

Riley J Rampton, DPM¹; Daniel J Hatch, DPM, FACFAS²; Meghan Hurley, DPM³; Aniela Cordoba, DPM⁴

¹PGY 3 Banner North Colorado Medical Center, ²Foot and Ankle Center of North Colorado, ³Table Mountain Foot and Ankle, ⁴Summerlin Foot and Ankle

Introduction

Charcot Marie Tooth (CMT) disease is a progressive neuromuscular disorder that is often the underlying condition involved in the development of a symptomatic cavovarus foot. This foot deformity is usually corrected with a triple arthrodesis. This case-series reviews several mid-stage cavovarus deformities in which a realignment subtalar joint arthrodesis was utilized to improve Kite's angle, reduce heel varus and re-establish subtalar joint instability to prevent the need for a triple arthrodesis.

Literature Review

One of the more common etiologies of cavus foot deformity is an underlying neuromuscular disorder, and of those, greater than 50% have CMT. CMT is a progressive disease that usually involves an imbalance of the antagonistic muscles inserting into the foot which results in a complex cavovarus deformity(1). Nonoperative treatment of the disease is recognized as producing unsatisfactory results (2). Reconstructive surgery is generally complicated and routinely staged. Depending on the stage and severity of deformity, procedures can consist of tendon transfer/lengthening procedures, osteotomies, arthrodesis or a combination of procedures (3). Although there is little evidence in the literature directly comparing various surgical options, recent literature favors joint preservation surgery. This is due to poor long-term results of patients who have undergone a triple arthrodesis in childhood due to CMT. Wukich and Bowen reported that in 22 patients with severe pes cavovarus secondary to CMT who underwent triple arthrodesis 24% of ankles and 62% of feet demonstrated evidence of degenerative joint disease. However, 86% of these patients were satisfied with their result. Wetmore and Drennan reported less satisfactory results with only 24% of 16 patients reporting good or excellent results and 6 patients required ankle arthrodesis.

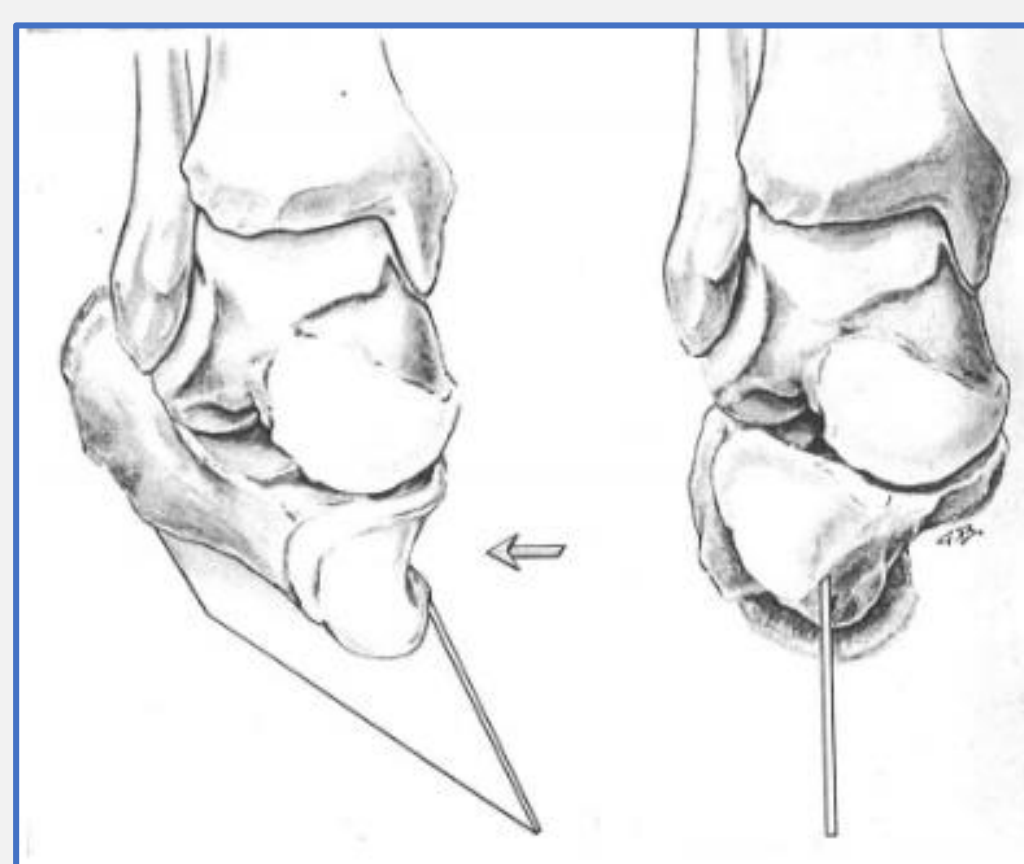


Figure 1. Reduction of deformity by using Ponseti biomechanical principles.

Case Presentation

This case series reviews mid-stage cavovarus progression in 3 patients with pedal instability and gait disturbances secondary to CMT disease. In each case, tri-planar correction of the deformity was achieved with a STJ arthrodesis. Pre-operative and postoperative radiographs were compared as well as functional outcome.

Case number one is a 37 year old male with CMT who presented with significant weakness and gait instability. He was not able to wear ankle foot orthoses (AFO) due to the amount of deformity and subsequent abrasions. Clinical evaluation revealed bilateral severe hammertoe deformities, plantarly deviated first rays and varus heel positions. To address the right foot deformity, the following procedures were performed: Jones tenosuspension with hallux interphalangeal joint fusion, dorsiflexory 1st tarsometatarsal joint arthrodesis, hammertoe repair by fusion of the proximal interphalangeal joint (PIPJ), tibialis posterior tendon transfer, subtalar joint realignment arthrodesis and tendo-achilles lengthening (TAL). (see figures 2 and 3) Note the improvement in Kite's angle on the AP and lateral radiographs of the right foot versus the left. Additionally note the significantly improved calcaneal alignment in the postoperative long leg axial views.



Figure 2. Preoperative radiographs, right foot



Figure 3. Post-operative radiographs, right foot

Case Presentation

Case number 2 is a 53-year-old male with CMT presenting with bilateral cavovarus foot deformities.(Figure 4) He especially complained of recurrent instability of the left ankle. Dynamic evaluation with fluoroscopy demonstrated significant subtalar joint instability. Therefore, subtalar realignment arthrodesis was performed along with: 1st tarsal metatarsal joint dorsiflexory arthrodesis; tibialis posterior tendon transfer, plantar fascial release and TAL bilaterally. (figure 5).



Figure 4. Preoperative axial views and stress of ankle and subtalar joints. Note lateral gap of STJ.

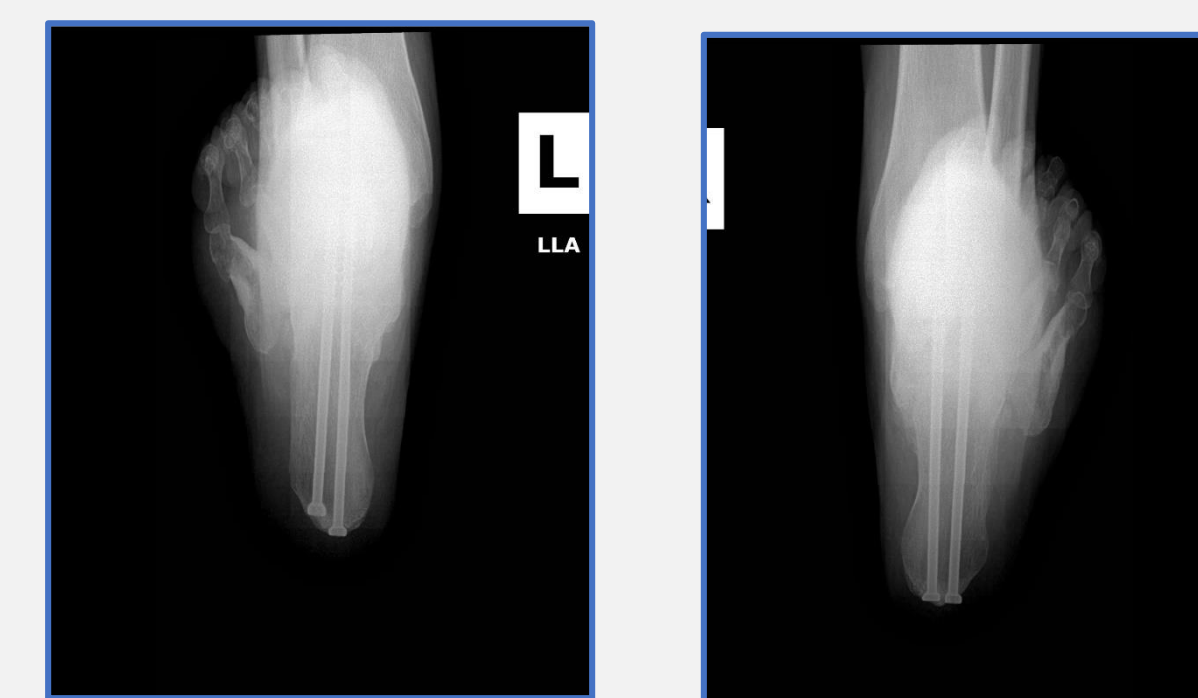


Figure 5. Postoperative radiographs.

Case 3 is a 29-year-old male who developed bilateral cavovarus foot deformities secondary to CMT. (figure 6) In order to correct the left foot we performed a tibialis posterior tendon transfer, Dwyer calcaneal osteotomy, peroneus longus to brevis tendon transfer, plantar fasciotomy and gastrocnemius recession. (figure 7) Note in the AP projection post operative the poor alignment of the talar calcaneal joint (kite's angle). Additionally in the axial views, note the lack of calcaneal alignment to the distal tibial axis.



Figure 6. Preoperative radiographs



Figure 7. Postoperative radiographs.

Discussion

The cases presented displayed mild to late stage cavovarus deformities without evidence of arthritic involvement. The consideration of STJ realignment arthrodesis is multifactorial. While all joints are involved in the rearfoot complex in CMT; we advocate that arthrodesis of the subtalar joint offers an ideal apex from which to address the triplane deformity while preserving some midtarsal joint motion. In order to appropriately correct a deformity, we support Paley's theory of a three-dimensional approach that allows for correction at the CORA. In this case, we are addressing the mechanical axis of the subtalar joint. Most studies evaluate this condition in two dimensions therefore a common recommendation to address heel varus is a calcaneal osteotomy. The varus heel position commonly seen in CMT patients is notably similar to the calcaneal position in patients with clubfoot deformity. When we apply the biomechanical principles as advocated by Ponseti, the deformity will reduce during surgical repair. (Figure 1) During subtalar realignment, the beak of the calcaneus is abducted, dorsiflexed and rotated about the subtalar axis producing a valgus positioning of the calcaneal tuber along with improvement of kite's angle.

Conclusions

All three patients related good functional outcomes and increased stability after a minimum 5 year follow up. Realignment arthrodesis of the subtalar joint offers improved talocalcaneal alignment, calcaneal re-positioning and stability in the mid to late stage CMT patient.

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Contact

Riley J Rampton DPM
Email: riley.rampton@gmail.com
Phone: 801-505-3463