Long-Term Results of Reconstruction for Treatment of a Flexible Cavovarus Foot in Charcot-Marie-Tooth Disease

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Background: Cavovarus foot deformity is common in patients with Charcot-Marie-Tooth disease. Multiple surgical reconstructive procedures have been described, but few authors have reported long-term results. The purpose of this study was to evaluate the long-term results of an algorithmic approach to reconstruction for the treatment of a cavovarus foot in these patients.

Methods: We evaluated twenty-five consecutive patients with Charcot-Marie-Tooth disease and cavovarus foot deformity (forty-one feet) who had undergone, between 1970 and 1994, a reconstruction consisting of dorsiflexion osteotomy of the first metatarsal, transfer of the peroneus longus to the peroneus brevis, plantar fascia release, transfer of the extensor hallucis longus to the neck of the first metatarsal, and in selected cases transfer of the tibialis anterior tendon to the lateral cuneiform. Each patient completed standardized outcome questionnaires (the Short Form-36 [SF-36] and Foot Function Index [FFI]). Radiographs were evaluated to assess alignment and degenerative arthritis, and gait analysis was performed. The mean age at the time of follow-up was 41.5 years, and the mean duration of follow-up was 26.1 years.

Results: Correction of the cavus deformity was well maintained, although most patients had some recurrence of hindfoot varus as seen on radiographic examination. The patients had a lower mean SF-36 physical component score than age-matched norms, and the women had a lower mean SF-36 physical component score than the men, although this difference was not significant. Smokers had lower mean SF-36 scores and significantly higher mean FFI pain, disability, and activity limitation subscores (p < 0.0001). Seven patients (eight feet) underwent a total of eleven subsequent foot or ankle operations, but no patient required a triple arthrodesis. Moderate-to-severe osteoarthritis was observed in eleven feet. With the numbers studied, the age at surgery, age at the time of follow-up, and body mass index were not noted to have a significant correlation with the SF-36 or FFI scores.

Conclusions: Use of the described soft-tissue procedures and first metatarsal osteotomy to correct cavovarus foot deformity results in lower rates of degenerative changes and reoperations as compared with those reported at the time of long-term follow-up of patients treated with triple arthrodesis.

Level of Evidence: Therapeutic Level IV. See Instructions to Authors for a complete description of levels of evidence.

C avovarus foot deformity negatively impacts a patient's quality of life and inhibits his or her ability to perform activities of daily living. Approximately 80% of cavovarus foot deformities can be attributed to a neurologic disorder. The most common cause is Charcot-Marie-Tooth disease (also known as *hereditary motor sensory neuropathy Type 1*), which is estimated to affect one in 2000 individuals in the United States[1]. Although the natural history of cavovarus foot deformity is not well documented, many authors have agreed that, in most patients with Charcot-Marie-Tooth disease, a flexible deformity of the foot develops during childhood or adolescence and gradually progresses to a fixed deformity[2-7]. Cavovarus deformity leads

to pain and callus formation under the metatarsal heads, foot fatigue, difficulty wearing normal shoes, lateral ankle instability, and tripping. An abnormal gait, with a tripod pattern of weight-bearing on the heel and first and fifth metatarsal heads and a toe-heel gait-line pattern, often develops[7-9].

A wide variety of surgical procedures have been described for the treatment of cavovarus foot deformity, and varying degrees of success have been reported. These procedures include plantar release[4,5,10,11], calcaneal osteotomy[2,4,12,13], metatarsal osteotomies[2,4,11,14-16], tarsometatarsal osteotomies[6,17], tarsal osteotomy[18,19], and various tendon transfers[15,20,21]. Typically, patients undergo any number of soft-tissue procedures while the deformity is flexible and additional osseous procedures when the deformity becomes rigid. Despite the many reports on cavovarus foot deformity, few authors have described the long-term results of surgical procedures. In the three studies in which patients were followed for more than five years, no single procedure or combination of procedures was found to offer consistently good long-term results[19,22,23]. In the past, many patients with Charcot-Marie-Tooth disease and cavovarus foot deformity have undergone triple arthrodesis, which was previously considered to be a definitive procedure to create a well-aligned, functional foot[15,23-25]. However, long-term follow-up studies have shown a high incidence of osteoarthritis of the remaining foot joints following this procedure[22-24,26,27].

For more than thirty years, surgeons at our institution have performed a unique combination of tendon transfers and osteotomies developed by the senior author (R.R.C.) to reduce cavovarus deformity and create a functional foot. Patients with a cavovarus foot deformity and a flexible hindfoot undergo plantar fasciotomy, transfer of the peroneus longus to the peroneus brevis tendon, and first metatarsal osteotomy. We believe that the deformity is driven by a plantar flexed first ray, as described by Paulos et al.[5]. Transferring the peroneus longus to the peroneus brevis removes a deforming force on the first ray and reinforces the weak eversion strength of the peroneus brevis. The plantar fascia release also reduces the cavus deformity. In most cases, the foot is not plantigrade after a peroneus longus transfer and plantar fasciotomy, and a first metatarsal osteotomy is performed to further decrease plantar flexion of the first ray. If there is clawing of the great toe, an extensor hallucis longus recession (Jones procedure) is also performed. If the preoperative strength of the tibialis anterior is at least 4 of 5, this tendon is transferred to the lateral cuneiform to supplement eversion strength. We believe that this transfer decreases the contribution of the tibialis anterior to the cavus deformity and foot inversion and augments foot eversion, while preserving its dorsiflexion function.

Although portions of this procedure have been described in other studies[2,4,5,10,11,14-16,20], to our knowledge the outcomes of this particular combination of procedures have not been reviewed or reported in the literature. The purpose of this study was to use reproducible outcome measures to systematically review the long-term results of this treatment approach with regard to patient function, radiographic changes, and gait parameters.

Materials and Methods

Study Sample

This study was approved by our institutional review board, and informed consent was obtained from all patients. The study included patients with a documented diagnosis of Charcot-Marie-Tooth disease (made by a neurologist) who had undergone surgery for a cavovarus foot deformity with a flexible hindfoot between 1970 and 1994 at our institution. A search of the medical records revealed forty-three patients (seventy-one feet) who met these criteria. Four of the forty-three patients had died. We were able to locate thirty-five of the remaining thirty-nine patients by using information from the medical record, free Internet database searches as previously described[28,29], and a fee-based Internet search[30]. Four patients could not be located, and one patient was a prison inmate. Seven patients declined to participate in the study. Two patients agreed to participate, but failed to return surveys despite multiple attempts to encourage them to do so. Of the remaining twenty-five patients (forty-one feet), seventeen (with twenty-nine affected feet) returned for clinical and radiographic evaluation and eight (with twelve affected feet) completed the clinical questionnaires and returned them by mail. Thirteen patients underwent gait analysis.

Medical records were reviewed to determine the symptoms at the initial presentation, age at surgery, postoperative complications, and details of any previous or subsequent treatment. Eleven patients were female, and fourteen were male. Ten patients had simultaneous bilateral procedures, six patients had staged bilateral procedures, and nine patients had unilateral procedures. Only two patients (two feet) had undergone surgery on the same foot prior to our surgical intervention; the prior operation consisted of a plantar fasciotomy and calcaneal osteotomy in one patient and a midfoot wedge osteotomy in the other. Two patients had undergone triple arthrodesis on the contralateral foot at another institution before presenting to our clinic. At the time of the preoperative evaluation, all patients had flexible hindfoot varus that was passively correctable with use of the test described by Coleman and Chesnut[31] or by Price and Price[32].

Surgical Technique

Surgical treatment includes plantar fasciotomy, transfer of the peroneus longus to the peroneus brevis tendon, and first metatarsal osteotomy in most patients. If the foot is plantigrade after peroneus longus transfer and plantar fasciotomy, a first metatarsal osteotomy is not performed. If there is clawing of the great toe, an extensor hallucis longus recession (Jones procedure) is also performed. Prior to 1980, a tibialis anterior transfer was not always included, although several patients subsequently underwent this transfer. Since 1980, we have routinely performed a transfer of the tibialis anterior to the lateral cuneiform if the patient has muscle strength of at least 4 of 5 preoperatively.

The plantar fasciotomy is accomplished through a 1-cm incision made on the medial side of the midfoot over the palpable plantar fascia. A hemostat is used to bluntly dissect

TABLE I Demographics of Those Who Did and Did Not Participate in the Study		
	Participants	Non-Participants
Sex (M:F)	14:11	14:4
Mean age at surgery (range) *(yr)*	15.5 (8.7-25.1)	16.7 (9.3-30.3)
Bilateral surgery *(no. of patients)*	16 (64%)	12 (67%)

above and below the fascia, which is then divided with a number-11 blade.

A curvilinear incision is then made laterally over the peroneal tendons. Careful dissection reveals the sural nerve, which is gently retracted inferiorly. The sheaths of the peroneus longus and brevis tendons are opened longitudinally with a scalpel. The peroneus longus tendon is transected as distally as possible through the same incision; it is then woven through the peroneus brevis tendon with a tendon passer and sutured with multiple number-0 Vicryl sutures (polyglactin; Ethicon, Somerville, New Jersey).

A longitudinal incision is made over the dorsum of the great toe, over the extensor hallucis longus tendon, and carried proximally to the tarsometatarsal joint. The extensor hallucis longus tendon is divided at its insertion. A drill hole is made through the neck of the first metatarsal.

The same incision is used to perform the first metatarsal osteotomy. A dorsal closing wedge osteotomy is created in the proximal third of the metatarsal shaft. A Kirschner wire is used to drill multiple perforations in the bone, and the osteotomy is then completed with a sagittal saw. The wedge of bone is removed. A dorsiflexion force is applied to the forefoot to close the osteotomy site. The extensor hallucis longus tendon is then passed through the drill hole from medial to lateral and sutured back on itself. The extensor hallucis longus tendon transfer is usually sufficient to secure the osteotomy site, but a Kirschner wire may also be placed for temporary fixation.

When a transfer of the tibialis anterior tendon is indicated, a dorsomedial incision is made over its insertion on the navicular. Dissection is carried distally in order to obtain as much length as possible on the tendon stump, and it is divided at its insertion. A number-2 nonresorbable suture is passed through the tendon stump in a Bunnell fashion. The lateral cuneiform is localized fluoroscopically, and a 2-cm longitudinal incision is made over it. An 8 or 9-mm drill bit is passed through both cortices of the cuneiform, from dorsal to plantar. With use of Keith needles, the suture ends are passed through this hole and are tied over a padded button on the plantar surface of the foot. The foot must be held with the hindfoot in neutral in the varus/valgus plane and at least neutral dorsiflexion while the tendon sutures are tied over the button.

A short leg cast is applied, and the patient is allowed to walk with toe-touch weight-bearing for the first six weeks, followed by three weeks of weight-bearing as tolerated while wearing the short leg cast. The cast and button are removed approximately nine weeks postoperatively, and the patient can wear a normal shoe again.

Clinical Evaluation
The clinical evaluation consisted of completion of a questionnaire, clinical examination, and gait analysis with use of the GAITRite System (CIR Systems, Havertown, Pennsylvania). The questionnaire contained the Short Form-36 (SF-36)[33] and Foot Function Index (FFI)[34] elements as well as several additional questions addressing shoe fit and brace wear. For the gait analysis, the patient was asked to walk across the 30-ft (9.1-m) GAITRite mat at three different speeds: slow, quick, and his or her self-selected pace. The order of walking speeds was determined randomly with use of a Latin square method. Each walk was repeated, and the values from the two trials were averaged.

Radiographic Evaluation
The radiographic evaluation consisted of standing lateral and mortise views of the ankle, anteroposterior and lateral views of the foot, and a hindfoot alignment view as described by Saltzman and el-Khoury[35]. Complete sets of radiographs were available for fifteen patients (twenty-six feet), and all radiographs except the hindfoot alignment view were available for an additional patient (one foot). The talo-first metatarsal (Meary), calcaneal-first

TABLE II Summary of Patient-Reported Outcome Measures*				
	Female	Male	Smokers	Nonsmokers
FFI pain score *(points)*	40.7 (0-67.9)	29.5 (0-67.9)	52.6 (2.5-67.9)	18.2 (0-49.4)
FFI disability score *(points)*	44.1 (8.3-66.7)	37.0 (0-67.9)	54.4 (43.2-67.9)	27.3 (0-60.5)
FFI activity limitation score *(points)*	22.1 (0-57.8)	22 (0-53.3)	36.1 (13.3-57.8)	8.7 (0-26.7)
SF-36 MCS *(points)*	52.9 (30.7-65.8)	47.3 (20.5-62.4)	45.6 (20.5-65.8)	53.1 (46.3-52.4)
SF-36 PCS *(points)*	34.2 (17.3-52.7)	40.4 (24.9-59.5)	28.8 (17.3-42.4)	44.6 (23.6-59.5)
Body mass index *(kg/m²)*	24.4 (17.3-39.8)	31.3 (20.5-49.3)	24.6 (17.3-39.1)	31.1 (19.5-49.3)
*The values are given as the mean with the range in parentheses.				

Fig. 1-A

Figs. 1-A through 1-F Clinical photographs of a male patient with Charcot-Marie-Tooth disease who underwent reconstruction of the left foot at the age of eight years and eight months. In the preoperative photographs (Figs. 1-A, 1-C, and 1-E), the right foot appears to be relatively unaffected by deformity, while the left foot has pronounced hindfoot varus and cavus. The patient was lost to follow-up but returned to the clinic twelve years later, at which time he requested surgery for the right foot. Photographs made at that time (Figs. 1-B, 1-D, and 1-F) show slight recurrence of hindfoot varus and excellent maintenance of the cavus correction of the left (operatively treated) foot and substantial progression of the right (untreated) foot deformity.

Fig. 1-B
Postoperative photograph.

Fig. 1-C
Preoperative photograph.

Fig. 1-D
Postoperative photograph.

metatarsal[9], and calcaneal inclination angles were measured on the lateral radiograph of the foot[36]. In addition, the foot height-to-length ratio was calculated as the ratio of the height of the navicular to the length of the foot[36,37]. Varus-valgus alignment was determined from the hindfoot alignment view[35].

Two orthopaedic surgeons and one musculoskeletal radiologist reviewed the radiographs for evidence of degenerative changes in the lesser metatarsophalangeal, first metatarsophalangeal, naviculocuneiform, talonavicular, subtalar, and ankle joints. Arthritic changes of the joints were rated on a scale of 0 to 4 as described by Kellgren and Lawrence[38]. The

mode of the three scores was used as the final score. For the ankle and talonavicular joints, which were clearly visible on two orthogonal radiographs, separate ratings were obtained for each view and the higher of the two scores was reported as the final score for the joint. One patient had undergone an ankle arthrodesis by the time of our radiographic review, and radiographs made prior to that fusion were used for the study.

Statistical Methods

Descriptive statistics were calculated for all variables. In addition, Pearson correlation coefficients were used to describe

Fig. 1-E
Preoperative photograph.

Fig. 1-F
Postoperative photograph.

the relationships between continuous variables, including the age at the time of surgery, duration of follow-up, body mass index, SF-36 scores, FFI scores, radiographic alignment measurements, and gait parameters.

Results

Non-Participation Bias

In order to evaluate possible non-participation bias, we compared preoperative characteristics of the patients who participated in the study with those of the patients who did not. There was no significant difference between the groups with regard to the age at the time of the surgery or the per-

centages who had bilateral or unilateral procedures. The average age of the patients who participated in the study was 15.5 years (range, 8.7 to 25.1 years) compared with 16.7 years (range, 9.3 to 30.3 years) for the non-participants. Seventy-three percent of the eligible female patients participated in the study compared with 50% of the eligible male patients. A similar percentage of patients had bilateral surgery in both groups (Table I).

Demographics and Treatment Variables

A chart review revealed that nine patients had initially presented to the orthopaedic clinic for evaluation of foot deformity and six

Fig. 1G

Fig. 1-H

Postoperative radiographs confirm the findings demonstrated by the photographs. The right (untreated) foot has substantial cavus and is shortened compared with the left foot.

patients had been referred to the clinic by their neurologist because of known Charcot-Marie-Tooth disease. The remaining patients presented because of foot pain or spasm (four patients), ankle instability (one patient), weakness (one patient), or difficulty with wearing shoes (one patient). We were unable to determine the presenting symptoms of three patients.

In the study group, the average age was 15.5 years (range, 8.7 to 25.1 years) at the time of surgery and 41.5 years (range, 20.5 to 53.8 years) at the time of follow-up. The average duration of follow-up was 26.1 years (range, 9.9 to 33.5 years).

In addition to the described operation, procedures done at the time of the index surgery included arthrodesis of the

TABLE III Radiographic Alignment		
Alignment Measurement	Study Group*	Normal Group†[35-37,39]
Calcaneal inclination (deg)	25.3 ± 3.3	21
Calcaneal-first metatarsal angle (deg)	128.9 ± 5.4	132
Navicular height:foot length ratio	0.30 ± 0.03	0.25
Talo-first metatarsal angle (deg)	6.2 ± 4.32	1
Hindfoot varus (mm)	15.9 ± 9.9	3.2

*The values are given as the mean and standard deviation. †The values are given as the mean.

interphalangeal joint of the great toe because of a rigid clawtoe deformity of the great toe in four patients (six feet), Achilles tendon lengthening in one patient (one foot), and transfer of the tibialis anterior tendon to the cuboid or middle cuneiform in nine patients (fourteen feet). Three additional patients (three feet) had the tibialis anterior tendon transferred as a second procedure. Seven patients (ten feet) did not undergo recession of the extensor hallucis longus, and four patients (six feet) did not have a first metatarsal osteotomy.

Seven patients (eight feet) had a total of eleven subsequent foot or ankle procedures after the index operation (see Appendix). As mentioned above, three patients (three feet) had a transfer of the tibialis anterior tendon, and one of them required a second procedure to release adhesions around the tendon transfer. The remaining subsequent foot or ankle procedures included, in one patient each, a bilateral tibialis posterior tendon transfer to the third cuneiform, a midfoot osteotomy, an extensor digitorum communis tendon transfer to the cuboid in a patient who had had a previous tibialis

anterior transfer, a calcaneocuboid fusion with a tibialis posterior tendon transfer to the third cuneiform and a subsequent revision tibialis posterior tendon transfer, and an ankle fusion. Six of the seven patients who had repeat surgery regularly used tobacco products.

Shoe Wear and Foot Appearance

At the time of follow-up, eleven of the twenty-five patients used some sort of orthosis for one or both feet. These orthoses included an insert (ten feet), an ankle-foot orthosis (six feet), and an ankle brace (one foot). Regular fashion footwear was worn on ten feet; comfort shoes, on twenty-four feet; and custom shoe wear, on one foot. No information on shoe type was given for six feet. Clinically apparent calluses were present on ten of twenty-seven feet. The callus was located under the fifth metatarsal head of six feet, the second metatarsal head of two feet, the first and fifth metatarsal heads of one foot, and along the lateral border of one foot. No ulcers were seen on any of the feet, and all patients stated that they had not had foot ulcers. The mean hindfoot alignment angle was 16.1° of varus for the patients with a callus under the fifth metatarsal head and 15.1° of varus for those who did not have a callus under the fifth metatarsal head.

Health and Functioning

Two patients reported a history of diet-controlled diabetes mellitus. Eleven patients reported using tobacco products on a regular basis. Fourteen patients worked outside the home, and five patients received Social Security disability income. Of the remaining patients, one was a homemaker, one was unemployed, and four did not list an occupation.

The average mental component score (MCS) of the SF-36 was 49.8 points (range, 20.5 to 65.8 points), and the average physical component score (PCS) was 37.7 points (range, 17.3 to 59.5 points). The mean PCS was numerically (but not significantly) lower for women than men, although the mean MCS values were nearly equal. Patients who smoked tended to have a lower mean body mass index as well as a lower MCS and a lower

TABLE IV Radiographic Arthritis Grades*					
Joint	Grades 0-1	Grade 2	Grade 3	Grade 4	Total
Talonavicular	23	2	1	1	27
Tibiotalar	21	4	1	1	27
First metatarsophalangeal	24	1	2	0	27
Medial cuneiform-first metatarsal	17	4	5	0	26†
Naviculocuneiform	21	5	0	0	26‡
Subtalar	27	0	0	0	27

*The values are given as the number of joints with each Kellgren grade. †One joint had been fused in a procedure at another institution before the patient underwent the index procedure for this study. ‡One joint was not adequately visualized on radiographs so it was not possible to evaluate for arthritis.

TABLE V Summary of Gait Analysis		
	Study Patients*	Normal Group†[40]
Gait velocity (cm/sec)	103 (70-122)	145-159
Cadence (steps/min)	102 (93-111)	111-122
Stance phase (% of gait cycle)	65 (61-68)	60-61
Swing phase (% of gait cycle)	35 (32-39)	39-40
Single support (% of gait cycle)	35 (30-39)	38
Double support (% of gait cycle)	30 (24-36)	24

*The values are given as the mean with the range in parentheses.
†Mean values for normal individuals between thirty and fifty-five years old.

PCS than the nonsmokers. However, only the difference in the PCS was significant (p = 0.0003).

The Foot Function Index (FFI) consists of three subscale scores: pain, disability, and activity limitation. Each subscale has a possible range of 0 to 100 points, with higher scores indicating more loss of function. The average subscores for pain, disability, and activity limitation were 35.0, 40.5, and 22.1 points, respectively. The mean FFI pain and disability scores were numerically (but not significantly) higher for women than men, although the mean activity limitation scores were nearly equal. Smokers had significantly higher FFI pain, disability, and activity limitation scores (p < 0.0001 for all three) (Table II).

There was a strong correlation between the FFI pain subscale and the FFI activity limitation and disability subscales (r = 0.66 and r = 0.63, respectively). The FFI activity and disability subscores were also strongly related (r = 0.77). The FFI activity and disability scores correlated with the SF-36 PCS (r = −0.76 and r = −0.82, respectively) as well. With the numbers studied, there was no significant correlation between either outcome measure and body mass index, age at surgery, or age at the time of follow-up.

The seven patients who had undergone a second operation on the foot or ankle had a significantly lower mean SF-36 MCS (40.2 points compared with 51.5 points for the patients who did not have repeat surgery [p = 0.024]) and a significantly higher (worse) mean FFI disability score (59.3 compared with 37.7 points [p = 0.002]) and FFI activity score (39.1 compared with 19.5 points [p = 0.021]). With the numbers studied, there was no significant difference in SF-36 or FFI scores between those with and those without a tibialis anterior transfer.

Radiographic Variables
The average calcaneal inclination, navicular height:foot length ratio, and calcaneal-first metatarsal angle were all slightly higher than those previously reported for normal patients[37]. The average talo-first metatarsal angle was also slightly greater (more cavus) than normal values, which range from 1° to 5°[36,39]. Photographs and radiographs of selected cases are presented in Figures 1-A through 1-H. The average hindfoot varus was substantially larger than that in normal individuals. According to Saltzman and el-Khoury, 80% of patients should have hindfoot varus of between +4.8 and −11.2 mm[35], but only five (19%) of the twenty-six feet in the study had a value that fell into this range (Table III).

Patients who underwent a secondary operation had a significantly higher mean calcaneal-first metatarsal angle and lower talo-first metatarsal angle than those who did not (136.3° compared with 128.0° [p = 0.009] and 0.7° compared with 7.0° [p = 0.014], respectively). The feet that had a tibialis anterior transfer had a significantly lower mean talo-first metatarsal angle than those that did not have a tibialis anterior transfer (5.2° compared with 8.8° [p = 0.007]).

Radiographically, osteoarthritis was most often seen at the medial cuneiform-first metatarsal joint. Arthritis with a Kellgren grade of 3 or higher was found in eleven joints in eight feet (Table IV). Patients who had at least one joint with osteoarthritis with a Kellgren grade of 2 or higher had higher FFI pain subscores (mean, 38.1 points) than those with no or grade-1 arthritis (mean, 23.5 points). There was little difference in the mean FFI activity limitation and disability subscores between the two groups. There was no significant correlation between any of the other radiographic measures and the outcome scores.

Gait Analysis
The average self-selected gait velocity was 103 cm/sec with a mean average cadence of 102 steps/min. The average self-selected stride length was 122 cm. During the gait cycles at the self-selected speed, an average of 35% of the total time was spent in swing phase and 65% was spent in stance phase. Thirty-five percent of the time was spent in single support (per foot), and 30% was spent in double support. When compared with published average values for normal individuals, the study patients had a slower gait velocity and lower cadence, but the proportion of time spent in stance and swing phase was near that of normal individuals (Table V)[40].

Interestingly, there was a moderate correlation between calcaneal inclination and cadence (r = 0.58), with increased calcaneal inclination associated with a greater number of steps per minute. There was a moderate negative correlation (r = −0.55) between hindfoot varus and stride length and between self-selected gait velocity and double-support stance time (r = −0.62). There was also a moderate correlation between the body mass index and the double-support stance time when the patient walked at a self-selected speed (r = 0.67) and at a fast speed (r = 0.72). Patients who had undergone a transfer of the tibialis anterior tendon spent less time in double-stance phase during both the self-selected-speed test (28.8% compared with 32.4% for those who had not undergone the transfer [p = 0.014]) and the fast-speed test (26.2% compared with 29.6%

[p = 0.014]), although patients who had had a tibialis anterior transfer also had a lower body mass index (25.6 compared with 30.9 kg/m^2 [p = 0.033]).

Discussion

A wide variety of procedures for the treatment of cavovarus foot deformities have been described, but no single combination of procedures has gained wide acceptance. Many previous reports[2,4-6,12,14,19] have included a heterogeneous group of patients with multiple etiologies of deformity, and there have been few studies on the long-term results of treatment. We were unable to find any studies in which patients had been followed for more than five years after soft-tissue surgical treatment for cavovarus foot deformity. Other than triple arthrodesis, the only procedure for cavovarus deformity in Charcot-Marie-Tooth disease for which outcomes have been reported after a minimum of five years of follow-up is cuneiform osteotomy combined with plantar release and calcaneal osteotomy[19].

The prevalence of additional foot or ankle surgery in our study (20% of forty-one feet) is lower than that found after long-term follow-up of previously reported treatments[19,22,23]. Seven of our patients had subsequent surgical treatment: three underwent a tibialis anterior tendon transfer, which is now part of the standard surgical treatment, and no patient underwent a triple arthrodesis. Wicart and Seringe performed a calcaneal osteotomy, plantar release, and an opening-wedge cuneiform osteotomy in sixteen patients with Charcot-Marie-Tooth disease who had cavovarus feet and hindfoot flexibility before skeletal maturity[19]. Eight of those sixteen patients (eleven of twenty-six feet) had secondary operations, and eight feet (31%) required triple arthrodesis at a mean of 6.9 years after the index operation. Reported reoperation rates after triple arthrodesis have ranged from 36% to 50%[22,23,26].

Radiographic analysis revealed that most patients in our study had little residual cavus, although some hindfoot varus recurred in most patients. On the basis of clinical notes that stated that all patients had a neutral hindfoot and some surviving immediate postoperative clinical photographs, we believe that the hindfoot deformity was fully corrected at the time of surgery and that the hindfoot varus seen in our patients represents recurrent deformity. Unfortunately, no patient had both preoperative and immediate postoperative radiographs available for comparison. Plantar flexion of the first ray no longer appeared to be driving the hindfoot varus in these patients, as evidenced by the lack of cavus. Because Charcot-Marie-Tooth disease causes progressive neurologic deterioration, muscle strength and balance probably change over time. The soleus muscle may be the source of the hindfoot varus, as soleus function is relatively well preserved in patients with Charcot-Marie-Tooth disease and the soleus is a strong hindfoot invertor[41]. Interestingly, recurrent hindfoot varus was one of the major causes of treatment failure in the cohort of patients treated with cuneiform osteotomy reported by Wicart and Seringe[19]. Loss of correction has been observed in patients with Charcot-Marie-Tooth disease even when the hindfoot was

fused[26]. Prevention of recurrent varus remains a treatment challenge.

As we gained experience with this treatment approach, transfer of the tibialis anterior tendon became part of the routine procedure for all patients who had tibialis anterior strength of at least 4 of 5 preoperatively. The tibialis anterior normally functions to invert and dorsiflex the foot. By transferring it to the lateral cuneiform, the tendon supplements weak everters. Patients who had undergone tibialis anterior transfer had a lower mean talo-first metatarsal angle than those who had not, suggesting that, in its native position, the tibialis anterior may contribute to cavus. Transferring the tendon results in a decrease in its overall strength. The tibialis anterior also typically loses strength with progression of the Charcot-Marie-Tooth disease, and over time the transfer often becomes nonfunctional. We have performed a transfer of the tibialis posterior tendon in some patients with Charcot-Marie-Tooth disease (not included in this study because of an insufficient duration of follow-up) in whom the strength of the tibialis anterior was <4 of 5, with anecdotal good early results.

Several authors have advocated the use of calcaneal osteotomy in the treatment of cavovarus foot deformity[4,11,12,19]. We believe that flexible cavovarus deformity is driven by excessive plantar flexion of the first ray, and the hindfoot varus is compensatory[5]. In all of our patients, we were able to obtain a plantigrade foot at the time of surgery with a combination of tendon transfers and first metatarsal osteotomy, and we did not think that a calcaneal osteotomy would be beneficial. In other studies, the use of calcaneal osteotomy in patients with progressive neurologic disorders did not always prevent recurrence of varus foot deformity[19,42]. As discussed earlier, Wicart and Seringe used a calcaneal osteotomy in combination with a cuneiform osteotomy and 31% of the feet required triple arthrodesis because of recurrence of deformity[19]. Similarly, Lariviere et al. reported recurrent deformity in twenty-one of thirty-four patients at an average of five years after treatment with a Dwyer calcaneal osteotomy[42]. In that study, ten of thirteen feet affected by progressive neurologic disease (Charcot-Marie-Tooth or Dejerine-Sottas disease) underwent a repeat operation, leading Lariviere et al. to conclude that calcaneal osteotomy does not provide long-lasting results in patients with progressive neurologic disorders. In our patients, who were followed for an average of twenty-six years, we observed good correction of cavus with some recurrence of hindfoot varus deformity. A calcaneal osteotomy might now be beneficial for those patients, but we do not have any clinical experience to support that conclusion.

Despite some recurrence of deformity, few patients had clinically relevant degenerative changes in the foot and ankle joints. Although we cannot make direct comparisons with the results after triple arthrodesis, our study group had less ankle and midfoot arthritis than has been previously reported in long-term follow-up studies of triple arthrodeses. Wetmore and Drennan followed, for an average of twenty-one years, patients with Charcot-Marie-Tooth disease who had undergone triple arthrodesis at an age that was similar to the age at

the time of the operations in our study[26]. Thirty feet were treated with triple arthrodesis in the study by Wetmore and Drennan, and a subsequent ankle arthrodesis for the treatment of degenerative joint disease was done in six cases (20%), compared with one of forty-one cases in our study. Wetmore and Drennan also reported that degenerative changes of the ankle and midfoot were seen in twenty-three (77%) of the thirty cases, although the degree of radiographic change was not described. In comparison, moderate or severe osteoarthritis (Kellgren grade 3 or 4) of any joint was seen in only eight (31%) of twenty-six feet in our study, including the one that had undergone ankle arthrodesis. Radiographic evidence of arthritis was associated with a higher FFI pain subscore in our study, which suggests that prevention of arthritis may have a greater impact on patient outcome than achievement of radiographic alignment.

Compared with age-matched norms, the SF-36 PCS for our patients was lower and gait, as measured with gait analysis, was slower. The impact of surgical intervention on patient function could not be determined in our study because of its retrospective nature. The patients had a progressive systemic disease that can result in substantial disability regardless of foot function. A survey of 121 Italian patients with Charcot-Marie-Tooth disease revealed a mean PCS of 39.3 points[43], compared with our patients' mean score of 37.6 points. As in our study, the mean PCS for the female Italian patients was lower than that for their male counterparts. In the absence of a control group of patients treated nonoperatively, the results of this study should be considered as descriptive.

Smoking was associated with lower SF-36 PCS and FFI scores. This difference was much larger than the previously reported difference in SF-36 PCS between smokers and non-smokers[44]. In addition, six of the seven patients who underwent secondary procedures were smokers. We could not find any prior study addressing the impact of smoking on disability associated with Charcot-Marie-Tooth disease. This finding may suggest that smoking accelerates the progression of Charcot-Marie-Tooth neuropathy, although we could find no report in the literature implicating smoking in disease progression. On the other hand, these lower functional outcomes could also be due to comorbidities related to smoking, such as heart disease and pulmonary dysfunction, that were not specifically evaluated in this study.

One limitation of this study is the lack of a control group. As described earlier, the true natural history of cavovarus foot deformity in Charcot-Marie-Tooth disease has not been well defined. Although many agree that the deformity seems to be progressive, we could not assess what each patient's foot function would have been in the absence of surgical intervention. In addition, there is great variability in the severity and distribution of musculoskeletal involvement in patients with Charcot-Marie-Tooth disease, even among those with the same genetic mutation[45,46]. The Charcot-Marie-Tooth Neuropathy Score, which is a validated system for scoring disability in patients with Charcot-Marie-Tooth disease, was published[47] after our study was completed, and it could be useful in future

studies. However, even if severity could be determined at a given point in time, the degree of progression of weakness varies from patient to patient.

A second limitation of our study is the variability of the surgical procedure during its development. Although all patients had a transfer of the peroneus longus to the peroneus brevis and a plantar fasciotomy, four patients did not have a first metatarsal osteotomy and seven patients did not have an extensor hallucis longus recession. A metatarsal osteotomy was performed in all patients who did not have a plantigrade foot after the peroneus longus transfer and plantar fasciotomy. We reserved extensor hallucis longus recession for patients with clawed toes. Although every patient did not undergo the same surgical procedures, each was treated with the same approach, so other surgeons could replicate this process for their own patients.

In addition, many of the early surgical procedures did not include tibialis anterior transfer, although it was performed as a second operation in several patients. There was no difference with regard to patient-reported outcome measures between those with and those without a tibialis anterior transfer. At this time, we utilize tibialis anterior transfer for all patients who have strength of at least 4 of 5 at their preoperative evaluation. Although it is known to weaken with time, the tendon transfer can also serve as a tenodesis, holding the foot closer to neutral dorsiflexion and thus in a more functional position.

A third limitation of our study is the low participation rate. Most of the subjects had had the surgery as adolescents, and locating patients ten years or more after surgery is a challenge[29]. Many patients had moved or changed their name since their last medical record information had been recorded. However, we were able to locate most patients (thirty-nine of forty-three), and if the four patients who died are excluded our participation rate was 64%. The functional and radiographic outcomes of the individuals who did not participate in our study could be worse than those of the study participants, resulting in bias in our study. The demographics of the non-participants with regard to age at surgery and percentages of bilateral and unilateral procedures were similar, but there were proportionally more female patients in the study group. Because the female patients tended to have a lower SF-36 PCS both in our study group and in the previously reported literature[43], the non-participants may actually have had better outcomes than our study group.

In conclusion, the described reconstruction for the treatment of a flexible cavovarus foot is associated with a lower rate of degenerative changes in the foot and ankle and a lower prevalence of reoperations at the time of long-term follow-up compared with previous reported outcomes in studies of patients with Charcot-Marie-Tooth disease treated with triple arthrodesis[22,26]. Although most patients have some recurrence of hindfoot varus, most are able to wear normal shoes. The standardized and reproducible patient-reported functional, radiographic, and gait parameter outcomes reported in this study may be used as a benchmark for comparison with the long-term results of other treatments.

Appendix

(eA) A table showing details on all study subjects is available with the electronic versions of this article, on our web site at jbjs.org (go to the article citation and click on "Supplementary Material") and on our quarterly CD/DVD (call our subscription department, at 781-449-9780, to order the CD or DVD). ■

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References

1. Holmes JR, Hansen ST Jr. Foot and ankle manifestations of Charcot-Marie-Tooth disease. Foot Ankle. 1993;14:476-86.

2. Sammarco GJ, Taylor R. Cavovarus foot treated with combined calcaneus and metatarsal osteotomies. Foot Ankle Int. 2001;22:19-30.

3. Alexander IJ, Johnson KA. Assessment and management of pes cavus in Charcot-Marie-Tooth Disease. Clin Orthop Relat Res. 1989;246:273-81.

4. McCluskey WP, Lovell WW, Cummings RJ. The cavovarus foot deformity. Etiology and management. Clin Orthop Relat Res. 1989;247:27-37.

5. Paulos L, Coleman SS, Samuelson KM. Pes cavovarus. Review of a surgical approach using selective soft-tissue procedures. J Bone Joint Surg Am. 1980;62:942-53.

6. Jahss MH. Tarsometatarsal truncated-wedge arthrodesis for pes cavus and equinovarus deformity of the fore part of the foot. J Bone Joint Surg Am. 1980;62:713-22.

7. Sabir M, Lyttle D. Pathogenesis of Charcot-Marie-Tooth disease. Gait analysis and electrophysiologic, genetic, histopathologic, and enzyme studies in a kinship. Clin Orthop Relat Res. 1984;184:223-35.

8. Metaxiotis D, Accles W, Pappas A, Doederlein L. Dynamic pedobarography (DPB) in operative management of cavovarus foot deformity. Foot Ankle Int. 2000;21:935-47.

9. Aktas S, Sussman M. The radiological analysis of pes cavus deformity in Charcot Marie Tooth disease. J Pediatr Orthop B. 2000;9:137-40.

10. Sherman FC, Westin GW. Plantar release in the correction of deformities of the foot in childhood. J Bone Joint Surg Am. 1981;63:1382-9.

11. Gould N. Surgery in advanced Charcot-Marie-Tooth disease. Foot Ankle. 1984;4:267-73.

12. Dwyer FC. Osteotomy of the calcaneus for pes cavus. J Bone Joint Surg Br. 1959;41:80-6.

13. Mitchell GP. Posterior displacement osteotomy of the calcaneus. J Bone Joint Surg Br. 1977;59:233-35.

14. Watanabe RS. Metatarsal osteotomy for the cavus foot. Clin Orthop Relat Res. 1990;252:217-30.

15. Jahss MH. Evaluation of the cavus foot for orthopedic treatment. Clin Orthop Relat Res. 1983;181:52-63.

16. Thomas FB. Levelling the tread: elevation of the dropped metatarsal head by metatarsal osteotomy. J Bone Joint Surg Br. 1974;56:314-9.

17. Giannini S, Ceccarelli F, Benedetti MG, Faldini C, Grandi G. Surgical treatment of adult idiopathic cavus foot with plantar fasciotomy, naviculocuneiform arthrodesis and cuboid osteotomy. A review of thirty-nine cases. J Bone Joint Surg Am. 2002;84 Suppl 2:62-9.

18. Wilcox PG, Weiner DS. The Akron midtarsal dome osteotomy in the treatment of rigid pes cavus: a preliminary review. J Pediatr Orthop. 1985;5:333-8.

19. Wicart P, Seringe R. Plantar opening wedge osteotomy of cuneiform bones combined with selective plantar release and Dwyer osteotomy for pes cavovarus in children. J Pediatr Orthop. 2006;26:100-8.

20. Bentzon PGK. Pes cavus and the m. peroneus longus. Acta Orthop Scand. 1932;4:50-2.

21. Farill J. A tendon transfer for the treatment of certain cases of cavus deformity of the foot. J Bone Joint Surg Am. 1963;45:1779-80.

22. Wukich DK, Bowen JR. A long-term study of triple arthrodesis for correction of pes cavovarus in Charcot-Marie-Tooth disease. J Pediatr Orthop. 1989;9:433-7.

23. Mann DC, Hsu JD. Triple arthrodesis in the treatment of fixed cavovarus foot deformity in adolescent patients with Charcot-Marie-Tooth disease. Foot Ankle. 1992;13:1-6.

24. Levitt RL, Canale ST, Cooke AJ Jr, Gartland JJ. The role of foot surgery in progressive neuromuscular disorders in children. J Bone Joint Surg Am. 1973;55:1396-410.

25. Siffert RS, del Torto U. "Beak" triple arthrodesis for severe cavus deformity. Clin Orthop Relat Res. 1983;181:64-7.

26. Wetmore RS, Drennan JC. Long-term results of triple arthrodesis in Charcot-Marie-Tooth disease. J Bone Joint Surg Am. 1989;71:417-22.

27. Saltzman CL, Fehrle MJ, Cooper RR, Spencer EC, Ponseti IV. Triple arthrodesis: twenty-five and forty-four-year average follow-up of the same patients. J Bone Joint Surg Am. 1999;81:1391-402.

28. King PJ, Malin AS, Scott RD, Thornhill TS. The fate of patients not returning for follow-up five years after total knee arthroplasty. J Bone Joint Surg Am. 2004;86:897-901.

29. Lovell ME, Morcuende JA. Patient location strategies for pediatric long-term follow-up studies. Iowa Orthop J. 2006;26:91-5.

30. Digimedia, Inc. http://www.peoplefinder.com. Accessed 2008 Aug 18.

31. Coleman SS, Chesnut WJ. A simple test for hindfoot flexibility in the cavovarus foot. Clin Orthop Relat Res. 1977;123:60-2.

32. Price BD, Price CT. A simple demonstration of hindfoot flexibility in the cavovarus foot. J Pediatr Orthop. 1997;17:18-9.

33. Ware JE Jr, Kosinski M, Keller SD. SF-36 physical and mental health survey scales: a user's manual. Boston: The Health Institute, New England Medical Center; 1994.

34. Budiman-Mak E, Conrad KJ, Roach KE. The Foot Function Index: a measure of foot pain and disability. J Clin Epidemiol. 1991;44:561-70.

35. Saltzman CL, el-Khoury GY. The hindfoot alignment view. Foot Ankle Int. 1995;16:572-6.

36. Steel MW 3rd, Johnson KA, DeWitz MA, Ilstrup DM. Radiographic measurements of the normal adult foot. Foot Ankle. 1980;1:151-8.

37. Saltzman CL, Nawoczenski DA, Talbot KD. Measurement of the medial longitudinal arch. Arch Phys Med Rehabil. 1995;76:45-9.

38. Kellgren JH, Lawrence JS. Radiological assessment of osteo-arthrosis. Ann Rheum Dis. 1957;16:494-502.

39. Vanderwilde R, Staheli LT, Chew DE, Malagon V. Measurements on radiographs of the foot in normal infants and children. J Bone Joint Surg Am. 1988;70:407-15.

40. Sutherland DH, Kaufman KR, Moitoza JR. Kinematics of normal human walking. In: Rose J, Gamble JG, editors. Human walking. 3rd ed. Philadelphia: Lippincott Williams and Wilkins; 2005. p 33-51.

41. Perry J. Anatomy and biomechanics of the hindfoot. Clin Orthop Relat Res. 1983;177:9-15.

42. Lariviere JY, Miladi L, Dubousset JF, Seringe R. [Failure of Dwyer's procedure in internal pes cavus in children. Physiopathological considerations and therapeutic deductions]. Rev Chir Orthop Reparatrice Appar Mot. 1985;71:563-73. French.

43. Vinci P, Serrao M, Millul A, Deidda A, De Santis F, Capici S, Martini D, Pierelli F, Santilli V. Quality of life in patients with Charcot-Marie-Tooth disease. Neurology. 2005;65:922-4.

44. Laaksonen M, Rahkonen O, Martikainen P, Karvonen S, Lahelma E. Smoking and SF-36 health functioning. Prev Med. 2006;42:206-9.

45. Thomas PK, Marques W Jr, Davis MB, Sweeney MG, King RH, Bradley JL, Muddle JR, Tyson J, Malcolm S, Harding AE. The phenotypic manifestations of chromosome 17p11.2 duplication. Brain. 1997;120:465-78.

46. Carter GT, Abresch RT, Fowler WM Jr, Johnson ER, Kilmer DD, McDonald CM. Profiles of neuromuscular disorders. Hereditary motor and sensory neuropathy, types I and II. Am J Phys Med Rehabil. 1995;74(5 Suppl):S140-9.

47. Shy ME, Blake J, Krajewski K, Fuerst DR, Laura M, Hahn AF, Li J, Lewis RA, Reilly M. Reliability and validity of the CMT neuropathy score as a measure of disability. Neurology. 2005;64:1209-14.