A Classic Approach in the Evaluation and Resolving Pediatric Lower Limb Angular Deformity: Educational Corner

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Background

Most pediatric knee angular deformities are physiologic which are resolved spontaneously without surgical intervention (1). However, these deformities are a common finding and, in some cases, need to be addressed with some interventions such as bracing, growth modulation, and corrective osteotomy. There are some reasons why we should address angular deformities, including unacceptable appearance, altered knee biomechanics, gait disturbance, knee pain, and induced early osteoarthritis (2).

The goal of the surgical interventions is to restore normal lower limb mechanical axis and alignment as well as equal limb lengths and horizontal knees by skeletal maturity. Based on the age and skeletal maturity of the patients, there are different techniques for the correction of pathological angular deformities, such as growth modulation and corrective osteotomy (3). Corrective osteotomies are often considered as the treatment choice in skeletally mature patients or in the older ages of children when there is not enough time for growth modulation. Another option is the growth modulation technique by manipulating normal bone growth patterns. Growth modulation by hemiepiphyseodesis was performed for pediatrics with a considerable amount of angular deformity in lower limbs (4-6). There are several techniques implants for lower and limb including hemiepiphysiodesis, tension-band plate method (TBP), percutaneous transphyseal screw (PETS), stapling, 8-plate, and 3-hole 3.5 mm reconstruction plate (7-11). Previous studies revealed that growth modulation had effective results in restoring lower limb normal alignment by all the implants with different complication rates which were favorable in terms of safety, cost, use of hospital resources, speed of correction, and most importantly, patient acceptance (3, 12, 13).

Therefore, in this educational corner, the aim is to address the angular knee deformity in the coronal plane in the pediatric population. To achieve this goal, the treatment methods and recent literature on lower limb angular deformity were reviewed step by step to achieve the best decision.

Case Presentation

A 12 years-old girl presented with bilateral idiopathic genuvalgum who denied any comorbidities and past medical history was examined in this study. We considered her knee deformity as a pathologic genu valgus because of 9° valgus and her age was more than 7 years. In additional assessments, it was confirmed that there was no metabolic disorder and dysplasia. On a full standing alignment view radiography, MAD was on zone +2 (0-25 percent) and the deformity was considered as a moderate genu valgus. Based on the age, healthy physes and bilaterality of deformity, hemiepiphyseodesis was performed and was followed with 3 month intervals. After 18 months, the patient was examined with a 3-joint x-ray and all the lower limb deformities were resolved and MAD was in -1 zone (50-75 percent) as a normal position.

Step one: Physiologic or Pathologic?

A newborn or infant normally has varus knee or bowlegs, and at months 6 to 12, the knee is in the highest grade of varus. At the 18 to 24 months of age, the knee has a neutral tibiofemoral angle (TFA) near zero (when the infant begins to stand and walk). With growth, genu valgum (knock-knee) appears, and maximal valgus deformity is around the age of 3-4 years with a mean TFA of 12°. Continuously, the degree of genu valgum is decreased spontaneously by aging until seven years, and normal degrees are 7 and 8 in the female and male gender, respectively (1, 14) (Figure 1).

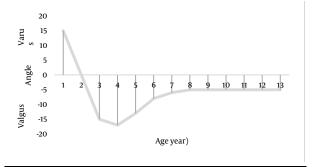


Figure 1. Normal development of the knee angular deformity

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It is so important to be careful about the evaluation and diagnosis of angular deformity in pediatrics because there are different treatment methods based on the etiology and degree of deformity. The physiologic angular deformities often need no treatment except for observation and reassurance of the parents (3). A physiologic genu valgum has some features including, lack of symptoms, normal structure, symmetric deformities, and prevalence in ages between 2 and 5 years.

Age at onset, stage of disease, condition of epiphyseal femoral head and extent of involvement, and the extrusion of the femoral head are the most important criteria considered to determine the prognosis of the disease and outcomes. The disease onset after the age of six, female gender, obesity, and hip joint stiffness, especially limited abduction, are associated with poor results (8,9).

The pathologic genu valgum is defined as a condition presented in the ages out of physiologic valgus (ages > 7 or < 2 years) in association with some conditions including posttraumatic, systemic, and metabolic conditions, dysplasia, and neoplasms as seen in table 1 (15-17). However, some conditions can induce pathologic varus deformities such as metabolic disorders, dysplasia, neoplasm, and previous trauma (Table 1).

A pathologic knee deformity needs further evaluation such as history, radiological, and bone metabolic assessments. A full history examination should be performed for the patients, including assessment of growth and development, progression, associated complaints (pain, limping), traumatic history, family history, onset, and previous treatments (18). All the patients with pathologic knee deformities should undergoe a full standing lower limb radiography (3-joint or alignment view), anteroposterior (AP) and lateral knee x-ray, and mechanical angles must also be measured. The metabolic evaluations (19) such as calcium, phosphorus, magnesium, vitamin D 25 OH, parathyroid hormone (PTH), and thyroid hormones should be assessed in all patients with pathologic knee deformity.

| - 11 | | | | | |
|------|---|--|--|--|--|
| | e1. Skeletal affection presented as bowlegs and knocked-knee | | | | |
| Gent | Genu varum or bowlegs | | | | |
| 1 | Apparent genu varum | | | | |
| 2 | Physiologic genu varum | | | | |
| 3 | Congenital familial tibia vara | | | | |
| 4 | Tibia vara (Blount's disease) | | | | |
| 5 | Asymmetric growth arrest of the medial part of the distal femur and | | | | |
| | proximal tibia due to infection, fracture, or tumor | | | | |
| 6 | Rickets-vitamin D deficiency or refractory (hypophosphatemia) | | | | |
| 7 | Bone dysplasia, such as achondroplasia and metaphyseal dysplasia | | | | |
| 8 | Fibrocartilaginous dysplasia (FCD) | | | | |
| 9 | Congenital longitudinal deficiency of the tibia with relative overgrowth of | | | | |
| | the fibula | | | | |
| 10 | Lead or fluoride intoxication | | | | |
| Gent | u valgum ir knock-knee (15-17) | | | | |
| 1 | Physiologic valgus (most common) | | | | |
| 2 | Posttraumatic (e.g., Cozen fracture, distal femoral physeal fracture, | | | | |
| | proximal tibial physeal fracture) | | | | |
| 3 | Systemic/metabolic conditions (eg, rickets, mucopolysaccharidosis type IV) | | | | |
| 4 | Skeletal dysplasias (e.g., chondroectodermal dysplasia) | | | | |
| 5 | Neoplasms [e.g., Hereditary multiple exostoses (HME)] | | | | |
| ~ | Iliotibial band tightness (due to paralytic conditions, such as | | | | |
| 6 | myelodysplasia, spastic diplegia, or spinal cord injury) | | | | |
| 7 | Knee arthritis (rheumatoid, hemophilia) | | | | |

Step Two: Need for Correction

In a pathological knee angular deformity , there is a mechanical axis deviation (MAD) in the lower limb (5). Paley et al. divided the knee into 6 zones (-1, -2, -3, +1, +2, +3) based on MAD, with positive and negative values related to the lateral to the midline or valgus and medial to the midline or varus, respectively. Zone 1 is centered over the tibial spines, zone 2 is within the tibial condyle, and zone 3 is beyond the cortex. A normal mechanical axis falls within zone 1 (20) (Figure 2).

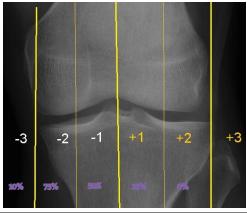


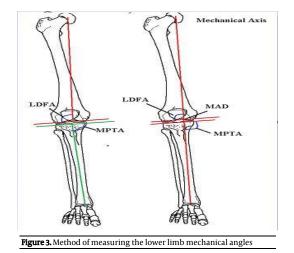
Figure 2. Mechanical axis deviation in the knee

As seen in figure 2, Tercier et al. divided the knee based on the Paley classification to zones from 0 to 100% laterally relative to the medial knee (21). The zone between 50 and 75% (negative side) is considered as the normal zone for the mechanical axis. The aim of growth modulation is to deviate the mechanical axis to 50% or near zero zones of MAD. The knee is in valgus deformity when MAD is in the positive zones (0 to 50%); in these cases, a medial hemiepiphyseodesis is performed and in varus deformities (MAD more than 75%) patients undergoe a lateral hemiepiphyseodesis (22).

The indication for intervention in this situation is deformities that are asymmetrical and associated with pain, joint stiffness, systemic disorders, or syndromes that may indicate a serious underlying cause requiring treatment (3). Concerning idiopathic genu valgum after the age of eight years old, correction of excessive physiologic (idiopathic) genu valgum may be indicated when there is gait disturbance, difficulty running, knee discomfort, patellar malalignment, evidence of ligamentous instability, or excessive cosmetic concern (23).

Step Three: Location of Deformity

Evaluating the location of knee deformity needs a full standing lower limb AP x-ray (3-joint) in order find the mechanical axis of limb and measure the lateral distal femoral angle (LDFA) and mechanical femorotibial angle (mFTA) (Figure 3).



The mechanical axis of the lower extremity in the frontal plane consists of two components: collinear centers of the femoral head, knee joint, and ankle joint. Based on the lower limb mechanical axis, there are some angles which could guide us to know the location of the deformity. With measuring both mechanical (mLDFA) and mechanical proximal tibial angles in full standing lower limb AP x-ray, one can evaluate the location of the deformity and address the deformity with the best approach. The normal range of these angles is $87 \pm 3(17, 20)$.

Therefore, in the cases that the medial proximal tibial angle (MPTA) is beyond the normal range, the location of deformity is in the tibia bone, and if LDFA is abnormal, the location of deformity lay in the femoral bone. Besides, if both the MPTA and LDFA angles are abnormal, the deformity originates from both tibial and femoral bones. **Step Four: Treatment Options**

There are several treatment options for lower limb angular deformities, such as conservative treatment with a brace, growth modulation, and corrective osteotomy. Generally, correction osteotomy is used for children within the end ages of growth as well as for adults, but growth modulation or hemiepiphyseodesis can be used for pediatrics in growth ages (23, 24). This method is the one adopted in the current study in pediatrics because of its less associated risk, no need for immobilization, and less hospital stay. Table 2 presents the indication and disadvantages of each method.

In cases with bilateral lower limb deformity, the growth modulation is the preferable treatment in most studies because of the early knee range of motion (ROM) and weight-bearing in this technique (25). However, corrective osteotomy needs no weight bearing period and limits the surgeons to performing the bilateral osteotomy. The criteria suggestable for performing hemiepiphyseodesis in angular knee deformity are:

- Clinically unacceptable deformity in a patient with open physis (26)
- a physis with adequate growth remaining (approximately one year) to allow correction (27)
- Appropriateness for bilateral knee deformity (25)
- Heathy physis and absence of skeletal dysplasia, trauma, and irradiation (26)
- Age of 12 years old or younger (28)

Step Five: Time Indications

It is important to estimate the remaining growth based on the skeletal age as a major step of angular deformity correction. Children with significant growth remaining including girls less than ten years old and boys less than 12 years old are suitable cases for acting these procedures on (26). In general, the remaining growth time is estimated by subtracting the age of the patient on the day of plate implantation from 17 years for males and 15 years for females.

Based on the previous studies (23, 35), the average correction angles are 7 and 5° per year in the femur and tibia, respectively. The timing of plate removal is a major challenge in preventing under- or over-correction. Growth modulation is a favorable option in patients with less remaining growth time because of no limitation in plate removal and the corresponding rebound effects (26). An overcorrection amount of 5° has been reported to be favorable among surgeons in previous studies to prevent the deformity rebounding (5, 6, 36).

Step Six: Techniques and Devices

There are two types of hemiepiphyseodesis, including permanent and reversible, with the permanent hemiepiphyseodesis consisting of ablation or curettage of one side of physis that is an irreversible method, which is indicated in specific cases (37, 38). Another type is reversible, which saves the growth plate that is not associated with the destruction of physis. This method could be utilized by several implants, including stapling, PETS, TBP, and 8-plate or 3-hole 3.5 mm reconstruction plate (Figure 4).

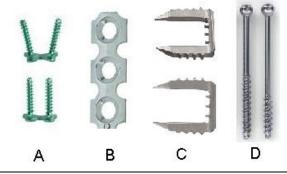


Figure 4. Implants used for growth modulation, 8-plate (A), Reconstruction plate (B), Rigid staple (C), and Transphyseal screw (D)

Several reports have shown the complication and high failure rate of rigid staple and PETS, including the slower correction of deformity, implant migration, and also more fatigue failure rate compared to the plates such as the reconstruction and eight plates (6, 10, 11, 39-42).

Stapling is the first device used extensively for hemiepiphyseodesis among surgeons (5). However, previous studies reported some complications of using staples, such as physeal arrest and device breakage (43, 44). Previous studies hav indicated that TBP and PETS are equally effective compared to stapling in hemiepiphyseodesis (7-9, 17). Park et al. compared the efficacy of TBP and PETS and concluded that TBP was as effective as PETS in hemiepiphyseodesis; however, the PETS method was faster in angular deformity correction (41). Limited studies reported complications following TBP insertion such as screw loosening and need to revision, for instance, Schroerlucke et al. reported a high failure rate (45%) regarding the use of TBP (45).

However, in growth modulation by submuscular or subfascial plates technique, in which one should preserve the periosteum in or just posterior to the midsagittal plane to avoid genu-recurvatum, there is no need for postoperative immobilization (Table 3).

| | Indications | Disadvantage |
|-----------------------------------|---|---|
| Bracing | Unilateral involvement (29) Applied for children younger than 3 years old (30) Mild deformity Absence of progression risk factors such as ligamentous laxity and obesity (30,31) | Inappropriate for children older than 3 years (29) Inappropriate for bilateral involvement (29) |
| Growth modulation (24, 32) | Late-onset with mild to moderate deformity Age of 12 years or vounger Heathy physis and absence of skeletal dysplasia | High failure in BMI greater than 45 Physeal damage Breakage and extrusion Not applied for cases with growth arrest |
| Corrective osteotomy (23, 33, 34) | Near or after skeletal maturitv Case of excessive physiologic deformity Cases with growth arrest | Nerve palsv Vascular damage Compartment syndrome Delav union Infection Immobilization |

| Implant | Advantages | Disadvantages | Study |
|----------------------|--|--|---|
| Stapling | Shorter operation time Easy to apply | Migration (2-8%) Physeal arrest Average rebound of 5° | Blount et al. (40) Cho et al. (4) Gottliebsen et al. (8) Mielke and Stevens (22) |
| PETS | Noninvasive Shorter operation time Faster correction 8% need to postoperative physiotherapy | Migration Physeal arrest Average rebound of 2° | De Brauwer and Moens (7) Metaizeau et al. (36) Nouth and Kuo (46) Liotta et al. (47) |
| TBP | Average correction rate of 5° per year | High failure rate Screw loosening 40% need to postop Physiotherapy | Schroerlucke et al. (45) Park et al. (41) Liotta et al. (47) |
| Eight plate | Lower device failure rate Early knee motion | High cost (around 200\$) Invasive surgery | Burghardt and Herzenberg (48 Das et al. (49) Vaishya et al. (42) |
| Reconstruction plate | Lower device failure rate Early knee motion Verv low cost (around 10\$) | Invasive surgery | Aslani et al. (39) Baghdadi et al. (11) |

PETS: Percutaneous transphyseal screw; TBP: Tension-band plate method

Hemiepiphyseodesis was performed by plating in patients with the knee angular deformity and two years remaining growth using several methods such as TBP, eightplate, and three-hole 3.5 mm reconstruction plate. Baghdadi et al. reported the outcomes of 198 limbs corrected with hemiepiphyseodesis using a 3-hole reconstruction plate (11).

This method seems to be more feasible in developing and poor countries because of the low cost of the plate (11) (Figures 5 and 6).



Figure 5. A 12-year old girl who was a candidate for bilateral hemiepiphyseodesis due to valgus deformity of her knees, the angles measured showed that the location of the deformity was in femoral bone.

Step Seven; Degree of Correction

By the growth modulation technique, the average correction angles were obtained as 0.77 and 0.5° per month in mLDFA and mMPTA angles, respectively (35). Regarding the correction angle and goal of treatment by preoperative planning, it is important to choose the appropriate method for the patients with angular deformity. It is suggestable in some studies to overcorrect as the amount of 5° to avoid deformity rebound (5, 6, 36).



Figure 6. Pre-operative and 18th month post-operative three joint alignment view, the 12-year old girl who underwent bilaterak bilateral hemiepiphyseodesis

There are two methods for identification of chronological age, including the Westh and Menelaus arithmetic method (50) and the Paley multiplier method (51). Westh and Menelaus described the arithmetic or "rule-of-thumb" method based on four basic rules and chronological age, including:

- a. The proximal tibia physis grows 6 mm per year
- b. The distal femoral physis grows 10 mm per year
- c. The growth stops in 16 years old in boys
- d. The growth stops in 14 years old in girls

It is important to know that the "rule-of-thumb" method could not be appied in the early childhood. Paley et al. first described the multiplier method in 2000. This method can be used to calculate the remaining growth in a particular segment of the limb based on the chronological age, gender, and length of the affected bone. Paley's method is easy and favorable to determine the growth remaining with no need for an additional x-ray.

Step Eight: Follow-up and Implant Removal

Radiographic assessment for follow-ups at the time of clinical correction and then approximately at 3-4 month intervals should be performed as described in previous studies (11, 26). Once the mechanical axis passes through the central third of the knee joint (full correction) and MAD perch on zone -1, the plate should be removed to avoid overcorrection.

Conclusion

Growth modulation is manipulating normal bone growth to correct the angular deformity of lower limbs in pediatrics. This method was performed with several implants such as stapling, PETS, TBP, eight plate, and reconstruction plates with various advantages and complications, as seen in table 3.

There are several factors and indications in using hemiepiphyseodesis in the patients with angular deformity, including the absence of bony diseases, open and healthy physes, 12 years old and younger ages, and remaining growth more than two years. It is so important to close postoperative follow-up to avoid under- or overcorrection. In the present educational corner, a stepwise and classic approach was proposed to pediatric angular deformity with eight steps to achieve the best alignment (Figures 7).

Conflict of Interest

The authors declare no conflict of interest in this study.

Acknowledgments

None.

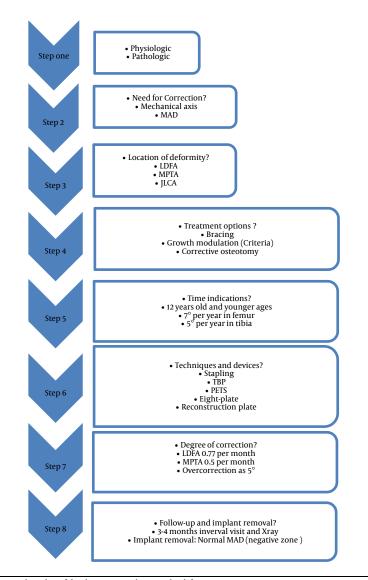


Figure 7. Stepwise algorithm of the classic approach to angular deformity MAD: Mechanical axis deviation; LDFA: Lateral distal femoral angle; MPTA: Medial proximal tibial angle; JLCA: Joint line convergence angle; TBP: Tension-band plate method; PETS: Percutaneous transphyseal screw

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