NEUROMUSCULAR DISEASE AS THE CAUSE OF LATE CLUBFOOT RELAPSES: REPORT OF 4 CASES

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ABSTRACT

Following correction with the Ponseti method some idiopathic clubfeet still will relapse even after six years of age. A better understanding of the cause for these late relapses will greatly help in the management of this condition. We evaluated a consecutive case-series from 1948 through December 1984 including 209 patients (321 clubfeet). Patients were treated following the Ponseti method. Initial number of casts, age at relapse, neurological evaluation, and final treatment for the late-relapses were recorded. There were 12 patients (6%) having a relapse after the seventh birthday. In 4 of these patients (6 clubfeet) a neuromuscular disease was diagnosed, representing 33% of the late relapses. These patients were initially treated with an average of 4 casts (range: 2-6) with 2 requiring an Achilles tenotomy. Patients used the brace for an average of 4 years. The average age at the relapse prior to the suspicion of neuromuscular disease was 9 years (range: 8-11 years). Two patients had family history of neuromuscular disease (myotonic dystrophy and multiple core disease). In the other two cases (Charcot-Marie-Tooth Disease type IA and myasthenia gravis) neuromuscular disease was not suspected. All four patients required an anterior tibialis transfer, three had a plantar fasciotomy, and two had peroneus longus to brevis transfers. One patient required a subsequent posterior tibialis transfer and another patient a triple arthrodesis (myotonic dystrophy). In conclusion, late relapses in patients with idiopathic clubfoot may represent the onset of a previously undiagnosed neuromuscular disease, and should be thoroughly evaluated.

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INTRODUCTION

A small number of clubfeet treated at our institution relapsed after six years of age following correction with the Ponseti method. Interestingly, several neuromuscular diseases may imitate a clubfoot relapse by causing cavus and equinus deformities. A late relapse may represent the onset of a previously undiagnosed neuromuscular disease as suggested by the following four cases.

METHODS

Consecutive case-series from 1948 through December 1984 including 209 patients (321 clubfeet). Patients were all treated following the Ponseti method. Of these patients there were 13 patients (19 feet) noted to have a clubfoot relapse after the age of six years. Four of these patients (6 clubfeet) were diagnosed with a neuromuscular disease after their clubfoot relapse. No other cases have had a neuromuscular disease diagnosis. Initial number of casts, age at relapse, neurological evaluation, and final treatment for the relapse were recorded (Table 1).

CASE 1

Patient presented as a 3.5 month old white female, product of normal delivery. Pregnancy was uneventful except for a cold during 2-3 month. Baby was otherwise normal at birth except for a right clubfoot deformity. Subsequently, the child was found to have a ventricular septal defect at 3 months of age. Neurologic and neuromuscular exams were normal at this time. Clubfoot casting at an outside institution was started shortly after birth and involved 7 casts before referral to our institution at the age of 3.6 months. She was treated with 4 casts and a tenotomy resulting in 10 degrees of dorsiflexion. The brace was consistently used until the age of six and a half years. A year after discontinuing the brace she had her first recurrence consisting of a tight heelcord and mild supination during gait which was treated with a tendoAchilles lengthening and an anterior tibialis tendon transfer to the 3rd cuneiform. Four months later it was felt that she had an excellent result, with the exception of tight heelcord. A year later, she had begun to slump forward, was unable to sit with her legs outstretched due to tight hamstrings, and developed a progressive equinus deformity on the right foot. She has a history of chronic constipation, occasionally soiling herself, but

TABLE 1

Side	Age Initial visit (weeks)	# of Casts	Tenotomy	Age Relapse 1	Rx	Age Relapse 2	Rx	Age Relapse 3	Age Last F/U	Neuromuscular Disease
R	0	4	yes	7.4	ATT, TAL	8.8	TAL, PFR	11.4	28	Charcot-Marie-Tooth Disease Type IA
L	250	2 and ATT	yes	6.1	PTS, PFR, PLB LTEM	7.6	PTT		17	Multiple Core Disesase
R	8	6	yes	4.3	3 casts	9.6	ATT		20	Myasthenia gravis
L	8	6	yes	4.3	3 casts	9.6	ATT, TAL		20	
R	16	3	no	10.9	PLB, PFR, 1MO, ATT, Triple				17	Myotonic Dystrophy
L	16	3	no	10.9	PLB, PFR, 1MO, ATT				17	
R	4	4	yes	5.9	ATT	9.7	Triple		40	
L	4	4	yes	5.9	ATT	9.7	Triple		40	
L	21	4	yes	9.4	ATT				17	
R	1	10	no	11	MR				11	
L	1	10	no	11	MR, CCO				11	
R	4	12	yes	8.5	Cast, TAL, PC, PFR, ATT, LTEM				44	
L	4	12	yes	8.5	Cast, MB, MR, PTL, FDCL, ATT				44	
R	1	5	yes	6.4	ATT, TAL				17	
L	1	5	yes	8.1	ATT				17	
R	4	8	no	7.4	ATT, TAL				18	
L	4	8	no	7.4	ATT, TAL				18	
R	4	7	yes	5.1	6 casts	8.3	ATT		39	
L	4	7	yes	5.1	6 casts	8.3	ATT		39	
L	2	4	yes	8.2	-	10.5	ATT, TAL		30	

ATT = Anterior Tibialis Transfer, TAL = Tendo Achilles Lengthening, Triple = Triple Arthrodesis, PLB = peroneus longus to brevis transfer, PFR = Plantar Fascia Release, 1MO = First metatarsal Osteotomies with recession of extensor hallucis longus, PTS = Posterior Tibialis Slide, LTEM = Transfer of the large toe extensor to the metatarsal neck, PTT = Posterior tibialis transfer, MR = Medial Release, CCO = Calcaneo-cuboid osteotomy, PC = Posterior Capsulotomy, PTL = Posterior tibialis lengthening, MB = Modified Brockman (postero-medial release), FDCL = Flexor digitorum communis lengthening.

had no urinary incontinence. She had a rectal sphincter with 50% normal tone. Motor strength was symmetrical at 4+. There were no muscle fasciculations or wasting. No sensory deficit was observed. Deep tendon reflexes demonstrated: biceps 0, triceps 2+, brachioradialis 2+, patellar 0, ankle 2+, with no clonus. She was diagnosed with Dejerine-Sottas Disease (Hereditary Hypertrophic Polyneuritis / Charcot-Marie-Tooth Disease Type IA). At the age of 11 years she had a right Achilles tendon lengthening with a percutaneous plantar fasciotomy for right equinocavus foot. At 24 years of age she still had 5 degrees of equinus, weakness of her dorsiflexors, and marked clawing of all toes. She underwent a left total hip replacement at the age of 26 due to early onset degenerative joint disease of unknown etiology.

CASE 2

Patient with a unilateral left clubfoot and a positive family history of clubfoot was initially treated elsewhere with casts and a tenotomy followed later by an Achilles tendon lengthening at the age of two. He presented to our institution at nearly five years of age for clubfoot deformity. He required two casts to obtain correction and had a left anterior tibialis transfer to the 3rd cuneiform, and the foot abduction brace was started. He wore the brace faithfully, but had a recurrence at the age of 6, involving a varus foot, which was corrected with a left posterior tibialis slide. A year later he had a left plantar fasciotomy, peroneus longus to brevis transfer, transfer of the large toe extensor to the metatarsal neck for pes cavus deformity. Nine months after this surgery he was diagnosed with the same type muscular dystrophy as his mother. On examination of strength, he has grade 3/5 anterior tibialis, 2/5 peronei, 4/5 extensor digitorum communis, and 0/5 extensor hallucis longus activity. He had marked, fixed heel varus. At the age of 8.5 years he underwent posterior tibialis transfer through the interosseus membrane to the cuboid. At 20 years of age, it was determined he had multiple core disease by muscle biopsy. Several of his relatives had biopsy-confirmed multiple core disease as well. In his last follow up at age of 21, he was working as a security guard and walking approximately 7 miles per day, without evidence of further progression of his neuromuscular disease.

CASE 3

Female patient with bilateral clubfoot initially seen at our institution at the age of 8 weeks. Clubfeet were corrected with 6 casts and an Achilles tenotomy, after which she began to use the Mitchell brace. Brace wear was not consistent and she had a relapse at the age of 4 years, which was corrected with 3 additional casts.

The brace was started again and worn until she was 8.5 years of age. One year after stopping the brace she had another relapse with forefoot supination and tight heelcords treated with left tendoAchilles lengthening and bilateral anterior tibialis tendon transfer to 3rd cuneiform. Two and a half years later she was diagnosed with myasthenia gravis. No further clubfoot treatment has been required.

CASE 4

Patient with bilateral clubfeet and a medical history significant for mitral valve prolapse, a large left parietal cephalohematoma, and mental retardation. Treatment was started at 16 weeks of age and consisted of 3 casts, followed by bracing until she was four years old. She developed a recurrence at the age of 11 years consisting of bilateral cavovarus feet without significant heelcord tightness. She was diagnosed with myotonic dystrophy by electromyography. At the age of 12 years she had surgery consisting of bilateral peroneus longus to brevis transfers, plantar fascia releases, 1st metatarsal osteotomies with recession of the extensor hallucis longus, and anterior tibialis tendon transfer to the 3rd cuneiform. At 13 years of age she had a right triple arthrodesis to correct persistent, progressive varus and cavus.

DISCUSSION

For the first few years after full correction of a clubfoot during childhood, there is a tendency for relapse. However, it is very rare to observe a relapse after age 6. Unfortunately, there are no clinical criteria that will help in the differentiation of these cases.

In this study we evaluated all patients treated under Dr. Ponseti until 1984, and we found that 13 cases (6%) had a relapse after 7 years of age. Evaluating these patients it was observed that 4 (31%) had a diagnosis of a neuromuscular disease, suggesting that this may have caused the relapse.

Initial correction of the deformity was easily accomplished in all cases with a few casts and the patients were mostly compliant with the bracing protocol. Interestingly, patients wore the brace until 4 years of age or older; a time that has usually been considered enough to prevent relapses.[1,2] In 3 cases the neuromuscular disease was suspected early after the relpase and the patient was referred for a neuromuscular examination immediately. However, in the 4th case (myasthenia gravis) neuromuscular disease was not suspected until 2.5 years later. The relapse that immediately preceded the diagnosis of neuromuscular disease occurred at an average age of 9.2 years (range: 7.6 to 10.9 years). Neuromuscular disease was diagnosed an average age of 9.9 years (range: 7.6 to 12.1 years).

The neuromuscular diseases represented were myotonic dystrophy, myasthenia gravis, multiple core disease and Charcot-Marie-Tooth Type IA. Myotonic dystrophy has been associated with clubfoot deformities and Charcot-Marie-Tooth disease has been associated with foot drop and inversion.[3-5] Myasthenia gravis, while classically affecting the bulbar muscles and proximal limbs, is occasionally associated with distal limb weakness and fatigability.[6] Clubfoot deformity has been associated with multiple core disease, scoliosis and joint contractures.[7]

In conclusion, late relapses in patients with idiopathic clubfoot may represent the onset of a previously undiagnosed neuromuscular disease, and should be thoroughly evaluated.

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