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OPERATIVE TREATMENT OF THE FOOT DEFORMITY IN CHARCOT-MARIE-TOOTH DISEASE

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A particular type of progressive muscular atrophy was independently described in 1886 by both Charcot-Marie and Tooth. It is characterized by symmetrical paresis, particularly of the peroneal musculature, for which reason the disease is frequently referred to as progressive peroneal muscle atrophy. There may also be paresis of the anterior tibial muscle, the extensor muscles of the foot and the gastrocnemius muscle. The posterior tibial muscle, on the other hand, generally displays a normal function. The imparied muscular balance results in a foot deformity consisting of an equino-varus-excavatus position. As the disease spreads, symmetrical paresis may also arise in the muscles of the hand and the forearms. The paresis develops in a proximal direction but stops at the knee and elbow joints. This centripetal progression gives the affected limbs a form that is usually likened to an upturned champagne bottle. Advanced cases involve a considerable degree of invalidity—the patients can neither stand nor walk nor, if the arms are affected, can they perform manual work.

The etiology of the Charcot-Marie-Tooth disease is unknown. It is an hereditary complaint. Two main groups have been distinguished, with a somewhat different prognosis, according to the age at which the symptoms first appear (*Allan* 1939). In dominant heredity the symptoms appear around the age of 30, progress slowly and lead to a moderate impairment of the function of the muscles concerned. In recessive heredity the paresis appears before the age of 8, the muscular atrophy progresses rapidly and severe invalidity may have developed by the age of 15–20 years.

The literature to date has chiefly been concerned with the hereditary

and neurological problems of the Charcot-Marie-Tooth disease. Less attention has been paid to the orthopedic aspects and there has been little discussion of the possibilities for surgical correction. Treatment of the foot deformity with a stabilizing subtalus arthrodesis in four cases has however been reported (*Miller* 1924).

In a large series presented more recently (*Jacobs & Carr* 1950), the surgical technique comprised lengthening of the Achilles tendon, plantar fasciotomy and triarticular subtalus arthrodesis according to Lambrinudi. In addition, the anterior tibial tendon was transferred to the middle of the dorsum of the foot in some cases, while in others the posterior tibial tendon was passed through a hole in the interosseous membrane and attached to the middle or lateral part of the dorsum of the foot. Out of 44 patients (*i.e.* 88 operated feet), 32 were reported to have "excellent" or "good" results and the remainder "poor" results. However, those described as "good" included 6 cases with a recurrence of the varus deformity due to hyperactivity of the posterior tibial tendon, which had been left in situ. The authors regarded the subtalus arthrodesis as the most important part of the correction but emphasized that the deformity tended to recur if tendon transfer was not performed as well.

OWN SURGICAL TECHNIQUE

In order to avoid bone surgery, *i.e.* subtalus arthrodesis, particularly in young patients, the intention was to correct foot derformity by means of tendon surgery alone. The equinus position was dealt with by lengthening the Achilles tendon, the excavatus by plantar fasciotomy. The varus position was countered by transferring the powerful posterior tibial tendon. At the suggestion of one of us (S.K.), the previous technique was modified in that the posterior tibial tendon was transferred via a subcutaneous tunnel *in front of* the ankle joint and attached to the cuboid bone (Figure 1). When lengthening the Achilles tendon, one must make sure that the tibial tendon can thereby reach its new insertion. After the operation the foot was kept in a plaster cast for 6 weeks in an overcorrected position, *i.e.* dorsal extension and pronation.

CASES

This report concerns 4 patients (2 girls, 2 boys), all of whom have undergone a bilateral surgical correction of typical foot deformities in Charcot-Marie-Tooth's disease; 8 feet have thus been operating upon. The age at operation varied from 5½ years to 19 years, the symptoms having been present for about 3 years. Heredity was confirmed for 3 of the patients, the disease having afflicted both the mother, the maternal grandmother and a sibling of one of them. No heredity could be demonstrated in the fourth case.

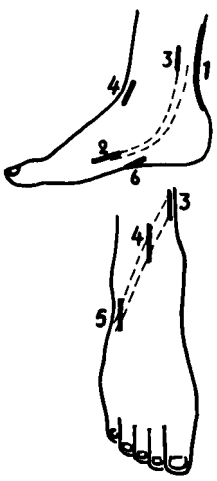

Figure 1. Incisions for the operative technique.

1. *Lengthening of the Achilles tendon.*
2. *Division of the insertion of the posterior tibial tendon.*
3 *and 4. Subcutaneous tunnel,* anterior *to the ankle joint for the posterior tibial tendon.*
5. *Attachment of the posterior tibial tendon to the cuboid bone.*
6. *Plantar fasciotomy.*

A preliminary report on these cases has already been published (*Karlholm & Nilsonne* 1964) The average period of observation was short at that time but now ranges from 4½ years to 8½ years.

RESULTS

The primary results may be described as "excellent" in every case. Thus the transferred posterior tibial tendon functioned strongly, enabling active pronation of the foot. Inspection showed normal weight bearing by the feet in standing, though there was possibly a slight residual excavatus position in one case. Walking capacity was also more normal and ordinary shoes could be used instead of supports or special shoes.

The primary improvement has been maintained throughout the period of observation in 3 cases (6 feet) (Figs. 2 and 3). The stability of the feet is still good and the functional strength of the transferred posterior tibial tendon is unimpaired. In the fourth case—a girl who was operated upon at 5½ years of age—there has been recurrence and deterioration. The reason for this is a very marked progress of the muscular atrophy. The patient now suffers from severe invalidity with almost total paralysis of the feet and lower legs as well as marked paresis in the hands and forearms. The transferred posterior tibial tendon does not function actively, though it does serve as a tenodesis.

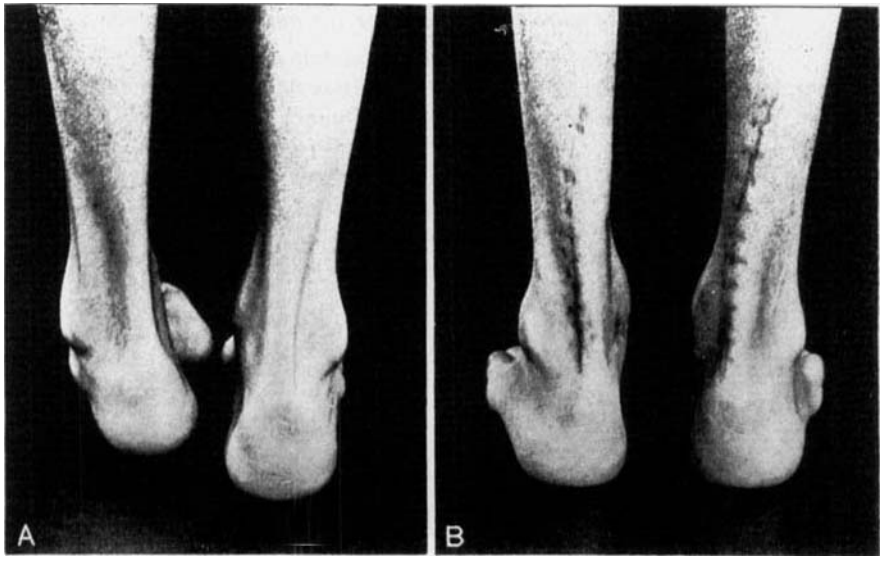

Figure 2 A. Typical deformity of the left foot in Charcot–Marie–Tooth disease; the right foot later developed the same degree of equino-varus-excavatus position. Figure 2 B. The same case as in Figure 2 A, the left foot 4½ years, the right foot 8½ years after operative correction. Both feet are well-balanced at weight-bearing.

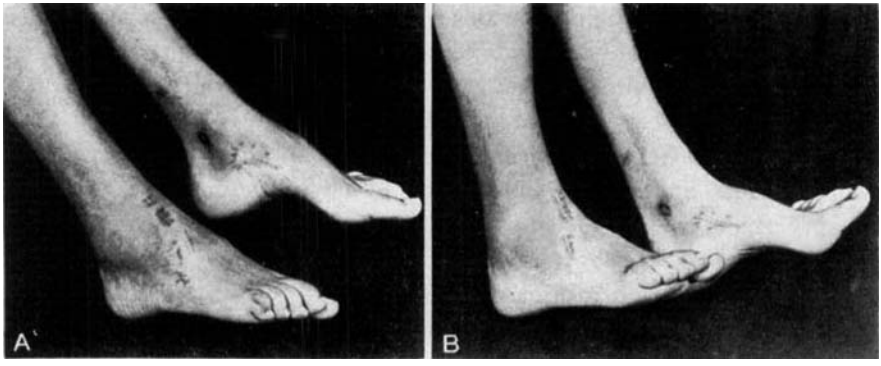

Figure 3. The same case as in Figures 2 A and B showing active dorsal extension and plantar flexion of both feet.

DISCUSSION

One must always bear in mind the diagnosis Charcot-Marie-Tooth's disease when considering foot deformities involving an equino-varus-excavatus position in children and adolescents. This applies particu-

larly to cases with symmetrical changes, especially if the peroneal muscles are found to be weak. We have not found any figures for the incidence of this disease in the literature. Having had our attention drawn to the condition, however, we have been able to diagnose an increasing number of definite cases. The complaint is perhaps not so uncommon as is stated in the usual textbooks. When confronted with a case of Charcot-Marie-Tooth's disease, one should call in the patient's closest relatives for an examination. Besides providing an opportunity to analyze the hereditary pattern, which may be of prognostic value, this enables one to assess and treat mild or undetected deformities of the foot.

Concerning the present surgical technique, we wish to emphasize the particular importance of transferring the posterior tibial tendon. Placing this in front of the ankle joint gives a better leverage and hence an increased power of pronation compared with the technique of passing the tendon through the interosseous membrane. Moreover, a subcutaneous tunnel undoubtedly provides freer passage for the tendon compared with a hole through the interosseous membrane, which may lead to tenodesis. On the other hand, transferring the posterior tibial tendon is not sufficient by itself. The equino-excavatus position must also be corrected by lengthening the Achilles tendon and plantar fasciotomy.

The result of the operation was "excellent" in 3 cases (6 feet) and "poor" in 1 case (2 feet). It is thus possible for tendon surgery alone to correct and stabilize deformities of the foot in Charcot-Marie-Tooth's disease. One can of course argue that equivalent results could have been achieved with a subtalus arthrodesis. In our opinion, however, and in view of the findings of Jacobs & Carr, one must nevertheless transfer the strong posterior tibial tendon in order to avoid a recurrence of the deformity. We therefore conclude that bone surgery can be avoided and that tendon transfer can be sufficient.

Even in the case in which the result was classified as "poor", the tenodesic function produced a relative stabilization of the foot deformity. The muscular atrophy was obviously highly progressive in this case. The symptoms had appeared when the patient was only about 2 years old, which suggest a poor prognosis, and hence the indications for a tendon transfer were doubtful. The result of a subtalus arthrodesis would probably not have been any better. Moreover, it has been shown that an early subtalus arthrodesis in congenital pes equinovarus does not produce the desired effect (*Lemperg* 1965).

SUMMARY

The typical foot deformity in the Charcot-Marie-Tooth disease consists of an equino-varus-excavatus position. The deformity can be treated by soft-tissue surgery alone. The operative technique is described. The prognosis depends not only upon the result of surgical corrections but also upon the inherent progressiveness of the muscular atrophies.

RESUME

La déformité typique du pied dans la maladie Charcot-Marie consiste en une position equino-varus-excavatus. Cette déformité peut être traitée par la chirurgie des tissus mous uniquement. La technique opératoire est décrite. Le pronostic ne dépend pas seulement du résultat des corrections chirurgicales mais aussi de la progression des atrophies musculaires qui s'y rattachent.

ZUSAMMENFASSUNG

Die typische Fussdeformitet bei der Charcot-Marie-Tooth Krankheit besteht in einer equino-varus-cavus Stellung. Die Verbildung kann mittels Weichteilchirurgie ellein behandelt werden. Die Operationstechnik wird beschrieben. Die Prognose ist nicht nur von dem Ergebnis der chirurgischen Korrektur sondern auch von der zugehörigen fortschreitenden Muskelatrophie abhängig.

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