Functional Assessment in Tibial Hemimelia (Can We Also Save the Foot in Reconstruction?)

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Background: The congenital absence of the tibia is a rare disease, and an orthopaedic surgeon may not encounter such cases during the course of his/her career. This is the largest report to date of the management of such cases by a single surgeon. The foot and leg were persevered in the majority of the cases, and a functional evaluation system was used to report outcomes.

Methods: Thirty-six patients with tibial hemimelia, who had been under the direct care of the authors since infancy, were evaluated clinically and radiographically. The patients or their parents filled out the Pediatric Quality of Life and the parents' satisfaction forms. The surgical interventions performed, and their effects on school attendance and, and also the shoe type they wore were documented.

Results: Thirty-six patients (19 girls and 17 boys) with 48 tibial-deficient limbs (19 right, 5 left, and 12 both right and left sides) were studied. The patients were assessed at 12 years (2.5 to 32.5 y), with a mean follow-up of 9 years (2 to 23 y). The 48 limbs included 14 type I, 16 type II, 11 type IV, and 7 unclassified by using the Jones classification; and 6 type I, 11 type II, 16 type III, 1 type IV, and 14 type VII by using the Weber classification. Primary amputation was performed in 8 patients (10 limbs) and limb preservation surgeries on 38 legs (28 patients). Tibiofibular synostosis, centralization of the ankle, and Ilizarov lengthening were the most common procedures. Nonunion of tibiofibular synostosis (2 cases) and knee stiffness (6 cases) were the main complications. Among the reconstructed limbs, 12 were in regular and 18 in modified shoes. The Pediatric Quality of Life of 68 points in the reconstructed group was a significant achievement, and it was also better than the score of patients who had undergone amputation.

Conclusion: Reconstruction of tibial hemimelia with foot preservation provides good functional outcome in the majority of cases.

Level of Evidence: Level IV.

Key Words: tibia, congenital limb deformities, lower limb, abnormalities, congenital defects

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The congenital absence of the tibia is a rare anomaly and is usually accompanied by congenital abnormalities in other parts of the body.[1,2] Familial incidence of this deficiency is also seen not infrequently.[1,3,4] Tibial hemimelia is seen in several syndromes, and gene foci have been also assigned to this anomaly.[5–7] The associated foot anomalies, including tibial ray(s) deficiencies or tarsal coalitions, have been reported, as well as hand malformations.[8–11]

Several investigators, including Jones and colleagues,[12–14] have classified this preaxial longitudinal tibial deficiency. Weber[15] proposed a different classification, changing the Jones' 4 groups into 7 groups to encompass some of the rare forms of the disease.

The common treatment strategy has been amputation of the limb or the foot over the years.[1,10–16]

We would like to report the largest reported series of tibial hemimelia from 1 center where foot and leg preservation is the main focus of treatment—partly because parents refuse amputation for their children. The functional status of the limb-preserving surgeries has also been assessed in this study.

METHODS

All cases of tibial hemimelia that were treated by the senior author (G.H.S.) during the last 23 years (1989 to 2012) were collected from the medical records at the Namazee Hospital—the main teaching hospital of Shiraz Medical University. The patients were called in for clinical examination and radiographic assessment. The original radiographs were reevaluated and classified in accordance with both the J (Jones) and W (Weber) classifications.

The treatment plan was limb reconstruction, including foot preservation in all the cases except for type I Jones's cases with no evidence for quadriceps function, and when foot had only 1 or 2 rays and was very misshapen.

The patients and/or their parents filled out questionnaires relating to their daily living activities, sports activities, social and psychological issues, schooling, etc.

Pediatric Quality of Life Inventory (PedsQL) questionnaires were also filled. The PedsQL is a scoring system that has various disease forms and has been used in many different pediatric conditions.[17] The musculoskeletal section, which has been validated in Persian language,[18] consists of 4 major issues of physical, emotional, social, and schooling functions. We employed the points such that the maximum would be 100.

The possible use of prosthetic limbs and their problems, as well as the use of orthoses or custom-made shoes were also assessed. The cosmetic concerns, limb alignment issues, and patients' and/or parents' satisfaction were also assessed. Only the cases that had already received treatment in our center and had at least 2 years of follow-up were included. The data were analyzed using SPSS 22 software.

The medical ethics committee of Shiraz Medical University approved the research. No financial support was received from any financial group, and no grant was provided by any financial or academic center.

RESULTS

Among the 40 tibial hemimelia patients who had received treatment by the senior author, 36 (48 legs) could be reached and were evaluated. They included 17 male and 19 female patients, with a mean age of 12 years and 1 month (range, 2.5 to 32.5 y) at follow-up. Nineteen legs had right tibial deficiency: 5 left and 12 both sides. The period of follow-up was 9 years (minimum 2 y, maximum 23 y). In accordance with the J classification,[13] the cases included 14 type I, 16 type II, 0 type III, and 11 type IV. Seven cases did not fit any of the J categories. The W classification system,[16] however, had 6 type I, 11 type II, 16 type III, 1 type IV,14 type VII, and 0 types V or VI. The 7 cases that could not be classified according to the J classification could be classified according to the W classification as type I or VI (Table 1).

Although tibial deficiency was an isolated musculoskeletal anomaly in 81% of cases, associated other abnormalities (more commonly upper limb) were present in 7 patients, (19%), which included radial club hand, cleft hand, 1 bone forearm, syndactyly, and polydactyly, developmental dysplasia of the hip, and proximal femoral focal deficiency.

Four patients (11%) who had type I J deficiency had positive family history for tibial hemimelia. Six patients (17%) had concomitant other congenital musculoskeletal deformities.

Forty-eight limbs had undergone a total of 77 operations as shown in Table 2. Ten limbs (8 patients) had undergone amputation—9 knee disarticulation, all in type I Jones (type VII W) and 1 Syme. (The amputations had been all done over the age 5, as the parents could not be convinced to consent to amputation earlier.) The other 38 limbs (28 patients) had undergone reconstructive surgeries for limb (including foot) preservation: tibiofibular synostosis (20 limbs), centralization of the ankle underneath the tibia (23 limbs), Ilizarov lengthening (18 limbs), and fibular centralization over the femoral condyle—Brown procedure (5 limbs). The remaining corrective procedures on the feet or contralateral epiphysiodesis for leg equalization are depicted in Tables 1 and 2.

The knees in the 5 type I were stiff: the 3 type Ia had 20 to 50 degrees of flexion contracture, but from 100 to 130 degrees of flexion; whereas the 2 type Ib had from 10 to 130 degrees of flexion motion. Two of type Ia were using occasional knee brace, and the other 3 walked without any brace or support.

The 38 limbs that received reconstructive surgery rather than amputation are reported in more detail as follows.

Tibiofibular synostosis was performed in 13 limbs of type II, 6 limbs of type IV, and 1 of type Ib J deficiencies, followed by limb-lengthening procedures according to the Ilizarov principles. The initial synostosis surgery would not unite in 2 patients and additional surgery was required for union. (These 2 cases received their initial surgery around 3 y of age, and had been fixed with screws.) The Ilizarov lengthening average of 10.8 cm (3.5 to 19 cm) was achieved in 18 limbs (15 cm in type I J, 10.7 cm in type II J, 11 cm in type IV J, and 6.5 cm in type I W). In 5 limbs, lengthening was achieved in 2 different stages. Contralateral epiphysiodesis was performed in 3 cases.

In terms of utilization of ambulatory aids, 34 cases were walking independently, that is without walking aids. Among the shoe type worn by the 30 reconstructed limbs that had completed treatment, 12 wore regular shoes and the remaining 18 wore modified shoes or orthoses (Tables 1 and 2). The orthoses, in the ones who used them, were mainly shoe fillers or were providing stability in long-distance walking, or providing lift in the 8 limbs that were waiting for their secondary operation.

The knee and ankle/foot motion in the reconstructed group were as follows: the knee motion was full in all, except for 5 type I J and 2 type II J cases that had abnormal-shaped knee joints from the beginning. The ankles were very stiff in 14, and 22 had around 15 degrees of motion arc. The subtalar joint was almost nonexistent before or after surgery, considering the fact that subtalar abnormality and fusion was present in the majority of cases.

The position of the limb on ankle was centralized, and nicely dome-shaped distal fibulae was seen in the types I and II (16 ankles). Six of the 11 type IV cases had been surgically centralized, that 5 remained centralized and in the remaining case the centralization was partially lost in follow-up.

In the reconstructed group 14 patients were quite active in sports, and the other only occasionally participated in any sports appropriate for their age.

The quality-of-life questions were analyzed in more detail. The overall PedsQL score was 68 points for the reconstructed group, 64.6 for the amputated group. (Amputation scores, of course, reflected the fact that only 2 type IA limbs were unilateral and 6 were part of the bilateral deformity.) The data were analyzed using *t* test, the Mann-Whiney test, and the Kruskal-Wallis analysis for comparing the functional outcome in different categories of tibial deficiency and also for correlating the surgical procedure with the outcome. The reconstructed group had a better functional score than the amputated group in the 4 groups of physical, social, psychological, or schooling scores when assessed separately—noting again that most amputated cases were part of bilateral hemimelia cases (Table 3). The lowest scores in the physical

TABLE 1. Characteristics and Treatments in Accordance With Type in 48 Limbs With Tibial Hemimelia

Type	Number	Follow-up	Associated Anomaly	Family hx for Tibia Hemimelia/Congenital Leg Anomaly	Type of Surgery	No. Surgery Per Limb in Recostruction	Footwear
I (J) VII (W)	14	7 y	0	5	KD + B + AC + S + L	1.4	9P, 4O, 1RS
II (J) III (W)	16	9 y + 7 mo	4	1	AC + S + L + E + Fem.ost + Syme	2.2	1P, 7O, 3RS, 3MS
III (J) IV (W)	0	0	0	0	—	—	—
IV (J) II (W)	11	10 y + 9 mo	2	2	AC + S + L + FO	1.8	3O, 4RS, 4MS
? (J)	7	8 y + 6 mo	1	2	AC + S + L + ATL	1.3	3O, 5RS, 1MS
I (W)	6	9 y + 10 mo	0	2	L + ATL + E +	1.5	
VI (W)	1	18 mo	0	0	AC + S + Post.cap	1	

? indicates unclassified in Jones classification.

AC indicates ankle centralization; ATL, Achilles tendon lengthening; B, Brown; E, epiphysiodesis; Fem.ost, femoral osteotomy; FO, foot osteotomy; J, Jones; KD, knee disarticulation; L, lengthening; MS, modified shoe; O, orthotics; P, prosthesis; Post.cap, posterior capsulatomy; RS, regular shoe; S, Synostosis; TT, tendon transfer; W, Weber.

group of questions were noted for long-distance walking or running; in the social issues, it was mixing with or being as active or competitive as peers in sports; in the psychological questions, it was losing temper quickly; and in the schooling questions, it was being attentive during school home works. Interestingly, they did not have any issues with regard to being teased or bullied by other children or having fallen behind in any school education because of their surgeries.

In terms of satisfaction with the treatment, the results—recognizing that 8 cases still await repeat lengthening—were as follows: in the amputated cases, 4 patients were fully and 4 were partially satisfied. In contrast, from the 28 patients (38 limbs) who had undergone reconstructive surgery, in terms of function, 8 (29%) were fully satisfied, 19 (67%) almost satisfied, and 1 (4%) unsatisfied. This unsatisfied case is a 12-year-old boy who plays soccer following his ankle centralization, tibiofibular synostosis, and 4-cm lengthening, and he is waiting to undergo another 7-cm lengthening procedure. The same satisfaction status applies for esthetic appearance. In all, 32% were fully satisfied, 50% were almost satisfied, and 18% were unsatisfied with the esthetic appearance of the limb. None of these patients would want an amputation at any level or would have opted for one, having known the outcome.

DISCUSSION

Tibial hemimelia is a very rare, genetically linked deficiency, which is frequently associated with other musculoskeletal anomalies in addition to lower limb shortening.[19–22] The limb preservation option in tibial hemimelia is dependent on the following issues: (1) knee stability; (2) the expected limb shortening; and (3) the correct ability of foot deformity.

The possibility of lengthening a very short bone segment with the Ilizarov technique and principles makes length equalization in tibial hemimelia a more effective treatment option, whether with 1- or 2-stage lengthening, with or without contralateral epiphysiodesis.

We aimed at obtaining and maintaining a plantigrade foot with a good heel pad for weight-bearing, in equal-length lower limbs—that are so important for walking on carpeted floor in regular Persian rooms (Fig. 1). Amputation, in our culture, is not a well-perceived procedure and is not often accepted by the families, similar to Egyptian society.[23] The authors frequently encounter older children and adolescents with various types of congenital limb deficiencies who walk on their knees and use wheelchair and still would not consent to any amputation. Having a foot rather than a Syme is psychologically more acceptable to the parents; it also provides a wide landing area, and there will often be no need for orthotics[24] (Fig. 2).

Seventy percent of our patients were using regular or slightly modified shoes and were comfortably walking barefooted at home.

Knee stability is the main concern in tibial deficiency type I J (type VII W) (Fig. 3). Five patients who refused amputation and underwent the Brown procedure with the secondary lengthening procedures had stiff but stable knees, and despite the deformity, often walked with no knee support and were satisfied with the outcome. Epps et al[25] reported on 14 patients who had undergone Brown fibular centralization. Seven of those cases required later knee disarticulation, and the rest had awkward gait owing to knee flexion contracture. The interesting point is that 13 of those cases also had Syme amputation at the time of centralization. Christini et al[26] and Simmons et al[27] found satisfactory results in patients who underwent the Brown procedure in the presence of strong to moderate strength quadriceps function. Their cases had also undergone Syme amputation. We, however, believe that with a plantigrade and functional foot, the Brown procedure, with preservation of the foot followed by fibular lengthening, is an acceptable option[3] (Fig. 4). Long time use of orthotics during early childhood can stop the knee flexion deformity, and the

TABLE 2. Case Details of 36 Tibial Hemimelia Patients

	Sex	Side	Type	Treatment	Age at First Surgery	No. Surgery	Ambulatory Aid	Footwear	Follow-up	PedsQL Score	Future Surgery
1. D.M.	M	Rt	Ia (J) VII (W)	KD	11 mo	1	N	P	9 y	50.8	N
2. K.T.	F	Rt	Ia (J) VII (W)	KD	12 mo	1	N	P	3 y + 6 mo	73.1	N
3. N.S.	F	Rt	Ia (J) VII (W)	KD	8 y	1	N	P	3 y	60.5	N
		Lt	Ia (J) VII (W)	KD	8 y	1	N	P	3 y	60.5	N
4. A.G.	M	Rt	Ia (J) VII (W)	KD	3 y	1	N	P	4 y	60.1	N
		Lt	Ia (J) VII (W)	B + AC	3 y	1	N	O	4 y	60.1	Fem.ost + TT
5. N.R.	F	Rt	Ia (J) VII (W)	KD	5 mo	1	N	P	2 y + 6 mo	71.9	N
		Lt	IV (J) II (W)	AC + S	5 mo	1	N	O	2 y + 6 mo	71.9	S
6. A.M.	F	Rt	Ia (J) VII (W)	D	7 mo	1	Y	P	3 y + 6 mo	79.4	N
		Lt	II (J) III (W)	AC + S	1 y + 4 mo	1	Y	O	3 y + 6 mo	79.4	N
7. A.R.	M	Rt	Ia (J) VII (W)	KD	1.5 y	1	Y	P	17 y + 6 mo	76.3	N
		Lt	II (J) III (W)	S + Syme	16 y	2	Y	MS	17 y + 6 mo	76.3	N
8. S.Z.	F	Rt	Ia (J) VII (W)	B + AC + L	3 mo	1	N	O	11 y + 9 mo	46.6	LE
9. A.N.	M	Rt	Ia (J) VII (W)	B + AC	5 mo	1	N	O	3 y	75.2	LE
		Lt	IV (J) II (W)	AC + S	1 y + 2 mo	1	N	O	3 y	75.2	N
10. A.H.	M	Rt	Ia (J) VII (W)	KD	1 y + 6 mo	1	Y	P	2 y + 6 mo	56.5	N
		Lt	II (J) III (W)	AC + S	7 mo	1	Y	O	2 y + 6 mo	56.5	Ost
11. S.F.	F	Rt	Ia (J) VII (W)	B + AC + L	5 mo	2	N	RS	23 y + 6 mo	–	N
12. Y.E.	F	Rt	Ib (J) VII (W)	B + AC + S	4 mo	2	N	O	4 y	89.8	LE
13. S.S.	M	Lt	II (J) III (W)	S + L + E	4 mo	5	N	MS	23 y + 6 mo	59.7	N
14. A.B.	M	Rt	II (J) III (W)	AC + S + L	9 mo	2	N	MS	6 y	92.2	N
		Lt	? (J) I (W)	ATL + Post.cap	9 mo	2	N	MS	6 y	92.2	N
15. M.D.	M	Rt	II (J) III (W)	AC + S + L + E	6 mo	4	N	O	15 y	95.9	LE
16. R.B.	F	Rt	II (J) III (W)	AC + S + L + FO + Fem.ost	6 mo	4	N	RS	13 y + 6 mo	64.2	N
		Lt	? (J) I (W)	E	12 y	1	N	RS	13 y + 6 mo	64.2	N
17. R.D.	M	Rt	II (J) III (W)	AC + S + L	12 mo	2	N	O	13 y	43	N

TABLE 2. (*continued*)

	Sex	Side	Type	Treatment	Age at First Surgery	No. Surgery	Ambulatory Aid	Footwear	Follow-up	PedsQL Score	Future Surgery
18. R.Z.	F	Rt	II (J) III (W)	AC + S + L	1 y + 6 mo	3	N	O	6 y	44.2	N
19. K.G.	F	Rt	II (J) III (W)	A.C + S	1 y + 3 mo	1	N	O	2 y + 9 mo	64	S
20. Z.A.	F	Rt	II (J) III (W)	AC + L	12 mo	2	N	RS	5 y + 6 mo	–	N
21. V.Z.	M	Lt	II (J) III (W)	S + L	12 mo	2	N	MS	20 y	55	N
		Rt	IV (J) II (W)	FO	12 mo	2	N	RS	20 y	55	N
22. A.A.	M	Rt	II (J) III (W)	S	9 mo	1	N	O	5 y + 3 mo	46.9	LE
23. A.V.	M	Lt	II (J) III (W)	L	8 y	1	N	MS	2 y	87.1	N
		Rt	IV (J) II (W)	ATL	8 y	1	N	RS	2 y	87.1	N
24. A.A.	M	Rt	IV (J) II (W)	AC + S + L	6 mo	3	N	MS	11 y + 6 mo	40.5	LE
25. S.N.	F	Rt	IV (J) II (W)	AC + S + L + E	7 mo	3	N	RS	14 y + 6 mo	58.5	N
26. F.T.	F	Rt	IV (J) II (W)	AC + S + L + TT	1 y + 2 mo	2	N	O	13 y	–	LE
27. O.D.	M	Lt	IV (J) II (W)	AC + S	4 y	1	N	RS	1 y + 6 mo	76.7	N
28. Z.S.	F	Rt	IV (J) II (W)	L + FO	27 y	2	N	MS	3 y	45.8	N
29. F.F.	F	Rt	IV (J) II (W)	L	20 y	2	N	MS	12 y	70.3	N
30. A.M.	M	Rt	IV (J) II (W)	ATL	6 mo	1	N	MS	31 y + 6 mo	72.2	LE
31. E.H.	F	Lt	? (J) I (W)	L	16 y	1	N	RS	4 y	59.5	N
32. H.J.	F	Rt	? (J) I (W)	ATL + L	12 mo	2	N	RS	6 y	54.8	N
33. M.J.	M	Lt	? (J) I (W)	E	13 y	1	N	RS	19 y + 6 mo	72	N
34. Z.D.	F	Rt	? (J) I (W)	L	6 y	1	N	RS	18 mo	90.8	N
35. A.S.	M	Rt	II (J) III (W)	AC + S	3 y	1	N	O	2 y	61.5	N
		LT	II (J) III (W)	AC + S	3 y + 6 mo	1	N	O	1 y + 6 mo	61.5	N
36. Y.H.	F	Lt	? (J) VI (W)	AC + Fib fusion	5 mo	1	N	O	1 y + 6 mo	–	LE
Total						77					

? indicates unclassified in Jones classification.

AC indicates ankle centralization; ATL, Achilles tendon lengthening; B, Brown; E, epiphysiodesis; Fem.ost, femoral osteotomy; Fib, fibula; FO, foot osteotomy; J, Jones; KD, knee disarticulation; L, lengthening; LE, limb equalization; Lt, left; MS, modified shoe; N, no; O, orthotics; P, prosthesis; Post.cap, posterior capsulatomy; RS, regular shoe; Rt, right; S, Synostosis; TT, tendon transfer; W, Weber; Y, yes.

TABLE 3. Four-Group PedsQL (Pediatrics Quality of Life) in Reconstruction and Amputation Groups

	Physical Score	Social Score	Psychological Score	Schooling Score	Total Score
Reconstructed group	61.6	71.2	62.5	76.1	67.8
Amputated group	54.9	63.7	75	65	64.6
Reconstructed and amputated group	54.7	73.7	73.7	–	67.4

resultant weight-bearing limb is better than a knee disarticulation.[2,24,26] Early amputation at the knee level is only indicated in cases of type 1 tibial deficiency and very severe foot deformity and deficiency. In our culture, the parents look at the Syme or Boyd amputation the same as knee disarticulation.

For most other types of tibial hemimelia, amputation at the foot or knee level has been the recommended treatment over the years.[10,25] There have been, however, several recent reports on limb preservation.[23,24] The multicenter study of Schoenecker et al[10] had 86% ablative surgery either at the foot or knee level. Only in 14% of cases, the foot had been preserved, and even their type I cases that had originally been centralized at the knee and had developed severe knee flexion contracture were later disarticulated. In the present report, we have shown that 78% of the limbs and feet were preserved and are functioning well. The 5 type I cases that underwent reconstruction with fibula-femoral centralization are too few for any statistical comparison. Hosny[23] reported successful results in 4 cases of Brown procedure.

Type IV deficiencies, in contrast to previous reports,[10,11,28,29] were treated successfully with limb (including foot) preservation in our series. Tokmakova et al[11] reported on 11 type IV cases, and only 3 received ankle reconstruction and 7 had Syme or Boyd amputation. We believe that retaining the foot and keeping it plantigrade is

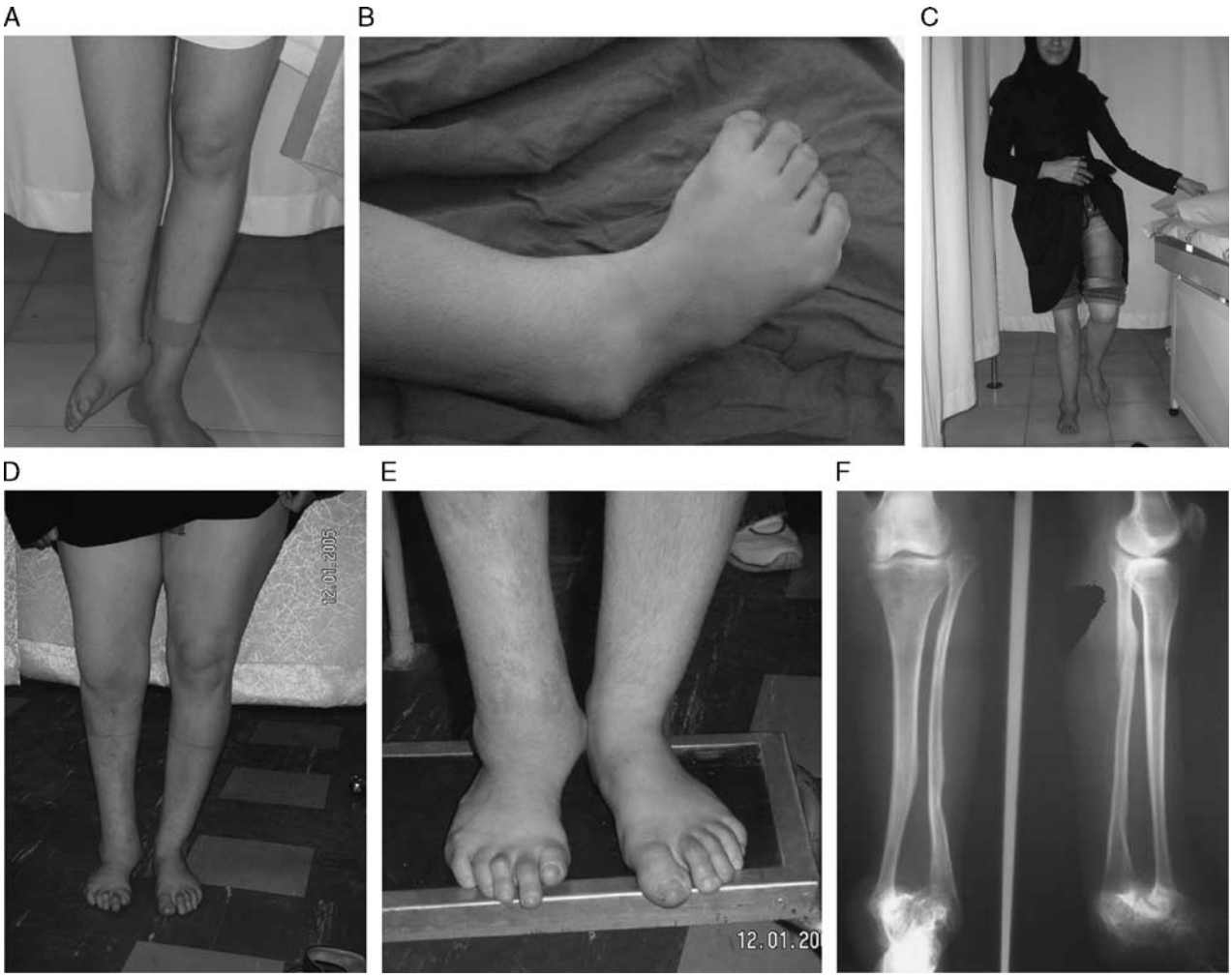

FIGURE 1. Type IV tibial hemimelia. A and B, Severe foot deformity and shortening. C and D, Standing photographs following 8 cm lengthening and correction of foot deformity. E, Broad ankle with plantigrade foot, using regular shoe. F, Radiograph of well-aligned leg with ankle diastasis.

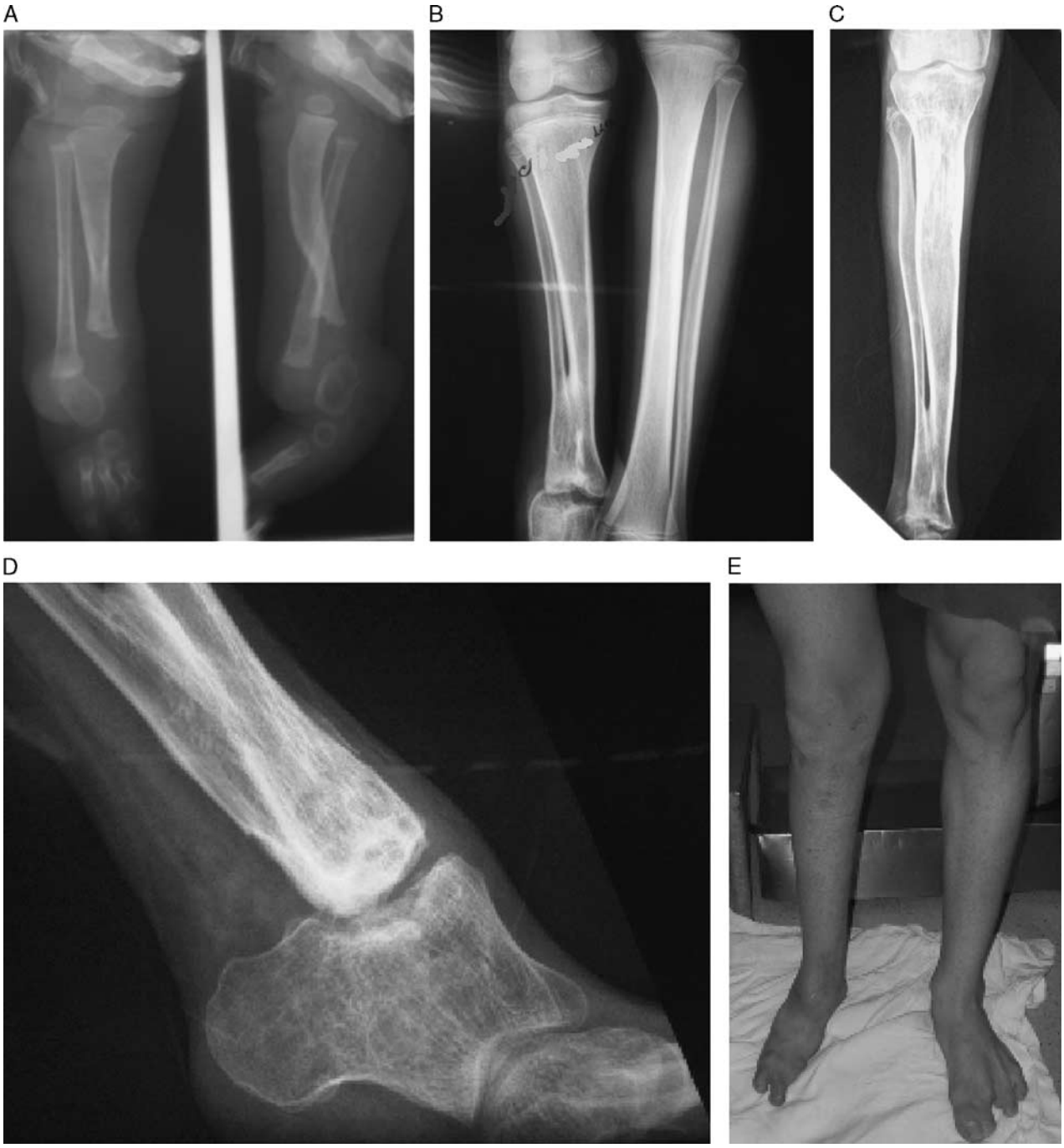

FIGURE 2. Unilateral tibial hemimelia type II. A, Radiographs at infancy, showing a single hind foot bone. B, At age 10, following tibiofibilar synostosis and ankle centralization. C, At age 17, following 12 cm lengthening with Ilizarov fixator. D, A lateral view of the ankle on the single hind foot bone. E, Clinical picture of the leg and the plantigrade foot, fitting in a regular shoe.

necessary for a functional limb and may allow a regular shoe wear instead of prosthesis. The ankle with tibiofibular diastases in the type IV cases would function well and can be improved using tibiofibular synostosis, differential distal epiphysiodesis, and osteotomy. The foot and ankle are important elements in function despite some comments to the contrary.[29] The appropriate use of Ilizarov techniques and principles enable successful reconstruction.[23,24]

We encountered no neurovascular complications or need for secondary foot ablation or knee disarticulation. The complications that required secondary surgical remedies were foot and ankle deformities in 1 and failure in achieving synostosis in 2 limbs. Spiegel et al[28] noted skin

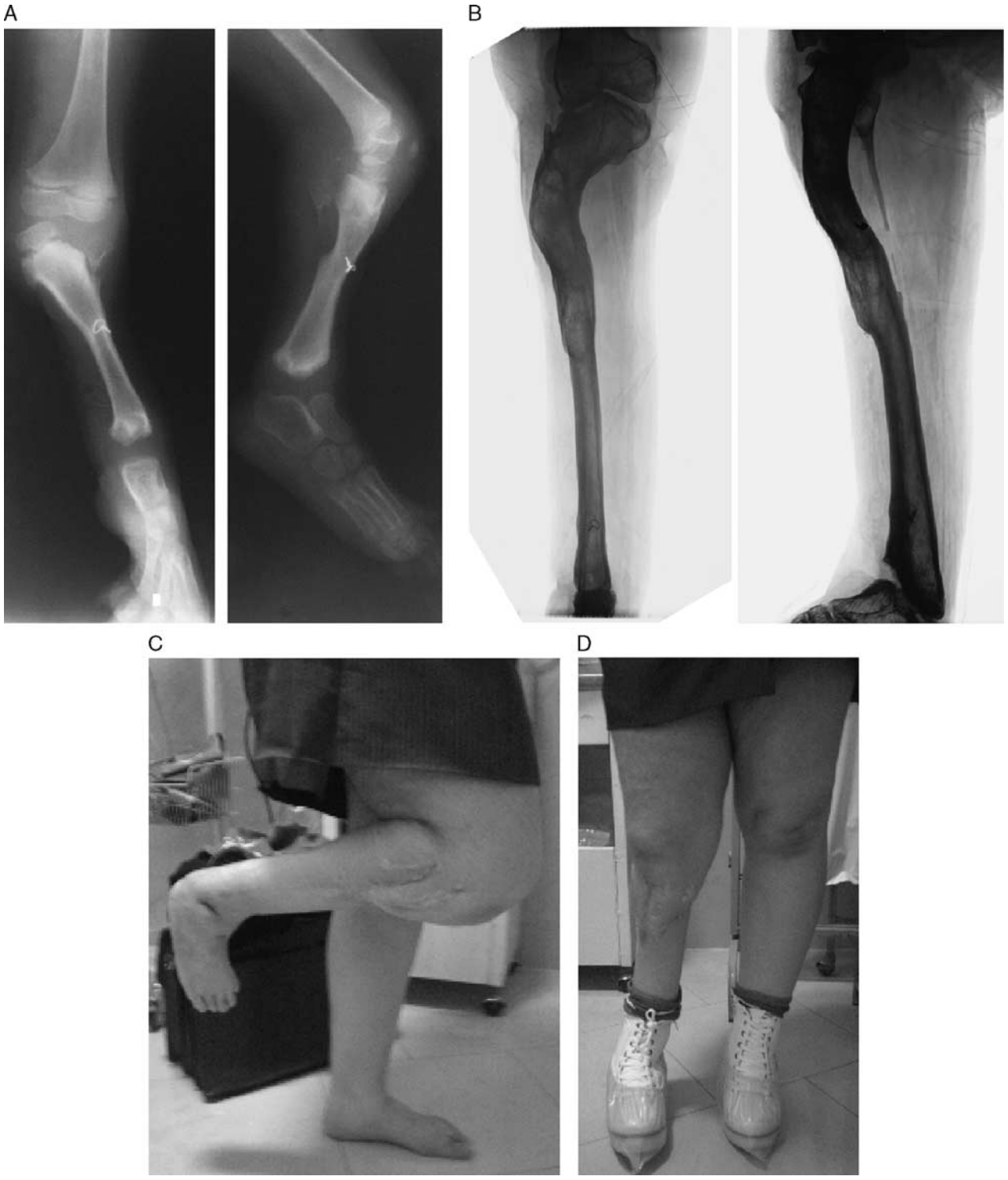

FIGURE 3. A 27-year-old lady with type IB tibial hemimelia and Brown procedure at infancy, and later fibular lengthening in 2 stages for a total of 18 cm and a contralateral proximal tibiofibular epiphysiodesis. A, At age 3, after Brown procedure. B, At age 27, after 2 stages of lengthening. C and D, Preserved foot, good range of motion of knee and regular shoe wear.

problems when the foot is ablated and also in varus deformity. Epiphysiodesis of the proximal fibular to avoid prominence at knee level and also varus deformity with prosthetic fitting problems have been mentioned in the literature.[28,30] Lately, we have been resecting the proximal fibular segments for our tibiofibular synostosis cases to

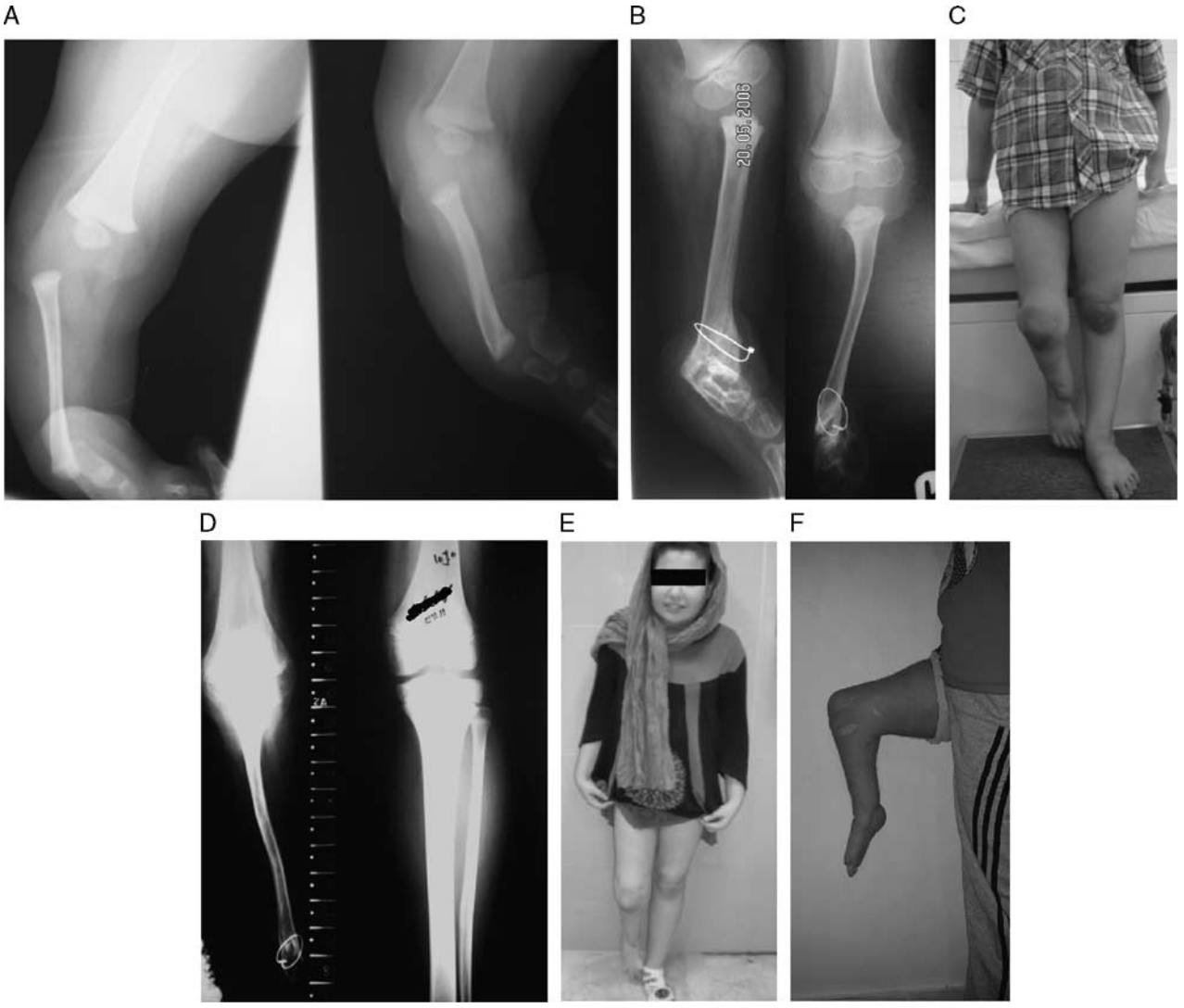

FIGURE 4. Type I tibia hemimelia. A, Preoperative x-ray. B, Radiograph at age 4. She received Brown procedure and ankle centralization at age 1.5 years. C, Clinical picture at age 4. D–F, Radiograph and clinical pictures at age 10, 1 year after first stage of Ilizarov legthening.

avoid the distraction force of fibular growth. This has been effective in decreasing pseudarthrosis in synostosis sites. The nonunion at tibiofibular synostosis has not been covered much in the literature.[10,26]

The functional outcome of the reconstructed limbs has not been adequately studied for tibial hemimelia in the past. Christini et al[26] reported on satisfactory functional outcomes for type I with the Brown procedure, which had been combined with foot ablation. The PedsQL was a very detailed questionnaire that covered all the relevant issues.[17] It is conceivable that longitudinally tibial deficiency, especially when it is often associated with other limb deformities as well, cannot be a fully normal individual. The PedsQL, when used in diabetic, juvenile idiopathic arthritis, congenital heart disease, etc., has been around 71 to 80 points.[31–33] The average score of around 68 is therefore a very good achievement. The preserved limb and foot cases—when specifically questioned—would have all chosen to keep the foot and the leg were they to decide again.

Thus far, this is the largest group of tibial deficiency to be reported by 1 surgeon's experience, in addition to a detailed functional outcome scoring. Foot and limb preservation was the main goal in these cases and was achieved in 78% of the cases. Knee disarticulation is suggested only for the type I deficiency that is associated with the severely deformed and not easily correctable foot. Centralizing the tibia or fibula on the ankle, fusing the tibia to the fibula, and then lengthening the limb in accordance with the Ilizarov technique and principles can manage all other types. The use of regular shoes or slightly modified shoes is attainable in the majority of cases. It will be also associated with complete or near-complete satisfaction of patients and parents.

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