Rotational Deformities of The Lower limbs in Children

Essay

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Abstract

Torsion abnormalities frequently demonstrates a familial tendency, therefore, one should question and even examine family members of patients in whom torsional malalignment is of concern, examination of family members may actually predict the natural history of the child and illustrate the frequency of functional problems related to mild torsional malalignment, intrauterine positioning may affect rotational malalignment and is believed to be responsible for some cases of medial tibial torsion and metatarsal adductus.

Key word

Deformities the lower limbs rotational

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INTRODUCTION

Rotational deformities of the lower limbs in children are the most common problems seen by the orthopedist in the pediatric age group. Although this is a relatively benign problem, a lack of understanding of the pathology and the prognosis often creates confusion in the minds of parents [(1)].

Considerable controversy exists among orthopedists regarding the natural history, disability and management of these conditions. A clear understanding of the deformity and its natural history is important to care for children with these conditions since they may correct spontaneously [(2)].

In the past, rotational problems in the transverse plane have often been ignored because they were difficult to diagnose. This is in contrast to frontal plane deformities (valgus or varus deformities) and sagittal plane deformities (flexion contracture or hyperextension), which are easily visualized and measured on conventional radiographs. These difficulties in assessment have lead to varied opinions and controversy regarding the evaluation and management of rotational deformities [(3)].

Abnormal lower limb torsion may present as in-toeing or out-toeing gait, this essay aims to disclose this problem, identifying its nature and etiology, discussing the diagnosis of its various types, causes and the accurate evaluation, analysis of such deformities and their resultant disabilities. It also discusses the prognosis and management of such conditions and how to tackle them wisely and effectively with new lines of management [(2)].

NATURAL HISTORY OF LIMB DEVELOPMENT

1- Intrauterine period:

At 28 days after fertilization, the lower limb elevation appears just caudal to the level of the umbilical cord, and develops similarly, but slightly later than the upper limb[4].

The limb bud initially consists of loose mesenchymal tissue enclosed in an epithelial ectodermal sheath; the limb bud is formed from mesenchymal cells of the lateral plate, and then augmented by cells from the adjacent somites. The skeletal elements and tendons develop from the lateral plate mesenchyme, whereas limb muscles arise from somitic mesenchymal cells that migrate into the limb bud. This mesenchymal swelling is covered by ectoderm, the tip of which thickens and becomes the apical ectodermal ridge (AER), (Fig. 1) [4].

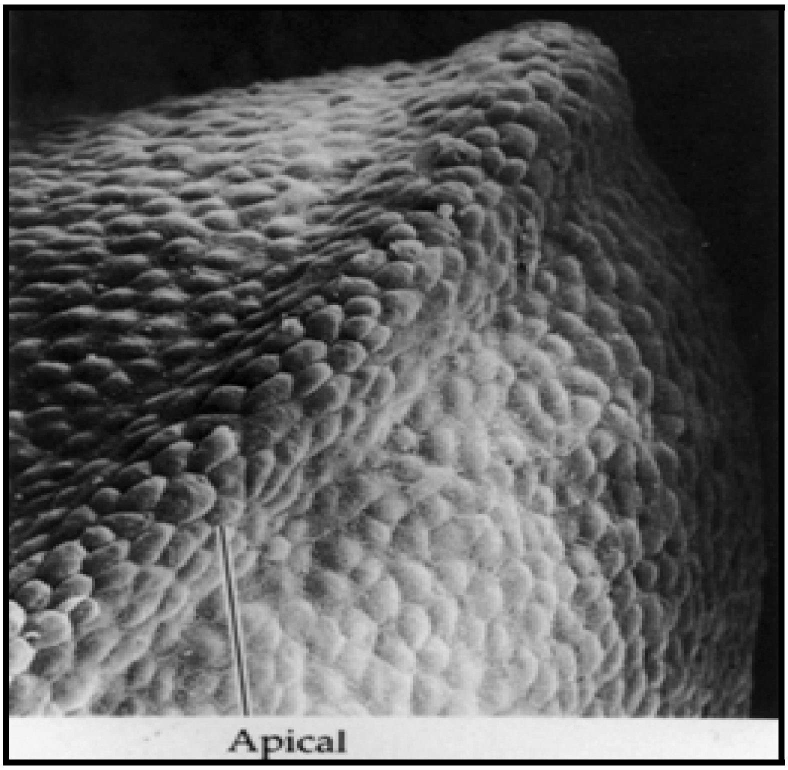

Fig. 1: **Scanning electron photomicrograph of an early chick forelimb bud, with the apical ectodermal ridge at the tip of the limb bud [4].**

Underlying the AER are rapidly proliferating, undifferentiated mesenchymal cells that are called the progress zone (PZ). Proliferation of these cells causes limb outgrowth. Cells begin to differentiate only after leaving the PZ. The interaction between the AER and the undifferentiated mesenchymal cells underlying it is crucial for limb development. Experimental procedures on chick embryos reveal the following about the limb bud mesenchyme:

1- If removed, no limb develops;

2- When grafted under the ectoderm at a location other than the normal limb area, an AER is induced and a limb will develop;

3- Lower limb mesoderm will induce leg formation when placed under an upper limb AER.

Grafting experiments with the AER reveal that:

1- AER removal aborts further limb development, the later in limb development the AER is removed, the less severe is the resulting limb truncation (limb elements develop from proximal to distal);

2- An extra AER will induce a limb bud to form supernumerary limb structures;

3- Nonlimb mesenchymal cells placed beneath the AER will not result in limb development, and the AER withers (4).

The implications of these experiments are that; the AER is necessary for the growth and development of the limb, whereas the limb bud mesenchyme induces, sustains, and instructs the AER. In addition to biochemical influence on the PZ, the tightly packed columnar cells of the AER perform a mechanical function, directing limb shape by containing these undifferentiated cells in a dorsoventrally flattened shape. The length of the AER controls the width of the limb, as well, when all limb elements have differentiated, the AER disappears (4).

The anteroposterior (AP) axis determination is under the control of an area of tissue in the posterior aspect of the limb bud called the zone of polarizing activity (ZPA), or polarizing region. If this tissue is grafted onto the anterior aspect of a limb bud, a duplication of digits in a mirror image to the normally present digits occurs. Cells for the new digits are recruited from the underlying mesoderm, and the distal part of the limb widens, as does the AER. If less tissue from the polarizing region is grafted, fewer new digits develop. This and other experiments suggest that a morphogenic gradient of a diffusible signal originating from the ZPA determines AP axis (4).

The dorsoventral (DV) axis is under the control of both the mesoderm and ectoderm of the limb bud at different stages of development. The mesoderm specifies the axis initially, but, very early after limb bud formation, the ectodermal orientation becomes eminent. If the ectoderm of a right limb bud is transplanted onto the mesenchyme of a left limb bud, the distal limb that develops will be that of a right limb, with respect to muscle pattern and joint orientation (5).

The proximodistal (PD) axis seems to be determined by the length of time a mesodermal cell remains at the tip of the limb bud in the progress zone under the influence of the AER. Once a cell leaves the tip, its position in the limb is fixed. Young tips grafted onto older limb buds will duplicate existing limb elements, whereas older tips grafted on young buds will only form distal elements. The best hypothesis as to how this information is passed is that the number of rounds of cell division that occur while under the influence of the AER determines the PD fate of a cell. Support for this hypothesis comes from experiments in which the limb bud is irradiated. The surviving cells of the irradiated tip have to undergo several extra rounds of mitosis before they can escape the influence of the AER and thereby gain positional determination. In these experiments, intermediate limb elements are not formed only the preexisting proximal elements and newly formed distal elements (5).

Cellular differentiation of the homogenous, undifferentiated-appearing mesenchymal cells in the limb bud results from signals different than those conveying the axis/positional information as described above. The center of the limb bud develops a condensation of cells that prefigures the skeletal elements the chondrogenic core, which begins at the body wall and progresses distally with limb elongation. A rich vascular bed surrounds the chondrogenic core, immediately adjacent to the vascular bed is a thick avascular zone that extends to the ectodermal sheath of the limb bud. Although the signaling mechanism has not been discovered, the ectoderm appears to control initial mesodermal differentiation by maintaining the adjacent mesenchymal cells in a flattened configuration, which prevents differentiation into chondrogenic cells. The central mesenchymal cells assume a rounded shape and form the chondrogenic core [(4)].

This process of differentiation occurs from proximal to distal, early in the 8th week, cartilage anlage of the entire lower limb skeletal elements, with the exception of the distal phalanges, is present, foot plates have been formed by the end of the 7th week, and condensations of cells have formed identifiable digital rays in the foot. The cells between the digital rays are a loose mesenchyme that undergoes programmed cell death (apoptosis) to create the separate toes [(4).]

During the embryonic period, all four limbs are similar, with parallel axes. The preaxial borders are cephalad and the postaxial borders are caudad. The thumb and hallux are preaxial; the radius/tibia and ulna/fibula are homologous bones occupying the same positions in the limb bud. The longitudinal axis at this stage passes through the long finger and the second toe. During the fetal period, the upper limb rotates 90 degrees externally (laterally), and the lower limb rotates 90 degrees internally (medially). The forearm flexors come to lie ventrally and the forearm extensors, dorsally. The leg extensors lie ventrally, and the leg flexors lie dorsally (Fig. 2, 3). [(5)]

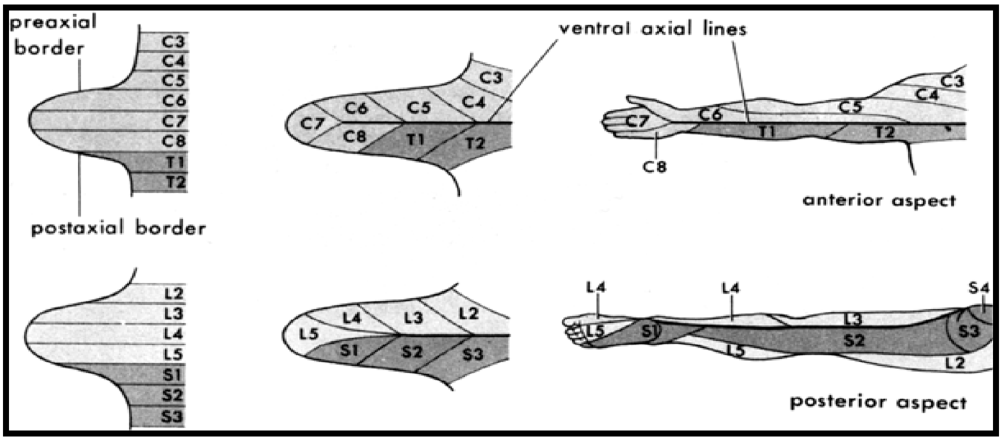

Fig. 2: **Development of the dermatome pattern in the limb. A and D: diagram of the segmental arrangement of dermatomes in the 5th embryonic week. B and E: the pattern is shown one week later as the limb bud grows. C and F: the mature dermatome pattern is shown. The original ventral surface becomes posterior in the mature leg and anterior in the mature arm, due to the normal rotation of the limbs** [5].

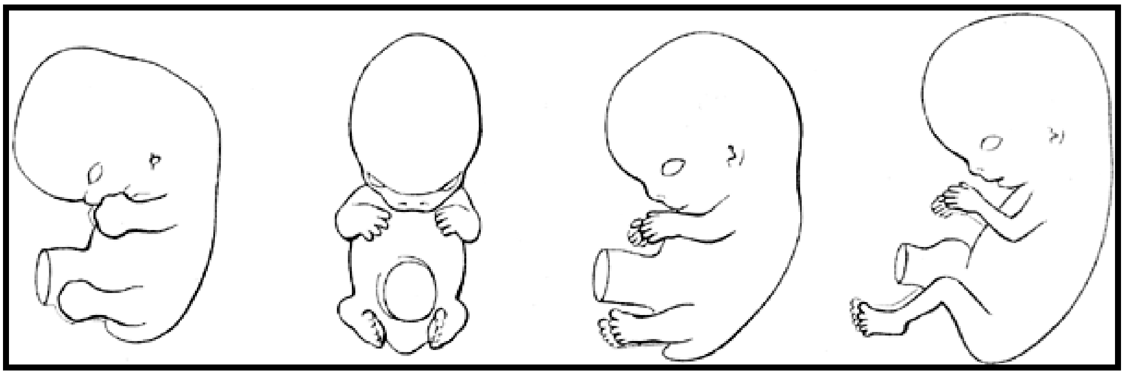

Fig. 3: **Normal limb rotation is depicted. A: 48 days, the hand and foot plates face each other. B: 51 days, elbows are bent laterally. C: 54 days, the soles of the feet face each other. D: lateral rotation of the arms and medial rotation of the legs result in caudally facing elbows and cranially facing knees.** [5]

Thus, by the 8th week, the task of tissue differentiation is largely completed and growth is the major task ahead [5].

Fetal position in uterus strongly affects the limb rotation (Fig. 4), during the late months of pregnancy; developmental changes are associated with increasing anterior inclination of the acetabulum which continues after birth. The tibia rotates laterally, anteversion of the femoral neck is about 5 degrees in the 3rd month of intrauterine life, and it increases to about 15 degrees in the 4th month, reaching up to 40 degrees at birth [6].

Fig. 4: **Intra uterine position affects the rotation of the lower limbs**. [6]

2- Limb rotation after birth:

The intrauterine position in the last months of pregnancy determines the position of the lower extremities at birth. They usually lie in external rotation with the hips and the knees flexed. (Fig. 4) [6].

If the hips are extended, external rotation is greater than internal rotation; this is due to soft tissue contracture. Just after birth there is about 40 degrees of femoral anteversion, the tibia is rotated internally between 5-30 degrees and associated with mild lateral bowing, and also, the feet are turned inwards. But, these attitudes undergo spontaneous correction in the course of development as long as extrinsic factors don't interfere [7].

Later on, anteversion decreases during childhood to a value between 5-16 degrees in adults [8] (Table: 1).

Age	Mean values
1 year	39 degrees
2 years	31 degrees
10 years	24 degrees
16 years	16 degrees

Table 1: Showing the angle of femoral anteversion decreasing with age. [8]

The largest amount of anteversion correction occurs between the first and second year of life, with another spurt occurring between 14 and 16 years of age [9].

An older child with excessive femoral anteversion usually has increased internal rotation of the hip and limited external rotation. however, during the first two years of life, when femoral anteversion is decreasing spontaneously, the internal rotation of the hip also decreases, but other structural components are important in determining the rotation of the hip e.g.: soft tissue adaptation to intrauterine position, hip contractures, acetabular shape as well as inclination and relative position of the pelvis and acetabulum to the femoral neck and head may also be factors affecting hip rotation [10].

The development of adult femoral anteversion angle (nearly 16 degrees) is the outcome of several factors including erect posture of the human being which depends on weight bearing and forces exerted by the glutei muscles (gluteus medius& minimus). Thus, in certain pathological conditions as congenital dislocation of the hip (CDH) and in paralysis of the gluteal muscles, the neck-shaft angle remains valgoid as in infants. Tibial torsion also changes, so that; there is about 20 degrees of lateral tibial torsion at the end of the growth period [11].