Clinical Neurology
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Foreword

There have been many attempts over the years to distil the knowledge needed for medical students and young doctors to begin to engage in neurological diagnosis and treatment. This book by Professor Peter Gates is one of the best books developed to date. Peter Gates is an outstanding clinical neurologist and teacher who has been acknowledged in his own university as one of the leading teachers of undergraduates and registrars in recent times. He takes a classical approach to neurological diagnosis stressing the need for anatomical diagnosis and to learn as much as possible from the history in developing an understanding of likely pathophysiologies and aetiologies. In this book he sets out the lessons of a lifetime spent in clinical neurology and distils some of the principles that have led him to become a master diagnostician.

The first chapter is devoted to neuroanatomy from a clinical viewpoint. The concept of developing diagnosis through an understanding of the vertical and horizontal meridians of the nervous system is developed and intriguingly labelled under latitude and longitude. All the key issues around major anatomical diagnosis are distilled in a very understandable way for the novice. This chapter (and subsequent chapters) is widely illustrated with case studies and the illustrations are excellent. Key points are emphasised and important clinical questions stressed. A great deal of thought has gone into the clinical anecdotes chosen to illustrate major diagnostic issues. These reflect the learnings of a lifetime spent in neurological practice.

Subsequent chapters take the reader through the neurological examination and major neurological presentations and neurological disorders. Key aspects are illustrated with great clarity. This is a book that can be consulted from the index to get points about various disorders and their treatments but, more importantly, should be read from cover to cover by young doctors interested in coming to terms in a more major way with the diagnosis and treatment of neurological disorders. It also contains a lot of material that will be of interest to more experienced practitioners. The book has a clinical orientation and the references are comprehensive in listing most of the relevant key papers that the reader who wishes to pursue the basis of clinical neurology further may wish to consult. The final chapter is an excellent overview of how one can approach information gathering and keeping up-to-date using the complex information streams available to the medical student and young doctor today.

This book is clearly aimed at medical students and young doctors who have a special interest in developing further understanding of the workings of the nervous system, its disorders and their treatments. I would recommend it to senior medical students, to young doctors at all stages and also to those beginning their neurological training. It also has some information that may be of interest to the more senior neurologist in terms of developing their own approach to teaching young colleagues. It is the best introduction to the diagnosis and treatment of nervous system disorders that I have seen for many years and contains a font of wisdom about a speciality often perceived as difficult by the non-expert.

Professor Edward Byrne AO
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Preface

This book was written with two purposes in mind: firstly, it is an introductory textbook of clinical neurology for medical students and hospital medical officers as well as neurologists in their first year(s) of training and, secondly, it is designed to sit on the desk of hospital medical officers, general practitioners and general physicians to refer to when they see patients with the common neurological problems.

This book is the culmination of 25 years of clinical practice and teaching in neurology and is an attempt to make neurology more understandable, enjoyable and logical.

The aim is to provide an approach to the more common neurological problems starting from the symptoms that are encountered in everyday clinical practice. It describes how best to retrieve the most relevant information from the history, the neurological examination, investigations, colleagues, textbooks and the Internet.

This book in no way attempts to be a comprehensive textbook of neurology and as such is not intended for the practising neurologist. There are and will always be many excellent and comprehensive books on neurology.

There are chapters on the examination technique as well as a DVD that demonstrates and explains the normal neurological examination together with some abnormal neurological signs.

Investigations and treatments in the text will very quickly be out of date but the basic principles of clinical neurology developed more than 100 years ago are still relevant now and will be for many years to come. The clinical neurologist is like an amateur detective and uses clues from the history and examination to answer the questions: ‘Where is the lesion?’ and ‘What is the pathology?’

Although it is not intended for the experienced neurologist, the author is aware of some neurologists who have found some of the techniques in this textbook (e.g. the Rule of 4 of the brainstem) useful in their teaching.

The original title of this book was Neurology Demystified, after a general physician commented to the author that the ‘Rule of 4’ of the brainstem had demystified the brainstem for the general physician. It encapsulates what this author has been attempting to achieve over the past 30 years of teaching neurology: to make it simpler and easier to understand for students, hospital medical officers, general practitioners and general physicians.

One of the most rewarding things in life is teaching those who are interested in learning and to see the sudden look of understanding in the eyes of the ‘student’.
Acknowledgements

I would like to thank my colleagues John Balla, Ross Carne, Richard Gerraty and Richard McDonnell for reviewing sections of the manuscript. At Elsevier, I also wish to thank Sophie Kaliniecki for accepting my book proposal, Sabrina Chew, Eleanor Cant and Linda Littlemore for all the support and encouragement they provided during the writing of the manuscript, and also to Greg Gaul for the illustrations. I would particularly like to thank Stephen Due and Joan Deane at the Geelong Hospital library who have been a tremendous support over many years, especially but not only during the writing of this book. Also, I thank the radiologists and radiographers at Barwon Medical Imaging for providing most of the medical images. My thanks also to the many patients and friends who generously consented to have pictures or video taken to incorporate in this book. Kevin Sturges from GGI Media Geelong, a friend and technological whiz, helped me with all the images and video production.

Thank you also to the students and colleagues who anonymously reviewed the manuscript for their many wonderful suggestions and words of encouragement.

I have indeed been fortunate to have been taught by many outstanding teachers during my training and, although to name them individually runs the risk of omission and causing offence, there are a few that I would like to acknowledge: Robert Newnham, rheumatologist at the Repatriation General Hospital in Heidelberg who, in 1975, first taught the symptom-oriented approach while I was a final-year medical student; at St Vincent’s Hospital in Melbourne the late John Billings, neurologist, who introduced me to the excitement of neurology and John Niall, nephrologist, for challenging me to justify a particular treatment with evidence from the literature; Arthur Schweiger, John Balla, Les Sedal, Rob Helme, Russell Rollinson and Henryk Kranz (neurologists) for the opportunity to enter neurology training at Prince Henry’s Hospital Melbourne where John Balla encouraged me to write my first paper; Lord John Walton, neurologist, for the opportunity to work and study in Newcastle upon Tyne; Peter Fawcett, neurophysiologist, for the opportunity to study neurophysiology; Dr Mike Barnes, neurologist in rehabilitation, who helped in 1983 at the Newcastle General Hospital to make the video of John Walton taking a history.

I also wish to thank Henry Barnett, neurologist, in London, Ontario, for the opportunity to work on the EC-IC bypass study; and Dave Sackett, Wayne Taylor and Brian Haynes in the department of epidemiology at McMaster University for opening my eyes to clinical epidemiology and evidence-based medicine. And last but not least my long standing friend Ed Byrne, currently Vice-Chancellor of Monash University, for the appointment at St Vincent’s Hospital in Melbourne on my return from overseas and for his friendship and wise council over many years.

This book is dedicated to my children, Bernard, Amelia and Jeremy, and my wife Rosie, for without their support over the many years this project would not have been possible.
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After the History and Examination, What Next?

Upon completing the history and examination, the next step is determined by the following factors:

- diagnostic certainty
- the availability of tests to confirm or exclude certain diagnoses
- the potential complications of those tests
- the severity and level of urgency in terms of the consequences of a particular illness not being diagnosed and treated promptly
- the benefit versus risk profile of any potential treatment
- the presence of any social factors or past medical history that could influence a course of action or treatment in this particular patient.

This chapter will discuss each of these aspects and how they influence the course of action.

LEVEL OF CERTAINTY OF DIAGNOSIS

There are three possible scenarios:

1. A particular diagnosis seems obvious.
2. One particular diagnosis is not apparent, and there are several possible diagnoses.
3. You have no idea what is wrong with the patient.

A particular diagnosis seems certain

In most instances the diagnosis is apparent. In the general practice setting almost 90% of diagnoses are established at the completion of the history and examination [1]. In one outpatient clinic this figure was 73% (history 56% and examination 17%) in patients with cardiovascular, neurological, respiratory, urinary and other miscellaneous problems [2]. In patients with neurological problems the initial diagnosis is less obvious and was correct in only 60% of patients presenting to an emergency department [3]. In this setting the appropriate course of action is to initiate investigations that can confirm the diagnosis, exclude alternative diagnoses with potentially more severe adverse outcomes and institute a plan of management taking into account factors in the past, social and medical drug history that would influence management in this particular patient.

A word of caution: being absolutely certain is potentially the most dangerous scenario. Doctors are strongly anchored by their initial diagnoses [4] (see Case 6.1) and are at risk of closing their minds to possible alternatives, often in the presence of clinical features or results from investigations that should raise doubt about the diagnosis.
Doctors recognise patterns of familiar problems with respect to critical cues [6]. Doctors who are more experienced appear to weigh their first impressions more heavily than those who are less experienced and at risk of closing their minds early on in the diagnostic process [7]. Even experienced clinicians may be unaware of the correctness of their diagnoses when they initially make them [8].

If there are tests to confirm the diagnosis, it is appropriate to perform those tests, provided the patient is informed of the risks associated with them. When ordering tests and reviewing the results, it is most important to be aware of the sensitivity and specificity and the influence of the prevalence of the disease on the positive predictive value and the negative predictive value of those tests [9] (for a discussion of these concepts, refer to the section ‘Understanding and interpreting test results’ below).

If there are no tests, one can proceed cautiously with management, but it is most important to review the response to therapy. A lack of response to therapy or the emergence of unexpected side effects (the latter is a personal observation) is often a clue that the diagnosis is incorrect. Conversely, a response to a therapy does not prove the diagnosis. This author has seen patients with vertebral artery dissection, viral meningitis and pituitary cysts ‘respond’ to treatment for migraine. This is discussed in more detail below.

There are several possible diagnoses

It is imperative to keep the diagnostic options open by making provisional diagnoses while keeping alternatives in mind. Be circumspect and take action to minimise the possibility of missing other critical diagnoses [10]. Once again, if there are tests that can differentiate one particular diagnosis from another, it would be most appropriate to perform those tests. If a specific diagnosis cannot be made following the investigations, the approach is similar to that discussed in the following section.

You have no idea what is wrong

In the setting of uncertainty there are several possible courses of action. A particularly useful strategy is to start again: take a more detailed history and repeat the examination.¹ This is the approach recommended when you have absolutely no idea what the diagnosis is. In this situation performing many tests is often misleading because of the sensitivity and specificity of tests.

If you have elicited a detailed history, but still have no idea what is wrong with the patient, there are several options including:

• wait and see
• perform investigations

¹ Recommended to the author by Dr Arthur Schwieger in 1985 and, to this day, remains one of the most powerful clinical tools available.
• seek a second opinion
• consult a textbook
• search for an answer on the Internet.

These various approaches will be discussed in terms of their relative merits and deficiencies.

WAIT AND SEE
In resolving uncertainty, time is a very powerful diagnostic tool. The idea is to wait for a period of time in the hope that the diagnosis becomes clear or the patient gets better [10], [11]. The effective use of this approach requires considerable skill, however. Often in this situation a doctor may order unnecessary tests in the hope that a diagnosis may be established; most often it is not. If the ‘wait and see’ approach is adopted, it is important to:
• inform the patient that there is uncertainty
• advise them of the possible outcomes
• recommend that they report immediately should symptoms worsen or if any new symptoms develop.

Shared medical decision making is a process in which patients and providers consider outcome probabilities and patient preferences and reach a healthcare decision based on mutual agreement. Shared decision making is best employed for problems involving medical uncertainty [12]. However, it is important to consider the fact that not all patients wish to be involved in shared medical decisions [13].

UNDERTAKE INVESTIGATIONS
In most cases there are tests that can confirm or exclude a particular disease. In this situation it is important to understand the concepts of the sensitivity and specificity of tests and the importance of prevalence of the disease. The essential questions to ask when considering investigations include:
• In what way will the results, whether positive or negative, alter the management of this patient?
• What is the risk of undertaking the test?

There is a more detailed discussion of investigations later in this chapter.

There are no tests for some diseases and the diagnosis is based entirely on the clinical features. When there are several possible diagnoses or when one has absolutely no idea what the diagnosis might be, performing numerous tests in the hope of making a diagnosis is a wonderful way of giving the illusion that something useful is being done when often all that may be achieved is stalling or buying time. It is a tactic that is used by a number of clinicians in the hope that a diagnosis will be made by a test result (unlikely), the illness will progress so that the diagnosis will become apparent or the patient’s problem will resolve. A reasonable approach is to think of the worst case scenario (the most serious diagnosis that the symptoms could represent, a diagnosis that if missed could result in an adverse outcome) and proceed accordingly.

OBTAIN A SECOND OPINION
Although doctors prefer to obtain information from journals and books, they often consult colleagues to get answers to clinical and research questions [14], [15]. Even for doctors whose first choice of information source was the medical literature – either books or journals – the most frequent second choice was consultations [14].

In a study of 254 referrals seen by a neurologist there was a significant change in diagnosis in 55%, and in management in nearly 70% [16].
There are several ways of obtaining a second opinion:

- the ‘informal consultation with a colleague’
- telephone advice
- the ‘curbside’ conversation in the corridor without actually seeing the patient
- presenting at meetings and seeking several opinions, often but not invariably with the patient at the meeting
- the ‘formal second opinion’ when a colleague is asked to see the patient.

**Telephone advice**
Telephoning a colleague for advice is common. The recipient of the call is in a very difficult situation as providing the correct advice very much depends on being given the correct history and examination findings. An experienced clinician often knows the particular questions to ask and can decide if and when they should actually see the patient. It is probably wiser for an inexperienced clinician to arrange to formally see the patient in consultation.

**Corridor or curbside consultation**
‘Corridor or curbside consultation’ is another approach used often [17]. Unfortunately, and sometimes with dire consequences, this is used by medical practitioners to seek informal advice about their own medical problems. The model of a good curbside consultation ‘is to say what you know and what you don’t know. Then you hope the person you are consulting with will treat you with respect’ [17]. Requesting doctors who could not present relevant information, frame a clear question or answer consultant questions in a well-informed manner were generally asked to formally refer the patient [17]. Perley et al [17] commented that tacit rules govern curbside consultation interactions, and negative consequences result when the rules are misunderstood or not observed.

Once again, the correct advice very much depends on being given the correct information. The neurologist providing advice will want to know the mode of onset and progression of the symptoms of the current illness together with the EXACT nature and distribution of the symptoms and the abnormal neurological signs, if present. It is difficult for inexperienced clinicians to perform detailed neurological examinations but there should be no reason why, as outlined in Chapter 2, ‘The neurological history’, an inexperienced clinician cannot obtain a detailed history. Finally, the neurologist would want information about the social, past and drug history that may influence any subsequent course of action.

**Clinical meetings**
Second opinions are often sought for the purpose of diagnosis and/or treatment in clinical meetings. There is the perception that one is obtaining multiple opinions. Although this can be a valuable tool, particularly if one of the participants identifies the problem, in a more complex case often what is said in meetings is very different to what is said in a formal consultation. Thus, the advice obtained in clinical meetings should be viewed circumspectly. A brain biopsy is often recommended in clinical meetings, but not often performed despite the recommendation.

**The formal second opinion**
The formal second opinion is probably the most effective method of dealing with diagnostic or therapeutic uncertainties. *There is no shame in asking a colleague to formally see the patient for a second opinion.* If you do, it can be a learning experience; if you do...

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2 Personal observation.
not and the patient perceives a lack of progress, they will independently seek a second opinion and you will miss out on a learning opportunity. If a patient requests a second opinion, NEVER hesitate to arrange one.

Remember a second opinion is simply that; you are the clinician caring for the patient and the ‘buck stops with you’. You must decide if the second opinion is useful or not and act accordingly – this may include obtaining a third opinion!

CONSULT A TEXTBOOK
Yet another approach is to consult textbooks. This is useful if you are looking at the clinical features of a particular disease(s) or to learn what investigations would be appropriate. However, therapy is evolving so rapidly that recommended therapy in textbooks is soon out of date.

SEARCH THE INTERNET
An increasingly popular and useful strategy is to search the Internet3 [18–20]. Patients frequently look for answers on the internet [21]. In the author’s own experience many patients bring the results of their searches to the consultation. In one study [19] Google was able to make the correct diagnosis in 58% of the cases in the *New England Journal of Medicine* clinical-pathological conferences. In a comparison of PubMed, Scopus, Web of Science and Google Scholar, the keyword search function of PubMed was superior. While Google Scholar could retrieve the most obscure of information, its use was marred by inadequate and less frequently updated citation information [22]. Searching in Google Scholar can be refined by adding + emedicine to the search [23]. For example, ‘trigeminal neuralgia’ yields 48,000 ‘hits’ while ‘trigeminal neuralgia + emedicine’ retrieves 478 references. Many remain skeptical [24] and, as recently as 2 years ago, Twisselmann stated that the jury is still out on whether searching for symptoms on the Internet is the way forward for doctors and consumers [25].

The author has adopted the practice of frequently consulting the Internet even in the midst of a formal consultation.4 It is a useful way to look for any new advances in therapy, to provide information to the patient or referring practitioner by adding the abstracts and references to the letter or even to search for an obscure diagnosis (see Case 6.2).

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**CASE 6.2  A trumpet player with nasal escape**

This example illustrates how this approach can be very useful. A young trumpet player developed nasal escape of air after 30 minutes of playing and could not continue to play. The symptoms took too long to develop and persisted for too long after cessation of playing to be related to myasthenia gravis. A quick search of ‘trumpet player and nasal escape’ revealed the diagnosis of a very rare condition termed velopharyngeal incompetence [26].

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3 Websites with instructions for searching the Internet for medical information are at the end of this chapter. Chapter 15, ‘Further reading, keeping up-to-date and retrieving information’, lists many relevant websites together with their URLs.

4 It is important to inform the patient that you are looking up something that might help with their problem and not to spend too long or the patient will feel neglected. If you cannot find a ready answer, continue the search at a later time.
An online information retrieval system [27] was associated with a significant improvement in the quality of answers provided by clinicians to typical clinical problems. In a small proportion of cases, use of the system produced errors [27]. Despite ready access to the Internet many doctors do not yet use it in a just-in-time manner to immediately solve difficult patient problems but instead continue to rely on consultation with colleagues [28, 29]. One major obstacle is the time it takes to search for information. Other difficulties primary care doctors experience are related to formulating an appropriate search question, finding an optimal search strategy and interpreting the evidence found [29].

Computer programs that can be used as an aid in diagnosing multiple congenital anomaly syndromes have been used for many years and are designed to aid the paediatrician diagnose rare disorders in children [30]. Other computer-aided software systems for diagnosing neurological diseases exist [31], and it is likely that more software will be developed in the future. Such software will always be dependent upon the information provided by the user.

AVAILABILITY OF TESTS TO CONFIRM OR EXCLUDE CERTAIN DIAGNOSES

This section discusses the general principles of investigations or tests. Essentially it will cover why tests give the ‘wrong’ or unexpected result and what to do when this occurs. There are many excellent books that discuss the interpretation of tests in great detail [32–34].

Understanding and interpreting test results

SENSITIVITY, SPECIFICITY, POSITIVE AND NEGATIVE PREDICTIVE VALUES

In order to understand how to interpret investigations correctly, you need to understand some basic principles. All tests have an associated sensitivity, specificity and positive and negative predictive values and are very much influenced by the prior likelihood that the disease is present in a particular patient. The usefulness of a test is very dependent on the prior probability that a patient has a particular disease, i.e. the prevalence of the disease.

- **Sensitivity** refers to how good a test is at correctly identifying people who have the disease.
- **Specificity** is concerned with how good the test is at correctly excluding people who do not have the condition.
- **Positive predictive value** refers to the chance that a positive test result will be correct.
- **Negative predictive value** is concerned only with negative test results.

For any diagnostic test, the positive predictive value will fall as the prevalence of the disease falls while the negative predictive value will rise. In practice, since most diseases have a low prevalence, even when the tests we use have apparently good sensitivity and specificity, the positive predictive value may be very low.

Table 6.1 shows the results of a test with a sensitivity of 90% and a specificity of 80%. When the test is performed on 100 patients with the suspected diagnosis, 10 patients with the diagnosis will have a negative test while 20 patients who do not have that particular diagnosis will have an incorrect positive test. The ideal test would be one with 100% sensitivity and 100% specificity, but this does not occur.

The pre-test likelihood of a patient having a particular diagnosis also greatly influences how a test result should be interpreted. Using the same values for sensitivity and
specificity, if 100 patients are tested for a particular diagnosis when only 50 of them have that diagnosis (see Table 6.2), a positive test will detect 45 of the patients with the disease (90% of 50) but the test will also be positive in 10 (20% of 50) patients who do not have the disease!

If the prior probability of a particular diagnosis being present is even lower, the results will be even more dramatic. If the patient is very unlikely to have the disease, say a 10% chance (i.e. 10 in every 100 patients tested), with the same sensitivity and specificity of 90% and 80%, respectively, a positive result will correctly identify 9 patients with the disease but will incorrectly diagnose 18 patients without the disease (90% of 10 = 9 and 20% of 90 = 18). The rarer the problem, the more certain we can be that a negative test excludes that disease, but less certain that a positive test indicates an abnormality (see Table 6.3).

In this setting a negative test in the presence of a strong suspicion of a diagnosis may lead inexperienced clinicians to dismiss that diagnosis. The antithesis of this is a patient being incorrectly diagnosed with a particular illness because of a false positive test.

The variability in prevalence of a particular disease between one study and another means that predictive values found in one study do not apply universally [35]. A common practice of inexperienced doctors is to repeat borderline abnormal tests simply because the result is ‘outside the normal range’ even when the test result is irrelevant to

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**TABLE 6.1** The influence of sensitivity (90%) and specificity (80%) of a test on the results for 100 patients with the suspected diagnosis

<table>
<thead>
<tr>
<th></th>
<th>+ve Test</th>
<th>–ve Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient has disease</td>
<td>90</td>
<td>10</td>
</tr>
<tr>
<td>Patient does not have disease</td>
<td>20</td>
<td>80</td>
</tr>
</tbody>
</table>

**TABLE 6.2** The effect of a 50% likelihood that the patient has a disease and the influence of the sensitivity and specificity on the number of correct diagnoses (n = 100)

<table>
<thead>
<tr>
<th></th>
<th>+ve Test</th>
<th>–ve Test</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient has disease</td>
<td>45</td>
<td>5</td>
</tr>
<tr>
<td>Patient does not have disease</td>
<td>5</td>
<td>45</td>
</tr>
</tbody>
</table>

*Note: An equal number of patients with the disease will have a negative test as patients without the disease will have a positive test.*

**TABLE 6.3** The number of positive and negative test results when the likelihood of a particular diagnosis is low (10%), i.e. doing tests to exclude rare conditions causes more problems than it solves (n = 100)

<table>
<thead>
<tr>
<th></th>
<th>+ve Test</th>
<th>–ve Test</th>
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<tr>
<td>Patient has disease</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>Patient does not have disease</td>
<td>18</td>
<td>72</td>
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the clinical problem. In this situation it is better to discuss the result with the relevant pathologist or radiologist.

INCIDENTAL AND IRRELEVANT FINDINGS ON TESTS: THE ‘INCIDENTALOMA’

An ‘incidentaloma’ is the finding of an abnormality on a test, usually some form of medical imaging, which is not related to the patient’s symptoms and signs. Typical examples of this include the presence of a degenerative disease in the cervical, thoracic or lumbar spine with advancing age that may be completely asymptomatic, an asymptomatic lacunar cerebral infarct or an unidentified bright object on MRI scan in a patient being investigated for headaches. Often only experience can teach and provide the confidence to ignore such incidental findings.

WHY TESTS GIVE THE WRONG RESULT AND WHAT TO DO WHEN THIS OCCURS

Although the sensitivities and specificities of investigations largely explain why tests may be negative or positive in the wrong setting, a test may also be negative for a number of other reasons. Patients with intermittent disturbances of neurological function will usually have normal tests between events; the test will only be positive if the examiner happens to capture an event and, if episodes are infrequent, this is unlikely. Symptoms can also precede the development of abnormalities that can be detected with currently available investigations, e.g. carpal tunnel syndrome, acute inflammatory demyelinating peripheral neuropathy, where the nerve conduction studies can be normal in the early stages, or patients with a cerebral infarct, where a CT scan of the brain can be normal for several hours after the onset of the infarction.

Other causes of a ‘negative test’ are when the test is directed to the wrong part of the body or the test that is ordered is not suitable for detecting abnormalities in that region. For example, a patient may present with difficulty walking and a normal CT scan of the lumbosacral spine when they have a problem in the cervical or thoracic spinal cord, and a CT scan of the thoracic spine is negative because it is not a sensitive enough test for detecting abnormalities in this region.

The relative ‘fallibility’ of tests emphasises the importance of the detailed history and examination. If you are absolutely certain that a patient has a particular diagnosis, then a negative test should not dissuade you from that diagnosis. The corollary of this is a positive test should not imply a diagnosis if the symptoms and signs are not consistent with that diagnosis.

HOW QUICKLY SHOULD TESTS BE PERFORMED?

This is discussed below in the section ‘Severity and urgency: the potential consequences of a particular illness not being diagnosed and treated’.

THE POSSIBLE COMPLICATIONS OF TESTS

There are very few tests that are not associated with risk. Venesection is perfectly safe in close to 100% of patients but rarely can be associated with injury to a nerve that can result in long-term pain and dysaesthesia. Although this complication is extremely rare (<0.02% [36]), the result can be very distressing.

When ordering any investigation it is important to consider the potential complications of the test in relation to the seriousness of the illness that is being investigated. A patient with a life-threatening illness might be willing to consider a potentially life-threatening investigation if it could make a significant difference; on the other hand,
a patient with symptoms without disability would be concerned about any investigation that might cause harm.

It is most important that the patient is fully informed of the risks versus benefits of the procedure beforehand.

SEVERITY AND URGENCY: THE POTENTIAL CONSEQUENCES OF A PARTICULAR ILLNESS NOT BEING DIAGNOSED AND TREATED

In everyday clinical practice if a diagnosis is clearly established, knowledge of the natural history of this condition would dictate how quickly one would investigate and treat the patient. Clearly, patients presenting comatose or with status epilepticus (a seizure that lasts more than 30 minutes, or recurrent seizures without return of consciousness between seizures) require urgent intervention.

The difficulty arises in the patient with a neurological problem when there is uncertainty as to the diagnosis. There is very little in the literature that can provide guidance in this area. Scoring tools for priority setting for general surgery and hip and knee surgery were useful but were not particularly good for MRI scanning [37]. The discussion below contains observations made by this author during many years of clinical practice and observations from colleagues who were asked specifically, ‘What do you think constitutes an urgent problem?’

The overriding principle is to consider the worst case scenario. It is prudent to consider the most serious possible diagnosis that, if left untreated, could result in significant morbidity or mortality. This will dictate the ‘level of urgency’ and how promptly a doctor should act (see Case 6.3).

CASE 6.3 Assessing urgency in a patient with an uncertain diagnosis

An example is a patient who presents with symptoms that suggest cerebral ischaemia, but the diagnosis is far from certain. Discuss with the patient that there is uncertainty regarding the diagnosis and that, if it is not treated as cerebral ischaemia when it is (i.e. an inappropriate course of action is taken), the consequences could be potentially disastrous. On the other hand, if investigation and treatment on the basis that it may be cerebral ischaemia can be undertaken without significant risk, then often that is a reasonable approach until the diagnosis can be clarified with more certainty.

Experienced clinicians can often accurately assess the level of urgency in a particular clinical setting. Although this may well relate to their level of expertise and ‘having seen it before’, more often it is because they use the tempo of the illness (the rapidity of development of symptoms and signs) to dictate how quickly they should act.

- Rapidly evolving weakness dictates immediate action.
- Although not all patients with symptoms related to the spinal cord have urgent neurological problems, disorders in this region can result in devastating neurological deficits and the degree of recovery is very dependent on the severity of the spinal cord lesion [38]. The investigations should be prompt if one suspects spinal cord disease. One would consider spinal cord problems with leg weakness and, particularly if there is associated sphincter disturbance, with bilateral leg weakness if the

5 'The author was once told by a senior consultant 'the spinal cord is unforgiving'.
lesion is in the thoracic spinal cord and four-limb weakness if the lesion is in the cervical spinal cord.

• Similarly, patients with symptoms related to the brainstem such as diplopia, dysphagia and vertigo, particularly if combined with ataxia or limb weakness, should be investigated as a matter of urgency.

• Patients with recurrent symptoms within a short period of time should also be dealt with as a matter of urgency. As a general rule, symptoms of weakness are more likely to imply significant neurological problems rather than isolated sensory symptoms.

• Similarly, symptoms associated with loss of function are more likely to be significant than symptoms without functional loss. Patients with multiple symptoms not associated with any loss of function, particularly if also associated with non-neurological symptoms, are less likely to have a serious illness requiring urgent intervention. Transient symptoms lasting seconds are also unlikely to be of any significance. One study found that higher numbers of physical symptoms and the complaint of pain were indicators of possible non-organic disease [39].

A summary of urgent and non-urgent presentations is given in Table 6.4. A simple rule is: ‘if in doubt do not hesitate to ask for help’.

### THE BENEFIT VERSUS RISK PROFILE OF ANY POTENTIAL TREATMENT

All medical interventions, whether they are pharmacological or surgical, have the potential to cause harm.

• Most patients can tolerate most drugs with few or no side effects. When a diagnosis is clearly established, the choice of the appropriate treatment would primarily be dictated by the knowledge that one particular therapy has greater efficacy than another.

• On the other hand, if there are several treatments with equal efficacy, the choice of therapy would then depend on the risk profile and the patient’s willingness to consider particular side effects. For example, there may be two or three drugs that could be used to treat a patient who suffers from epilepsy; the drugs that may cause weight gain or interfere with the oral contraceptive pill would be most unacceptable to a young female patient.

• In the setting where the diagnosis is uncertain and one is instituting empirical therapy, it is important to inform the patient of the perceived benefits of the therapy prescribed but also to alert the patient to the potential risks of that therapy.
importantly, carefully monitor the response to therapy and be willing to reconsider
the diagnosis and/or choice of therapy.

SOCIAL FACTORS AND PAST MEDICAL PROBLEMS THAT MAY
INFLUENCE A COURSE OF ACTION OR TREATMENT

This has already been discussed briefly in Chapter 2, ‘The neurological history’, where
the importance of not using information about the past history, family history and
social history to make a diagnosis was stressed. Once a diagnosis is established, however,
the subsequent management of the patient is very much influenced by their past medical
history, their social circumstances and, more importantly, the drugs that they are
currently taking.

- Ten to thirty per cent of admissions to hospital are due to iatrogenic drug-related
  problems [40, 41]. Computer software can alert clinicians to the potential for drug
  interactions and should be consulted when prescribing a new medication.
- Other medical problems will have a major impact on subsequent management and
  may limit the therapeutic options as a choice of therapy could be contraindicated
  in that condition.
- Similarly, an elderly patient who has the support of spouse and family can be man-
  aged at home as opposed to the patient who has no support and who develops an
  illness that would prevent them from living independently.

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WEBSITES
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UC Berkeley Library Internet Searching Tutorial: http://lib.berkeley.edu/TeachingLib/Guides/Internet/FindInfo.html