

Chronic Childhood Disorders

A Handbook of Medical Care
in the Community

Edited by
Gwilym Hosking
and
Ruth Powell

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With a Foreword by
F. S. W. Brimblecomb

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PREFACE

The challenge we face together in the 1980s is to improve the community aspect of the medical care of children with special needs. They have disorders which because of their chronicity, severity or complexity have a major impact upon their lives and those of their families. For most there will have been a need for the involvement of specialist hospital services. This handbook is primarily concerned with many of the disorders which are rare when considered alongside childhood infections, accidents and emotional problems – sometimes even in the experience of the hospital specialists who have been involved.

With very few exceptions children with chronic disorders live with their families in the community and the doctors designated to advise and manage the day-to-day problems are based within the community. These are the general practitioners and doctors working in the child health service who are likely to have had limited experience of the less common disorders of childhood. The opportunities for the community-based doctor to discuss a particular child with the hospital specialist are usually rare. Thus, we may have a doctor occupying a crucial position in relation to the child and his family who has little or no knowledge of the disorder in question. This doctor also advises those who are responsible for the child's education, social welfare and recreation activities. We question how useful is the doctor whose knowledge of the condition is no more or perhaps even less than that of the parent?

The details of hospital-based treatments do not figure highly in this book, but technical information is provided where it is felt that it will be of value. Hospital reports and summaries may be just that little bit more intelligible!

In the clinical section of this book we have provided brief details of many disorders, the complications that arise and aspects of their management. We have included practice points which we feel should emphasize important aspects worthy of particular notice.

We hope that this handbook fills the definite gap that exists in the provision of realistic information for those doctors working in the community.

To encourage a total approach towards the management of these childhood disorders we have included some information regarding the identification of potentially treatable disorders, the current attempts being made to provide education that meets the needs of children with chronic conditions, and, just as important, the modern medical practice which aims to prevent disabling or handicapping conditions.

It is inevitable that this book with its predominantly British contributors should reflect British practice and the services established

to meet needs in Britain. It is hoped that it will also be of value to those working outside Britain in other developed and developing countries, as with improved facilities for travel we come closer together and learn more from each other.

G.H. and R.P.
Sheffield, 1985

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We are also grateful to Professor Freddie Brimblecombe for writing the Foreword, and would like to thank him for his kind words and sentiments.

G.H.
R.P.

FOREWORD

Frederic S. W. Brimblecombe CBE, MD, FRCP
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The need for information for family doctors and clinical medical officers about chronic childhood disorders is increasing. As the tide of acute illness in childhood recedes in the U.K., the emphasis on optimal care of children with chronic illness demands an increasing priority. There are two particular reasons for this.

First there is the trend towards an increased integration of hospital and community care for these children and their families. For each child there is a need for a more clearly defined contract between hospital specialists and doctors working in the community so that both are more clear about their respective roles in clinical management than they have been in the past. If this is not done, there is a danger that duplication and possibly conflicting advice will be given; alternatively there will be omissions of care if either side assumes that particular functions are being carried out by the other.

Secondly, children with chronic illness are, as a result of improved medical knowledge, surviving in increasing numbers into adulthood. The transition from childhood to adulthood for these young people and their families is an area of medical care that has received insufficient attention in the past, as has the need to ensure that the medical contribution to the quality of life which they lead is coordinated with that provided by other agencies.

For both these reasons, hospital specialists, clinical medical officers and family doctors need to explore improved ways of ensuring that the needs of these children receive the best possible medical care. The responsibility of doctors is primarily for the health of these youngsters, but no 'good doctor' can neglect the vital question of the quality of life that they experience despite the presence of a chronic or handicapping illness.

There is a major need for increased attention to this aspect of medical practice in all its aspects. For these reasons this book edited by Dr Gwilym Hosking and Dr Ruth Powell, like other contributions to this aspect of medical knowledge, is to be highly commended. It is a subject that is going to require increasing attention in the remaining years of this century.

CONTENTS

Foreword	xv
Section I Introduction	
1 The children who have 'special needs'. The size of the problem <i>Shelagh Tyrrell</i>	3
2 Identifying children who have special needs <i>Ruth Powell</i>	14
Section II Clinical	
3 Seizure disorders <i>Gwilym Hosking</i>	25
4 Cerebral palsy <i>Gwilym Hosking</i>	42
5 Neuromuscular disorders <i>Gwilym Hosking</i>	54
6 Neurodegenerative disorders <i>Gwilym Hosking</i>	75
7 Spina bifida and hydrocephalus <i>Gwilym Hosking</i>	95
8 Mental retardation <i>Gwilym Hosking</i>	108
9 Visual handicap: The deaf child: Speech and language disorders: Disorders of learning <i>Lewis Rosenbloom</i>	122
10 Respiratory disorders <i>Brendon Watson</i>	136
11 Congenital heart disease <i>Barry Keeion</i>	146
12 Diabetes and inborn metabolic errors <i>Stefan Herber and David Milner</i>	163
13 Malignant diseases and chronic blood disorders <i>John Lilleyman</i>	177
14 Gastroenterological problems in childhood <i>Stuart Tanner</i>	188
15 Liver problems in childhood <i>Stuart Tanner</i>	201
16 Renal diseases and hypertension <i>Leonard Taitz</i>	208
17 Musculoskeletal disorders in childhood <i>Ian Griffiths</i>	221
18 Tropical disorders and problems of children from tropical countries <i>Michael Chan</i>	238
19 Personality and behavioural disorders: Autism <i>Robert Bugler</i>	253
20 Child abuse and neglect (Non-accidental injury) <i>Mary Lindsay</i>	266

Section III Services in Britain to Meet the Needs of the Child and the Family	
21 Family and child services for those with special needs. Social support: Recreation: Education <i>Ruth Powell</i>	279
Section IV The Next Generation. The Prevention of Handicapping Conditions	
22 Antenatal screening and care <i>Sheila Duncan</i>	299
23 Perinatal and neonatal problems <i>Richard Pearse</i>	316
Envoi:	
24 'Communicating' <i>Gwilym Hosking and Ruth Powell</i>	331
Appendix	333
Index	339

The Children who have 'Special Needs'

The Size of the Problem

SHELAGH TYRRELL

Few people would disagree that children in any society form a powerless group lacking the capacity to speak for itself. It was not until the twentieth century that they were seen in the West as having particular needs which must be met if they were to develop their personalities and intellect to the full. At the same time within medicine, paediatrics evolved as a specialty concerned with the medical care of children. More recently paediatrics has become concerned with the prevention of disease, including that affecting adults when the roots lie in childhood.

The basic needs of all children are for love (and belonging), food, shelter, protection against danger and disease and an opportunity for continuous learning. The needs of the child for food and shelter cannot be separated from the need of the family for money. The State recognizes this in terms of allowances of various kinds which may or may not be adequate. But in some cities in Britain, where families speak little or halting English, the mysteries of acquiring grants and benefits may be considerable and there is a need for an informed friend to turn to for advice; this may be the health visitor (public health nurse).

For a large number of families in Britain who are undergoing a new and relative form of economic hardship, full-time or part-time day care provisions for children are a high priority as mothers want and need to go to work. Although the State may be concerned about the quality of such provision families seldom wait until they find something which will satisfy the criteria of the providers (and their research colleagues).

Some families may require more or a different kind of help, and for this one of the voluntary bodies, who do much valuable work, may be appropriate. A health visitor who introduces families to such a resource



Equality through sport: on the left a girl with Down's syndrome; on the right a girl without legs and hands.

Copyright of the *Sunday Times*. Photograph by David Lavender.

can then provide the required liaison so that the care received by the family is cooperative and not competitive.

THE CHILD WITH SPECIAL NEEDS. WHO IS HE?

From the medical viewpoint the child with special needs is likely to be one who suffers from some defect, disability or handicap. In effect it is only a combination of these that will ensure that he has 'special needs' as should become obvious from the definition:

Defect - 'absence of something necessary for completeness'

Disability - 'want of ability'

Handicap - 'a child is said to be handicapped when a disability puts him at a disadvantage in his particular environmental circumstances'

A girl may have suffered an accident and lost one foot (defect). She is in consequence unable to run as well as her friends (disability). But none of them is particularly keen on competitive sports so she suffers little or no handicap as a result of her injury. In contrast, another child has a damaged cochlea (defect) because his mother had rubella early in her pregnancy. He is profoundly deaf (disability - unable to hear). His

handicap is that he cannot communicate well with his peers, so they stop playing with him and his hearing loss is so severe that he has to be educated with other, similarly affected children. Such a child will have special needs for life because his deafness cannot be cured.

A child may be born with a serious heart condition which handicaps him considerably, yet at an appropriate time this is amenable to surgery. Such a child will then only have special needs for a limited period. Other children may have handicaps which are of an episodic nature and conductive deafness is perhaps the most common. Attention here to his special needs is no less important because the problem is temporary. Children who cannot hear for a time may be at a significant disadvantage in school. As well as this, if the deafness occurs when they are very young, the experience may dishearten them and they may switch off their interest in 'listening'.

Some social and emotional factors within the home, especially depression, may compound a handicap and inhibit mental development in several ways. Conversely, emotional factors which are positive towards the child may mitigate the effects of his handicap. Many parents may learn to supply not only the needs but the special needs of their children. There are mothers who have learned and use successfully the skills of teachers and therapists. One parent with a handicapped child wrote, 'Professional help cannot be wholly effective, if at all so, unless it builds upon the parents' understanding of their children's needs and upon the parents' capacity to be involved.'

The Warnock Committee, when looking at the child with special needs from an educational point of view, stated:

To describe someone as handicapped conveys nothing of the type of educational help and hence of provision that is required. We wish to see a more positive approach and we have adopted the concept of special educational needs, seen not in terms of a particular disability, which a child may be judged to have, but in relation to everything about him: his abilities as well as his disabilities - indeed all the factors which have a bearing on his educational progress.

THE SIZE OF THE PROBLEM

A child's development may be so unpredictable and early estimates of handicap consequently imprecise that it is only at school age that the size of the problem can be effectively considered. The Warnock Committee, who were concerned with appropriate provisions for children with special needs, found that the rate of 'ascertainment' of children for special school varied between 120/10000 pupils in rural areas to over 300/10000 in larger cities. There was no evidence to suggest that such children were overwhelmingly concentrated in the towns. It was concluded that the number of children ascertained was dependent upon

the number of places available to receive them in school. It was ultimately felt that one child in five in the population is likely at some time in a school career to require special educational provision.

From the medical standpoint the pattern of disability varies at differing ages in childhood. Children with a severe degree of spina bifida or major chromosomal defects may not survive infancy. It is generally thought that 0.8 – 1 per cent of children may live with a severe handicap whether this is physical, mental or, more often, a combination of the two. Three per cent of children are likely to suffer from a moderate but significant disability or handicap, the commonest being learning difficulties that affect educational performance.

Cerebral palsy, requiring comprehensive care, is a motor disability resulting from damage somehow sustained by the immature brain. As it affects the person for life it has significant resource implications. The incidence of cerebral palsy is approximately 2.4/1000 live births in Britain.

The incidence of major spina bifida lesions would appear to be somewhere between 1.6 and 4 per thousand live births in Britain but with a considerable variation from one geographical area to another.

22/100000 children are registered as being 'blind'. Most have some vision but this is insufficient for learning purposes and they have to be taught by methods dependent upon touch.

More common than children with severe visual handicaps are children with moderate to severe hearing loss, requiring the use of aids. These probably represent about 2/1000 children but as many as 4 per cent at any one time may suffer from a milder but still significant temporary conductive hearing loss.

Of three-year-old children in urban areas 15 – 20 per cent have significant language delay. Less than 1/1000 are likely to suffer from a severe disorder of language development.

Ten per cent of ten-year-old children in the Isle of Wight study had significant psychiatric disturbances of one form or another. In the same study it was realized that 10 per cent of children also had a reading problem which was not necessarily related to their overall level of intelligence.

The figures given can be taken as a very approximate guide to the provisions required and what follows is a closer look at the needs of these children and their families.

THE INDIVIDUAL NEEDS

The Children

The child with a severe disability still has the same basic needs as other children. It is his opportunity for continuous learning that is most likely

to be affected. Any child with a potential problem requires early diagnosis and assessment of a possible disability followed where appropriate by immediate medical and remedial treatment and management.

The *Education Act* of 1981 in Britain made the early identification of special needs a key issue. But as the child's development depends both upon his genetic make up and his environment few doctors or psychologists will make precise predictions early in the child's life. Besides which, parents need time and an understanding approach before they can accept that their child has any such special needs.

The need for early learning is met when possible by peripatetic teachers who can help the parents to understand aspects of their child's development. They can combine with therapists to help create the optimum environment for the child. The blind baby may need a teacher to help his mother to develop his touch and listening skills, an occupational therapist to help arrange the home so that it is safe, interesting and predictable for a child without sight, a physiotherapist to help with encouraging movement and independence and a speech therapist to help the development of language. In some countries with widely separated communities there is a single visiting therapist combining all these roles.

The place of education for disabled or handicapped children is an important issue, although the concept of special needs which can be met in differing situations has helped to reconcile some of the opposing views. Before full integration is possible, the public has to be educated in the simplest truths of disabilities and handicaps. For instance, handicap is not 'catching', standards in schools which accept pupils with special needs will not automatically be lowered, the weaker brethren among the children will not automatically start 'copying' the athetoid movements of the cerebral palsied child. For the child with a disability, integration into the neighbourhood school with his peers, even if only for part of the day, will help him and everyone else to face up to the disability and its implications. However good a special school may be the child will often emerge at the end of his time at school to lead the lonely life of the adult handicapped person, with none of the sympathy that may be afforded by society to a little child holding a walking frame, or the toddler with Down's syndrome peering attractively from the poster.

Independence is a key goal for children as part of their search for new worlds and new discoveries. A disabled child may not have the will to seek such independence, or it may have been burnt out in the great effort of daily living. Several studies have shown that disabled children may have poor language development which is quite unrelated to their disability or their intelligence. This could seem surprising when immobile children would theoretically have more opportunity to follow intellectual pursuits. There is the suggestion that disabled children are