Correction of Clubfoot Deformity Associated with Weber Type I Tibial Hemimelia Using the Ponseti Method

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Abstract

The clubfoot deformity associated with Weber type I tibial hemimelia, a rare congenital disorder, is rigid and difficult to correct. Surgeons have utilized a variety of treatment methods. Since the 1960s, some adopted the Syme amputation to produce a weightbearing lower limb. Others began to explore alternatives such as the Ilizarov technique, ankle reconstruction, and casting, which salvage the foot but have produced mixed results. The current investigators suggest that the Ponseti method, a minimally invasive technique, can produce a functional weightbearing foot. Two cases were treated with the Ponseti method, including a percutaneous Achilles tenotomy and post-cast bracing. After a minimum of 2-years follow-up, both are ambulatory.

Clubfoot deformity, or talipes equinovarus, can be associated with tibial hemimelia, including hypoplastic tibia, Weber type I (Fig. 1).1 Previous classifications by Jones and colleagues,2 in 1978, and Kalamchi and Dawe,3 in 1985, did not describe this form of tibial hemimelia. Tibial hemimelia is an extremely rare congenital disorder, making it hard to collect sufficient cases to investigate the effectiveness of various treatments. No method to treat the clubfoot deformity associated with tibial hemimelia is universally accepted, since these feet are typically rigid and difficult to correct. Treatments have included amputation, ankle reconstruction, Ilizarov external fixation, casting, soft tissue releases, and osteotomies.

Amputation became increasingly popular in the 1960s and 1970s, because this procedure led to a weightbearing limb. In a case report of a 1-month-old infant with bilateral tibial hemimelia (probably Weber type VII1) and equinovarus deformities, the limb with a small tibial anlage was treated with knee disarticulation, and the limb with a larger tibial anlage was treated with forefoot amputation and fusion of the os calcis to the fibula, performed with future plans of fusing the fibula to the anlage.4 Loder and Herring5 included a case study of a 3-year-old infant with complete absence of the tibia (probably Weber type VII1) and an equinovarus deformity, which had been treated previously with fibular centralization, talectomy, and foot centralization. The recommended treatment was a knee disarticulation, due to the presence of a knee flexion contracture and inadequate foot function.

The Ilizarov method has been used to address the deformities, including equinovarus, associated with tibial hemimelia. Choi and coworkers6 used the Ilizarov method to simultaneously correct the lower leg and clubfoot deformities of a 5-year-old male and an 8-year-old male, both of whom had congenital inferior tibiofibular diastasis (probably Weber type II1). Previously, each patient’s foot had been treated unsuccessfully, one with serial casting (5 years old) and the other (8 years old) with a posterior release. At latest follow-up, the feet of both patients were in a plantigrade position, but ankle motion was limited.

Serial casting has produced limited success in treating clubfoot deformity associated with tibial hemimelia. All of the described cases involved congenital inferior tibiofibular diastasis (probably Weber type II1). In a case with 2-year follow-up, Bajuifer and Letts7 reported correction of a clubfoot deformity during early infancy, using six 1-week
casts, followed by a Denis Browne brace started at 3 months of age, to maintain the correction and manage the internal tibial torsion. Dhammi and associates,8 in another case, that of an 18-month-old infant, after four previous trials of serial casting had failed, a posteromedial soft tissue release was performed with ankle mortise reconstruction to attain a plantigrade foot. At 2 years follow-up, the deformity had not recurred. Wolfgang9 initially treated the equinovarus deformity of a 1-month-old male infant with serial casting and an orthosis. Although the patient was ambulating independently at 11 months of age, the orthosis was ineffective in controlling the progression of equinovarus, resulting in the performance of a modified Syme amputation and the use of a prosthesis. In another case, the previously serially casted bilateral clubfeet deformities of an 8-month-old infant were recast without correction, followed by posteromedial release on one side, posterior release with fibular osteotomy on the other side, and orthoses at 3 years of age.10 At 3-years follow-up, both feet were in plantigrade. Gislanz and colleagues11 performed a Syme ankle disarticulation in a 15-month-old infant after the failure of serial casting and subsequent ankle mortise reconstruction.

The Ponseti method12,13 has gained acceptance over the last 20 years for treating the idiopathic clubfoot.14,15 Recently, others have had some success using this method to treat non-idiopathic clubfoot deformity.16-19 In 18 patients (33 clubfeet) with myelodysplasia, who were treated with casts and percutaneous Achilles tenotomies (PATs), 31 feet required no further treatment; the remaining two had posterior medial releases to correct recurrences.16 Although Wright and coworkers17 reported a 46% (23 feet) recurrence rate in 50 clubfeet associated with neuromuscular and syndromic disorders, only 14 feet needed open surgical releases after repeat casting (repeat Achilles tendon tenotomies in two feet). In two series, after initial correction of the clubfoot of 12 children (24 feet)18 and 10 children (19 feet)19 with arthrogryposis, recurrences occurred in three patients (six feet)18 and four patients (eight feet),19 respectively, and were treated with repeat casting with or without a PAT. Only two feet (8%)18 and four feet (21%)19 within the two series required surgery beyond a PAT at a minimum follow-up of 2 years18 and 13 months (range, 13 to 70 months), respectively.19

We present two cases of Weber type I tibial hemimelia with clubfoot deformity that were corrected using the Ponseti method. At 2-year follow-up, the patients’ feet remained corrected and both children were ambulating independently, one with a brace. We suggest that the Ponseti method may be used to correct the clubfoot deformity and salvage the ankle in Weber type I tibial hemimelia.

Case 1

A 5-day-old male presented with a left clubfoot deformity with Weber type I tibial hemimelia, probable short femur, and an unstable knee. Initial treatment plans focused on correcting the clubfoot deformity with the Ponseti method. Prior to the first manipulation and cast, the patient’s clubfoot was rated using both the Dimeglio-Bensahel10 and Catterall-Pirani21,22 systems, scoring an 18 out of 20 and 6 out of 6, respectively, with higher scores representing a more severe deformity. At each subsequent visit, the foot was scored and recasted. After the 5th cast was removed, a PAT was performed, and a final 3-week cast was applied. Upon removal of the last cast, 2 months after the initial visit, the Dimeglio-Bensahel score was 3 and Catterall-Pirani score was 0.5. After a total of six casts, the patient was given a foot abduction orthosis (FAO) to wear 23 hours per day for
two months, followed by nighttime wear only.

At 1-year follow-up, the patient was beginning to ambulate, the Dimeglio-Bensahel score was 0 and the Catterall-Pirani was 0.5. The patient continued to use the FAO nightly, and a shoe lift was prescribed as a temporary solution for the approximately 2-cm limb-length inequality. No further treatment was required for the clubfoot deformity at this time. At latest follow-up, 2 years post-initial visit, the clubfoot deformity remained corrected, and the patient was ambulating independently. The patient continued wearing the FAO at night to prevent recurrence of the clubfoot deformity. The limb-length inequality of approximately 2.5 to 3 cm in the tibia and 0.5 cm in the femur, knee instability, and bowing will need to be addressed in the future.

Case 2
A 7-month-old female presented with severe right clubfoot deformity (Fig. 2) with Weber type I tibial hemimelia. Plain radiographs demonstrated tibial hypoplasia and clubfoot deformity (Fig. 3). She also had a prominent kyphosis of the lower spine, occult spina bifida, limited abduction of the right hip, and laxity of the right knee. The Ponseti method was started to correct the clubfoot deformity that initially had a Dimeglio-Bensahel score of 17 and Catterall-Pirani score of 5.5. A PAT was performed after the first cast was removed, which is earlier than is typically done with Ponseti casting. After a total of six casts, the patient’s Dimeglio-Bensahel and Catterall-Pirani scores had improved to 2 and 1, respectively. The patient was placed in a knee-ankle-foot orthosis (KAFO), which provided support to her unstable knee, to be worn all day and removed at night. She started ambulating with a KAFO at 15 months of age.

At 11-months follow-up, the family had been noncompliant with using the brace. The patient had a recurrence with a Dimeglio-Bensahel score of 7 and a Catterall-Pirani score of 1.5. The patient was recasted, and, after three casts, she was returned to the KAFO for daytime use and given an ankle foot orthosis (AFO) with dorsiflexion straps (Fig. 4) for nighttime. She ambulated with the KAFO and in-toeing on the right.

At 20-months follow-up, the family was once again found to be noncompliant with bracing. Equinus had recurred, requiring a repeat PAT and recasting. After two casts, the patient resumed using the KAFO with the knee locked in extension. Two months later, a pelvic band was added to control the in-toeing during ambulation, and a 1-cm shoe lift was prescribed to compensate for a 1.5-cm leg-length inequality.

At latest follow-up, 35 months after the initial visit, the...
patient was ambulating independently but with marked in-toeing, primarily due to internal tibial torsion. The child’s parents reported: 1. using the KAFO with pelvic band, which was observed to only minimally improve her in-toeing during the day, inconsistently; 2. not having obtained the prescribed shoe lift; and 3. using the AFO with dorsiflexion straps at night, intermittently. The Dimeglio-Bensahel score was 10 and the Catterall-Pirani was 2.0, indicating a mild residual equinus, valgus, supination, forefoot adductus, cavus, and the presence of abnormal musculature (Fig. 5). She had -5° of passive dorsiflexion, knee instability in all directions, proximal fibular subluxation, and a 1.5 cm shortening. At this time, her foot position remains braceable and does not interfere with her functioning. In the future, additional treatment may be needed to maintain her clubfoot deformity correction and to address her other orthopedic conditions, including internal tibial torsion, knee instability, and limb length inequality.

Discussion

Both of our cases achieved successful initial correction of their clubfoot deformities associated with Weber type I tibial hemimelia using the Ponseti method. Idiopathic clubfoot deformities are considered, by the authors, to have been corrected if they achieve a Dimeglio-Bensahel score of 6 or less and a Catterall-Pirani score of 1.5 or less.15 Upon completion of the casting, case 1 had a Dimeglio-Bensahel score of 3 and a Catterall-Pirani score of 0.5, while case 2 had scores of 2 and 1, respectively. In our second patient, the last Dimeglio-Bensahel score of 10 reflected a mild recurrence. She has gained flexibility in her foot, compared to initial presentation, and is ambulatory.

A PAT was performed in the initial series of casts for both children to expedite the correction process by correcting the equinus deformity, thereby decreasing the total number of casts required.23 As Scher and associates23 had found, 85% of patients with an initial Catterall-Pirani score of 5 or greater had received a tenotomy; it was not unexpected that our cases, with scores of 6.0 and 5.5, would require one. Typically, tenotomies are done prior to the last cast if dorsiflexion is less than 20º. In case 2, a PAT was done after only one cast in order to release the calcaneus from the tibia to increase flexibility about the ankle joint.19 Recently, van Bosse and colleagues19 reported using this modified Ponseti method to correct the rigid clubfoot associated with arthrogryposis, avoiding extensive surgery in the infant and young child.

Compliance with bracing has been found to be an important factor in the maintenance of correction post-casting in the idiopathic clubfoot15,18,24-26 and also may help to prevent recurrence in the clubfoot deformities associated with hemimelia. Of our two cases, one patient was compliant and continued to show improvement in his scores, while the second case was not compliant and had recurrences. As occurred with our case 2, Lehman and colleagues15 reported that recasting and a PAT (or a repeat PAT) after recurrence can once again produce a successfully corrected foot. Bracing after recasting remains essential for maintaining the correction.

As our patients had only a minimum of 2 years of follow-up, continued monitoring will be necessary to determine the long-term effectiveness of the Ponseti method in treating their clubfoot deformity. In the future, their other orthopedic conditions, such as limb-length inequality, will need to be addressed, probably surgically. Following these patients

Figure 5 At latest follow-up, case 2 (A) in supine and (B) in standing, showing right internal tibial torsion and mild recurrence of clubfoot deformity.
through these procedures will be important to observe for changes in the foot. With lengthening, recurrent equinus is possible and repeat tenotomies may be required. Also, as the patient grows, the soft tissue and bony structures may require further treatment.

Clubfoot deformity associated with tibial hemimelia is typically rigid and difficult to correct. Of the seven types of tibial hemimelia defined by the Weber classification, we report here the treatment of clubfoot deformity associated with type I, the mildest form. To our knowledge, this is the first report of casting with this type of tibial hemimelia and the only report of the use of the Ponseti method with tibial hemimelia. Further studies need to include other Weber types of tibial hemimelia to evaluate the effectiveness of the Ponseti method in the treatment of the associated clubfoot deformity in conjunction with other treatments, such as the Ilizarov, to correct the tibial deformity.

Conclusion

Clubfoot deformities associated with Weber type I tibial hemimelia can be corrected with the Ponseti method. Using the minimally invasive Ponseti method provided a functional weightbearing foot for ambulation in our two cases, but they will need to be monitored as each child grows.

Disclosure Statement

None of the authors have a financial or proprietary interest in the subject matter or materials discussed, including, but not limited to, employment, consultancies, stock ownership, honoraria, and paid expert testimony.

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