Case report

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The Ilizarov method for treatment of severe foot deformity in a patient with Charcot-Marie-Tooth disease

A.S. Neretin, S.S. Leonchuk, G.P. Ivanov
Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Kurgan, Russian Federation

The treatment result in a patient with a complex deformity of the feet due to neural Charcot-Marie-Tooth amyotrophy is presented in the article. A combined use of the Ilizarov transosseous osteosynthesis method and transfer of the tibialis posterior muscle tendon by anchoring to the cuboid bone allowed us to achieve a successful long-term result of treatment and restore ability of active dorsiflexion of the foot. Functional outcome was assessed using the American Orthopedic Foot and Ankle Society (AOFAS) scale. AOFAS score increased significantly from 26 points preoperatively to 80 points at 18 months after intervention. The ankle motion increased from 25° preoperatively to 50° on the left side and 60° on the right side at long term. In our opinion, this approach allowed the patient to start walking earlier, restored weight-bearing ability of the foot and reduced treatment time.

Keywords: foot deformity, Charcot-Marie-Tooth disease, Ilizarov frame, anchor

Charcot-Marie-Tooth disease (CMT) (neural amyotrophy) is a group of hereditary disorders characterized by motor sensory neuropathy and belongs to severe chronic diseases of the nervous system, being the most common clinical hereditary polyneuropathy [1]. The disease occurs due to defects in the genetic code of the protein of the peripheral myelin sheath [2]. The average incidence in the population is one case per 3,000. Incidence in the Russian Federation varies from 7.14 to 13.3 per 100,000 and differs in regions but accounts for about 80 % of all hereditary neuropathies [3]. The disease progresses gradually, usually in the second or third decades of life. The general phenotype of Charcot-Marie-Tooth disease includes foot deformities and painful callus formations [2, 4, 5]. Currently, despite a significant progress in elucidating the genetic molecular mechanisms of this disease, its effective treatment has not been found [6].

Due to muscular imbalance, neutralization of deforming forces decreases when an affected person loads his/her limbs and a multicomponent foot deformity develops. Similar foot deformations that develop due to neuromuscular diseases and injuries with damage to the peripheral nerves of the lower extremities constitute a significant number of orthopedic pathology. Severe deformity and accompanying trophic changes impair foot functions and frequently lead to disability [7].

The goal of surgical treatment is to correct the deformity, restore the static and dynamic function and improve the appearance of the segment. The ability to move fully and independently, if restored with a complex treatment, assists in patient's social communication and improves quality of life.

Most authors emphasize that effective methods of surgical correction of severe foot deformities in this pathology imply simultaneous intervention on bones and soft tissues. In this case, achillotomy, dissection of plantar aponeurosis, tendon and muscle plasty are used as a supplement to interventions on the joints and bones of the foot. Among the latter, wedge resections in combination with triple- or four-joint arthrodesis with subsequent fixation by internal metal structures and a plaster bandage are the main means. This approach allows deformity correction but leads to significant shortening and makes additional deformity correction, if necessary, impossible. Moreover, in some cases, transposition of tendons of various muscles is performed to restore the muscular balance of the foot [7-12].

It should also be noted that some authors use external fixation devices for a single-step [9] or dosed correction of severe foot deformities followed by dismantling of the external apparatus and subsequent stabilizing operation applying internal elements [7, 13].

Long-term outcomes of surgical treatment of foot deformities in patients with CMT have been described only in several studies [10, 14, 15]; therefore, there is not enough evidence of optimal surgical interventions in patients with this pathology.

In our view, it is advisable to use a two-stage approach in a pronounced deformity, applying only the Ilizarov apparatus as an external fixator. At the first stage, gradual correction of foot deformity is performed with this fixator. It completely eliminates the risk of trophic soft tissue complications. At the second stage of treatment, immediately after the deformity has been corrected, we recommend to perform a stabilizing operation (triple arthrodesis) and a transfer of the tendon of the posterior tibial muscle to the cuboid bone. Fixation of the tendon to the bone is possible both by suturing through the canal in the cuboid bone, or by a more convenient technique of anchor fixation. Due to disease persistence, this procedure restores not only active dorsal flexion in the ankle joint but also prevents deformity recurrence at long term. The function of the muscles of the posterior tibial group including the tibialis posterior, as a rule, remains at a sufficient level in CMT patients (innervated n. tibialis). The anterior tibial muscle (as well as the peroneal muscles) rapidly loses its force in the process of patients’ growth due to progressing functional disorders of n. peroneus and its branches. Given the fact that muscular imbalance of varying degree is present in all the patients with this disease, we believe that the indications for transposition of the tendon of the posterior tibial muscle should be widened.

In our opinion, stable fixation of the lower leg and foot in the Ilizarov apparatus enables to mobilize the patient and to bear weight on the foot starting from days 2 or 3 after the operation. Additional immobilization, as a rule, is not required after dismantling of the device upon completion of the fixation period.

Further on, preservation of the treatment result should be provided by a complex of rehabilitation measures and supporting courses of neurotropic therapy.

Our approach can be illustrated with a clinical case.

Patient S., aged 28, was admitted to our orthopedic department No. 5 of the Ilizarov Center with the diagnosis: Charcot-Marie-Tooth neural amyotrophy, equinovarus deformity of the foot.

Earlier, the patient had been rejected to be operated at his residence hospital and was recommended only to wear orthopedic shoes, have massage and exercise therapy courses.

Clinically, upon admission, the patient complained of severe foot deformity, inability of active dorsal flexion and coordinated walking, pain and hyperkeratosis on the outer surface of both feet. The range of active movements in the ankle joint (extended knee joint) was the following: on the left side – 25º (135–160º), on the right one – 25º (145–170º). Adduction of the anterior part of the left foot was 20º, and of right one – 15º; equinus on the left side was 135º, and on the right – 125º; supination of the front foot on the left – 45º, on the right – 30º; heel varus on the left side – 10º, on the right – 5º. There was a pronounced tensioning of the Achilles tendon and plantar aponeurosis as well as hammer toes. Muscular strength of the plantar flexors of both feet was estimated at 5 points, of the dorsal ones at 2-3 points; the active abduction of the feet was absent. Walking was possible only in orthopedic shoes using crutches. During ambulation, the anterolateral part of the foot was overloaded (Fig. 1, a, b). X-rays also showed a pronounced deformity of the feet, subluxation in the talonavicular joint, signs of osteoporosis of the calcaneus (Fig. 1, c). According to the AOFAS scale, the functional state of the feet was rated at 26 points.

![Fig. 1](image-url)

**Fig. 1** Photos and radiographs of patient's feet before treatment: a – side view (sitting); b – view from the plantar surface; c – radiographs of the feet in the lateral projection and a frontal view of midfoot
Due to a complex and rigid deformity of the feet, the tactics of gradual elimination of the deformity with the Ilizarov apparatus was chosen after a preliminary achillo-, planotomy and tenotomy of the flexors of the toes followed by diafixation of the toes with wires. Deformity correction in each foot continued 31 days. This approach did not cause trophic disorders. Diafixation of the toes with wires made it possible to avoid flexion contracture of the toes during the correction process. After correction, a stabilizing operation was performed (triple arthrodesis) along with a transfer of the tendon of the posterior tibial muscle onto the cuboid bone. And, on the right foot, the tendon was fixed with an anchor. Stages of surgical treatment were performed sequentially on each foot (Fig. 2).

The patient began full weight-bearing after deformity correction had been completed, and from the second day after the 2nd (stabilizing) operation he continued to load the limb. Physical exercises and a course of massage were administered. The patient ambulated, actively loading all parts of his feet, using one crutch as an auxiliary means of support. The Ilizarov apparatus was dismantled 45 days after the operation. Additional immobilization with a plaster splint was not required. The patient was recommended to wear hard back shoes, orthopedic insoles with a pronator under the forefoot. He was recommended to have thigh and shin massage, exercises for ankle joints, electrostimulation of the anterio-lateral group of leg muscles.

At a follow-up examination 18 months after the removal of the Ilizarov apparatus, the patient did not have any complain. Clinically and radiologically, the result of the treatment was preserved (Fig. 3). The feet were in normal correction. There were no deformities and toe contractures; the support was restored to all parts of the feet. The range of active movements in the ankle joint (with an extended knee joint) was as follows: on the left – 50° (80–130°), right – 60° (80–140°). The muscular strength of the dorsal flexors was 3–4 points and the plantar strength was 4–5 points. The patient walked in regular shoes supplied with a hard back and the recommended orthopedic insole with a pronato and did not use additional means of support. He continued visits to a neurologist at the place of his residence, received courses of neurotropic therapy, physiotherapy, exercise therapy and massage. The patient was completely satisfied with the result of treatment. According to the AOFAS scale, the functional state of the feet was 80 points.

Fig. 2 Photo and radiographs of patient’s feet during treatment: frontal view: a – left foot; b – right foot; c – lateral X-rays of the feet
CONCLUSION

The use of the Ilizarov apparatus for severe foot deformities in patients with Charcot-Marie-Tooth disease enables to restore the normal static and dynamic functions as well as the appearance of the segment without the risk of trophic disorders and shortening. In addition, the transfer of the tendon of the posterior tibial muscle to the cuboid bone during the stabilizing operation facilitates the restoration of foot dorsal flexion, improves the balance between the flexors in general and prevents deformity recurrence.

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Information about the authors:

1. Andrei S. Neretin, M.D., Ph.D.,
   Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Kurgan, Russian Federation
   Email: wasp75@mail.ru

2. Sergei S. Leonchuk, M.D., Ph.D.
   Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Kurgan, Russian Federation;
   Email: leon4yk@mail.ru

3. Gennadii P. Ivanov, M.D., Ph.D.,
   Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Kurgan, Russian Federation