Fibular Hemimelia

Subjects: Orthopedics
Contributor: Dror Paley

Fibular hemimelia (FH) is the most common lower-extremity congenital longitudinal deficiency, with occurrence between 1:135,000 and 1:50,000 births. It is associated with a constellation of deformities that fall into five general categories: tibial deformity, genu valgum, knee instability, leg length discrepancy (LLD) and foot and ankle deformities and deficiencies. Both deformities and leg length discrepancy in FH can present with a wide spectrum from mild to severe .

Keywords: fibular hemimelia ; SHORDT ; SUPERankle procedure ; Paley classification ; leg length discrepancy

1. Introduction

Fibular hemimelia (FH) is the most common lower-extremity congenital longitudinal deficiency, with occurrence between 1:135,000 and 1:50,000 births. It is associated with a constellation of deformities that fall into five general categories: tibial deformity, genu valgum, knee instability, leg length discrepancy (LLD) and foot and ankle deformities and deficiencies. Both deformities and leg length discrepancy in FH can present with a wide spectrum from mild to severe [1][2][3].

In addition to being hypoplastic, the tibia often presents with a diaphyseal valgus-procurvatum deformity with a skin dimple over the apex of angulation. Independent of the tibial diaphyseal valgus, the knee joint often presents with valgus orientation as well, which can originate in the proximal tibia, distal femur or both [4]. The cruciate ligaments can be hypoplastic or aplastic, creating anterior and posterior knee instability [5]. This is often asymptomatic in childhood but can become more problematic as the child grows.

The majority of FH cases present with unilateral involvement and are associated with a leg length discrepancy that originates from growth inhibition of the tibia and foot. Many children with FH have associated congenital femoral deficiency as well, in which femoral growth inhibition contributes to the overall leg length discrepancy. Combined with femoral inhibition this can become greater than 30 cm. The LLD at skeletal maturity is predictable using standard prediction methods including the Paley multiplier method and it follows the Shapiro 1a curve [6][7][8][9][10].

The foot and ankle deformities in FH have traditionally been the most challenging and disabling problems. In addition to absent rays and bracket metatarsals and syndactyly, the foot in more involved cases often presents with rigid and severe equino-valgus deformity. This fixed equino-valgus originates from a dysplastic and valgus distal tibia, a subtalar coalition malunited in equino-valgus, or both. The combination of rigid equino-valgus with a significant LLD has historically resulted in poor outcomes from reconstruction, resulting in ablative surgery commonly being recommended.

2. Foot and Ankle Reconstruction

Each Paley type is accompanied by a reconstructive treatment for the tibia, foot and ankle deformities. Most cases of Paley type 1 FH do not need any foot or ankle surgery before lengthening as the ankle is stable. In contrast, most cases of Paley type 2–4 FH will need foot and ankle reconstructive surgery to stabilize and/or create a plantigrade foot. Foot and ankle reconstruction usually needs to be performed prior to or can be combined with the first lengthening.

The SHORDT procedure involves a shortening and realignment tibial supramalleolar osteotomy to correct ankle valgus and procurvatum malorientation and lengthen the fibula relative to the tibia. By shortening the tibia relative to the fibula, the fibula is effectively lengthened, restoring the buttressing effect of the lateral malleolus against dynamic ankle valgus. This addresses the foot and ankle deformity and instability in Paley type 2 FH and prepares the patient for concomitate or future leg lengthening. The SHORDT procedure does produce an acquired leg length discrepancy by the amount shortened that must be accounted for in future limb equalization.

This is common in Paley type 3 FH, where there is fixed equino-valgus and in Paley type 4 FH, where fixed equino-varus is present. The SUPERankle involves fibular anlage resection with a supramalleolar shortening and realignment osteotomy of the tibial and/or a subtalar osteotomy in order to achieve a plantigrade and stable foot and ankle. The

surgical technique and specific SUPERankle variations designed for Paley FH classification type 3, its subtypes and type 4 are described in detail in his publication in 2016 [11].

Since its original description, the SUPERankle procedure has evolved, with the senior author (D.P.) modifying it in 2008 to perform a shortening osteotomy of the distal tibia osteotomy instead, to avoid lengthening tendons. This modification avoided loss of push-off strength and development of a supination midfoot deformity that can occur with weakness that results from lengthening of the Achilles and peroneal tendons, respectively [12].

However, a medial approach is now preferred. Only the distal fibular anlage requires routine resection, and the proximal fibular anlage is not routinely released. Although counterintuitive, as from a medial approach it would seem the tibia would block exposure of the fibular anlage for resection, the interval posterior to the tibia and anterior to the posteromedial neurovascular bundle is easy to expose, leading directly to the fibular anlage. Once excised, the remainder of the procedure involving the tibial osteotomy is much easier to complete through a medial approach than the original lateral approach.

3. Knee Valgus Deformities

This genu valgum can originate in the proximal tibia, distal femur or both. Determining whether the genu valgum originates from the proximal tibia or distal femur is determined by measuring the lateral distal femoral angle (LDFA) and medial proximal tibia angle (MPTA) However, recurrent knee valgus in the proximal tibia is common in FH and likely related to the Cozen phenomenon [13] and repeat hemi-epiphysiodesis may be necessary. In this scenario, recurrent knee valgus in the tibia can be prevented by intentionally deforming the tibia into 10–15° of varus at the end of the lengthening to compensate for the expected rebound valgus.

4. Knee Ligaments

Cruciate ligament hypoplasia or deficiency of the knee is common in FH. The SUPERknee procedure was designed by the senior author as a comprehensive procedure to address the congenital deficiency and deformities in the knee often present in both FH and congenital femoral deficiency (CFD) If the knee instability in FH is symptomatic or if the knee remains subluxated with the knee in full extension, then ligament reconstruction with the SUPERknee using either the iliotibial band or allograft tendon may be necessary. As opposed to femoral lengthening in congenital femoral deficiency, when lengthening the tibia in FH stabilization with the SUPERknee procedure is not mandatory before proceeding with lengthening if the knee does not subluxate in full extension and is otherwise asymptomatic.

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