ANGULAR KNEE DEFORMITIES IN CHILDREN - A REVIEW

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Introduction

Angular knee deformities are a common sight when one watches a large group of playing children. This 'picture' illustrates what is probably the most important issue, which is that the parents often are more disturbed about the deformity than the kids and usually these deformities resolve with time i.e. by far the commonest deformity is the physiologic type.

The physician treating these patients should remember to reassure the parents and carry them along. Their greatest fears are usually that their child will need surgery and that a required procedure is delayed. These paradoxical fears make these parents ready victims in any environment, of charlatans and traditional bone setters. Luckily, most cases resolve without direct intervention.

Historical background

The treatment of these patients helped define the specialty of orthopaedic surgery, as orthon from Greek means “straight, free from deformity” and paidion means “child”.

Professor Nicolas Andry (1658 to 1742) who introduced the term “orthopaedic” through his book “L'Orthopedie”, recommended treatment of a “crooked” leg in a child, by fastening the limb to an iron plate on the hollow side of the leg and then tightening the plate against the leg daily until the leg became straight[1]. His illustration of a similar method to straighten “the crooked trunk of a young tree” by tying it to a stake became the symbol for the specialty of orthopaedics.

Böhm, in 1933, was the first to describe the normal physiologic evolution of the lower limbs[2].

Measurement of angular knee deformities

Terms used in describing angular knee deformities, denote the position of the distal segment of the deformity, relative to the proximal segment. Varus indicates an angulation towards the midline of the body, distal to the part named; while Valgus indicates angulation away from the midline, distal to the part named.

In angular knee deformities, both measurement in degrees and linear measurement are used. The commonest method is the linear measurement, with the patient standing, if this position can be maintained unsupported by the patient. Infants have their knee angles measured in the supine position, before they start standing. In genu valgum, the distance between the medial malleoli is measured with the knees in full extension, patellae facing exactly forward, and the medial condyles of the femur brought together with moderate firm pressure, to compress excessive subcutaneous fat. The distance between the medial femoral condyles is measured in genu varum, with the medial malleoli brought together firmly. The patellae must face exactly forward, as medial rotation of the lower limbs at the hips will cause apparent bowlegs. Another method used to measure the degree of deformity is to determine the angle between the lateral surface of the thigh and leg (lateral thigh-leg angle), using a goniometer.

Natural history of knee angles

Vankka and Salenius[3] studied the development of the tibiofemoral angle by clinical and radiographic measurement in 1,480 normal children, and are credited with defining the physiologic course of these conditions. In the newborn and the infant up to one year of age, the tibiofemoral angle was in marked varus, with a medial deviation of 15 degrees. The tibiofemoral angle straightened to 0 degrees at about one and a half years of age. Subsequently, up to the third year, there was progressive valgus deviation of the tibiofemoral angle, reaching 12 degrees. By age 7 years, the valgus alignment gradually corrected to that of the adult angles of 7 degrees in the male and 8 degrees in the female. A review paper by Bruce RW[4]

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noted a similar course of knee angle development. The findings were similar in boys and girls. Omololu B. et al[5] studied the knee angles in 2166 normal children in southwestern Nigeria, in the age range of 1 to 10 years. They noted that the maximum varus angle was at ages 1 - 3 years and this reduced to neutral or 0° at the age of 5 years in girls and age 7 years in males. The maximum valgus angle in their series was between the ages of 7 and 9 years. They also noted that gender was not a significant variable in knee angle evolution.

Physiologic knee angular deformity

The physiologic deformities of the knee angle are the commonest type of knee deformity seen in all the papers reviewed and my experience is no exception to this. The wide space between the knees, the rolling gait and toeing-in, which is due to an associated medial tibial torsion, are the points of concern to most parents of children with genu varum. Less common is marked genu valgum, in which the child walks awkwardly swinging one leg around the other, to avoid banging his knees, with pronated feet. In severe cases of genu valgum, because of the malalignment of the quadriceps mechanism, the patellae may subluxate laterally.

Evaluation

The initial objective is to exclude a pathological cause for the angular deformity. The history should note the nature, age of onset, progression, prior treatment and response to treatment of this deformity. History of rapid progression suggests a pathologic condition. The family history of knee deformity helps in detecting familial cases. Overall growth and development of the child may be a pointer to patients with skeletal dysplasia. Is there a history of direct sunlight deprivation, trauma, milk allergy or special dietary habits? The wavelength of ultraviolet waves in sunlight required for conversion of ergosterol in skin to 7-dehydrocholesterol is in the 296-310nm range and it is impeded by ordinary window glass[6].

On examination, the height and weight are noted for percentile calculations. The alignment of the lower limbs is inspected in stance and gait. In stance, measure the deformity as earlier described. The patellae must face straight forward. Note where the center of gravity falls relative to the center of the foot, which is the second ray, when projected downward from the anterior superior iliac spine. Normally it passes between the first and second metatarsals, but in genu varum it shifts laterally between the third to fifth metatarsals. Note the foot position. It may be normal, pronated or in postural metatarsus varus. Associated deformities should be sought. For example, absent or hypoplastic lateral rays of the foot and skin dimpling may suggest fibular hemimelia. The degree of tibial torsion should be measured. The stability of the knee collateral ligaments and the cruciate ligaments should be assessed. During gait, measure the foot progression angle (FPA). This enables a diagnosis to be established and provides a baseline for future comparison. In the stance phase of gait, observe for the presence of medial or lateral thrust at the knee, which indicates medial or lateral collateral ligament incompetence respectively, and a pathologic etiology. It also indicates a high risk for progression of the deformity and must be taken into account during the repair. Furthermore a rotational profile should be done i.e. in the prone position, assess medial and lateral hip rotation in knee flexion, and the thigh-foot angle (TFA).

The place and use of x-rays

An x-ray film of the knee is sometimes recommended by the referring doctor, but is not necessary in all cases. It is indicated when a pathologic condition is suspected, as in cases that have one or more of the following: (1) stature below the 5th percentile (2) significant asymmetry between the two sides (3) severe deformity (4) failure to follow a normal developmental sequence (5) rapid progression of the deformity (6) family history of pathologic conditions (7) associated clinical abnormalities. A single anteroposterior radiograph of the entire lower extremity is adequate. The mechanical axis of the knee should be measured and the deformity localized. In my practice I focus mainly on defining the knee

deformity, as the size of the x-ray film is usually too small to include the entire lower extremity. Does the deformity involve mainly the femur or tibia or both?

In trauma and osteogenesis imperfecta, the radiographic abnormality is in the diaphysis, while it is in the physeal plate in rickets and in the epiphysis in Blount's disease. In the bone dysplasia's the radiographic features could be found in any or all the zones from epiphysis to metaphysis.

The metaphyseal-diaphyseal (MD) angle should be determined. In physiologic genu varum it is less than 11 degrees, but more than 11 degrees in Blount's disease (tibia vara).

Radiologic findings in physiologic genu varum are: (1) the transverse planes of the knee and ankle joints are tilted medially (2) the tibia is angulated medially at the junction of its proximal and middle thirds, and the femur at its distal third (3) the medial cortices of the tibia and femur are thickened and sclerosed (4) the epiphyses, physis and metaphysis have a normal appearance, and there is no evidence of intrinsic bone disease (5) involvement is usually bilateral and symmetrical. Physiologic genu valgum has similar radiographic features in reverse.

Differential diagnosis

These are pathological causes of knee angular deformity. The commonest of these in Nigeria, is rickets[7,8]. In the paper by Solagberu BA[7], all the subjects had nutritional rickets and none was associated with the Islamic practice of purdah restricting the wife to the inner compartment of the house, which tends to keep her young children indoors and hence deprived of direct sunlight. Salawu SAI[9] noted a rickets rate of 70% in northern Nigeria, where the practice of purdah is common, but he did not relate this high incidence to purdah. The diagnosis of rickets is usually clinical, supported by x-ray findings. Jackson[10] had noted that serum chemistry is unnecessary in the management of these patients, which was confirmed by Solagberu[7]. Clinical features include a varus or valgus knee angular deformity, wrist bulge and swollen costochondral junctions of the rib. The x-ray findings are physeal widening; splaying, cupping and fraying of the juxtaphyseal metaphysis and cortical thinning, which gives the "inverted" Champaign-glass appearance on an x-ray of the distal femur. Tibia vara, Blount's disease, is the most common cause of pathologic varus deformity of the leg in children in developed countries, where nutritional rickets has largely been eliminated[11]. The condition affects mainly those of negroid descent and is bilateral in 80% of cases[12]. Its features are a steep medial angulation in the medial cortex of the tibia in its proximal metaphyseal region, while the lateral cortex remains nearly straight. The femur is normal, except late in the course of the disease. The MD angle is greater than 11 degrees. There may be internal rotation of the tibia. The pathology is a growth disorder of the posteromedial part of the proximal tibial physis, epiphysis and metaphysis. Erlacher reported the first case of tibia vara in 1922[13].

W.P. Blount, in 1937, presented 13 new cases and reviewed the 15 cases in the literature; he delineated the similarities between infantile and adolescent tibia vara and emphasized the differences in their etiology[14]. He coined the term tibia vara, which is used descriptively. However, the eponym Blount's disease is more commonly used. The term osteochondrosis deformans tibiae has been abandoned, as no avascular necrosis is present. Langenskiöld, in 1952, classified tibia vara into six progressive radiographic stages[15].

Other differential diagnosis is as listed:

Metabolic bone disease

Rickets

Renal disease

Asymmetrical growth arrest or retardation

Tibia vara (Blount's disease)

Trauma

Infection

Tumour

Bone dysplasia

Metaphyseal dysplasia, camptomelic dwarfism,

achondroplasia, enchondromatosis
Multiple epiphyseal dysplasia, chondrometaphyseal dysplasia

Tumour
Fibrous dysplasia
Multiple hereditary osteochondromata

Congenital
Congenital pseudoarthrosis of the tibia
Congenital posteromedial bow
Fibular hemimelia
Congenital tibia vara angulation at middle third of tibia, often positive family history, prognosis for spontaneous cure is guarded

Metal intoxication
Flourosis excessive fluoride intake during pregnancy, may result in severe bowlegs in infancy

Osteogenesis imperfecta

Treatment
Physiologic genu varum will nearly always resolve with weight-bearing and skeletal growth. The parents should be informed of the natural history, including the temporary knock-knees at about three years which corrects spontaneously by seven years. A six monthly follow-up for reassessment, will afford the opportunity to determine the course of events and allay the parents anxiety. The use of orthosis is contraindicated.

Physiologic genu valgum resolves spontaneously in 95% of cases, for the two to six year age group[16]. Six monthly follow-up is also advised. The patients who have significant valgus by the age of ten years and those whose deformity becomes excessive in adolescence, may benefit from surgical correction.

The surgical options available are stapling of the physis or hemiepiphysiodesis (fusion of one half of the growth plate) on convex side of the deformity, realignment osteotomy or callous distraction (Ilizarov method). The osteotomy is usually done in the distal femur for valgus deformity and the proximal tibia for varus.

Treatment of Blount's disease is usually some form of realignment osteotomy in the proximal tibia, with correction of both the angular and rotational deformities. Slight overcorrection, about 5-10 degrees should be aimed for, as some recurrence is inevitable. During skeletal growth the physeal plate should be protected and when growth is completed, a high tibial osteotomy may be done, being careful not to cause an intra-articular fracture. In severe cases, it may be necessary to elevate the depressed medial tibial plateau or excise a bridging bony bar if one has formed (Langenskiöld Stage VI) and replace it with a free fat graft. Corrective osteotomies in the proximal tibia should be accompanied by fasciotomy, to reduce the risk of a post-operative compartment syndrome. Skeletal stabilization can be by splintage using either a cast alone or external fixator, or fixation with a plate or crossed k-wires. Internal fixation still requires protection with a POP cast. I usually use a POP cast alone for children and a plate for adolescents / adults. Schoenecker et al in a retrospective study found that one osteotomy before five years of age achieved complete and permanent correction of the deformity in 83% of the children[17]. In patients with unilateral disease and significant limb length discrepancy(LLD) (4cm or more), tibial lengthening is advised but should be done in centers that specialize in the procedure, as it is fraught with problems and complications. For LLD of 1 to 2cm, an open wedge osteotomy usually suffices, with bone graft in the gap and preferably stabilized with a plate.

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