John Patten

Neurological Differential Diagnosis

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an illustrated approach
with 288 figures

JOHN PATTEN

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PREFACE

The majority of doctors are ill at ease when confronted by a patient with a neurological problem. Candidates for qualifying examinations and higher diplomas dread that they will be allocated a neurological "long case".

This is a serious reflection on the adequacy of training in neurology. It is still possible in some medical schools for a student to go through his entire clinical course without an attachment to the neurological unit. Increasing competition for teaching time has led to the situation where in most U.S. medical schools and at least one new medical school in the U.K., a two-week clinical attachment to the neurology service is considered adequate. Those fortunate enough to attend a post-graduate course find a minimum of three months' intensive training is necessary before any confidence in tackling a neurological problem is achieved.

Unfortunately, neurological textbooks seldom seem to recognise the intensely practical nature of the subject. There are many short texts that achieve brevity by the exclusion of explanatory material; these are difficult to read and digest. At the opposite extreme are the neurological compendia, often unbalanced by excessive coverage of rare diseases and all based on the assumption that patients announce on arrival that they have a demyelinating, heredofamilial, neoplastic, etc., etc., disorder. These texts are useful only to those who already have a good working knowledge of neurological diseases.

Patients present with symptoms that need careful evaluation before physical examination which in turn should be firmly based on the diagnostic possibilities suggested by the history. Understanding neurological symptoms and signs requires a good knowledge of the gross anatomy of the nervous system, its blood supply and supporting tissues; and yet an almost universal feature of neurological texts is the paucity of instructive diagrams. Most students readily admit that this is an insoluble problem as their poorly remembered neurological anatomy is often both inadequate and inappropriate to the clinical situation.

The present text is a personal approach to this complex and fascinating subject and basically reflects the way the author coped with this difficulty. The subject matter is dealt with in two ways:

(1) A symptomatic, regional anatomical basis for those areas where local anatomy determines symptoms and signs, allowing what might otherwise be regarded as "small print" anatomy to be understood and appreciated.

(2) A full discussion of the historical diagnostic clues in those conditions that are diagnosed almost exclusively on symptoms such as headache, face pain and loss of consciousness.

Throughout, an attempt has been made to preserve the "common things are common" approach which is widely rumoured to be the secret of passing examinations, and is also the basis of good clinical practice. Rare disorders are discussed briefly, but it is important that beginners realise that rare diseases are usually diagnosed when it becomes apparent that the disorder does *not* fit into any of the common clinical pictures. The first aim therefore should be to gain a very good grasp of the common disorders rather than attempting to learn long lists of very rare diseases.

The text is profusely illustrated by the author and although anatomical accuracy has been preserved, artistic licence has been taken whenever necessary to illustrate an important point. Each diagram is drawn from a special angle of view that enables the reader to visualise the area under discussion *in situ* in the patient. It is easy to construct a "diagram" so remote from actual anatomy that it becomes incomprehensible. It is hoped that this problem has been avoided.

Neurological terms are defined whenever they occur and neurological "jargon" is explained, although the beginner is well advised to keep to factual statements until he is very sure of himself. As an example of the incorrect use of jargon the following is abstracted from a report by a physician. The patient actually had a left sixth nerve palsy and a mild hemiparesis due to cerebal metastases.

> *He has the spastic dystonic gait of the multiple sclerotic. Also he has a third nerve paraplegia with ocular movements to the left and with nystagmus. He has cogwheel rigidity of the upper extremities and an absence of patellar reflexes. His speech is beginning to slur and there is the mask facies of the multiple sclerotic . . . I do not believe . . . further studies to be necessary as the clinical signs are* all too obvious*!*

Specific references are not given; this book is intended for the novice who wishes to develop a "feel" for the subject and in the author's opinion references are unnecessary. This is not intended to indicate that the writer claims originality for all the information provided. The knowledge contained in this text is a distillate of wisdom gained from many teachers and, indeed, generations of teachers who have passed on their own observations. The personal part of this text is the attempt to organise this information around the anatomy of

the nervous system to reinforce this knowledge and make the subject less intimidating to the beginner. In particular I would like to acknowledge my debt to my first teacher, Dr. Swithin Meadows, who aroused my enthusiasm for the subject and then inspired me to attempt to emulate his skill as a clinical neurologist.

I am also indebted to many undergraduate students at Westminster Medical School, University College Hospital and the University of Texas Medical Branch, and post-graduates at the National Hospital for Nervous Diseases whose questions, suggestions and enthusiasm originally encouraged me to bring this approach to a wider audience.

Dr. John R. Calverley, M.D., Associate Professor of Neurology in the University of Texas Medical Branch at Galveston, provided enormous encouragement in the early development of this project and allowed me to try the format on several generations of post-graduate students in his Department.

I would like to express my thanks to him and his colleagues for making my stay in his Department so instructive and enjoyable. Dr. M. J. Harrison read the original manuscript and made many useful suggestions which have been incorporated into the text. I am very grateful for his help and encouragement.

I would like to thank my wife and family for their continued support during the years this text has been in preparation and Miss Gillian Taylor for typing the often illegible manuscript.

John Patten

Guildford, 1977

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CONTENTS

Chapter 1

HISTORY-TAKING AND PHYSICAL EXAMINATION

By tradition the first chapter in any textbook of C.N.S. diseases deals with history-taking, and this necessity appears inescapable. Yet in neurology the history is so critical, in indicating both the site and the nature of the lesion, that adequate coverage is impossible in a single chapter. The features of the clinical history appropriate to each region will form a major part of each of the subsequent chapters. At this stage discussion will be confined to some broad generalisations.

The secret of good history-taking is to be a good listener. However rushed the doctor may be, it is vital that the patient feels that he has the whole of the doctor's time and attention during the interview. The doctor must also be constantly aware that the majority of patients are extremely frightened, even though their actual behaviour may range from tongue-tied, tremulous anxiety to the blustering type of patient who insists that he came only because his wife was worried about him! The patient requiring most care, attention and caution is the one who is self-effacing and apologetic for wasting your time; with remarkable regularity these are the patients who have a serious disease.

Every effort should be made to put the patient at ease. Several minutes spent discussing inconsequential subjects such as the weather may seem to be time-wasting to the uninitiated but the relaxation and the insight gained into the patient's mood, his reaction to his disease and his intellectual ability is often greater than one can obtain with several minutes of formal testing, or more direct questioning.

As soon as the patient is talking freely the subject of his symptoms should be introduced. Although a referring letter may be of help in guiding the questioning, it is always important to ask the patient to relate the *entire* history again. Otherwise, half-digested views expressed by other doctors, relatives or workmates may intrude into the history as facts rather than supposition. The importance of cross-checking the history with a relative or friend is vital. In diseases affecting the patient's consciousness or intellect the history may be confused or impossible to obtain without the help of a third party. In other conditions patients frequently understate the duration and severity of their disability and again a completely different story may emerge from enquiries made of a relative. In general these supplementary interviews are best conducted in another room while the patient is undressing, particularly in the case of epileptic attacks in children, where the parents are often loath to reveal any of the details of ictal behaviour in front of the child. Peculiar behaviour traits at any age clearly cannot be discussed easily in front of the patient and a private interview with relatives, fellow-employees and others in contact with the patient is vital.

In all cases an accurate history of the entire course of the illness is essential. The diagnostic differences between weakness of an arm coming on overnight, over a week or over several months are so important that vague statements such as "gradually" should not be accepted at face value. A clerk with a radial nerve palsy present on waking one Sunday morning was cautiously asked if he had been drinking. With considerable reluctance, anxiety and embarrassment he admitted that he had become drunk for the first time in his life the night before. Yet with a single question one could exclude a stroke (the diagnosis made elsewhere) and explain both the aetiology and excellent prognosis of a "Saturday night radial nerve palsy" to the relieved patient.

In many patients bizarre historical events may be deliberately concealed for fear of embarrassment or of moral judgements made by the doctor. The onset of symptoms during sexual activity or alcohol indulgence are frequently suppressed for this reason. The family of an elderly patient with periodic confusion and amnesia (who had been extensively investigated elsewhere) were asked about her drinking habits. An initial angry denial was later retracted following a family conference, with the admission that her wardrobe was full of empty sherry bottles! Considerable tact is called for in these instances and yet the matter must be pursued if unnecessary investigations and incorrect diagnoses are to be avoided.

In neurological medicine, in addition to the importance of making the diagnosis, in many instances the doctor's subsequent role will be to help the patient accept and cope with what may prove to be a lifelong disability. The establishment of an easy confident relationship at the first interview will help ensure a useful supportive role in the future.

When one sees the comment "hopeless historian" on the notes it is really as much a comment on the doctor taking the history! The doctor must tailor the interview to suit the patient and the skill to do this can only be acquired; it cannot be taught. The ability to sit back and listen does not come

easily to many, but it is only the eventual realisation that one cannot force the pace of an interview without losing much of value that leads to the ability to take a good history.

The circumlocutory historian and the patient who insists on including irrelevant detail can be the most trying of all. Any attempt to alter the line of questioning will start another sequence of irrelevances. It is best to sit and listen and wait for the useful pieces of information to emerge. These are often the facts regarded as the least important by the patient! Considering how infinitely variable symptoms could be, it is amazing how often patients eventually describe their feelings and symptoms in almost identical phrases. The diagnosis of several classical syndromes depends on typical symptom descriptions. For example, the "red hot needle" jabs of pain in tic douloureux; the feeling of "*déjà vu*" often associated with temporal lobe attacks, or the "icy cold bandage around the waist or legs" described by patients with spinal cord lesions. Some of the feelings are so bizarre that the patient hesitates to describe them for fear of being accused of imagining things. A useful rule in neurology is that the more bizarre and unusual a symptom, the more likely it is to be organic. An arrogant approach—assuming that a symptom which is not known to the examiner must be functional or imaginary—is very likely to lead to misdiagnosis. It is dangerous to presume universal knowledge.

The above suggestions may not appeal to those who think that a doctor ought not to adopt a passive role and who see themselves as too busy and too important to be slowed down by vague patients. Such a person would be ill advised to embark on a career in neurology.

The usual sequence of a medical consultation is that the history leads on to the physical examination. It is only at this stage that the full value of the history becomes apparent. If a *full* neurological examination were performed on each patient it would take up to an hour for its completion and would prove extremely boring. The history serves to indicate those parts of the examination that should be performed with special care, skill and finesse. Few would perform exactly the same physical examination on a patient with headache as they would on a patient with pain in the leg, and yet the ability to tailor the examination to the situation is based entirely on the history. If a confident diagnosis of the site and probable nature of the lesion has not been made by the completion of the history it is very unlikely that the diagnosis will suddenly become apparent during the physical examination.

Having made a provisional diagnosis based on the history, the physical examination should be performed in the same relaxed way. A warm room with reasonable privacy is essential for a neurological examination. Although the doctor is accustomed to the sight of a tendon hammer and tuning fork, he must remember that the patient may not have seen either previously. He should always explain what he is about to do *before* doing it. Suddenly flashing a light in the patient's eyes or sticking a pin into a limb without warning hardly inspires patient confidence or patient co-operation. When attempting to elicit the plantar responses the examiner should warn the patient that it will hurt. Patients will tolerate considerable discomfort if invited to do so, but do not take kindly to unannounced aggression! Elicitation of both the supinator and ankle jerks is quite painful for the patient and yet one sees doctors repeatedly striking the tendon and wondering why the patient will not relax. Notes such as "patient will not relax" or "plantars impossible" can usually be regarded as due to poor examination technique rather than an adverse comment on the patient's co-operation.

The correct method of performing the various parts of the neurological examination will be detailed in the following chapters. This will always be in the setting of diseases that actually cause abnormalities.

In the author's view the only way to understand the mechanism and correct elicitation of physical signs is in relation to the situation where correct technique actually matters. For example, in a patient with backache, correct testing of the corneal response may not be critical, but in a patient with a giddy attack, facial weakness or face pain, the sign must be understood and elicited correctly, as the depression or absence of a corneal response may be the only physical sign of an underlying lesion in the cerebello-pontine angle.

Throughout this book the emphasis is on the recognition of the diagnostic features in the history, and the planning of the clinical examination to confirm or refute the provisional diagnosis *at the bedside*. The ultimate objective is to arrive at the correct diagnosis in the least number of moves and at the least risk to the patient; this must always be the overriding consideration.

Chapter 2

THE PUPILS AND THEIR REACTIONS

Due to the long intracranial and extracranial courses of the nerve pathways controlling the size and reactions of the pupils, lesions in many areas may cause pupillary abnormalities. Important differential diagnostic information may easily be overlooked by a cursory examination of the pupils or unnecessary investigations may be performed when a physiological inequality in pupil size is mistaken for a physical sign. In the unconscious patient the size and reactions of the pupils are of paramount importance in both diagnosis and minute-to-minute management of the patient.

BASIC EXAMINATION TECHNIQUE

1. The size, shape and symmetry of the pupils in moderate lighting conditions should be noted.

2. The direct and indirect responses of the pupils to a bright light should be elicited. An inadequate light source is the most frequent cause of absence of the light reflexes. The direct light reflex is the constriction of the pupil that occurs when it is directly illuminated. The indirect response or consensual light reflex is the simultaneous constriction of the opposite pupil.

3. The accommodation reaction should be tested. This reaction is the constriction of the pupil that automatically occurs as the patient attempts to converge the eyes. Failure to elicit this response is usually due to the patient's failure to converge the eyes. Convergence is most easily achieved if the patient tries to look at the end of his own nose, or at the examiner's finger brought in from below the line of the nose. The majority of people find it much easier to converge while looking downwards. It is helpful if the examiner holds the patient's eyelids up so that the pupils can be easily observed.

General Considerations

1. It is essential to establish whether any drops have been put in the patient's eyes. Many patients have been investigated for what later proved to be pharmacological pupillary inequality because of a failure to observe this simple rule.

2. If the pupils are unequal it is important to decide *which* is abnormal. A frequent mistake is to investigate for the cause of a dilated pupil on one side when the patient actually has a constricted pupil on the other side due to a Horner's syndrome!

3. If the pupils are unequal in size there are two additional features that may help establish the cause. If there is ptosis of the eyelid on the side of the *small* pupil the patient has a Horner's syndrome on that side. If there is ptosis on the side of the *large* pupil the patient has a partial third nerve lesion on that side. The light reflex and accommodation reflexes will be normal in Horner's syndrome and impaired in a partial third nerve lesion. In the absence of ptosis if both pupils react normally to light and accommodation the patient has "physiological anisocoria" (i.e. he has probably always had pupillary inequality). Many normal people have slight asymmetry in the size of the pupils.

4. Whenever a patient is found to have a widely dilated pupil that is fixed to light and accommodation without ptosis, the possibility that the patient has deliberately instilled atropine drops into the eye should always be considered. The writer has encountered this situation twice; both the patients were nurses.

5. Pupils are usually small in infancy, become larger in adolescence and are "normal" size in adulthood. In old age they again become small and the light reaction is accordingly difficult to see. Many elderly patients are incorrectly suspected of having Argyll–Robertson pupils because of this normal change in pupil size with age which is called "senile miosis" (miosis—pupillary constriction; mydriasis—pupillary dilation).

6. The pupils of many patients show a phasic constriction and dilation to light of constant intensity. This phenomenon is called "hippus" and has no definite pathological significance.

PUPIL SIZE AND REACTIONS

Pupil size is controlled by a ring of constrictor fibres innervated by the parasympathetic nervous system and a ring of radially arranged dilator fibres controlled by the sympathetic nervous system.

The resting size of the pupil is governed by the amount of light falling on either eye and depends on the integrity of the parasympathetic nerves. Increased activity in the sympathetic is reflected in slight pupillary dilation as occurs in an

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anxiety state. It is unusual for changes in pupil size to affect vision so that the majority of pupillary abnormalities are asymptomatic.

Parasympathetic Pathways (Figure 2.1)

The intensity of light falling on the retina is conveyed in the optic nerve to the optic chiasm. The impulses are then split and conveyed in *both* optic tracts to *both* lateral geniculate bodies. Some ten per cent of the fibres reaching the geniculate bodies subserve the light reflex and are relayed in the periaqueductal grey to both Edinger–Westphal nuclei and therefore light falling on *either* eye inevitably excites *both* nuclei and causes constriction of *both* pupils—the anatomical basis of the consensual light reflex.

The Edinger–Westphal nuclei are also stimulated by activity in the adjacent third nerve nuclear mass which controls the medial rectus muscles. therefore when both medial rectus muscles are activated in an attempt to converge the eyes, the Edinger–Westphal nuclei become active and constrict the pupils, this being the suggested basis of the accommodation reflex.

The parasympathetic fibres are carried in the third nerve to the orbit; and lie in a superficial and dorsal position which may explain the variable abnormalities of the pupil in third nerve palsies to be discussed later (Figure 2.1A).

The final relay of the pathway is in the ciliary ganglion in the posterior orbit, which gives origin to eight to ten short ciliary nerves, which sub-divide into sixteen to twenty branches, that pass around the eye to reach the constrictor muscle of the pupil.

Clinical Lesions affecting the Parasympathetic Control of the Pupil

1. The light reflex and the resting size of the pupil depend on adequate light perception by at least one eye. There is *no* direct light reaction in a completely blind eye, but the resting pupil size will be the same as the pupil size in the intact eye. If both eyes are blind both pupils will be dilated and fixed to light *if* the cause is located anterior to the lateral geniculate bodies. If, however, bilateral blindness is the result of destruction of the occipital cortex the light reflex pathways will be intact. Therefore, it is possible for a patient to be completely blind with preserved light reflexes in both eyes. Furthermore, if there is *any* perception of light in an eye that is for practical purposes blind, the light reaction may well be intact.

Lesions in the retina, optic nerve and chiasm and the optic tract of minimal degree, in particular optic nerve damage due to multiple sclerosis, cause what is known as an "afferent pathway lesion". This results in an abnormal pupillary response known as the Marcus Gunn pupil. When the normal eye is stimulated by a bright light there is no abnormality. When the affected eye is stimulated the reaction is slower, less complete, and so brief that the pupil may start to dilate again (the pupillary escape phenomena). The reaction is best seen if the light is rapidly alternated from one eye to the other, each stimulus lasting about a second with two or three seconds between. The reaction is thought to be due to a reduction in the number of fibres subserving the light reflex on the affected side.

2. A lesion of one optic tract does not affect the resting size of the pupil due to the consensual light reflex. In this situation a better light reflex may be seen if the light is shone on the intact half of the retina (see Chapter 3 for details of the field defect). This is called the Wernicke pupil reaction. It is very difficult to elicit this sign due to dispersion of light in the eye.

3. Lesions compressing or infiltrating the tectum of the mid-brain (the area of the superior collicular bodies) will interfere with the decussating light reflex fibres in the periaqueductal area. This results in pupils that are dilated and fixed to light. This is often coupled with loss of upward gaze and is known as the Parinaud syndrome (see Chapter 7).

4. The Argyll–Robertson pupil is also traditionally ascribed to damage in the periaqueductal area. The typical Argyll–Robertson pupil is small, irregular and fixed to light, but reacts to accommodation. It is the latter feature that suggests that the light pathways leading to the Edinger–Westphal nuclei are damaged, but this cannot explain the small size of the pupil or its irregularity. It is also worth noting that accommodation is a much stronger stimulus to pupillo-constriction than light and the apparent dissociation may merely reflect very minimal pupil reactivity. It has been suggested that a local lesion of the iris must be responsible. (The Argyll–Robertson pupil reaction is a classical sign of meningo-vascular syphilis.) Other causes of Argyll–Robertson pupils are pinealomas, diabetes and brain stem encephalitis. These conditions usually cause fixed *dilated* pupils; a *small* irregular fixed pupil is usually due to neurosyphilis. The Argyll–Robertson pupil cannot be dilated by atropine (Figure 2.2). Other causes of small pupils that are apparently fixed to light include senile miosis and the instillation of pilocarpine drops in the treatment of glaucoma.

5. Epidemic encephalitis lethargica, which last occurred in the 1920s, caused many cases of Parkinsonism associated with loss of the ability to converge the eyes. This disability produced pupils that reacted to light but not to accommodation. This "reversed" Argyll–Robertson pupil has become a clinical rarity.

Figure 2.1. The Parasympathetic Control of the Pupil

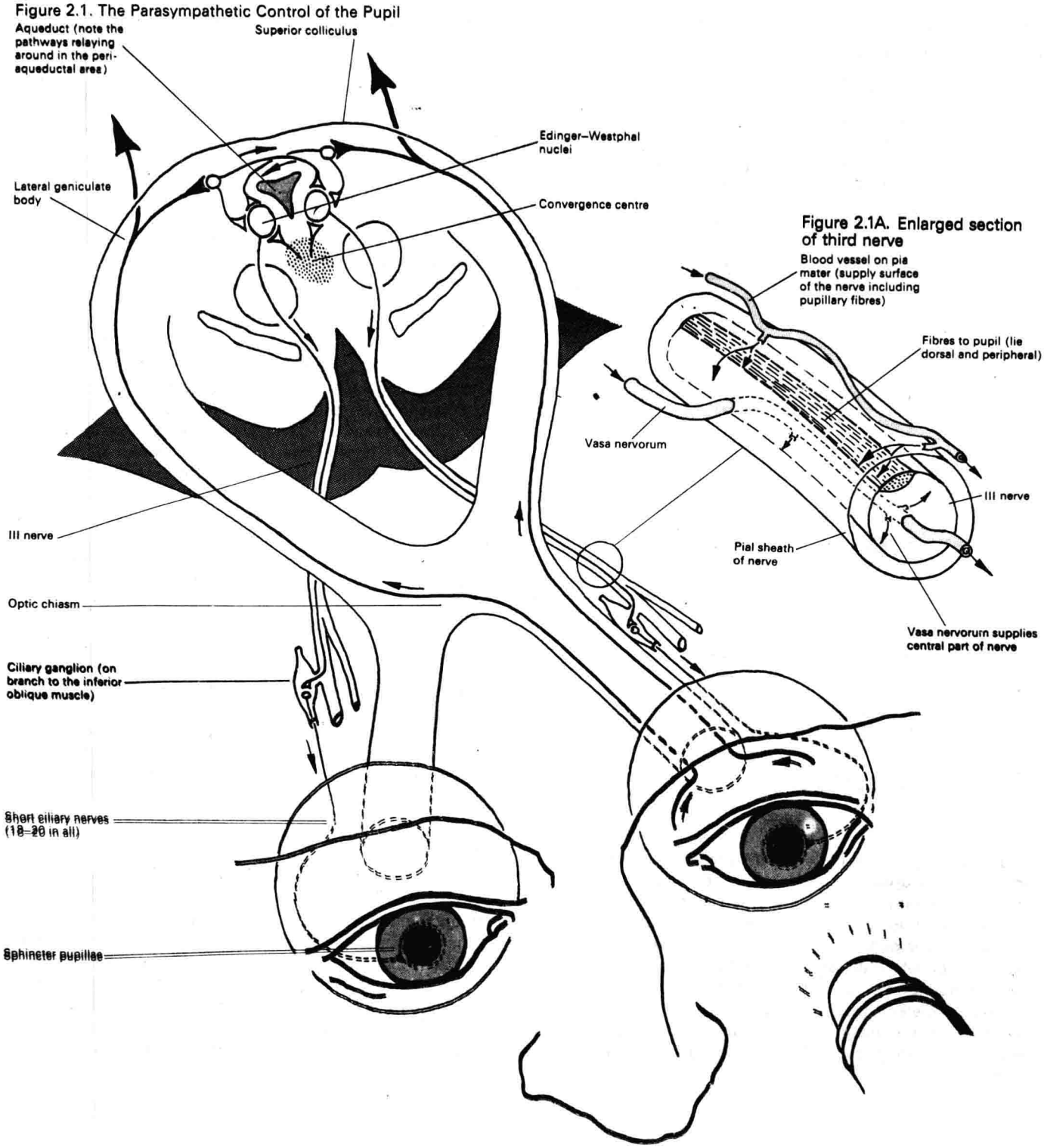


Figure 2.1A. Enlarged section of third nerve

Figure 2.2. Argyll–Robertson Pupil

Note. Ptosis, pupil is small, irregular, does not react to light, but does to accommodation

1% atropine solution in both eyes
No effect on A—R pupil
Wide dilatation of the normal pupil

(for illustrative purposes only, left pupil is shown as normal. A—R pupils are usually bilateral although they can be asymmetrical)

Figure 2.3. Holmes–Adie Pupil

Note. No ptosis, pupil is large, regular, does not react to light (other than prolonged exposure) but may show some response to accommodation

2.5% methacholine (mecholyl) in both eyes
The sensitised H—A pupil promptly constricts
No effect on the normal pupil

(Holmes–Adie pupils are usually unilateral—another feature to distinguish them from the A—R pupil)

6. Lesions affecting the third nerve may or may not involve the pupillary fibres. This is of great differential diagnostic value and will be discussed in detail in Chapter 5. At this point it is sufficient to note that if the nerve trunk is infarcted the superficial pupillary fibres may well be spared. As a general rule, if the pupil is affected the cause is surgical (i.e. compressive); and, if spared, the cause is medical (either diabetes, meningovascular syphilis or arteriosclerosis). The whole investigative approach to the patient with a third nerve lesion is influenced by the state of the pupil.

7. Degeneration of the nerve cells in the ciliary ganglion causes a Holmes–Adie or "tonic" pupil. The cause of this condition is unknown but it is often associated with loss of knee jerks and impairment of sweating. The Holmes–Adie pupil is a widely dilated, circular pupil that may react very slowly to very bright light, and shows a more definite response to accommodation. This dissociation again probably demonstrates the greater constrictive effect of accommodation. Both reactions are minimal and thought to be produced by slow inhibition of the sympathetic and *not* by any residual parasympathetic activity. The Holmes–Adie pupil is usually unilateral and more frequently found in females. It is often unnecessarily confused with Argyll–Robertson pupils which are small, irregular and usually bilateral. Congenital syphilis can cause fixed dilated pupils, but these are bilateral and other signs of congenital neurosyphilis will usually be found.

Confirmation of the diagnosis is best made by the pupillary response to $2\frac{1}{2}$ per cent methacholine drops. This chemical is too rapidly hydrolysed by acetylcholine-esterase to have any effect in the normal eye. In the denervated pupil, post-denervation hypersensitivity (due to enzyme depletion) allows the pupillo-constrictor effect to be seen. It has recently been shown that 80 per cent of diabetic patients react to methacholine in this way and 8 per cent of normal subjects also show a response. Therefore the test cannot be regarded as entirely specific for the Holmes–Adie pupil (Figure 2.3).

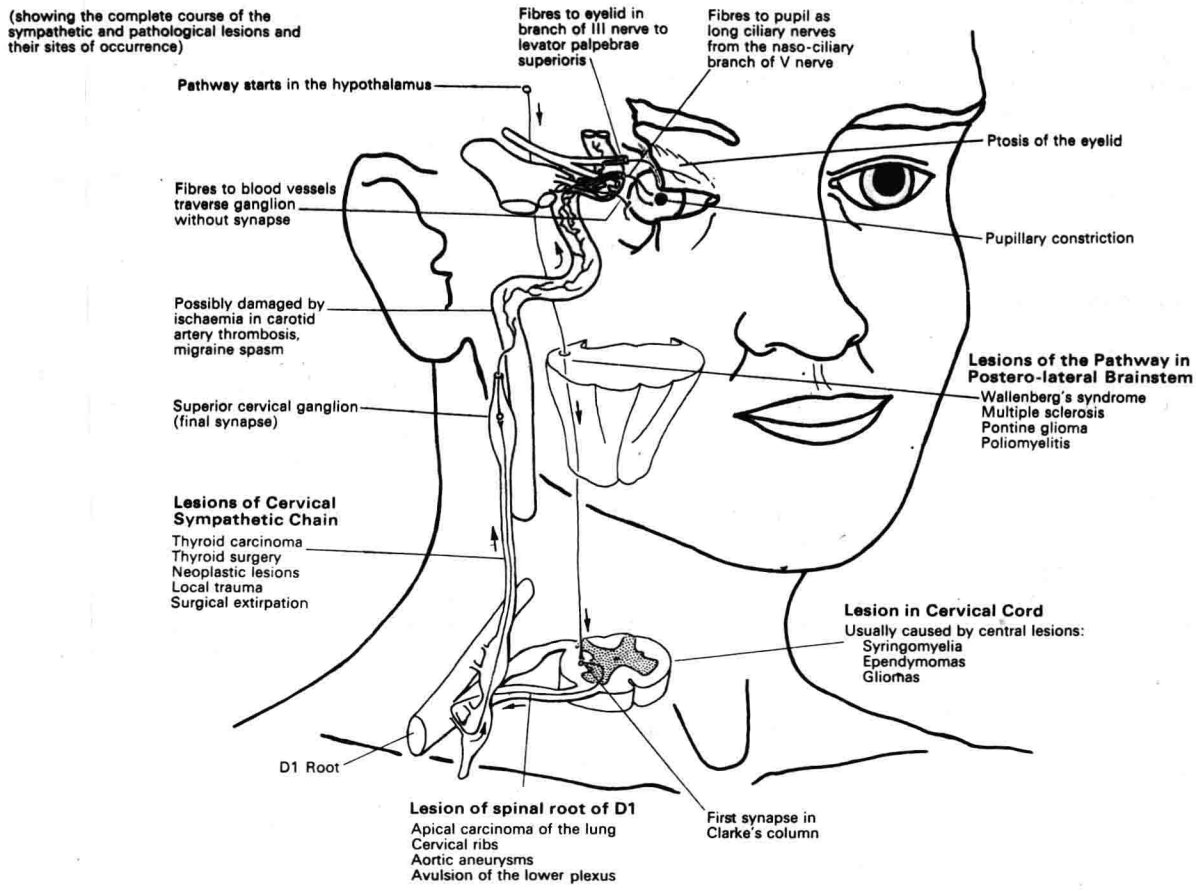
8. Blunt trauma to the iris may disrupt the fine short ciliary nerve filaments in the sclera causing an irregularly dilated pupil with impairment of the light reaction. A history of trauma is diagnostic. This is called post-traumatic iridoplegia (paralysis of the iris).

9. Diphtheria is a rare cause of pupillary dilatation due to damage to the ciliary nerves. It usually occurs in the second and third weeks of the illness and is often combined with the palatal paralysis. The pupillary abnormality usually recovers.

Sympathetic Pathway

The course of the cervical sympathetic pathway is shown in Figure 2.4. Although the pathway apparently starts in the hypothalamus there is a considerable degree of *ipsilateral* cortical control (see Chapter 10). A lesion anywhere in the pathway on the right side will affect the right pupil. There are three neurones. The first passes from the hypothalamus to the lateral grey in the thoracic spinal cord; the second from the spinal cord to the superior cervical ganglion and the third from the superior cervical ganglion to the pupil and blood vessels of the eye.

Figure 2.4. Horner's Syndrome



The third neurone enters the cranial cavity on the surface of the carotid artery and reaches the eye and eyelid as follows:

1. Fibres carried in the third nerve innervate the levator muscle of the eyelid.
2. Fibres in the nasociliary nerve traverse the ciliary ganglion without synapse to supply the blood vessels of the eye.
3. Other fibres branch from the nasociliary nerve as the long ciliary nerves to innervate the pupil by passing around the eye.

Abnormalities of the Sympathetic Pathway

Wherever the site of the lesion, a single physical sign results from damage to the sympathetic pathway and is known as Horner's syndrome. Associated physical signs allow the location of the causative lesion to be identified in some cases.

Horner's syndrome can be extremely subtle and the sign is easily overlooked by the less than obsessional examiner.

The features are as follows:

1. The affected pupil is slightly smaller than its fellow, due to reduced pupillodilator activity. This asymmetry is minimal in a bright light and exaggerated in darkness. The pupil reacts *normally* to light and accommodation but over a reduced range.
2. There is a variable degree of ptosis of the eyelid. In severe cases the lid may reach to the edge of the pupil, in other patients the ptosis may be barely detectable; and may vary from time to time during the day.

3. The conjunctiva may be slightly bloodshot (loss of vasoconstrictor activity).
4. Sweating over the forehead may be impaired, depending on the site of the lesion (see later).
5. In congenital Horner's syndrome the iris on the affected side fails to become pigmented and remains a blue-grey colour.
6. Enopthalmos (sunken eye) is not an easily detected feature of Horner's syndrome in man.

Causes of Horner's Syndrome

1. *Hemisphere Lesions*

Hemispherectomy or massive infarction of one hemisphere may cause a Horner's syndrome on the same side.

2. *Brain Stem Lesions*

The sympathetic pathways in the brain stem lie adjacent to the spinothalamic tract throughout its course. Hence Horner's syndrome due to a brain stem lesion is often associated with pain and temperature loss on the opposite side of the body. Vascular lesions, multiple sclerosis, pontine gliomas, and brain stem encephalitis may all cause a Horner's syndrome at this level (see Chapter 11).

3. *Cervical Cord Lesions*

Due to the central position of the pathway in the lateral column at D1 level the sympathetic is often involved in central cord lesions. Syringomyelia, cord gliomas or ependymomas will usually cause loss of pain sensation in the arms, loss of arm reflexes and often a *bilateral* Horner's syndrome. This can be very hard to detect as it is only the ptosis that draws attention to the condition—the pupils being small, but symmetrical and reactive! (see Chapter 15).

4. *D1 Root Lesions*

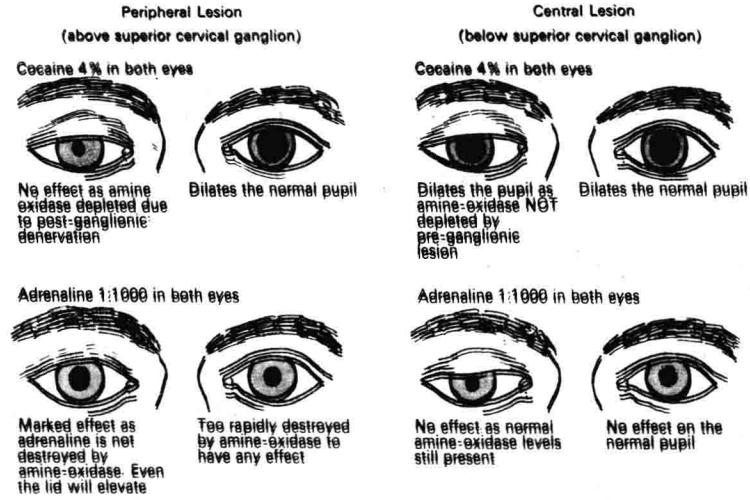
The D1 root is rarely affected by simple disc lesions, or degenerative disc disease. The root lies on the apical pleura where it may be damaged by primary or metastatic malignant disease. The classical syndrome of Pancoast, usually due to a cancer of the lung apex, consists of pain in the axilla, wasting of the small hand muscles and a Horner's syndrome, all on the same side. Other causes include cervical rib (usually in young females) and avulsion of the lower brachial plexus (Klumpke's paralysis). (See Chapter 18.)

5. *The Sympathetic Chain*

Throughout its course in the neck the sympathetic may be damaged by neoplastic infiltration, during surgical procedures on the larynx or thyroid, or surgically extirpated for a

Figure 2.5. Horner's Syndrome

Note. Slight ptosis. Small but normally reacting pupil



number of indications. Malignant disease in the jugular foramen at the skull base causes various combinations of Horner's syndrome and lesions of cranial nerves IX, X, XI and XII (see Chapter 6).

6. *Miscellaneous*

Congenital Horner's syndrome is not rare and has been mentioned previously. Horner's syndrome may occur during migraine headache. Lesions in the cavernous sinus or orbit usually damage both the sympathetic and the parasympathetic leading to a semi-dilated pupil that is fixed to light combined with other extra-ocular nerve palsies (see Chapter 5).

Other Features of Horner's Syndrome

The associated physical signs or history usually leave little doubt as to the site and cause of Horner's syndrome. There are some other useful diagnostic pointers.

Central lesions usually affect sweating over the entire head, neck, arm and upper trunk on the same side. Lesions in the lower neck affect sweating over the entire face. Lesions above the superior cervical ganglion may not affect sweating at all, as the main outflow to the facial blood vessels and sweat glands is below the superior-cervical ganglion.

The presence of three neurones in the pathway leads to some useful pharmacological tests based on the phenomenon of denervation hypersensitivity.

The decrease in amine-oxidase caused by a lesion at or beyond the superior cervical ganglion, sensitises the pupil to adrenaline 1 :1,000, which has no effect on the normal pupil.

Conversely, the effect of cocaine on the pupil *depends* on its blocking effect on amine-oxidase, therefore cocaine has no effect on a distally denervated pupil. It will only dilate the pupil in a Horner's syndrome if the lesion is below the superior cervical ganglion and there is amine-oxidase at the nerve endings for it to block.

These tests may be useful in the absence of other localising neurological signs in indicating the approximate site of the lesion. The pattern of responses are summarised in Figure 2.5.

PUPILLARY ABNORMALITIES IN THE UNCONSCIOUS PATIENT

The management of head injuries and the unconscious patient are discussed in Chapter 23.

1. *Normally Reacting Equal Pupils*

In an unconscious patient normal pupils are a reassuring sign indicating that no immediate surgical action is necessary. In the absence of a history of head trauma an immediate search for metabolic causes of coma should be initiated. Seventy per cent of unconscious patients have *not* had an intra-cranial catastrophe, but are in diabetic coma, hypoglycaemic coma, other metabolic coma, or have taken a drug overdose. Normal pupillary reactions are an important pointer to these possibilities. Only glutethamide and amphetamine (dilated pupils) or opiates (constricted pupils) cause misleading pupillary changes.

Table 1

PUPILLARY ABNORMALITIES

Reaction to Light	*Small Pupils*	*Large Pupils*
Non-reactive	A–R pupil	Holmes–Adie pupil
	Pontine haemorrhage	Post-traumatic iridoplegia
	Opiates	Mydriatic drops
	Pilocarpine drops	Glutethamide overdose
		Cerebral death
		Atropine poisoning
		Amphetamine overdose
Reactive	Old age	Childhood
	Horner's syndrome	Anxiety
		Physiological anisocoria

2. *Unequal Pupils*

This is the single most important physical sign in the unconscious patient. Until proved otherwise a dilated pupil indicates that a herniated temporal lobe is stretching the third nerve on that side and prompt surgical action is required. Problems occur if the eye was directly damaged in the injury, or if someone has put mydriatic drops in the eye in the pointless search for papilloedema (pointless because patients with acute head injury will die long before papilloedema can appear).

3. *Bilateral Dilated Pupils*

The final stage of progressive tentorial herniation is heralded by progressive dilatation of the previously unaffected pupil. The chances of the patient recovering from this stage are remote. This is also used as one of the signs of irreversible cerebral damage in cardiac arrest. Glutethamide, atropine (mushroom) and amphetamine poisoning are the only metabolic causes of bilateral dilated pupils.

4. *Bilateral Pin-point Pupils*

This situation is the hall-mark of another lethal neurologi-

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