

Type III longitudinal deficiency of the tibia and outcome of reconstructive surgery in a female patient

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Key words: congenital dysplasia of tibia; reconstructive surgery; children.

Summary. Type III longitudinal deficiency of tibia according to Kalamchi and Dawe denotes the presence of distal hypoplasia of the tibia with diastasis. We report a case of type III longitudinal deficiency of the tibia in a female patient who later underwent reconstructive surgery. The first reconstruction of the leg was done when child turned 4 months of age. Surgical procedures included foot reconstruction and ankle stabilization with twice lengthening by the Ilizarov method (14 cm in total). During the follow-up, both the tibia and fibula of the affected leg showed the same lengthening and regression due to preserved distal growth zone cartilage. After surgical correction, the acetabulum was satisfactorily configured with an acetabular angle of 23 degrees.

Explanation for surgical success was that osteotomy and distraction were done in the proximal part of the crural region where the growth potential was better. The tibia remained lean and hypoplastic while the fibula was incrassated. The function in the area of the knee joint was preserved, while the distal part of the leg served as good stand on. When the child was 18 years old, on check-up, the acetabular angle was 23 degrees while the Wiberg angle was 24 degrees.

Introduction

Type III longitudinal deficiency of the tibia (LDT) is a rare pathological condition with an incidence of one per one million newborns (1, 2). It is often associated with other anomalies (3–5). Earlier proposed classifications of LDT by Jones et al. and Kalamchi and Dawe are based on radiographic findings (6, 7). Jones et al. stress out that type III LDT presents with an overall short tibia, proximal and distal epiphysis with relatively normal fibula in shape and development (6) while Kalamchi and Dawe classified type III LDT as distal hypoplasia of the tibia with diastasis (7). A new classification of tibial deficiency was proposed by Weber pointing out the importance of detection of cartilaginous anlage by magnetic resonance imaging or sonography (8). Since our patient underwent the first examination before newly proposed classification categories, we diagnosed type III LDT according to the Kalamchi and Dawe classification.

Clinical presentation of type III LDT depends on the type of the deficit (9). Treatment depends on the type of the deficit, radiographic findings, functional status of the quadriceps muscle, and whether deficit is unilateral or bilateral (10).

When shortening of the lower limb is significant and ankle diastasis is pronounced, talectomy and synostosis of the tibia and fibula with Boyd's amputation are recommended (11). Difference in the length of the lower extremities can be corrected by length equalizing procedures (12).

Case report

A female newborn patient on labor presented with severe deformity of the right lower limb. Physical examination at birth revealed a 3-cm shortening of the right leg with contracture of the knee between 5 and 10 degrees and well-developed quadriceps mechanism. Valgus of the right knee was in minimal position, and the right foot was in equinovarus position. Medially separated "finger-like processus" with bone incorporated within was described as most likely to be the tibia. There were 3 missing fingers on foot with outer hypertrophic finger (Fig. 1).

Radiographic findings revealed the presence of the tibia with poorly developed distal part and significant diastasis of the tibia and fibula distally. The fibula was banded, proximally displaced with the foot on it and completely separated from the tibia (Fig. 2). The

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Fig. 1. The crural region of the leg before surgery



Fig. 2. Radiographic image of both legs before surgery



Fig. 3. Angulated tibia and angulated fibula when the patient was 18 years old

foot consisted of the calcaneus, cuboid bone, and 2 metatarsal bones. The knee joint was satisfactorily developed with hip dislocation.

Reconstruction of the leg was done when child was 4 months old. As the first step, transaxial extension throughout the talus was done in order to enable the foot and fibula to be at the level of the tibia. To bring the foot on the distal part of the tibia, blood vessels (posterior tibial artery and vein) and tibial nerve were moved to its anatomical position beside the reconstructed tibia. It was necessary to cut and elongate the Achilles tendon and the peroneal muscles. After bringing the foot in its physiological position, fixation with a Kirschner wire throughout the talus was performed. Plaster immobilization lasted for 6 weeks.

After plaster removal, shortening of the right leg was 1.5 cm. At the age of 18 months, the patient underwent hip surgery. Open reposition and acetabuloplastic technique according to Dega were performed resulting in satisfactory postoperative period. Radiographic evaluation revealed irregularity of the femoral head epiphysis as seen in Perthes' disease that was diagnosed as type III postreductional osteochondritis according to Kalamchi.

In 1992, radiographic evaluation revealed diastasis including the calcaneus but not talus. It was recorded that the tibia started to develop gaining physiological shape and the fibula also started to develop gradually achieving physiological shape. When the child turned 6 years of age, the difference in length between legs was 6 cm. The distal part of the leg was lengthened by 8 cm using the Ilizarov technique when we achieved hypercorrection that was corrected by shoes for the left foot. On control examination, when the patient was 158 cm tall, the difference between

legs was 6 cm: 2.5 cm above the knee and 3.5 cm below the knee. Repeated correction by the Ilizarov method was done with hypercorrection of 10 mm due to protection of the deformed leg. The Trendelenburg sign was negative, and the patient could not stand steady on the deformed leg without holding to a static object.

Flection within the right hip was 110 degrees, while bilateral abduction on both sides was 60 degrees. Rotation was within physiological range as well as flection and extension of both the knees. No movement was possible at the level of the ankle joint. Artificial joint was between the calcaneus, tibia and fibula. Re-examination of the patient at the age of 18 years showed that reconstructive surgery yielded good results and was considered effective mainly because functional walking status was maximally preserved and improved (Fig. 3).

Discussion

The paper presents our experience in the treatment of unusual congenital type III LDT associated with luxation of the hip and foot anomalies. Evaluation of initial condition, changes during growth as well as functional assessment 18 years later allowed us presenting this study as a possible contribution to the treatment of such anomaly.

Some previous studies point out soft tissue reconstruction as a part of the treatment (13), while other denote talectomy and closure of the diastasis (14). The severity of such pathological state is presented in the study by Schoenecker et al., where the ankle joint was reconstructed, and the foot was retained in 9 of the 10 patients, but later 5 of these patients had a Syme amputation (15).

There are authors stressing out opinions that discrepancy of the leg length, foot anomalies and knee status can influence the decision to perform reconstructive surgery (16).

Simmons et al. performed follow-up study, where they used a modified Brown's procedure in the treatment of tibial hemimelia. In their study, a prerequisite for surgery was satisfactory quadriceps function. After a Syme amputation of the foot, the authors reported satisfactory function of the patient's lower extremities. Further, they found that a fibular centralization procedure, when quadriceps function is preserved, can give good functional results (17).

Our surgical approach included several surgical techniques: foot setting down, foot reconstruction, and ankle stabilization with twice lengthening by the Ilizarov method (14 cm in total). Both the tibia and fibula of the affected leg showed the same lengthening and regression due to preserved distal growth zone cartilage.

Explanation for such success in our case was that osteotomy and distraction were performed in the proximal part of the crural region where the growth

potential was better. The tibia remained lean and hypoplastic while the fibula was incrassated (Fig. 2). Knee joint function was preserved, while the distal part of the leg served as good stand on. The ankle joint was completely ankylotic and stable, while the right foot remained smaller. Due to the presence of acetabular dysplasia even after operation, the acetabulum in general was satisfactorily configured. In 2007, re-examination showed the acetabular angle of 23 degrees and the Wiberg angle of 24 degrees.

Conclusion

Different approaches in surgical techniques indicate that patients with diagnosed longitudinal deficiency of the tibia should be evaluated using morphological classification rather than radiological only. With improvement in medicine, modern and precise classifications are made giving new insights into potential treatment options in order to gain and preserve maximal functional status. This will improve quality of life for children with longitudinal deficiency of the tibia.

Trečiojo tipo išilginio blauzdikaulio defekto rekonstrukcinė operacija

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Raktažodžiai: įgimta šlaunikaulio displazija, rekonstrukcinė operacija, vaikai.

Santrauka. Trečiojo tipo išilginis blauzdikaulio defektas, pagal A. Kalamchi ir R. V. Dawe, rodo distalinę blauzdikaulio hipoplaziją su diastaze. Mes pateikiame klinikinį atvejį, kai pacientei buvo diagnozuotas trečiojo tipo išilginis blauzdikaulio defektas. Pirmoji kojos rekonstrukcija atlikta, kai vaikas buvo 4 mėn. amžiaus. Atliktos šios chirurginės procedūros: pėdos rekonstrukcija, kulkšnies stabilizacija ir du apatinės galūnės pailginimai Ilizarovo metodu (iš viso 14 cm). Stebėjimo laikotarpiu paveiktos kojos šlaunikaulis ir šėivikaulis pailgėjo vienodai dėl išsaugotos kremzlės distalinės augimo zonos. Po chirurginės korekcijos gūžduobė suformuota patenkinamai su 23 laipsnių gūžduobės kampų.

Operacijos sėkmė gali būti paaiškinta tuo, kad osteotomija ir distrakcija atlikta proksimalinėje blauzdos dalyje, kur augimas buvo geresnis. Blauzdikaulis išliko plonas ir hipoplastinis, o šėivikaulis sustorėjo. Funkcija kelio sąnario srityje išsaugota, o distalinė kojos dalis atstojo gerą ramstį. Kai pacientei suėjo 18 metų, patikros metu nustatyta, kad gūžduobės kampas buvo 23 laipsnių, o Wiberg kampas – 24 laipsnių.

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