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Non Standard Deviation - Managing angular deformities around the knee in young age

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Abstract

Angular deformities around the knee in children are common presentations in day to day practice. While some of the deformities are physiological that need careful observation, the others are pathological and need timely medical or surgical intervention. Systematic evaluation of the deformity is the first step in making the correct decision about the management. The right choice of treatment, the correct timing and perfect execution of the technique are necessary for optimum outcome. This article describes the aetiology, clinicoradiological evaluation and management of angular deformities around the knee in children.

Keywords: Knee, Angular deformity, Children

Introduction

Angular deformities around the knee, known as genu varum (bow legs) or genu valgum (knock knees) are common during childhood and are usually a cause of serious concerns for the parents. Many of these deformities are physiological and resolve spontaneously. But some of these deformities may develop from various clinical conditions and may lead to cosmetic problem, gait disturbance, joint instability and activity related pain. If untreated, they can also lead to early degenerative arthritis due to abnormal joint overload. In order to avoid these future consequences of genu varum and valgum in children, accurate assessment of the angular deformities and timely appropriate intervention is essential. The first step in approaching a child with angular deformity around the knee is to differentiate between physiological and pathological deformity and for this, one must understand the normal development of knee angle (Femoro tibial angle-FTA) in children. [1]

Normal development of Knee angle and Normal alignment of lower limb

Selenius and Vankaa have described the physiological development of the FTA in children. There is varus alignment of 10-15^0 in the newborn. By 18-20 months, the alignment becomes neutral. From 3 years onwards, there is progressive valgus alignment upto 8-10^0 and by approximately 7-8 years, the valgus decreases to mature FTA of around 6-8^0. (Fig. 1)

Hence, persistence of genu varum beyond 3 years and valgus deformity >12-15^0 beyond 8-10 years may be considered abnormal and rarely corrects itself with growth. [2]

The mechanical axis of the lower limb is the line connecting the centre of femur head and the midpoint of tibiotalar joint. Normal mechanical axis passes through or immediately medial (8 +/- 7 mm) to the centre of the knee joint line. Genu varum is defined as an increased FTA with the mechanical axis passing through the medial portion of the knee joint. Genu valgum is defined as a reduced FTA with the mechanical axis passing through the lateral portion of the knee joint.[2]

Assessment of a child with angular deformity

History:

A child with genu varum or genu valgum usually presents with the parental concern of cosmetic deformity. Some of the children may have anterior knee pain or discomfort, gait disturbance or instability symptoms.

A careful medical history is important to differentiate between physiological and pathological angular deformity. The age of onset and progress of the deformity should be noted. A family history of short stature, varus or valgus alignment and genetic disorders need to be ascertained. Dietary history and history of milk allergy may require further investigations

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Various causes of angular deformities around the knee	
Genu Varum	**Genu Valgum**
Physiological **Bow legs**	**Physiological** **Knock – Knees** **Apparent genu valgum (Fat thighs, rotational deformity)**
Pathological	**Pathological**
Idiopathic	Idiopathic
Post – traumatic -Malunion -Physeal arrest	Post – traumatic -Malunion -Physeal arrest -Tibia metaphyseal fracture(Cozen's phenomenon)
Metabolic -Rickets -Renal osteodystrophy	Metabolic -Rickets -Renal osteodystrophy
Neuromuscular	Neuromuscular -Cerebral palsy -Paralytic conditions (eg. Poliomyelitis)
Infection (Osteomyelitis)	Infection (Osteomyelitis)
Skeletal dysplasia -Achondroplasia -Metaphyseal dysplasia	Generalised disorders -Juvenile arthritis -Osteochondrodysplasia -Osteogenesis imperfecta

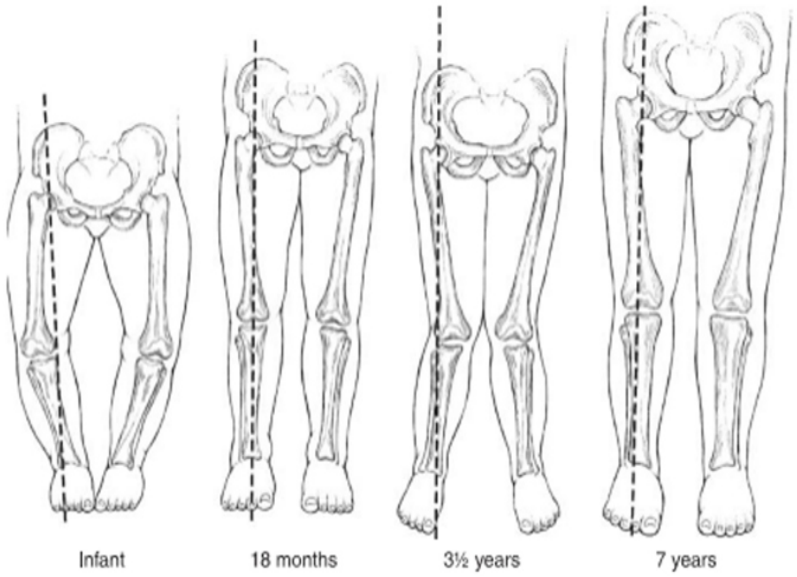

Figure 1: Normal development of knee angle in children

for metabolic cause of the deformity. Past history of trauma, infection or rheumatic conditions is important to understand other causes of the deformity.

Clinical examination:

If the underlying cause of the deformity is not evident, a careful clinical examination can help to elucidate the possible cause.

•Unilateral deformities are almost always pathological and the cause is likely to be local pathology in the femur or tibia. Bilateral symmetrical deformities are more likely to be the result of metabolic disorders or skeletal dysplasia, especially if associated with short stature (Fig. 2). Measurement of sitting and standing height, general appearance and family history of short stature is important in case of skeletal dysplasia and hypophosphatemic rickets.

• Measurement of inter- malleolar distance in genu valgum and inter - condylar distance in genu varum deformity is important to assess the severity and progress of the deformity (Fig. 3).

Specifically, persistent varus deformity with an inter – condylar distance >5 cm in children older than 3 years and valgus deformity with inter – malleolar distance >10 cm. in children older than 8-10 years should be considered abnormal.

• Site of the deformity should be determined by knee flexion test (Fig. 4). If the angular deformity disappears on knee flexion, the origin of deformity is in distal femur, but if the deformity persists on knee flexion, the origin of deformity is in proximal tibia.

•In physiological genu varum, there is a gentle curve involving both the thigh and the leg with more pronounced bowing in distal femur and U/3-M/3 junction of tibia.

In Blount's disease, the deformity is commonly at the proximal tibia metaphysis with an acute medial angulation immediately below the knee. 'Cover up' test is a useful screening test to assess the alignment of upper part of the lower leg in children presenting with bow legs between the age of 1 and 3 years.[3] (Fig. 5)

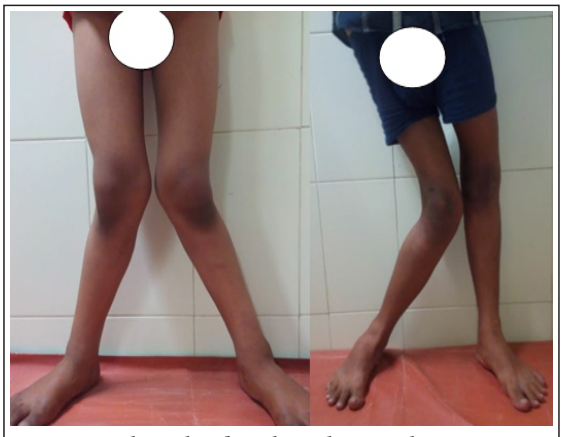

Figure 2: Bilateral and Unilateral genu valgum

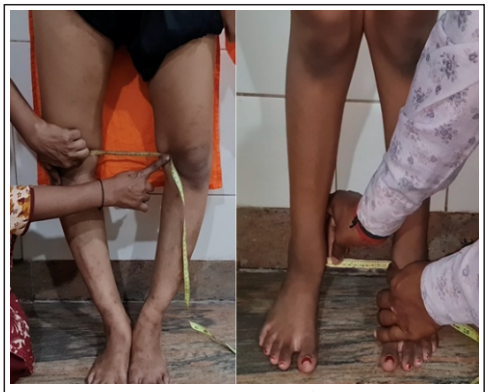

Figure 3: Measurement of inter-condylar distance in genu varum and inter-malleolar distance in genu valgum

• Ligamentous laxity may be the cause of genu valgum, therefore evaluation of generalized ligament laxity and stability of collateral and cruciate ligaments of the knee should be done.

• Angular deformities around the knee are often associated with torsional deformities e.g. genu varum is mostly associated with internal tibial

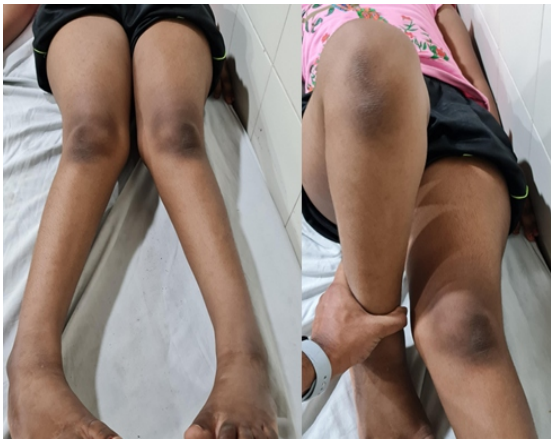

Figure 4: Genu valgum disappearing on knee flexion suggesting the deformity in distal femur.

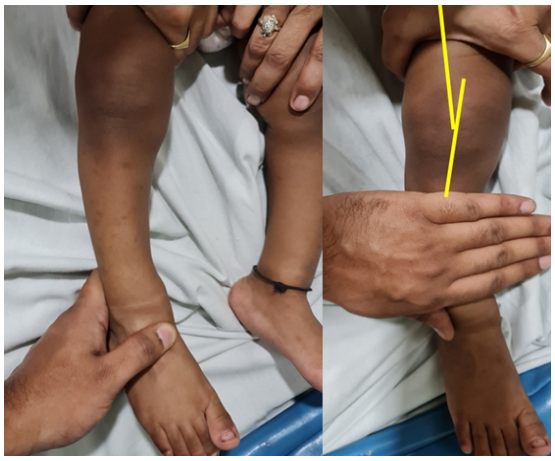

Figure 5: Negative 'Cover up test' (upper tibia in valgus) in physiological bowing.

torsion, hence proper evaluation of hip rotation and thigh foot angle are important for assessment of femoral and tibial torsion.

• Children must be examined for generalized features of rickets to rule out metabolic cause for angular deformity (Fig. 6).

• Iliotibial band contracture may cause genu valgum and its presence should be ruled out by Ober test.

• Inspection of gait and determining the foot progression angle is important. When laxity and incompetence of the lateral collateral ligament are present, the lateral head and upper tibia shift laterally during gait, causing a lateral thrust while walking. In physiological bow legs, there is no such lateral thrust. In severe genu valgum, child may walk with the knees rubbing together, feet apart and one knee swinging around the other. Such abnormal gait may lead to pain in the thigh and / or calf and easy fatigability.

Imaging

• Full length standing, weight bearing radiograph of both lower limbs from top of the iliac crest to the ankle with knee straight and patella facing forward should be obtained to assess the angular deformity. Paley and Tetsworth have standardized the approach to the radiographic evaluation of the lower limb deformity and advised the malalignment test as a step by step method to assess the site and magnitude of the deformity[4,5]. The MAD(Mechanical Axis Deviation), mLDFA (mechanical Lateral Distal Femur Angle), mMPTA (mechanical Medial Proximal Tibia Angle), JLCA (Joint Line Congruence Angle) need to be measured.(Fig.7) CORA needs to be determined and osteotomy site and angle needs to be measured.

• Aetiology : AP and lateral X – ray of the knee may reveal features of rickets (cupping , fraying, splaying) (Fig. 8a). They are also helpful to detect any bony physeal bar (post-traumatic/ post-septic) or condyle hypoplasia.

• In the suspected case of skeletal dysplasia, skeletal survey is recommended.

• MRI : It is the modality of choice for physeal bar assessment. A 3-D spoiled gradient echo sequence with fat saturation is used to perform physeal bar mapping.

• While planning for growth modulation surgery, the elbow X-ray can be helpful to know the skeletal age and growth remaining by Sauvegrain method.

Blood Investigations

If a metabolic cause is suspected for the angular deformity, blood investigations like Serum Calcium/ Phosphorus/ Alkaline Phosphatase/ Vitamin D / PTH/ Renal function test and Urine

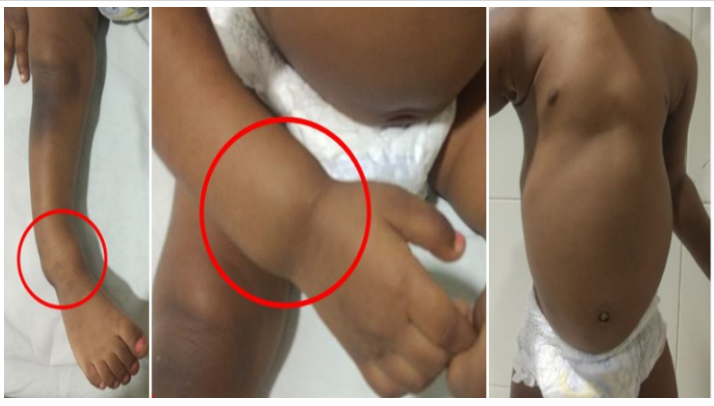

Figure 6: Metaphyseal widening around ankle and wrist , pigeon chest-clinical features of rickets.

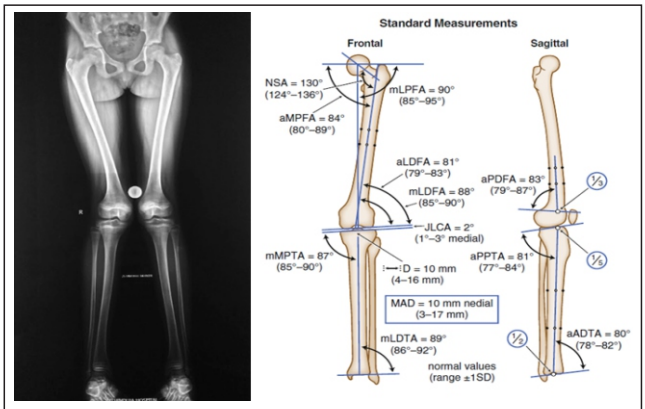

Figure 7: Full length AP X-ray of legs with normal angle measurements

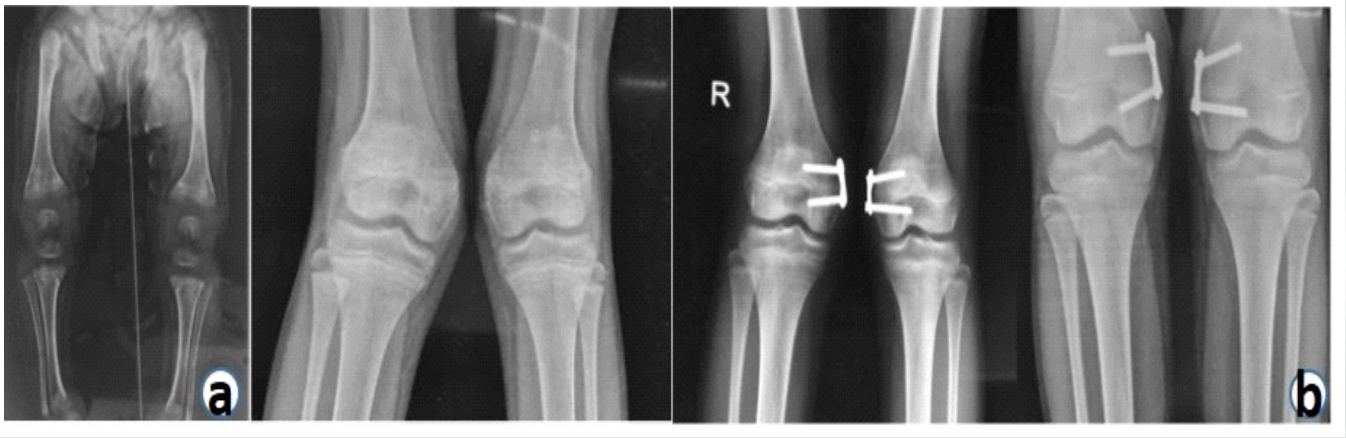

Figure 8: (a) Radiological features of rickets. (b) Bilateral genu valgum treated with distal femur eight plates

Calcium may be required.

Treatment :

The goal of treatment for angular deformities around the knee is restoration of normal mechanical axis alignment and joint orientation.

Observation:-

Children with physiological varus or valgus knee (i.e. who fall within two standard deviations from the normal value for their age or within the second zone on the X- ray) require no treatment other than observation and regular follow up at 3 months for clinical evaluation of the deformity. Parents need to be counseled that it is not a true deformity, but a variant of normal development of the knee which usually resolves spontaneously. X-ray needs to be repeated if there is clinical worsening(10).

Bracing or shoe wedges are not effective in correcting the deformity. They are poorly tolerated and unnecessary in case of physiological deviation.

Medical Treatment :

Genu varum or valgum deformities secondary to rickets should be treated with calcium and vitamin D supplements and dietary advice. The progress of the deformity should be monitored and surgical intervention may be done when indicated.

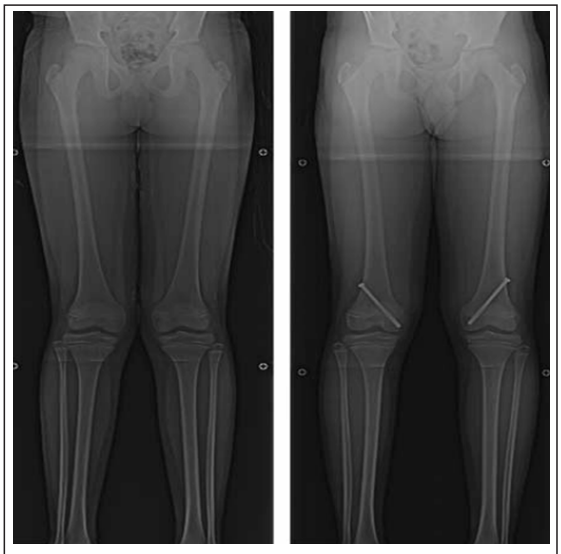

Figure 9: Bilateral distal femur PETS in a child nearing skeletal maturity

Surgical Treatment:

Persistent severe deformity with an inter- condylar distance > 5 cm in children older than 3 years and valgus deformity with inter -malleolar distance >10 cm. in children older than 8-10 years should be considered abnormal and are indications for surgical treatment. The choice of surgery depends on the skeletal maturity of the child (skeletal age), severity and site of the deformity.

Guided growth (reversible hemiepiphysiodesis) :

Principle:

It is a method of gradual correction of deformity in a skeletally immature child, where tethering of the physis is done on the convex side of the deformity, while allowing the physis on the concave side of the deformity to continue to grow.(6) The anticipated amount of correction depends on the age of the child and the location of the physis. The prerequisites include a minimum of 2 years of growth remaining and a deformity originating from or in the vicinity of the growth plate.

Technique:

This technique of guided growth was first used in children by Blount and Clarke in the 1940s using staples, but there was a high complication rate including staple migration and failure. Peter Stevens et al. in 2007 introduced tension band plate commonly known as eight plate. The eight plate is a two – hole non locking extra- periosteal plate that is fixed to the deformed bone at the level of the physis with one screw each inserted proximal and distal to it(Fig. 8b). There is some debate in the literature about the angle of the screws during implantation. In a recent clinical study, Eltayeby et al. showed that the initial screw angle (0-30^{0}) does not significantly affect the rate of correction and they recommend inserting screws taking account of anatomical restrictions to avoid the

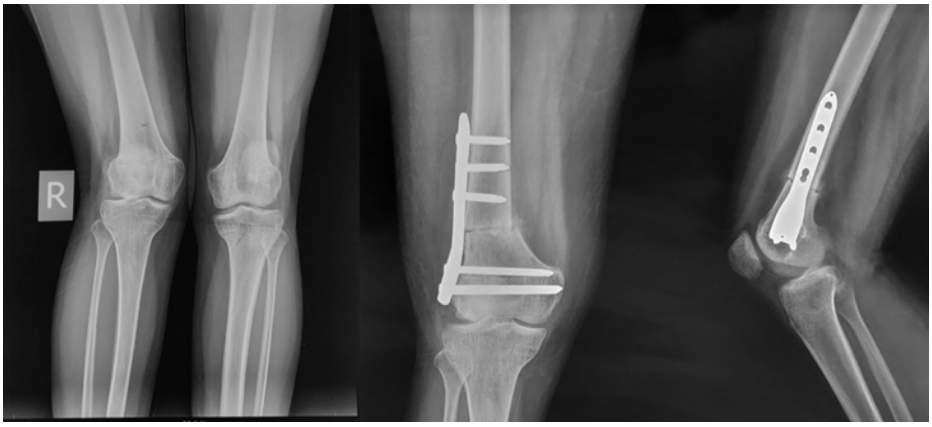

Figure 10: Right genu valgum treated with distal femur closing wedge osteotomy and plate fixation

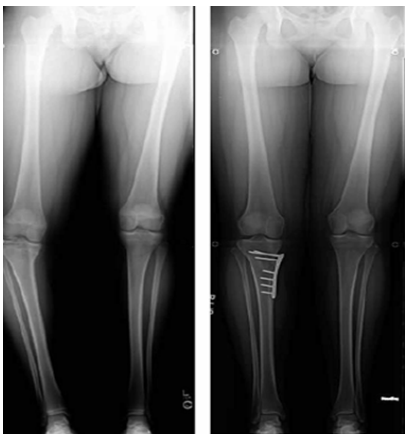

Figure 11: Right adolescent tibia vara treated with corrective osteotomy

physis, rather than favoring parallel, divergent or widely divergent configuration.(7) After insertion of eight plate, the child is followed up clinic-radiologically every 3 months and once desired deformity correction is obtained, the plate is removed. Some authors use a modified surgical technique called the 'sleeper plate' where after the deformity correction, only the metaphyseal screw is removed and the plate with the epiphyseal screw is left in place. If the deformity recurs, only the metaphyseal screw needs to be reinserted.

Outcomes and complications of growth modulation surgery:

The success rate of growth modulation surgery is almost 100% in idiopathic cases and slightly lower in pathological cases. Some complications are

1) Failure to achieve the correction: Related to BMI, age and underlying aetiology

2) Overcorrection : Related to poor patient compliance and failure to report on time for implant removal

3) Hardware failure : Implant breakage, loosening, migration or extrusion

4) Rarely, premature physeal closure

5) Rebound : More likely in severe initial deformity undergoing early hemiepiphysiodesis

In 1998, Metaizeau et al described PETS (Percutaneous Epiphysiodesis using Transphyseal Screw) where fully threaded cannulated screws are inserted from the metaphysis across the physis into the same side or opposite of the physis.(8) The physis fuses very quickly after screw insertion, hence this method is more suitable for a child nearing skeletal maturity(Fig. 9).

Corrective Osteotomy

It is indicated in children close to or at skeletal maturity or in presence of a physeal bar. A meticulous pre-operative planning is essential based on the site and magnitude of deformity. A weight bearing full length AP radiograph from hip to toe with knee in extension and patella facing forward is helpful in pre-operative planning. Acute correction of deformity can be done using opening wedge, closing wedge, reverse wedge or dome osteotomy(Fig.10). Internal fixation is usually done using plates and sometimes Kirschner wires.(9) Complications of acute correction are injury to neurovascular structures, delayed union or non-union of osteotomy. Alternative method is gradual correction using an external fixator or distraction osteogenesis.

Special Conditions

Post-traumatic / Post-infective physeal bar leading to angular deformity:

The treatment depends on the age of the patient, the location, the aetiology and the extent of deformity. Physeal bar resection is indicated when there are at least 1 or 2 years of remaining growth and the bar involves < 50% of the growth plate. Physeal bar resection consists of removal of the bone bridging the metaphysis and the epiphysis and filling the gap with an interposition material like fat, methyl methacrylate or polymeric silicon, to prevent reformation of the bony bar. Additional osteotomy or hemiepiphysiodesis may be indicated in addition to physeal bar resection if a clinically unacceptable deformity is present.

Tibia Vara / Blount's Disease:

Tibia vara is a developmental condition, characterized by multiplanar deformities like proximal tibia varus, procurvatum and internal rotation, caused by a growth alteration of the medial proximal metaphysis, physis and epiphysis of the tibia. In 1937, Blount was the first one to identify two forms based on the age of onset- infantile form (age < 4 years) and adolescent form (> 10 years).

Infantile tibia vara is bilateral in almost 50%, but rarely symmetrical varus and with variable internal tibial torsion. In some cases, lateral ligament instability with a 'varus thrust gait' may be present. Levine and Drennan have measured the metaphyseal-diaphyseal angle (MDA) on AP X-ray for early diagnosis of infantile tibia vara and reported that

MDA>16^0 is suggestive of infantile tibia vara. It is staged using Langenskiold's classification with six progressive stages, based on radiological appearance of medial proximal tibia and epiphyseal-physeal-metaphyseal alterations.(10)

Growth modulation surgery is effective in younger children whereas corrective osteotomy, sometimes with hemi plateau elevation is used in adolescent tibia vara(Fig.11).

Conclusion

Angular deformities around the knee i.e. genu varum and genu valgum are common in childhood. Physiological variations in the growth and development of children need no treatment other than observation. True deformities need proper management to avoid future consequences. Treatment should be tailored to the underlying aetiology and associated deformities on the other axis(rotation and shortening) should be ruled out. Growth modulation surgery should be the treatment of choice in patients with sufficient residual growth , whereas deformities in skeletally mature children require corrective osteotomy. Meticulous pre-operative planning of the site and magnitude of the deformity is essential for a successful outcome of the surgery.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the Journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Conflict of Interest: NIL; **Source of Support:** NIL

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