Severe progressive deformities after limb lengthening in type-II fibular hemimelia

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Until recently the accepted treatment of choice for severe type-II fibular hemimelia has been Syme's or Boyd's amputation. The alternative of distraction lengthening using the Ilizarov technique is now available.

We report three patients (four limbs) with type-II fibular hemimelia who were treated by the Ilizarov technique and followed up for two to six years. Severe progressive procurvatum and valgus deformity of the tibia and valgus deformity and lateral subluxation of the ankle were found in all four limbs. Multiple additional soft-tissue and bony surgery was necessary. In view of these problems we feel that reappraisal of the indications for lengthening in type-II fibular hemimelia is necessary.

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Fibular hemimelia is the commonest congenital deficiency of the long bones. It is estimated to occur in about 7 to 20 per million live births. Most cases are sporadic, but are occasionally associated with complex syndromes. Clinically, it has a wide range of severity from minor shortening to total absence of the fibula and associated anomalies of the lower limb. In the more severe cases the femur is also shortened.¹⁻⁹ The commonly used classifications are those of Achterman and Kalamchi¹ and Coventry and Johnson.⁴ The former is based on the involvement of the fibula. In type IA, the entire fibula is present but is shortened and smaller and in type IB it is partially absent. In type II the fibula is either completely absent or there is only a distal vestigial remnant.

Treatment of the severe type with absence of more than one lateral ray of the foot, complete absence of the fibula,

and an anticipated leg-length discrepancy of over 7 to 8 cm remains controversial. Syme's or Boyd's amputation has been reported to give good long-term functional results if performed in early childhood.^{3,6,10-13} The introduction of the Ilizarov leg-lengthening technique has led to a reappraisal of the indications for amputation and the technique has been proposed for the severe group.¹³⁻¹⁸ There are few reports on lengthening with only short-term results and a high rate of complications.^{3,11,18,19} We now review our results in three patients (four limbs) with severe type-II Achterman-Kalamchi fibular hemimelia (Fig. 1).

Patients and Methods

We reviewed prospectively three patients (four limbs) with Achterman-Kalamchi type-II fibular hemimelia and a leg-length discrepancy of over 8 cm who had had Ilizarov lengthening. The mean follow-up was four years (2 to 6). The preoperative assessment had included standard standing full-length scanograms and segmental radiographs of the tibia and fibula. All patients had had previous soft-tissue surgery including excision of the fibular anlage, posterolateral soft-tissue release and lengthening of tendo Achillis before the age of one year (Table I). The age at lengthening ranged from five to eight years. All three patients had tarsal coalition and deficiency of the anterior cruciate ligament. The leg-length discrepancy (LLD) at the time of lengthening was 8 to 9 cm in all four limbs.

Technique of lengthening. We used the Ilizarov technique in all four limbs. The rings were assembled to allow correction of the anteromedial angulation first with a distraction hinge, and then transformed into a lengthening assembly. At both the proximal and distal ends three to four 1.5 mm Ilizarov wires were attached to the rings with purchase of six to eight cortices. Extra care was taken to avoid damage to the common peroneal nerve. The wires were tensioned to 110 N. In all cases the position of the foot and ankle was controlled by a foot ring connected to the lower tibial ring. Distraction were started four to five days after surgery. All patients had unifocal lengthening of the proximal tibia at an average rate of 0.25 mm four times a day. The knee was stabilised by an external tailor-made lockable hinged brace to prevent the development of a flexion contracture during lengthening. This allowed mobil-

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Fig. 1

Case 1. Left-sided type-II fibular hemimelia.

isation of the knee under supervision during the day. Vigorous physiotherapy with active and passive joint mobilisation and a weight-bearing programme was maintained throughout the period of treatment. At the time of radiological corticalisation the frame was removed and further protection of the leg continued by an external hinged weight-bearing KAFO brace for up to two years. All patients were followed up in a special clinic by the authors with regular radiographs and clinical assessment at monthly intervals in the first six months and then at two- to three-monthly intervals. Special attention was paid to the functional status of the limb, its alignment and the range of movement of the foot and ankle. The KAFO brace was regularly checked and good compliance noted in all three patients.

Results

We achieved lengthening of 8 to 9 cm in all four limbs. The mean lengthening index was 34 days and the mean time in the frame ten months (Table II).

Mild procurvatum of the proximal tibia was seen in all four limbs at the end of lengthening. The ankle was maintained in the preoperative position with mild equinovalgus of the foot. There was no change in the knee from the preoperative state.

Acute and early complications. In all limbs minor complications occurred such as pin-track infection which was controlled conservatively.

Major complications such as knee contracture and subluxation did not occur. The range of movement of the ankle was similar to that preoperatively with mild equinovalgus of the foot.

Late complications. Progressive valgus deformity and lateral subluxation of the ankle and foot with associated procurvatum and valgus deformity of the tibia were observed about one year after lengthening. At the time of the final assessment at two to six years, there were considerable deformities in all four limbs. One patient (case 1) emigrated and was seen only six years after the original lengthening when she presented with severe deformity (Fig. 2). She was virtually walking on the end of her tibia.

Table I. Details of the three patients (four limbs) with type-II fibular hemimelia

Case	Type	Age at operation (yr)	Foot condition	Knee condition	Previous surgical procedure
1	II	8	Tarsal coalition	ACL deficiency	Excision of anlage
2	II	8	Bilateral tarsal coalition Absent 5th rays	ACL deficiency	Excision of anlage
3	II	5	Tarsal coalition Absent 4th and 5th rays	ACL deficiency	Excision of anlage

Table II. Details of lengthening in the three patients

Case	Limbs	Leg-length discrepancy (cm)	Simultaneous angular correction	Lengthening achieved (cm)	Months in frame	Lengthening index (days/cm)
1	Unilateral	8	Yes	8	10	37.5
2	Bilateral	9	Yes	9	10	33.0
3	Unilateral	8	Yes	8	10	32.5

Fig. 2a

Fig. 2b

Figure 2. Case 1. AP (a) and lateral (b) views showing a valgus ankle and foot and anteromedial tibial bowing.

Further surgery involving soft-tissue release of the posterolateral structures of the ankle, corrective osteotomy at the supramalleolar region and repeated Ilizarov lengthening was performed. Similar soft-tissue procedures have been necessary in cases 2 and 3 because of uncontrollable and progressive deformity. Further bony procedures are likely to be required.

Discussion

Fibular hemimelia has a wide range of severity ranging from mild hypoplasia of the fibula with minimal functional disturbance to complete absence and associated deficiency of lateral rays of the foot and tarsal bones. The ultimate leg-length discrepancy may be more than 10 cm and up to 17 cm. In the past both Syme's and Boyd's amputation has been the recommended treatment for the severe form, preferably before walking age, to lessen the psychological trauma and to allow better adaptation to a walking prosthesis. Many medium- and long-term follow-up studies have shown consistently good functional results with the use of this treatment.^{4,6,10-13}

The early results of lengthening using Wagner's technique were disappointing and there was a high rate of complications. Choi et al³ showed that only 55% of patients had satisfactory results with Wagner lengthening compared with 88% with amputation. The introduction of the Ilizarov technique stimulated a reappraisal of treatment.¹³⁻¹⁸ Reports by Miller and Bell, Catagni et al and others¹³⁻¹⁸ have shown

Fig. 3a

Fig. 3b

Figure 3. Case 1. AP radiograph six years after lengthening. There is asymmetry of the distal tibial epiphysis.

Fig. 4a

Fig. 4b

Figure 4. Case 3. AP radiograph two years after lengthening showing an asymmetrical distal tibial epiphysis.

that significant lengthening with foot preservation is achievable by the Ilizarov method. There are, however, few reports of the long-term effects and complications. Gibbons and Bradish¹⁸ described ten limbs in eight patients which had satisfactory lengthening, but there were four major and nine intermediate complications. The major complications included knee subluxation, permanent stiffness and persistent valgus deformity. Sharma et al¹⁹ reported severe growth retardation of the lengthened tibia in seven skeletally immature patients followed up until skeletal maturity. The average growth rate of the tibia decreased from a mean of 82.5% before to 39% after operation.

In our series all four limbs achieved the desired lengthening with good bone formation in the distraction gap. The lengthening index compares favourably with that of other studies. Foot and knee subluxation was well controlled by careful external splintage and physiotherapy during the lengthening. Later, however, progressive valgus deformity of the foot and ankle together with recurrent anteromedial bowing of the lower tibia occurred. Additional procedures including soft-tissue and bony surgery were necessary to correct these recurrent cosmetic and functional problems. A possible explanation of these deformities is that the abnormal posterolateral soft tissues cannot grow and elongate at the same rate as the lengthened tibial segment. This gives rise to posterolateral tethering and subsequent progressive valgus deformity and lateral subluxation of the ankle. In addition, the lengthening may exert asymmetrical pressure on the abnormal distal tibial epiphysis resulting in asymmetrical epiphyseal growth. One patient (case 1) was found

to have the most severe deformity at a follow-up of six years when she was entering the pubertal growth spurt. The asymmetrical distal tibial epiphysis can be seen in cases 1 and 3 (Figs 3 and 4).

We have not experienced these complications after lengthening in patients with type-I fibular hemimelia. Our results, together with some of those reported, suggest that it is necessary to reappraise the indications for lengthening in severe type-II fibular hemimelia. There appears to be a place for lengthening in patients with the milder form of deficiency and with length discrepancy of a minor degree. For those with severe length discrepancy, associated with tarsal coalition and absence of lateral rays, the indication is less certain in view of the complications seen in our series. The possible complications and the necessity for subsequent multiple lengthening and corrective procedures should be clearly explained to the parents and other treatment options discussed.

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