Surgical Treatment of Charcot-Marie-Tooth Neuropathy X

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Purpose

The purpose of this study was to present a novel surgical approach to the treatment of drop foot and neuro muscular hypertrophy in Charcot-Marie-Tooth disease Type X. The approach used by the authors consisted of combining rigid osseous correction with an adjustable treatment to lessen corrective soft tissue procedures.

Background

Charcot-Marie-Tooth neuropathy Type X (CMTX) is characterized by a more severe motor and sensory neuropathy, male predominance, and tend to be symptomatic while females are carriers of the gene [1]. To address digital contracture and cavus foot type isolated hallux is the most common description of treatment for Type X contractures as well as with drop foot symptoms and gait disturbances [2]. Combination procedures have been described [3], however there is little in the literature that describes significant osseous stabilization to increase effectiveness of known soft tissue procedures. Multiple studies have described the benefit of physical therapy and bracing [4,5] however few have shown this benefit in addition to surgery. Among the numerous approaches, the posterior tibial tendon transfer has been the mainstay in treatment of drop foot, which is the most gait modifying deformity of advanced CMTX [6].

The patient in this study has an axonal dominant form of Charcot-Marie-Tooth disease (Type X) and presented with difficulty ambulating and pain in his left foot. Symptoms included range of motion and strength in the foot and ankle, steppage gait, claw toes, and cavovarus foot deformity resulting in overlapping the metatarsus along the fifth ray accompanied by painful/hyperkeratotic lesions. The patient had suffered from an inversion avulsion fracture of the left ankle, which was successfully treated with conservative care. The patient displayed no anterior drawer instability, but had inversion stress instability about the subtalar joint and hindfoot.

The preoperative exam of the patient revealed muscle weakness, including decreased strength in all lower extremity muscles with the exception of small hand weakness of tibialis posterior. Preoperative planning discussed that the presence of a plantarflexed first metatarsal associated with increased power and minimal deformity. Therefore, a more advantageous and improved mechanical transfer in the foot [7,8] outweighing the pure plantarflexor caused by a stronger peroneus longus causes plantarflexion of the first metatarsal and anterior pes present in a varus foot. The hyperpronated foot is an internal rotation at the tibia and tarsal hyponatrotic is the primary complaint of the patient was his steppage gait and as a result a tibial posterior tendon transfer was recommended and performed through the tibial tendinous perforation of the lateral ankle, which was superiorly overpowered by a stronger peroneus longus causing plantarflexion of the first metatarsal and anterior peroneus longus [9]. Our patient presented with a classic weak peroneus brevis and anterior tibial tendon, a relatively stronger tibialis posterior causing adduction of the forefoot and inversion of the hindfoot [7]. In addition to the anterior being overpowered by a stronger peroneus longus causing plantarflexion of the first metatarsal and anterior peroneus longus [9]. Our patient presented with a classic weak peroneus brevis and anterior tibial tendon, a relatively stronger tibialis posterior causing adduction of the forefoot and inversion of the hindfoot [7].

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Methods

X-ray eval

Pre Op Angles

Post Op Angles

Calcaneal

Inclination

Simmons(AP)

Kites(AP)

Meary's

(19.4)

10

12.2

14.8

15.7

15.4

12.7

16.8

Results

1. Triple arthrodesis
2. Distal ray osteotomy, first metatarsal
3. Hallux interphalangeal joint fusion
4. Proximal interphalangeal joint fusion second, third, fourth toe
5. Achilles tendon lengthening
6. Tibialis posterior tendon transfer to medial cuneiform
7. Hilde lenis suspension

Case Study

A 37 year-old male with a past medical history of CMTXII with attempted surgical correction as a child, presented initially in February 2015 for surgical consultation for his left lower extremity deformity. This case study demonstrates a unique combination of osseous and collateral procedures including tarsometatarsal, calcaneal, and digital tendinous osteotomy and multiple tendon transfers to achieve stability and improve function. The patient has 18 months of follow up, and reported improvement of function, decreased pain, and the patient was completely satisfied with the outcome at final follow up in August 2016.

Discussion

The hyperpronated foot is an internal rotation at the tibia and tarsal hyponatrotic is the primary complaint of the patient was his steppage gait and as a result a tibial posterior tendon transfer was recommended and performed through the tibial tendinous perforation of the lateral ankle, which was superiorly overpowered by a stronger peroneus longus causing plantarflexion of the first metatarsal and anterior peroneus longus [9]. Our patient presented with a classic weak peroneus brevis and anterior tibial tendon, a relatively stronger tibialis posterior causing adduction of the forefoot and inversion of the hindfoot [7].

The purpose of this study was to present a novel surgical approach to the treatment of drop foot and neuro muscular hypertrophy in Charcot-Marie-Tooth disease Type X. The approach used by the authors consisted of combining rigid osseous correction with an adjustable treatment to lessen corrective soft tissue procedures.

For a prospective gait video scan the QR code

References