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# Surgical Management of Pes Cavus Deformity with an Underlying Neurological Disorder: A Case Presentation

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ABSTRACT

Charcot-Marie-Tooth disease is a complex group of motor and sensory disorders presenting with varying levels of deformity dependent on the chronology and specific subgroup of the disease. In this report, we discuss a 19-year-old man with Charcot-Marie-Tooth 1A, a progressive and aggressive form of hereditary sensorimotor neuropathy, with rigid forefoot and rearfoot deformity. The authors discuss the etiology, tests, and sequential surgical management of this condition, focusing on a triple arthrodesis including a closingwedge subtalar joint fusion and a dorsal closing wedge osteotomy of the first metatarsal.



The surgical management of patients presenting with a pes cavus deformity can be challenging for any professional because of the underlying complex mechanisms of deformity. The degree of pathology and morbidity is unique to each patient, and the clinical presentation can vary from advanced digital deformities to plantar buckling of the metatarsophalangeal joints, with associated painful plantar callosities and cutaneous ulceration. If the condition progresses, lateral ankle instability can have a profound impact on gait and lower limb stability (1). When planning any surgical intervention, it is important to determine the following: 1) whether an underlying neurological condition exists, because this is central to the prognosis and therefore management; 2) the keystone or apex of the deformity; and 3) which components are a reflection of compensation rather than primary pathology. In this report, we present a case study describing a 2-stage surgical course of management of pes cavus in a patient with an underlying neurological disease, which highlights the aforementioned considerations and processes.

## Overview of Pes Cavus

Pes cavus is a descriptive term and not a specific diagnosis (2). In its simplest form, pes cavus is characterized by an excessively high medial longitudinal arch, an inverted rearfoot, and an associated true or pseudo ankle equinus (3). The majority of patients have an underlying neurological component, and initially the deformity occurs in the sagittal plane with increasing degrees of plantarflexion of the forefoot on the rearfoot (4). McGlamry and Kitting (5) first coined this presentation with the term forefoot (anterior) equinus, to describe the plantarflexed attitude of the forefoot on the rearfoot. They further subdivided this into 4 types based on the apex of the deformity. Whitney and Green (6) further described this forefoot attitude as pseudoequinus, to differentiate it from a traditional rearfoot or ankle equinus.

If the condition progresses, as a result of deterioration associated with a neurological presentation, the peroneus longus muscle tends to overpower the weakened tibialis anterior muscle, thereby creating frontal plane inversion as the first ray plantarflexes below the plane of the lateral metatarsal heads. This results in increasing degrees of rearfoot supination and frequently concomitant lateral ankle instability. Ultimately, transverse plane metatarsus adductus occurs because of the mechanical advantage of the strong tibialis posterior muscle over the weak peroneus brevis. This, in concert with the other planal components, results in the failure of the foot to function in a plantigrade position (6–9). Mann and Missirian (10) further commented that muscle imbalance between the evertors and invertors of the foot was the underlying cause for the pes cavus deformity in Charcot-Marie-Tooth (CMT) disease.

## Etiology of Pes Cavus

Historically, there have been many hypotheses as to the etiology of pes cavus (11). However, in a study by Brewerton et al (12), who

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reviewed 77 patients with a pes cavus deformity, 51 (66%) had clinical evidence of neurological involvement. This increased to an overall 75% after electromyographic studies. The remaining 25% were felt to have had an idiopathic cause to their conditions. Ibrahim (13) subsequently classified the etiologies of pes cavus deformity into neurological, congenital, and traumatic origins (Table 1). Of all of the underlying neurological causes for pes cavus deformity, CMT is the most common neurological disorder (14). CMT has been further subdivided into 4 classes (Table 2), the most significant of which is primary or CMT1, which represents the most progressive form and exhibits slow nerve-conduction velocities (NCV); and secondary or CMT2, which tends to be more indolent, with normal NCV. This opinion is further supported by the work by Krajewski et al (14). Advances have been made over the past decade with a clearer understanding of the molecular basis of several forms of CMT disease, via genetic blood screening. This has increased the number of patients diagnosed with an underlying neurological component rather than being labeled as having idiopathic pes cavus (15, 16). It is our belief that only with a thorough understanding of these spectra of disorders and presentations can appropriate surgical planning and management be instigated.

## Case Report

A 19-year-old man, who complained of painful feet and difficulty walking, was referred to our department by his general practitioner. He reported that his feet had not grown since the age of 13 years, although his pedal arches had increased in height. Physical examination revealed bilateral pes cavus with retraction of the lesser digits and rigid plantarflexed first rays with triggered great toes. His hindfoot assumed an unstable, uncompensated varus position bilaterally. The Coleman (8) block test, which is based on the principle that when the lateral foot and heel are positioned on the block, the first ray is off loaded, showed that when the plantarflexed first ray was compensated, the patient's rearfoot remained in a varus position. The lower limb musculature appeared underdeveloped, and to a lesser extent his hands displayed wasting of the intrinsic muscles. On further questioning, the patient reported that his father had similar foot deformities. His patella reflexes were absent, no plantar response (Babinski) was established, and vibration perception was reduced throughout the lower limb. Manual muscle power testing, using the Medical Research Council's Oxford scale (17), was graded as dorsiflexion 4, plantarflexion 5, inversion 5, and eversion 2, bilaterally. Weight-bearing plain radiographs revealed an increased calcaneal inclination angle (38.2°) and a correspondingly high forefoot (metatarsal-talar) declination angle (62.9°). The dorsoplantar radiograph showed substantially increased metatarsus adductus (Figure 1).

**Table 1**
Etiology of pes cavus*

| Class | Cause |
|---|---|
| I—Neuromuscular | |
| a) Muscle disease | Muscular dystrophy |
| b) Afflictions of peripheral nerves and lumbosacral spinal nerve roots | Charcot-Marie-Tooth disease, spinal dysraphism, polyneuritis, intraspinal tumor |
| c) Anterior horn cell disease of the spinal cord | Poliomyelitis, spinal dysraphism, diastematomyelia, syringomyelia, spinal cord tumor, spinal musculature atrophy |
| d) Long tract and central disease | Friedreich's ataxia, Roussy-Levy syndrome, primary cerebellar disease, cerebral palsy |
| II—Congenital | Idiopathic cavus foot, residuum of clubfoot, arthrogryposis |
| III—Traumatic | Residua of compartment syndrome, severe burn, fracture malunion |

* From: Ibrahim K. Pes cavus. In *Surgery of the Musculoskeletal System*, p 4015, edited by CM Evarts, Churchill Livingstone, New York, 1990. Reprinted by permission of Oxford University Press.

**Table 2**
Classification of Charcot-Marie-Tooth disease*

| Class | Features and Subgroups |
|---|---|
| CMT1 | |
| Approximately 50% of cases, autosomal dominant, NCV range 10 to 30 m/s | CMT1a: Most common form, accounting for 80% of CMT1, associated with segmental trisomy for peripheral myelin protein 22 (PMP-22) on chromosome 17<br>CMT1b: Accounts for 5% to 10% of CMT1, associated with point mutation in myelin $P_o$ gene, phenotype display aggressive degeneration<br>CMT1c: Less common, genetic defect remains unknown |
| CMT2 | Approximately 20% of cases, autosomal dominant, indolent, NCV near normal, no demyelination, 4 separate chromosomal loci identified, no offending proteins identified |
| CMT3 | X-linked, inherited in males |

Abbreviations: CMT, Charcot-Marie-Tooth; NCV, nerve conduction velocity; m/s, meters per second.

* Adapted from Krajewski KM, Lewis RA, Fuerst DR, Turansky C. Neurological dysfunction and axonal degeneration in Charcot–Marie–Tooth disease type 1A. Brain 123:1516–1527, 2000.

Based on the aforementioned findings, a primary diagnosis of acquired pes cavus deformity with an underlying neurological deficit was made, and a referral to the consultant neurologist was expedited. The neurologist confirmed peripheral muscle weakness, wasting of both hands and feet with areflexia, and abnormal sensation. There were no other neurological symptoms of note, notably no visual disturbances, speech impediments, dysphagia, or hearing loss. NCV and amplitude of the motor responses tests revealed reduced peripheral responses and absent sensory potentials (Table 3). Blood tests confirmed chromosome 17p 11.2 duplication. These findings were consistent with hereditary motor sensory neuropathy type 1 (CMT1a) with over 70% locus and allelic heterogeneity (18). The hallmark of this more progressive form of CMT disease often shows electrical evidence of nerve dysfunction as early as 2 years of age with selective tibialis anterior muscle involvement, while the extensor hallucis longus muscle is often initially spared. Within the lateral muscle compartment of the leg, the peroneus brevis muscle is usually the more severely affected skeletal muscle, whereas the peroneus longus muscle is usually initially spared (19). After preparing the patient, the decision was made to pursue surgical intervention to correct the debilitating pes cavus deformity.

### *Surgical Procedures—Operation 1*

The patient identified the right foot as his most unstable, and it presented with a triggered hallux and associated interphalangeal joint pain, a rigid plantarflexed first ray, rigid rearfoot varus, and a forefoot equinus. The patient's chief concerns were with the triggered hallux being repetitively irritated by footwear and difficulty walking or standing for periods greater than 2 to 3 hours, especially when unshod. We opted for a 2-staged approach to correct the misalignment, placing primary attention on addressing the lateral instability with a triple arthrodesis and a closing wedge osteotomy to elevate the first ray.

The operation was performed with the patient under general anesthesia as a day case. The patient was positioned supine with a thigh tourniquet. In concert, a popliteal nerve block was administered for postoperative pain management. Two incisions were used to