Association of Telescopic Rod and Ilizarov Frame in Fibular Hemimelia Surgical Treatment

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Abstract: Fibular hemimelia is a rare congenital malformation, but one of the most common long-bone length deficiencies, without a clearly known cause. It can occur as an isolated anomaly or as part of a malformation syndrome. The presentation may vary from mild fibular hypoplasia to fibular absence. We reviewed the case of a 4 year old male who referred to our clinic at the age of one year, presenting with leg length discrepancy, bowed leg and with an absence of the peroneal malleolus, the fifth toe and equinovalgus deformity. After further investigations and presenting the family the treatment options, they ruled out amputation and surgical treatment was started with two circumferential periosteal releases followed by a telescopic rod implant and an Ilizarov frame fixator. The purpose of the present report is to highlight the good outcomes of combining the telescopic rod and Ilizarov frame fixator in complete fibular hemimelia.

Keywords: Fibular hemimelia, treatment, telescopic rod, Ilizarov frame.

1. INTRODUCTION

Fibular hemimelia has an estimated incidence between 5.7 to 20 cases per 1 milion births, being more frequent than tibial hemimelia, with a ratio of 4:1 [1, 2]. The syndrome is complex and involves abnormalities of the foot, ankle, tibia, knee and femur. The presentation may vary from mild fibular shortening, knee valgus and instability, shortening of the femur to partial or complete absence of the fibula with a short and bowed tibia, small foot, equinovalgus deformity and missing rays. Achterman and Kalmachi classified fibular hemimelia in two types. Type I is subdivided in IA, in which the proximal fibular epiphysis is distal to the proximal tibial growth plate, and the distal fibular epiphysis is proximal to the talar dome; type IB in which the proximal fibula is absent for 30-50% of its length and the distal part does not provide ankle support. In type II fibular hemimelia the fibula is completely absent [3, 4]. Surgical treatment has to be individualized taking into consideration the severity of the deformity and the other associated deformities. The most used method for tibial lengthening is progressive distraction with an external fixator [5]. The lengthening process can be divided in two phases, distraction, during which the bone is lengthened and consolidation, which represents the formation of new bone. As soon as the gap created during the lengthening phase is sufficiently healed, the fixator can be removed. The consolidation phase is different in children than adults, it usually takes two times longer than the distraction phase, unlike adults where the consolidation phase takes three to four times longer [6].

2. CASE REPORT

We present the case of a 4 year old male patient, with no family history of congenital anomalies who referred to our clinic at the age of 1 year.

Clinical examination revealed anomalies of the right lower limb, with a leg length discrepancy of 4cm, the right leg being shorter, anteroposterior bowed leg, absence of the peroneal malleolus, fifth toe absence and equinovalgus deformity.

After the clinical examination, X-rays were taken and a bowed right tibia was found, along with the absence of the fibula and the fifth foot ray (Figure **1**).

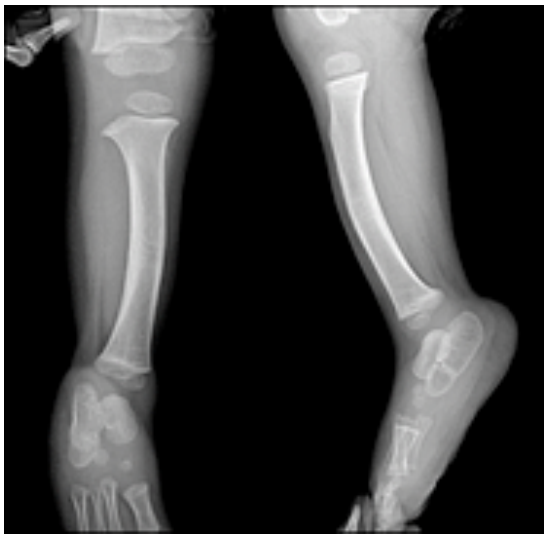

Figure 1: Fibula and fifth ray absence.

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Taking into consideration the clinical examination and X-rays the diagnosis of fibular hemimelia type II, according to Achterman and Kalamchi classification, was established [4].

After presenting the parents the treatment options and ruling out amputation, although early amputation with prostetic support offers a good long-term outcome [7], surgical treatment was started with 2 consecutive circumferential periosteal releases in order to strengthen the bone for future surgeries.

At 3 years of age, tibia osteotomies were made and fixed with a telescopic rod, which was advanced through the talus to provide ankle stability and compensate for the peroneal malleolus absence (Figure **2**), the follow-up x-rays at ten months showing complete bone healing, rod lengthening by 2.5cm and a straight leg axis (Figure **3**).

Figure 2: Pos-op follow-up.

Figure 3: 10 months follow-up.

At the age of 4 the overall limb length discrepancy was of 6cm and the decision of using an Ilizarov frame for leg lengthening was made, keeping the telescopic rod in place, maintaining the leg axis and the ankle stability (Figure **4**).

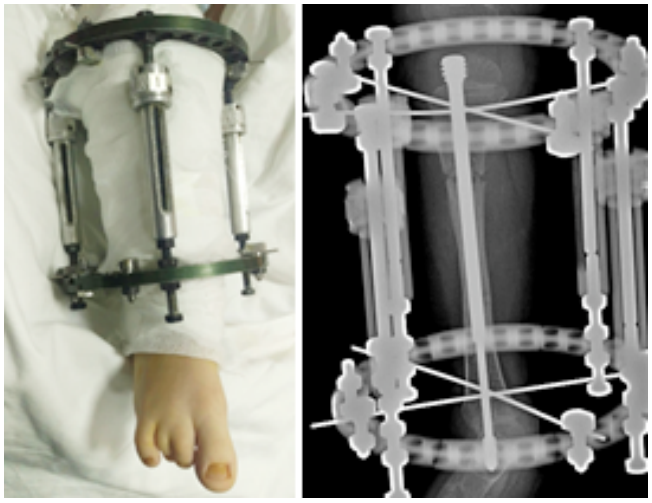

Figure 4: Clinical and X-ray post-op.

In surgery, the osteotomy tranche was opened to 0.5cm.

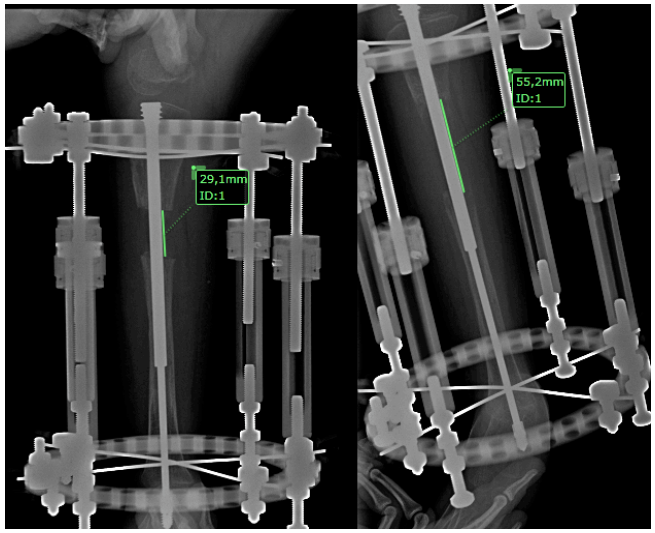

Figure 5: 25 days and 2 months post-op follow-up.

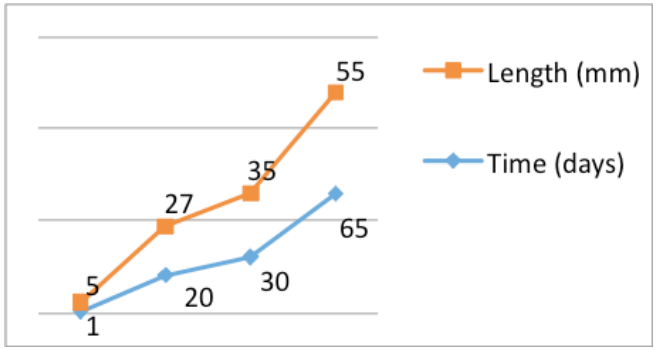

Figure 6: Lengthening progression.

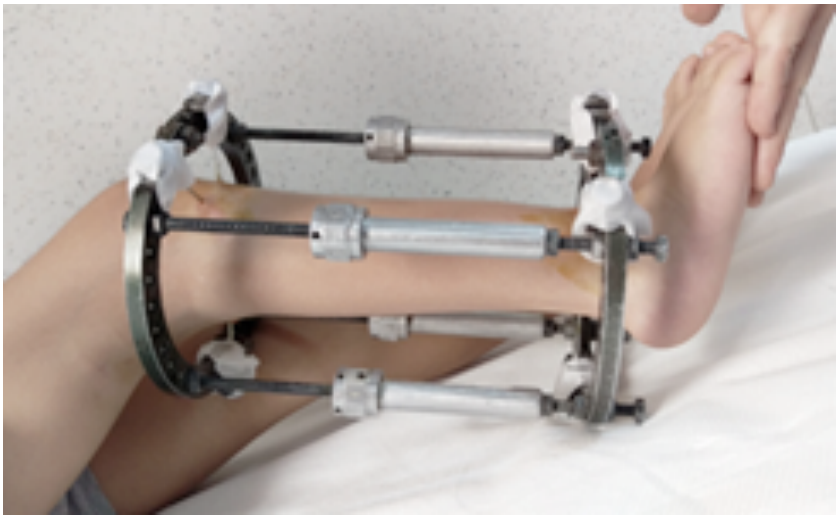

Figure 7: Side view comparison.

The lengthening procedure was started 3 days after the surgery at a pace of 1mm per day, in two turns of 0.5mm at 12 hours and continued for 65 days (Figure **5** and **6**).

After the lengthening procedure was stopped, the clinical residual discrepancy of the leg was of 0.5cm (Figure **7** and **8**).

Figure 8: Front view comparison.

The fixator was kept longer than usual and removed 7, 5 months after the surgery, due to a slower consolidation caused by the nature of the deformity and it's poor vascularization (Figure **9**).

Figure 9: X-ray after fixator removal.

3. DISCUSSION

Fibular hemimelia is a condition with a great physical, social and emotional impact on the child and the parents as well. Treatment is difficult as it has to address the associated deformities such as limb length discrepancy and the instability of the foot and ankle, deformities that will only worsen with growth. Treatment options are few and consist of nonsurgical or surgical (amputation, lengthening) treatment and the outcome is often poor.

Some of the complications that can arise when using the Ilizarov fixator are pain, infection, swelling of

the limb, neurovascular complications, contractures of the soft tissues and angular deformities [8, 9].

A method that was proven effective in reducing the complication rate and the period of external fixation is lengthening over an intramedullary nail (LON). The concept is not new and in early procedures Kuntscher nails were used [10].

The angular deformities and ankle instability, in this case, were adressed by using the telescopic rod, however, during the lengthening procedure swelling of the leg occured and was solved by stopping the procedure for 5 days.

After completing the procedure, the patient suffered a flexion knee contracture of 30 degrees, which will have to be resolved with future surgery and physio-therapy.

CONCLUSION

The decision of using a telescopic rod and an Ilizarov fixator for this patient with congenital fibular hemimelia was made in the prospect of obtaining a stable ankle and an adequate limb length after ruling out the amputation option.

Using the Ilizarov fixator and the telescopic rod can reduce the overall complications of the lenghthening process.

After performing the procedures and removing the fixator, good bone healing and a satisfying clinical outcome was obtained. The patient is currently undergoing physiotherapy for the knee deformity, however long term follow up along with other surgical interventions will still be necessary.

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