Paediatric Diagnosis and Treatment

N M Jacoby

Paediatric Diagnosis and Treatment

N.M. Jacoby MD, FRCP

Hon. Paediatrician, Pembury Hospital, Kent
Hon. Physician, Queen Elizabeth Hospital for Children, London

with a contribution on
Diseases of the Cardiovascular System by

Douglas Pickering MB, MRCP, DCH

Consultant Paediatrician to the Oxford Area Health Authority (Teaching);
Consultant Paediatric Cardiologist to the Oxford Area Health Authority;
Clinical Lecturer in Paediatrics to the University of Oxford

PITMAN MEDICAL

First published 1978

Catalogue Number 21 1802 81

Pitman Medical Publishing Co Ltd
P O Box 7, Tunbridge Wells,
Kent, TN1 1XH, England

Associated Companies

UNITED KINGDOM
Pitman Publishing Ltd, London
Focal Press Ltd, London

CANADA
Copp Clark Ltd, Toronto

USA
Fearon Pitman Publishers Inc, California
Focal Press Inc, New York

AUSTRALIA
Pitman Publishing Pty Ltd, Carlton

NEW ZEALAND
Pitman Publishing NZ Ltd, Wellington

British Library Cataloguing in Publication Data
Jacoby, N M
Paediatric diagnosis and treatment.
1. Children—Diseases
I. Title
618.9'2 RJ45

ISBN 0-272-79510-0

Set in 11 on 12 pt. IBM Journal by
Gatehouse Wood Limited, Cowden, Kent
Printed by offset-lithography and bound
in Great Britain at The Pitman Press, Bath

Preface

It is hoped that this book will be of interest and use to senior students and doctors who are not paediatric specialists but have the care of children in their practices. It deals with the diagnosis and treatment of diseases but does not include the special care of neonates nor child assessment, subjects that require textbooks of a kind different from this one.

The main purpose of any textbook is to instruct and to pass on information the author has acquired over years of experience. As a rule, it rarely contains startling new discoveries or, in modern parlance, 'breakthroughs', and in that respect none are claimed for this book. Nevertheless, nearly everything that is written in this one is based on personal experience, and I have felt under no obligation to anyone to accept without question information and advice that, although widely taught, to my mind is wrong or misleading. Readers may therefore expect to find here viewpoints on some subjects that may be regarded as controversial; for instance those on opium and steroids.

I have taught students for many years that the two most certain ways of failing an examination are to seduce the examiner's wife, or tell him that you would use the above-mentioned drugs in the treatment of a child with asthma. Yet, I have discovered, a number of paediatricians do indeed treat patients in that way but decline to teach their students the benefits patients derive from the treatment. That barrier should be broken down.

However, the essential *raison d'etre* for this book lies in something I wrote in *The Lancet* in a 'Personal Book List'. I asked why textbooks had to be so boring, and I was challenged by Mr David Dickens

of Pitman Medical to write one that wasn't. During my teaching career I discovered that what those at the receiving end seemed to appreciate most were the facts, put as succinctly as possible, interspersed with periods of relaxation when matters of history, light entertainment, and strongly controversial views were propounded. This is the formula I have followed here in the hope that instruction and pleasure can be combined.

I have not included in this book any information on diseases of the eye or of the skin, or on psychiatry. They are all specialist subjects which are best dealt with at greater length than could be permitted here and which call for a wealth of illustration; at least the eye and the skin do.

Lastly, I am indebted to Dr Douglas Pickering, a one-time colleague of mine at Pembury Hospital, who has contributed the chapter on cardiovascular diseases, a subject in which I am not sufficiently competent, and thus enabled me to round off the general background of paediatric diagnosis and treatment.

I would express my thanks to generations of students, both undergraduate and postgraduate, who have put up with me and at the same time moulded my teaching, and to so many junior colleagues who served and instructed me. To the influence of them all this book owes much. Finally, my gratitude to Mrs D. Barton, my one-time amanuensis at Pembury Hospital, who volunteered to undertake the typing and administrative work of the book in her spare time.

Edenbridge, 1978 N.M.J.

Contents

	Preface	vii
1	Introduction	1
2	Diseases Due to Infection	8
3	Diseases of the Alimentary Canal	57
4	Diseases of the Respiratory System	119
5	Diseases of the Heart	171
6	Diseases of the Central Nervous System	218
7	Diseases of the Blood	287
8	Diseases of the Renal Tract	316
9	Diseases of Metabolic Origin	347
10	Diseases of Bones and Joints	372
11	Diseases of the Liver	396
12	Diseases of the Endocrine Glands	404
13	Diseases Associated with Chromosome Abnormalities	430
14	Neoplastic Diseases in Children	436
15	Diseases of the Newborn	444
	Index	462

1

Introduction

Health and Disease

There has been a fashion, in fact almost a rule, in modern times that Departments and Professors of Paediatrics shall be called Departments and Professors of Child Health. This gives a good impression and is also an indication of the ultimate goal of all those concerned; but the title is misleading as to the nature of the work concerned. *Health is an exclusion diagnosis,* and only those with an extensive knowledge of disease can decide whether a person is healthy. Insurance companies, who have to back the 'diagnosis' with their money, need to know whether those they propose to insure are healthy or not. They do not refer them to 'physical fitness practitioners', or 'body builders', or 'health food stores', but to medical practitioners, who have an extensive knowledge of disease that might reduce life expectancy. Thus, it happens that the main work of those in departments of child health is concerned with child disease, its treatment and prevention.

Paediatrics and General Medicine

The human infant, if carefully nurtured, has an uncanny knack of developing into an adult human being. It is therefore obvious that paediatrics and general medicine cannot be very different, though 'the thousand natural shocks that flesh is heir to' may differ at different ages. The adult is mature in body and mind, both of which with the passage of time deteriorate and degenerate. The child is

still developing and, as such, is less stable and more labile. It is this lability that sometimes allows a child to run the course of a disease at a frightening pace and it is a matter of getting accustomed to the different tempos, though basically the two disciplines are much the same.

Diagnosis

In paediatrics, as in any other branch of clinical medicine, accurate diagnosis is the foundation on which everything else is built. It is the be-all and end-all; the alpha and omega of the proper practice

For the Paediatrician	For the Racegoer
The Immediate Matter of Concern	
1. As an example, let us say headache in a ten-year-old boy	As an example. The Epsom Derby for 3-year old horses
2. *The Differential Diagnosis* Those diseases and conditions that might cause such a symptom at such an age.	*The List of Runners* Those horses of the correct age and status, thought likely to have a chance of winning.
3. *Family History*	*Breeding*
4. *History and Social Conditions*	*History—the Nature of the Course, and the State of the Going,*
5. *The Probabilities* After considering all the factors concerned and bearing in mind that 'common things commonly occur', one chooses the condition most likely to cause the complaint. This becomes the Diagnosis.	*The Probabilities* After considering all the factors concerned, especially how often the horse has won in previous races, the one considered most likely to win this race is called the Favourite.
6. *The Diagnosis* is what one thinks is wrong with a patient, which is not necessarily the same thing as what *is* wrong with a patient.	*The Favourite* does not always win the race.

of one's profession. The treatment, the prognosis, the expected complications and the prophylaxis can be correct only if the diagnosis is correct. Too often there is a tendency, which must be resisted, to listen to the patient's history and then prescribe treatment purely on this basis. Such a method is paved, not with good intentions but with stones, to the undertaker and the cemetery.

Doctors are apt to think of themselves as allied to the other higher professional callings, especially the Church, and in many ways this is true; but it is as well to know that when engaged in the business of diagnosis, the calling closest to the medical profession is that of the 'racegoer' trying to find the winner of the Derby, or any other race for that matter.

So similar are the two exercises, and such are the lessons to be learned from them by the doctor, that they are set down side by side on the page opposite.

Oliver Cromwell, addressing his officers after the battle of Drogheda, said to them, 'Gentlemen, in the bowels of Christ, I beseech thee rememberest that thou mightst be wrong'. A doctor should always do the same.

The Diagnosis of 'I Just Don't Know'

If it is accepted that diagnosis is the foundation of medicine, it follows that the wrong diagnosis makes a shaky foundation; in view of the imperfection of man this is a possibility that has to be accepted. To err is human. What is not acceptable is making a diagnosis which implies that the aetiology and pathology are known, whereas in fact nothing could be further from the truth. This misleads everyone, most of all the diagnostician, and leads to all sorts of unsatisfactory results. To emphasise the point it will help if some examples are quoted, especially where the pathology is now known, as against what was implied previously.

Prior to 1945 there were diagnoses such as ruminating; posseting and habit vomiting, grouped together as the gastric neuroses and applied to infants who vomited but had no discernable pathology even after death and an autopsy. As the name suggests, it was said with authority that the infants vomited because they enjoyed doing it, and there was even a suggestion of bloodymindedness in some cases. Serious clinicians stated that when these infants died as a result of their neurosis they still had a postmortem smile on their faces. It is now known that this was a lot of rubbish and what

these patients suffered from must have been either one of the metabolic dyscrasias, or hiatus hernia, or oesophageal reflux. It would have been rather better had the ignorance been acknowledged.

Another example, still in current vogue, is the diagnosis of such things as mesenteric adenitis and chronic subacute appendicitis. It is not suggested that these diseases cannot exist; but short of a laparotomy one cannot be sure that the diagnosis has any validity, and the disease, if any, might be something quite different. It would be more satisfactory to confess ignorance in such cases, for to borrow from Dr Johnson, there is nothing so concentrates a doctor's mind as knowing that he doesn't know. This must be one's lot only too frequently and there should be no loss of face in admitting it.

Treatment

Treatment, which is so vital to the patient, is relatively unimportant to the doctor. Anybody can look it up in a book but the effect it will have depends on the accuracy of the diagnosis and the treatability of the disease. When a line of treatment does not have the expected result, it is wise to consider whether the diagnosis may be wrong before assuming the treatment is ineffective.

Where there are differences of opinion about treatment, it can be assumed that none of the alternatives is perfect, because, if it were, everyone would be agreed. It is impossible to convey in writing exactly why, when, and how one person sometimes uses one form of therapy and sometimes another. Nor can it always be explained how different people achieve comparable results using different methods. It all depends on that indefinable factor, clinical acumen and experience.

Finally, it should be borne in mind that, just as there is no such thing as a completely safe operation, any drug worthy of its name usually has disadvantages or dangers, or both.

Balance

In medicine there is no more satisfying experience than diagnosing correctly some rare and obscure disease, especially when others have failed to recognise it. By the nature of things, this does not happen very often because rare diseases are, by definition, rare. The desire to achieve such distinction more frequently than is possible may lead

to a tendency to find the unusual where it does not exist, and everyone has to resist the temptation.

To maintain a correct balance in these matters there is an old saying, part of which is very well known and part hardly ever heard any more: it is 'Common things commonly occur; but if you see an elephant walking down Regent Street, don't say it isn't an elephant because elephants do not usually walk there'.

In other words, the commonplace and mundane conditions are one's usual fill; but one must be prepared for the esoteric if and when it appears.

There is another totally different matter of balance that is becoming more distorted year by year. It is between clinical acumen, on the one hand, and special investigations, on the other. Too often, the attitude is 'What investigations shall we do?' rather than 'Are investigations really necessary, and if so, which ones?' Very frequently the tests are an expensive waste of time and money, none more so than the routine blood picture. Sometimes such tests can be grossly misleading, of which there is no better example than the routine urine specimen sent for culture. Unless it has been taken with special precautions it is almost certain to grow *Escherichia coli*, and a second specimen, taken equally carelessly but after a course of antibiotics or urinary antiseptics, will almost certainly prove to be sterile. This will be taken to indicate that the diagnosis of pyelitis was not only correct, but proven so, whereas the exact opposite is probably the case. In paediatrics, and especially in infants when a collecting bag is used, the specimen may be nothing better than a 'perineal wash' teeming with *E. coli* whose origin has nothing to do with the urinary tract. It must also be remembered that the result of any investigation presented to a clinician is subject to experimental error and, though rare, the much greater divergence is caused by human error. Thus, interpretation of the results of investigations may be a matter of fine balance even when they are not misleading. In their proper place, and given the weight they deserve, they can be decisive but they must be the servant and not the master of medical practice; at present this is becoming progressively *not* the case.

Iatrogenetic Illness

It is a sobering thought that most sick people would, and in fact do, recover without any treatment. In days gone by this was obvious since most therapy was totally ineffective and the medical profession

knew the course of a disease was little affected by whatever they did. Fortunately, the patient put his trust in his doctor and his 'bottle of physic' and no doubt attributed his recovery to their help. The nastier the taste of the medicine, the more effective it was supposed to be and, provided this was the only unpleasantness, little or no harm was done. If matters had stopped there, iatrogenesis might have been stillborn; but, unfortunately, other therapies were introduced that could only do harm, sometimes disastrously. No better example can be quoted than the operation, fashionable at the beginning of the twentieth century, of total colectomy for supposed intestinal toxaemia. No such disease existed. With the primitive anaesthesia of that time, plus the actually damaging pre- and postoperative measures then in use, the mortality rate was appalling, and all for no benefit in the unlikely event of a patient surviving.

In the archives of this sad saga may be counted venesection, breaking the nipple strings, cutting the lingual fraenulum, lancing the gums, purgation, high colonic washouts, and nephropexy, and removal of the teeth, the gallbladder and the appendix for supposed focal sepsis, which never existed. In a different sphere, children were kept in bed, often at complete rest and for periods up to sixteen weeks for rheumatic fever and nephritis and, sometimes, very much longer for other diseases such as tuberculous glands in the neck. None of this, as we now know, did any good at all.

It would be nice to think this grisly compendium was all a matter of the past but, regretfully, it is not so. The worst offender by far is tonsillectomy; still performed by hundreds of thousands a year. Apart from the tragedy of those who actually die from the operation there is no evidence to show that the survivors derive any benefit that might compensate the bereaved.

The perfection of techniques for intravenous therapy has blinded people to the fact that 'drips carry risks'. There is no doubt that the benefits far outweigh any disadvantages but it is no excuse for using intravenous therapy when there is no indication for it. The wrong blood group, the wrong electrolyte, overloading the circulation, and accidental infection are always possible.

Another area where great potential damage may be done is in the use of X-rays. These are just as much radiation as the aftermath of exploding nuclear devices. Nobody can foresee the future of the world in that respect or, more immediately, the future of the child being X-rayed, and who knows but that the unnecessary X-ray taken now might just be the item to tip the balance to malignancy in later

life. To fail to X-ray when necessary can be criminally negligent but to do so repeatedly when there is no indication may be equally negligent. Since nobody can know at that stage, the temptation is very great.

Whatever a practitioner is doing, be it investigation or treatment, restriction or enforcement, he or she should question whether it is really required or, perhaps, damaging. The fact that something has been done for aeons by generations of doctors does not, by implication, exclude it from this test.

2

Diseases Due to Infection

A vast number of diseases are generated in infections which, in turn, are derived from a variety of sources. In the classification of these diseases, the acute infectious fevers are grouped together because they have certain clinical features in common.

THE ACUTE INFECTIOUS FEVERS

To most people, some of these fevers are thought of as an inevitable occurrence at some time during childhood and they have come to be known as 'the childish diseases', the word childish in this context meaning unimportant. It might amaze such people to know that the term could be applied equally to commemorate the fact that in the course of history many children have been killed or maimed by these fevers.

The features common to all are—

Infectivity. This varies, although in all cases it can cause epidemics.
A fixed incubation period.
A specific clinical illness from which, if uncomplicated, recovery is complete.
An attack confers prolonged, usually lifelong, immunity.

In these illnesses it is helpful to memorise the various incubation periods, and the day of the illness on which a rash, the exanthem appears. There are many occasions in medicine when ignorance can

be masked by temporisation until one has had an opportunity to look it up. Infectious fevers are not in that category, as both the incubation period and the day on which the rash appears are essentials to accurate diagnosis, and there will be loss of face or a missed diagnosis or both, if the facts are not to hand.

Incubation Periods In the table below the grouping is sufficiently accurate for easy memorisation.

One to seven days:	scarlet fever diphtheria
Nine to fifteen days:	measles poliomyelitis roseola infantum
Twelve days:	smallpox
Two to three weeks:	rubella varicella pertussis typhoid
Two to four weeks:	mumps

Day of Illness on which the Rash Appears

This is most easily memorised by the old classic mnemonic (*see* below) which is read from above downwards, the initial letter of each word of the mnemonic and the disease being the same.

Rash appears	*Mnemonic*	*Infectious fever*
Day 1	Really	Rubella
Day 2	Sick	Scarlatina
Day 3	People	Pox (smallpox)
Day 4	Must	Measles
Day 5	Take	Typhus
Day 6	No	None
Day 7	Exercise	Enteric (typhoid)

Infectivity

The degree of infectivity of each of these diseases varies a good deal and is of importance in assessing the chances of spread. The list below gives an overall picture.

Extremely Infectious:	measles smallpox pertussis
Moderately Infectious:	rubella varicella mumps poliomyelitis
Low Infectivity:	scarlet fever diphtheria roseola infantum

Duration of Infectivity

Scarlet fever:	24 hours prior to start of antibiotic treatment.
Diphtheria:	from onset until three cultures taken on consecutive days are negative.
Measles:	from onset of fever for approximately ten days.
German measles:	one week from onset.
Smallpox:	from onset of rash until all scabs are shed.
Chickenpox:	for one week from onset.
Pertussis:	for three to four weeks from onset.
Mumps:	one week before and two weeks after appearance of parotid swelling.
Poliomyelitis:	first week by droplet infection; up to four weeks in the faeces.

Isolation Period

The recommendations vary according to local views. At one time 'measles parties' were fashionable to spread the disease and have done with it. Little heed was given to the price paid by some. The modern view is a compromise between unnecessary risk, on the one

hand, and disorganising life and schooling, on the other. Thus, with mumps, only non-immune children beyond puberty should avoid contact, and in measles (apart from the question of vaccines) children under the age of three should be protected.

Diphtheria, typhoid, poliomyelitis and smallpox require strict isolation until the risk of infection has ceased.

Symptomatology

Were one to analyse an average textbook of medicine it would cause surprise to realise how much space is given to repetition of the symptoms of acute fever. For practical purposes, whatever the aetiology the picture is the same and may be summarised as follows—

headache,
pain in the back and limbs, sore throat
vomiting,
a rash, perhaps.

In addition, there is raised temperature and pulse rate.

SCARLET FEVER (scarlatina)

This once common, important and sinister disease is now neither common, important nor sinister. It was never really a disease in its own right, just a haemolytic streptococcal infection with a characteristic rash. Prior to the introduction of sulphonamides and penicillin, ENT wards were the prime example of the failure to recognise the true relationship. A child with streptococcal mastoiditis would be admitted for operation, and within a week several other children developed scarlatina. The ward was then closed to further admissions for three weeks, whereupon the whole process repeated itself with the result that the wards were closed more often than they were open.

Clinical Picture. This consists of marked signs of fever and an acute pharyngitis. The tonsils are intensely inflamed with a purulent exudate which on culture grows beta haemolytic streptococci. The tongue is heavily coated and the inflamed papillae show through, producing, in the typically medical habit of comparing the delectable with the revolting, 'strawberry and cream tongue'. The cervical glands are markedly enlarged.

On the second day of the illness the rash appears, first in the