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CASE REPORT

Case Reports

Lengthening and Reconstruction of Congenital Leg Deficiencies for Enhanced Prosthetic Wear

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Abstract Congenital limb deficiencies with severe shortening and/or deformity can be difficult to fit with a prosthesis. We report two patients in whom gradual lengthening and deformity correction with the Ilizarov/Taylor spatial frame™ was used to improve prosthesis fit, comfort, and gait.

Introduction

Congenital limb deficiencies are a rare but important problem. Problems with the residual limb can include deformity and/or shortening that can make prosthetic fitting difficult. Traditional treatments have included the use of prosthetics to accommodate the deformity or amputation above the level of the deformity as needed to facilitate prosthesis fitting and function. There is a distinct advantage to preserving the knee and avoiding an above-knee amputation. Energy expenditure is substantially greater in patients with an above-knee amputation compared with a below-knee amputation [2]. If the prosthesis is accommodated to a deformed residual limb, there can be problems with comfort, gait, and force transmission across the knee [4, 7].

We describe the use of distraction osteogenesis to correct and lengthen residual limb deformities for enhanced prosthesis wear.

Patient 1

A 21-year-old woman presented with femoral shortening and tibial and fibular shortening, deficiency, and deformity secondary to intrauterine amniotic band syndrome. Her toes and part of the foot were amputated at birth secondary to gangrenous changes. She wore a prosthesis, walked with a limp, and her prosthesis was uncomfortable.

She had obvious shortening of the entire left lower extremity with a proximal left knee when compared with the right side and varus deformity of the residual leg (Fig. 1A). Gait examination revealed a valgus thrust with weightbearing of the left lower extremity. There were some superficial abrasions on the pressure points of her residual limb. Knee motion was from full extension to 130° flexion; motor power was 5/5 of both knee flexors and extensors. She had mild anteroposterior instability but no varus or valgus instability. She had a 30° varus deformity of the proximal tibia, and the prosthesis was made to accommodate the varus deformity of her tibia. Her substantial varus deformity was accommodated by internal rotation of the thigh during walking. It appeared that the entire prosthesis was molded around a compensatory internal rotation of the thigh. For the kneecap to point straight, the prosthesis foot needed to point into 30° external rotation. This was an intentional adaptation to fit the prosthesis on such a deformed tibia.

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Each author certifies that his or her institution has approved the reporting of this case report, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.

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Fig. 1A–G (**A**) A preoperative clinical picture and (**B**) anteroposterior and (**C**) lateral radiographs of the left knee show the left residual leg deformity. (**D**) Frame and leg after correction. The frame is extended to compensate for the leg-length discrepancy to allow ambulation. (**E**) Anteroposterior and (**F**) lateral radiographs at the 1-year followup show consolidation with no recurrence of deformity. (**G**) A clinical picture at the 1-year followup shows full knee extension, good alignment, and enhanced fit of the prosthesis.

Radiographic assessment included a 51-inch standing X-ray obtained with the patient wearing her prosthesis. We analyzed the deformities according to the method developed by Paley [13]. The right lower extremity length was 80.4 cm, femoral length was 43.8 cm, and tibial length was 36.7 cm. The lateral distal femoral angle was 89° and the medial proximal tibial angle was 87°. There was a 7-mm medial mechanical axis deviation. The total length of the left lower extremity was 41.8 cm, femoral length was 30.2 cm, and tibial length was 11.8 cm. The lateral distal femoral angle was 74° and the medial proximal tibial angle was 63°. It was not possible to measure the mechanical axis deviation, as it is measured from center hip to ankle. We do not believe this measurement is reliable or even meaningful when the patient wears a prosthesis. Her leg-length discrepancy was 38.6 cm (discrepancy from femur, 13.6 cm; discrepancy from tibia, 24.9 cm). An anteroposterior radiograph showed a proximal tibial varus deformity measuring 29° (Fig. 1B). A full extension lateral radiograph (Fig. 1C)

Table 1. Summary of leg lengths, angles, and deformity parameters

Patient	Side	Total leg length (cm)	Femoral length (cm)	Tibial length (cm)	Δ Femur (cm)	Δ Tibia (cm)	MAD (mm)	LDFA	MPTA	PPTA	Deformity parameters
1	Right	80.4	43.8	36.7			7 medial	89°	87°		LLD = 38.6 cm short
	Left*	41.8	30.2	11.8	13.6	24.9	Immeasurable	74°	63°	114°	Left proximal tibial varus = 29°
2	Right*	65.5	37	30.2	12.2	8.3	80 lateral	95°	123°		LLD = 22.2 cm short
	Left	87.7	49.2	38.5			15 medial	87°	83°		Left proximal tibial valgus = 30°

* Diseased leg; Δ = difference in measurements; MAD = mechanical axis deviation; LDFA = lateral distal femoral angle; MPTA = medial proximal tibial angle; PPTA = posterior proximal tibial angle; LLD = leg-length discrepancy.

showed no hyperextension and the posterior proximal tibial angle measured 114° (Table 1).

An above-knee amputation had been recommended by other surgeons to improve prosthetic wear. We believed that saving her knee and lengthening and straightening her tibia would improve her prosthetic wear and function. We planned a proximal tibial osteotomy to correct the deformity and add modest length to her residual leg. She then would be fitted with a prosthesis to fit the longer and straighter residual leg. We corrected the deformity by osteotomizing the tibia and fibula and applying an Ilizarov/Taylor spatial frame^{TM} (TSF) (Smith and Nephew Inc, Memphis, TN) (Fig. 1D). The distal femur was not approached, which left the knee line with some obliquity. The posterior proximal tibial angle of 114° suggested there was extension deformity in the bone with a flexion contracture of the knee. This was not changed because full knee extension was achieved on physical examination. The total time she wore the frame was 4 months. A total length of 5 cm and full correction of the deformity were achieved (Fig. 1E–F). At the latest followup 24 months posttreatment, the range of motion in the left knee was 0° to 130°. The residual limb measured 15 cm in length. She had no functional limitation arising from difference in knee height. A new prosthesis was applied with complete relief of pressure symptoms at the distal part of the residual limb and marked improvement in gait was achieved (Fig. 1G).

Patient 2

A 35-year-old man presented with a right congenital leg-length discrepancy, secondary to a congenital femoral deficiency in combination with fibular hemimelia. Instability of the knee was not present. Surgical history included a distal femoral osteotomy, partial growth plate closure, and removal of some prominences of his distal femur. He used a foot-in-foot prosthesis (the foot is placed into a socket in the equinus position) since childhood. He had mild occasional knee pain.

The distance from the heel to the floor was approximately 20 cm. When standing, he had a large valgus deformity (Fig. 2A). He felt comfortable with a 14.5-cm block under his right foot but had an equinus deformity and his heel was approximately another 5 cm off the block. Knee motion on the right was –5° extension to 125° flexion and 20° valgus deformity clinically. Knee motion on the left was full extension to 130° flexion. Ankle motion on the right was –30° dorsiflexion to 70° plantar flexion. On the left, there was 15° dorsiflexion to 40° plantar flexion. Neurologic and vascular examinations were normal.

Radiographic assessment included a 51-inch standing radiograph (Fig. 2B). We analyzed the deformities according to the method developed by Paley [13]. The right lower extremity length was 65.5 cm, femur length was 37 cm, and tibia length was 30.2 cm. The mechanical axis deviation was 80 mm lateral to the midline. The lateral distal femoral angle was 95° and medial proximal tibial angle was 123°. The patient had a dysplastic knee with a well-maintained joint space. On the normal left side, the femur length was 49.2 cm, tibia length was 38.5 cm, and mechanical axis deviation was 15 mm medial to the midline. The lateral distal femoral angle was 87° and medial proximal tibial angle was 83°. His overall leg-length discrepancy from hip to ankle was 22.2 cm (femoral discrepancy, 12.2 cm; tibia discrepancy, 8.3 cm). There was a 30° valgus deformity that stemmed predominantly from the proximal tibia (Table 1).

Our treatment goal was to place the mechanical axis of the right leg through the center of the knee to improve prosthesis fit, gait, and knee pain and to prevent progression of arthritis. The treatment plan included correction of the valgus deformity at the tibia using a TSF. The leg shortening and the equinus contracture were not corrected because the patient felt comfortable using the foot-in-foot prosthesis. The option of equinus correction and greater limb lengthening was discussed, but the patient preferred the current

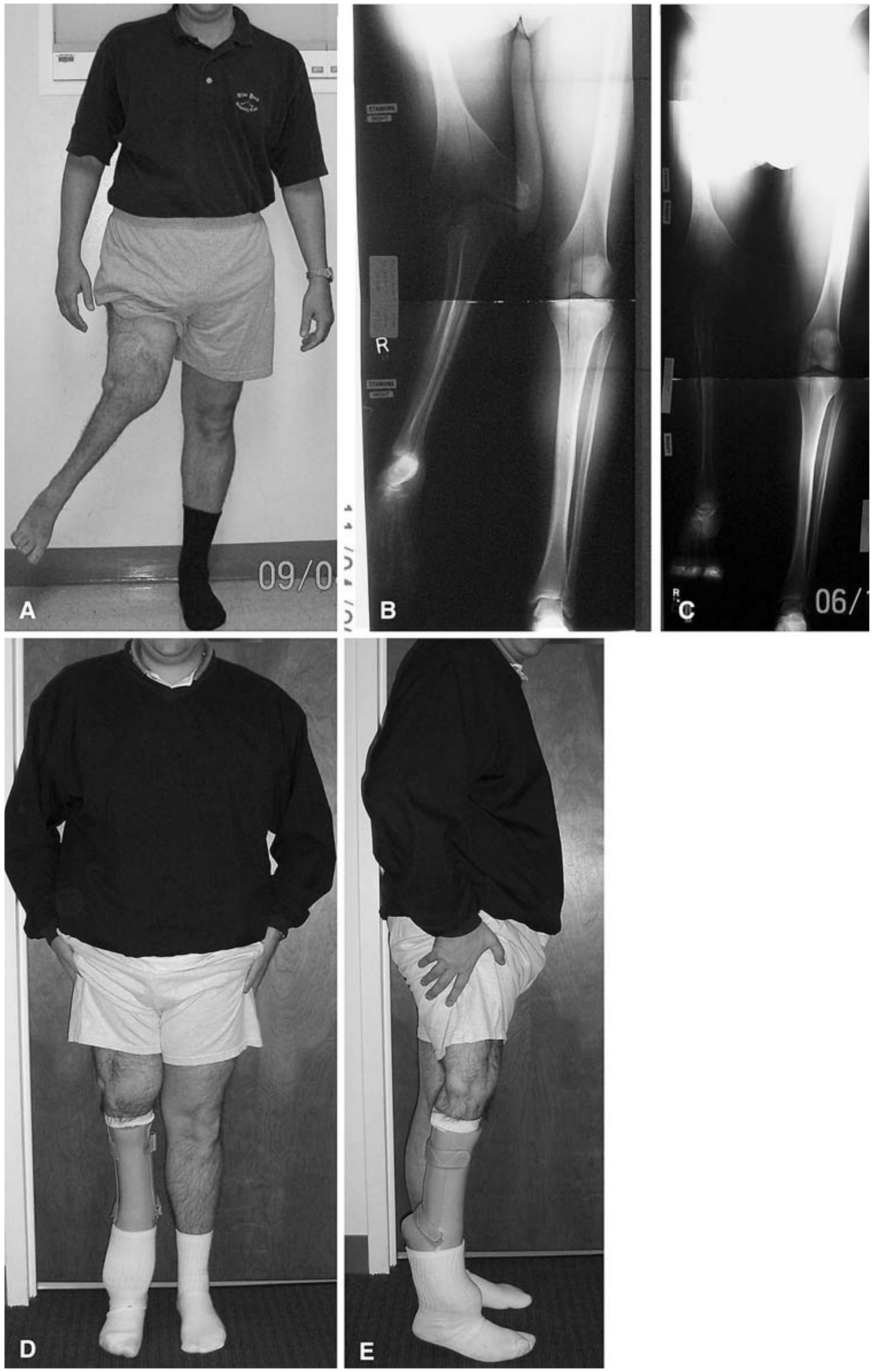

Fig. 2A–E (**A**) A preoperative clinical picture and (**B**) a standing radiograph show the right leg deformity. (**C**) A standing radiograph shows correction of the right leg deformity. The mechanical axis line runs through the center of the knee. (**D**) Frontal and (**E**) side views show the patient with a well-aligned leg and well-fitted foot in the foot prosthesis.

treatment. He had limited time available to wear the TSF and the ankle position was not a problem for him. The total time wearing the frame was 4 months, with uneventful treatment and followup. At the latest followup 30 months posttreatment, the range of motion of the right knee was 0° to 125° with no instability. Mechanical axis deviation was 0, and leg-length discrepancy was 18.5 cm. Full deformity correction was obtained. The difference in knee height introduced no apparent functional limitation. His ability to wear a prosthesis and his gait were improved (Fig. 2C–E).

Discussion

Adequate length and alignment of the residual limb are paramount for optimal prosthetic wear. Factors that determine the efficiency of using a prosthesis include sufficient power to perform the gait cycle and sufficient residual limb surface area to transmit load without skin breakdown. Adequate range of movement of the joint adjacent to the residual limb also is important because no benefit would be gained from lengthening a residual limb with a stiff joint [3]. The Ilizarov method can be used for lengthening and reconstruction of a short, deformed residual limb [1].

The TSF is an evolution of the original Ilizarov frame and uses the same concepts of distraction osteogenesis as the classic frame. It is used with a computer program that produces a numerical schedule that can be dialed on the struts to simultaneously correct length and all aspects of deformity, including angulation, translation, and rotation. The computer program is fed with deformity and frame-mounting parameters to produce the schedule.

Several reports suggest lengthening is a reasonable option for improving prosthetic wear and helps avoid the need for revision surgeries that normally involve amputation of the limb at a higher level to overcome the deformity [9–12]. Studies suggest below-knee amputation is superior to above-knee amputation, with better gait efficiency and prosthesis fit [8, 14]. In addition, optimizing alignment of the preserved knee is important to prevent abnormal force transmission across the knee with resultant pain and arthritis progression. Preventing knee arthritis in congenital deformities is important because standard arthroplasty options may not be applicable. Lengthening and deformity correction can only be performed safely in the presence of a well-contoured soft tissue envelop and supple movement of the adjacent joint. During distraction, the patient is at risk of having contractures and stiffness develop. Therefore it is important to follow a strict passive and active physical therapy regimen to avoid loss of knee extension or flexion [5].

The problem that arises from lengthening in patients with limb deficiencies is difference in knee heights. We believe having the knee heights at different levels is not ideal but is second in priority to equal leg length and normal alignment. We are not aware of specific information regarding gait and different knee heights. The first patient was offered femoral lengthening to decrease knee-height discrepancy and decrease prosthesis length and weight, and she is considering these recommendations.

We believe distraction osteogenesis using the TSF is a reliable method for correction and lengthening of congenital limb deficiencies, with improved prosthesis fit, cantilever function in lifting the prosthesis, and gait efficiency.

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