Limb Salvage in Tibial Hemimelia

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Objective: *To study the results of treatment of tibial hemimelia with limb salvage procedure in term of patient satisfaction, clinical results and complications.*

Material and Method: *From 1993 to 2007 the authors treated six cases of tibial hemimelia with limb salvage procedures. Three legs of type Ia and four legs of type IV tibial hemimelia classified by Jones classification. The age at the operation ranged from 2 to 11 years. For type Ia cases, the Brown procedure, foot centralization and ilizarov lengthening of the fibula were used to correct limb length discrepancy. For type IV, the foot centralization, soft tissue release and ilizarov lengthening were used to correct limb length discrepancy. The follow-up range from 4 to 10 years.*

Results: *In two patients with type Ia, one patient could bear weight without gait aids, the other walked with orthosis and axillary crutch because this patient had bilateral Ia type and knee instability with progressive flexion contracture due to weakness of the quadriceps muscle. All patients with type IV can walk independently without gait aids. Three patients were performed limb lengthening. One case was fibular lengthening following Brown procedure in Ia type. Two cases were tibial lengthening in type IV. The mean lengthening was 5.1 cm. Mean lengthening index was 2.4. Satisfactory functional and cosmetic results were achieved in all patients with partial deficiency, whereas in patients with completely deficiency of the limbs, none of the 3 knees treated by fibular transfer achieved a satisfactory functional result because of insufficient quadriceps strength, progressive knee flexion contracture and persistent ligamentous instability. Nevertheless, in these 3 legs, all patients were ultimately able to withstand weight bearing.*

Conclusion: *Patients and families were satisfied even though patients must have multiple surgery to correct deformities of the foot and the knee joint, as well as leg-length discrepancy and also a prolong treatment time. Limb salvage procedure in tibial hemimelia is appropriate in Thai culture because patients can weight with bare feet in the house and have sensation in the feet.*

Keywords: *Tibial hemimelia, Brown procedure, Foot centralization, Limb length discrepancy*

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Congenital deficiency of the tibia (tibial hemimelia) is a very rare deformity of unknown etiology, occurring in 1/1,000,000 live births[1]. Although most cases occur on a sporadic basis, familial occurrence has occasionally been reported[2,3]. Patients with congenital deficiency of the tibia often have a wide range of accompanying congenital anomalies, including cleft lip and palate, spinal deformity, dislocation of the hip joint and hand deformity such as cleft hand, polydactyly, syndactyly, or absent fingers[2,3]. Flexion contracture of the knee and a skin dimple overlying the proximal tibia region are commonly present. The foot is rigid with varus and supination and frequently associated with medial ray defects[2]. Based on radiographic data the classification of Jones et al divides the type I tibial deficiency into 1a and 1b. In type 1a it is actually absent and without an extensor mechanism, whereas in type 1b it is present as a cartilaginous remnant that will later ossify, suggesting that the extensor mechanism may be intact. A very unusual variant with a diaphyseal and distal remnant of tibia but no proximal tibia is known as type 3. The diastasis of the distal tibiofibular joint is type 4.

Early amputation has generally been recommended for congenital deficiency of the tibia because it usually requires only one surgical procedure and allows the children to undergo straight forward prosthetic rehabilitation[2-6].

On the other hand while surgical correction of this deformity may obviate the need for a prosthesis it is very difficult and challenging[4,7-9]. Multiple surgical procedures are required to correct deformities of the foot and the knee joint, as well as leg-length inequality. However, there are many reports of considerable advantages to retaining the foot in countries where people live with bare feet inside the house or where people do not easily accept amputation because of

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their culture and faith[15-17]. The purpose of the present study was to demonstrate the results of limb salvage treatment of congenital deficiency of the tibia by reconstruction of the foot and knee joint followed by limb lengthening.

Material and Method

Between 1993 and 2007, 7 limbs salvage treatment were performed in 6 patients with congenital deficiency of the tibia (types Ia and IV). The age of patients at first operation ranged from 2 to 11 years with an average of 5.5 years. The details of the 6 patients were shown in (Table 1).

All 7 feet showed equinovarus deformity and in type Ia were centralized by placing the distal fibula into the posterior facet of the calcaneus at ages ranging from 2 to 12 years. Soft tissue release also performed to corrected the deformity of the feet. In one case with fixed equinous at the age of 12 years, the midfoot close wedge osteotomy was performed in order to corrected the deformity. The fibular transfer (Brown procedure) was performed in 3 limbs with complete deficiency of the tibia type (Ia) at ages ranging from 2 to 3 years. Operative treatment of each patient shown in (Table 2).

Limb lengthening with ilizarov was performed for leg-length discrepancy on the fibula in one case and two tibial lengthening in type IV. The details of limb lengthening shown in (Table 3). Postoperative radiographic evaluation was used to assess the amount of lengthening required. The lengthening index was calculated by dividing the healing time in months by the amount of lengthening achieved in centimeters.

Foot centralization technique

A complete posteromedial release of the foot was performed by making a curved incision from the medial to the lateral side. The distal end of the fibula was sharpened. The distal epiphysis of the fibula was

Table 1. Demographic Data of 6 patients

case	sex	side	Age at consultation (yr)	type	Age at first operation	F/U (MO)
1	M	R	3	Ia	3	130
2	F	R	1	IV	2	48
3	F	R	8 mo	Ia	2	50
		L		Ia	2	
4	M	L	11	IV	11	60
5	F	R	11	IV	11	60
6	M	R	1	IV	2	48

Table 2. Operative Procedure

case	side	type	Age at Operation (yr)	Procedure
1	R	Ia	3	Brown procedure
			3	Foot centralization + soft tissue release
			9	Fibular lengthening
2	R	IV	2	Distal tibiofibula fusion
			2	Foot centralization + soft tissue release
3	R	Ia	3	Brown procedure
			3	Foot centralization + soft tissue release
	L	Ia	3	Brown procedure
			3	Foot centralization + soft tissue release
4	L	IV	11	Foot centralization + soft tissue release
5	R	IV	12	Foot centralization + soft tissue release
				Midfoot closed wegde osteotomy
			12	Ilizarov tibial lengthening
6	R	IV	2	Foot centralization + soft tissue release
			2	Ilizarov tibial lengthening

Table 3. Result of Bone lengthening with Ilizalov

case	procedure	Total lengthening (cm)	Age at lengthening yr	Lengthening Index (cm/mo)
1	Fibular lengthening	1^{st} = 5 cm	9	1.4
		2^{nd} = 4 cm	11	4.2
5	Tibia lengthening	1^{st} = 5.7 cm	12	1.6
		2^{nd} = 7 cm	15	1
6	Tibia lengthening	4 cm	2	2

preserved carefully to promote good longitudinal growth of the fibula after surgery. The distal fibula was placed into the posterior facet of the calcaneus. The foot was held in position with Kirschner wires and a plaster cast, which were removed at 8 weeks after surgery.

Fibular transfer technique

The authors performed fibular transfer for complete deficiency of the tibia, using the technique first described by Brown[2,3]. Anterolateral incision was performed to approach the knee, distal femur and fibula. Quadriceps, patella and distal femur were inspected. The common peroneal nerve was explored. The biceps tendon was released from fibula. The fibula was then contoured to match the shape of the distal femur. The knee was allowed to fully extended and fibular was centralized to the distal femur. Kirschner wire 2.0 mm was fixed with 2 or 3 Kirschner wires and a plaster cast. The plaster cast and Kirschner wires were removed at 8 weeks[13].

Leg-lengthening technique

The fibula was lengthened in one patient by callus distraction using ilizarov and two tibial lengthening in type IV patients. At each level two 1.8 mm K wires were mounted to the rings. Weight bearing was allowed at the first day after the operation. After 7 days, callus distraction was initiated at a rate of 1 mm/d (0.25 mm 4 times daily). The patients or family members performed the distraction themselves as outpatients. If poor callus response was observed, distraction was stopped for 1 to 2 weeks to allow better callus formation. The ilizarov was removed when bone consolidation was confirmed by radiographs.

Results

The families and the patients were satisfied in all cases. The duration from the first operation to the final follow-up ranged from 4 to 10 years. Two cases of type Ia, who were not ambulating before the operation, one case could walk using knee-ankle-foot orthosis because of the residual instability of the knee joints (a bilateral Ia type case 3). Another case of Ia type could walk independently. They could perform unrestricted household activities without pain. The ankles were stiff in all cases but no pain were reported. The four cases of type IV could walk independently and perform normal daily activities without pain.

Foot centralization

All 7 feet in the present study showed severe equinovarus deformity, and all were corrected by foot centralization with soft tissue release. The ankles were stiff in all cases but no pain was reported. The four cases of type IV could walk independently and perform normal daily activities without pain (Fig. 2A, 2B and Fig. 3A to 3F).

Fibular transfer

All knee joints in type Ia did not have strong quadriceps function, resulting in poor range of motion and progressive knee flexion contracture. In patients with poor extensor mechanism, progressive knee flexion contracture was carefully observed and treated as necessary by using the appropriate corrective orthosis. In case 1, whose quadriceps tendon and patella were absent, the knee finally ankylosed and our final treatment for this patient was a femoral-fibulocalcaneal arthrodesis (Fig. 1A to 1D). In a patient (case 3) who had bilateral type Ia, the authors performed bilateral fibular transfer but eventually resorted to knee orthosis because of persistent ligamentous instability. Based on the criteria established by Jayakumar and Eilert[3] (active range of motion, 10 to 80 degrees; varus or valgus instability not more than 5 degrees; no flexion contracture), none of the 3 knees in the present study achieved a satisfactory result.

Leg-lengthening technique

The details of the 3 callus distraction lengthening are shown in (Table 3). One fibula was

Fig. 1A Anteroposterior radiographs of a child with type Ia tibial deficiency of the right leg

Fig. 1C&D Radiographic and Clinical after 8 years follow-up

lengthened by callus distraction and another two were tibial lengthening. The total amount lengthened in each patient ranged from 4 cm to 12.7 cm. Bone consolidation was completed in all cases. Neither nonunion nor neurovascular complications were observed in any of the patients. The lengthening index ranged from1 to 4.2.

Complications

Pin tract infection occurred in all the cases with ilizarov lengthening and was treated by local or parenteral antibiotics.

Fig. 1B Brown procedure, foot centralization and lengthening of fibular to correct limb length discrepancy

Fig. 2A Anteroposterior and lateral views of tibial deficiency type 4 of Jones classification. Diastasis of the ankle mortise and clinically looks like clubfoot. Shortened tibia and the disruption of the normal relation between the tibia and the fibula

Discussion

Although the treatment of the type Ia of Jones is knee disarticulation in many literatures which has claimed that children who underwent early amputation were more active, had less pain and were more satisfied[2-5]. The authors tried to centralize the fibula under the femur by Brown procedure[7,8].

Most previous reports have described relatively poor results of fibular transfer[3,5,6,8,10-13]. Brown and Brown and Pohnert initially reported good early results in patients undergoing this technique but later showed poor results in 18 of 40 patients needing further surgical procedures[7,8].

Epps et al reported the results of twenty

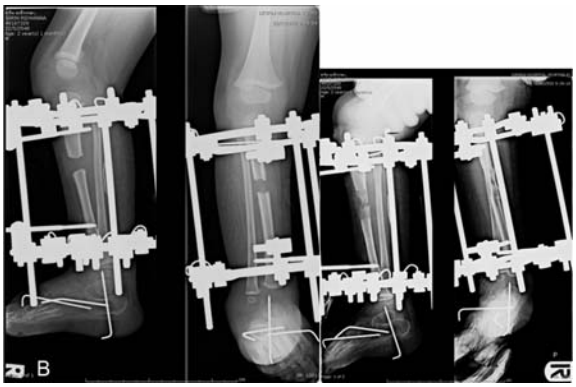

Fig. 2B Foot centralization and limb lengthening is performed to correct the deformity

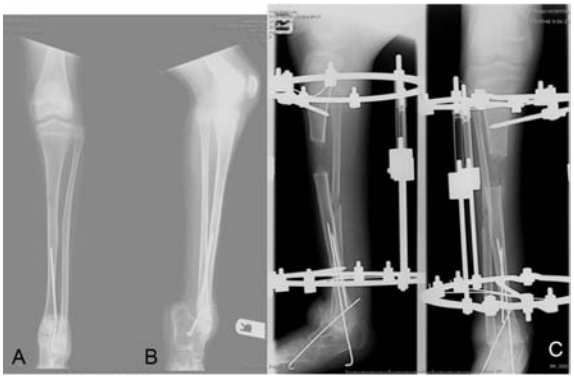

Fig. 3A-C (A-B) Anteroposterior and lateral views of tibial deficiency type 4 of Jones classification. Notice the shortened tibia and severe equinous of the ankle, (C) Foot centralization and limb lengthening is performed to correct the deformity

operations of fibular centralization. A progressive flexion deformity of the knee developed after all procedures. Twenty-six procedures were needed, including disarticulation at the knee, posterior release, extension osteotomy, femorofibular arthrodesis, a biceps to quadriceps transfer and one patient had a second attempt at centralization of the fibula[6].

Schoenecker et al reported secondary amputations after the fibular transfer in more than 50% of their cases. Most failures were due to marked knee instability and the progressive knee flexion contracture because of insufficient quadriceps strength[4].

Loder[13] examined 87 cases from the literature using the minimal requirements for a good result, as suggested by Jayakumar and Eilert[3]. He found that 53 of the 55 cases of Jones type Ia deficiency treated by Brown's procedure had a poor result because of flexion contracture. This echoed of most others and emphasizes the need for strong, active knee extension, which is usually not present without a remnant of the proximal tibia.

Simmons et al were satisfied with the results from their evaluation of Brown's procedure[15] and concluded that a fibular centralization procedure in patients with at least grade III quadriceps function could give good functional results that did not appear to deteriorate with time. Their satisfaction was based more on the patients' feelings than objective assessment.

In view of the poor results obtained by fibular transfer, primary knee disarticulation has been widely preferred.

Fig. 3D-F Results after foot centralization and limb lengthening in tibial deficiency type 4 after 4 years follow-up

In the present study, on 3 limbs with complete deficiency of the tibia (type Ia), the foot centralization, soft tissue release and fibular transfer were performed followed by repeated callus distraction lengthening. All of the 3 knee joints did not have strong quadriceps function, resulting in poor range of motion and progressive knee flexion contracture, which was consistent with the results of other previous reports[3,5,6,8,10-13]. In case 1, whose quadriceps tendon and patella were absent, the knee finally ankylosed. One of our patients (case 3, bilateral type Ia), who had poor quadriceps function, underwent fibular transfer bilateral, but the patient resorted to knee orthosis because of persistent ligamentous instability.

Of these, 2 cases were sufficiently able to withstand weight bearing, but none of the cases achieved a satisfactory functional results, as suggested by Jayakumar and Eilert[3].

After the feet and knees were centralized, one fibulae and two tibia were lengthened by callus distraction. Neither the amount lengthened nor the healing index differed significantly between the tibia and the centralized fibula at the first time of callus distraction but was markedly different in the second callus distraction, especially in fibula lengthening. In case I, the authors performed two events of fibular lengthening; at first lengthening, the lengthening index was 1.4 for total length 5 cm and at the second lengthening the lengthening index increased to 4.2 which was shown as the delay in distraction time in the second time of fibula distraction. Very few cases of distraction lengthening for the centralized fibula have previously been reported[9,10] and it remains unclear whether the centralized fibula can be effectively lengthened. Our results demonstrated that the centralized fibula gradually widened and lengthened and was sufficiently able to withstand weight. Healing Index was not different between the tibia and the centralized fibula in first distraction, but delayed the lengthening index of fibular lengthening in second lengthening. Some reports showed osteogenesis relatively slow during fibular lengthening in a case of tibial hemimelia which required gradual compression twice, as well as bone grafting[10]. The authors did not face this problem, as there was good regenerate formation. The progressive knee flexion deformity, may develop during tibial lengthening of the type IV and fibula lengthening in type Ia case and carries the risk of knee subluxation. On the contrary Javid et al in 2002 reported 15 cm fibular lengthening in a type II case without any knee flexion deformity[10]. However, our (case 1) type Ia case developed flexion contracture of 20 degrees which might be because of both fibula lengthening and poor quadriceps function.

In the present study, the authors had the opportunity to treat a patient with bilateral type Ia with Brown procedure and foot centralization bilaterally. Although she had poor knee function (assess by Jayakumar and Eilert criteria) due to ligamentous instability and progressive knee flexion contracture, she could withstand weight with orthosis.

The present study also has 4 patients with type 4 in the Jones classification. The foot in these cases deformed, often appearing like a clubfoot and also has tibial shortening. All of this made it difficult for the parents to accept amputation. Many reports suggested the true type 4 deficiency with diastasis of the ankle joint should be treated with amputation[4].

Schoenecker et al reported on ten patients with Jones type 4, of which nine had initial reconstruction of the foot[4]. A Syme amputation was subsequently done in six of them, usually at the parents' request. Of the four who retained the foot, two had contralateral deficiencies in which the prosthesis accommodated the length discrepancy. One had a lengthening of 4.6 cm and one remained 4.8 cm short. From the available information it would seem reasonable to attempt to retain the foot, if the deformity was at the less severe end of the spectrum, or if there was a significant contralateral deficiency.

Problems to obtained plantigrade foot and limb-length discrepancy are common in these patients. In this present study one patient followed-up to the age of 15 years (case 5) and was described as having stable ankle after foot centralization and posteromedial release, another followed-up to the age of 16 years (case 4,6 years after reconstructive surgery) and was reported as having a stable ankle and plantigrade foot.

To date, only a few authors have described the effectiveness of foot centralization by means of talofibular arthrodesis[1,4,11]. In the present study, the authors centralized the foot by placing the distal fibula into the posterior facet of the calcaneus. Although the operated fibular and calcaneus lacked mobility and finally ankylosed. Patients had stable ankles and can withstand weight bearing. This was a major functional improvement for it did not limit their household activities; as the patients were bed-ridden before the operation.

Treatment of tibial hemimelia with limb saving procedure is complicated. Reconstruction of these cases described here is difficult, lengthy and requires

experience. Amputation is an easy and effective solution. The authors could not compare it with our results due to the different parameters used in evaluation. The authors believe the method of treating tibial hemimelia described in this series might be appreciated in instances in which amputation is refused, as marked functional improvement can be expected.

Potential conflicts of interest

None.

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การรักษาภาวะกระดูกหน้าแข้งสร้างไม่สมบูรณ์โดยไม่ตัดรยางค์ขา

พีระจิตร เอี่ยมโสภณา, กมลพร แก้วพรสวรรค์

วัตถุประสงค์: *เพื่อศึกษาผลการรักษาภาวะกระดูกหน้าแข้งสร้างไม่สมบูรณ์ที่ได้รับการรักษาโดยวิธีการไม่ตัดรยางค์ขา ในด้านความพึงพอใจ, ผลการรักษาและภาวะแทรกซ้อนจากการรักษา*

วัสดุและวิธีการ: *ผู้ป่วยจำนวน 6 ราย ที่ได้รับการรักษาด้วยวิธีการไม่ตัดรยางค์ขา ที่ได้รับการรักษาในโรงพยาบาลศิริราชตั้งแต่ปี พ.ศ. 2536-2550 ในจำนวนนี้ประกอบด้วยชนิด Ia จำนวน 3 หน้าแข้ง และชนิด IV จำนวน 4 หน้าแข้ง, อายุที่ผู้ป่วยได้รับการผ่าตัด ตั้งแต่ 2 ปี ถึง 11 ปี ในผู้ป่วยชนิด Ia เป็นผู้ป่วยที่ไม่มีหน้าแข้งอย่างสมบูรณ์ร่วมกับการทำงานของกล้ามเนื้อในการเหยียดข้อเข่าทำงานได้ไม่ดี จะได้รับการผ่าตัดเพื่อย้ายกระดูกฟิบูลามาทำหน้าที่แทนกระดูกทิเบียและจัดกระดูกเท้าและข้อเท้าให้อยู่ในแนวตรง รวมทั้งทำการยืดขาเพื่อแก้ไขภาวะขาสั้นยาวไม่เท่ากัน ในผู้ป่วยชนิด IV ผู้ป่วยจะมีกระดูกหน้าแข้งแต่มักมีปัญหาข้อเท้าไม่มั่นคงและขาสั้นยาวไม่เท่ากัน จะได้รับการผ่าตัดเพื่อจัดกระดูกเท้าและข้อเท้าให้ตรงและยืดขาเพื่อแก้ไขภาวะขาสั้นยาวไม่เท่ากัน ทำการศึกษาและตรวจติดตามการใช้งานโดยติดตามตั้งแต่ 4-10 ปี*

ผลการศึกษา: *จากการศึกษาพบว่าผู้ป่วยจำนวนสองคนของชนิด Ia หนึ่งคนสามารถลงน้ำหนักได้โดยไม่ต้องใช้ไม้ค้ำยันอีกหนึ่งคนที่เป็นชนิด Ia สามารถเดินได้โดยใช้อุปกรณ์พยุงเท้าและไม้ค้ำยัน เนื่องจากผู้ป่วยรายนี้ เป็น ชนิด Ia ทั้งสองข้างและมีขาข้างหนึ่งที่ข้อเข่าไม่มั่นคง ในผู้ป่วยที่เป็นชนิด IV ทุกคนสามารถเดินได้โดยไม่ต้องมีอุปกรณ์ช่วยเดิน มีผู้ป่วยจำนวน 3 คน ได้รับการยืดขาหนึ่งคนเป็นการยืดขาในชนิด Ia ผ่านกระดูกฟิบูลาและอีกสองคนเป็นการยืดกระดูกทิเบียในชนิด IV เฉลี่ยความยาวของขาที่ยืดอยู่ที่ 5.1 เซนติเมตร และค่าเฉลี่ยจำนวนเดือนต่อการยืดขาได้ 1 เซนติเมตร (lengthening index) เท่ากับ 2.4.*

สรุป: *ความพอใจของผู้ป่วยและญาติพบว่าอยู่ในเกณฑ์ดี แม้ว่าการรักษาโดยวิธีการไม่ตัดขาจะต้องมีการผ่าตัดหลายครั้งและใช้เวลานานในการรักษามากกว่าวิธีการตัดขาและใส่ขาเทียม แต่วิธีการนี้เหมาะกับสังคมไทยที่นิยมถอดรองเท้าอยู่กับบ้านไม่นิยมใส่รองเท้าหรือขาเทียมเดินในบ้านและผู้ป่วยยังมีความรู้สึกของเท้าขณะเดิน*