Pediatric Forefoot Pathology

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KEYWORDS

Pediatric forefoot
Polydactyly
Macrodactyly

KEY POINTS

- The management of pediatric pedal deformities of any kind requires extreme vigilance.
- The pediatric bone has unique characteristics that warrant special consideration.
- Many times, at initial presentation, the only complaint of pain comes from the child's parent. A careful physical examination and history-taking can help sort through the decision tree.
- Approach with a vision of how ones intervention will affect the child later as he/she reaches skeletal maturity.

INTRODUCTION

Surgical management of the pediatric forefoot often brings challenges to the foot and ankle surgeon. It requires a thorough understanding of the pathologic abnormality and underlying causes involved, which include the contributing genetic conditions. Albeit most of the deformities carry a rare level of incidence, they do however have a significant level of psychological component and stress on the pediatric patient. The goals of managing those pathologic abnormalities are ultimately to improve function while achieving a cosmetically acceptable outcome. In this article the common forefoot pathologic abnormalities in the pediatric population are reviewed with an added focus toward management of forefoot trauma in that patient group.

POLYDACTYLY

Polydactyly is a common congenital condition involving the hands and feet. It is characterized by the presence of one ore more supernumerary digits, and in some cases metatarsal/metacarpal bones (**Fig. 1**). Involvement in the hand is typically twice as often as in the foot.¹ The condition is equally frequent among male and female patients with bilateral involvement seen in 25% to 50% of the patients.^{1,2} In a study of 120,000 live births, Frazier found an incidence of 1.7/1000 births.^{3,4}

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The pattern of inheritance and exact cause of polydactyly are not fully understood. Appoximately 15% of patients with polydactyly of the feet have an associated anomaly.² In addition, the condition is seen with autosomal-recessively inherited syndromes, such as Pallister-Hall, Lawrence-Moon-Bardet-Biedl, Ellis-Van Creveld, and various trisomies. A family history of polydactyly has also been reported in 10% to 30% of the subjects.^{2,4}

Classification

Swanson in 1976⁵ classified congenital limb malformations. The main categories included failure of formation of parts, failure of differentiation of parts, duplication, overgrowth (gigantism), undergrowth (hypoplasia), congenital constriction band syndrome, and generalized skeletal abnormalities.

Temtamy and McKusick classified polydactyly by determining whether the presentation was isolated or part of a syndrome. Within those categories, further subclassification is based on the anatomic location of the duplicated digits as either preaxial or postaxial. Preaxial refers to the tibial side of a line bisecting the second ray, whereas postaxial refers to the fibular sides. Postaxial polydactyly are subdivided into types A and B. Type A digits are fully developed with articulating skeletal structures, whereas type B are rudimentary digits. Postaxial type A polydactyly is the more common pattern.⁶

Venn-Watson⁴ provided another classification based on the morphology of the corresponding metatarsal. The patterns from least differentiated to most differentiated were as follows: soft tissue duplication, wide metatarsal head, T-metatarsal, Y-metatarsal, and complete duplication.

Watanabe and colleagues² classified polydactyly by the type of ray involvement and level of duplication. The anatomic pattern types in medial-ray polydactyly are tarsal,

metatarsal, proximal, and distal phalangeal. Central-ray types are metatarsal, proximal, middle, and distal phalangeal. Lateral-ray are subdivided into medial and lateral supernumerary toes.

Treatment

Surgical management of infants can often be addressed by early suture ligation or surgical excision.⁷ General principles recommend saving the digit that is the most developed, has the most normal or anatomic MTP articulation, is most cosmetically acceptable, and is the one that will give the best contour to the foot.

Management of preaxial polydactyly is more complex with less optimum long-term results. Typically, the most medial digit is excised and additional tendon lengthening procedures, osteotomies, or other first ray soft tissue procedures may be used. Temporary pin fixation may be required to allow for soft tissue healing. However, complications of recurrent hallux varus and splaying of the first metatarsal are possible outcomes.

Postaxial polydactyly management has better and more predictable long-term results.^{4,5} In most cases, the lateral-most digit is excised, usually through a racquettype incisional approach. In addition, a block-metatarsal or wide-metatarsal head should be narrowed to a standard size to stabilize the joint. In the cases of T-shaped or Y-shaped metatarsals, the extra bone is typically resected to create a more standard metatarsal. Furthermore, in the cases of duplicated metatarsals, the entire ray should be removed. The transverse intermetatarsal ligament is also typically repaired in those cases to prevent splaying of the foot.

Central ray duplication has a more rare presentation but typically supernumerary central digits can be excised through a racquet-shaped incision with a dorsal arm. Similar to postaxial polydactyly in the cases of metatarsal duplication, the entire ray should be resected and a primary intermetatarsal ligament repair should be performed to prevent splaying.

MACRODACTYLY

Macrodactyly is considered a rare congenital deformity characterized by an increase in size of osseous and soft tissue structures of one or more digits.⁸ Unilateral presentation of digits is the most common presentation and is typically seen with the first, second, and third toes. In addition, soft tissue overgrowth is also associated at the distal plantar aspect of the involved digit.⁹ Although presentation in the foot is less common than in the hand, the deformity is more progressive.¹⁰

Although macrodactyly frequently occurs as an isolated congenital defect, literature supports association with neurofibromatosis-1, Klippel-Trenaunay-Parkes-Weber syndrome, hemihypertrophy, hemangiomatosis, arteriovenous fistula, lymphangiomatosis, and congenital lipofibromatosis.

The goals of surgical management of macrodactyly include achieving a plantigrade foot similar in width and length to the contralateral side. Treatment options include amputation, epiphyseal plate arrest, and different debulking procedures of the soft tissues. Potential complications include but are not limited to wound dehiscence, prolonged edema, and sensory disturbances.^{11,12}

SYNDACTYLY

Syndactyly and polysyndactyly of the digits are common congenital deformities of the foot (**Fig. 2**). Unlike the hand, syndactyly is typically a cosmetic problem without any functional impairment. Thus, the treatment goals are indicated for cosmetic, psychological, and practical reasons.^{13,14}





Classification

Davis and German¹⁴ classified syndactyly into 4 main categories: (1) incomplete, the skin webbing between the 2 digits does not extend into the distal aspect of the digit; (2) complete, the skin webbing extends to the most distal aspect of the digits; (3) simple, no phalangeal involvement; and (4) complex, phalangeal bones are abnormal.

A well-accepted technique in the surgical management of syndactyly is the use of a dorsal rectangular flap and full-thickness skin grafts to provide additional skin cover, especially at the base of the digits. However, complications of donor site morbidity, hypertrophic scarring, web creep, pigmentation problems, contractures, and hair growth are possible. Other methods for surgical repair include the use of a xenograft for tissue coverage and various plastic surgical techniques, such as the V-flap or rectangular flap.¹⁵

BRACHYMETATARSIA

Brachymetatarsia was described by Kite as shortening of one or more metatarsals because of a premature fusion of the epiphyseal line at the distal end of the metatarsal (**Fig. 3**).¹⁶ The causes range from traumatic, iatrogenic, and systemic to disorders that include pseudohypoparathyroidism, Turner syndrome, Albright hereditary osteodystrophy, and Down syndrome.^{17,18} Females are affected by a 25:1 ratio and typically the fourth metatarsal has the highest rate of incidence.

Surgical correction is used to alleviate pain, establish acceptable cosmesis to the foot, and restore functional metatarsal parabola. Those include soft tissue correction such as the Kelekian syndactylization of lesser toes 4 and 5, use of joint spacers, bone-grafting techniques, and the Ilizarov method of callus distraction.¹⁹

METATARSAL FRACTURES

Fractures of the metatarsals in children can present several treatment dilemmas. Many practitioners abide by the "if the bones are in the same room they will heal" philosophy



Fig. 3. Skeletal mature pediatric patient with a brachymetatarsal of the fourth metatarsal.

and others take a more aggressive stance. They also comprise most fractures seen by foot and ankle providers in children. In a study performed by The Cincinnati Children's Hospital they found that metatarsal fractures comprise 90% of all skeletal trauma in the foot in children with the fifth metatarsal being the most common of the 5 metatarsal bones to be acutely fractured and the second metatarsal being the most common bone subjected to stress fractures. A typical history of a child with a metatarsal stress fracture is that of a recent increase in activity or change of shoe or cleat. Radiographs are only diagnostic of about half the stress fractures seen in children so high suspicion is needed to diagnose an occult fracture. Magnetic resonance imaging may be used if suspicion is high. These injuries respond to immobilization in a cast for 3 to 6 weeks (Crawford AH, unpublished data, 1991).

Metatarsal Neck Fractures

Although metatarsal neck fractures are rare injuries, if there is no cartilaginous or epiphyseal involvement, most authors recommend a below-knee cast for 3 weeks. If surgical intervention is necessary, smooth K-wires are to be used to prevent physeal arrest or the development of growth aberrations.²⁰

Metatarsal Shaft Fractures

These fractures occur usually from direct trauma. Transverse plane deviation is well tolerated; however, sagittal plane mal alignment can result in abnormal weight distribution and future metatarsalgia, stress syndromes, and digital deformities.²⁰ When casting with or without closed reduction can be performed, it is indicated otherwise,

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open reduction, internal fixation with internal fixation that does not violate the physis generates good results.

Metatarsal Base Fractures

Metatarsal base fractures rarely cause long-term problems except for the first metatarsal due to the proximal location of its epiphysis. These fractures are referred to as "buckle base" fractures owing to their tendency to buckle at the proximal epiphysis. If there is no displacement of the physis, these fractures heal uneventfully with short leg casting.²⁰ Fractures of the base of the fifth metatarsal become more common as children reach the age of playing competitive sports, namely sports that require repetitive jumping. Avulsion fractures of the fifth metatarsal base must be differentiated from a painful secondary ossification center or os vesalianum. Riccardi and colleagues²¹ found that 47% of painful ossification centers of the fifth metatarsal base were misdiagnosed as fractures. Lawrence²² expounded on the Stewart classification system to reflect pediatric injuries. The system is separated into 6 categories: (1) open growth plate apophyseal avulsions; (2) open growth plate apophyseal stress fractures (Iselin disease); (3) closed or closing growth plate tuberosity avulsion fractures; (4) Jonestype fractures through the metaphyseal-diaphyseal watershed area; (5) acute diaphyseal fractures; (6) stress fractures of the diaphysis. These fractures respond well to conservative care in a below-knee cast. If nonunion occurs, removal of the fragment may be needed but is rarely necessary in the pediatric population.

Phalangeal Fractures

Phalangeal fractures in children rarely warrant surgical intervention. Van Vliet-Koppert and colleagues²³ found that 95% of digital fractures were minimally or nondisplaced. In the review performed by Cincinnati Children's Hospital, it was found that the proximal phalanges were the most commonly fractured digital bone followed by the middle and distal (Crawford AH, unpublished data, 1991). Treatment consists of buddy splitting with a stiff-soled shoe. Displacement of the first proximal phalangeal base may warrant a closer look because of the propensity for the development of hallux limitus later in skeletal maturity. Open fractures in these spaces should be treated as any other stage 1 open fracture with irrigation and debridement; if the insult occurs around the nail complex, removal of the nail is warranted. These wounds are often closed primarily.²⁴

Iselin's Disease

Overview

Traction epiphysitis of the base of the fifth metatarsal was originally described by Iselin in 1912. He noted this condition arose during adolescence. Iselin's disease is one of several osteochondroses found in the foot. Anatomically, the osteochondroses are categorized as articular, nonarticular, or physeal. This secondary center of ossification appears at about 10 years of age in girls and 12 years of age in boys.²⁵ This secondary ossification center can be seen well on medial oblique radiographs of the foot but is masked by the anterior-posterior and lateral projections.²⁶ The anatomic area in which Iselin's disease arises is unique in that the base of the fifth metatarsal fills all anatomic criteria for nonarticular osteochondrosis, being (1) the site of tendon attachment; (2) the site of ligamentous attachment; and (3) the site of impact.^{25–27} Clinical diagnosis can be made with a typical history of pain on weight-bearing, participation in sports that require running, cutting, and jumping. The traction results from the pull of the peroneus brevis while the foot is inverted. Oftentimes edema can be noted around the

base of the fifth metatarsal. Radiograph findings show fragmentation of the epiphysis and TC-99m bone scans will show increased uptake around the apophysis.

Treatment

Treatment consists of relative rest, icing, and anti-inflammatory medication. If no resolution of symptoms is noted with these modalities, the next stage in the treatment ladder is immobilization in a cast or CAM walker in the acute stages. A surgical shoe is not an ideal treatment because it cannot control the movement of the peroneus brevis muscle-tendinous unit. This condition has been noted to resolve spontaneously with the cessation of skeletal growth but nonunion has been noted.^{26,28} Failure to recognize this diagnosis can cause long-term pain into adulthood. Differentiating a chronic or acute-on-chronic Iselin's disease from an avulsion fracture or stress fracture requires a thorough history because treatment likely will differ. Proximal stress fractures occur in diaphyseal bone, which is a key anatomic difference; however, Jones fractures have the potential to occur closer to the physis given their metaphyseal-diaphyseal location. Treatment of a base avulsion would be casting; however, if a nonunion has occurred that has been aggravated later in life, removal of the fragment would be the treatment of choice.²⁶ Differentiating chronic Iselin's nonunion versus a painful of vesalianum is not as critical in that the treatments are similar.

Freiberg Disease

Overview

Osteochondrosis of one or more metatarsal heads was originally described by Freiberg in 1914. It is more common in female teenagers than male teenagers and the second metatarsal head is most commonly affected, followed by the third metatarsal head.²⁷ Repetitive microtrauma is the most accepted etiologic theory despite Freiberg's original theory of a single traumatic event being the causative factor. It was a common misconception that increased plantar pressure from an elongated second metatarsal was the cause of this transient osteonecrosis; however, Gauthier and Elbaz²⁹ in their study found no correlation between second metatarsal length and risk of osteonecrosis. Some authors think that a genetic predisposition is present in the development of this osteonecrosis. Blitz and Yu³⁰ published a case report of identical twins developing Freiberg infarction in which they used this data to elucidate a genetic root cause. Radiographs typically show flattening of the involved metatarsal head with sclerosis, fragmentation, and early joint space widening²⁹ and over time the joint will show typical signs of arthrosis. Magnetic resonance imaging is reserved for atypical radiographic presentations or to differentiate stress fracture or osteomyelitis. Usually, depending on the level of necrosis, the involved metatarsal will show low signal intensity on both T1 and T2 sequence.³¹

Treatment

Acute therapy involves a period of non-weight-bearing with icing and antiinflammatory medications. If this condition becomes chronic, conservative measures include offloading with a custom-molded orthoses that offload the metatarsal heads. Surgical interventions include chilectomy and joint remodeling, dorsiflexory closing wedge osteotomy of the involved metatarsal head that brings unaffected cartilage into the joint space, total joint arthroplasty, metatarsal head resection, and osteochondral autograft transfer system (OATS) transplant. Arthrodiastasis has also recently been described to treat chondrocyte damage either alone or as an adjunct. DeVries and colleagues³² described a surgical technique in which they combined osteochondral autograft transfer system (OATS) transfer of the involved metatarsal head from the femoral distal lateral condyle, which was augmented with mini-rail external fixation for 6 weeks. Interpositional arthroplasty using the extensor digitorum longus tendon has also been described for surgeons wary of total joint arthroplasty.³³

INSERT SMILIE CLASSIFICATION Longitudinal Epiphyseal Bracket

Overview

Longitudinal epiphyseal bracket (LEB) is a congenital anomaly affecting the tubular bones of the hand and foot in which the proximal epiphysis is L-shaped or C-shaped and runs along the medial border of the bone; this results in the formation of a short, trapezoidal bone caused by the bone growing in more of a mediolateral direction instead of longitudinally.^{34,35} The great toe is affected in 11% of all cases of LEB, or "Delta Phalanx," as is it was originally described.³⁶ Conditions associated with LEBs include the following: hallux varus, polydactaly, Rubenstein-Taybi, diastrophic dwarphism, Apert syndrome, syndactyly, ulnar and cleft hands, and tibial hemime-lia.^{35,37} Radiographically, the bracket can be visualized from 2 to 11 years of age.

Treatment

The stalwart of treatment of this condition is surgical resection of the bracket. Timing of surgical intervention is ideally during the first year of life to allow the affected bone to experience normal longitudinal growth. Further reasoning for resection of the bracket early in life is because there is still no bone overgrowing the bracket. Once ossification occurs over the bracket, bone must be resected to reach it, which increases the chance of disturbing the longitudinal physis. Adjunctive procedures include opening wedge osteotomy, which may be performed as part of the original surgery or as a staged procedure. There is also a role for callus distraction with external fixation.³⁸ Also, tendon lengthening may be needed as the tendinous unit may have modeled around the shortened bone.

JUVENILE HAV

Overview

Ninety percent of hallux valgus is hereditary (Fig. 4).³⁹ Pediatric hallux valgus presents several unique treatment dilemmas. Many times the child will present, pre-empted by a worried parent, with no pain whatsoever. Other times, the child may present with a hallux valgus deformity as part of a larger scale limb length discrepancy, metatarsus adductus, skewfoot, or painful flatfoot deformity that sincerely limits activity. The physis of the first metatarsal is located proximal along the bone unlike the other 4 metatarsals. Occasionally, a pseudoepiphysis will appear at the distal metaphysealepiphyseal junction. In male children, the proximal physis can appear from age 5 months to as late as 2.3 years in and from birth to 2 years in female children and closes anywhere from 13 to 22 years of age. The first step in any treatment algorithm for juvenile HAV should be a thorough radiographic, neuromuscular, and biomechanical examination.⁴⁰ McCluney and Tinley found significant radiologic parameters for differentiating juvenile HAV from a normal foot to be a 15 degree or more metatarsus primus adductus angle and a positively met protrusion index.⁴¹ A pediatric bunion will invariably have some component of hypermobility, especially when dealing with pediatric hypermobile flatfoot. A Silfverskiold test should be performed to determine the role of equinus in driving the deformity.

Treatment

The first step in the treatment of juvenile patients is to support the foot with orthoses. In some cases, a deep-seated heel cup may be used to control a flexible flatfoot. A heel

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Fig. 4. An anterior-posterior radiograph demonstrating a significant hallux abducto valgus deformity in a pediatric patient.

lift should be placed on the contralateral limb to stop abnormal subtalar joint pronation of the affected side if a limb length difference is measured. If there is a larger scale abnormality driving the hallux valgus such as a rigid flatfoot caused by tarsal coalition, this condition will dictate treatment. Watchful waiting is also an acceptable treatment algorithm if the child is in no pain and still has many formative years ahead.⁴⁰ In this philosophy, the surgeon waits until the physis is near closure at the time of adolescence and at that time a modified Lapidus is performed. In the adolescent population, a hallux valgus deformity with an associated hypermobile first ray can present a treatment challenge. This mechanical abnormality, when presenting in adults, can be corrected with predictability using a modified lapidus bunionectomy.^{42,43} In pediatric patients, however, this procedure has great potential to violate the proximal physis of the first metatarsal in patients who are not near skeletal maturity; thus, there is a certain popularity of the long-arm chevron osteotomy in this patient population that still has growth potential from the proximal physis. Some surgeons in the past have opted for soft tissue rebalancing procedures in this population; however, these procedures are fraught with recurrence.⁴⁴ Other options include an oblique closing base wedge osteotomy placed distal to the physis. This osteotomy does involve diaphyseal bone; however, the pediatric bone potential for healing should be considered. Medial cuneiform opening wedge osteotomy, originally described by Cotton in 1935, has also been used in the pediatric population as a procedure to maintain length of the first ray, although this procedure should not be performed until the age of 6 when the medial column has fully ossified.⁴⁵ Some surgeons choose to use the proximal physis to their advantage with epiphysiodesis procedures in an attempt to correct the angle of growth before most longitudinal growth occurs in the first metatarsal; however, this is a challenging surgical option and the potential for undercorrection or overcorrection

is high and it must be performed on the younger pediatric population for any success to be achieved. Achilles tendon lengthening or gastrocnemius recession should always be considered an adjunctive procedure if equinus contracture is present; if this driving force is not corrected, recurrence will inevitably occur because of a tight heel cord causing abnormal subtalar joint pronation and unlocking of the transverse tarsal joint.

DISCUSSION

The management of pediatric pedal deformities of any kind requires extreme vigilance. The pediatric foot should not be considered a miniature adult foot. The pediatric bone has unique characteristics that warrant special consideration. Many times, at initial presentation, the only complaint of pain comes from the child's parent. A careful physical examination and history-taking can help sort through the decision tree. In this article, an overview of the pathologic pediatric digits and metatarsals has been given as well as treatment algorithms for several digital and metatarsal derangements. Any deformity in children should be approached with a vision of how one's intervention will affect the child later as he/she reaches skeletal maturity.

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