CHAPTER I

INTRODUCTION

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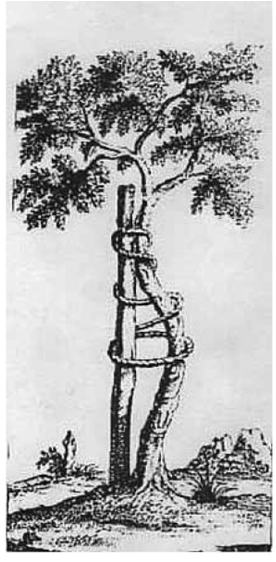
Orthopædic derives from *L'Orthopedie* (1741) by Nicholas Andry de Bois-Regard (1658–1742). It is derived from Greek ορθος: "straight" and παις (root παιδ–): "child." Andry was a Professor of Theology before becoming a Professor of Physick in the Faculty of Medicine at the University of Paris. The remainder of the title explains his purpose: "*Or, the art of correcting and preventing deformities in children: by such means, as may easily be put in practice by parents themselves, and all such as are employed in educating children*." The book emphasized simple remedies even a child's caretaker could administer, such as straightening by bracing [A]. Nowhere is surgery mentioned.



A Tree. The enduring symbol from an engraving on the frontispiece of *L'Orthopedie*. A straight stake is tied to a crooked sapling to partially correct it and guide its growth.

GROWTH

Growth distinguishes the child [B].

Joint

Joints may be fibrous, for example, syndesmosis and symphysis, or synovial, in which the skeletal elements are in contact but not in continuity: they are draped by hyaline cartilage of which the purpose is motion and not structure. Innervation of synovial joints is of two types.

- Myelinated group A fibers in the capsule detect joint position and motion.
- Unmyelinated group C fibers in blood vessels of the synovial membrane transmit pain. Joint injury or disease results in effusion, which stretches these fibers and hurts. The child accommodates by placing the joint in the position that maximizes volume and thereby reduces pressure, for example, flexion, lateral rotation, and abduction in an infected hip.

In children, a third type of articulation occurs, synchondrosis, during coalescence of ossification centers or bony segmentation: persistence may cause dysfunction, for example, tarsal coalition.

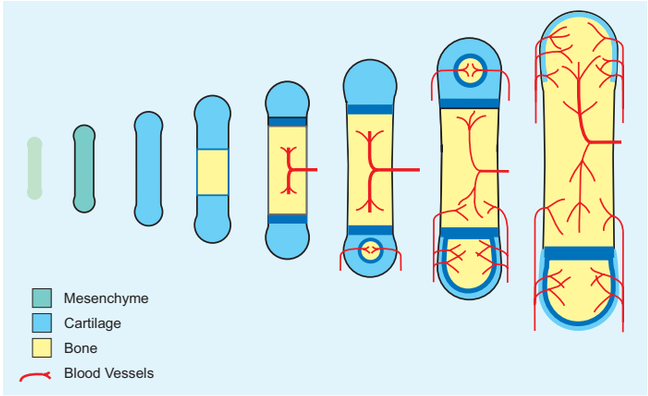
Joints develop first as a cleft in mesenchyme, which chondrifies and cavitates by the end of 3rd month. The process is regulated by the HOX family of genes. Joint development requires motion; hence, joint dysplasia in neuromuscular conditions characterized by akinesia or dyskinesia, such as arthrogryposis.

Bone

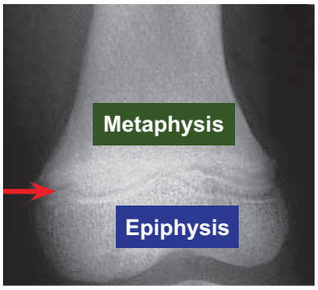
Timing of ossification influences care of the child. Imaging of hip dysplasia is facilitated by appearance of the proximal epiphysis of the femur, absent at birth. Patellar ossification after the 2nd year may delay diagnosis of its disorders. The multiple secondary ossification centers of the elbow appearing at different ages obfuscate the unfamiliar.

Stage	Definition	Comment
Early embryo	0–2 weeks	Before implantation
Embryo	2–8 weeks	Organogenesis Anomalies — problems of formation
Fetus	8 weeks to birth	Problems of growth
Infant	Birth to 2 years	Latin *in-* "not," *fari*: "to speak"
Child	2 years to puberty	Growth influences treatment Treatment influences growth
Adolescent	Transition to maturity	Childhood disease and treatment resemble adult

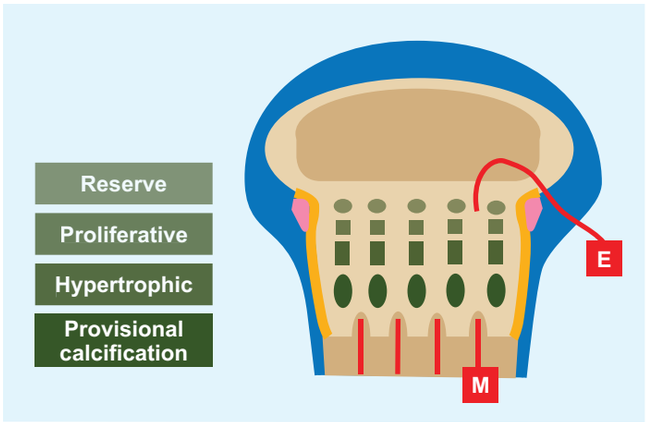
B Stages of growth.



C Endochondral ossification Mesenchyme turns into cartilage that turns into bone.



D Physis Röntgenogramme shows physis (*red*) interposed between metaphysis and epiphysis.



E Physis architecture E: epiphysial artery supplies the resting zone. Interruption of this may explain growth disturbance after fracture. M: metaphysial blood supply loops back acutely at the impervious physis, leading to sludging that increases likelihood of bacterial concentration and infection. The physis is supported by the ring of La Croix (*orange*), which is continuous between the periosteum at the metaphysis and the epiphysis and which is constricted by the groove of Ranvier (*pink*).

Intramembranous ossification Mesenchymal stem cells give rise to osteoblasts that secrete osteoid that mineralizes directly without an intervening cartilage model. In achondroplasia, this process is spared.

- Relatively normal size of clavicles results in broad shoulders; of fibulæ, it results in ankle varus; of skull, it results in a large head.
- Disordered endochondral ossification is seen as a "champagne pelvis," resulting from constriction at the triradiate cartilage amidst flat bones produced by intramembranous ossification.
- Constriction of the midface with unfettered surrounding osseous growth disrupts breathing.
- Constriction of the foramen magnum compresses the brainstem.

Intramembranous ossification also is the mechanism responsible for periosteal appositional growth (bone width) and fracture healing.

Endochondral ossification During the 6th week of gestation, mesenchymal stem cells differentiate into chondrocytes to form a model of the future skeleton. During the next week, periosteum forms, and by the 8th week, vascularization is under way [C]. Vascularization ushers in ossification and the fetal period. Secondary ossification of the epiphysis begins after birth, with the exception of the distal femur, which is present at birth. In between the primary and secondary ossification centers is interposed the physis.

Physis

This also is known as growth plate and epiphysial plate [D]. On one side is the secondary ossification center that sits "upon" it (Greek επι-), called epiphysis. The epiphysis forms the articular end of a bone: when alone, it is referred to as "head," when paired, it is known as "condyle." On the other side of the physis is the metaphysis (Greek μετα-: "beside," next to). Where it separates an ossification center that grows "away" from the main bone under traction from an attached muscle, this is known as an apophysis (Greek απο-: "away").

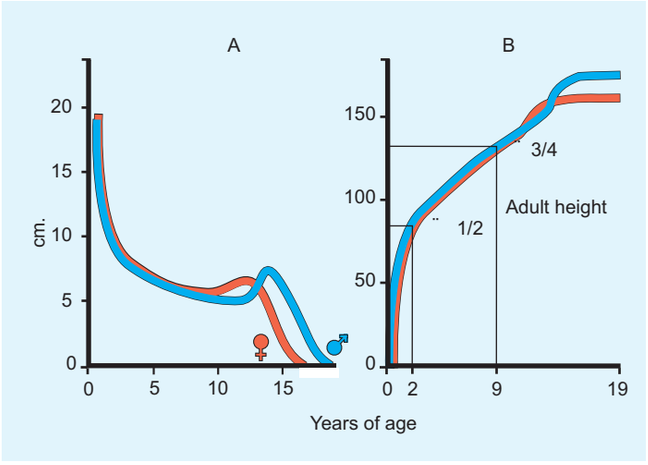
The physis has four zones [E]. In the resting zone, SOX-9 is coexpressed with COL2A1, its regulatory target, resulting in a high concentration of type II collagen. Proliferation is under the influence of several regulatory factors such as insulin-like growth factors and fibroblastic growth factors. Due to stress concentration between cartilage and bone, physial fracture occurs through the zone of provisional calcification, which may be distinguished from the hypertrophic zone. Injury to the reserve and proliferative zones, such as by a crushing mechanism (Salter-Harris V) or at an irregular physis with irregular shape (e.g., Poland hump at the distal tibia), will cause growth disturbance.

Longitudinal growth occurs by bone deposition at the metaphysis. After the 1st year of life, the physis becomes impervious to metaphysial vessels, which are turned back in sharp loops: this results in slowing of flow that concentrates bacteria and increases the likelihood of infection. The physis is surrounded by a ring of perichondrium (La Croix): thinning of this at the commencement of puberty may destabilize the physis in slipped capital femoral epiphysis. The perichondrial ring is constricted by a groove (Ranvier), which supplies physial chondrocytes and epiphysial osteoblasts.

Rate of Growth

The child is growing most rapidly at birth [F]. By 2 years, growth plateaus, increasing again at the pubertal growth acceleration. A child is half adult height by 2 years, and three-fourth by the end of the 1st decade. The head is disproportionately large at birth: accommodate for this when immobilizing a child with a suspect neck injury, so that the cervical spine may be neutralized and not flexed. The trunk grows earliest, while the lower limbs lag behind, achieving half of adult length by 4 years. The upper:lower body segment ration is 1 by maturity.

Growth may be influenced mechanically according to the Heuter-Volkmann law: compression of the physis retards growth, while distraction accelerates it. This may be harnessed for limb lengthening. Growth also is retarded physiologically by denervation, for example, hemiplegic cerebral palsy, and accelerated by hyperæmia, for example, congenital vascular malformation.



F Growth Rate A: absolute rate. B: variation of height with age.

PRINCIPLES OF DISEASE

Definition and classification aid understanding.

Syndrome A group of anomalies that present in a predictable manner and due to a single cause.

Association A group of anomalies that occur together but are not related by a common cause.

Malformation This refers to an abnormality that arises during the period of organogenesis. There are five types [A].

Dysplasia This is used for abnormal structure due to an intrinsic tissue defect, for example, diastrophic dysplasia, or for abnormal shape due to an extrinsic cause, for example, of the hip

Disruption Abnormal event occurs late in gestation after normal growth and development established, for example, amniotic band syndrome.

Deformation Extrinsic factors produce disease superimposed upon a normal part, for example, idiopathic scoliosis.

Sequence A group of anomalies that arise downstream from a single initial event.

Field defect A group of anomalies that are geographically linked or restricted.

Diseases affecting the musculoskeletal system have been variably classified by the salient clinical feature, for example, Friedreich ataxia; geographically, for example, nail–patella syndrome; by röntgenographic appearance, for example, chondrodysplasia punctata; by pathogenesis, for example, osteogenesis imperfecta; by eponym, of which countless examples exist. We are evolving toward a fundamental and unifying approach based upon the molecular basis of disease [B].

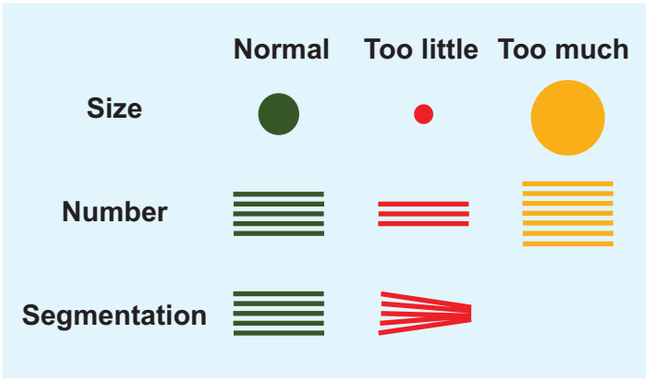
Structural gene The product has a mechanical function. Osteogenesis imperfecta is caused by a mutation in type I collagen, which is the principal collagen supporting the extracellular matrix of bone.

Tumor regulatory gene The product stimulates cell growth and differentiation or prevents cell death, affecting the cell cycle. Neurofibromatosis type I is caused by a mutation in neurofibromin, which is a tumor suppressor inhibiting p21 ras oncoprotein.

Developmental gene The product is involved in cell development or patterning. Nail–patella syndrome is caused by a mutation in LMX1B, which plays a rôle in dorsoventral patterning of the vertebrate limb.

Nerve and muscle function gene Mutation in dystrophin causes muscular dystrophy.

Protein processing gene The product is an enzyme, of which mutation results in accumulation of precursors or abnormal products. The quintessential example is mucopolysaccharidoses, which are characterized by intracellular accumulation and urinary excretion of mucopolysaccharides due to deficiency in degradative lysosomal enzymes.



A Malformation types. This simplified method includes too little, too much, too few, too many, and failure of segmentation.

Gene	Examples	Clinical features
Structural	Marfan syndrome Osteogenesis imperfecta	Phenotypes change with time as structure wears out, superimposing degenerative changes on congenital abnormalities
Tumor regulatory	Multiple exostosis Neurofibromatosis	Overgrowth of cells and tissue type in which gene is expressed. Malignant potential
Developmental	Achondroplasia Cleidocranial dysplasia Nail–patella syndrome	Abnormal embryonic development results in malformation present at birth
Nerve or muscle	Charcot-Marie Tooth Duchenne syndrome Rett Spinal muscular atrophy Friedreich ataxia	Musculoskeletal system normal at birth, develops abnormalities secondary to neuromuscular imbalance Muscle and central nervous disorders more severe for example, shortened life-span, than peripheral nervous disorder
Protein processing	Gaucher syndrome Mucopolysaccharidosis Osteopetrosis	Enzyme replacement may alter natural history
Chromosomal defect	Down syndrome Turner syndrome	Multiple genes affected, resulting in multiple abnormalities in multiple systems

B Classification by type of gene mutation (Alman) An attempt to bring order to a disparate group of conditions.

Distinguishing features of pædiatric orthopædic surgery
The original
Horizontal
Growth
More nonoperative care
More casts
More diseases
More multidisciplinary
Less long-term follow-up
Better surgical outcomes in the uninvolved child
Parents

A Distinctive features of pædiatric orthopædic surgery.

Chromosomal defect This affects multiple genes on a whole, such as trisomy 21, or part of a chromosome.

Not all mutations can be classified. Some products can be included in more than one category. How a given mutation exerts its effect is understood in some and unclear in others.

CARE OF THE CHILD

Pædiatric orthopædic surgery is a horizontal discipline [A]. Its domain is the child as a whole, traveling across the broad landscape of the body and navigating disparate diseases within a defined age echelon. This contrasts with the direction of our discipline as a whole, where specialization has turned it vertical, where the surgeon is focused on a single system or part "from cradle to grave." Delicate is the balance between caring for the child as a whole, interconnected person and developing sufficient skill and experience to care most optimally for the affected part.

The approach to a child, and to the family, is unique in many ways.

- Sit down, at eye level with a small human being (child), and take some time to ease anxiety brought on to a child by an authority figure.
- Minimize exposure. Uncover only the part necessary to examine, and limit traffic, to reduce the sense of vulnerability in the child.
- Retain the parents, whose presence will reassure the child and whose input is essential when a child cannot or will not speak.
- Close the door so that the child feels safe.
- Focus on the child, even when the parent is giving the history. Talk to the child first and last.
- Ask children to come in sports clothes or a bathing suit. A gown is a foreign object.
- Listen to the child. While parents advocate and consent for their children, their concerns and perceptions may differ from their children's. Additionally, children often bring a simple clarity to complex decision making that eludes their parents.
- Take every and all opportunities to examine the child. Watch the child walk into the office (gait, affect, pain, stature, proportion). Assess how well a child who complains of pain moves about in the room, or surmounts the examining table. How does the child who is supine use or rest the limbs? Ask the child to walk on the heels and toes, perform a deep knee bend, and walk "like a duck," thereby testing strength against body weight and doing so without touching the child.
- Start by checking the normal side, or perform a screening examination, so that the child is assured you are not there to inflict more pain.
- Consider as an examining table using a parent's lap, which will comfort and calm a baby.
- Choose your words carefully. The language of a 12-year-old is simple and direct, not technical and not convoluted.
- Involve the child in decision making; sequestration engenders suspicion.
- Avoid unnecessary or excessive treatment, which will invade childhood and may expose the child to risk.
- Weigh hope and a desire for parental agency against "medicalization" (devices, therapies, consultants, new treatments) of a handicapped child.
- Enlist the parents. They are an ally who will care for the child at home, who will comply with instructions, and who will ensure scheduled follow-up. Their vigilance is normal and in the interest of the child.
- Pay attention to a parent's intuition. They know the child better than you do.
- Acknowledge and accept that often the most benign presentation will be the most time consuming. It takes time and patience to educate and convince parents of normal variants such as flexible flat foot and in-toeing, which is the most reason for consultation of a pædiatric orthopædic surgeon.
- Measure childhood in dog years. Every year in a child's life represents a big change, physiologically, emotionally, socially. How much does

treatment of a dislocated hip change between 6 months of age (soft tissue) and 18 months of age (bone)? A child's rapid evolution presents many children within the same childhood. How different is the same child at 10 years of age and 2 years later during puberty?

- Observation is a form of active management. It is not neglect, benign or otherwise. It admits that sometimes the entire picture may not be revealed at an initial encounter. It allows time for a family to come to grips with a diagnosis and elective treatment options when these are serious and to become comfortable with their child's surgeon. It gives natural history, which is very dynamic in a growing child, some more time to declare itself. Insisting on finding out an answer by evaluating further may not be benign, for example, an MRI may require an anæsthetic. Remember the English poet and statesman John Milton: "They also serve who only stand and wait" (*On His Blindness*, 1655).