Paediatric Orthopaedics and Fractures

IN TWO VOLUMES

VOLUME 1

W.J.W.SHARRARD

SECOND EDITION

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Preface to the Second Edition

Advances that have taken place in the last seven years have shown that orthopaedic surgery continues to be a progressive discipline. In almost every aspect of paediatric orthopaedics, new findings, advances in technique and better results of treatment have been discovered. In preparing the first edition, one of my aims was to ensure that the text was as up to date as possible; in this edition I have tried to maintain this standard. Every chapter has been progressively revised in the light of current publication and my own continuing experience.

Some sections, particularly those in which considerable changes have taken place, as in the surgery of the spine, have been recast, and, it is hoped, improved. Other parts have needed less drastic change, but few have remained static.

There are those who say that further editions of a book should not be larger than their predecessors. No apology is offered for the fact that this edition is larger and more profusely illustrated than before. Those who use the text as a reference wish for more completeness, for example, in the cataloguing and illustration of abnormalities, diseases and syndromes. Those who use it for practical surgery have asked for additional content of operative techniques. I have tried, at least in part, to meet both requirements. Wherever time or trial has shown that methods of management or operative procedures are no longer useful they have been eliminated except for historical mention.

W.J.W.SHARRARD

Preface to the First Edition

The last 20 years have shown a remarkable advance in orthopaedic surgery in children. Better understanding and knowledge of intra-uterine development, of the genetics of orthopaedic conditions, of the causation, diagnosis and treatment of congenital dislocation of the hip, talipes equinovarus, scoliosis, poliomyelitis, cerebral palsy, spina bifida, osteomyelitis and tuberculosis has made much that was previously written in orthopaedic textbooks outdated. The trend of orthopaedic treatment has moved away from management by manipulation and splintage to the rational use of surgery, aided by improved surgical and anaesthetic techniques and by the better protection afforded by antibiotic drugs.

The realization that some conditions normally correct themselves with growth and are not, strictly speaking, true deformities but variations of normal development has meant that some of the measures previously employed in treatment can be discarded.

From my own experience and from the writings of others, I have tried to put together all that is currently useful in the treatment of orthopaedic conditions in childhood. The aim of this book is to help the practising orthopaedic surgeon and orthopaedic surgical trainee, but I hope that it will be read and used for reference by any who practise in the field of paediatric work.

If there is a theme that is common to almost all orthopaedic conditions in childhood, it is concerned with the effects of growth and development. It is this that separates the measures that are used in childhood from those in adults and it is the main reason why it is appropriate that there should be an orthopaedic textbook devoted entirely to paediatric conditions.

I have tried to balance the content fairly between all aspects of work as they come to the orthopaedic surgeon. Some diseases, such as tuberculosis and poliomyelitis, no longer form a large part of orthopaedic surgery in Europe and North America but substantial portions of the text deal with these subjects because of their continuing prevalence in many parts of the world. In some fields, such as cerebral palsy and spina bifida, there is need for the use of sophisticated techniques and multidisciplinary management that may, as yet, only be applicable in centres in which all facets of medical and surgical expertise are available. The complex conditions of today are the commonplace conditions of tomorrow and there are few hospitals where it is not possible to gather together a team of individuals capable of treating major abnormalities by combined action. For those who thirst for additional and more detailed information, each chapter is provided with a list of references that seem to the author to be authoritative or useful.

W.J.W.SHARRARD

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Chapter 1

Diagnosis and General Management of Orthopaedic Conditions in Childhood

The management of diseases and injuries of the musculoskeletal system in childhood is dominated by concern with continuing growth and development of the child as a whole and of the component tissues of the trunk and limbs. Primary and secondary abnormalities of growth and development constitute the major part of orthopaedic and traumatic conditions in childhood and, in contrast with deformity in adults, deformity in children is often progressive. Conditions such as osteomyelitis, rheumatoid arthritis or fractures affect children and adults but evoke a different response in childhood and methods of treatment must be varied accordingly. Orthopaedic and traumatic conditions in childhood should not, therefore, be considered in isolation from, but in comparison and contrast with, those in adults. The failures of treatment in childhood often become the problems of treatment in adults; the orthopaedic surgeon who cares for both is more sensitive of this and keen to avoid future disability that may still remain on his doorstep.

The child is not to be regarded as a small version of an adult and the younger the child, the more differences there are in his anatomical form, physiological function and response to disease, injury and surgical assault. Between birth and the completion of growth, a child passes through several stages of development. Each requires a different approach in diagnosis and management and in the nature of the disease or abnormality that is likely to be present. The stages can be conveniently divided into:

1. The neonatal period during the first 4 weeks of life.
2. The period of infancy, during the first 2 years.
3. Early childhood, between the third and fifth year.
4. Later childhood, between the sixth and tenth year.
5. Pre-adolescent and adolescent period.

It is not appropriate to a postgraduate text to describe the basic details of history-taking, examination and treatment, but matters of particular importance in each of these periods of life and development will be discussed briefly in this chapter.

HISTORY-TAKING

In the neonatal period, the commonest orthopaedic problem is likely to be that of a congenital deformity. A history may have to be sought from the mother about similar or

related deformities in relatives and about any illness or medication during pregnancy. The obstetrician may be able to give information about complications during pregnancy or difficulty in labour or delivery, particularly if there have been any sequelae of obstetrical trauma. The paediatrician or general practitioner who has made a general examination of the newborn baby is likely to be the individual who refers for an orthopaedic opinion and his findings, particularly of evidence of congenital dislocation of the hip, may be of great importance if the infant is not seen until the third or fourth day when some of the relevant signs may have disappeared or altered. In the later months of life, the paediatrician or general practitioner may again be the individual who notices a deformity or abnormality at a postnatal examination, or the mother may have noticed limited abduction of the hips or asymmetrical limb movements.

The period of infancy, during the first 2 years of life, is one in which there is often parental anxiety about the development of temporary deformity such as bow leg, flat foot, knock knee or in-toeing when the child starts to walk. At this time of life it is the parents who are likely to provide the history and their knowledge of their child's normal abilities and behaviour should not be ignored if they discover differences in gait or evidence of pain that warrants orthopaedic examination.

During early childhood, the child himself can contribute to the history. The parents still take precedence because the child at this period of life is unable to recognize the passage of time with any accuracy though he will usually indicate the site of pain fairly precisely. A history of injury, on the other hand, may not be obtained from either parents or child and may sometimes have to be assumed.

In later childhood, after the sixth year, the child himself usually gives a clear, accurate and unbiased history of his condition. He answers questions concisely and almost invariably truthfully. A complaint of pain should never be disregarded during this period of life. Children may sometimes fake illness to parents to avoid school but they seldom continue to do so when they are seen by a doctor. The same is not necessarily true in adolescence, when anxiety state, hysteria and other neurotic manifestations may occasionally present as orthopaedic complaints.

EXAMINATION

Examination in childhood by inspection, palpation, percussion (over bone), determination of active and passive joint movement and measurement do not vary much at any period of life and follow the same lines as in adults. In infants and young children the limbs need to be handled gently, the child being watched to make sure that pain is not being elicited and that excessive force is not being used. Active movements at this period of life can only be discovered by allowing the child to move spontaneously and the baby who is sleepy or a young child who is reluctant or embarrassed may need to be examined on more than one occasion to make sure that active movements are present.

A knowledge of the normal anatomy and physiology of body form and function at each period of life is vital if abnormality is to be discovered. For example, the prominent bases of the fifth metatarsals in children during early adolescence and arising because of the more rapid growth of the forefoot compared with the hindfoot need to be recognized to

avoid the hazard of removing what appear to be exostoses. Overall function in the limbs also needs to be measured in relation to the age of the child and in this respect the orthopaedic surgeon should be aware, as the paediatrician is, of exactly what a child should be capable of doing at each period of life.

The maintenance of good records of the results of examination applies to all aspects of medical and surgical work. In childhood, the description of a deformity must be such that it can be compared with the state at a later date to determine whether it is progressing or altering. Clinical photography is a valuable adjunct to a medical record in this context. Ranges of joint movements should be registered in a standard form as recommended by the American Academy of Orthopaedic Surgeons (1965) and preferably measured by a goniometer, so that a decreasing or increasing range of movement can be identified at subsequent examinations.

Neurological abnormality is so commonly the cause of orthopaedic disability in childhood that an adequate neurological examination should be part of the orthopaedic assessment of any child in whom a neurological cause is suspected. The sequential development and disappearance of reflexes such as the tonic neck reflex, grasp reflex, Landau trunk extension reflex when the child is prone, Galant's reflex incurvation of the trunk, stepping and placing reflexes and the extensor plantar response should be known to the orthopaedic surgeon concerned with the management of children and particularly those who may have cerebral palsy. The determination of motor power in muscle groups and in individual muscles is essential both for diagnosis and for decisions about treatment in all kinds of limb paralysis. In the neonatal period, it should be possible to determine whether any individual muscle group is showing spontaneous activity or not though it may be difficult to know whether such activity is voluntary or involuntary. It may also be a matter of judgement whether a muscle is regarded as strong or weak until the child is able to respond to instruction reliably, which is not achieved until later childhood. The recognition of intrauterine paralysis as a cause of congenital deformities previously labelled arthrogryposis implies that muscle activity in the newborn should be assessed to try to discover any gross evidence of unbalanced muscle action. The absence of normal skin creases in, for example, the sole of the foot or on the dorsum of the ankle, is a useful indication that movement has not been occurring *in utero* and that the muscles concerned may be paralysed or abnormal. After the fifth year, it is normally possible to record the power of individual muscles or movements in a system of grading (Medical Research Council 1942) against which subsequent estimates can be compared in paralytic conditions.

Sensory testing is almost always unreliable in infancy and early childhood and is not completely reliable until a child is over 9 or 10 years old but complete sensory loss can usually be discovered by the obvious response or absence of it to pin-prick.

Finally, the circumstances of examination may make a great difference to the findings that can be elicited. A child who is frightened or embarrassed may become almost impossible to examine. Many children are frightened of white coats and it is the author's practice never to wear a white coat when doing a children's clinic. To insist on undressing a child completely, except an infant, is likely to invite disaster and as much of the examination as possible should be made with the younger child sitting on his parent's knee. Only after this should the child be undressed and it is reasonable to respect his wish for privacy and his or her reluctance to remove all underclothing in public.

RADIOLOGICAL AND ALLIED INVESTIGATIONS

Just as assessment of abnormality in a child depends on a knowledge of normal anatomy and function for his age, so does the interpretation of radiographs depend on a knowledge of normal bone form and architecture and its modifications with age and growth. In infancy, the absence of osseous epiphysial nuclei and the variation in age at which epiphysial centres may appear increases diagnostic difficulties. It is well to remember that the radiological shape of a bone is not necessarily a reliable guide to its true form. An appearance of sloping of the acetabular roof in infancy does not necessarily indicate that the acetabulum itself is shallow, just as the appearance of the epiphysial nucleus of the head of the femur is not necessarily a guide to the shape of the femoral head. Arthrography can be usefully employed to determine the true shape of articular surfaces at any joint though it is most often used to determine the condition of the hip. Radiographs taken with decreased penetration may sometimes be valuable to indicate the form of cartilaginous structures.

Cine-radiography, possibly combined with arthrography, can also contribute useful information about joint movement in certain situations. Special techniques may be needed to investigate specific abnormalities in childhood such as anteversion of the femoral neck or tarsal coalition and serial radiographic measurement is almost essential in the management of scoliosis and inequality of limb length. Tomography may sometimes be needed in the diagnosis or localization of lesions in bone. Computerized axial transverse tomography (Emiscan) has its main application in the investigation of intracerebral lesions, but there is a potential use in children for the assessment of femoral or acetabular anteversion and of lesions within the spinal canal. Myelography using water-soluble contrast medium is now acceptable for use in childhood, replacing oil-soluble media which were not advisable because of their failure to be absorbed and their tendency to irritate the tissues.

In the management of some lesions, such as congenital dislocation of the hip, scoliosis or in multiple abnormalities, the need for repeated radiographic examination must be weighed against the dangers of excessive irradiation, particularly of the abdominal and pelvic contents or of growing epiphyses.

Radiological diagnosis is the proper domain of the radiologist and, in certain aspects of paediatric work, the radiologist with a special knowledge of paediatric radiology can give valuable assistance to the orthopaedic surgeon. In lesions of complexity or doubt, particularly in general affections of the skeleton, texts such as those of Fairbank (1951) or Caffey (1967) are invaluable for reference.

Bone scanning with radioactive isotopes can be used to study a wide variety of bone lesions, especially neoplastic deposits before they are visible in radiographs, in the localization of infective lesions, osteoid osteoma or occult fracture, or in the investigation of bone changes in Perthes' disease. Strontium 85 and 87 and fluorine 18 have been used in bone scanning but are not suitable for use in children. Technetium-labelled phosphate compounds are probably the most satisfactory since they are safe, with a radiation dose equal to a plain radiograph, and give high quality scans with a variety of scanning apparatus.

HAEMATOLOGICAL, BIOCHEMICAL AND OTHER SPECIAL INVESTIGATIONS

Routine haematological investigations such as the erythrocyte sedimentation rate and differential white-cell count are applicable to the diagnosis of inflammatory or neoplastic conditions in paediatric orthopaedics as in other branches of medical work. The special blood investigations required for the diagnosis of bleeding diseases may be needed for the first time in a child presenting with haemophilic haemarthrosis or in the diagnosis of haemoglobinopathies or reticuloendothelial lesions.

Biochemical investigations may aid the diagnosis of nutritional, metabolic or general skeletal abnormalities, the most important measurements being those of serum calcium,

TABLE 1.1. Blood chemistry in skeletal disease

Disease	Serum calcium (normal 2·1–2·6 mmol/l = 8·5–10·5 mg/100 ml)	Serum phosphorus (normal 0·8–1·4 mmol/l = 2·6–4·6 mg/100 ml)	Alkaline phosphatase (normal 40–140 u/l = 5·5–20·0 KA units/100 ml)
Vitamin-D-deficiency and phosphaturic rickets	Normal	Diminished	Normal or increased
Malabsorption rickets	Diminished	Normal or diminished	Increased
Renal osteodystrophy	Diminished	Increased	Sometimes increased
Scurvy	Normal	Normal	Diminished
Polyostotic fibrous dysplasia	Normal	Normal	Often increased
Osteogenesis imperfecta	Usually normal	Diminished	Slightly increased
Osteopetrosis	Normal or increased	Normal or diminished	Normal
Hypothyroidism	Normal	Normal	Markedly diminished

phosphorus and alkaline phosphatase (Table 1.1). Estimations of serum enzymes such as the transaminases, aldolase or creatine phosphokinase are needed in investigation of diseases of muscle. Very occasionally, estimation of blood lead may be needed when radiographs show suspicion of chronic lead ingestion.

Investigation of fluid aspirated from joints may be valuable in the differential diagnosis of joint disease in childhood. Some characteristic synovial fluid changes found are shown in Table 1.2 (Ropes & Bauer 1953).

In bone or joint lesions in which an infection is suspected, apart from blood culture or culture from aspirated joint fluid in acute osteomyelitis or septic arthritis, the diagnosis in subacute or chronic infective lesions may require a battery of tests to indicate previous infection by a relevant organism. A negative tuberculin test is especially valuable in the exclusion of all but a very few tuberculous lesions and altered titres may aid the

TABLE 1.2. Characteristics of normal and abnormal synovial fluid (from Ropes & Bauer 1953)

Joint conditions	Appearance	Erythrocytes	Leucocytes	Predominant cell type	Mucin clot test	Sugar (mg per cent)	Total protein (mg per cent)	Organisms
Normal	Clear	A few	100	Mononuclear	Normal	90	1·7	
Traumatic arthritis	Slightly turbid	2,000	1,500	Mononuclear	Normal	90	4·0	
Haemophilic arthritis	Turbid and red	2,500,000	5,000	Polymorphonuclear	Poor	60	5·9	
Rheumatoid arthritis	Clear	2,000	15,000	Polymorphonuclear	Poor	78	4·7	
Tuberculous arthritis	Turbid	28,000	20,000	Polymorphonuclear	Poor	27	5·3	
Septic arthritis	Very turbid	30,000	80,000	Polymorphonuclear	Poor	21	4·8	+

+ = present.

diagnosis of earlier infection by streptococci, staphylococci, salmonella organisms or brucellosis. The Wassermann test and treponema immobilization test may be needed for the diagnosis of syphilis or yaws. Bacteriological culture or animal inoculation of bone, joint or lymph gland biopsy material is almost essential in the diagnosis of a difficult inefective lesion, especially if tuberculosis is suspected.

Investigation of amniotic fluid obtained during pregnancy allows prenatal diagnosis of certain congenital abnormalities. The test is applicable when previous pregnancy, family history or foetal assessment gives rise to the possibility of an abnormality of the foetus that could justify termination of the pregnancy. The amniotic fluid is obtained between the 14th and 20th week of pregnancy and investigated by direct study or after tissue culture (Edwards 1972). The fluid may contain abnormal amounts of bilirubin, amino-acids or proteins. Excessive amounts of alphafetoprotein are present in anencephaly and open myelomeningocele, an observation first made by Brock & Sutcliffe (1972). Leek *et al.* (1973) indicated that alpha-fetoprotein estimation in maternal serum might be of value as a diagnostic test that could be used in any pregnancy for possible anencephaly or neural tube defects. Initial results have shown that many, but probably not all abnormal foetuses can be identified and subsequently confirmed by amniocentesis. It is likely that more than half of all pregnancies associated with a foetus with a major neural tube defect will be able to be identified prenatally in this way. The position is fully reviewed by Brock (1976). Cells in amniotic fluid can be studied for sex chromatin to reveal the sex of the foetus, of importance in serious sex-linked diseases such as Duchenne muscular dystrophy. Amnion cell culture (Laurence & Gregory 1976) allows chromosome abnormalities such as the trisomies to be defined and conditions with abnormal enzyme activity such as hyperphosphatasia or homocystinuria to be recognized. Generalized skeletal diseases such as mucopolysaccharidosis may be diagnosed by the discovery of an enzyme deficiency specific to the disease.

BIOPSY

Histological examination of biopsied material is an essential part of the diagnostic investigation of some diseases of the musculoskeletal system in childhood. In a number of bony lesions with cystic, destructive or reactive changes on the radiograph, bone biopsy may establish the diagnosis, especially when osteosarcoma is suspected; conversely, the histological demonstration of a benign lesion such as a chondroblastoma or chondromyxoid fibroma may occasionally avert the tragedy of an amputation for a non-malignant bone condition. Electron microscopy may allow conditions that cannot be distinguished by light microscopy to be differentiated, for example, Ewing's tumour from secondary bone deposits in malignant neuroblastoma.

Muscle, muscle and nerve, or nerve biopsy may establish the diagnosis in diseases of muscle or in affections of the upper or lower motor neurone. Some techniques of histochemical examination require that the sample should be fixed in special reagents immediately after removal and the biopsy should ideally be taken from a partially affected muscle. Ultramicroscopy, histochemistry or tissue culture may help diagnosis in some diseases of bone, cartilage or muscle, and fibroblast tissue culture from skin biopsy allows biochemical study of enzyme defects to be made.

ELECTRODIAGNOSTIC METHODS

Electrical tests are useful adjuncts to neurological examination in the diagnosis of paralytic lesions and sometimes in the assessment of recovery or of the effects of treatment. The simplest, and the most useful, general test is the response of the muscle to long-duration (galvanic) or short-duration (faradic) stimulation. In children it may be difficult to determine the presence of lower motor neurone paralysis following possible nerve injury or in association with developmental anomalies of the neuraxis, and the results of the stimulation may help to confirm clinical estimates of the paralysis. In lesions such as obstetrical paralysis, the plotting of intensity-duration curves may give additional information about recovery or may indicate progressive denervation in lesions such as lipoma of the cauda equina. Nerve conduction techniques may occasionally be useful in the diagnosis of spinal cord lesions from peripheral neuropathy in children presenting with obscure muscle weakness.

Electromyography can be employed in the diagnosis of myopathic lesions from lesions of the lower motor neurone and will indicate the impending re-innervation of a muscle after nerve injury at least a month before clinical tests can do so. In infancy, electromyography is much more difficult to use because the patient cannot be asked to perform specific actions to cause muscles to contract and, in a small child, needle electrodes are preferable to surface electrodes if results are to be conclusive. A concentric needle through which a stimulus can be applied to determine into which muscle the needle has been inserted is extremely helpful in the performance of electromyography in small babies. Cortical evoked responses have a place in the assessment of sensory function in babies and young children and can help to distinguish between lower motor neurone paralysis and loss of continuity in the spinal cord in lesions of the neuraxis.

DIAGNOSIS

The art of diagnosis lies in the weighing of the significance of facts and observations. Deformity or abnormality of musculoskeletal function is the most common reason for orthopaedic consultation. Most deformities are not painful, and a child, even when old enough to do so, seldom complains about the deformity. If the deformity has been present since early childhood, he will have accepted it as part of himself and, even if it does impair function or cause him pain, he has no standards by which to judge his own degree of disability. A child, for instance, whose feet ache after he has walked for a distance, may not make any complaint because his feet have always behaved in this way.

It is often an abnormal appearance in a child's limb, or an abnormality of gait of which the child is unaware, that is observed by a parent and is the reason for orthopaedic assessment. The parents are, rightly, concerned with the effect that the abnormality may have if it persists into adult life and their views may be coloured by personal experience of a similar abnormality, such as a curly toe, that was disregarded in their own childhood.

Diagnosis in childhood is much more dependent on the results of clinical examination than it is in adult life. In many adult orthopaedic conditions, the history alone is sufficient to allow a provisional diagnosis to be made; in childhood, and particularly in infancy, the diagnosis may be made almost entirely on the results of clinical examination.

A child has a much greater capacity to compensate for abnormality or defect than does an adult. Even a single abnormal physical sign must be given serious consideration, especially if it is asymmetrical; sometimes the only abnormal physical sign in an established Perthes' disease may be limitation of medial rotation of one hip.

A complaint of pain or the discovery of tenderness should always be given very serious consideration. Normal growth is never painful and even when severe deformity is present it is remarkable how seldom there is any complaint of pain. A joint that is painful to move or a bone that is tender is almost always indicative of significant damage to bone or soft tissues by injury, inflammation or neoplastic change.

TREATMENT

The success of treatment of any condition depends on an understanding of its causation and a knowledge of the effects of the application of therapy. In recent years, considerable advances have been made in knowledge of the causation of deformity and disease in childhood. It is no longer appropriate to regard a deformity such as talipes equinovarus or dislocation of the hip as a consistent entity for which a particular form of management is always appropriate or effective. Equinovarus deformity of the foot, for example, may arise *in utero* as a result of primary malformation of bone and soft tissue, as an intra-uterine deformity secondary to paralysis, contracture or maldevelopment of muscles, tendons or soft tissues or by the effects of intra-uterine posture. The same deformity may arise after birth as a result of paralysis secondary to poliomyelitis, cerebral palsy, spinal cord abnormalities, nerve or soft tissue injury or even bony injury or infection. The application of treatment has been rationalized by better understanding of what can be achieved by splintage, physical therapy or surgery and, in some instances, by the recognition of the fact that improvement that was previously thought to be the result of treatment was the result of spontaneous correction by the normal mechanisms of growth.

Splintage

Splints and supports by external apparatus are applicable for three purposes:

1. To immobilize a limb or part of a limb in the management of injury, infection or inflammation. The techniques and application in childhood do not differ in principle from those in adults. Function is usually restored more easily in childhood and, provided that anatomical continuity is maintained, joints will mobilize more readily than in adult life. It is possible, for example, to immobilize the elbow in the extended position in the treatment of a flexion supracondylar fracture for a limited period in childhood in the confidence that elbow flexion will recover.

2. To support a paralysed or hypermobile joint to improve its function. A flail ankle or knee associated with paralysis due to poliomyelitis or complete loss of function in the lumbosacral spinal cord or roots is an absolute indication for external splintage to make walking possible. Its function in this respect must not be confused with the use of splintage to attempt to prevent deformity in the presence of partial paralysis, nor should splintage be used if it is possible to obtain or restore sufficient activity by physical therapy or surgery to avoid the need for external splintage. If, for example, dorsiflexor power is