudden onset of dystonic pos turing on performing motor activity, and is not predictable. Idiopathic generalized torsion dystonia (dystonia musculorum deformans) ffij q H IP In this state, the patient's trunk and limbs undergo very forcible, wide amplitude, writhing movements, mainly at the proximal joints, with quicker, smaller movements superadded. Marked hypertrophy of the dystonic muscles may develop. The slower movements consist of arching of the back and neck, and strong rotation of the neck and arms into positions of extreme distortion, which are held at their maximum for 5-10 seconds. The more rapid movements are seen at the beginning and end of each slow movement, especially the neck turning. There is usually grimacing, grunting and protrusion of the tongue. All features are greatly increased by nervousness, are absent during total relax ation and sleep and become progressively worse as repeated attempts at voluntary movement fail. Such patients may be able to feed them selves at home, and yet in the outpatient department may be unable to grasp the examiner's hand. Intelligence may again be much higher than the superficial appearance suggests. (Intellect is, in fact, entirely preserved.) These cases can be associated with mutation in DYT1 gene. Spasmodic torticollis This consists of forced turning of the head to one side, or even backwards, with elevation of the chin and dropping of the occiput. There may be a long sustained spasm, a series of rapid spasms or short-lived spasms with intervals of normality, often accompanied by grimacing, overaction of the platysma, cracking noises from the neck and considerable pain. It is made worse by nervousness and embarrassment, and relieved by total rest. In long-standing cases, there is hypertrophy of the stemomastoid or other cervical mus cles. It may be part of a widespread dystonic condition or entirely 'focal'. Its onset is often undeniably associated with clear psychopa thology and other psychogenic history and symptoms. But a stress ful experience may trigger an underlying organic disturbance, and, here again, the 'functional' versus 'organic' argument is unhelpful. Tremors Q They are the most common form of involuntary movement. Tremor is a rhythmic oscillatory to-and-fro movement which results from alternating contractions of antagonist muscles in at least one part of 176 Chapter 19 Involuntary movements the body. They can be classified on the basis of the phenomenology (rest or action), distribution (head, trunk, voice, hand, ankle etc.), frequency (low frequency and high frequency) or aetiology. Tremors are classified on the basis of the circumstances they are seen under: Rest tremor - when the body part is fully supported against gravity Action tremor - when volitional muscles contract Postural tremor - when the arms are outstretched against grav ity in front of the body Re-emergent tremor - appears in the outstretched hand after a latency of few seconds Kinetic tremor - when a movement of finger or limb is attempted to reach a target Task-specific tremor - appears on performing a particular task such as handwriting Essential tremor W t ftn irff It is the commonest movement disorder seen in the general popu lation. Classic essential tremor is characterized predominantly by postural and action tremors that are slowly progressive over years. For diagnosis, the patient should have bilateral action tremor of the hands and forearms that is visible and persistent with no dystonic posturing. There should not be any rest tremor and other neuro logical signs with the exception of cogwheeling. The duration of symptoms is more than 3 years with a positive family history and benefit from ethanol. The patient may also have head tremors with no abnormal posture. The tremor can be tested by asking the patient to trace an Archi medes spiral or join two or three discrete points on a paper which are placed in a line. Parkinsonian tremor 4-xnrnT This is a rhythmical 4-8-H z tremor varying from the simple move ment of one thumb to a state in which so gross is the shaking of the whole limb and body that the tremor can be heard as well as seen as the extremities thump the chair or floor. In the earliest stages, the movement is seen in the tip of the thumb as a flexion-extension movement. The next stage occurs when the thumb comes into opposition with the forefinger, which itself produces 177 Part 3 The motor system a flexion tremor at the first interphalangeal joint. As the tremor advances, the fingers are moved en masse, rather than individ ually, and the flexed position becomes permanent at rest. The tremor is absent in total relaxation, or in sleep, and often imme diately after waking. It is increased by emotion, and is present at rest providing some form of posture is being maintained and usually suppressed by the initiation of voluntary movements. It will return as soon as that movement becomes maintained or a new position taken up. It is accompanied by cogwheel rigidity at the wrist and fingers and slowing of each movement. Masking of the face may be obvious even when the thumb alone is involved. Occasional patients with Parkinson's disease show an exaggera tion of the tremor with movement. In other words, there are fea tures of 'essential' tremor. Red nucleus tremor This tremor, usually unilateral as in Benedikt's syndrome, is slow, coarse and rhythmical, present at rest and also throughout a volun tary movement and, by virtue of its site of origin, usually associ ated with ataxia on that side. Rubral tremor, so-called, is really a severe cerebellar tremor with the responsible lesion more likely in the superior cerebellar peduncles. (The frequency is 2.5-4.0 Hz.) Tremor in Wilson's disease ('wing beating' tremor) Characteristically, on holding the hands outstretched, this tremor is seen as an irregular, vertical, flapping movement at the wrist, with large amplitude movements from the shoulder, asymmetrically on the two sides. It is grossly increased by further voluntary move ment, and may then become a wild, incoordinate, choreic flinging of the arm in all directions. It is absent on total relaxation, so that the patient may be able to conceal it, but it appears as soon as any attempt is made to maintain posture. The youth of the patient, the family history, increasing dementia and, of course, the pathogno monic Kayser-Fleischer ring confirm the diagnosis, and evidence of liver disease usually being present only at a late stage. Perioral tremor A constant, coarse tremor of the orbicularis oris and chin is seen in the general paralysis of the insane, and may be the only site of the tremor. 178 Chapter 19 Involuntary movements Titubation This is a vertical oscillation of the head, present when the head is maintained in an upright position, and therefore seen when the patient sits or stands, and disappears on lying down. It is indicative of disease of the cerebellar connections, and is seen in multiple scle rosis and some variants of ataxias and essential tremor. It differs from the isolated head nodding forming part of the torticollis spec trum, and is always accompanied by marked signs of other neuro logical disease, although there is an idiopathic variety associated with the so-called essential tremor. Enhanced physiological tremor Anxiety and nervousness The tremor is rapid, varying from fine to coarse, affecting mainly the fingers, but capable of spreading to the whole arm or body. It is present at rest, increased by any voluntary movement which the patient fears he may not do correctly or quickly enough (such as the finger-nose test, or undressing), not increased during auto matic movement, made worse by speaking sharply to the patient, reducing towards the end of the examination as he realizes that it is not a very alarming experience and often absent on a second examination for the same reason. A 'physiological' tremor is pres ent whenever a muscle contracts (due to subtetanic contractions of motor units). This is not visible, but becomes so when 'enhanced' by (3-adrenergic stimulation of the segmental stretch reflex. The fre quency is fast, 10-15 Hz, and this is the tremor of nervousness. Thyrotoxicosis This is a fine rapid tremor, present constantly, greatly influenced by emotion and accompanied by sweating and tachycardia, but the extremities are very warm; there is lid retraction, possibly exoph thalmos, and loss of weight. Other toxic trem ors Almost any drug taken in excess over a prolonged period may give rise to a tremor that resembles alcoholic tremor. Inorganic mercury compounds are particularly liable to produce coarse tremor, but are rarely a problem these days. Addiction to stimulant drugs, includ ing some of those used as antidepressants, is now more commonly responsible than industrial toxins. The tremor caused by with 179 Part 3 The motor system drawal from alcohol and opiates is an 'enhanced physiological' tremor again. Lithium, nicotine and L-dopa are further examples of agents that can induce this form of tremor. Toluene abuse ('sol vent encephalopathy') is a recent problem, producing an acute ill ness with coma, ataxia, behavioural disturbance and convulsions. Withdrawal from such 'glue-sniffing' can provoke tremor, and an intention tremor occurs during intoxication along with other 'cer ebellar' signs. Pseudoathetosis or 'sensory wandering' This is an important physical sign, and is seen when a patient lies with his eyes closed and his hands either held outstretched in front of him or resting on a flat surface such as a bed table. Slow wandering movements of the fingers occur, accompanied by flexion at the metacarpophalangeal joints so that the palm is drawn away from a flat surface. As this happens, the fingers gradually close, the wrist flexes and there is internal rotation of the pronated forearm. In many ways, the movements resemble athetosis, but it is suppressed when the patient watches his hand, and he is usually unaware of its occurrence. It is always accompa nied by gross loss o f postural sensibility. Occasionally, the toes, or even the legs, may show similar involuntary movements. When such a limb is moved passively, the examiner gains the impression (incorrectly) that the patient is resisting voluntarily. Any lesion causing very severe loss of position sense may produce this phe nomenon. When tabes dorsalis was a common cause for loss of position sense, these movements were called 'tabetic athetosis', but nowadays it is most common in those cases of cervical spon dylosis where the spinal canal is very narrow, so that backward displacement of the cord results in the compression of its poste rior aspect. In this condition, it may affect only the arms, but it is also seen in carcinomatous sensory neuropathy where it may affect the arms and legs equally. It occurs with cerebellar ectopia at foramen magnum level and very occasionally, unilaterally, in parietal lobe lesions. It can occur, but it is rare, in multiple sclero sis, and here it may remit completely. Other involuntary movements of the face and neck Almost any of the movements described may involve the face and neck, but in certain conditions, the abnormality is limited to this part of the body. 180 Chapter 19 Involuntary movements Facial tics Generally, tics are stereotyped, repetitive movements which are easily produced voluntarily, such as blinking, screwing up the face and pursing the lips. They are present when under observation, sometimes absent when concentrating on something else, common in childhood, increased by nervousness, and though they may remain stereotyped for months or years, singly or occasionally, multiple tics may persist or even evolve throughout life. Volun tary suppression is possible for a while, but increasing inner urge ultimately causes their return, with associated momentary relief from tension. Though commonest in the face, such movements frequently involve the shoulder girdle, causing shrugging move ments, retraction of the neck and, at times, contraction of individ ual muscles such as the platysma, the pectorals or even one-half of the abdominal muscles. The whole muscle is always in action and the movement is in every respect similar to voluntary contrac tions of those muscles. A rare disorder, the syndrome of Gilles de la Tourette, is characterized by multiple persistent tics often accom panied by inarticulate cries or barks or compulsive utterance of obscenities. Hemifacial spasm In this condition, the muscles in one part of the face go into spas modic contraction, drawing the mouth towards that side, with a series of fine twitches after it is drawn up. The muscles around the eye are similarly involved. Each spasm starts suddenly and stops suddenly, is very embarrassing, made worse by nervousness, stopped by concentration or by sleep, is always the same in type if not in degree and after some years may be accompanied by facial weakness. The aetiology is uncertain, but irritation of the facial nerve by an aberrant arterial loop, aneurysm or acoustic neuroma can be responsible. A somewhat similar twitching is seen long after a facial palsy, but the facial muscles show contracture already, the spasm is initiated by voluntary movement even though very slight and fasciculation is often present at rest. Focal facial fits may simulate facial spasm, but the contrac tions are much slower and coarser and, though the onset may be similar, they do not cease suddenly, but through a series of lessening twitches, and, of course, are frequently accompanied or followed by further manifestations of focal or generalized epilepsy. 181 Part 3 The motor system Facial myokimia This differs from myokimia in the orbicularis muscles in being of sudden onset, affecting the whole of one side of the face by undu lating flickering waves of contraction on a background of constant contraction. It may occur bilaterally, is of self-limiting course and has been seen in vascular lesions of the stem, multiple sclerosis and pontine gliomata. Short runs of lesser degree affecting only a small portion of the facial musculature are not very uncommon and have the same benign significance as periorbital myokimia. Facial dystonia and tardive dyskinesia Bizarre grimacing of the face, associated with intermittent protrusion of an apparently hypertrophied tongue, occurs certainly in general ized dystonic states, but also as an isolated phenomenon. This may be seen in some varieties of Huntington's chorea, and in the Gilles de la Tourette syndrome, where involuntary noises or utterances may occur. Facial (cranial) dystonia otherwise forms into a pattern known as Meige's or Brueghel's syndrome. There is usually a combination of blepharospasm and oromandibular dystonia. They may some times occur singly but are usually in combination. It is a condition of middle age, and can be associated with spasmodic torticollis. The movements are distinguished from drug-induced orofacial dyskine sia (see below) by their more sustained and spasmodic nature. Tardive dyskinesia is a phenomenon not uncommon after pro longed heavy dosage of phenothiazine-type drugs, particularly in females. It is seen more commonly with increasing age. A dyskine sia of the face (orofacial) may be isolated from generalized dyskine sia, and occurs during treatment of parkinsonism by L-dopa, being relieved by lowering the dose, but possibly recurring on progres sively smaller dosage. R/Iovements U n ited to the m u scles If there is tremor, the muscles of that limb will show this move ment. It is therefore essential to look at the limbs as a whole when any abnormal movement is noted in one of its muscles. There are, however, certain movements confined to the muscles. Fasciculation This term, by common clinical usage, is applied to an irregu lar, non-rhythmical contraction of muscle fascicles, the result 182 Chapter 19 Involuntary movements of random firing of motor units; it is sometimes fine, sometimes coarse, most easily seen in the larger muscles such as deltoid or calves, and present at rest, stopping during, but increased after, voluntary movement, stimulated by smacking or warming the muscle, but usually not felt by the patient. When spontaneously noticed by the patient, commonly in arms or calves, unaccompa nied by other signs, then it is benign and often related to muscle fatigue or anxiety. Otherwise, it varies greatly in degree and may be so coarse and gross as to be visible through the clothes, when the whole muscle seems alive and like a bag of worms. Fasciculation is a better word than fibrillation, which is too fine a movement to be seen, though it can be measured on the electromyograph. It indicates a lesion of the lower motor neuron, usually degeneration of the anterior horn cell, or irritation of the anterior root, when the fasciculation is very coarse, and repetitive in muscles having the same root supply. The finer fasciculation is seen classically in motor neuron disease, where it normally is associated with wasting, but exaggerated reflexes, and is almost diagnostic of this disease when affecting the tongue and accompanied by an exaggerated jaw jerk. It may also be seen in the tongue in syringobulbia, but the jaw jerk is then absent. It occurs also in muscles recovering from poliomy elitis; in calves after lumbar disc lesions or laminectomies which have caused much arachnoiditis. If seen in wasted muscles with absent reflexes, thyrotoxic myopathy, syphilitic amyotrophy or a polyradiculopathy must be considered. Widespread fasciculation can be produced in non-myasthenic individuals by an injection of 2.5 mg of neostigmine, sometimes after edrophonium (Tensilon), ■ and in the unaffected muscles in some myasthenics. In pathological states, a much smaller dose will greatly increase any fasciculation already present. (See Chapter 36 for electromyographic definition.) Myokymia This, the most common involuntary movement of the muscles, is seen in two forms. 1 As a fine, very rapid rippling of muscle fibres persisting in the same group o f fibres for minutes at a time, most commonly in the orbicularis oculis, and easily felt by both the patient and the observer. If a fold of the skin below the eye is held between the thumb and forefinger, a sensation is experienced similar to touching a purring kitten. It is usually a manifestation of fatigue, but is common in anxiety states. 2 The second type is a much coarser contraction of bundles of muscle fibres, again both visible and palpable, and though Part 3 The motor system usually not moving a limb, if near enough to a joint is strong enough to do so. This is common in the outer aspect of the thigh or upper arm, but occurs in any muscle, including the pectorals and intercostals. It is not due to organic disease, but is common in fatigue. This is synonymous with 'benign fasciculation' (see also facial myokimia, p. 183). Shivering This is quite frequently seen in the muscles of patients who are cold or nervous, without the rest of the limb or body being involved. It may, therefore, be mistaken for fasciculation. Its rapid, regular movement of the whole muscle, its appearance in a series of bursts and its cure by warmth and correcting the other simple causative factors distinguishes it from more serious conditions. Clonus It is possible for individual muscle bundles to go into clonus with out any active measure having been taken to elicit it arid without movement of the limb as a whole. It is entirely regular, occurs only in a hypertonic limb with exaggerated reflexes, is present when that muscle is in some degree of tension and is stopped immediately by altering the position of the limb so that the muscle is in relaxation. Do not assume that abnormal movements are of psychogenic origin if they disappear when not under direct observation. Most involuntary movements are least obvious on complete relaxation, a state rarely achieved during physical examination. 184 Part 4 The Sensory System 2 © Basic principles for examination of sensation There is probably nothing more frustrating and fatiguing than the detailed examination of sensation, particularly in a patient who is so unreliable that answers are too variable to be of value, or in one so determined to report minor differences that the end result is equally confusing. The complete sensory examination may not be desirable on one session. It is often wise to carry out sensory examination in several stages, testing different parts of the body, or different modalities, on different occasions. Whichever modality is being examined, try to form a mental picture both of the pathway that that particular sensation fol lows from the site of stimulation to appreciation in consciousness and the relationship of that pathway^to^neighbouring nervous structures. This entails a little anatomical knowledge, but though there are many varieties of sensation, long practice has taught the value of studying those forms whose tests are simple and whose anatomical pathways are well established. It is also important to bear in mind that sensory loss is of far greater importance in some conditions than in others, e.g. in sus pected syshvgoftvyelia, or in searching for the level of a lesion caus ing a paraplegia, or in delineating a peripheral nerve lesion, exact assessment is vital. In a hemiplegia, however, the detailed analysis of superficial sensory loss rarely adds much information of value. To find significant and important sensory signs in the absence of appropriate symptoms is very rare. The modalities of sensation to be tested 1 Pain, light touch and temperature. These are the exteroceptive sensations derived from sources outside the body. 2 Sense of position, passive movement, vibration and deep pain. These are the proprioceptive sensations derived from the body itself. 3 Stereognosis, graphaesthesia and two-point discrimination. These are the combined and cortical sensations. Visceral or interoceptive sensation is a fourth group, but is rarely examined as a clinical bedside routine. In fact, the word 'interoceptive' is unknown to most examiners. 187 Part 4 The sensory system Essential features of the sensory pathways In the following paragraphs, a very complex subject is oversim plified in order to state briefly some anatomical facts that form the basis for the localization of lesions producing sensory distur bances. 1 All form s o f sensation must travel via a peripheral nerve and a sensory root to the spinal cord or, for cranial nerves, the brain stem. A nerve or root lesion will, therefore, cause loss of all forms of sensation from the area that it supplies. 2 Fibres serving pain and temperature sensation enter the posterolat eral aspect of the spinal cord, travel upwards a few segments and then cross to the opposite anterolateral spinothalamic tract. A superficial cord lesion will cause loss of these sensations on tire opposite side of the body, or a central cord lesion on both sides but over a limited area. This tract ascends to the brain stem, where it lies lateral to the medial lemniscus, and is joined by the quintothalamic tract (p. 86) in the pons. These fibres pass dorsal to the red nucleus and end in the ventrolateral nucleus of the thalamus. A lesion here will cause loss of sensation through out the whole of the opposite side of the body. It is, however, now thought that many pain fibres end in the reticular forma tion of the brainstem. From the thalamus, sensory impulses pass through the posterior limb of the internal capsule and the thal amoparietal radiations to the post-Rolandic cortex, but lesions at cortical level cause little disturbance of pain and temperature. 3 Fibres carrying the sense o f light touch ascend the posterior col umns of the spinal cord on the same side as they enter, as far as tire nuclei gracilis and cuneatus, and further fibres then cross the midline to ascend the brainstem in the medial lemniscus, where they are joined by touch fibres from the face. They then pass to the thalamus and on to the post-Rolandic cortex. Other elements of touch, however, on entering the cord, ascend sev eral segments, and then relay across the mid line to follow the course of the spinothalamic tract in its anterior portion. It is for this reason that some cord lesions, especially central lesions, affect pain and temperature but not light touch. 4 The fibres carrying sense o f position, o f passive movement and o f vibration ascend the cord in the posterior columns on the same side, as far as the nuclei gracilis and cuneatus, then syn apse, decussate and form the medial lemniscus, continuing as described above. A posteriorly situated cord lesion will cause loss of these sensations below the lesion on the same side - this applies to 5 also. 188 Chapter 20 Basic principles for examination of sensation 5 Stereognostic and discriminative sensations follow the same pathway as proprioceptive sensation, but they should not be considered as separate tracts, for stereognosis depends on the sense of touch and position, and two-point discrimination on the sense of touch. 6 Facial sensation is described in Chapter 9. A rrangem ent o f th e sensory fibres 1 In the posterior columns: Fibres from the lower part of the body are displaced medially as more fibres enter. 2 In the spinothalamic tract: Fibres from the lower part of the body are displaced to lie superficially to those from the upper part. 3 In the thalamus: Fibres from the lower part of the body lie later ally to those from the trunk and arms, fibres from the face lying most medially of all. ‘ 4 In the sensory cortex: Fibres from the lower limbs terminate near the superior longitudinal fissure, and those from the face in the lower part of the post-Rolandic gyrus. The hand and mouth occupy a very much larger area in relation to size than other parts of the body. Sen sory derm atom es A visual image must constantly be borne in mind of the spinal segmental supply to each area under stimulation, the shape and extent of the sensory dermatome in which it lies and which of the peripheral nerves supplies it. Avoid testing at the fringes of these dermatomes because con siderable overlap occurs. Unfortunately, such patterns can only be learnt by heart and, though not necessarily the same in different textbooks, Fig. 20.1 gives a practical workable scheme. Doctors, more than most, use mnemonics to help memory. In this context, it is better to memorize certain key facts, and this applies to the following notes regarding sensory dermatomes, because if some facts are indelibly imprinted on one's mind, the others can be deduced. 1 Tire patient is always considered to be standing with the palms of the hands facing forwards. 2 C l gives no supply to the skin, the occiput being supplied by C2. 3 C5 supplies the outer aspect of the shoulder tip. 4 C7 (the longest cervical spinous process) supplies the middle finger (the longest finger). 189 Part 4 The sensory system Greater , occipital nerve Lesser C2 Greater auricular nerve Anterior cutaneous nerve of neck t3 Supraclavicular nerves X , ----------- Axillary nerve it Post. A I N cut. • rami | Lat. of JCUt. .r fv thor. •a \ nerves mi W t V -----------Radial nerve Med. cutaneous nerve of forearm I Poshj rami o.f lumbar, sacral i zoccygeal nerves',. S3 Med. cutaneous nerve of arm and intercostobrachial nerve Musculocutaneous nerve Iliohypo gastric nei Radial nerve Median nerve N Ulnar nerve S5 Perforating cutaneous nerve Lat. cutaneous nerve of thigh S2 Obturator nerve Posterior cutaneous nerve of thigh Femoral nerve Lateral popliteal nerve Femoral nerve lL! Lateral popliteal nerve L4- Medial popliteal nerve S1 Calcanean branches of sural and tibial Nerves Fig. 20.1 Outlines of the sensory segmental dermatomes and average areas of peripheral nerve supply. Individual variability is considerable. (Redrawn from Wolf, H.G. & Wolf, S. (1958) Pain, courtesy of Charles C. Thomas, Illinois.) 190 Chapter 20 Basic principles for examination of sensation Great auricular nerve Anterior cutaneous nerve of neck Supraclavicular Nerves Axillary nerve Med. cutaneous nerve of arm and intercostobrachial nerve Radial nerve Med. cutaneous nerve of forearm Musculocutaneous nerve Iliohypogastric nerve Ulnar nerve Radial nerve Median nerve Femoral nerve Dorsal nerve of penis Perineal nerve Obturator nerve Lat. cutaneous nerve of thigh Fermoral nerve Lateral popliteal nerve Medial popliteal nerve Posterior tibial nerve Fig. 20.1 (continued) 191 Part 4 The sensory system 5 T3 dermatome lies in the axilla. 6 T8, T10 and T12 supply the rib margin, the umbilicus and the pubis respectively. 7 L3 dermatome lies at the knee. L5 runs diagonally from the outer aspect of the tibia to the inner aspect of the foot. 8 SI includes the little toe. 9 S3, S4, S5 are in concentric rings around the anus. Skin areas supplied by peripheral nerves are more difficult to remember (Fig. 20.1). Fortunately, though any nerve may be damaged, comparatively few are injured or diseased, with any degree of frequency. These are the Vth cranial nerve, the circum flex branch of the axillary nerve (see Fig. 24.1), the radial, median and ulnar nerves (see Fig. 24.2), the femoral nerve, the lateral cuta neous nerve of the thigh, the sciatic nerve and its branches, the lateral popliteal (common peroneal) and the anterior tibial nerves (see Fig. 24.1). The o b jectiv es of sensory te s ts 1 To demonstrate clearly and consistently the limits of any areas of abnormal sensation. 2 To determine which modalities are involved within those limits. 3 To compare the findings with known patterns of abnormal sen sation. 0 Tire more experienced one becomes, the more one learns to recognize the type of case in which sensory impairment is a vital diagnostic feature and must be minutely examined; and conversely the case in which, even if present, its importance is minimal and barely justifies a tedious analysis. • Tire less experienced must complete the examination, but must also ask himself 'is this apparent sensonj impairment rel evant to the diagnostic problem, or might it be a red herring drawn across the path by too minute attention to detail'? • For instance, in a spastic hyperreflexic paraparesis, spend ing a lot of time over a very vague and inconsistent sensory level may be completely wasteful if one has not appreciated that the jaw jerk is grossly exaggerated so that the lesion must therefore be above the midbrain stem, and spinal sensory lev els are irrelevant. Sensory examination may be time-consuming. It is important to pose a clear question and tailor the sensory examination to answer it. For example, the questions may be as follows: 1 Is there a sensonj loss at all (e.g. in patients with muscle or ante rior horn cell disease)? This can be answered by testing a patient 192 Chapter 20 Basic principles for examination of sensation with eyes closed using a wisp of cotton to detect any loss of superficial touch sensation and/or by testing with a pin to detect whether the patient can distinguish between its sharp and blunt end. 2 Is there a sensory level and if yes, at what spinal segment? The exam iner should move a pin or cotton from the spot of impaired sensation to that of the normal sensation, asking the patient to immediately report when it becomes normal. 3 Is there a graded sensory loss? Tire examiner should first establish that there is definite sensory loss (as above in Q l), and then ask the patient to compare normal (taken as 100%) with abnormal areas. hr the following chapter, each modality of sensation is considered in turn and advice is given on the methods of testing most likely to produce the clear, consistent results without which this part of the examination is pointless. 193 21 Pain, touch and temperature Pain Modern investigative procedures have to a large extent supplanted the very detailed sensory testing considered so important in years gone by. But such procedures are not necessarily available to the first person who examines a patient. All sensory tests are open to faulty observation, by both the patient and the examiner. To try and produce uniform or graduated stimuli while test ing pain, numerous pieces of apparatus have been suggested, but in practice the simplest remains the best, and a sharp pin with a rounded head will serve most purposes. The following are note worthy. 1 The shaft of the pin should be long enough to allow the exam iner's index finger and thumb to slide downwards on impact. 2 A short pin held with the index finger on the head of the pin usually produces an indelicate and variable stimulus. 3 A gentle, but firm and uniform touch is required. 4 Puncturing the skin is not only heavy-handed and most unlikely to produce an accurate response from an alarmed patient, but does of course risk transmitting disease. Hollow needles should never be used. Preliminary screening 1 First choose a part of the patient's body that, from the history, is expected to be normal and touch him precisely, but not too firmly, several times with the point of the pin. 2 Ask him (a) if he can feel anything (b) what it is that he can feel (c) if he says that he can feel a point ask if it is sharp or blunt Remember that a single prick may not always register pain. Having established that the patient recognizes the stimulus, compare quickly the appreciation of the sensation in a number of areas, including the face, the shoulders, the inner and outer aspects of the lower forearms, the thumb and little finger, the upper and lower chest and abdomen, the front of the thighs, the lateral and medial aspects of the lower legs, the dorsum of the feet, the little toe and the buttocks. Bearing in mind the aide-memoire in the previous 194 Chapter 21 Pain, touch and temperature chapter, this will give a very good general idea of the overall pat tern of sensory defect. From the history, attention may have been drawn to areas likely to be abnormal. The next stage, therefore, whether the preliminary screening revealed any abnormality or not, is to test these areas particularly and in the same way. M o r e detailed analysis If possible, it is wise to postpone closer analysis to a later examination. ° One should now go at once to the area previously roughly defined as abnormal, and, moving the pin from the centre of this area, ask the patient to say immediately where the sensation changes and to describe the type of change. ° Mark this point and change the direction of the pin, always mov ing from impaired sensation to normal sensation, for the change is easier to detect. Do not stimulate too rapidly or the slowly react ing patient may still be referring to the last prick but one. Avoid asking 'Is it sharp' and 'Is this sharper than that?', for a nervous patient may agree with something he does not really mean. A patient may also truthfully answer 'Can you feel that?' by say ing 'Yes' even though no pain is produced. It is quite sensible to ask if there is any difference between different areas, and then what the difference is. Touch Practically, all the previous remarks apply equally to the examina tion of light touch. Again, many methods are used, but a small piece o f cotton zvool answers most needs, does not cause sufficient pres sure to stimulate deep sensibility and produces a sensation familiar to the patient. After similar preliminary screening, tell the patient to shut his eyes and to say 'Yes' each time he feels anything. The cot ton wool is shaped to a point and the skin is then touched lightly, but not too lightly, testing again in dermatome areas, and map ping out abnormalities more clearly later by the methods already described. Remember that a 'dab' with cotton wool rather than a 'stroke or tickle' is a more reliable test for pure tactile sensation. A light touch with a fingertip is perfectly acceptable. Temperature Testing temperature sensation is not required in ordinary clinical practice: it provides no more useful information than testing pain. Part 4 The sensory system Nevertheless, it can provide a colourful adjunct to teaching by case demonstration. 1 For preliminary screening, the patient can compare the temper ature of a cold object such as a tuning fork on the main sensory areas of the body. 2 After this, use test tubes containing hot water (44°C), and cold water (30°C). Extremes of heat and cold should not be used, because these stimulate pain fibres, but the temperatures must be maintained throughout testing. The same principles again apply. The patient has the eyes closed, is first asked what he can feel, whether there is any difference when the other tube is used and what that difference is. A note on sensory levels In the cord, the spinal segments are not necessarily at the level of the vertebra bearing the same number. In the highest cervical region, they do correspond; in the low cervical region, there is one segment difference, i.e. C8 spinal segment is opposite C7 vertebra. In upper thoracic regions, the difference is of two segments, in midthoracic, three, and the lumbar and sacral segments are all opposite T l l - L l vertebrae. In cord lesions, there may be a clear-cut upper level of sensory abnormality, defined by a zone of hyperaesthesia. Remember always to test for a lower level also, because sacral sparing is com mon. When testing for a sensory level by moving the pin from lower to higher spinal segments, there is a danger of error. If one passes from one skin area to an adjacent area supplied by a much higher spinal segment, at the point of segmental change the patient thinks there has been a sudden increase in the intensity of the stimulus. This is easily shown on the chest, where T3, T4 and C4 are adjacent, around the upper arm (T2-C5) and the thigh (S2-L2). Unless one is aware of it, a false impression may be gained of an abnormal sen sory level. Try it on yourself - it is quite normal, and has been called the summation o f sensory stimuli. Difficulties and fallacies These are all 'subjective' tests; one is at the patient's mercy to some extent and it is difficult to give strictly comparable stimuli. No appa ratus has found permanent favour over those described above, but care, concentration and patience are the essential qualities for both the examiner and the patient. 196 Chapter 21 Pain, touch and temperature Remember that the pulp of the fingers is relatively insensitive to pinprick, but very sensitive to touch. Stimulate with the pin just proximal to the nail. Simulation of sensory impairment is usually fairly easy to detect. Tire sensory loss does not correspond to known anatomical pat terns; it is often confined to the whole of one limb; variability of margins and suggestibility are frequent and the disability that such a severe sensory loss would produce is not apparent. Turning the patient over and repeating the tests may reverse the side affected. A sudden, painful stimulation in the analgesic area while talking about something else may produce a definite reaction, if only mus cle contraction, a blink or dilatation of the pupils, yet it is enough to show that the stimulus has been appreciated. When testing touch, the well-tried method of asking the patient, whose eyes are closed, to say 'Yes' each time he feels a touch, and 'No' each time he does not, still works in a high proportion of the less-alert patients. Abnormal sensory patterns are described in Chapter 24. 197 22 The proprioceptive sensations Position sense and sense of passive movement These two closely related sensations are examined together. Where most people refer to tests of position sense, or joint sense, they are usually, in fact, referring to the sense of passive movement. Position sense The patient's eyes should be closed throughout the examination. 1 Place the patient's arm in a particular position, then move it away and ask him first to replace it himself, and then to place the opposite limb in a similar position. 2 Ask him to touch the forefinger of one hand with the forefinger of the other, and make it harder by moving his finger to differ ent positions. 3 Let him try adopting similar positions with his legs, and ask him to raise one leg to touch his own outstretched hand with Iris big toe. 4 Ask him to place his forefinger accurately on the tip of his nose, and his heel accurately on his knee (see Chapter 26). Sense of passive movement Tire patient's eyes must still be closed. Tire digit (thumb, finger or big toe) is held firmly and moved up and down, while the patient is asked if he can feel any movement. If so, he should be told that he is going to be asked whether Iris thumb (or toe) has been moved upwards or downwards. Move the digit widely in tire appropriate directions so that he understands which movement he is to call 'up' and which 'down'. This apparently elementary instruction is very necessary. • Hold the sides of the digit between the finger and thumb, so that uneven pressure above or below does not reveal the direc tion of the movement, and make a clear and precise movement in one or other direction. • Repeat the test several times, avoiding alternate movements, and if any error is made, the test should be continued until at least six successive correct responses are given, or until one is satisfied that the defect is constant. 198 Chapter 22 The proprioceptive sensations 9 If digit movement could not be detected in the first place, the same test is carried out at the wrist, elbow and knee. 9 If one suspects a very minor defect, the test can be varied by asking the patient to say 'Now' at the moment he first feels the toe moving. The movement is then made slowly, but precisely, and a remarkably slight degree of displacement is normally detected immediately. Defects of the sense of passive move ment may be so gross that the patient has no knowledge of the wide displacement at the shoulder or hip. 9 Subjective awareness o f posterior column deficit. It is common for a patient with posterior column defect to complain of a 'numb ness' in the affected limb - a numbness that cannot be demon strated objectively as loss of touch, pain or temperature. Difficulties and fallacies The most frequent difficulties are of one's own making, when the test becomes so automatic that the patient is expected to know exactly what he has to do without preliminary explanation. Wag gling the toe vigorously and then suddenly moving it in one direc tion may be satisfactory with an intelligent and cooperative patient, but merely confuses the less alert, his responses and the physician. The movement should always be clear and precise. Some patients lapse blissfully into a rhythmical, alternate repeti tion of 'up' and 'down'. If one draws the patient's attention to this by saying firmly 'Now, think hard, I am not just going to move it up and down, up and down', a simple measure such as this, surpris ingly enough, will almost invariably overcome the difficulty, and it is rarely necessary to record the findings as 'too unreliable to be of value'. However, the patient who consistently and with unerring accuracy places his finger 2 cm to one side of his nose during the finger-nose test is unlikely to have a genuine postural loss, particu larly if he does the same thing when the eyes are open. A patient who briskly and invariably gives an answer opposite to the actual movement made need not necessarily be simulating a non-existent fault. He may quite genuinely have misunderstood the directions - hence the importance of the preliminary demon stration, which may need to be repeated. Vibration sense A tuning fork (128C0 or 256C1) of well-maintained vibration is shown to the patient and then placed on his clavicle to allow him to identify the sensation of vibration. 199 Part 4 The sensory system • He then closes his eyes, the fork is struck and placed on bony points, starting peripherally at the internal malleolus and the lower end of the radius. • If there is gross deficiency here, it can then be placed on the tibial tuberosity and the elbow, the anterior superior iliac spine and the clavicle or the ribs. • Placing it in turn on the spinous processes o f the vertebrae and mov ing upwards until it is appreciated may, on rare occasions, give the only clear sensory level o f a posteriorly situated spinal tumour. The patient is first asked if he can feel the vibration. Do not say 'Can you feel th at? because he probably will, in fact, be able to feel the fork. Then ask him to say immediately when the vibration stops. The fork is stopped by touching it and the speed with which he recognizes this is noted. The two sides are now compared, first by asking if the degree of vibration feels the same, and then by comparing the promptness with which he notes the cessation of vibration. Next, allow the fork to rim down by itself, asking the patient to say when he can feel it no longer. Move it then quickly to the other limb, where normally vibration will still be detectable for 3-5 seconds. In minor degrees of abnormality, it is not detected when transferred from normal to abnormal side, and persists lon ger than usual when moved from abnormal to normal side. Some experts note the length of time for which the vibration is felt at various sites and compare with the normal values for age or the examiner. Difficulties and fallacies Always test the vibration of a strange fork first; it may be barely perceptible. A bony point must be used. On soft tissue, it may be quite imper ceptible. In fat people, it may be difficult to find any bony points, but in oedematous legs, the oedema can be compressed away before starting. Unless the test is clearly explained, many patients think it is the sound that they are meant to detect and they may well hear that when vibration sense is lost. Do not confuse the issue by using the word 'humming'. In increasing age, vibration sense diminishes steadily and over the age of 65 some deficit at the ankles is common. In diabetes, it is It is a wise neurologist who strikes his fork on his percussion hammer or other resilient material, for constant striking on his own knee is capable of producing effusion in the joint. 200 Chapter 22 The proprioceptive sensations reduced at a much earlier age, even in the absence of other signs of a polyneuropathy. Muscle sensitivity To assess the degree of discomfort produced, the thumbs are firmly pressed into the muscles of the forearm and calves. Normally, by the time one has to strain the hand muscles a little, the patient will complain of discomfort. If the muscles are abnormally tender, there will be obvious distress under lighter pressure and the pain will continue for a few seconds after release. In states of diminished sensitivity, the patient will allow all possible force to be exerted without complaint. Squeezing the tendon Achilles between the finger and thumb will allow the diminished deep sensation to be assessed. Increased muscle tenderness is found in some polyneuropathies, subacute combined degeneration of the spinal cord, myositis and in some psychogenic states. Diminished tenderness occurs in tabes dorsalis, syringomyelia, carcinomatous neuropathy and in lesions of the posterior roots and root entry zones of the spinal cord. 201 23 Stereognosis, discriminative sense and graphaesthesia Stereognosis This is the ability to recognize an object purely from the feel of its shape and size. Test objects must be familiar, easily identifiable and large enough for a weak hand to feel. The patient should close his eyes and the object is placed first into the hand suspected of abnor mality and he is asked to identify it. If he fails, or takes a long time to decide, it should then be placed in the other hand and compari son made of both the accuracy and the speed of response. Abnormalities It will usually be obvious at once whether the patient recognizes the object or not. If stereognosis is defective, the normal skilled movements of exploring an unknown object are absent. The hand moves as a whole instead of as a series of small joints. Often it is evident that the patient is not aware that anything has been placed in the hand and may perhaps allow it to fall out, and yet go on apparently trying to feel it. Stereognosis is defective if touch and position sense are defec tive and may, therefore, be defective in severe lesions of the sen sory pathway at any point. Its particular value in localization arises when other forms of sensation are normal or only slightly affected, yet there is astereognosis. The lesion responsible then lies in the parietal lobe. Difficulties and fallacies A very weak hand may be unable to make the movements required to feel the object properly. In this case, the patient's fingers must be held closed over it. A hand that is partly paralysed, oedematous or deformed by some process such as rheumatoid arthritis may be unable to close over a small object and may give a false impression unless a large enough object is used. As stated above, severe loss of sensation will include astere ognosis, so that a parietal lobe lesion must not be automatically diagnosed. Stereognostic sensation travels in the posterior columns, 202 Chapter 23 Stereognosis, discriminative sense and graphaesthesia which means a cord lesion can produce it. It is a finding that must be taken in conjunction with the rest of the examination. The 'dumb hands' syndrome of marked cervical spondylosis in the elderly is the commonest cause nowadays. Tw o -p o in t discrimination This is the ability to detect that a stimulus consists of two blunt points when they are simultaneously applied. • Explain the test to the patient first, and illustrate the sensation for him by touching his finger with the two points widely sepa rated. • He must then close his eyes and the pulp of the finger is touched firmly with either one point or two, starting with them far apart, and approximating them until he begins to make errors. • The threshold is thus determined and the other hand can be compared. Tine same test can be carried out on the feet, prefera bly stimulating the dorsum of the foot. Avoid areas of calloused skin both on hands and feet. • The normal ability to distinguish the two points from one varies in different parts of the body. On the fingers, it should be pos sible at less than 5 mm separation, but on the dorsum of the foot, separation of even 5 cm may be required and still be normal. Two-point discrimination depends on the integrity of light touch, but if this is only slightly deficient or normal, and no signs of severe posterior column disease are present, then impairment of two-point discrimination indicates a parietal lobe lesion. Difficulties and fallacies The usual cause of faulty interpretation, however, comes as a result of forgetting how different the discriminative threshold is in dif ferent parts of the body, and expecting the foot to be as sensitive as the hand. Graphaesthesia 1 This is the ability to recognize letters or numbers written on the skin with a blunt point. 2 The patient closes his eyes and letters or numerals are traced out on the palm of the hand or the anterior aspect of the fore arm, thigh or lower leg. 3 Clear figures such as 8 ,4 and 5 should be used first and, if cor rect, the more difficult 6 ,9 arid 3 can be used as finer tests. 203 Part 4 The sensory system If peripheral sensation is lost, graphaesthesia will be absent, but if peripheral sensation is normal, the absence of graphaesthesia is then a sign of a parietal cortical lesion. Localization of touch The patient closes his eyes and then is touched on some point of the body with the examiner's finger or a pin. He is asked to indicate the point touched with his own forefinger. The significance of the test is the same as for graphaesthesia and two-

point discrimination. Other sensory abnormalities associated with parietal lobe disease are described in Chapter 30. A pair of compasses can be used for two-point discrimination test. These are useless unless the points have been well blunted, for one must not stimulate the pain fibres. 204 24 Common patterns of abnormal sensation No two patients are exactly the same. Their size, their faces, their eyes and the colour of their skin varies, and one might expect their neuroanatomy to vary similarly. It is, however, quite extraordinary how constant, within only a slight range, are the signs on the body of a lesion of the sensory pathways. This of course is of utmost value to that fundamental initial stage in diagnosis - the location of the lesion. In addition, this reproducibility is vital in detecting a sensory abnormality that is genuine, because sensation is after all a purely subjective phenomenon and alleged sensory loss is com mon, but anatomical knowledge is not! The most common patterns of sensory abnormality are now illus trated, remembering that though anatomical pathways may not vary, lesions do vary greatly, so that the intensity of sensory abnor mality may range from total loss to very slight reduction, or even hypersensitivity, and it is in these latter cases that great experience is needed for judgement of the significance of any abnormality that may be found. Total unilateral loss of all forms of sensation (Fig. 24.1(a)) This indicates an extensive lesion of the thalamus or its immediate neighbourhood. 'Pure sensory stroke' from a lacunar infarct in the thalamus or internal capsule is an example. If all forms of sensa tion are truly lost, there must be gross disability. For instance, total postural loss is incompatible with normal use of a limb, and if this is claimed, and the limb used normally, the sensory loss must be non-organic. One must, however, be wary of the late result of a long-standing thalamic lesion (such as a vascular episode) when motor function may have returned, but sensory diminution, not total loss, may remain. Unilateral loss confined to sensation a W exteroceptive This can be caused by a partial lesion of the thalamus, or a lesion laterally situated in the upper brainstem. Associated signs, such as 205 Part 4 The sensory system motor impairment in thalamic lesions, or cranial nerve palsies, or perhaps red nuclear tremor if in the midbrain, are likely. When no other signs are present, this also is often a hysterical sign, but the sensory impairment then usually stops exactly at the midline or some way over it, instead of a short distance from it, on the normal side. The hysteric may allege that he cannot feel the vibration of a tuning fork on the anaesthetic side of his skull or sternum; bone conduction makes this impossible on an organic basis. Indeed, the tuning fork placed exactly in the midline may provoke the response that one side of the fork is vibrating but not the other! Unilateral loss confined to all proprioceptive sensation This can be caused by a partial lesion of the thalamus, or a lesion medially situated in the upper brainstem. It is a very uncommon finding and grossly disabling. Unilateral loss of position sense and cortical sensation, with disturbance of light touch and the quality of pain This indicates a parietal lobe lesion between the thalamus and the cortex. As the lesion becomes more superficial, so it would have to be increasingly large to affect all the diverging fibres. Super ficial lesions, therefore, tend to affect one localized area only. Responses are characteristically variable from one moment to the next. Unilateral hyperalgesia and hyperaesthesia This commonly follows partial lesions of the thalamus. It is usually of vascular origin and often associated with thalamic pain. Loss of pain and temperature on one side of the face and the opposite side of the body (Fig. 24.1(b)) This indicates a lesion of the medulla affecting the descending root of the Vth nerve, and the ascending spinothalamic tract from the rest of the body. This 'lateral medullary syndrome' used to be thought due to thrombosis of the posterior inferior cerebellar artery, but is more 206 Chapter 24 Common patterns of abnormal sensation f ig. 24.1 (a) Total hemianalgesia - thalamic, or upper brainstem lesion, (b) Lateral medullary lesion (usually vertebral artery deficiency)- (c) Transverse lesion of the cord, (d) Brown-Sequard lesion - hemisection of the cord, (e) Cuirasse analgesia - central cord lesion, (f) Sacral sparing - cord compression, often extrinsic, (g) Saddle analgesia - cauda equina or conus medullaris lesion. (h) G love and stocking analgesia - polyneuropathy, developing cervical cord lesion or hysteria. 207 Part 4 The sensory system Upper cervical nerves Circumflex Lateral cutaneous of forearm Medial cutaneous of forearm Combined sciatic and posterior cutaneous Vth cranial nerve Total brachial plexus High radial Posterior cutaneous Femoral Sciatic (i) Fig. 24.1 (continued) (i) Average areas of sensory loss resulting from the more common peripheral nerve lesions. Variation is considerable and in incomplete lesions the area involved may be greatly reduced. often due to atheroma of a vertebral artery and particularly likely to occur if there is, as occurs not too infrequently, a major difference between the size of the two vertebral arteries. Bilateral loss of all forms of sensation below a definite level (Fig. 24.1(c)) This indicates a gross lesion of the spinal cord. The upper level of the lesion may be indicated by a zone of hyperaesthesia and it is the upper level of this that should be taken as showing the highest spinal segment affected. Note that this segment is not necessarily at the same level as the equivalent vertebra (see Chapter 21, p. 196). If the upper level is vague and there is no zone of hyperaesthesia, the actual level of the lesions may be many segments higher than the sensory level suggests. See the note on sacral sparing below. 208 Lower roots of brachial plexus Lateral cutaneous of thigh Lateral popliteal Anterior tibial Chapter 24 Common patterns of abnormal sensation If pain and temperature only are affected, it will be the anterior aspect only of the cord that is involved. Unilateral loss of pain and temperature sensation below a definite level (Fig. 24.1(d)) This indicates a partial unilateral lesion of the spinal cord. The Brown-Sequard syndrome of hemisection of the cord consists of ipsilateral motor and proprioceptive impairment and contralateral loss of pain and temperature, while at the highest level on the side of the lesion there is a thin band of analgesia representing involve ment of the root entry zone. It is more common, however, for com pression injury or demyelination to produce incomplete examples of this syndrome. Impairment of pain and temperature sensation over several segments with normal sensation above and below (Fig. 24.1(e)) This indicates an intrinsic lesion of the cord, placed near its cen tre and so involving the crossing fibres. If placed more posteriorly than anteriorly, proprioceptive sense may be lost over a similarly limited area. It is a common lesion in syringomyelia and intrinsic cord tumours. Loss of sensation over many segments (e.g. T6-S2) with normal sacral sensation is, however, common in extrinsic cord compression also (Fig. 24.1(f)). Loss of sensation of 'saddle' type (Fig. 24.1(g)) This is the description given to impairment of sensation over the lowest sacral segments. If affecting all forms of sensation, accom panied by loss of leg reflexes and sphincter control, it indicates a major lesion of the cauda equina. If touch is preserved, the lesion is in or near the conus medullaris, in which case the plantar reflexes may be extensor and the knee jerks may be retained. Glove and stocking anaesthesia (Fig. 24.1(h)) Peripheral loss of sensation affecting both hands and both feet is common in polyneuropathy of any cause. In such cases, there is normally impairment of reflexes as well, and this type of sensory defect with no other signs or symptoms, or with symptoms relating to some totally different part of the body, is often hysterical. There cm dorcrp't' cut-oit' just’below the elbow and knee joints. 209 Part 4 The sensory system Genuine 'glove and stock' distribution sensory abnormality is also common in incomplete cervical cord lesions, either multiple sclero sis or early compression. Loss of all forms of sensation over a clearly defined area in one part of the body only (Figs. 24.1(i) and 24.2) This could be due to a lesion of a sensory root or of a peripheral nerve. To differentiate, comparison must be made with the known sensory dermatomes and peripheral nerve distributions (Fig. 20.1). Few peripheral nerve lesions are purely sensory, while pure poste rior root lesions will be. Quite apart from common disorders (such as diabetes), poly neuropathies - and one must include multiple isolated peripheral nerve lesions (mononeuritis multiplex) - are seen in the collagen diseases, especially, as regards the latter, in polyarteritis nodosa, but these days they must also be carefully looked for in patients on one of the many cytotoxic preparations, because they may repre sent an early toxic manifestation. Loss of vibration sense alone If affecting the lower limbs, this is common in intrinsic cord lesions such as those produced by multiple sclerosis and syringomyelia, and has led some to suggest that vibration sensation has a differ ent pathway from other proprioceptive sensations. Impairment is common in old age. Fig. 24.2 Area supply of (a) radial, (b) m edian and (c) ulnar nerves. In each, the shaded area represents the average areas of loss to touch, the lines the degrees of variation and dashed that are common. Pain is often lost over less than the smaller area of supply. 210 Chapter 24 Common patterns of abnormal sensation Loss of position and vibration sense alone This indicates a lesion of the posterior columns which is often dif ficult to localize. It is common in tabes dorsalis, subacute combined degeneration, Friedreich's ataxia, carcinomatous neuropathy and certain toxic states such as those due to organic mercurial com pounds. If lost only below a definite level, there may be compression of the posterior part of the cord. If the arms are affected to a much greater extent than the legs, and asymmetrically, the lesion may be due to a combination of cervical spondylosis and a very narrow vertebral canal; or to foramen magnum lesions, especially with ton sillar prolapse (see also p. 166). Miscellaneous features Patchy areas o f’loss cfi ■ yei ration.heJnw a certain level are common in intrinsic lesions of the spinal cord secondary to such conditions as cervical spondylosis. They are also found during recovery from a Brown-Sequard lesion. Patches of sensory loss irregularly scat tered throughout the body occur in chronic polyneuropathies, and in leprosy. Patches on the face, the forearms, the lower legs and bands of abnormality on the trunk occur in tabes. Intercostal mono neuropathies are not uncommon in diabetes. Except in defining a mononeuritis multiplex, the exact charting of very vague patches of sensory loss rarely gives clinical information of a value comparable to the time taken to complete the task. It is the overall picture and pattern that needs to be assessed. Areas of hypersensitivity, if corresponding to the segmental der matomes, indicate lesions of the posterior root or the root entry zone of the cord. If these areas correspond with the distribution of the peripheral nerve, they usually mean an incomplete or recover ing lesion of the nerve. Very transient, very localized areas of hyperaesthesia are not uncommon in normal people; their aetiology is uncertain but they do not represent a disease process.

Part 5 The Motor-Sensory Links 25 The reflexes Any reflex action requires a stimulus, a sensory pathway, a link with a motor unit, a motor neuron and, filially, a contractile or other effector element. Any breach in this arc results in an absent reflex. Most reflex arcs are also influenced by higher centres, and where tnere'is a’Dreacn'in'tne patnways'rrom tnese centres, tne’Denaviour of the reflex is fundamentally changed. Purpose of the examination No matter which reflex is being examined, one must determine: 1 Whether the reflex is present or absent. 2 If present, whether it is normal, or showing signs that influ ences from higher centres are defective. 3 If absent, whether the arc is breached on the motor or sensory side. 4 Whether any abnormalities are unilateral, bilateral, affecting all reflexes, or whether a definite level can be detected in the ner vous system at which abnormalities first appear, because reflex 'levels' may be as helpful as sensory levels. TSie tendon reflexes Any skeletal muscle will contract reflexly when suddenly stretched, the tendons merely being convenient structures with which to pro duce this stretching. Certain muscles are, however, easy of access and by long experience their normal and abnormal responses have been defined. For clarity of results, there are certain requisites: 1 A good percussion hammer (p. 18). 2 An examiner with flexible wrists who allows the weight of the hammer to decide the strength of the blow. 3 A patient who is warm, comfortable and relaxed. 4 A muscle placed in the optimum position, slightly on stretch, but with plenty of room for contraction. 5 A constant mental picture of the segmental and peripheral nerve supply of each muscle as its reflex is being tested. 215 Part 5 The motor-sensory links The jaw jerk This is described under the Vth cranial nerve (see Chapter 9). It rep resents one of the most helpful tests in finding a 'level' of reflex abnormality. The upper limbs If possible, the patient should at first be partially propped up, the elbows slightly flexed, the hands lying loosely across the abdomen and the fingers just not touching (Figs. 25.1 and 25.2). In this posi tion, the biceps and supinator jerks can be tested. The biceps jerk (Fig. 25.1) Technique: Press the forefinger gently on the biceps tendon in the antecubital fossa and then strike the finger with the ham m er. Normal result: Flexion of the elbow and visible contraction of the biceps muscle. Segmental innervations: C5 Peripheral nerve: Musculocutaneous The supinator jerk (Fig. 25.2) , J' ^ ^ * * h m e < X Technique: Strike the lower end of the radius about 5 cm above the wrist. Watch the movement of the forearm and fingers. Normal result: Contraction of the brachioradialis and flexion of the elbow. The biceps often contract as well and slight flexion of the fingers may occur (see p. 224 for the inverted supinator reflex) Segmental innervation: C5, C6 Peripheral nerve: Radial The triceps jerk (Fig. 25.3) Technique: By holding the patient's hand, draw the arm across the trunk and allow it to lie loosely in the new position. Then strike the triceps tendon 5 cm above the elbow. If there is no response, repeat two or three times either side of the original point. Normal result: Extension of the elbow and visible contraction of the triceps Segmental innervation: C6, C7 Peripheral nerve: Radial 216 Chapter 25 The reflexes Fig. 25.1 The biceps jerk. Fig. 25.2 The supinator jerk. I ig. 25.3 The triceps jerk. 217 Part 5 The motor-sensory links Fig. 25.4 The finger flexion reflex. The pectoral reflexes T echnique: With the patient's arm in midposition between adduc tion and abduction, hook your index finger on the tendon of the pectoralis major muscle in the anterior fold of axilla and strike with the hammer. N orm al resu lt: Adduction of the arm and visible contraction of the pectoralis major. S eg m en tal in n erv ation : C5-T1 P erip h eral n erv es: Lateral and medial pectoral The finger flexion reflex (Fig. 25.4) T echn iqu e : Allow the patient's hand to rest palm upwards, the fin gers slightly flexed. The examiner gently interlocks his fingers with the patient's and strikes them with the hammer. N orm al resu lt: Slight flexion of all fingers and of the interphalangeal joint of the thumb. S eg m en tal in n erv ation : C6-T1 P erip h eral n erve: Median The lower limbs The patient should lie in the same position as described for the upper limbs, but care should be taken to see that the legs do not extend beyond the foot of the couch. The knee jerk (Fig. 25.5) T echn iqu e: The eye should not be too high above the patient's knees, and so if the bed is too low the examiner may have to kneel. To 218 Chapter 25 The reflexes compare the two sides, the left arm (for a right-handed examiner) is placed under both knees in order to flex them together. If the patient holds the legs rigidly, instruct him to lie back and let his heels fall on the bed. This often produces complete relaxation. The patella tendon can then be struck lightly on each side, increasing the strength if there is no response. A light tap is best for compar ing the two sides. Watch the movements of the lower leg and of the quadriceps muscle. Normal result: Extension of the knee and visible contraction of the quadriceps. Segmental innervation: L3, L4 Peripheral nerve: Femoral Alternative methods Place the finger just above the patella and, while the legs are extended, strike it in a peripheral direction. The quadriceps con tract and the patella will be pulled upwards. This method detects slight differences between the two sides and can be used when the lower leg has been amputated, or the patella removed. Another well-known method is to seat the patient on the edge of the couch so that the legs are dangling. This may produce a reflex in unrelaxed patients, but often the rigidity persists. It will, however, clearly show the pendular character of the reflexes in cerebellar dis ease. The ankle jerk (Fig. 25.6) Technique: This reflex appears to present technical difficulties, and is frequently recorded as absent when correct technique produces 219 Part 5 The motor-sensory links a perfectly normal response. The patient's leg should be externally rotated and slightly flexed at the knee. The examiner uses the left hand to dorsiflex the foot (Fig. 25.6(a)). For the left leg, the examiner moves to the other side of the bed (Fig. 25.6(b)). The Achilles tendon is then struck and both the movement of the foot and the contraction of the calf muscles are observed. Normal result: Plantar flexion of the foot and contraction of the gas trocnemius Segmental innervation: SI Peripheral nerve: Medial popliteal (b) 220 I i;4- 25.6 The ankle jerk. Chapter 25 The reflexes Alternative method A patient who is fit enough can kneel up on a cushioned or blan keted chair, with his back to the examiner and the feet projecting over the edge. The Achilles tendon can then be struck from above, and it is very easy to compare the responses on the two sides. Examioisitiioin) with reintforcemeinill If a tendon reflex appears reduced or absent, this need not necessar ily be of pathological significance. Such a situation is often found in patients involuntarily tensing themselves, and in those who are very relaxed, such as a happy young child, and perhaps rather sur prisingly in very muscular individuals. If other muscles are placed under strain, it often becomes possible to obtain a normal reflex. For the upper limbs, the patient should clench his teeth tightly, or while one arm is being examined he should clench the fist of the other. For the lower limbs, these measures can still be used, but the well-tried method of Jendrassik is more reliable. The patient inter locks the flexed fingers of the two hands and pulls one against the other at the moment the reflex is stimulated. Whatever the method used, the patient must make the move ment at the moment that the reflex is tested and relax afterwards. O ther allied reflexes The Hoffman reflex (Fig. 2 5 .7) The terminal phalanx of the patient's middle finger is flicked downwards between the examiner's finger and thumb. In states of hyperreflexia, organic or emotional, the tips of the other fingers Fig. 25.7 Testing the Hoffman reflex. 221! Part 5 The motor-sensory links flex and the thumb flexes and adducts. It is finding it on one side only that is useful because this can sometimes be an early sign of unilateral pyramidal tract disease. VUartenberg's sign (Fig. 25.8) The patient supinates his hand, slightly flexing the fingers. The examiner pronates his hand and links his similarly flexed fingers with the patient's. Both then flex their fingers further against each other's resistance. Normally, the thumb extends, though the termi nal phalanx may flex slightly. In pyramidal tract lesions, the thumb adducts and flexes strongly. Unfortunately, this is not a constant sign, but if present on one side only, it can indicate an early stage of pyramidal tract disease. IRossolimo's reflex The patient lies supine with the leg extended and the foot partially dorsiflexed. The ball of the foot is then struck with the hammer and in hypertonic states there is a brisk contraction of all toes. The same reflex can be obtained by flicking the toes in an upwards direction. Its significance is the same as the finger flexion reflex in the upper limb. Abnorm alities of the tendon reflexes Only by repeated examination of reflexes can sufficient experience be gained to judge a particular response as normal or abnormal. It is therefore very important, as in examination of the fundus, for tendon reflexes to be examined in all patients, not only those i ig. 25.S Wartenberg' sign; normal result. Note the extension of the patient's thumb. Chapter 25 The reflexes suspected of neurological disease. Gross abnormalities are easy to recognize, but when deviations from normal are slight, it is neces sary to re-examine several times before a decision is reached. Exaggeration 1 A reflex may be excessively brisk, the movement being a sudden, short-lived jerk. Sometimes its amplitude may be very small and unless both limb and muscle are watched, it may be thought to be reduced. This type of reflex is seen in lesions of the upper motor neurons, and in clinical practice is generally taken to be one of the manifestations of a lesion of the pyramidal system. Similar reflexes occur in agitation, fright, anxiety and after vio lent exercise, but they return to normal on rest and relaxation, and there are no other features of pyramidal lesions. 2 The reflexogenic zone may be increased. For example, if the biceps reflex is exaggerated, it may even be obtained by tapping over the clavicle due to the increase in the reflexogenic zone. 3 A reflex may be clonic. The muscle that has been stretched goes into clonic contractions until the stretch is relieved. This is most commonly seen in the knee and ankle jerks and some times in the finger jerk. It indicates a marked degree of reflex excitability and usually means pyramidal system disease, although unsustained clonic beats are common enough in tense individuals. Reduction or absence Reflexes appreciably reduced or absent in normal individuals become normal on reinforcement. Pathologically, reduction occurs under the following circumstances: 1 When there is a breach in any part of the reflex arc, e.g. lesions of the sensory nerve (polyneuropathy), the sensory root (tabes dorsalis), the anterior horn cell (poliomyelitis), the anterior root (spinal compression), the peripheral motor nerve (trauma), the terminal nerve endings (polyneuropathy) or the muscle itself (myopathies, periodic paralysis). There are, of course, many other possible disorders at these various sites. 2 ha the state of the so-called cerebral or spinal shock which occurs immediately after a severe cerebral catastrophe or spinal injury. Spasticity and reflex overactivity may be delayed for hours, days or weeks. An extensor plantar response often appears long before the tendon reflexes return. 3 When great rigidity, spasticity or muscle contracture more or less splint the joints so that movement cannot occur. Advanced Part 5 The motor-sensory links spastic paraplegia and severe extrapyramidal rigidity usually cause this situation, but it can be found in normal individu als completely unable to relax. In the latter, re-examination at another, less anxious, time usually produces satisfactory reflexes. An ankylosed joint may not move, but the muscle whose tendon is being tapped will show the contraction. Tw o im portant combinations of reflex abnormality Th e inverted deep tendon jerks When the supinator jerk is tested, there is often a slight flexion of the fingers. This may become very obvious when all the reflexes of the upper limb are exaggerated. If, however, finger flexion is the only response, contraction of the brachioradialis and elbow flexion being absent, then the reflex is said to be 'inverted'. In practice, this is usually associated with an absent biceps jerk, and exaggeration of the finger and triceps jerks. The difference between the reflex activity of the biceps and triceps may be so marked that striking the biceps tendon produces extension of the elbow (inverted biceps jerk), and striking the triceps tendon produces flexion of the elbow (inverted triceps jerk). This response indicates damage to the affer ent arc of the triceps reflex, as in lesions of the seventh and eighth cervical segments, particularly in presence of lesions of adjacent pyramidal tract. Inverted biceps jerk has the same significance as the inverted supinator jerk (see below). The inverted supinator reflex indicates a cord lesion at the fourth or fifth cervical level, causing a lower motor neuron lesion of C5 and an upper motor neuron lesion of reflexes innervated below this level. It is very common in cervical disc disease, syringomyelia, cer vical trauma and sometimes in cervical neoplasms. It is invaluable in localizing lesions responsible for a spastic paraparesis which have no sensory abnormality or clear-cut level. This applies par ticularly to cervical spondylosis. Lesions of the conus medullaris Classically, the ankle jerks only are absent in lesions of the conus medullaris. In practice, as these lesions frequently extend into the conus from higher in the cord, there may however be loss of both knee and ankle jerks, but, at the same time, an extensor plantar response. This is associated with symmetrical dissociated saddle anaesthesia, spontaneous segmental sweating in the sacral seg ments and loss of bladder and rectal control. In combined upper and lower motor neuron lesions, such as that occur in subacute combined degeneration of the spinal cord, or Chapter 25 The reflexes taboparesis, a similar combination of reflex changes may be found, but there is not the characteristic segmental sensory loss and loss of sphincter control. Other abnormalities of deep tendon reflexes include 1 Pendular jerk. In cerebellar lesions, particularly if there is some element of upper motor neuron disease as well, the reflex is not so much a brisk one as a large one - the amplitude being greater, the speed reduced, hi certain positions of the limb, it may sway back beyond its starting point and oscillate in a pendular fash ion. Normally, the number of oscillations is three. Three to five is borderline and more than five is considered abnormal. 2 Hung-up reflex o f hypothyroidism. The movement is retarded, especially during delayed relaxation, so that the impression of a slightly slow-motion film is obtained, the reflex appearing to 'hang-up' compared with a normal jerk. 3 'Hung' reflex' as in chorea. When the patellar tendon is tapped while the foot is hanging free, the knee is held in extension for a few seconds before relaxing. This is probably due to the super imposed choreic movement over the contracting quadriceps. Special situations Demonstrating asymmetry of deep tendon reflexes. Sometimes, the vital clinical question is whether the deep tendon reflexes are asymmetrical (compressive versus degenerative cause), and the above examination is imclear. In this situation, midline deep ten don reflexes may help reveal the asymmetry. They are: 1 Sternal reflex. With both forearms relaxing over the abdomen, place your index finger over the manubrium sterni and tap with the hammer. Normally, there is either no response or symmetri cal movement flexion of both the forearms with and without finger flexion. Any asymmetry will be clearly visible. 2 Symphysis pubis reflex. With the patient supine, abdominal mus cles relaxed and thighs in mild flexion, abduction and external rotation tap over the symphysis pubis. When there is spasticity, you will observe adduction of both legs. Any asymmetry will be clearly visible. Difficulties and fallacies Failure of relaxation is the major difficulty in examining tendon reflexes. Re-examination is wise just before leaving the patient, because, knowing that it is all over, he will be in a greater state of relaxation than at any other time, providing the techniques used have been gentle and sympathetic. Part 5 The motor-sensory links Faulty positioning of the limb, especially for the ankle jerk, is another cause of difficulty and advice has been given at some length on this point. Many patients make a confusing, semi-voluntary jerk of the limb shortly after the reflex has occurred. Pointing out to the patient that it is he who is doing it, that it is confusing and bluntly asking him to stop it seems to be effective. Arthritic patients often fear that the test will be painful, hold themselves rigid and jump violently when the tendon is struck. Start with a very gentle tap and gradually work up to the normal strength of stimulus. Always watch the muscle itself as well as the limb movement. When the biceps reflex is absent, on striking the tendon the elbow may appear to flex, but a glance at the biceps itself will show no contraction at all. The jaw jerk is often thought to be of little importance, but its value is, in fact, very great (see p. 89). In amputees, the reflex muscular contraction may be normal, but the amount of limb to be moved is so small that there is apparently a very brisk response. In a normal patient with a below-the-knee amputation, the stump may even go into a form of clonus. No apology is made for repeating that clonus is only a manifesta tion of heightened muscle tone, and it may occur in very tense or frightened individuals, after straining, or after exercise. It is, how ever, rarely long-sustained as in pyramidal system disease, is usu ally equal on the two sides and truly clonic reflexes are not obtained. Th e superficial reflexes For practical purposes, only four of the many superficial reflexes are routinely tested - the abdominal reflexes, the cremasteric reflexes, the anal reflex and the all-important plantar reflexes. The abdominal reflexes (including the ep ig astric reflex) Technique: The patient should first lie flat. Palpate the abdomen gently to assess the degree of relaxation and the sensitivity of the skin. Then explain that something is about to be drawn across the stomach, illustrating the manoeuvre on the chest. Any physician who has unexpectedly had his abdominal reflexes examined will appreciate the value of this warning. Lightly stroke the abdomen with a pencil, key or two-point discriminator, from without inwards, stimulating each of the Chapter 25 The reflexes four quadrants of the abdomen and the lower margins of the thoracic cage in turn. However, if the objective is segmental localization, then the stimulus may be applied along the derma tomes. Normal result: The muscles in the quadrant stimulated contract and the umbilicus moves in that direction. Segmental innervation: Epigastric T7-T9; upper abdominals T9-T11; lower abdominals T il, T12 (sometimes also LI). Abnormal responses 1 Exaggerated abdominal reflexes occur in psychoneurosis, often in the absence of overt anxiety, and may also be brisk in simple nervousness. 2 Absent abdominal reflexes may be due to: (a) Defects of techniqu^ rela^St^eft or^qjps^j^afionQr (b) A breach of the appropriate reflex arc, due to lesions such as herpes zoster, or surgical operations which have damaged the peripheral nerves or the muscle itself. (c) Pyramidal system lesions above the upper level of segmen tal innervation. This is more common in spinal lesions (ipsilateral or bilateral) than in cerebral lesions (contralateral), but the reflexes are not necessarily absent in all such cases, or in one case on all occasions. 3 Easily fatigued reflexes, ha the young, this may be a sign of early pyramidal disease. 4 An inverted response. Occasionally, when the abdominal reflexes are absent on one side, stimulation on that side produces mus cular contraction on the normal side, so that the umbilical devi ation takes place towards the normal side. Difficulties and fallacies If the patient has a very fat abdomen, one scarred by enthusiastic surgeons, or exhausted by frequent pregnancies, muscular contrac tion may not be visible or may not occur. Slight contraction of the external oblique may still be visible, and stimulation near to the ribs and inguinal ligaments often produces a local response not vis ible elsewhere. If a patient has difficulty in relaxing, it may be possible to obtain these reflexes by testing them with the patient sitting on the edge of the bed, or even standing upright. The stimulus should be light and never unpleasant. Pins should not be used. An abdomen resembling a problem in Euclid carries the trade mark of a bad examiner. Part 5 The motor-sensory links Heavy stimulation may produce a deep abdominal reflex, which, being a tendon reflex, is often exaggerated when the superficial reflexes are absent. A big fallacy is a belief that abdominal reflexes are rarely retained in multiple sclerosis. They are, indeed, often absent, but their presence in no way invalidates the diagnosis, and they may persist in the advanced stages of the disease. The cremasteric reflexes Technique: The upper inner part of the thigh is stroked in a down wards and inwards direction, the patient lying down or standing up. Watch the movement of the scrotum and testicle. Normal result: The contraction of the cremasteric muscle pulls up the scrotum and testicle on the side examined. Segmental innervation: LI, L2 Abnormal responses Exaggeration has no special importance. The reflex is often absent in: 1 Elderly men and those who have hydroceles, or have had scro tal operations. 2 Any breach of the reflex arc, including impaired sensation over the skin of the thigh. 3 Pyramidal tract disease, but there is the same inconstancy of response shown by the abdominal reflexes. The anal reflex Technique: Lightly scratch the perianal skin. Normal result: Contraction of the external sphincter Segmental innervation: S4, S5 The plantar reflex This is the most important reflex in the body, and yet the most fre quently misinterpreted. Technique: Many methods are recommended; even more are used. If the following instructions are observed, many of the difficulties are overcome. Start by positioning the patient so that the knee is slightly flexed, and the thigh externally rotated. The outer aspect of the foot should now rest on the couch as for testing the ankle jerk. Warn the patient Chapter 25 The reflexes that the sole of the foot will be scratched and ask him to try to let the foot remain loose. The outer aspect of the sole is then firmly stroked with a blunt point such as a key, the end of the percussion hammer handle or the two-point discriminator. The stimulator should move forwards and then curve inwards towards the middle metatarsophalangeal joint (Fig. 25.9). The stimulus must be firm, but not frankly painful. A pin should not be used, nor should the stimulus be a tickle, for this produces abnormal responses in nor mal individuals. Do the stimulation slowly and allow yourself time to see what is happening. Watch the first movement of the metatarsophalangeal joint of the great toe, which in this position is immediately visible. Watch also the movement and behaviour of the other toes. The test should now be repeated with the knee in extension, and in case of doubt the knee may even be pressed downwards. In this new position, early abnormalities may be detected which disappear when the knee is flexed. The two methods serve as a check on each other. Normal result: Normally, no matter what its shape may be, the great toe will flex at the metatarsophalangeal joint, even if the terminal joint appears to extend. At the same time, the other toes will flex and close together. The true reflex movement normally does not start when the stimulus is started, but when the instrument is about one-third of the way along the foot, or sometimes not until the end of the movement. Abnormal responses No response at all usually indicates that the patient is not relaxed or is very cold. Sometimes total loss of sensation interferes with Fig. 25.9 The plantar r-eflex. The arrows in d icate the d irectio n taken by the instrum ent. Part 5 The motor-sensory links the sensory side of the reflex arc, or total paralysis of the muscles makes any movement impossible. The Babinski response The Babinski response consists of extension of the great toe at the metatarsophalangeal joint, and usually the interphalangeal joint as well, while the other toes open out in a fanwise manner and are dorsiflexed. The movement is usually a slow one, but may be jerky and repeated. This may particularly happen if the stimulus is ended in a quick flick. If only the great toe extends, this may be a true extensor response; if the whole foot extends, it is often a voluntary movement. In advanced cases, the whole foot dorsiflexes, the knee and hip flex and the contraction of the muscles responsible can be felt. In cases where the toe does not move, there may be palpable and visible contraction of the tensor fas ciae latae. The Babinski response indicates the disturbance of the function of the pyramidal system, a fact on which there is both clinical and experimental agreement. It does not necessarily mean a structural lesion of the pyramidal tract and may, for instance, be found in coma from almost any cause. In advanced pyramidal disease, this response may be obtained from stimulating over a wide area, even as high as the thigh. Difficulties and fallacies The main difficulties arise from failure of relaxation, and from vol untary movements made as a result of the unpleasant sensation. The preliminary warning and avoiding the use of traumatic stimuli are the first steps in overcoming this problem. Some patients, however, are truly unable to tolerate the sensa tion. Stimulating the outer side of the foot, even slightly on the dor sum, causes less distress, or the stimulator may be placed on the sole and pressed inwards, then drawn slowly forwards without altering the pressure. This will often allow the response to be seen before the patient withdraws his foot. If, during the test, the patient takes a deep breath, attention is distracted enough to allow a few moments of relaxation. In marked pes cavus, it is difficult to assess the movement of the big toe. At times the movement may be frankly extensor, but this need not be a sign of a new pathological process. The toe may be so retracted as to appear to be in the extensor position to start with and if the terminal joint only is observed, a further extension may be quite misleading. Always watch the metatarsophalangeal joint. 230 Chapter 25 The reflexes Deformities may prevent any movement of the big toe at all. This applies particularly to a surgically corrected hallux valgus. In such cases, the type of movement of the other toes must be observed. Complete paralysis of the extensors of the toes may make Babinski's response impossible. This is only rarely of importance, how ever, because such total paralysis is likely to be of lower motor neuron origin. The contraction of the tensor fasciae latae must be observed. The plantar response is normally extensor below the age of 6 months, and this may persist up to 12 months in about 75% of children. After this, however, it should be flexor. A Babinski response is an extensor plantar response. 'Babinski flexor' is a contradiction of terms. Several other methods of obtaining the plantar reflex have been described. The three most commonly used are: 1 The Oppenheim reflex. A firm stroke with the finger and thumb down either side of the anterior border of the tibia, greater pres sure being applied to the medial side. 2 The Gordon reflex. A hard squeeze of the calf muscles. 3 The Chaddock reflex. A light stroke below the external malleolus, on the outer side of the foot. Each has the same significance as the Babinski response, but each is less reliable. These methods can, however, be applied to: (i) patients whose soles are extremely sensitive; (ii) some patients whose cooperation is poor and (iii) children, while their attention is directed elsewhere. Other important reflexes The grasp reflex First place the first and second fingers of one hand between the thumb and forefinger of the patient's hand and try to draw them lightly away. Then place these fingers on the palm of the patient's hand and move them peripherally towards the tips of the fingers. If a grasp reflex is present, the fingers will be held tightly by the hand in the first instance, the hold increasing as the effort to draw away is increased. In the second instance, the patient's fingers will flex strongly. This reflex is normal in babies under 4 months. It persists in men tal deficiency and birth injuries; reappears in tumours and vascular accidents in the premotor cortex, particularly on the medial sur face of the brain; and in lesions of the corpus callosum, probably due to neighbourhood involvement of the medial frontal lobes. It 231 Part 5 The motor-sensory links is not infrequently found in very diffuse lesions of the brain when it is difficult to say which part is responsible, so that bilateral grasp reflexes are not of great localizing value. In practice, they are most likely to be found in neurodegenerative disorders with cortical or subcortical atrophy: Alzheimer's disease, forms of parkinsonism and multi-infarct states. They can occur in 'normal ageing'. The embarrassing situation of being unable to extract one's hand (embarrassing also to a conscious patient) can be overcome by mas saging the ulnar aspect of the dorsum of the patient's hand, when the grasp will be released. Tlhe forced] giropninig re fle x Lightly and repeatedly touching the side of the palm results in the hand following the stimulus and perhaps attempting to grasp it. This is usually demonstrated in states of lowered consciousness and severe mental defect due to widespread cerebral lesions and is not of great localizing value. Tlhe syckirag re fle x Touching the corner of the mouth produces a sucking movement of the lips and deviation of the mouth in the direction of the stimu lus. This is seen in advanced and diffuse cerebral atrophic lesions, and in states of stupor from widespread encephalitic or traumatic lesions. It is normally present in infants. uhe g la b e lla r tap Althugh not specific for Parkinson's disease, it is usual to find it in Parkinsonian disorders and can be helpful in early diagnosis. Repeated tapping of tire bridge or at tire root of the nose is accompanied by syn chronous blinking. Tire tapping finger is brought from behind, hr the 'normal' response, blinking disappears after four to five taps. three classic signs C lw o s te k 's slgirn Tapping over the facial nerve or the branches of the facial nerve pro duces marked twitching and retraction of the corner of the mouth. This is present in tetany, and other states of muscular irritability. It may be seen to a slight degree in motor neuron disease. A similar movement occurs during regeneration of a damaged facial nerve. Chapter 25 The reflexes ilroysseau's sigra If the upper arm is compressed by a sphygmomanometer band for not more than 4 minutes, the hand goes into the main d'accoucheur position. This is an important sign of latent tetany, and can be increased and accelerated by hyperventilation. Lherm itte's pfoeiniomeimoira Forwards flexion of the neck results in a burst of electric shock-like paraesthesia shooting into all four limbs, or down the centre of the back. Sometimes this may occur in lower limbs only, or even on one side only. It was once thought to be pathognomonic of multiple sclerosis, and is most commonly found in this disease, but it also occurs in high cervical compression from spondylosis, or cerebellar ectopia, and is also seen in the early stages of radiation myelopathy and occasionally in subacute combined degeneration. The number of reflexes, named and unnamed, is legion. One textbook lists over 700. Fortunately, few possess any special advantages over those described, which are easy to elicit and whose reliability has earned their place in the routine examination.

’ ed 26 Coordination # Coordinate movement requires that both motor and sensory systems should be intact, because any satisfactory voluntary effort requires: 1 Efficient, smooth, adequate, but not excessive, movement of a group of muscles. 2 Appropriate and smooth relaxation of the correct antagonists, together perhaps with the contraction of other muscles, such as joint fixators, to ensure the efficiency of the movement. 3 Knowledge of the position of the moving part before, during and at the end of each movement. 4 Knowledge of the position of the point to which the part is to be moved, and the relationship between the two at any moment during the movement. Coordinate action of the muscles is under cerebellar control, and influenced by the extrapyramidal system, but intact proprioceptive sense combined with an accurate image of one's own body and its relationship to the environment are equally essential for the move ment to be completed satisfactorily. Lesions in many sites may therefore produce incoordination. Purposes of the tests To give the patient simple movements to perform which the exam iner can, from past experience, compare with the normal. A certain stereotyping of tests is required, because the possible number is, of course, uncountable. The aim of each test is to decide whether any defect demon strated is due to: 1 Cerebellar dysfunction 2 Proprioceptive deficiency 3 Some other factor, such as muscular weakness, simple lack of comprehension as to what the patient is supposed to do, or, and very important, perfect comprehension, but a determina tion to do it wrongly for a variety of reasons, none representing organic disease M ethods of testing coordination Coordination of ocular movements, speech and gait are dealt with in their appropriate sections. Chapter 26 Coordination rhe upper limb Preliminary observations Ask the patient to hold both arms outstretched in front of him. Watch the movement, and how well the posture is maintained. Then ask him to close his eyes and continue to watch the posture. Next, telling the patient to keep his arms up, tap each arm briskly and observe the displacement and return to the previous positions, both when Inis eyes are open and closed. Next, press downwards on each arm in turn and suddenly release the pressure. Normally, very little displacement occurs. With elbows flexed, ask the patient to tap briskly with the middle finger on his thumb. Note the speed and rhythm, which are usually swifter and smoother in the dominant hand. Next, ask him to tap the thumb with each fingertip in sequence, back and forwards from index to little finger. Abnormalities On first raising the arms, if there is cerebellar disease, the arm on the affected side will usually overshoot and have to be brought back to be parallel with the other. Frequent correction of its posi tion has to be made so that the arm appears to bounce and often deviates towards the other. Tire hand may show hyperextension of the metacarpophalangeal joints with partial flexion of the wrist. When the eyes are shut, these features may increase in cerebellar disease, but if the arm now begins to drift upwards and sideways (possibly outwards), this usually indicates postural deficiency. If the arm falls slowly, this may also indicate postural deficiency, but more commonly is due to simple muscle weakness. External rotation of the hand, bizarre posturing of the fingers and little writhing movements of the thumb and fingers may also be seen in severe postural deficiency, as well as in dystonic states (see p. 180). The arm is easily displaced on tapping due to: (a) motor weakness, when it remains in the displaced position, each tap displacing it further; (b) cerebellar hypotonia, when it tends to return after several bounces to the previous position; and (c) postural deficiency, when, with the eyes closed, the arm may be grossly displaced without the patient realizing it, and if told to replace it, he assumes an entirely incorrect position. 0 In cerebellar disease, with the eyes open or closed, after releasing pressure, the arm will fly upwards, sometimes 235 Part 5 The motor-sensory links so vigorously as to hit the patient (or the examiner) in the face. 0 In postural deficiency, when the eyes are closed, the arm tends to remain in the new position without much rebound. • Finger tapping is slowed by spasticity and extrapyramidal rigidity and by weakness, whether organic or 'functional'; the rhythm is disturbed by cerebellar disease, 'functional' disturbance or genuine involuntary movements such as chorea. It is the most useful screening test in the examination of the motor system. The finger-nose test Unless illustrated clearly for them, many patients for some reason have difficulty in following the first simple instruction which is to 'point the forefinger'. They may then perform the next movement in a quite unnatural manner, making assessment very difficult. 0 Each arm in turn is drawn out to full abduction and the patient is told to place the fingertip on his own nose and hold it there (Fig. 26.1). Again, if the instruction is not clear, a surprising number of patients try to touch the examiner's nose, a gesture that causes general embarrassment. ® Note the ability to point the correct finger, the smoothness of the movement, the accuracy of placing and the steadiness with which the finger is held on the nose to a count of five. Repeat the test with the eyes closed. 0 Minor abnormalities can be emphasized if the patient touches his forefinger to the examiner's outstretched finger and then moves it backwards and forwards from nose to finger quickly, the examiner holding his finger in a different place for each movement. Fig. 26.1 The fingernose test: (a) correct and (b) incorrect. The arm must be held in abduction or many of the abnormalities will be concealed. Chapter 26 Coordination Abnormalities In cerebellar disease, the arm on the side of the lesion may first be flung wildly outwards, striking nearby objects. Tire finger moves to the nose in a wavering side-to-side or up-and-down, but not a jerky, fashion, at the last moment being brought on to its object fairly accurately. This is increased slightly by eye closure, and emphasized by the harder tests. In milder lesions, all that is seen is the wavering to either side of a straight line during the course of the movement, with a tendency to overshoot the object at first, towards the side of the lesion. This wavering must not be confused with intention tremor, which is a side-to-side oscillation that develops as the finger approaches the nose, or occurs shortly after the finger touches the nose. It may be so wild as to make it impossible for the movement to be completed, or so slight as to consist merely of two or three oscillations while the finger is being held on the nose for a few seconds after touching it. In postural ataxia, the movement from start to finish can be a smooth one, though it is usually hesitant, and the finger hesitates before finally touching the nose. When the eyes are closed, how ever, the finger is unable to find the nose and, after first touching somewhere else, is then dragged along the skin of the face towards its object. If the finger is briskly, constantly and confidently placed on the cheek a little to one side of the nose, in the same place each time, this almost without exception reflects a non-organic state, and completely without exception if it is still constantly done with the eyes open. The Sower limb Preliminary observations Exactly the same principles are used as in the arms, but such fine movements cannot be expected. ° The patient is told first to raise one leg to touch the examiner's outstretched hand with his big toe. 0 A patient with normal muscle strength will be able to do this with only a little 'bounce' at the end of the movement. 0 Tapping time with the foot on the examiner's hand follows the same principles as finger tapping in the upper limbs. Abnormalities A patient with cerebellar disease is likely to overshoot, perhaps wildly, the leg will need several bounces before it reaches its object and the knee may be unnecessarily flexed. 237 Part 5 The motor-sensory links The heel-knee test o The heel is placed on the opposite knee and when the move ment is completed, the patient should be told to rim the heel down the front of the shin to the top of the foot. The whole test is then repeated on the other side, and finally it is carried out again with the eyes closed. 0 It is wise to touch initially the appropriate heel and knee, because otherwise some patients whose comprehension is slow flounder helplessly at a simple verbal instruction: others always perform the movement with the opposite limb, and some even attempt the discouraging manoeuvre of trying to place the heel on the knee of the same leg. Abnormalities 0 In marked cerebellar disease, the heel overshoots the knee side ways, and develops a rotary oscillation as it approaches it, which is the equivalent of the intention tremor in the upper limb. As the heel moves down the shin, it oscillates from side to side and finally shoots off the opposite foot in an uncontrolled manner. • In mild degrees o f cerebellar ataxia, the side-to-side oscillation may be the only obvious defect. ® In sensory ataxia, the heel is lifted too high in the first place and the patient raises his head to see what the relationship is of the two limbs. Deficiencies in the rest of the movement are not rhythmical, though the leg may fall off the other several times during its course. When the eyes are then closed, the patient is only able to find the knee by allowing the heel to land on some part of the thigh and then sliding it downwards. • In combined ataxia, elements of the two types of abnormality are present and the movement may be equally disorganized whether the eyes are open or closed. Again, in non-organic states, the heel is brought down confidently and repeatedly, eyes open or closed, in exactly the same place, usually about 15 cm below the knee. Dysdiadochokinesis This is a failure to efficiently perform rapidly alternating move ments. Tine patient should sit with his arms flexed at the elbow, holding the forearms vertically with the palms facing inwards. He is then Chapter 26 Coordination told to rotate the hands rapidly at the wrists. This will require both illustration and encouragement. Most people are less skilful at this with the non-dominant hand, and children are less adept at it than adults. The patient with cerebellar disease, however, makes a movement that is coarse, irregular and slow, with the hand dorsiflexed and the fingers extended, so that the whole palm is being shaken rather than the wrist rotated. If repeated with the fist clenched, a jerky flexion-extension of the wrist is superimposed on the attempted rotation. hi motor weakness, especially due to pyramidal tract lesions, the movement may be clumsier and slower on the affected side, but the particular features mentioned above are not seen. Past-pointing tests Past-pointing is a sign found both in cerebellar and labyrinthine disease. It must not be confused with the inaccurate placing of a position sense defect. ° The patient sits opposite the examiner holding his arm for wards horizontally, so that his fingers touch the fingers of the examiner's similarly outstretched arm. 0 He should then either lower the arm to his side or raise it above his head, and bring it back to the original position, first with the eyes open and then when they are closed. The test should be repeated several times with each arm. In cerebellar disease, the arm on the side of the lesion will deviate outwards towards the side of the lesion instead of accurately regaining its original position. hr unilateral labyrinthine disease, both arms will deviate towards the side of the lesion. This will be in the same direction as the slow component of the nystagmus, which undoubtedly will be present (q.v.). Additional tests These are the tests that are altered in cerebellar, pyramidal and extrapyramidal disease, and, though demonstrating ataxia, are of less value in its analysis. Rapid hand tapping Hand tapping is a good method of detecting a unilateral motor deficit, but does not fully analyse it. The back of one hand is tapped 239 Part 5 The motor-sensory links rapidly with the fingers of the other. Again, the non-dominant side is normally less skilful. In cerebellar disease, the tap becomes a rotary stroking movement. Such dysmetria can be emphasized by telling the patient to rotate the hand while tapping so that alternate taps are carried out by the palmar and dorsal surfaces. This test can also be adapted for the feet, but the speed of tapping as normally as possible is much less. Tapping in a circle A circle 1 cm in diameter is drawn and the patient, given a pencil, is asked to tap out a series of dots, all within the circle. In any ataxia, the patient will spread the dots irregularly over a wide area, out side as well as inside the circle. In unilateral cerebellar disease, more of the dots may be found displaced to the side of the lesion. This test is a good method of recording in the notes a deteriora tion or improvement in ataxia. Spiral drawing (Archimedes spiral) Ask the patient to draw a spiral. This is totally impossible in severe ataxia, tremor or chorea. It is not a specific test, but serial drawings offer a useful method of comparing improvement or deterioration. It is a useful test in essential tremor and primary writing tremor. See Video of Archimedes spiral drawing in the free companion CD-ROM of this book. Difficulties and fallacies Poor performance of these tests, particularly the heel-knee test, is not necessarily a sign of true ataxia. They are not easy tests to do for the first time and an agitated patient must be encouraged to take his time and to do them slowly. Remember, this may be the tenth patient that afternoon you have told to put his right heel on to his left knee, but for the patient this may be the first time in 65 years that he has been given this improbable instruction. Note the presence of any static tremor in the hands at rest, because at the end of the finger-nose test, this might be mistaken for intention tremor. Marked muscular weakness may interfere seriously with the heelknee test, the heel repeatedly falling off the shin, but the difficulty in raising the thigh and maintaining the knee in extension will have been noted in the early stages of the test. 240 Chapter 26 Coordination Pyramidal and extrapyramidal lesions slow up the performance of tests requiring rapid fine movement, but the execution of the individual stages of the movement is correct and there is not the decomposition seen in cerebellar ataxia. Always correlate the degree of ataxia demonstrated on the couch with your observations of what movements the patient is able to perform when he does not realize he is being specifically examined. Precise coordinated movements may be seen which are not merely automatic, and a suspected simulated ataxia may be confirmed. The failure to demonstrate abnormalities of coordination when the patient is lying in bed must never be considered to exclude cer ebellar disease, for in a midline posterior fossa or cerebellar vermis lesion, or in one in which the cerebellar tonsils are displaced through the foramen magnum (e.g. in the Chiari deformity and other anom alies in this area), gross ataxia of gait may be the only physical sign, tests of coordination while recumbent being normal. Before this was realized, many patients with gross ataxia of gait were wrongly labelled as hysterical, and even these days many still are.

t 241 Part 8 Examinations of Particular Difficulty TH The unconscious patient 'Normal consciousness' has not, so far, been satisfactorily defined and it is easier to describe abnormalities of the conscious state. There are two aspects of the conscious state that can vary inde pendently in different types and distribution of brain disease. One is the content of consciousness, the sum of mental function; the other is called the state or level of consciousness, which is the degree of alertness or arousal. So, there are conditions in which the patient, though awake and responsive, is imperfectly aware of himself, his actions and his environment. Here, the content is affected. Alternatively, in other states of impaired or lost con sciousness, we see defects of arousal of varying degree. Anatomi cally, the central reticular formation of the brainstem is the basis or arousa/; ’the content of consciousness depends on the activities of the cerebral cortex and thalamus. Full conscious behaviour requires intact cerebral hemispheres and a normal brainstem. In both cases, an acute lesion is more likely to influence conscious ness than a slowly developing one. Destructive lesions of the hemispheres, if slowly evolving, have to be extensive before con sciousness is impaired. On the other hand, discharging lesions, as in epilepsy, are often small. Similarly, only relatively small, bilateral lesions of the brainstem are required to disturb conscious ness profoundly. Purpose of the examination 1 To determine the state or level of consciousness. 2 To identify or exclude systemic disease known to cause dis turbances of consciousness without prim ary disease of the brain. 3 If due apparently to a neurological lesion, to determine its site in the brain or brainstem. One thinks in terms of 'north or south', rostral or caudal. The attitude towards investigation is bound to differ in units fully equipped for intracranial emergencies, and in the more general hospitals. The majority of the remarks in this chapter are based on the fact that most comatose patients are not primarily admitted to neurological or neurosurgical units, but in the final paragraphs there is an indication of the steps such units would undertake. 245 Part 6 Examinations of particular difficulty History So much information can be gleaned from the history that no amount of trouble should be spared to obtain the fullest details. Relatives, friends, workmates, police, ambulance men or other wit nesses must be searched out and questioned, making certain that the information obtained is precise, relevant, factual and not sup position, and determining whether it comes from direct knowledge or hearsay. The points of particular importance in the history are: 1 The mode o f onset. Was the loss of consciousness abrupt as in cerebrovascular catastrophes or epileptic states; rapid, over a period of a few hours, as in some cases of intracranial haemor rhage and in toxic states, or gradual, over days, as in expanding intracranial lesions? 2 Premonitory symptoms. Complete absence of premonitory symp toms would suggest a primary intracranial vascular accident or an epileptic attack. Previous episodes of a similar nature sug gest either epilepsy, the recurrent administration of drugs or, very rarely, the blocking of cerebrospinal fluid (CSF) pathways by some intermittent obstruction to the ventricles, such as an intraventricular mass or cyst. Headaches, with vomiting, progressive mental change, increas ing weakness or unsteadiness of the limbs would all suggest an expanding intracranial lesion. Progressive severe loss of weight, anorexia and asthenia suggest metastatic disease, while symptoms such as a cough, dyspnoea, anorexia, melaena, lumps in the breast, past mastectomy or gas trectomy may indicate the probable site of the primary lesion. Any active infection in the ears, chest or sinuses may suggest intracranial infection. A history of severe psychological disturbance, especially depres sion, raises the possibility of self-administered drug intoxication. A history of alcoholism may be obtained, but it is often very dif ficult to get an honest assessment, even from would-be helpful rela tives. Most important is the question of trauma, remembering that the elderly are prone to intracranial bleeding after relatively slight degrees of trauma. The cause and type of the injury must be established, its severity, the length of the interval between its occurrence and the loss of consciousness, i.e. whether imme diate, or after a delay of minutes, hours, or days; if gradual or abrupt in onset and whether there was initial recovery and then relapse. 246 Chapter 27 The unconscious patient It is also important to know if there has been any associated frac ture of long bones which might have been the source of cerebral fat embolism. Examination Much of the examination will consist of careful observation, car ried out repeatedly and perhaps over a long period of time, both in the patient's immediate vicinity and also from a position where a patient who might be feigning unconsciousness is not aware of being under observation. Assessment of the degree of altered consciousness This must be decided at once, because the future examination will be largely governed by the conscious level, which should be described in detail. There are infinite gradations of altered con sciousness characterized by defects of arousal, but the terms drowsi ness, stupor and coma usefully describe the three major stages that can be clinically recognized. D row siness This is a state resembling normal sleepiness. Stimulation rouses the patient to a state of complete wakefulness and cooperation, but he tends to sink into sleep again if stimulation ceases. Normally, the full neurological examination can be carried out. This state is com mon in high brainstem disturbances, direct or indirect, and in drug toxicity. Stupor Left alone, the patient appears to be completely unconscious, but nevertheless may be restless. On vigorous stimulation, he can be roused sufficiently to resist painful stimuli, or even for short peri ods to respond to commands or to answer simple questions. No satisfactory cooperation is obtained and as soon as stimulation ceases, the patient reverts to Inis original state. Bilateral cerebral hemisphere disease (anoxic, toxic or traumatic) and compression or disease of the upper brainstem may all cause stupor. In the variety called akinetic mutism, the patient lies motionless and speechless but with eyes open as if awake. He is not unrespon sive or resistive and Iris eyes may appear to gaze about him. One has the curious impression that these patients are just on the very brink 247 Part 6 Examinations of particular difficulty of saying something, but never quite achieve it. Yet, there is always an amnesia for the duration of this state. It occurs in association with lesions in the neighbourhood of the third ventricle, or subacute encephalopathies, and is probably a consequence of involvement of the reticular formation in the upper brainstem. It illustrates the dissociation that may occur between wakefulness and awareness. There is another similar condition but having important differ ences. All movement save for blinking and vertical gaze is para lyzed; but consciousness is retained. The patient is aware of himself but physically 'locked-in'. When the cause is a peripheral disorder such as acute polyneuritis (Guillain-Barre syndrome), it is not dif ficult to recognize that the patient is conscious. But when central processes lead to this 'de-efferented' state, coma may be readily simulated. The responsible lesion is in the ventral pons, either vas cular or demyelinating. Vertical eye movements are served from the superior collicular region of the midbrain and are thus preserved in the presence of a tetraplegia and faciobulbar paralysis. Coma The patient is deeply unconscious. He will respond only in the most elemental way to painful stimulation and cannot be roused to any degree of cooperation. He is usually lying immobile and incontinent. There is some justification in describing a coma that is bordering on stupor as 'light' and one in which no form, even of reflex activity, exists, as 'deep'. The deeper degrees of unconsciousness are com mon in pontine and low brainstem lesions; pupillary, corneal and swallowing reflexes may be abolished. Three particular aspects of the behavioural response have been used in the now widely accepted Glasgow Coma Scale (Fig. 27.1). Various grades of eye opening, ver bal and motor response can be recorded on a special chart to pro vide consistent standard of appraisal. Such methods help to avoid the errors that often result from the imprecise use of language by the medical or nursing staff. (Never use the phrase 'semi-coma'.) In addition to these three states, there are other abnormalities of awareness (content) that must be recognized though they need not necessarily be accompanied by any disturbance of arousal. Diffuse disturbance of the cerebral hemisphere is responsible. Confusion and disorientation The confused patient may be quite alert and even cooperative, but is incorrect in his comprehension and assessment of his own state or his environment. It is usual to test in three dimensions. 248 Chapter 27 The unconscious patient 4 To sp eech c 0 N one 3 2 1 O rie n ta te d 5 M A C o n fu se d 4 In ap p ro p riate w o rd s 3 In co m p reh en sib le so u n d s 2 N one O b ey co m m a n d s 1 6 W ith d ra w a l 5 Lo calize pain 4 Flexion to pain Exten sio n to pain 3 2 N one 1 Eyes open S Fig. 27.1 Glasgow Coma Scale. In each square is marked the 'score' for each test on consecutive examinations annotated either as a figure on a scale of 1 to 4 or simply by a tick. S p o n ta n e u o u sly C Best verbal response A L E Best m o to r response To pain 1 Time. Ask his knowledge of the day, date, month, season, year and how long he has been in his present place. 2 Place. Ask where he is, what room, what building, what floor, what town and what country. 3 Person. Ask who he is, what is his work and what is his age. Finally, ask if he knows who the doctors, or nurses, or relatives are who may be around the bedside. Delirium In this state, the patient appears out of touch with his surround ings and is spontaneously producing evidence of his confusion and disorientation by muttering, rambling, shouting, often offensively, often continuously, with evidence of delusion and hallucination, and often with so much associated motor activity that physical exhaustion overcomes him. This is to be seen in toxic and infective states, and most strikingly in the delirium tremens of alcoholism, a condition that once seen will never be forgotten. Catatonia This may be a symptom of a psychotic state in which the patient, otherwise entirely normal, lies mute, immobile and unresponsive. He does not follow movements; does not appear to be paying any attention to his surroundings and will often have a plastic rigid ity of the limbs, which may remain in any position in which they 249 Part 6 Examinations of particular difficulty are placed, however bizarre that may be. Modem drug therapy has greatly reduced its frequency. Similar states are more frequently seen in organic frontal and hypothalamic lesions, including mutism, but other neurological signs are present. Having assessed the level of consciousness and the degree of cooperation to be expected from the patient, a general inspection of the face, skin, position and movements of the body must be carried out before passing on to the examination of the cardiovascular and central nervous systems. General observations The purpose of the following paragraphs is to indicate a train of thought set up by certain observations and they must not be taken as a suggestion that these observations give the diagnosis. Colour and condition of the skin 1 Pallor and sweating occur in syncope, severe blood loss and hypoglycaemia, but may be seen in brainstem vascular defi ciency, and following some epileptic attacks. 2 Suffusion o f the face is seen in hypertension, alcoholism and sometimes in cerebral haemorrhage. 3 Cyanosis o f the face and neck accompanies respiratory obstruc tion, epilepsy and some intracranial vascular accidents. It is, of course, a crucial sign requiring correction as a matter of urgency if possible, and should therefore, until proved otherwise, be assumed to be due to respiratory obstruction. 4 Cyanosis o f the limbs is seen in peripheral circulatory stagnation, severe coma and collapse. 5 Jaundice may suggest multiple metastases, but may also indi cate hepatic coma due either to primary liver disease or to drug toxicity. 6 A cherny red colour is seen in carbon monoxide poisoning. 7 Petechiae and ecchymoses are found after a fit; in bacterial endo carditis, severe septicaemia, collagenoses, blood dyscrasias (primary or secondary to drug toxicity) and in patients on high cortisone dosage. In small children, the possibility of multiple inflicted trauma must not be overlooked. 8 Corrosion o f the lips and mouth indicates poisoning. 9 Needle marks may indicate drug abuse or medication. 10 Emaciation suggests advanced malignant disease, or severe deficiency disease, which might, of course, be misleading and merely due to prolonged unconsciousness. 250 Chapter 27 The unconscious patient Position of the body 1 Neck retraction, especially in children, usually indicates menin geal irritation, but it may sometimes occur when there has been a cerebellar tonsillar pressure cone. 2 Neck retraction and arching o f the back (opisthotonos). This is not uncommon in small children with meningeal infections, or advanced degenerative lesions. It may occur in severe menin geal irritation and in tetanus in adults. Extreme degrees of opis thotonos in adults with extension (not flexion) of the hips and knees is rarely organic. 3 Curling o f the body away from the light is seen in meningeal irrita tion by infection or blood. 4 The patient who lies with the head drooped to one side, one upper limb pronated and adducted and one lower limb externally rotated, usually has developed an acute and profound hemiplegia. 5 When all limbs are extended, the upper limbs are pronated and the feet plantarflexed, this represents the decerebrate attitude and occurs in lesions of the midbrain between the superior collicu lus and pons. 6 If, however, the upper limbs are flexed and the lower limbs extended, the decorticate position is seen in extensive lesions at the level of the basal nuclei, or between this level and the cortex. Both postures may first be revealed by response to cutaneous stimulation. Involuntary movements of importance iin states of disturbed consciousness Convulsions Coma accompanies and follows isolated fits or status epilepticus, of 'idiopathic' or symptomatic origin. The degree of uncon sciousness may be far greater than the severity of the preliminary convulsions may lead a layman to expect that the cause is benign in nature - so the history again is

vital. If convulsions are known to be limited to one part of the body, for all practical purposes this indi cates structural disease of the opposite hemisphere. Repeated focal convulsions involving a little more of the limb each time, and par ticularly if then spreading to the other side, are highly suggestive of a spreading cortical thrombophlebitis or an
encephalitis. Minor twitching of the extremities, if unilateral, has localizing value, but if bilateral, it merely indicates diffuse cerebral disorder. Unilateral myoclonic jerks have a similar significance; see the myoclonus section (p. 171) for their importance in subacute encephalitis. 251 252 Examinations of particular difficulty Attacks of generalized
rigidity and decerebrate attacks These may occur when the brainstem is the site of primary dis ease, when it is under compression, subjected to trauma or has had haemorrhage into its substance either primarily or as a result of tentorial herniation. They may also be seen in very advanced dif fuse cerebral and basal nuclear disease, especially in
children, in whom they may also accompany meningitis. Rigors and tremors Intermittent rigors without rise of temperature occur when there is an irritative lesion of the ventricular walls, e.g. by rupture of an abscess or haematoma, and a constant coarse twitching tremor of the hands and arms accompanies this ventriculitis. Further general
examination Respiration (Fig. 27.2) Stertorous breathing, accompanied by stridor and cyanosis if there is also respiratory obstruction, is common in epileptic fits and cere brovascular accidents. One of the commonest disorders of respiration is the periodic (Cheyne-Stokes) type in which there is alternating hyperpnoea and apnoea. This usually
indicates a bilateral cerebral or high brain stem lesion or metabolic dysfunction. The causal lesion is rarely as low as the upper pons. Other brainstem lesions of the midbrain or mid-pons cause central hyperventilation or ataxic irregular breathing patterns. The latter are most commonly seen in lesions of the medulla and lack the rhythmical waxing
and waning of the Cheyne-Stokes type. Shallow, rapid breathing occurs in shock, haemorrhage and hypoglycaemia. The odour of acetone in the breath in diabetic comas is classical, but not detectable by many people. Pulse The pulse is: (a) Rapid and weak in haemorrhage, shock, some stages of syncope and circulatory collapse. (b) Slow and weak in
some stages of syncope and in vasovagal attacks. Chapter 27 The unconscious patient A. Forebrain and diencephalic lesions B. High brain stem lesions C. Bilateral pontine lesions ••• D. Pontomedullary junction lesions E. Medullary lesions A . F o r e b r a in a n d d ie n c e p h a lic le s io n s / ' /IMMi Ai ---------- /WlWf Tr+ j i l AI Alt i . ■ i l l AI M i l . ~
—W w * !** -------- Cheyne-Stokes respiration B . H ig h b r a in s t e m le s io n s Central neurogenic hyperventilation C . B ila t e r a l p o n t in e le s io n s s — ~\— / a ____ Apneustic breathing r D . P o n t o m e d u lla r y ju n c t io n le s io n s feMw'frW— I-------------------------- 1 60 seconds Cluster breathing or ataxic breathing E . M e d u lla r y le s io n s
Apnea Fig. 27.2 Abnormal respiratory patterns associated with lesions at different levels of the brain. 253 Part 6 Examinations of particular difficulty (c) Very slow and full in complete heart block (when Stokes-Adams attacks occur), and in severely increased intracranial pressure. A weak, irregular pulse is found in low brainstem lesions and a
fibrillating pulse raises the possibility of cerebral embolism. Examination of the head Injuries may be hidden by hair, but careful palpation may reveal a fracture line, a depression or a 'soggy' area of a severe contu sion. A bony lump may suggest an underlying exostosing tumour. Percussion of the skull in children with hydrocephalus produces a high-pitched, cracked-pot note, and in comatose infants a bulging fontanelle indicates increased intracranial pressure. If burr holes or a decompression are present, they should be palpated, because if tense and bulging this means high intracranial pressure. Signs of meningeal irritation Test for neck rigidity and carry out Kernig's test, both at the beginning and the end of the examination, when the stimulation may have roused the patient. Neck rigidity is the more important sign. It may, however, indicate cerebellar tonsillar herniation at foramen magnum level, rather than frank meningitis. This is important for it would contraindicate a lumbar puncture, and the history of the onset is vital. In
meningitis, Kernig's sign is usually positive as well. In the earliest stages of subarachnoid haemorrhage, neck rigid ity may not have developed, sometimes is delayed even 24 hours, and if coma or collapse is profound, e.g. after massive intracranial bleeding, or in advanced meningitis in childhood, neck stiffness may not develop at all. The ears
Examine carefully for signs of: • Middle-ear infection, which may indicate an intracranial abscess. • Tenderness and swelling over the mastoid, for the same reason. • Bleeding from the inner ear, as this often indicates a basal fracture. The tongue A common cause of coma being epilepsy, the tongue should be examined for evidence of having been
bitten. The surface and the inside of the cheek, palate, fauces and pharynx should be inspected for the effects of corrosive fluids. 254 Chapter 27 The unconscious patient Examination of the nervous system An attempt should first be made to raise the conscious level. This is done by rubbing the sternum vigorously with the knuckles, and by pressure in
the supraorbital notch. (The actual level or state of consciousness has already been recorded.) The more detailed examination of the nervous system is now designed to differentiate between the two basic causes of altered consciousness (brainstem reticular formation depression or bilat eral cerebral dysfunction together with brainstem depression)
and to seek any evidence for a specific structural lesion. There may be signs of diffuse disease or of a focal lesion in the supratentorial or subtentorial compartment. In addition to evaluation of the level of consciousness itself, and attention to the pattern of respiration, it is the examination of the pupils, ocular movements and motor func tion that
forms the most crucial part of the exercise. Th e cranial nerves The ordinary routine examination is not possible, for all observa tions have to be objective, and a somewhat different approach must be adopted. The fundi These must be most carefully examined in every case, the abnor malities having the same significance as described in Chapter 7.
Remember, however, that whereas true papilloedema (in the absence of marked hypertension) indicates increased intracranial pressure, its absence does not in any way exclude a space-occupying lesion. Visual fields These cannot be accurately tested, but in stuporose states, a menac ing movement towards the eye from one side and then the other
will normally produce a blink (menace reflex), which may be absent when the gesture is made from the side of a hemianopia. The pupils (Fig. 27.3) Note the size of each pupil, their comparative sizes and their reac tions. For full details of pupillary abnormalities, see Chapter 8. In states of disturbed consciousness, the following points are of greatest
importance: Part 6 Examinations of particular difficulty Small, reactive: Metabolic-toxic encephalopathy Small, reactive: Diencephalic Dilated, fixed : III nerve (uncal herniation) Midposition, fixed : Midbrain Pinpoint, reactive: Pons Fig. 27.3 Pupillary changes seen in various lesions at different levels in the brain. 0 In most cases of toxic or metabolic
coma, the pupillary light reflexes are spared. Their presence or absence is, in fact, the single most important sign in distinguishing metabolic from structural disease. • Widely dilated pupils (7-9 mm) occur in midbrain lesions that selectively affect the oculomotor complex or third nerves, and do not react to light. • If the sympathetic system, also, is
involved, pupils of medium size (4-7 mm), which are inactive to light, may result from mid brain lesions. These may be primary, or secondary to central pressure herniation as in cases of tumour or haematoma in the supratentorial compartment. Be certain any dilatation is not due to the use of atropine by a previous examiner. • Unilateral pupillary
dilatation with deteriorating consciousness should be considered as a sign of unilateral tentorial herniation until proved otherwise. It is commonly, but not invariably, on the side of the expanding lesion and demands prompt surgical Chapter 27 The unconscious patient action. By the time both pupils are dilated, there has usually been a secondary
haemorrhage into the brainstem, and even correction of the primary lesion may not save the patient. • Very small pupils (0.5-1 mm) are seen in pontine lesions (pinpoint but reactive - to be examined with the help of a magnifying glass), and in certain drug intoxications, especially morphia, but also during simple sleep. • Small pupils with loss o f light
reflex and upward gaze palsy are seen in thalamic haemorrhage of posterolateral type. Ocular position and ocular movements Note the position of the eyes, any deviation from the midline, how well sustained this is, any spontaneous ocular movements and any movements on sudden noise or on command. Abnormalities of conjugate deviation have an
important role in comatose states and are dealt with in Chapter 8. Remember that forced upward deviation of the eyes or oculogyric crises can be caused by toxic doses of the phenothiazine derivatives. Skew devi ation of the eyes occurs in pontine or cerebellar lesions. Because the pathways controlling oculomotor vestibular reflexes lie adjacent to
the reticular formation, good use can be made of testing these responses. The reflex eye movements following passive rotation of the head (oculocephalic or 'doll's head' manoeuvre) and caloric stimulation of the ears (oculovestibular) are noted. These observa tions are of particular value in distinguishing structural lesions of the cerebral hemispheres
from those of the brainstem, remember ing, of course, that the latter dysfunction is often secondary to cen tral herniation. But in the absence of secondary brainstem compres sion, cerebral hemisphere lesions do not affect either reflex. Similarly, metabolic suppression of the hemispheres and brainstem spares the oculomotor vestibular pathways until
late, when their abnormality can then be used to identify the so-called brainstem death. In the normal oculocephalic reflex (the 'doll's head' or 'doll's eye' manoeuvre) - brisk rotation of the head from side to side or flexion/ extension - the. eyes, show. conij way firom 'frie direc tion in which the head was moved, before then returning to the pri mary
position. It can be recalled from Chapter 11 that irrigation of the external auditory canal with cold (or warm) water excites nystagmus. (In the unconscious patient, ice-cold water is routinely used to cause maximum effect.) This forms the basis of the oculovestibular (caloric) reflex. In lesions of the diencephalon causing coma, the fast compo nent of
that nystagmus is depressed, so that there is, instead, just tonic conjugate deviation of the eyes towards the irrigated ear. Midbrain or Part 6 Examinations of particular difficulty upper pontine lesions can cause various abnormalities; deviation may be confined to the ipsilateral eye, indicating a contralateral intemuclear ophthalmoplegia, or be seen
only in the contralateral eye, suggesting a sixth nerve lesion on the stimulated side. Lower pontine lesions abol ish all responses and there .will also be absent corneal reflexes. Occa sionally, such absence of any ocular response is the result of bilateral end-organ poisoning by gentamycin or other related antibiotics. The face Facial muscles Voluntary
movements cannot be tested. First study the lines and contours of the face. Flattening and smoothing of the wrinkles on one side, with uncovering of the sclera, are seen in paralysis of lower motor neuron type and sometimes in the acute stages of severe upper motor neuron paralysis. The paralyzed side will be blown out and sucked in during
expiration and inspiration. Firm pressure on the supraorbital notch may produce a grimace. Normally, this is most marked on the side stimulated, but when there is facial paralysis it is reduced on the affected side, no matter which side is stimulated. Touching the comer of the mouth may produce a sucking reflex and deviation of the comer of the
mouth. This is seen in the adults in bilateral cerebral lesions of severe diffuse character. Facial sensation The corneal reflex must be tested in each case. It is often bilaterally absent in deep coma or depressed if the cornea has been exposed for a long time. Painful stimulation of the skin should be carried out by pinprick or pinching, and the relative
grimacing on each side compared. Great care in interpretation of corneal reflex and facial sensory tests is required in the presence of facial paralysis (see Chapter 10). Purely unilateral loss of sensation in comatose patients is usu ally part of a complete hemianalgesia and indicates a deep-seated hemisphere or upper brainstem lesion. Sensory loss on
one side of the face but on the opposite side of the body indicates a lower brainstem lesion, and is especially common in the lateral medullary syndrome, traditionally due to a posterior inferior cerebellar artery thrombosis, but in fact usually a sign of vertebral artery disease. The motor system A great deal will have been learnt during the preliminary
period of inspection. This applies particularly to the position of the limbs Chapter 27 The unconscious patient and the degree of movement carried out by each limb. The tone of the limbs can be tested in the normal way, but remember that in the acute stage of a severe intracranial lesion, the tone may be lost, rather than increased, on the side
opposite to the lesion. Raise both arms together and let them fall back. Normally, the fall is checked and slowed, but in unilateral paralysis the arm will fall unchecked. Next, raise the legs into the flexed position, so that the soles of the feet rest on the bed, and then allow them to fall back. Unparalyzed limbs will remain in that position or slowly
extend. A paralyzed limb will rapidly fall back to its origi nal position. Remember, midbrain lesions produce decerebrate posturing, and that this may at first only follow noxious stimulation, and may be unilateral if the other side is paralyzed. Th e reflexes Testing the reflexes, being an objective procedure, can be carried out in the normal way and the
usual deductions can be drawn, but in states of deep coma the interpretation of the results may be different. Tendon reflexes may be absent on the paralyzed side, or in deep coma, throughout. Superficial reflexes may be unobtain able and have little localizing value. The plantar responses may be absent, or bilaterally extensor, and so also have little
localizing value, except that if there is a unilateral extensor plantar, this indi cates the side of a pyramidal system lesion. Th e sensory system Sensation can be assessed by observing the patient's response to painful pinching or pricking of the face, arms, trunk and legs, on each side, and it may be necessary to stimulate vigorously. Dilata tion of the
pupils, grimacing or movements of the limb will occur if sensation is present. Failure to withdraw a limb may indicate paralysis, loss of sensation or both. In the former, there is usually movement of the normal limb even when the abnormal side is stimu lated. In deep coma, these tests are of little value, other than as an indication of the depth of the
coma. Coordination This cannot be tested in the ordinary way, but in stuporose states the patient will often attempt to fight off interference and to brush off the painful stimuli. During these movements, the presence of 259 Part 6 Examinations of particular difficulty a marked ataxia can be observed, and assessment can be made of the patient's ability
to localize a stimulus and to place his hand cor rectly on the painful point. Essential investigations Blood Obviously, the history and clinical assessment of the case will influ ence greatly the type of investigation to be undertaken, but in every case the sample must be sent immediately for a full blood count, estimation of glucose, urea and electrolytes
and often for plasma calcium and liver function tests. Once these have been done, a major screening process will have been completed, and such conditions as hypoglycaemia, diabetic coma, uraemia and severe electrolyte disturbances should have been detected. In suspected alcoholism, the blood alcohol may also help, and in hepatic failure there is
usually a raised blood ammonia level. Drug toxicity7 is bound to arise, and it is becoming increasingly necessary7 to screen for drugs and toxins and to know whether these levels are compatible with normal or excessive intake. Close liaison with the biochemical laboratory on this point is vital. If there is evidence of cardiac disease and the erythrocyte
sedi mentation rate or viscosity is raised, the coma may be due to infected emboli from bacterial endocarditis, and this will be one of the indications for a blood culture. In some situations, there will be a need for blood gases, cortisol levels or thyroid function tests. (Although the occurrence of myxoedema coma must now be very rare, the
characteristic facial appearance, fishlike skin, hypother mia and 'hung-up' reflexes should still be recognized.) It must be emphasized that in cases of suspected trauma or tumour, time should not be wasted awaiting the results of these investigations. They can be going on whilst scanning is being car ried out. Computerized tomography Head
computerized tomography (CT) is indicated in practically in all unconscious patients if the diagnosis is unclear, particularly if there is sign of structural, focal or lateralizing cerebral pathology or raised intracranial pressure. Further evaluation of the structural lesion close to skull base or posterior fossa may require magnetic resonance imaging (MRI).
260 Chapter 27 The unconscious patient M R I braem In case the CT brain is not able to reveal any gross abnormality, MRI brain may be useful in detecting the focal abnormalities by routine T l, T2 and fluid attenuation inversion recovery sequences. Other sequences like diffusion-weighted imaging (DWI), susceptibilityweighted imaging (SWI) and
others may be required to identify relevant lesions. Small lesions in the posterior fossa or brainstem (demyelination/infarcts) may be picked up by MRI. Whenever indicated, contrast MRI should be done. X-rays o f th e ch e st Radiological appearances in the chest of comatose patients can give indirect hints as to the possible cerebral lesion.
ElectroeracephaBography Electroencephalography (EEG) is most useful in cases of general ized encephalopathy, metabolic or infective. There may be specific clues suggesting, e.g. hepatic coma, Creutzfeldt-Jacob disease, her pes simplex encephalitis and non-convulsive status epilepticus. A focal abnormality, in the absence perhaps of clinical signs,
will alert one to the need for CT. Alternatively, a normal recording may indi cate psychiatric problems or intrinsic brainstem disease. Finally, the identification of subclinical epilepsy of any form is vital. Lwmfoar puncture Great care is required in the selection of comatose cases for lum bar puncture. No patient with papilloedema should have a lum bar
puncture carried out unless neurosurgical facilities are readily available and preferably in the same hospital. This applies also where, papilloedema or no papilloedema, it is strongly suspected that the lesion is a space-occupying one, and particularly if it is highly likely to be in the posterior fossa, or to be a subdural haematoma or a cerebral abscess.
If, however, the patient has signs of meningeal irritation, but nothing to suggest a dangerous intracranial space-occupying lesion such as papilloedema, a slow pulse, gross cerebellar signs, active middle-ear or mastoid infection, then the CSF should be examined. Hie details of CSF examination are given in Chapter 34. CT can confirm subarachnoid
haemorrhage when raised intracranial pres sure contradicts lumbar puncture. 261 Part 6 Examinations of particular difficulty Deteriorating consciousness is always a grave sign, but before embarking on extensive investigation, each case must be assessed as an individual problem, always asking oneself whether 'doing something' is, in fact, going to
benefit the patient. Remember also that deteriorating consciousness is not the same as failure to regain consciousness. In head injuries, e.g. the former is very much more of an indication for further investigation than the latter. 262 28 Disorders of speech This part of the neurological examination frightens many physicians, probably because the
volume and complexity of the literature on the subject is so great that it is assumed that the examination must be similarly prolonged and complex. Certainly, a minute analysis of a speech defect can be both, but for practical purposes the procedure can be both shortened and simplified. Assessment starts while tak ing the history, continues
throughout the physical examination and then is analysed in more detail by the use of specific tests. Speech defects fall into four main types: aphasia, dysarthria (anarthria), dysphonia (aphonia) and mutism. Before any detailed analysis is attempted, the disturbance must be placed in one of these groups. Methods of differentiation 1 If the patient is
conscious, but making no attempt to speak or make sound, this is mutism. It is usually part of a psychological disorder, but may be seen in lesions affecting the anterior part of the walls of the third ventricle and the posterior medial surface of the frontal lobe bilaterally. Total mutism is sometimes due to profound motor aphasia (Broca's aphasia), but
the patient then gives the impression both of paying attention and of trying to communicate even if unable to do so verbally. 2 If the patient, though speaking, fails to produce any volume of sound, or merely whispers, this is aphonia. It is due to disorders of the larynx and vocal cords. If, despite this, the patient is able to cough normally, it is probably
of non-organic origin. 3 If the volume of sound and the content of the speech are normal, but the articulation arid enunciation of the individual words and phrases are distorted, this is dysarthria. It is a disorder of control of the muscles producing speech, the lesion lying in upper or lower motor neurons, cerebellar system or the muscles themselves -
i.e. at many different levels. 4 If the patient is failing to put into properly constructed words or phrases the thoughts he wishes to express, even if articula tion is adequate, this then is aphasia. The lesion is one of the highest mechanisms of speech and must be in the dominant cerebral hemisphere. Aphasic states also include disturbances of writing
(dysgraphia), and failure to comprehend the spoken word (receptive aphasia), or the written word (dyslexia). Part 6 Examinations of particular difficulty If either dysarthria or aphasia is present, the defect needs closer analysis, but should not be attempted in any detail at the first inter view. If the analysis is difficult, it should not all be attempted at
one time, for fatigue easily affects both the patient and the examiner and militates against accurate results. The analysis of dysarthria Correct articulation requires correct bilateral coordination of the tongue, lips, palate, larynx and the muscles of respiration. These may suffer from lesions of the upper and lower motor neurons, the coordinating
influence of the extrapyramidal and cerebellar sys tems and the actual muscles themselves. Examination 1 Listen to the clarity of the patient's enunciation during history taking. 2 Ask the patient to repeat certain phrases. These can be cho sen from a newspaper. Many tongue-twisters are used, but it is wiser to start with easier words, for the normal
person may find great difficulty with some of the traditional test phrases. These include 'British Constitution', 'Methodist Episcopal', 'The Royal Devonshire Constabulary', 'Chakravarty Rajgopalachary', 'Tiruanantapuram', 'Muzaffarpur' etc. 3 Ask the patient to read a paragraph from a simple book. 4 Ask the patient to count successively to 30 or above
to test for muscle fatigue. Listen to the words, whether they slur into one another; whether the rhythm is jerky, explosive or monotonous; whether speech is too loud or too soft; whether particular letters present particular difficulty, and which are they; whether there is a nasal tone to the speech and whether the disturbance is constant throughout,
variable or increas ing towards the end of each sentence or on prolonged counting. Varieties of dysarthria Spastic dysarthria This is caused by bilateral upper motor neuron disease. The tongue is small and spastic; the speech is slurred; excursions of the mouth are limited; the letters 'b', 'p', 'd' and 'y' suffer particularly. The impression is gained that
the patient is trying to talk from the back of the throat. The jaw jerk and palatopharyngeal reflexes are exaggerated. 264 Chapter 28 Disorders of speech Hypokinetic dysarthria This is a result of extrapyramidal lesions producing rigidity of face or tongue muscles, without wasting, and without exaggeration of the reflexes. Speech is monotonous, all
inflections and accents dis appear, words run into one another and sentences start and stop abruptly. Excursions of the tongue and lips are greatly reduced. The later words in long sentences may come out in a rush. In severe cases, the phenomenon of palilalia, i.e. the constant repetition of a particular syllable, may be heard. Ataxic dysarthria Here
there is incoordination of the muscles of speech, including the respiratory muscles. Speech is irregular, slurred and drunken, sometimes too loud, or too soft, the words run into each other or are spaced too far, the rhythm is jerky, sometimes explosive, stac cato or scanning. There may be accompanying grimaces and ges ticulations. It may be quite
impossible for the patient to articulate the test phrases. Think of an intoxicated person's speech! Flaccid dysarthria Dysarthria due to lesions of the lower motor neurons and muscles Speech as a whole may be well preserved, but individual words and sounds cause difficulty, the distribution and extent of the muscular weakness governing the
particular variety of defect produced. Spastic ® Pseudobulbar palsy ® Motor neuron disease • Stroke (multiple/single) ° Upper brainstem tumours H yp o k in e tic ° Parkinsonism A ta x ic 8 8 • ° 8 Multiple sclerosis Cerebellar disease or tumours Hereditary ataxia Choreas Anticonvulsant and other drug toxicity 265 Part 6 Examinations of particular
difficulty Facial paralysis causes difficulty with the labials such as 'b', 'p', 'm ', and V . Tongue paralysis affects a large number of sounds, particularly those involving the letters T , 'd', 'n', 's', 't', V , V , and speech is profoundly distorted. Palatal paralysis produces nasal speech; V a n d 'd ' become 'm ' and 'n', 'g' becomes 'rh' and 'k' sounds like 'ng'. The
position of the head alters the degree of the defect, the patient's speech being worse when the head is bent forwards. Demonstration of which muscle is paralyzed also aids in dif ferentiation, but in diseases such as polyneuropathy or myasthe nia gravis, many different muscles are involved and the defects become more complex. In all muscular
weakness, there is a tendency for the disability to increase as the patient struggles to speak, but in true myasthe nia the voice may be normal at the beginning of each sentence, but abnormalities develop as the sentence progresses. These may be of any of the types mentioned above, but particularly pala tal weakness and including hoarseness. A few
moments of rest will return the voice to normal. This is usually demonstrated by asking the patient to count up to 30. The abnormality is tempo rarily cured by injections of edrophonium chloride (Tensilon) or neostigmine. Mixed dysarthria It is a combination of any of the above types of dysarthria. For example, it can be a combination of spastic and
ataxic types where it is not easy to distinguish each type separately. Dysarthria in aphasic states A degree of dysarthria may accompany expressive aphasia, non fluent aphasia (see below), and may occasionally be found in lesions producing severe apraxia of the muscles of articulation. It can also be seen in lesions involving pyramidal, cerebellar or
basal ganglia in varying degrees of overlap. A combination of aphasia and dysarthria may accompany internal carotid artery thrombosis, if the other vessels in the neck are diseased as well. It must always be remembered that in mixed speech defects, aphasia is the most important localizing element, because it must mean a dominanthemisphere
lesion, whereas dysarthria follows lesions at many sites. 266 Chapter 23 Disorders of speech The analysis of aphasia An attempt will be made here to produce a relatively simple scheme of examination by which the main types of aphasia can be detected. Unfortunately, the lesions that usually produce aphasia are not such as to justify attempts at
pinpoint localization, and for similar reasons, though classification of aphasia aids in its compre hension, in clinical practice, a mixture of the different types is the rule rather than the exception. Preliminary information One must first know the patient's nationality, native language and educational level; Iris previous ability to read, write, spell and calculate; his handedness; whether, if right-handed, this is genuine or was enforced at an early age and whether there is left-handedness in the family. One must, of course, determine if there is any mental deterioration, hemianopia or hemiparesis which might confuse the tests. Anything up to 99% of right-handed individuals have lefthemisphere
dominance for language, but bilaterality may be pres ent in some. The majority even (perhaps 60%) of left-handed people may have left-hemisphere dominance, but bilateral dominance is probably quite common. The matter is still unclear. Spontaneous speech Always be on the alert for aphasia. Many aphasic patients find their way to psychiatric
departments because a doctor has not realized what it is he is hearing when listening to a patient's apparently incoherent spontaneous speech. The patients are merely described as 'confused'. Most information is gained from just listening to the patient. Let him try to tell his own story without his relatives interrupting. Is his speech 'fluent' or 'non-fluent'? Listen carefully to the construc tion of each word and sentence. Are these correct? Is he making sense? Is he using the right and appropriate words? Is he using wrong words or words which are nearly, but not exactly, right (par aphasia)? Is he using word forms that do not exist (neologisms)? Is he involuntarily repeating the same words after
having once used them (perseveration)? Is he using long sentences to overcome the failure to find a particular word (circumlocution)? At the same time, note if he understands the question asked and the instructions later given during the course of the ordinary exam ination. Perseveration may be evident both in verbal reply and in 267 Part 6
Examinations of particular difficulty response to commands, e.g. a patient who has just shown his teeth may do so again when told to place his finger on his nose. Comprehension Note whether the patient's reaction and answers to ordinary con versation are normal and appropriate. With the exception of some patients with 'conduction' aphasia, even
the most severely affected patients should be able to answer questions designed to require only an 'Yes' or 'No' answer. For example, one could ask 'is it true that it is raining outside'. Then the patient can be asked to point, on command, to name the objects around the room. Lastly, three objects (e.g. your pin, cotton-wool and reflex hammer) should
be placed before the patient. He is then asked to place the objects around himself in positions designated by the examiner. These tests should accurately assess comprehension even in patients with gross motor speech deficits and dyspraxia. Naming objects Explain to the patient that he is about to be asked the names of a number of common objects.
Start by showing such things as a watch, comb, key, pen or pencil. Then go on to the component parts of the object, such as the strap, buckle, winder and second hand of the watch. Note if he gives the right name promptly or hesitantly; if he uses paraphrasia, or neologisms, or if he perseverates. Use about 20 objects before deciding that no
abnormality exists. If he fails to give a name, can he choose the right one from a list of alternatives, or if shown several objects, can he pick out the object named? Make a note at the time of any errors. Repetition Testing the ability to repeat words or phrases is important and often overlooked (see below). Ask the patient to repeat a complex phrase
and if he fails, simpler phrases or even single words should be given. Reading Having assessed the patient's normal capacity in this respect in relation to age, education, nationality and visual acuity, give indi vidual words to be read out first, then sentences, then longer pas sages and finally instructions to perform certain actions. This will link the
sensory and motor sides. 263 Chapter 28 Disorders of speech Writing For all practical purposes, writing is always abnormal in a truly aphasic person. Aphasia is a disorder of language, not just speech. The patient may be able to write his name and address, but not con struct linguistically correct sentences. Ask him to compose a piece about the
weather, or his job, and test the ability to write to dicta tion. If he has a right hemiplegia, he must try to write with Inis left hand. Calculation Ask the patient to perform simple sums. Start simply by easy addi tion - 2 plus 4, 3 plus 8, and make this progressively harder. How many paise are there in a Rupee? How many 10-paise pieces in 1 Rupee? How
much would 10 stamps of 1 Rupee each cost? Ask him to subtract 7 from 100, then 7 from the answer and so on progressively down the scale. Most people of average educa tion can do this in 40 seconds or less. One mistake should be over looked. Listen carefully to their method of doing this, and avoid confusing inability to concentrate with inability
to calculate. Varieties of aphasia The first and most noticeable feature of aphasia concerns the dis ruption of spontaneous, conversational speech. By and large, patients are either 'fluent' or 'non-fluent' in their speech output. Basically, patients with 'fluent' speech will have a lesion posterior to the Sylvain fissure, those with 'non-fluent' aphasia a lesion
ante rior to the fissure. There are some exceptions to this rule, however, in that some patients cannot be so simply classified and that also some patients' aphasia is initially 'non-fluent' only later becoming 'fluent'. A typical 'non-fluent' aphasic will utter few words, will struggle visibly, will not be able to use long phrases and there is often asso ciated
dysarthria (see p. 264). In contrast, the 'fluent' patient may even be excessively loquacious, but his speech is full of mistakes: paraphasias, neologisms and circumlocution will be present, with very few norms. When the immediate peri-Sylvian region, both anterior and posterior to the fissure, is involved in a lesion, then one of the sig nificant
disturbances concerns repetition. The presence or absence of normal ability to repeat spoken language is now regarded as an important distinction. Thus, patients with abnormal repetition 269 Part 6 Examinations of particular difficulty form an identifiable category of aphasics which include those with lesions in Broca's area, Wernicke's area and
between the two, in the arcuate fasciculus. Conversely, aphasia with normal rep etition indicates pathology sparing the peri-Sylvian region, fur ther away from the fissure. Aphasias with preserved repetition are, similarly, of three types, all forms of the so-called transcorti cal disturbances. Lesions anterior to the fissure, but superior to Broca's area,
cause 'transcortical motor' aphasia with non-fluent speech and normal comprehension. Lesions posterior and infe rior to Wernicke's area cause 'transcortical sensory' aphasia, in which repetition is again preserved, but in the presence of flu ency and poor comprehension. If there has been a watershed zone infarction (often following sudden
haemodynamic disturbances of perfusion), then the whole area surrounding the peri-Sylvian regions may be infarcted, and the latter becomes 'isolated' from other cortical structures. The aphasia that follows is non-fluent, comprehension is also poor, but repetition is still possible leading to persistent echolalia. Finally, it would have been noticeable to
readers that the above remarks do not include the words 'expressive' or 'receptive'. This is because firstly, as already emphasized in previous editions, most aphasias exhibit difficulties both of expression and compre hension of language, but also because the more recent classifica tions of speech disorders stress the importance to the clinician,
concerned as he is with anatomical localization, of considering patients' speech as being 'fluent' or 'non-fluent', with or without preserved repetition. On tills basis, the main types of aphasia can be defined. Broca's aphasia Speech is non-fluent, even to the point of mutism in extreme exam ples, with preserved comprehension and poor repetition.
Naming ability is also poor, and there is a distinct breakdown of correct syntax (grammar). The patient may have several words, which are used repeatedly and incorrectly to convey an astonishing variety of meanings. The usual cause is an infarct in the territory of the anterior part of the left middle cerebral artery. The anterior insular is probably
more often damaged than Broca's area itself. A patient encountered by Dr. Bickerstaff was only able to say 'South Af rica vica me' and yet by the use of this phrase was able to ask for a bedpan, complain about the food and indeed carry on a useful degree of conversa tion. A skill of this type indicates that the defect has been present for a very long
time. 270 Chapter 28 Disorders of speech Wernicke's aphasia Lesions in Wernicke's area result in a fluent aphasia, but with pro foundly affected comprehension and impaired repetition. 'Nam ing' is poor. The patient, failing to produce correct words or syntax, so fails to comprehend his own errors that he produces a jumbled string of meaningless
words and neologisms, in fact - jargon. Hence the older term 'jargon, aphasia'. Posterior left middle cerebral artery territory infarction is the commonest cause with damage to the posterior third of the supe rior temporal gyrus. Conduction aphasia As mentioned above, a lesion that disrupts the arcuate fasciculus connecting Broca's and Wernicke's
areas causes a third variety of aphasia characterized by poor repetition. Speech is fluent and paraphasic because Broca's area is disconnected, and comprehension is also relatively spared, thus distinguishing this aphasia from Wer nicke's type. But the distinction is not always absolutely clear and true conduction aphasia is probably rare. Transcortical
aphasia This is characterized by retained repetition, and subdivided accord ing to fluency and comprehension into 'transcortical motor' and 'transcortical sensory' types, as above. Stuttering and stammering This is too familiar to need description. When affecting children, predexterity or with the enforced use of the right hand in a left-handed child.
How much it has a psychogenic basis, and how much associ ated psychogenic features are secondary to the stutter, is still uncer tain, but there is no doubt it can occur as a manifestation of a mild aphasia, not infrequently in the recovery phase of a definite aphasia. There are other forms of speech disturbance considered formerly to have
localizational significance. Recent experience puts this in considerable doubt, but the terms have become familiar, and for the sake of completeness are included below, with the locations origi nally thought responsible. Echolalia This is the involuntary repetition of words and phrases, spoken by someone else without the intention of doing so, and
without 271 Part 6 Examinations of particular difficulty understanding their meaning. The lesion lies in the temporopari etal region. Apraxia of speech This form has the main features of effortful speech with trial-anderror attempts at speech and corrections, lack of prosody and varia tion in articulation errors and difficulty in initiating speech. Tire
testing is done by repetition of a set of phonemes (p /t/k). Rang ing from total inability to articulate to mere slurring of speech, this uncommon defect is accompanied by normal reading, writ ing, copying and ability to tap out syllables. The muscles unable to articulate are able to perform their other functions. This is an apraxic speech defect and the
lesion may be widespread in the left hemisphere. Pure word deafness In this, spontaneous speech, reading and writing are normal, but the patient cannot recognize the spoken word, even as words or as a familiar language. This is a rare defect and is due to a lesion in the middle of the first temporal gyrus. Alexia This, the inability to understand
written speech, is further divided into alexia without agraphia, or pure word blindness, and alexia with agraphia. Pure word blindness is rather rare. Tire patient is able to speak, under stand and write, both spontaneously and to dictation, but cannot understand or copy the written word. It is an example of a 'discon nection syndrome', similar to
conduction aphasia already described. In this instance, a lesion of tire left occipital cortex and splenium of the corpus callosum 'disconnects' tire language area of the left hemi sphere (in right-handed people) from both the visual cortices. There is an associated right homonymous hemianopia, and the usual cause is infarction in the territory of the
posterior cerebral artery. Alexia zvith agraphia, also known as visual asymbolia, is very much more common. The patient cannot read, or write spontaneously with either hand, but may be able to copy to some extent. The lesion lies in the left angular gyrus, and is usually accompanied by nomi nal aphasia, acalculia, hemianopia and possibly visual
agnosia. Developmental dyslexia is usually seen in children as a slowness in learning to read. Their vision being normal, they may be thought to be mentally retarded, though intelligence can be quite normal and Chapter 28 Disorders of speech all other speech functions normal, and mirror writing may be pres ent. The child may indeed try to read
words from right to left. The defect may be familial, and may possibly be due to an incomplete left-hemisphere dominance. Agraphia As one of the symptoms of other aphasic states, writing defects are common. A pure agraphia, without the other features of speech dis turbance, may occur in lesions that lie between the angular gyrus and the motor
area, but may sometimes be anterior or posterior to tills. An apraxia of the hand, due not to a disturbance of language so much as to an inability to make the correct movements for writing, produces a form of dysgraphia, the lesion lying in the region of the left angular gyrus, or if limited to the left hand, in the corpus cal losum. Acalculia The recent
development of an inability'' to manipulate figures may be part of any of the major aphasias. If acalculia is not associated with other speech defects, it indicates a lesion of the left angular gyrus (see Gerstmarui's syndrome). It will be seen that there is considerable overlap between these dif ferent states and the sites of the lesions responsible. Where
tumours are concerned, the lesion is usually much larger than the type of speech defect might suggest. At other times, the position of the main bulk of a lesion appears rather far from the area known to produce particular defects. It must be remembered that tumours, for instance, have remote effects due to displacement, vascular com pression or
oedema, as well as the effects of direct local invasion. For these reasons, attempts at pinpoint localization from dysphasic defects often become frustrating to the perfectionist because the pathological process involves, directly or indirectly, so wide an area. Computerized axial tomography sounded the final death knell of the would-be pinpoint localizer
- but the academic intellectual exercise in the attempt still has its merits in the logic of neurological appraisal. Stroke syndromes, on the other hand, are more precise and do allow very accurate localization. No apology is made for repeating the necessity of detecting apha sia in any speech disturbance - because the lesion responsible for this lies in
the dominant cerebral hemisphere, no matter what other lesions the patient may have as well. 29 Apraxia Apraxia is a failure of the ability to carry out well-organized vol untary movement correctly despite the fact that motor, sensory and coordinative functions are not significantly impaired. It is not the word, as is the modern tendency, to delineate
any improper motor response resulting from paresis or incoordination. Methods of testing First make sure that if there is any weakness, sensory defect or ataxia, it would not interfere seriously with voluntary movements, and that the patient understands instructions. Ask him to hold out his arms, put out his tongue, show his teeth etc. If he fails, note
whether, despite this, these movements are normal when they are automatic, e.g. licking the lips, smiling or responding to an obvious offer to shake hands. Next, ask him to make a fist at someone, to scratch his arm and, more difficult, to use a pair of scissors, a pen and a comb, making sure first that he recognizes these things. Ask him to open a box
of matches and to take out a match; to go through the motion of lighting a cigarette; to show how he would hammer a nail, play a violin, put in a corkscrew and take a cork out of a bottle. Any test requiring three or four different movements can be used, both with and without objects. Give him a series of matches and ask him to form a triangle, a
square or more complex patterns. If this appears difficult, do it for him and ask him to copy it. Ask him to write his name and address. Note how he manipulates buttons or studs; how he takes off his coat and jacket and puts them on again. Form s of apraxia Four types of apraxia are usually recognized, the first two of which cause most difficulty
because their names are similar and the differ ence between them is a fine one. Ideomotor apraxia The patient performs automatic acts normally, such as blowing his nose, shaking hands, pushing back hair etc., and is able to formulate 274 Chapter 29 Apraxia the idea of an act and to describe how it should be done, but when it comes to carrying out
the movement on command, he is unable to do it correctly. This is probably the commonest form of apraxia, and usually just referred to as 'apraxia'. The basic disturbance is an inability to imitate or mime an act involving the use of objects. There is a common tendency to substitute a body part fo r the object, e.g. using the index finger as a toothbrush
rather than pretending to hold one. Ideational apraxia The formulation of the method of carrying out the whole of a complex act is defective, though the execution of different parts of the complete act may be normal. The popular description is of the patient who, when told to do each of these actions separately, will take a match box, hold it correctly,
open it correctly, take out a match correctly, and do the same with a cigarette box, but when told to go through tine motion of lighting a cigarette, will be unable to do so or will try some manoeuvre such as striking the cigarette on the match box. This form of apraxia, therefore, is recognized by the patient's inability to use objects properly and not
tested for by imitation or miming. In clinical practice, ideomotor and ideational apraxia frequently coexist. Constructional apraxia The patient is defeated by attempts to make designs with, for instance, matches, either spontaneously or by copying. Often in writing he cramps everything into one small corner of the paper. Koh's blocks is a series of
blocks with colours occupying the whole or half of one side, which can be so arranged as to make simple or complex patterns. The patient with constructional apraxia is unable to make or copy the simplest design. The side of the figure opposite the lesion may be omitted altogether. Dressing apraxia The patient becomes hopelessly muddled in trying
to dress and undress, puts clothes on the wrong way round, or may be quite unable to start the necessary motions. This defect, which is also possibly present if ideational apraxia is present, may be found entirely by itself. Lesions producing ideomotor apraxia interrupt connections run ning between the dominant parietal cortex and the motor area,
and also with the opposite motor area through the corpus collosum. Therefore, a lesion situated: Part 6 Examinations of particular difficulty 1 In the dominant supramarginal gyrus should produce bilateral apraxia. 2 Between this gyrus and the left motor cortex should produce right-sided apraxia (in right-handed people). 3 In the corpus callosum
could produce left-sided apraxia. In practice, however, the lesions are often more diffuse, though the dominant parietal lobe is most heavily involved. Construc tional apraxia occurs in angular gyrus lesions of either hemisphere, but when isolated is usually from non-dominant parietal lesions. Dressing apraxia may also occur in a non-dominant
posterior pari etal lobe lesion. Vascular and degenerative diseases are more often responsible than neoplasms. 30 Agnosia and disorders of the body image Agnosia Agnosia, though a remarkable phenomenon, is not an unfathom able mystery, and it is, in short, a failure to recognize some object or sound when the sense by which it is normally
recognized remains intact. This defect may be present in the field of any of the normal sensations, but agnosia for smell and taste are doubtful entities. Examining for agnosia Visual recognition First show the patient a number of common small objects. Ask him to (i) name them, (ii) describe their use and (iii) pick out others named for him. If he is
unable to do so, allow him to feel the object in the hand in which the sensation is normal and ask the same questions. Next, show the patient several different colours and ask him (i) their names, (ii) to pick out duplicates from a separate set and (iii) to arrange them in shades of increasing lightness. Now show him some familiar objects, holding them
in one or the other homonymous visual field. If the patient is well enough, ask him to walk towards a par ticular point, having placed some chairs in the way, and observe whether he is able to find his way to the correct place and around the obstructions. Visual agnosia 1 The patient is unable to name or describe the use of the objects shown, but is
able to identify them when he touches them, or by their characteristic noise or smell. (A patient with nominal aphasia will be unable to find the exact name, whichever sense is used.) This defect is called visual object agnosia. It may vary according to the size of the objects, the patient's environment and even from examination to examination. 2 The
site of the lesion lies in the second and third occipital gyri and the adjacent subcortical white matter in the dominant hemisphere. 277 Part 6 Examinations of particular difficulty 3 The patient may be unable to identify, match or arrange colours. This is often associated with object agnosia and is termed visual agnosia fo r colours. The lesion is in a
similar situation. 4 The patient may be unable to find his way around an obstruc tion or to find his way to a given point. This is visual agnosia fo r space and results in visual disorientation. It is most marked in bilateral posteroinferior parietal lesions, but unilateral lesions can cause an agnosia for one-half of space, so that the patient will turn always to
the opposite side and is liable eventually to return to the point of starting. This is most clearly demonstrated when the non-dominant hemisphere is affected. Tactile recognition First ensure that sensation is normal in both hands. Ask the patient to close his eyes and then place a number of common objects in turn in one or the other hand. Ask him to
name them, to describe their shape, size and texture and to indicate their use. If he fails to do this, allow him to look at them or to hear or smell them, and see if he is then able to recognize them. Tactile agnosia The patient may be unable to recognize any detail of the object at all. In such a case, it is probable that discriminative or postural sense is
defective, and the defect becomes one of astereognosis. If he is able to describe its size, shape and texture, but is not able to give its name or its use, and particularly if on seeing it he is then able to name it, this is true tactile agnosia. It is a defect that com monly coexists with visual object agnosia. The site of the lesion is in the contralateral
supramarginal gyrus. It is possible that left-sided agnosia in a right-handed person may also be caused by a lesion of the corpus callosum. Auditory recognition First determine that the hearing in both ears is normal. Ask the patient to close his eyes and to identify sounds made by striking a match, ringing a bell, shaking money, tearing cloth etc.
Auditory agnosia The patient who is unable to recognize these sounds, but can rec ognize the objects on sight or touch, has auditory agnosia. As there is often word deafness as well, it may be necessary to write out the 278 Chapter 30 Agnosia and disorders of the body image instructions. If there is an associated aphasia which might confuse Inis
answers, he should be told to illustrate the sound himself, if he recognizes it. Tire site of the lesion is in the posterior part of the temporal con volutions of the dominant hemisphere. The parietal lobe and disorders of the body scheme A normal individual is able, without looking or having to give much thought to the matter, to tell at any moment where
each part of the body is, and where it lies in relation to objects around it. If there is a defect of postural sensibility, some other sense, such as vision, may be necessary to determine where the defective limb lies. There are situations, however, when, despite the use of other senses, the normal appreciation of the shape, size and position of one or the
other part of the body is disturbed, sometimes to such an extent that if that part is diseased the patient may be quite unaware of it, and may even deny that it is part of Iris own body. Examination First determine if the patient knows which is his right and left hand and leg. Then ask him to point to different major parts of his own body, using each hand
in turn if possible. Next ask him to point to the ring finger of his left hand, the forefinger of his right hand, to point to the little toe etc. Make this more difficult by asking him to place his fingers behind his back and pick out individual digits. Ask him then to pick out individual digits on the examiner's hand, and filially, the examiner should interlock his
fingers with the patient's and ask him to pick out the various digits. During the course of the general examination, observe whether the patient does or does not appear to be aware of any disability, such as a hemiplegia, which may be present. If he seems unaware of this, draw his attention specifically to the paralyzed limb and ask him to explain his
inability to move it. Disturbances of the body scheme Difficulty in remembering which is right and left is a common prob lem especially if the question is suddenly asked, but when once reminded, most people continue to answer correctly afterwards. Persistent right-left disorientation is seen in parietal lobe disorders. 279 Part 6 Examinations of
particular difficulty Patients may be unable to identify any part of their body. This is termed autotopagnosia. A lesser degree of this is found in the inabil ity to pick out individual digits and is termed finger agnosia. Gerstmann's syndrome, which consists of finger agnosia both for the patient's own fingers and the examiner's, acalculia, right-left
disorientation and agraphia without alexia, is found in lesions of the dominant hemisphere, in the region of the angular gyrus. hi lesions of the parietal lobe of the non-dominant hemisphere, which present an easier situation for examination as there is less intellectual defect and no aphasia, there may be complete lack of awareness of the opposite half
of the body. If that side is paralyzed, the patient may be unaware of, and even deny the existence of, the hemiplegia. He may say the limbs do not belong to him and may even complain that the arm in Iris bed belongs to someone else. This is known as anosognosia, and may exist in relation to hemiple gia, blindness, deafness or any other disability. At
the other end of the scale, there is the phenomenon of the phantom limb, so common in amputees as to be considered normal, which is a retention of the whole body image after the removal of one member, a retention sufficiently vivid for pain and parasthesiae to be felt in the fingers or toes. Over the years, the phantom tends to shrink in size.
Sensory inattention Neglect of one side may be present, but not in the gross degree described above. It is then necessary to present to the patient simul taneous bilateral stimulation of one of the common senses in order to detect the defect which we call 'inattention'. Visual inattention First determine by confrontation that the visual fields are
individu ally intact. Then face the patient, both the patient and examiner having both eyes open. Place the forefingers of both hands so that they can be seen in the patient's temporal fields equidistant from the midline. Ask the patient to point to the finger he sees moving. Move first one finger, then the other (not alternately) and finally, move both
together; the patient with visual inattention on one side will point to one finger as if that one alone had moved. Next use movements of the whole hand. If the attention defect still persists, tell the patient positively that at times both hands will be moved together. This will make no difference in the fully developed case. 280 Chapter 30 Agnosia and
disorders of the body image Now move both hands for a few moments and then stop moving the hand on the normal side. The patient may, after a short delay, appreciate the movement on the abnormal side which he had pre viously failed to do. Ask the patient to draw some symmetrical object, such as the head of a flower, a house, a clock face or a
bicycle. If necessary, give him a model to copy. If defeated by complexities of this type, simple geometrical designs could be copied. The patient with an attention defect will leave out, or incorrectly space out, the petals on one side of a flower, the figures on one side of a clock face, one wheel of a bicycle, the window on one side of a house or he may
fail to join up the angles to one side of a rectangle. If there is an associ ated constructional apraxia (see p. 275), this test may, of course, be completely impossible. Auditory inattention First determine that hearing in each ear is approximately equal. Then ask the patient to close his eyes and to point to the side from which he hears a noise. Shake a
bunch of keys, irregularly, first on one side and then the other and every now and then on both sides simultane ously. If auditor}' inattention is present, only the sound from one side will be appreciated. At times, unequal sounds can be used, quite a slight sound being heard on the normal side, while a loud soimd is simultaneously produced, but not
appreciated on the abnormal side. Tactile inattention First determine that the patient's superficial sensation is normal, and that he is able to recognize right from left. Then ask him to close his eyes and, with a pin, prick one or the other arm at irregu lar intervals. He should say, or move the arm that is being stimu lated. Assuming he locates the side
of these pricks correctly, then prick on both sides simultaneously. A patient with tactile inatten tion will indicate one side only. The stimuli can then be changed to stroking, pinching and quite painful tapping of a limb, and still only one side is appreciated. Ask the patient to hold both hands out, keeping the eyes closed, and to say into which hand an
object is placed. Put similar objects, e.g. oranges, into each hand simultaneously. The patient with tac tile inattention will only appreciate one. Take this one away and after a short time he will draw attention to the other. Note how he puts on his coat and jacket. He may be entirely satis fied with a situation in which one arm is left out. If dressing
apraxia is present, this is only of value if it is consistently on the same side. 281 Part 6 Examinations of particular difficulty This phenomenon of inattention is also described as 'extinction'. It is most commonly seen after vascular accidents, in cerebral atro phic lesions, sometimes in parietal lobe tumours, including meningiomata, when it may recover
after removal, and rarely in other conditions. Isolated cases have been described in which a similar phenomenon is present in lesions of a large variety of other parts of the nervous system, but usually it can be taken to mean a lesion of the parietal lobe of the contralateral hemisphere. 31 ¥Eie autonomic nervous system Only a limited amount of the
mass of information available regarding the autonomic nervous system (ANS) is applied with any frequency during the practice of clinical neurology. However, recent years have seen the evolution of several simple and noninvasive tests of both sympathetic and parasympathetic function. These can be applied with reliability using only straightforward
equipment and requiring little time. Detailed testing of auto nomic function remains highly complex and is really the pre serve of specialized units. New advances have revealed a system abounding with neurotransmitters within a very complicated neural organization such that full investigation involves exper tise in physiological, pharmacological and
biochemical labora tory techniques. Whereas the ANS is concerned with virtually every organ in the body, from skin to brain, it has been in the study of cardiovas cular reflexes that tests of a 'bedside' nature have been recently devised. That is not to forget, naturally, the many autonomic reflexes that are examined within the cranial nerves (see
pupils, lacrimal response, salivation etc.). Bladder and bowel dysfunction is prominent amongst the manifestations of autonomic failure, and there are also the changes in respiration, skin and sweating to record. General inspection Note the colour of the skin, especially of the extremities, both when first seen and after exposure. Look for local or
general flushing or cyanosis, and feel the temperature of the skin in dif ferent parts of the body. Recent sympathetic lesions produce warmth and redness in the affected areas. Note whether sweat ing is unexpectedly profuse or abnormally slight, and if there is a localized abnormality, what its distribution is in relation to sen sory dermatomes.
Complete sympathetic lesions result in the absence of sweat ing, but partial lesions may produce excessive sweating. Notice any changes in colour, texture and quantity of hair or nails. Look especially for signs of any well-recognized local autonomic lesions, such as Horner's syndrome (p. 68). Part 6 Examinations of particular difficulty Cardiovascular
reflexes The simpler physiological tests here are based upon the responses of the heart rate and blood pressure to various stimuli. Sympathetic and parasympathetic function is involved. Postural hypotension A fall in systolic BP below 20 mmHg and diastolic BP below 10 mmHg of baseline within 3 minutes in upright position is abnor mal. The causes
can be central or peripheral; if the latter, severe sympathetic dysfunction is present. Blood pressure response to pressor stimuli Mental arithmetic, sustained handgrip or exposure to cold cause a rise in blood pressure. The use of a handgrip dynamometer allows a quantifiable test. Less than 10 mmHg rise in diastolic pressure is abnormal. Both
central and peripheral (sympathetic) lesions affect these tests. Heart rate responses 1 In the normal person, a change of posture from lying to stand ing is followed by an immediate increase in heart rate and then a relative bradycardia. This can be quantified by using continu ous electrocardiograph (ECG) recording. A parasympathetic lesion will slow
down or abolish the response, a situation found iir diabetic or other autonomic neuropathies. 2 With an intact parasympathetic nerve supply, the heart rate var ies with respiration (sinus arrhythmia). This is reduced or abol ished iia the presence of an autonomic neuropathy. The valsalva manoeuvre Here, there is a response in both blood pressure and
heart rate, hr the first exhalation (against a closed glottis), the blood pressure drops and the heart rate increases. On releasing (by opening the glottis), the blood pressure 'overshoots' the resting value and the heart slows. The test can be performed by the patient exhaling into a mouth piece connected to a manometer or sphygmomanometer to hold
the pressure at 40 mmHg for 15 seconds. An ECG records the heart rate response. Afferent or efferent parasympathetic lesions impair the response. (Patients with lesions at the foramen magnum such as cerebellar ectopia develop a characteristic headache.) Non-invasive monitoring of finger arterial pressure has been an important recent advance
useful in all the above tests. They can be augmented by biochemical or pharmacological methods. Intravenous infusions of noradrenaline and atropine test for 284 Chapter 31 The autonomic nervous system sympathetic and parasympathetic function respectively. Plasma noradrenaline and renin can be measured directly. In practice, however, the
response to standing up is the single most useful test and all manner of other complex computations add remarkably little to the evaluation of an individual patient's problem. Skin resp o nses Erythema Scratching the skin will produce a line surrounded by a spreading flare and followed by a wheal with central pallor. This is often exag gerated below
the level of a transverse cord lesion, and, according to the lesion, may be unilateral or bilateral, often becoming appar ent after testing cutaneous sensation to pinprick. At the level of the lesion, even the normal response may be absent. Exaggerated responses, unfortunately, can occur in normal or psychoneurotic individuals - this 'dermatographia' is,
however, not simply limited to the skin below a segmental level. Temperature The measurement of skin temperature by means of accurate skin thermometers can show: 1 Increased temperature below a recently denervated level. 2 Decreased temperature following chronic neurological lesions which have caused prolonged immobility. 3 Increased
response to warming of the part or of distant parts over the denervated area in sympathetic lesions; but. 4 But decreased response to warming in areas where the deficit is circulatory rather than neurogenic. Pilomotor responses These are similar in significance to the erythematous responses. They are produced by sharp scratching, or by touching the
warmed body with cold metal. Scrotal response Touching the scrotum with a cold object normally results in a ver micular contraction of the dartos, without elevation of the testicles, thus differing from the cremasteric reflex. This is absent in sympa thetic paralysis. A sweating te s t Spoon test may be performed to check the adequacy of sweat ing. A
spoon is slid over the part to be tested and the resistance Part 6 Examinations of particular difficulty encountered is subjectively assessed. In areas of normal sweating, the spoon will encounter some resistance. On the contrary, in case of reduced sweating, the spoon moves easily over the skin. Alter natively, the areas to be tested are first thoroughly
dried, and then liberally dusted with Quinizarin powder. The patient is placed under a heat cradle, and given a hot drink combined with 0.5 g acetylsalycilic acid. Areas of sweat production are clearly outlined as the powder turns black when exposed to moisture. In sympathetic lesions, there will be a segmental loss of sweat ing corresponding to the
distribution of the affected sympathetic fibres. Below a transverse cord lesion, sweating may be absent in the early stages, but may later become abnormally profuse. At the segmental level of a recent transverse lesion, the response is often exaggerated. This is a good test for the results of sympathectomy, of section or injury to peripheral nerves and
for demonstrating and photograph ing denervated areas. It can show, objectively, areas of sympathetic denervation in syringomyelia. It is a capricious test, however, and requires a lot of experience before it can compare in value with the assessment of cutaneous sensation for the purpose of determining the level of a cord lesion. Examination of
bladder function Normal urinary continence and voiding requires a balance between the so-called forces o f expulsion and the forces o f retention. Detrusor and abdominal muscle contraction, on the one hand, needs to be balanced against the activity of the bladder neck (internal) and urethral (external) sphincters on the other. This balance does not
depend exclusively on nervous control - there are physical factors involved also, such as the inherent distensibility of a given detru sor muscle. Thus, not all disorders of micturition are the result of faulty nervous control; but they are certainly common in neurologi cal disease. 0 Lesions of the superior frontal and anterior cingulate gyri, whether the
result of stroke, tumour or hydrocephalus, reduce awareness of bladder function and cause incontinence. 9 More posterior lesions of the frontal lobes may cause spasticity of the striated muscle (external) sphincters and therefore pro duce retention. ° Incomplete lesions of the upper motor neuron pathways above the sacral spinal cord interfere with
the storage and voiding of urine. Frequency and urgency of micturition are the rule, with incomplete bladder emptying. 286 Chapter 31 The autonomic nervous system 0 A complete transection of the spinal cord in the first place causes retention with 'overflow incontinence' and then later a 'reflex bladder' may develop with automatic emptying
induced by abdominal pressure. 0 Lesions of the sacral cord itself or the outflow in the cauda equina lead to a flaccid neurogenic bladder which enlarges and again can cause 'overflow incontinence'. With impaired para sympathetic function in the sacral segmental supply, sensation is lost in the S2-S4 dermatomes. 0 hi tabes dorsalis, damage to the
sympathetic and parasympa thetic sensory side of the reflex arc results in painless bladder enlargement. In investigating bladder function, the exact history is abso lutely vital. All but the more complex problems can be reasonably assessed with a good history and appropriate neurological exami nation. Some questions would include: Is there normal
sensation of bladder filling and voiding? Can voiding be interrupted vol untarily by sphincter action? What is the force of the stream like? When does incontinence occur? The examination, naturally, should always include testing the sacral segments (see, Fig. 20.1). Bladder swelling may be palpable. The superficial anal reflex should be tested. The
condition of the motor and sensory pathway to the lower limbs will testify to any spinal cord, conus medullaris or cauda equina lesion. A useful assessment of detrusor function can be made by asking a patient while passing urine to breathe deeply, thus interrupting abdominal muscle straining. If the detrusor is not functioning, the flow will cease.
Residual urine can be assessed by a post-micturition ultrasonog raphy of the bladder. Urodynam ic studies In the narrower sense of the word, 'urodynamics' refers to the investigation and assessment of the bladder and lower urinary tract. In its broader concept, we now have a whole new discipline embracing the total management of bladder
disorders. Rather like good neurophysiological investigation of, say, a case of suspected muscle disease, the ideal urodynamic assessment is tailored to the clinical situation. The essential requirement, however, is to measure intravesical pressure during bladder filling (the 'filling cystometrogram'), and the relationship between intravesical pressure
and urinary flow rate during voiding (the 'micturition cystometrogram'). Sophisticated 287 Part 6 Examinations of particular difficulty urological departments will now use a four-channel recording of abdominal pressure (via rectal catheter), total bladder pressure (via bladder catheter), subtracted true intravesical (detrusor) pres sure and flow rate.
The filling volume and voiding volume will be recorded. A note is made of the patient's awareness of bladder sensation, the capacity before a strong desire to void is appreciated, the compliance of the bladder (the volume change for a given pres sure) and the contractility of the detrusor (e.g. spontaneous contrac tions before complete filling). If the
bladder is filled with contrast medium, then video-radiological screening is possible, detecting abnormalities of structure and visualizing the bladder contrac tion and sphincter mechanisms. These can then be interpreted alongside the pressure-flow measurements. In this way, the assess ment of neuropathic bladder dysfunction has become much
more detailed and accurate. Surgical intervention or other treatments can be advised on a more rational basis. The pressure in the normal almost empty bladder is about 1 cm H ,0 , and if quantities of about 50 ml are introduced at a time, the pressure steadily rises, with rhythmical variations as the bladder wall accommodates. Vesical sensation is felt
at about 100-150 ml when a pressure of about 6 cm H;0 should be registered. The bladder can usually be distended to 400-600 ml. The rhythmical contractions which are accompanied by a slight sense of urgency will build up until reflex micturition occurs accompanied by a sharp rise in pressure. This may, however, not occur in patients who are
tense and nervous. If the bladder is atonic and known to be insensitive, great care must be taken not to overdistend it and risk rupture. Results These are expressed in terms of the observed bladder sensation, capacity, contractility and compliance together with the pressure and flow measurements during filling and voiding. A 'functional profile' is

thus obtained. • In a complete lower motor neuron lesion, one finds a senseless, high capacity (flaccid), acontractile and hypercompliant blad der with low pressure. Voiding requires straining. • In a complete upper motor neuron lesion, one finds a senseless, low capacity (spastic), hypercontractile (hyperreflexic) and hypocompliant bladder with high pressure. There will be a hyperactive sphincter, and reflex voiding. • In practice, lesions are often not complete; they can be 'mixed' and the urodynamic findings can become modified by local Chapter 31 The autonomic nervous system changes in the bladder wall, chronicity of disease and the degree and duration of any bladder distension. However, some generalizations can be made: ® In bilateral lesions of the pyramidal system of some standing, i.e. cord or brainstem lesions, the bladder is small in capac ity, contracts rapidly on stimulation, so that discomfort is felt early, but the sensation is poorly localized. In recent lesions o f the spinal cord, there is often a period of paralysis of bladder musculature with atonia and easy distension before these fea tures develop. 3 In lesions of the cauda equina, conus medullaris, or peripheral nerves, there is loss of bladder sensation, the pressure is high, the capacity not greatly increased and contractions may occur infrequently. These may be large enough to cause evacuation, which, however, is incomplete. ° In severe lesions of the sensory roots or posterior columns, there is total loss of bladder sensation, the muscle is atonic, distends easily so that the pressure is low, contractions are absent, evacu ation is absent and the residual urine high, ha such cases, great care must be exercised. ° In cerebral lesions, the results are variable. There may be reten tion or incontinence at the outset, but as the cerebral lesion recovers, urinary symptoms also recover. Bladder sensation is normal, but the capacity is reduced and micturition is precipi tate. Retention with overflow is a rare finding in high spinal or cerebral lesions. ° Lesions of the dominant hemisphere, demyelinating lesions, and some diffuse cerebral lesions result in the so-called uninhibited bladder, in which the contractions, capacity and sensation are normal, but micturition occurs precipitately as soon as contrac tions reach a size large enough to produce evacuation. As a group, patients with multiple sclerosis often develop a 'mixed' picture of disturbance. The typical pyramidal tract-type disorder of bladder function may be combined with external sphincter occlusion (spasticity or dyssynergia). There will follow a combination of frequency and urgency of micturition with a ten dency to retention. Alternatively, the detrusor itself may be weak, with low intra vesical pressure. Symptoms will then vary in accordance with the state of the sphincters. Some patients will tend towards inconti nence, others towards retention. It will be seen that in any one given patient, a variety of mechanisms may underlie an appar ently simple complaint. Full urodynamic evaluation is then essential. 289 Part 6 Examinations of particular difficulty The rectum Acute lesions of the spinal cord, particularly of the sacral segments, cause laxity of sphincters and incontinence. If the sacral segments or the pelvic nerves are involved, the gripping of a gloved finger by the internal anal sphincter will be absent even in the chronic state. Stroking the skin near the external sphincter will not produce reflex contraction. Higher spinal lesions or pontine lesions of chronic type usually allow tonic contraction of the sphincters and result in con stipation. Sexual function Either diffuse autonomic failure, local spinal or root lesions may interfere with sexual function. The history alone may be of consid erable diagnostic value. There are occasions when potency is dis turbed before bladder function in a male with a developing spinal cord disorder. Relevant questions include enquiring whether erec tion is absent or occurs normally, or whether just reflexly (noctur nal tumescence)? If ejaculation is present, is it of normal force or not? Is orgasmic sensation normal? ° Proper, psychically induced erection is a function of the para sympathetic hypogastric nerves. Reflex erection is mediated by the pelvic nerves from sacral roots. Spinal lesions may cause initial priapism (reflex painful erection). Subsequently, spinal disorders above T12 may cause impotence with retained reflex erection but without ejaculation. 0 A complaint of impotence with retained reflex erection and ejaculation is characteristic of a psychological disorder. (Never forget the many drugs that may cause impotence.) Use may be made of both the cremasteric and scrotal reflexes (p. 228) in the assessment of impotence. Rarely performed, but also valuable, is the bulbocavernosus reflex. A pinch or stroking stimulus to the dorsum of the glans penis causes palpable contraction of the bulbocavernosus muscle at the base of penis or contraction of the anal sphincter which is best per ceived with a gloved finger in the rectum. Cauda equina lesions will usually result in failure of both erection and ejaculation with impaired sensation. 290 Part 7 The Investigation of Neurological Problems (Excluding Neuroradiology and Imaging) 32 rards a balanced attitude (some introductory observations) Over the last few years, there have been gradual changes in the attitudes of clinicians towards neurological investigations, which on the whole have been all to the good. However, a dark cloud is looming on the horizon warning of a potential further change which could be a serious reversal to the thoughtful physician, and to neurology as a whole. In the past, the neurologist had few measures at his command outside his clinical acumen, a number of X-ray procedures, the ability to do certain relevant blood tests and to examine the cerebrospinal fluid. So big a part did this latter element play that in the first edition of this book it was implied that the indications for lumbar puncture would be any neurologi cal condition in which the clear-cut contraindications did not apply. This is no longer an acceptable dogma either in respect of lumbar puncture or of any other investigation. As more diagnostic measures become available, one needs to ask oneself even more closely, 'Why, in this particular patient, am I sug gesting this particular investigation? Is it truly likely to give rel evant information, and is the information so obtained going to be of value in the management of the case? Indeed, is it so important to know that information, that if the investigation carries with it some risk to the patient, is that risk justified'? It often is (maybe in experienced hands usually is), but sometimes it is not, and if it is not justified, then it should not be carried out. This line of thought has brought new terms into medical lan guage. One that is heard on all sides is the classification of pro cedures into 'invasive' and 'non-invasive'. While all of us know more or less what we mean when we use these terms, they seem sometimes to have rather artificial definitions. For instance, to put a needle into an artery is invasive; to put it into a vein apparently is non-invasive (or sometimes the thoughtful person says 'relatively' non-invasive). Presumably, a skin electrode for electromyogra phy is non-invasive, an intramuscular electrode invasive. Shining a light in the eyes is non-invasive, but is it non-invasive to flash a stroboscope during electroencephalography to stimulate corti cal responses, and maybe on a rare occasion to precipitate a major epileptic fit? Part 7 The investigation of neurological problems Some sense of proportion needs to be brought to the use of these terms, because it is the implications of the very terms them selves that could be a foretaste of the further change of attitude referred to earlier. This is the development of the so-called defen sive medicine - i.e. doing everything one can think of in the way of investigation for fear of not doing something that might just con ceivably have been important, the fear being these days, unhap pily, not so much of missing information that might be of value to the management of the patient, but that of being exposed at some later date to litigation for not having done it. This is a major prob lem in some countries whose legal system encourages actions for so-called negligence against doctors, with the potential for enor mous damages if found proven; and, like a malignant growth, it is spreading. The total paid out in malpractice litigation in the US reached billions of dollars in the mid-1980s. In the UK, negli gence claims cost the National Health Service 150 million pounds annually. In those parts of the world where this fear has as yet not become so prominent, there is gradually developing a desire to find and limit methods of investigation to those which are of the least dis comfort to patients, and which expose them to minimal danger of complications. The trouble here is that these methods may or may not be as good at demonstrating what is required as those that are older and well established. So, one has to be convinced that one has used not only a relevant method, but one that has been enough. Nowhere more than in the field of neuroradiology does this apply, and each 5-year period has seen the most dramatic advances in this field. (It is felt that to cover advances in neuroradiology since the last edition is beyond the scope of this book and therefore omitted. Readers are advised to refer to a standard textbook in the subject.) With regards to intracranial lesions such as tumours, haematomata, infarcts, oedema, abscesses and atrophy, computerized tomography (CT) scanning is now standard. Most district general hospitals, let alone regional neuroscience units, will have the ben efit of CT. In their place, there is not only CT but magnetic reso nance imaging (MRI). It is now commonplace to observe that MRI is unequalled as an imaging technique in neurology. Worldwide availability of MRI does, of course, differ widely, there being signif icant financial implications both in terms of capital outlay and run ning costs. So not every physician with a case of suspected multiple sclerosis (MS) will have direct or swift access to MRI. On the other hand, are there instances in which easy accessibility to MRI may 294 Chapter 32 Towards a balanced attitude not necessarily be to the patient's benefit? There probably are. Any neurologist will have under his care a patient with relatively benign MS who, e.g. may have only suffered scattered episodes of barely disabling sensory disturbance spread over 25 years. Is it really to the patient's benefit, in this situation, to be told of a firm diagnosis of MS at the very outset? There is a small group of patients who genuinely do not wish to know. Of course, the majority nowadays are well informed and questioning, such that a proper and open discussion about the possible causes of the neurological problem will inevitably lead to an equally proper investigation and diagno sis. New investigation techniques, then, can pose ethical problems not hitherto encountered. Further ethical issues are raised by the great advances in genetics. Some of the newer tests now available are mentioned in the sec tion on general medical investigations following directly. The latter, of course, are frequently required in the investigation of a neuro logical complaint, but the larger part will be devoted to the special investigations most commonly used in neurology. The usefulness of straightforward haematology and biochemistry in the neurol ogy clinic is not that great, and has probably been exaggerated hi the past. Only one out of many hundred patients with blackouts or headache will be discovered to have any relevant abnormality on 'routine' blood tests. There is an argument that automated and relatively very cheap investigations can, without harm or undue damage to the hospital budget, form part of the 'normal' investiga tion of every new patient. I do not believe this to be the correct view any longer, any more than the ready availability of CT scanning should encourage a request to be sent on every patient with 'ten sion' headache 'just in case'. One problem is that an increasing number of patients with a chronic symptom such as headache now expect and demand a CT scan. There is adequate evidence to show that investigating such patients without due clinical indication is almost always fruitless and certainly very expensive. This is not even to mention the exces sive exposure to radiation involved in such practice. But a specific anxiety about, e.g. an apparent family history of cerebral tumour, should be received with sympathy, and on occasion only a CT scan will provide reassurance adequate enough to avoid further consul tations. Some general practitioners, indeed, may be helped in the management of their patients more by direct access to a CT scan ning department than an inordinate delay whilst their patient waits to see the neurologist. Unfortunately, it will almost certainly be eco nomics rather than a planned clinical consensus that determines the outcome of such issues. Part 7 The investigation of neurological problems The overall investigation of neurological problems is becoming easier and safer, but as is already stated elsewhere, this is no excuse for unthinking overindulgence. With the correct application of clinical skills allied to the judicious use of the following described investigations, the hope is that the reader will usually get it about right. 296 33 General medical investigations The good neurologist will use laboratory investigations as part merely of the collection of evidence to differentiate between the 'shortlist' of possible conditions into which the history and exami nation has grouped the patient's condition. Certain of the more simple examinations may arguably be car ried out in most cases. These include a full blood count, erythrocyte sedimentation rate (ESR) or viscosity, liver function tests, urea and electrolytes. Commonly to these will be added blood glucose and calcium studies. In particular circumstances, it will be important to include syphilis serology and testing for human immunodefi ciency virus (HIV). Thyroid function tests, vitamin B]2 and folate levels are, of course, time-honoured investigations in neurology, and there is the need to screen for vasculitic disorders in an increas ing number of clinical situations. The investigations appropriate to coma have already been mentioned (p. 260). Dementia, peripheral neuropathy and vascular disease all require particular care with general medical investigation. Mention has already been made of the role of metabolic disorder in muscle disease as well as distur bances of consciousness. The place of tests for specific bacterial antigens in the diagnosis of meningitis will be described (p. 310). Auto-antibodies in association with paraneoplastic cerebellar syn dromes are described, and tests available. Without in any way being comprehensive, there follow a further few remarks concern ing common 'medical' investigations. It is probably in the diagnosis of peripheral neuropathy and vascu lar disease that the greatest number of general medical investigations is required from the outpatient clinic or ward. Twenty years ago, not all neurologists would have been aware of tire antiphospholipid syn drome, nor would patients with, e.g. venous thrombosis have been subjected to such a wide-ranging screening for thrombophilia as is currently available. By way of example, typical schemes of general investigation for patients with peripheral neuropathy, suspected vasculitic disease or other vascular disorder are outlined below. Suspected thrombophilia By this term, we mean a primary disorder of enhanced coagulation as the cause of either venous or arterial thrombosis. Appropriate 297 Part 7 The investigation of neurological problems investigation would include at the outset a routine haemostatic screen: 9 Full blood and platelet count 9 Prothrombin time a Activated partial thromboplastin time 9 Thrombin time o Reptilase time 9 Fibrinogen concentration Further detailed study might then include: 9 Tests for activated protein C resistance 9 Assay for protein C and protein S 9 Assay for antithrombin III ° Test for lupus anticoagulant • Test for anticardiolipin antibodies (IgG and IgM) The presence of either lupus anticoagulant or anticardiolipin antibody defines an antiphospholipid syndrome. Although first described in patients with systemic lupus erythematosus (SLE), the majority present with thrombotic disorder, arterial or venous. Suspected vascuflitis ° A screening programme for vasculitic disorder would often run alongside investigations for thrombophilia, as above, and would be initiated by full blood count and tests for ESR or vis cosity and C-reactive protein. This would then be followed by tests for antinuclear antibody, antinuclear cytoplasmic antibody auto-antibody screen and complement C3/C4. 0 In addition to stroke disorder, systemic vasculitis may present to the neurologist with encephalopathy, peripheral neuropathy cranial nerve lesions (isolated or partial ocular motor disorder, optic neuropathy or even deafness) and myositis. At least seen from a neurological perspective, not many patients seem to conform to the well-known and characteristic disorders such as SLE and polyarteritis nodosa. Peripheral! neuropathy Initial investigations would include a full blood count and ESR or viscosity, urea and electrolytes, blood glucose, serum proteins and plasma electrophoresis and liver function tests. To this would normally be added serum B12 and folate levels, and then, if indi cated, urinary porphyrins, thyroid function studies and a search for urinary Bence-Jones protein. Further investigation might then include a search for neoplasia, or ultimately biopsy as later 298 Chapter 33 General medical investigations discussed (p. 328). A vasculitis screen might also be required, in par ticular with the presentation of mononeuritis multiplex rather than diffuse symmetrical neuropathy, ha either case, where indicated, testing for HIV may be required. Multifocal or diffuse polyneu ropathy may also be associated with antiganglioside antibodies. Reduction of antibody concentration whether by plasma evchange or in response to immunosuppressive therapy may possibl be associated with clinical improvement. Oth mto-antibodies may accompany paraneoplastic polyneuropatl particularly accompa nying small cell carcmoma of the bronchus. Genetic tests V£ In recent years, there have been great advances i n TCrcWmymg the genetic abnormality in some important neurological disorders. This applies to Huntington's chorea, myotonic dystrophy and McArdle's syndrome. In Duchenne muscular dystrophy, the loca tion of the gene on the X chromosome is known. There are assays available to detect the various mitochondrial DNA mutations asso ciated with mitochondrial diseases, whether myopathy or more widespread. A DNA test can now identify cases of spinocerebellar ataxias. Similarly, there is now a gene marker available for type I ChaicoV-Mane-Toodv disease (heieddaiy motoi and sensory neu ropathy) and this may be incorporated in the above screening pro gramme for peripheral neuropathy, if it is of chronic demyelinating type. A quite different example is that of narcolepsy. There are only very few patients who do not have particular histocompatibility antigens, and in the sometimes difficult sphere of sleep disorder, this can provide extremely useful corroborative evidence. Liaison with the local genetics unit is increasingly frequent and essential. The above-mentioned examples of new genetic tests for many neu rological diseases are by no means comprehensive, as molecular genetics forges ahead. 299 34 The cerebrospinal fluid In bygone years, analysis of the cerebrospinal fluid (CSF) was the main ancillary investigation available to the neurologist. In modem practice, the value of information gained from this examination needs to be assessed critically, because the advances of neuroradiology and neurophysiology, together with the increasing relative ease and safety of surgical exploration, have combined to make lumbar puncture a far less important manoeuvre than it formerly was. It can be dangerous, indeed lethal, under certain circumstances. The information gained is often of dubious value - a protein level of 0.8 g/1 imports one piece of information only, namely that the patient has a raised protein in tine CSF; it tells nothing of the cause of the rise, the site, nature or extent of the lesion, if such exists. Negative findings exclude very few diseases, and casually carried out lumbar punctures sometimes make more important subsequent investigations very much more difficult. This chapter is an attempt to show when this investigation is of value, when it may be misleading and when it is positively contraindicated. Lumbar puncture Indications for lumbar puncture In brief, a lumbar puncture should be carried out if some specific piece of information is likely to come from the CSF examination that would substantially contribute towards diagnosis, treatment or assessment of the progress of the disease, ami where the clear-cut contraindications are not present. In each case, the question must be asked, 'What exactly is the information I hope or expect to obtain from this procedure'? If one has no clear idea of the answer, then there is no justification for lumbar puncture. Contraindications for lumbar puncture The contraindications to the procedure are quite clear are of the greatest importance and yet are ignored with frightening regularity. Absolute ° Local sepsis. Infected material from a lesion such as a bedsore near the lumbar puncture site can be transferred to the menin ges, with disastrous results. 300 Chapter 34 The cerebrospinal fluid 0 Symptoms and signs suggesting a tumour in the posterior fossa, causing incipient brainstem compression whether there is papilloedema or not. This would be judged by occipital head ache, drowsiness, vomiting, slowing of the pulse or episodes of faintness on change of posture in addition to cerebellar or other localizing signs. The danger of tonsillar herniation and severe brainstem compression is acute. 0 Patients already showing signs of tentorial or tonsillar herni ation. This should be suspected when a patient with symp toms suggestive of an intracranial space-occupying lesion suddenly becomes drowsy, develops neck stiffness and the pupils dilate. There is danger here also to the patient with the Arnold Chiari malformation where the tonsils may already be well below the foramen magnum. Computerized tomog raphy (CT) scanning should be a preliminary step. Persistent disregard of this rule sooner or later causes avoidable fatali ties from cerebellar tonsillar herniation through the foramen magnum or uncal herniation through the tentorial hiatus. A special neurological or neurosurgical hospital resident becomes distressingly familiar with urgent requests from other hospitals for the admission of a previously alert patient with papilloedema 'who has a CSF pressure of over 300' and has suddenly become very drowsy. Relative ® High intracranial pressure as shown by bilateral papilloedema, or suspected because of drowsiness, focal signs or brain swell ing shown already on CT or magnetic resonance imaging (MRI) scanning. Antibiotic treatment must be managed without CSF examination if there is raised intracranial pressure. In suspected subarachnoid haemorrhage, an urgent CT scan is generally pref erable to a lumbar puncture, but emphatically so in the presence of impaired consciousness or focal signs. In meningitis, CT scanning must exclude suspected abscess or subdural empyema before contemplating lumbar puncture. 0 Patients whose attitude towards investigative manoeuvres is such that the lumbar puncture is likely to be blamed for all future ills. This is not uncommon and unless truly vital informa tion is likely to be obtained, house physicians should remember that their contact with the patients is unlikely to last more than 12 months, while their chief may have to suffer for a lifetime. 301 Part 7 The investigation of neurological problems Technique of lumbar puncture For the normal diagnostic procedure, the patient should lie on his side, the back at the extreme edge o f a firm bed. One iliac crest must be maintained vertically above the other. The body is curled up to separate the spinous processes, and two lines are drawn, one join ing the tops of the iliac crests, and the other running down the spi nous processes. These intersect at the L3-L4 space, and the punc ture can be carried out at this space, or the next above or below. The higher spaces are usually easier to enter. Mark the space, cleanse the skin thoroughly (asepsis is as impor tant as in any surgical operation), towel off the selected space, scrubbing the hands thoroughly before and after the skin prepara tion, and drying the hands on a sterile towel. After warning the patient that he will feel a stinging sensation, with a fine needle raise a bleb of local anaesthetic in the skin. Wait a few moments, and then inject a little more under the skin, into the ligament between the spinous processes and a little to either side of it. Withdraw the needle, massage the area and then allow at least 2 minutes to elapse for the anaesthetic to have its effect. The majority7 of painful lumbar punctures are due to failure to follow this last rule. Now warn the patient that he will feel pressure, and may at one point have a sharp stab of pain - on piercing the dura. Insert the needle exactly in the midline midway between two spines, and pointing very slightly towards the patient's head, remembering that in this lateral position the midline is not necessarily halfway between the upper and lower sides of the body. Push the needle slowly but firmly forwards. A distinct jerk will be felt as it passes the interspinous ligament, and a smaller one as it enters the dura. After this second point, withdraw the stiletto and allow a few drops of fluid to escape. If none appears, push the needle a few millimetres further in and turn it slightly. When fluid is obtained, the manometer is quickly attached to the needle either directly or by using a three-way tap, and an assistant can hold the upper end. The commonest errors of technique include failure to locate accu rately the midline and not introducing the spinal needle at 90° to the surface of the patient's back. Interpretation of findings The CSF pressure Normally, the fluid rises in the manometer to about 110-115 mm, moves up and down on respiration, jerks upwards on coughing and 302 Chapter 34 The cerebrospinal fluid moves freely upwards on abdominal compression. A pressure of up to 230 or even 250 nun may be found in 'normal' obese patients. A very low pressure is most commonly due to faulty placing of the needle, in which case abdominal compression will cause no rise. A slight alteration of its position by advancing, withdraw ing or rotating the shaft will be necessary. The pressure is also low below a complete spinal block and when there is a block at the fora men magnum. Lumbar puncture, even as part of myelography, is highly dangerous in the presence of a foramen magnum lesion. If suspected, MRI is the investigation of choice. A high pressure is most commonly due to tension, or to abdomi nal compression in a plump patient. Straighten the back and legs slightly and assure the patient that all discomfort is now over. If the reading still remains high, this may be due to a genu inely high intracranial pressure. (Pressure cannot be assessed by observing the speed with which the CSF drips out of the needle. A manometer must always be used.) CaU.ecU.aa o f tb.e fluid Disconnect the manometer and allow the fluid to drip slowly into a series of sterile containers, collecting up to 10 ml. If the glucose con tent is likely to be important and there is a delay before the analy sis is carried out, the fluid should be collected in a fluoride-coated tube. Generally, the specimens should be dispatched immediately to the (correct) laboratory. If the pressure should be unexpectedly high (over 200), then only the fluid in the manometer should be removed. If the pressure is extremely low, enough fluid to examine the cells and protein only should be removed. One cannot too strongly deplore the practice of withdrawing fluid by means of a syringe in such cases. This can make a previously incomplete spinal compres sion complete, or can convert incipient cerebellar tonsillar hernia tion into an irremediable impacted pressure cone. T h e ©bese pat'cerat In a fat patient, it may be impossible to identify the landmarks. It is then justifiable to carry out the puncture with the patient seated astride a chair, with pillows in front to cause the back to curve back wards. The exact midline can be better established and it is very rare for the spinous processes to be impalpable. Pressure readings are, however, of little value and the danger of a pressure cone forming in high-pressure states is even greater. Alternatively, a cooperative 303 Part 7 The investigation of neurological problems radiologist may help with a lateral cervical or lumbar approach under direct X-ray control. Cisternal puncture is thus unnecessary. Examination of the CSF Hold the container first up to the light and then against a white surface. A cloudy fluid • Indicates an increase in the cell count above 400 cells/m m 3. • If also smoky in appearance, this may be due to the presence of large numbers of red cells. A blood-stained fluid The first few drops must be watched most carefully. A blood-stained CSF is due most commonly to trauma to local vessels during the puncture. If the first few drops are clear and the fluid then becomes blood-stained, this is due to damage to the vertebral venous plexus. If the first few drops are blood-stained and the fluid then runs clear, it is due to damage to the superficial vessels. If the fluid remains evenly blood-stained, it may: 0 Still be due to a traumatic tap ° Represent genuine spontaneous subarachnoid haemorrhage. In order to distinguish between the two, the fluid must be cen trifuged immediately. If the supernatant fluid is colourless, this is unlikely to be anything but traumatic. If it is xanthochromic, blood is likely to have been in the fluid already. This is a good working general rule, but not invariably correct. If a perfectly conscious and alert patient has no signs of meningeal irritation, be very wary of attributing a very heavily blood-stained fluid to subarachnoid haemorrhage - it is nearly always traumatic. A xanthochromic fluid This is due to the entry of blood pigments into the CSF. It may occur in several ways: 0 Following haemorrhage into the CSF. It usually does not develop for 6 hours, but may persist up to 20 days. 0 ha a highly proteinous fluid, e.g. below a complete spinal block; in the vicinity of a neurofibroma and in some cases of the Guillain-Barre syndrome. 0 In the presence of jaundice. 304 Chapter 34 The cerebrospinal fluid A fluid that clots • The blood from a traumatic puncture usually clots. • A highly proteinous fluid may clot if it becomes contaminated by a drop of blood. • A cobweb clot may form in tuberculous meningitis. This is a rich source of tubercle bacilli, but not commonly seen. The cells If there are more than 5 cells/m m 3, or if any of them are polymor phonuclear, the fluid is abnormal. A wholly or predominantly polymorphonuclear pleocytosis indi cates acute meningeal irritation, but gives no indication as to the cause. A wholly lymphocytic pleocytosis occurs most commonly in subacute irritative or degenerative processes affecting the parenchyma of the nervous system, with or without meningeal involvement. Polymorphonuclear pleocytosis 0 Very high counts (many thousands) can be found in most pyo genic bacterial meningitides, and these are associated with a low glucose content. • High counts (thousands), in addition to the above, occur in leptospiral meningitis, in allergic meningeal reactions to parasitic infestation, and occasionally when cerebral haemorrhages or softenings have occurred in close relationship to the ventricles or surface of the brain, and in infarction of a pituitary tumour. In these cases, the glucose content is normal, but in other respects they may resemble a bacterial meningitis. • Moderately high counts (hundreds) are seen in the earliest stages of several conditions which are usually characterized by a lymphocytic pleocytosis, such as acute lymphocytic chorio meningitis, or tuberculous meningitis, and also in some very acute disseminated demyelinating lesions. • Raised polymorphonuclear counts (below 100) usually form part of a rise in both types of cells, and this can be found in a wide variety of subacute infective lesions (e.g. cerebral abscess or tuberculous meningitis), granulomatous lesions (e.g. syphi lis or sarcoid), metastatic neoplastic lesions and following the introduction of irritant substances such as blood into the sub arachnoid space. 305 Part 7 The investigation of neurological problems Needless to say, these divisions and those that follow are artifi cial. They are intended merely to indicate the train of thought that may be started when figures of a particular order are obtained. Lymphocytic pleocytosis Lymphocyte counts in the thousands are very rarely seen. 1 Hundreds of lymphocytes per cubic millimetre are found in viral meningitis and encephalitis, in tuberculous meningitis, in some very acute demyelinating conditions, especially in young children, in parasitic infestation and in the presence of a cere bral abscess. A rise in the polymorphonuclear cells frequently accompanies the infective lesions in the earliest stages. 2 A lymphocyte count of less than 100/m m 3 is very common in a variety of subacute degenerative and granulomatous lesions, the commonest of which are multiple sclerosis, neurosyphilis and in metastatic cerebral or spinal disease. More than 50 lympho cytes/m m 3 casts doubt on a diagnosis of multiple sclerosis; 99% of cases will have less than 20 cells, even when in acute relapse. 3 High cell counts may occur if a necrotic tumour lies in close contact with the ventricular system. A lvmphocytic pleocytosis with a glucose content below 0.4 g/1 (2 mmol/1) occurs, to all practical purposes, only in tubercu lous meningitis, carcinomatosis of the meninges and multiple spinal seedlings from a medulloblastoma. In early stages of tubercular meningitis, there may be polymor phonuclear leucocytosis and similarly some cases of pyogenic men ingitis may have lymphocytic pleocytosis. However, in untreated pyogenic meningitis, the cell number is usually in thousands and the interval between onset and altered consciousness is few days (usually less than 5), whereas in tubercular meningitis, the cell number is usually not above 500 and the interval between onset and altered consciousness is usually more than 10 days. The cell number along with history is more important than the cell type to distinguish between pyogenic and tubercular meningitis. 306 Chapter 34 The cerebrospinal fluid A raised lymphocyte count may be seen if another lumbar punc ture has been performed recently (as may be the case prior to transfer from a general hospital to a special unit). In these circumstances, it is most important to find out if the original CSF was normal. Distinguishing ceil types The routine laboratory examination will differentiate between polymorphonuclear and lymphocytic cells, but in carcinomatous or other malignant meningitides, special histochemical and immu nological staining techniques can be applied to the centrifuged deposit from a fresh CSF sample. Carcinoma cells from primary tumours of the bronchus or breast may be identified, along with evidence for melanoma, myeloma, lymphoma or leukaemia. The protein content Normally, this lies between 0.15 and 0.4 g/1. It is lower in the cisterna magna and lower still in the ventricular fluid. If, as is not infrequent, figures of 0.1 g/1 are consistently recorded in the lum bar fluid, there is something wrong in the laboratory and the meth ods and standards need reconsideration. This applies equally if figures are persistently above 0.6 g/1. A low protein In practice, this has only negative value. A raised protein This occurs in so many diseases of the central nervous system (CNS), the meninges or the blood vessels, that there is no purpose to be gained simply by listing them. Figures over 1.0 g/1 are usu ally only seen below a complete spinal block, in post-inflammatory polyneuropathy (the Guillain-Barre syndrome), carcinomatosis of the meninges and infective meningitis, particularly tuberculous or fungal. A rise in protein merely indicates abnormality and cannot be considered diagnostic of any one particular disease process. If the CSF is blood-stained, an allowance of 0.01 g/1 of protein should be made for each 750 red cells/m m 3. Electrophoresis of the CSF This allows quantitative measurements of the protein fractions after concentration of the CSF to be carried out, especially of the Part 7 The investigation of neurological problems globulins, and this has become diagnostically helpful in certain situations. If a patient is clinically suspected of having multiple sclerosis, a gammaglobulin of over 12% of the total protein is considerably in favour of this diagnosis, and of over 25% is very strongly in favour if there is no reason to suspect neurosyphilis or collagenosis. The importance is that it may be shown when the dis ease is clinically quiescent, when the total protein is normal, and a Lange curve, if carried out, normal. The gammaglobulin may contain two characteristic bands. Further analysis by immunoelectrophoresis will show it is the IgG fraction that is raised. These IgG bands are called oligoclonal, because when antibody is synthesized within the CNS, the response is limited to a small number of B-cell clones. Oligoclonal bands are present to some degree in all CNS infections and are an especially prominent feature iii chronic infection such as subacute sclerosing panencephalitis. (In this case, nearly all the IgG is measles specific.) There is bound to be elevated IgG in the presence of many CSF lymphocytes, hi multiple sclerosis, it is the finding of IgG bands in the appropriate clinical setting that is important, together with a lack of any abnormal IgG in the serum, hi established cases, up to 95% will show this response, and even after just one suspected episode, the figure is 50-60%. Electrophoresis is not the only means to demonstrate IgG bands, and hi recent years, more experience of isoelectric focusing has accu-mulated. This test is particularly sensitive for the detection of oligo clonal IgG; commercial 'kits' are available but the technique is more expensive than electrophoresis. Th e glucose content Normally, this lies between 0.5 and 0.75 g/1 or more than 50% of a simultaneous blood glucose estimation. The estimation must be carried out quickly, especially if there is a pleocytosis. If a highly cellular fluid stands overnight, the sugar content may be greatly reduced, unless a fluoride tube is used. A high glucose Usually, such a reading merely reflects a high blood glucose, e.g. in diabetes, and this diagnosis has been made hi this way on more than one occasion! A low glucose The glucose is lower in some, but not all, cases of meningeal infec tion. hi active pyogenic meningitis, the level is usually below 308 Chapter 34 The cerebrospinal fluid 0.4 g/1. In virus meningitis, it does not fall. A figure less than 50% of the blood glucose indicates pyogenic, fungal, tuberculous or malig nant meningitis. Subarachnoid haemorrhage is an alternative cause. Unless one of the other conditions mentioned in is obvious, a lymphocytic meningitis with a CSF glucose below 0.4 g/1 should be treated as tuberculous meningitis without waiting for confirma tory investigations, because these cause delay and delay may spell disaster. Bacteriology and virology If an infective condition is suspected, advice should be sought from the laboratory before the puncture is carried out, because immedi ate inoculation of culture media with the fluid often gives a better chance of identifying an organism. All cases of pyogenic meningitis must have the organism identified by every possible means, and its sensitivity to various antibiotics determined. Specific bacterial anti gens may be detected by countercurrent immunoelectrophoresis or agglutination techniques, and their presence may be diagnostic in cases where antibiotic treatment interferes with organism culture. Special culture media are required for certain organisms, e.g. myco plasma and listeria, and so again, full cooperation with the microbi ologist is essential. Repeated examination of serial CSF samples is often required to detect acid-fast bacilli. India ink stains and similar persistence is necessary if fungal meningitis is suspected. In viral disease, it is disappointingly rare to recover the virus from the CSF, but specific antibodies may be raised (e.g. herpes simplex type I). More value comes from isolation of virus from a variety of swabs or serological screening. Serology In patients suspected of neurosyphilis, the CSF sample should be evaluated by Venereal Diseases Research Laboratory (VDRL) test. It should be complemented by serum VDRL and fluorescent trepo nemal antibody-absorption (FTA-ABS) test. FTA-ABS is more spe cific, sensitive and reliable, although false-positive results do occur occasionally. In patients with encephalitis, paired sera should be sent for serol ogy of Japanese encephalitis, dengue and other arbo- and nonarbo viruses. The rising titre of antibodies is detected in positive cases. India ink staining of CSF is warranted in all immunocompro mised and also immunocompetent patients with chronic meningitis to detect cryptococcal fungal meningitis. 309 Part 7 The investigation of neurological problems For detecting other fungal meningoencephalitis, the CSF should be examined by KOH staining (Aspergillus and Mucormycosis) and fungal culture. Direct CSF wet mount m icroscopy Patients who develop acute meningitis with a recent history of swimming in fresh water should undergo CSF examination with direct wet mount microscopy to detect Acanthamoeba-related meningoencephalitis. To differentiate Naegleria fowleri infection and Acanthamoeba spp. infections, wet mount examination and culture of a large volume of CSF is required. Iatrogenic changes Blood introduced into the CSF by a traumatic lumbar puncture causes a white cell pleocytosis within 12 hours which may persist in decreasing degree up to 20 days, and long after the blood itself has disappeared. Xanthochromia appears here as well. 310 35 The clinical value of electroencephalography Although in the past electroencephalography (EEG) was used as a screening test for structural brain disease, it is first and foremost an investigation of cerebral function, most useful in the investigation and management of patients with epilepsy. Focal abnormalities on an EEG may suggest an underlying structural abnormality, but the diagnosis will require scanning with either computerized tomogra phy (CT) or magnetic resonance imaging (MRI). Tire fact that a right frontal lobe tumour causes slow waves to arise from the right frontal lobe does not mean that slow waves arising from the right frontal lobe necessarily means the patient has a right frontal lobe tumour, or even is most likely to have one. Although invaluable in the diag nosis of epilepsy, as many as 50% of patients who undoubtedly have suffered a tonic-clonic seizure will still have a normal EEG. In this case, and indeed under any circumstance, the EEG is not a shortcut around, or a substitute for, a proper and complete history from both the patient and a relation or any other witness to the alleged 'sei zure'. In patients who have an established seizure disorder, the EEG will help to classify the type of epilepsy, indicate the most appropri ate antiepileptic drug available and, together with special techniques discussed below, possibly indicate a focal source for the seizure dis order leading ultimately to the possibility of surgical treatment. Although encephalitis and encephalopathy generally may be rare, an EEG is crucial to accurate diagnosis and assessment, and in certain instances specific disorders may be recognized by their character istic EEG abnormality. These include herpes simplex encephalitis, Creutzfeldt-Jakob disease, subacute sclerosing panencephalitis (SSPE) and metabolic encephalopathy. Importantly, the EEG may also reveal non-convulsive epileptic activity underlying an enceph alitis or encephalopathy presenting as coma. Again, specific abnormalities may be found in certain metabolic encephalopathies, particularly hepatic failure. An entirely normal EEG in the presence of apparent unconsciousness will indicate hysterical pseudocoma, and in patients with behavioural disturbance or confusional state, defining any abnormality suggestive of metabolic encephalopathywill be extremely helpful, and occasionally epileptic discharges will be found. Tire EEG may further be useful in the diagnosis and management of sleep disorders, but an EEG, at least in the UK, is not included in the guidelines for the diagnosis of brainstem death. 311 Part 7 The investigation of neurological problems This is because an isoelectric (flat) EEG does not necessarily indi cate brain death, occurring in reversible drug-induced coma. Long-term monitoring with ambulatory recordings is being increas ingly used both in the assessment of frequency of epileptic activity and in tire often difficult differentiation between attacks of true petit mal, complex partial seizures of temporal lobe origin or attacks of no organic origin at all. hr this latter category - non-epileptic or pseudoseizures - com bined EEG and video recording of tire patient can be particularly help ful, and both tire physical and EEG events occurring during transient seizures or other attacks can be simultaneously studied. Before moving on to an outline of the technique of the EEG exami nation itself, a few further points require emphasis. In the investiga tion of a patient complaining of attacks of disturbed consciousness, the main purpose of the request should not be a vague hope that the EEG will decide whether the patient has epilepsy. This must conre from a detailed clinical history, correlated later, perhaps, with any abnormal EEG finding. It is an abuse of the investigation if the phy sician requesting the EEG is likely to be more influenced by a nor mal report than by his own assessment of the problem. Conversely, as it will be seen later, both sharp waves and spikes can occur in the absence of clinical epilepsy. With these caveats, the aim should be to learn if there is an abnormality suggestive of epilepsy, whether it is mild or severe, spasmodic or continuous, localized or diffuse. In an epileptic patient already under treatment, follow-up assessments can be made; on the whole, unchanged and continuing EEG abnor malities, despite appropriate medication, do not indicate a favour able prognosis. Any drug treatment, particularly benzodiazepines, must be indicated on the request form (or withdrawn) because certain drugs can fundamentally alter the EEG. Overall, with the exception of pseudocoma, the EEG will not exclude organic disease, epilepsy or tumour, and it will not, by itself, diagnose any disease condition, although it may suggest very strongly those particular conditions already mentioned. Waiting for the EEG report should not delay the initial treatment for definite tonic-clonic seizures, nor delay further investigation for structural disease. Alternatively/ EEG is equally or more important than CT scan in the investiga tion of encephalitis or encephalopathy, but is frequently omitted. A surprising number of younger doctors exclaim puzzlement at the notion of an unconscious patient with a normal CT scan. Technique of examination Only a bare outline of a highly technical procedure will be given here, in an attempt to make the reported findings more comprehensible. 312 Chapter 35 The clinical value of electroencephalography About 21 electrodes are held in place on the scalp, either by a cap of straps or by 'sticking on' with paste or gel according to the International 10-20 system. They are usually placed in standard positions, covering as much of the underlying brain as possible. A lend runs from each electrode to a headpiece and on to the ampli fiers. Each recording pen is connected to two electrodes leading to one channel so that the two channels record the activity from three adjacent electrodes, one electrode being common to each. Standard machines have 16 channels. Preset switching enables a large variety of recording patterns to be used, although in practice a relatively small number of standard patterns are routinely used. Physiological rhythms (Fig. 35.1) Channels recording from electrodes over the posterior part of the brain usually show an alpha rhythm when the eyes are closed. This is the original activity recorded by Hans Berger using a pair of elec trodes, one frontal and one occipital. The waves occur in runs of varying length, now defined as between 8 and 13 Hz, and these tend to disappear when the eyes are opened or during some form of thought process, such as mental arithmetic. Theta activity runs at 4-7 Hz, is seen in small amounts anteriorly, particularly in transverse recording patterns, and in children, it may be the dominant rhythm. Beta activity is above 13 Hz, present at low voltage at random in most records, common in tense apprehensive subjects and promi nent in patients on barbiturates. Lambda waves are seen posteriorly, conventionally recorded as an upward deflection and present in many subjects when looking at a patterned field. Mu activity is complex, being a basic rhythm of about 10-12 Hz with a harmonic component superimposed. It may represent the resting activity of the motor and sensory areas, and can be blocked by movements or stimuli applied to the opposite half of the body. K complexes, seen most commonly anteriorly, consist of one or more high-voltage slow or sharp waves followed by a burst of rhythmic activity (sigma activity), and are related to arousal stimuli during sleep, though not necessarily followed by arousal. Abnormal rhythms Excessive theta activity in adults can be abnormal, though it may be due to nothing more than drowsiness. A degree of theta activity is often found in young adults, but if not explicable by drowsiness, 313 Part 7 The investigation of neurological problems 1 second v/w V \ a /x/^/V V W ^ W /v^ aA / (b) F i g . 3 5 .1 N o rm al and a b n o r m a l r h y th m s . vwv^/\AM/VyvvM/" (c) in older patients, it may indicate either a focal or lateralized pathol ogy. Its evaluation is difficult, and requires particularly careful cor relation with the clinical situation. Delta activity consists of very7slow waves, less than 4 Hz, and outside sleep, its presence in adults is always abnormal. It can be generalized in many pathological processes and in long-standing epilepsy, but when localized, it is indicative of a local structural lesion. Focal discharges (Fig. 35.2). If the deflections of an abnormal discharge are in opposite directions at tine same moment in two channels having a common electrode, this is termed as phase reversal and indicates that tine origin of the discharge is near the common electrode (Fig. 35.2(a)). Spikes (Fig. 35.2(b)). These are very sharp waves, usually of high voltage, looking something like the QRS complex of an electrocar diogram. A spike is defined as having a duration o f 70 ms or less, whereas a sharp wave has a duration o f between 70 and 200 ms. While the pres ence of an interictal spike discharge is often suggestive of an epilep tic disturbance, such an activity7may occur in non-epileptic people. Caution is required in interpretation. If persistent at any one site, a spike discharge may indicate a focal cortical lesion. 314 (a) Alpha rhythm, note the suppression on eye opening at arrow; (b) theta rhythm and (c) delta rhythm (paper speed 3 cm /s). Chapter 35 The clinical value of electroencephalography Fig. 35.2 Focal discharges, (a) Focal delta activity in a case of cerebral abscess. (b) F o c a l s p i k e s in a n o ld c o r t ic a l in ju r y (p a p e r sp e e d 3 c m /s). Spike and wave discharges. These consist of a spike followed by a large delta wave, or vice versa. They may occur in isolation, in runs of irregular size and speed or in completely regular runs of abrupt onset and cessation. Generalized, bilaterally symmetrical and bisynchronous 2.5-3.0-Hz spike-wave activity is the charac teristic finding in patients with primary or idiopathic generalized epilepsy (Fig. 35.3(a)). It may also occur in patients with general ized seizures secondary to focal pathology, the so-called secondary generalized epilepsy. Atypical spike and wave runs of irregular fre quency are common in other forms of epilepsy, and may consist of one or more spikes, associated with irregular slow waves and biphasic waves, and usually occurring in episodic bursts. Myoclonus. Here we are referring to primary generalized epileptic myoclonus as a fragment of primary generalized epilepsy (above). Again, there are bilaterally synchronous and symmetrical bursts of spike and wave activity, but the spikes may be multiple, perhaps 4-6 together, and the discharges are usually relatively brief. These complexes may be referred to as 'polyspikes and waves'. Hypsarrhythmia is the term given to the EEG abnormality accom-panying encephalopathy in infants. Clinically, there are infantile spasms together with developmental delay, the causes of which include viral encephalitis or neonatal hypoxic damage. The EEG shows a diffusely abnormal background activity with superim posed high-voltage and multifocal spikes and slow waves. Periodicity. It has already been mentioned that in certain condi tions such as herpes encephalitis and SSPE, the EEG may show a characteristic periodic pattern (Fig. 35.3(b)). In herpes encephalitis, the periodic activity consists of high-amplitude, sharp-slow wave 315 Part 7 The investigation of neurological problems Fig. 35.3 Characteristic BEGs. (a) Petitmal attack- regular 3-Hz spikes and w a v es (paper speed 3 cm /s). (b) SSPE; regular periodic high-voltage complexes throughout record (paper speed 1.5 cm /s). complexes recurring every 2-4 seconds. In SSPE, there are repeated high-amplitude bilateral bursts of slow activity and sharp waves at 3-10-second intervals. Patients with hepatic encephalopathy or Creutzfeldt-Jakob disease may also show periodic complexes. M ethods of stimulation One of the problems of EEG is that abnormalities may not be pres ent at the particular time that the tracing is being taken. Various ways have been evok ed by which abnormalities may be evoked or slight abnormalities made more obvious and more clear-cut. Hyperventilation The patient breathes deeply and vigorously for about 3 minutes, or less if he cannot tolerate it. This may convert a dubious tracing into one that is definitely abnormal (Fig. 35.4). It may, however, produce a deviation from the normal in apparently normal subjects, but this is usually transient. It is particularly significant if the abnormal ity persists for over a minute after stopping the hyperventilation, and a very exaggerated response may be seen in hypoglycaemia, even in the absence of a low blood glucose level. This can be abol ished by oral or intravenous glucose in such patients. In epileptics, it may change a relatively normal record into one which is very unstable and abnormal. In the majority of patients suffering from untreated primary generalized epilepsy, runs of spike and wave activity occur during hyperventilation. There is also always the 316 Chapter 35 The clinical value of electroencephalography '■''VW

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