Common Orthopedic Problems in Children

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Raven Press ■ New York

Raven Press, 1140 Avenue of the Americas, New York, New York 10036

Made in the United States of America

Library of Congress Cataloging in Publication Data

Katz, Jacob F.
Common orthopedic problems in children.

Includes bibliographies and index.
1. Pediatric orthopedia. I. Title.
[DNLM: 1. Orthopedics—In infancy and childhood. WS 270 K19c]
RD732.3.C48K37 617.3'0088054 77-85071
ISBN 0-89004-273-X AACR2

Preface

Pediatric orthopedics has gradually assumed more importance as a specific body of information within the scope of general orthopedics. Just as pediatrics has developed from general medicine to respond to the unique needs of the growing and developing child, so have the other specialties in their pediatric relationships.

This volume encompasses the common conditions seen in the musculo-skeletal system of children from infancy through adolescence, including congenital anomalies, genetic diseases, developmental deformities, tumors, injuries, and infections.

Diagnosis, clinical course, and natural history are stressed in the discussion of orthopedic problems. Therapy is usually indicated in principle; technological details are omitted. The generous use of photographs facilitates identification of the various disorders. The organization of the text generally follows regional anatomical localizations. A comprehensive glossary at the end of the volume bridges the gap of technical language to make it readily available to the nonorthopedic surgeons, yet contains sufficient documentation to serve as a refresher to the orthopedic surgeon, whose major interests are other musculo-skeletal facets.

This volume will be of interest to pediatricians, orthopedists, family practice physicians, physiatrists, and particularly to medical students during pediatric and orthopedic rotations.

Jacob F. Katz

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Skeletal Growth and Development

The first 8 weeks after fertilization of the ovum is termed the *embryonic period*. During this time the primitive cell mass differentiates into layers of cells (ectoderm, endoderm, and mesoderm) that later give rise to the various organ systems. The musculoskeletal system develops mainly from mesenchyme. The axial skeleton appears as segmental condensations which ultimately approximate the definitive vertebral shapes. Subsequently, these are converted to cartilage models, which, in turn, develop ossific centers. The appendicular skeleton appears primitively as cranial and caudal limb buds, which differentiate into arms and legs. These also are converted to cartilaginous models of definitive outlines which then develop ossific centers. Teratologic malformations commonly start during this early period. The *fetal period* then follows, encompassing the growth and development up to the time of birth. New definitive rotational relationships develop between the limbs and the axial skeleton during this interval, bringing the horizontally oriented anlage to the more functional vertical position of the fully developed upper and lower limbs.

The position of the enlarging fetus within the uterine enclosure is determined by a variety of external compressive factors. Constraint ordinarily increases as the fetus distends the uterus during late gestation. This constraint is most obvious with a very large fetus, multiple fetuses, increased uterine muscle tone, uterine malformations that cause a decreased uterine volume, or diminished amniotic fluid. Constraints on the fetus may result in various limb entrapments, which produce developmental abnormalities.

NORMAL OSTEOGENESIS FROM CARTILAGINOUS PRECURSORS

Ossification of the limb cartilage model begins with vascular penetration at mid-diaphysis, where there is concomitant cartilage cell hypertrophy and destruction, matrix calcification, and finally bone formation. This process extends outward toward each bone end, stopping at a predetermined distance from each epiphysis, which remains cartilaginous. Until this time, growth occurs by cartilage cell multiplication throughout the limb. After bone is formed, however, growth occurs from the epiphysial growth plate at each end of the bone. Peripheral or circumferential growth derives from contributions to the surface, initially from the perichondrium and later from the periosteum, via membranous bone formation.

At later, predetermined times, a similar process of bone formation takes place within each epiphysis. This process also begins as bone conversion in the center; the bone gradually enlarges until only the subarticular zone of the epiphysis retains its growth potential, which remains until skeletal maturity.

IN UTERO POSITIONING

At birth the infant can usually be "folded" into its uterine position by the examiner. This position is obviously a comfortable, familiar pose that reflects any abnormal posture *in utero* and indicates the possibility of a subsequent deformity that will fail to resolve spontaneously.

In "small-for-age" infants, such deformities may vary from generalized compression to those resulting in peculiar head and face configurations, adducted and extended lower limbs associated with hip and knee dislocations, and a variety of foot deformities, from clubfeet to distorted toes.

2

Pediatric Orthopedic Examination

Although children may develop orthopedic abnormalities at any age, many such conditions appear with increased frequency during specific age periods. Fractures and bone infections may occur at any point during childhood, whereas congenital hip dislocation is commonly seen during early infancy, the osteochondroses mostly during mid and late childhood, and scoliosis and slipped capital femoral epiphysis primarily during late childhood and early adolescence. Pediatricians and orthopedists traditionally follow maturational developmental changes of children and so are in a position to detect musculoskeletal abnormalities when they first appear.

A careful orthopedic examination performed on the newborn infant will detect abnormalities of the prenatal and perinatal periods. Subsequent re-examinations are necessary to determine if any new problems have appeared as well as to monitor the status of those seen earlier.

ORTHOPEDIC EXAMINATION OF THE NEWBORN

Inspection

The normal unrestrained newborn infant actively moves all extremities when not sleeping; therefore any sign of asymmetrical limb flaccidity or immobility must be regarded with suspicion. Failure to move an extremity may result from local injury (e.g., fracture or epiphysial displacement) or from more distant neural damage (e.g., spinal cord or plexus injury).

Under ordinary circumstances limb attitudes at birth reflect fetal intrauterine positions (Fig. 2.1). In the normal infant, elbows, hips, and knees are in flexed positions; the hips are moderately abducted; and the feet may be fixed in either internal or external rotation, carrying the tibias along in

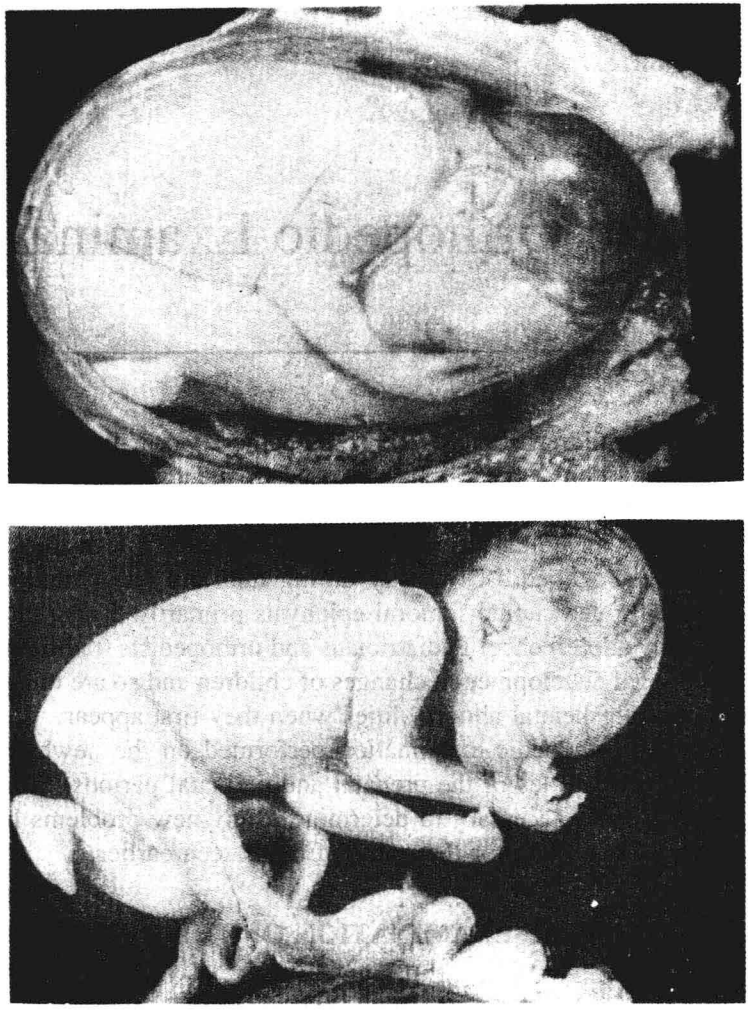

FIG. 2.1. Fetus demonstrating the effect of intrauterine positioning during late trimester development. **Top:** Position *in utero*. **Bottom:** Residual limb attitudes after delivery. Reprinted with permission from *Medical Radiography and Photography*.

this rotational relationship. There are individual variations in this entrapped position, so that one foot may turn inward and the other outward. The presence of internal and external tibial torsion as well as metatarsus adductus is diagnosed by examining these positions. When the feet, in mild external rotation, are forced into prominent dorsiflexion against the uterine wall,

they assume a calcaneovalgus attitude. These mild developmental variations must be differentiated from clubfeet and congenital flatfeet, more serious abnormalities which are discussed later.

Careful inspection reveals any disproportion between the trunk and lower limb lengths, as in achondroplastic dwarfism. Proportional dwarfism cannot be identified with certainty at this time, however. Absence of limbs or parts of limbs as well as angular distortions of extremities are easily diagnosed. Outpouchings over the lower back usually are consistent with myelomeningocele or meningocele; hydrocephalus is also occasionally associated with these abnormalities. Other spinal anomalies (e.g., scoliosis or absence of vertebra) may be suspected when there is trunk asymmetry.

Palpation and Manipulation

The portion of the examination wherein the neonate is palpated and manipulated must be conducted in a disciplined manner. The examiner starts at the head and neck, where to determine mobility the infant is examined via flexion and extension, lateral rotation, and lateral bending maneuvers. Palpation is then directed to the sternomastoid muscles as well as the adjacent clavicular areas. Neck restrictions may be associated with congenital anomalous development of the cervical spine as in the Klippel-Feil syndrome or, more commonly, with muscle tightness as in muscular torticollis.

The motion of the shoulders, elbows, forearms, wrists, and hands is tested by manipulation. Forearm restriction may signify radioulnar synostosis. Thumb flexion may be associated with "trigger" thumb, resulting from constriction of the thumb flexor tendon in its sheath. More extensive contractural processes frequently signify arthrogryposis, a prenatal condition associated with lack of motion on either a myopathic or a neurologic basis (see page 123).

Examining the hips to detect congenital dislocation or dislocatability is of prime importance. Although infants with teratological or syndrome-associated abnormalities may be born with prenatal dislocations, the most common form of dislocation is that which appears with hip joint laxity at birth. Usually hip motions are unrestricted at this early stage of development. Leg length discrepancies may be present in unilateral complete dislocations, but equal lower limb lengths do not necessarily signify a normal state. An adequate examination involves stressing the hips for dislocatability and relocatability with appropriate techniques (Barlow and Ortolani maneuvers; see *Glossary*).

Ordinarily the knees can be expected to flex fully but not extend fully because of physiological contracture resulting from the flexed fetal position. In certain unusual cases (e.g., intrauterine dislocation seen in Larsen's disease; page 120), the knees may be hyperextended. Gentle flexion often returns them to normal position, although more complicated maneuvers are sometimes required.

Mobility of the ankles and feet is determined. Equinus ankle contractures usually are associated with additional deformities. The normal ankle can be dorsiflexed and plantarflexed equally. In calcaneovalgus there is more than customary dorsiflexion, with mild restriction in plantar flexion along with abduction of the forefoot. In clubfoot as well as congenital flatfoot (vertical talus), the ankle cannot be brought into dorsiflexion. In these instances there are accompanying rigid foot deformities; these are seen as inversion and adduction in clubfoot, and as eversion and abduction in congenital flatfoot.

EXAMINATION OF THE YOUNG CHILD

As the infant grows and attains an upright posture and some degree of walking stability, new postural relationships develop. The examiner must now review all of the areas previously checked as well as study those brought into focus by the erect bearing and locomotion.

The child's symmetry in walking and lying is inspected. The position of the head and any spinal deviation are noted. Spinal asymmetries, although not common at this early age, are occasionally seen with congenital vertebral anomalies or, less frequently, with idiopathic scoliosis. Mild lower limb asymmetry, on the other hand, is common. As a rule, the newly walking child generally exhibits some degree of asymmetry in standing attitudes or gait patterns. These may be either habit patterns or postures resulting from tibial torsions or intrinsic foot distortions, or from a not yet fully developed gait pattern. Tibial bowing may be seen either alone or in combination with tibial torsion. Knock-knees are not likely to be present at this period of development; they usually appear after 1.5 to 2 years of age. Specific tests must be utilized to detect this condition (discussed 8–5 later).

Some children begin ambulating as "toe walkers," and it is important to determine if they have structural limitations to ankle dorsiflexion. This special gait may be due to spasticity (as in cerebral palsy) or shortening of the Achilles tendon. Often, however, toe walking is seen in otherwise normal children; it is unexplainable and usually disappears spontaneously.

By far the largest orthopedic concern of a new mother is that her infant has "flatfeet." The absence of a longitudinal arch and eversion of the hindfoot, one varying in direct proportion to the other, are commonly seen in loosely ligamented children. These children have normal-looking feet with full motion at all joints when not standing. Demonstration of unusual laxity at the knees, elbows, and wrists confirms the mechanism of the relaxed flatfoot. All joints should be examined for passive ranges of motion. A clinical estimation of tibial torsion and femoral anteversion should be done, as an abnormality here may explain why a child exhibits toeing-in or toeing-out (chapters 7–13).

EXAMINATION OF THE OLDER CHILD

Inspection of an appropriately disrobed child is necessary to determine trunk symmetry. Early evidence of scoliosis may be found in this age group by careful evaluation. The examiner stands behind the child. The position of the head and neck are noted in relation to the center of the pelvis. An actual or imaginary plumb line from the back of the head passes along the intergluteal cleft if proper centering is present (compensation); if the line falls to the right or left, loss of centering (loss of compensation) may be diagnosed. The level of the shoulders and scapulas are examined next. The flank creases are inspected and the space between them and the adjacent hanging upper limbs measured in order to estimate asymmetry. The child is then asked to bend forward so that posterior prominences of the dorsal and lumbar paravertebral areas may be noted. These are usually more obvious in the dorsal area where rib rotation adds to the prominence. Active trunk motion is determined for its range as well as its influence on improving or worsening any spinal asymmetry that may be present.

Palpation of the vertebral spinous processes sometimes helps to confirm a suspected spinal deviation. Pain is only occasionally provoked by such palpation. It is important to obtain radiographic confirmation of spinal asymmetry when it is first detected in order to have a base line for subsequent comparisons.

Evaluation of the child's general posture sometimes reveals the presence of "upper round back" (dorsal kyphosis) or "swayback" (increased lumbar lordosis). Occasionally the parent registers complaints of "poor posture" before the examination. It is important to determine, if possible, whether these spinal attitudes are fixed or spontaneously reversible. If a fixed spinal position is present, radiography is required to clarify the underlying status of the vertebral column.

The child's manner of walking is studied to determine if it is a smooth rhythmic progression or is interrupted by the presence of a limp. Again, the feet are examined for such local characteristics as presence of "arch," inversion or eversion of the hindfoot, and the presence of such deformities as bunions, hammertoes, or callosities.

With the child on an examing table, leg lengths are determined with a tape measure. fixed anatomical bony landmarks, usually the anterior superior iliac spine proximally and the medial malleolus of the ankle distally, are selected for this assessment. Circumferential measurements are taken at specific levels of the thighs and calves as indicators of muscle mass. Differences in such measurements indicate muscle atrophy from either disuse or disease and require explanation.

Ranges of motion are determined sequentially at all joints. Malalignments from bowing of the legs or knock-knees are noted. Tibial torsion is estimated and femoral anteversion suspected from collateral determinations.

3

The Spine

Primary abnormalities of the vertebral column may be derived from a variety of mechanisms, i.e., teratological, genetic, developmental: those occurring as malformations in the embryonic period often have neural concomitant involvement. Examples will be considered in the following discussion.

Secondary syndrome-associated deformities will be discussed with the specific entity, i.e., Hurler's disease and diastrophic dwarfism. Like long bones in general, the vertebral columns may be involved by infection, trauma, and neoplasia.

SPINA BIFIDA

Spina bifida (myelomeningocele) (Fig. 3.1) is a form of neural tube defect associated with failure of the posterior elements of adjacent vertebrae to fuse; there is usually protrusion of the spinal cord or its membranes.

Its incidence is 2 per 1,000 live births, with an unknown number among abortions, stillbirths, and early neonatal deaths. The sex ratio is male to female 1.5:1.

There is a familial relationship, with siblings affected; a higher proportion of concordance than discordance is seen in monozygotic twins, suggesting some genetic influence. There is also a higher incidence of neural tube malformations in the firstborn and with increased maternal age, suggesting intrauterine environmental influence.

Genetic counseling indicates that among normal parents with one affected child the risk to a subsequent child is 5%. When parents are normal and have two affected children, the risk to a third is 10%. Alpha fetoprotein

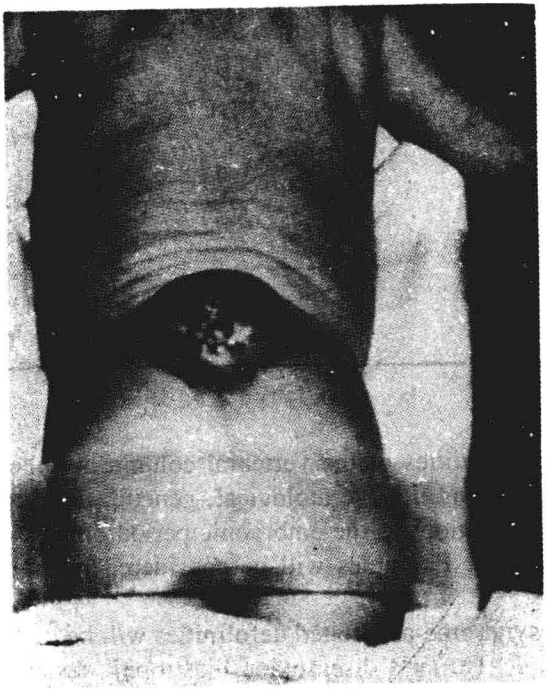

FIG. 3.1. Myelomeningocele is characterized by a midline protrusion of neural tissues through posteriorly deficient vertebral elements.

levels are elevated in amniotic fluid from women carrying fetuses with spina bifida. Hence amniocentesis offers a method of fetal screening.

Determination of maternal alpha-fetoprotein blood levels is being explored for its value as a first-line noninvasive screening mechanism.

The lesions occur at variable sites on the spinal column, but the lumbosacral region is most often affected. Lesions in the cervical spine seem to have the fewest neurologic sequelae, whereas those in the lumbar spine frequently carry the most severe consequences.

Spina bifida associated with a bulging mass may involve the skin, meninges, spinal cord, and vertebrae. The skin is commonly absent over the myelomeningocele prominence, leaving a fine translucent layer of arachnoid membrane as cover for the underlying conglomeration of cord and nerve roots. This fragile situation may be complicated by the development of overlying granulation tissue or necrosis with secondary infection. In all cases except the rare meningocele that contains no neural elements, some degree of spinal cord derangement is present in the form of failure of

differentiation, cystic changes, or degeneration. The vertebral deformity is lack of fusion or improper development of the laminae. There is associated widening of the spinal canal which can be identified radiographically as increased interpedicular distance.

Hydrocephalus commonly accompanies myelomeningocele and adds considerably to the morbidity of the disease. The additional clinical equivalents of spinal cord abnormality are muscle weakness at approximately the neurosegmental levels, impaired bladder and bowel control, and loss of sensation below the level of the disease process. Finally, there is a significant incidence of associated congenital anomalies including cardiovascular, gastrointestinal, and other skeletal malformations.

Successful management of such a complex disease process is twofold: (a) The immediate requirements for preservation of life are assessed; and (b) an integrated multidisciplinary approach to the associated anomalies is developed. Primary importance is given to closure of the myelomeningocele defect and ventricular shunting for control of the hydrocephalus. Secondary management includes establishing bowel and bladder control, a problem that assumes increasing importance as the infant reaches childhood. Finally, orthopedic measures are taken to preserve and enhance locomotor potential.

The level of cord involvement determines the degree of lower limb muscle weakness and imbalance. The potential for independent walking varies with the injury—ranging from total dependence on crutches and braces to total independence except for minimal splinting. Orthopedic therapy is begun with early maintenance of passive joint motions, and splinting is added later as the child approaches walking age.

The major orthopedic problems that usually require attention in the child with spina bifida are joint contractures, secondary acetabular dysplasia or hip dislocation, various joint instabilities, and secondary bony deformities. The vertebral column may also exhibit early lumbar kyphosis and subsequent paralytic scoliosis. If lumbar kyphosis is severe at birth, closure of the skin defect may be difficult, requiring immediate correction of the spinal angulation by resecting one or more vertebral bodies at the time of skin closure. Standard orthopedic techniques are used in the care of these patients, keeping in mind the need to protect against skin breakdown due to pressure because of loss of sensation.

IDIOPATHIC SCOLIOSIS

Idiopathic scoliosis, a form of spinal curvature (Fig. 3.2), consists of structural vertebral deformities with no known origins, in contrast to those