Mac Keith Press Courses

Principles of Child Neurology in Infancy

Clinical Practice of Child Neurology in Infancy

Mac Keith Press 2021

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Contents

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Course Overview & Process

Mac Keith Press and the European Paediatric Neurology Society (EPNS) have collaborated on two online learning courses that will enable you to achieve best clinical practice for all neurological problems in infancy.

EUROPEAN PAEDIATRIC NEUROLOGY SOCIETY

https://www.epns.info

Course 1, *Principles of Child Neurology in Infancy*, looks at the basic guiding principles behind achieving best practice and course 2, *Clinical Practice of Child Neurology in Infancy*, puts these principles into their practical context.

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Course Content

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Quick-Reference Guide

We believe in clinical excellence and accessible science.
That's why we've prepared this quick reference guide and useful resources list to help guide your future studies, even if you don't want to pursue a full Mac Keith Press course.
Below, you'll also find a free copy of chapter 1 *Principles and Practice of Child Neurology in Infancy 2nd edition*, edited by Colin Kennedy, a key text for the course.

Key Messages

·Providing excellent care to children with suspected neurological problems has many facets. The 'Principles' course will increase your knowledge and understanding of how to examine an infant with confidence, how to interpret MRI images and EEGs, how to use genetic testing to answer a clinical question, how to know the difference between a p value and a confidence interval, how to include care-givers in the formulation of a management plan, and much more.
·The 'Clinical practice' course outlines a practical approach to common clinical scenarios including neonatal and post-neonatal encephalopathies and metabolic disorders, Traumatic brain injury, stroke, epileptic and non-epileptic seizures, neurological illness with fever, macro- and micro-cephaly, disorders of sleep, neurological abnormalities at birth and early developmental impairment, the floppy infant, cerebral palsies, other motor disorders and progressive loss of skills.
·Key skills include careful history taking and examination and good communication both within the inter-disciplinary team and also with care-givers.
·The 'first do no harm' principle must inform treatment interventions.

Common Errors

·Being too focused on the medical aspects of treatment and forgetting the psychological and emotional well-being of the child, parents, and sometimes the health-professionals.
·Imprecise or incorrect use of terms for which precise definitions exist.
·Generalisation of uncommon conditions to common clinical situations.

When to Worry

·In the child: short term - deterioration in conscious level; long term - loss of skills.
·In your practice: when published experience contrasts with your personal experience
·In your health system: resistance to the evaluation of benefits and harms of current. treatments; screening for conditions for which the benefit of early intervention is unclear.

Quick-Reference Guide Cont.

USEFUL RESOURCES - INCLUDED FREE WITH COURSE

- Arzimanoglou A, editor. (2018) Aicardi's Diseases of the Nervous System in Childhood, 4th edition. London: Mac Keith Press. **Chapter 6**.
- Cioni G, Mercuri E (2007) Neurological Assessment in the First Two years of Life. Clinics in Developmental Medicine Series. London: Mac Keith Press. **Complete text**.
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- Einspieler C, Prechtl FR, Bos A, Ferrari F, Cioni G (2005) Prechtl's Method on the Qualitative Assessment of General Movements in Preterm, Term and Young infants. Clinics in Developmental Medicine. London: Mac Keith Press. **Chapters 1 & 2**.
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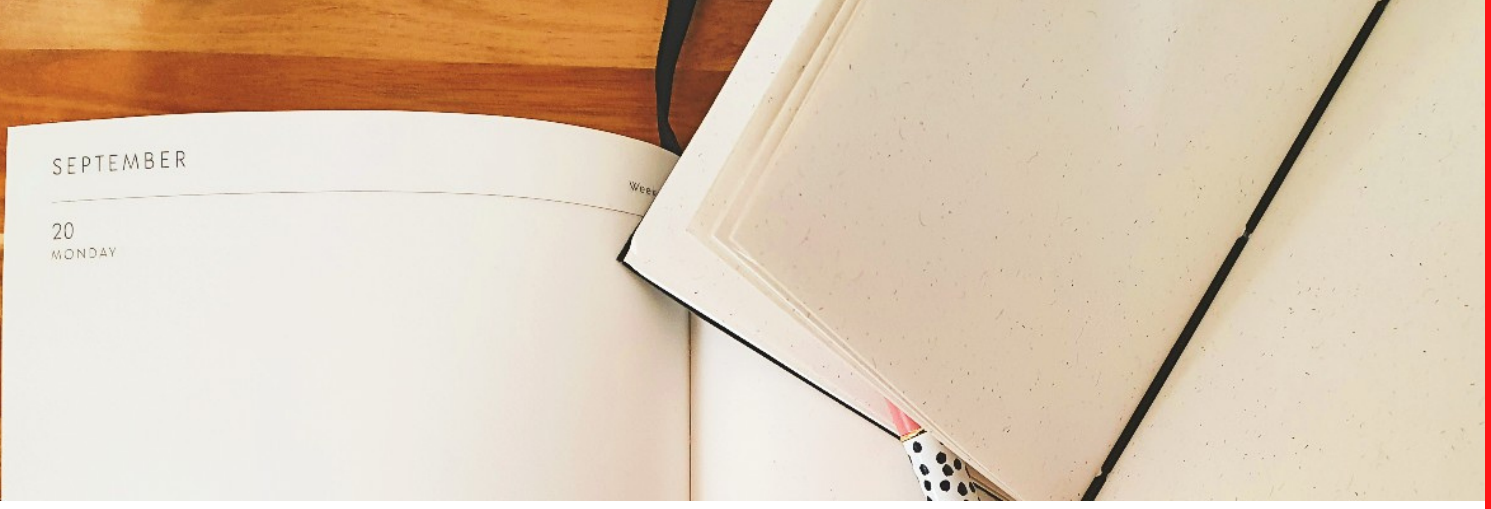
- King MD, Stephenson JBP (2009) A Handbook of Neurological Investigations in Children. London: Mac Keith Press. **Chapters 1.1 and 1.2**.
- Newton RW, Puri S, Marder L, eds (2015) Down Syndrome: Current Perspectives. London: Mac Keith Press. **Complete text**.
- Preece PM, Riley EP (2011) Alcohol, Drugs and Medication in Pregnancy: the Outcomes for the Child. Clinics in Developmental Medicine series. London: Mac Keith Press. **Complete text**.
- Stephenson JBP (1990) Fits and Faints. London: Mac Keith Press. **Complete text**.
- Sullivan PB, ed (2009) Feeding and Nutrition in Children with Neurodevelopmental Disability. London: Mac Keith Press. **Complete text**.



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Principles and Practice of Child Neurology in Infancy 2nd Edition

Principles and Practice of Child Neurology in Infancy 2nd Edition

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Foreword

This is the second edition of *Principles and Practice of Child Neurology in Infancy*, edited by Professor Colin Kennedy, whose first edition appeared in 2012. The primary objective of this book is to provide a symptom-based guide to the diagnosis and management of neurological disorders in infancy. Each chapter is preceded by a concise summary, updated according to the present best practice and subdivided into 'key messages', 'common errors', and 'when to worry'. In this way the reader has a bird's eye view of the content, points of interest, the most important clinical signs and symptoms, and the pitfalls to avoid.

As can be expected in a second edition, the chapters and references covering the different neurological symptoms have been thoroughly updated to reflect the progress that has been made in recent years in the fields of genomic analysis, neuroimaging pattern recognition, and neurophysiology as well as a concise description of the more recently discovered diseases. Contributions from the book come from widely recognized experts in the field of paediatric and developmental neurology. Many members of the original international working group are again authors in the second edition. However, some updates and a few new topics have been added and covered by the best experts in the field.

The first half of the book now contains 13 chapters and is dedicated to subjects not to be found in ordinary paediatric neurology textbooks such as the principles of working with families, clinical assessments, use of investigations, treatments, and evidence-based medicine. In addition, attention is paid to typical development and its variants. Chapter 4 on 'Promoting child development' has been thoroughly expanded: the topic has been put in a broader context and mentions not only the important role of families in infant development and other positive factors, but now also the common negative social factors for development such as psychoactive drugs, poverty, and stress, especially important when present together. An important new section on neurological aspects of vaccination has been added alongside the updated sections on prevention and screening. Two important new chapters are added that were sorely missed in the first edition:

first, a very helpful and practical chapter on 'Neurological examination beyond the neonatal period' and, second, a chapter, also much needed, on 'Genetic testing'. This last chapter offers a concise introduction to inheritance patterns, common types of genetic mutations, and an overview of currently available types of genetic testing, what to expect from them and their strengths and limitations. Many examples of everyday clinical situations where genetic testing can be of use are discussed.

The second half of the book addresses the major disorders that may present in neonates and infants in a symptom-based manner. Topics include neonatal encephalopathy, neonatal seizures, acute febrile and nonfebrile encephalopathies (now including malaria), epileptic and nonepileptic paroxysmal disorders, macrocephaly (now including brain tumours), cerebral palsy and movement disorders other than cerebral palsy, as well as progressive loss of skills. The chapters on microcephaly (with extended consideration of genetic aspects and inclusion of Zika virus) and the floppy infant have been largely revised for this edition. Also new for this edition are authoritative but concise chapters on stroke and on inherited neurometabolic diseases – with valuable information on the clinical manifestations, the approach in the diagnosis and therapy of both well-known and newly identified disorders. Another new and very useful chapter has been added covering normal and abnormal patterns of infant sleep and behaviour. This is an important topic because sleep disturbances are common and may be quite disruptive to family life. The Appendix provides WHO growth charts for skull circumference, length, and weight and a colour-coded chart for use in conjunction with Gross Motor Function Classification System.

The authors have filled the chapters not only with their knowledge but also with the wisdom of many years of clinical practice. They emphasize the importance of clinical assessment, adequate history taking, and skillful neurological examination as key diagnostic tools. Mastering these delicate skills is a prerequisite to successfully solving a neurological problem while involving parents and caretakers, investigating selectively, and avoiding over-treatment. This approach limits the burden on and increases the benefits to the child and parents. The book thus provides the reader with important tools to improve both technical and ethical aspects of acute and long-term care of the child.

The first edition of this book was an immediate and great success. This updated and extended second edition was, like the first edition, written to provide strong guidance for paediatric neurologists, whether in practice or still in training, in both resource-poor and resource-rich countries. The book will also provide a quick general update for more senior paediatric neurologists who mainly work in a highly specialized field – such as for example epileptology, sleep disorders, or neuromuscular diseases – and may serve as a reference guide for those who are teaching paediatric neurology in medical schools. Because of its strong clinical approach, the book is also very accessible and helpful for all physicians, either in training or practice, who need guidance in the care

of infants with neurological problems, be they paediatricians, neurologists, developmental specialists, or rehabilitation specialists.

The content of the first edition of the book was the basis of a series of very successful and much appreciated teaching courses in Eastern Europe (Tbilisi 2015, Astana 2012, 2017, Tashkent 2019) and of the European Paediatric Neurology Society (EPNS) teaching courses in western Europe. Thus, the chapter authors, and especially the book's editor Professor Colin Kennedy, have contributed to the training of a generation of young paediatric neurologists from all countries of Europe and beyond, not only by providing practical knowledge on neurological diseases in neonates and infants but also by stimulating a critical attitude to the diagnostic process based on clinical epidemiological data and evidence-based medicine.

The second edition will be no less influential given the fact that two very informative online courses based on the chapters of this book have been created by Mac Keith Press.

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Preface

The favourable response to the first edition of *Principles and Practice of Child Neurology in Infancy* created the challenge of updating the book to remain portable but also to include methods and diagnoses of increasing importance in 2020. Additions to the first edition include four chapters (*Neurological examination beyond the neonatal period, Genetic testing, Inherited metabolic encephalopathies of infancy,* and *Infant sleep and behaviour*) and five completely new sections within chapters (*Vaccination, Immune-mediated encephalitis, Malaria, Zika virus infection,* and *Intracranial space-occupying lesions*). All chapters have been updated and there have been major redrafts and a change or addition to the authorship of three other chapters (*Promoting child development, Microcephaly, including congenital infections,* and *The floppy infant*). The number of hyperlinks to online resources has been substantially increased.

Taken together, these improvements provide a major revision of the first edition. They will also provide the basis for two online courses related to the book which have been developed in partnership with Mac Keith Press and the European Paediatric Neurology Society (EPNS).

I am grateful to all the authors for updating chapters and providing new ones, and to Sally Wilkinson at Mac Keith Press for her inexhaustible patience.

Colin Kennedy, Southampton, UK
December 2019

CHAPTER 1

Terms, definitions, and concepts

Colin Kennedy

Key messages

- The precautionary principle, 'first do no harm', was established 2500 years ago. Justification for the use of a treatment remains the responsibility of the treating physician.
- For many centuries 'ecologies of care' rather than definition of illness was the predominant paradigm of medical practice and this continues to be relevant to the young because the relationships between a child, the family, and the wider environment remain important determinants of health outcomes, especially in infancy.
- Discussion of the management of disease is greatly facilitated by internationally agreed definitions of disease and these are available as the International Classification of Diseases, 11th Revision (www.who.int/classifications/icd/en/).
- Evidence-based medicine provides an objective method for the systematic evaluation of the evidence of the benefit and harm of medical interventions.

Common errors

- Use of imprecise terms for which international agreement is lacking, for example, the syndrome of raised intracranial pressure, hydrocephalus syndrome, myotonic syndrome, hyperexcitability syndrome.
- Imprecise or incorrect use of terms for which precise definitions exist, for example, perinatal encephalopathy, epilepsy.
- Generalization of uncommon conditions to common clinical situations, for example, attributing trembling of the chin, feeding problems, excessive crying, or febrile seizures to neurological disorders.

When to worry

- Separation of infants from their families (one should facilitate bonding between an infant and his/her main carer).
- Use of poorly evaluated, potentially harmful interventions in many infants to treat rare neurological problems (one should use common sense and look at international recommendations of good practice).
- Resistance to evaluation of the benefits and harms of current treatments (one should consider both the potential benefits and potential risks of all interventions).

THE BASIS OF MEDICAL PRACTICE

This book offers knowledge, only some of which is truly evidence-based, and a framework for incorporating evidence into the clinical care of infants in whom there is concern about neurological function or developmental progress.

Historically, the starting point was myth and wise myths will continue to have their place in medical practice. According to the ancient Greeks, Apollo was the god of healing and Asclepius, his son, was rescued by Apollo from the womb of his dying human mother, Coronis. Asclepius' daughters were Hygeia, the goddess of health, and Panacea, the goddess of cures. Asclepius also had sons and Hippocrates, according to myth, was a descendant of one of those sons. Hippocrates was a practising physician nearly 2500 years ago and author of the Hippocratic Oath (www.pbs.org/wgbh/nova/body/hippocratic-oath-today.html), the most famous text in Western medicine. The most widely quoted section of that oath states: 'I will use treatments for the benefit of the ill in accordance with my ability and my judgement, but from what is to their harm and injustice, I will keep them'. In addition to this statement of the precautionary principle (i.e. 'first do no harm'), other sections of the oath bind the practitioner to resist all temptations that their privileged position as physicians offer, to acknowledge the limits of their competence and refer to specialist practitioners when necessary, to leave surgery to the surgeons, to respect patient confidentiality, to treat one's professional teachers as one's parents, and to pass on the art of medicine to the next generation. Thus, many of the issues of key importance to clinical practitioners and the health systems within which they work are identified within the oath.

THE PRECAUTIONARY PRINCIPLE IN THE CONTEXT OF NEUROLOGICAL PROBLEMS IN INFANCY

The precautionary principle is especially relevant in the assessment and management of neurological and neurodevelopmental problems in infancy, when medical intervention may unwittingly hinder the role of the parents in the child's development,

whether typical or impaired; hospitalization or other institutionalization should be avoided whenever possible (Chapters 2 and 4). Any system of medical activity that involves surveillance of typically developing children should be based on explicit principles of screening (Chapter 7), including evidence that the benefit of early intervention, whether special investigation (Chapters 8, 9, 10 and 11) or treatment (Chapters 12 and 13), outweighs the potential for harm. The range of 'normal', better termed 'typical', neurological development in infancy is broad. In cases of doubt, continuing clinical surveillance and support for normal parenting is needed. This has less potential for harm than either enthusiastic separation into medical categories in the border zones of normality or the use of treatments for which benefit is not established or is outweighed by risk of harm. Any system of practice that categorizes more than a few per cent of infants as neurologically atypical must itself be suspect. Such a system is incompatible with the epidemiology of neurological disorders in childhood and will, by definition, expose many typical children to the risk of being wrongly categorized as impaired. This is a particular example of the need for any form of screening to fulfil several criteria additional to those that apply to the treatment of the illness (Chapter 7).

Neurological and developmental assessment and neurological examination of an infant (Chapters 5 and 6) is a practical skill of central importance that requires hands-on experience as well as knowledge. The importance of the physiological state of the infant (hungry or fed, wakeful or drowsy, contented or distressed), the need to rely on best performance (as opposed to poorer performance on a single occasion), and the extent to which clinical features are consistent over time are more important to bear in mind at this age than any other factor. Almost any finding with respect to deep tendon reflexes, other than complete absence of them, for example, is within the typical (i.e. normal) range in some physiological states or at some age within the first year.

ECOLOGIES OF CARE AND CATEGORIES OF ILLNESS

On the foundations expressed by Hippocrates, the art of clinical practice in Western countries evolved in the pre-scientific era using a system of knowledge based on the eminence and experience of senior practitioners. For many centuries before the more modern description of categories of illness as the basis of medical practice, ecologies of care for maintenance of health and for the treatment of illness acknowledged the importance of the relationship between the patient and the wider environment and provided the predominant paradigm of care. In the case of the child-patient, family relationships are of primary importance and are fundamental to the Head Start (USA) and Sure Start (UK) programmes for the improvement of the health and well-being of young children (Blair and DeBell, 2011). These issues are discussed in Chapters 2, 4, and 7.

THE INTERNATIONAL CLASSIFICATION OF DISEASES

The World Health Organization (WHO) was founded by international treaty in 1948 as a specialized agency of the United Nations with unique authority to establish global health standards and to secure international agreement on defining disease. The 193 member states of the WHO have agreed to use the International Classification of Diseases (ICD), the most recent version of which, the 11th Revision (ICD-11), was released on 18 June 2018 (www.who.int/classifications/icd/en/) and endorsed at the World Health Assembly in May 2019 for use from January 2022. Classification of mental disorders, which include neurological disorders, is complex and controversial, both because underlying pathophysiology cannot be observed directly and because many symptoms are contiguous with normal phenomena. Nowhere are these issues more relevant than in the neurology and neurodevelopment of infants. The ICD classification is predominantly driven by the clinical utility and public health outcomes of the disease entities and, despite these controversies, is, therefore, an appropriate framework for clinical practice (Reed et al. 2011).

The ICD provides the basis of the discussion for classifying the phenomena observed in the clinical contexts that are discussed in Chapters 14 to 29. A number of entities (e.g. brain tumours of infancy) are included in the differential diagnosis but not covered in detail in this volume for lack of space. Other entities do not appear because they are based on classifications of disease other than the ICD. Such diagnostic classifications, including some listed in the paragraphs below, may claim to identify disease entities requiring active management in a substantial percentage of neonates or infants. In some cases, the criteria for making such diagnoses are vague, their relationships with disorders of later childhood unknown, and the rationale for intervention is obscure (Mustafayev et al. 2020).

International clinical guidelines depend upon this shared nosology and classification of illnesses and disease: the foundation for rational management requires knowledge of what treatments are of benefit and what are harmful, which in turn requires specific disease definitions that are shared by all those involved in providing care.

RELEVANT TERMS AND DEFINITIONS

The brief discussion of terms below is intended to help the reader to navigate through later chapters of the book but is not intended to be exhaustive.

Encephalopathy is defined as 'a disease in which the functioning of the brain is affected by some agent or condition' (*New Oxford Dictionary*). Because this definition is so inclusive, it is of little practical value in clinical medicine. While 'acute' means 'of short duration' or 'experienced to an intense or severe degree' (*New Oxford Dictionary*), the medical definition of *acute encephalopathy* includes alteration in conscious level as an

essential criterion (Chapter 17) and to that extent, is a more clinically useful term for the formation of a plan of investigation and management.

The term *perinatal encephalopathy* does not indicate whether the observed effect on the functioning of the brain is of short duration or whether it involves an alteration of conscious level. Furthermore the term 'perinatal' includes the period before birth when the infant's level of consciousness is usually not known to the clinician. However, the general term 'encephalopathy' is no more useful in the perinatal period than it is generally, especially since there is often disagreement as to whether or not commonly observed neonatal or post-neonatal behaviour (e.g. tremor of the chin) indicates abnormal brain functioning. This carries a significant potential for harm in exposing many infants, the vast majority of whom will have no known subsequent medical disorder, to the risks associated with medical diagnoses of doubtful validity (Mustafayev et al. 2020). In practice therefore, the definition of neonatal (not perinatal) encephalopathy (NE) includes alteration of the level of consciousness (Chapter 14). It is, in effect, the special case of 'acute encephalopathy' in a newborn baby. Other more inclusive uses of the term *perinatal encephalopathy* are to be avoided and will not be further discussed here.

Hydrocephalus is used to mean an excess of cerebrospinal fluid within the head but excluding those situations where that condition has arisen purely from atrophy or failure of the brain substance to develop (sometimes called hydrocephalus *ex vacuo*). The presence of hydrocephalus cannot be confirmed or excluded based on the dimensions of the third cerebral ventricle alone (see Chapter 23 for further discussion).

Hydrocephalus and *raised intracranial pressure* frequently co-exist and specific clinical signs, often including disturbed consciousness, can be combined with cranial imaging to provide evidence for the presence of both entities (see especially Chapters 17 and 23). By contrast, 'the syndrome of intracranial hypertension' and 'hydrocephalus-intracranial-hypertension syndrome' are not internationally recognized as diagnostic entities (Mustafayev et al. 2020) and should not be confused with the rare condition of older children and adults known as pseudotumor cerebri syndrome (and also as idiopathic intracranial hypertension) (Matthews et al. 2017).

A *seizure* may be epileptic (Chapter 20) or nonepileptic (Chapter 21). Epilepsy is defined as recurrent unprovoked epileptic seizures. Febrile seizures are provoked by a rising fever and are not conventionally regarded as falling within the above definition of an epilepsy. These chapters also provide further discussion of these definitions and of the syndromes that constitute disease entities within them.

Myotonia is defined as the inability of muscle fibres to relax after muscle contraction and can be demonstrated by myotonic discharges on electromyography. This is a very rare phenomenon in infancy and even in an infant with congenital myotonic dystrophy (Chapter 24), myotonia is usually only demonstrable in an affected parent.

The term *myotonic syndrome*, in which abnormality of muscle tone is the dominant feature, is not an internationally recognized diagnostic entity in infants.

Hyperexcitability syndrome is not a generally accepted diagnostic disease entity term in infancy and internationally accepted criteria for its definition are lacking. Problems with one or more of sleeping, feeding, or excessive crying in infancy are reported in up to 30% of all infants and are usually transient. Most cases may, therefore, be regarded as falling within the spectrum of typical development and need not be conceptualized as indicating an underlying neurological disorder. Associations do, however, exist between multiple problems with these functions (often referred to as early regulatory problems) and long-term behavioural outcomes in childhood, including attention-deficit/hyperactivity disorder. These problems are attributable partly to biological predisposition in the infant, partly to parenting behaviours, and partly to interactions between the two. Interventions that alter parenting behaviours may help (see Chapter 25). Support to parents to help prevent or reduce early regulatory problems is typically given in the context of general paediatric nursing or medical assessment of the infant and advice to families rather than as treatment of a disease entity.

EVIDENCE-BASED MEDICINE

The accumulated wisdom of previous generations of medical practitioners has, since the 19th century, been progressively supplanted by the concept of 'evidence', of greater or lesser quality, to support the use of treatments. Myth has been progressively replaced by evidence, although the process has sometimes been hindered by political interference (McKee, 2007). Hopefully the value of certain myths, starting with Hygeia and Panacea, will continue to be recognized. Evidence-based medicine has only emerged within the second half of the 20th century and has been an increasing influence on medical practice in the 21st century. It is, as described in Chapter 3, the systematic construction of a body of knowledge about interventions for medical illnesses with explicit, objective criteria for rating the quality of the evidence upon which that knowledge is based. The great strength of evidence-based medicine lies in its capacity for constant improvement as new information comes to light and without *ad personam* arguments about the authority of the individuals advocating any particular treatment, which had dominated previous medical thought since Hippocrates, sometimes referred to as 'eminence-based medicine'.

Unfortunately, the quality of much of the evidence upon which we must currently rely for guidance in the treatment of neurological disorders in infancy is poor. Furthermore, the traditional measures of quality of evidence are sometimes difficult to apply when studying rehabilitation, including physiotherapeutic interventions (Rosenbaum, 2010; Autti-Ramo, 2011). The methodology of evidence-based medicine can also help us to

identify those situations where evidence is lacking and serve to remind us that justification is always required for medical intervention, especially in an infant, and the responsibility for this rests with the physician.

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