Ollier’s disease limb lengthening: Should intramedullary nailing be combined with circular external fixation?

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Accepted: 14 January 2010

Summary

Introduction: During progressive lower limb lengthening in the management of Ollier’s disease, the mean bone-healing index usually reported in the literature stands around 35 days/cm. One of the therapeutic objectives is to reduce the duration of the external fixation.

Hypothesis: The use of an elastic stable intramedullary nailing system (ESIN) combined with a circular external fixator significantly reduces the healing index.

Material and methods: Two groups of patients were compared. In group I, seven patients were operated on for progressive limb lengthening using a circular external fixator associated with an ESIN system: four monosegmental femoral lengthenings, one monosegmental tibial lengthening and two polysegmental femorotibial lengthenings. Nailing was performed via two intramedullary nails already used in traumatology. The date of external fixator removal coincided with that of radiographic healing. The nails were left in place. Group II included 37 patients who underwent limb lengthening by means of an external fixator only. The healing index was calculated and complications were analysed in both groups.

Results: The mean healing index (HI) values were: in group I: 23.3 days/cm for the femur, 22.4 days/cm for the tibia and 11.6 days/cm for polysegmental lengthenings; in group II: 31.6 days/cm for the femur, 35.7 days/cm for the tibia and 19.9 days/cm for polysegmental lengthenings. Group I demonstrated a statistically significant decrease in the HI for monosegmental femoral lengthenings.

Conclusion: A substantially reduced duration of external fixation, limited postoperative complications and prevention of later pathologic fractures are the reported advantages of the associated use of a circular external fixator with an ESIN system in the management of Ollier’s disease.

Level of evidence: Level III, comparative retrospective study.

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Introduction

Originally described in 1899, dyschondroplasia [1] or Ollier’s disease is a rare non hereditary skeletal disorder. This disease is responsible for various troubles linked to the development of multiple enchondromas secondary to growth disturbances. According to Maroteaux, surgery appears inefficient in the treatment of enchondromas [2]. This is why the related complications such as axial deviations and limb length discrepancies require a more specific management.

For the last few years, external fixation including monolateral fixators or circular fixators such as the Ilizarov fixator have become a popular treatment option in the correction of axial deformities and limb length discrepancies [3,4,5,6]. One of their drawbacks is the treatment period, which might sometimes reach 9.4 months [4]. In the treatment of Ollier’s disease, the healing index when using external fixation has often been reported to be over 35 days/cm [3,6,7,8], except in an interesting femoral case, which reported a 25 days/cm healing index [5]. One of the main objectives of this treatment is to significantly reduce the external fixation period. It contributes to the reduction of complications such as pin tract infections, external fixation loosening due to demineralization at the pin sites and joint stiffness.

Preliminary clinical studies have demonstrated the efficiency of elastic stable intramedullary nailing (ESIN) when associated with circular external fixation in limb lengthening. Flexible intramedullary nails provide a resistant and elastic system. The insertion of two nails, which do not completely fill the medullary canal also contributes to the development of an endosteal callus [9].

The aim of that study is to assess in lower limb lengthening and associated deformity-correction procedures, the efficiency of an ESIN combined with a circular external fixator such as the Ilizarov or Taylor Spatial Frame® (TSF), in patients suffering from Ollier’s disease. This retrospective study is compared to a series of patients managed with external fixation only and not combined with an ESIN.

Material and methods

In group I, we analysed seven progressive limb lengthenings performed using an ilizarov (3 cases) or a TSF® (four cases) fixator combined with an ESIN between 2004 and 2009. Four boys and three girls were included. The mean age of the patients was 13.4 years, range, 7 to 21 years. In two cases, the disease impact was general whereas it was unilateral in the five other cases. Monosegmental lengthening involved the femur in four cases and the tibia in one. Two polysegmental lengthenings femur and tibia consisted in a femoral osteotomy and two tibial osteotomies in a young man and a double femoral osteotomy combined with a tibial osteotomy in a girl.

Group II included 37 patients and involved 57 lengthenings using an ilizarov external fixator only without ESIN between 1972 and 2005. Twenty patients underwent two lengthenings. The mean age of these patients was 13.3 years, range, 4 to 24 years. The disease had spread in ten cases and was unilateral in 27 patients. The procedure was monosegmental 18 times for the femur and 11 times for the tibia. It was polysegmental in 28 cases for femur and tibia.

Operative technique

In both groups, the operative technique according to ilizarov always started with placement of the external fixator, followed by a percutaneous osteotomy, which aimed at preserving the periost and endomedullary vascularization, as much as possible [10,11].

In group I, the second phase consisted in the insertion of the ESIN: Through a 1–2 cm incision facing the metaphys.

Figure 1  Surgical technique [9]: two rings are placed, two precurved intramedullary nails are introduced after a percutaneous osteotomy was performed. Their maximum curve should be situated close to the osteotomy site on the diaphyseal side (a); insertion of external fixator additional wires (b); the apex of the nail curve is located at the bone regeneration site at the end of the lengthening phase (c); once the external fixator has been removed, the intramedullary nails are left in place (d).
Intramedullary nailing guide. The concave surface features a slot in which the nail slides during its insertion. The guide is supplied with a nail.

ysis, 2–3 cm from the physis, two cortical holes, one medial and the other lateral, were performed using a square point. This square point, of 3 to 4 cm diameter, was first introduced perpendicularly to the bone, then obliquely towards the diaphysis. For femoral lengthening, the selected ESIN was inserted in a retrograde fashion from the distal metaphysis. For tibial lengthening, the ESIN was inserted in an anterograde fashion from the proximal metaphysis. Both nails were carefully introduced up to the osteotomy site, then through this latter, and finally pushed toward the opposite metaphysis. Their curved part was orientated toward the diaphyseal side of the osteotomy to allow nail junction at a distance from the osteotomy site. Therefore, during the lengthening process, the curved section of the nail was progressively mobilized in order to face ideally the growth part of the bone regenerate at the end of the lengthening (Fig. 1).

The extraosseous end of each nail was curved against the metaphyseal cortex to achieve an angulation over $90^\circ$. Therefore, the nails could not slip outside the bone during the lengthening process but remained stable within the metaphyseal section. They were cut with a remaining 5 to 10 mm extraosseous portion then the skin was sutured.

Bifocal lengthenings were also secured by means of an ESIN (Fig. 3). In these cases, bipolar nailing was performed by combining an anterograde with a retrograde nail. During the lengthening phase, both nails were sliding one against the other like in a sliding nailing technique as described in the treatment and prevention of fractures in osteogenesis imperfecta [12]. After external fixator removal, each bone regenerate was secured at least by one intramedullary nail which curve was orientated in such a manner that it would resist the previous bone deformity tendency.

In group I, external fixation was completed after ESIN by adding wires and/or pins according to the external fixation requirements. Actually, it is easier to place the complementary external fixation once insertion of the ESIN has been performed rather than to introduce the ESIN nails between the external wires and pins.

In both groups, lengthening started about the 4–6th postoperative day when a percutaneous osteotomy had been performed and around the 10th postoperative day when open periosteal osteotomy was performed [13,14,15]. In all patients, potential associated deformities were progressively corrected during the lengthening process. When using the Ilizarov fixator, the hinges situated on the concave side of the deformity were placed on the axis of correction of angulation (ACA). When using the TSF®, deformity correction was simultaneously achieved with lengthening according to the instructions given in the correction programme.

Active and passive mobilization of the adjacent joint and partial then full weight-bearing were carried out in accordance with the lengthening procedure requirements.
In case of instability, joints were secured by performing a bridging technique and movements were made using the hinges.

The external fixator was removed as soon as the bone regenerate growth section had disappeared and when three or four continuous cortices could be seen on coronal and AP radiographs. The regenerate healing date was defined as corresponding to the date of the fixator removal while the intramedullary nails were left in place to protect the bone regenerate against any secondary deformity and/or pathologic fracture. The Healing Index was expressed in day/cm and calculated by dividing the number of external fixation days by the obtained lengthening in cm.

**Statistical methodology**

Both groups healing index was compared by using the Student *t*-test and the Wilcoxon Rank-Sum test for independent samples. These tests were performed to evaluate the Healing Index differences reported between the two independent groups of patients. The level of significance was set to *p* < 0.05. The descriptive statistical values describe the mean and standard deviation. The StatPlus® Professional 2008 software was used.

**Results**

In group I, the distraction bone regenerate always demonstrated a dense and clear aspect. From the third week, in six cases, the bone regenerate was well-structured and filled in the whole lengthening space between both bone fragments. The growth section was 2 to 6 mm and demonstrated numerous bone rows while at the same time, periosteal ossification was developing. The regenerate diameter was 2 to 6 mm greater than that of the bone. No cartilaginous islets could be seen or did persist within the regenerate. The mean rate of lengthening was 0.86 ± 0.13 mm/day for the femur and 0.80 ± 0.06 mm/day for the tibia.

In the six femoral cases, the mean lengthening was 5.78 cm (from 4.1 to 7.5 cm), that is 17.4% of the initial femoral length (10.7 to 25.0%) (Table 1). The mean Healing Index of the four femoral monosegmental lengthenings was 23.3 day/cm (19 to 28.2 day/cm) (Table 2).

In the three tibial cases, the mean lengthening was 5.83 cm (from 4.5 to 8.0 cm), that is 16.1% of the initial tibial length (10.5 to 24.2%). The Healing Index of the monosegmental lengthening was 22.4 day/cm.

In polysegmental femoral and tibial lengthenings, the mean HI was 11.6 day/cm (10.9 to 12.3 day/cm) (Fig. 3).

Full weight-bearing was initiated 4 to 6 weeks after external fixator removal and secured by a removable semicircular splint for a 2- to 4-week period. Knee range of motion recovery was obtained within 3 to 8 months after removal of the external fixator. In six patients, no nail-related complications were observed. In one patient, an intramedullary nail migrated 2 weeks after the TSF® fixator removal once the patient had initiated full weight-bearing after femoral lengthening. The nail was then removed. In all other cases, nails were left in situ. The follow-up period ranges from 2 months to 4 years. In group I, no complication such as delayed union, vascular impairment, non-union or fracture could be observed after fixator removal.

In group II, the mean lengthening rate was 0.94 ± 0.08 mm/day for the femur and 0.80 ± 0.11 mm/day for the tibia. The mean femoral lengthening was 6.4 ± 0.8 cm, that is 18.8 ± 2.1% more than the initial femoral length; in monosegmental lengthenings, the HI was 31.6 ± 5.3 day/cm. The mean tibial lengthening was 5.6 ± 1.4 cm, that is 20.1 ± 6.3% more than the initial tibial length; in monosegmental lengthenings, the HI was 35.7 ± 6.1 day/cm. In polysegmental femorotibial lengthenings, the HI was 19.9 ± 3.7 day/cm. Once the external fixator had been removed, immobilization was required for

**Table 1** Presentation of patients from group I.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age and gender</th>
<th>Method/segment</th>
<th>Lengthening</th>
<th>Lengthening (%)</th>
<th>Correction of axial deviation</th>
<th>Healing Index day/cm</th>
</tr>
</thead>
<tbody>
<tr>
<td>M.</td>
<td>11 Boy</td>
<td>Ilizarov + ESIN Tibia</td>
<td>5.0 cm</td>
<td>10.5</td>
<td>Valgus 12°</td>
<td>22.4</td>
</tr>
<tr>
<td>T.</td>
<td>15 Boy</td>
<td>Ilizarov + ESIN Femur</td>
<td>7.0 cm</td>
<td>23.3</td>
<td>Varus 16°</td>
<td>19.0</td>
</tr>
<tr>
<td>L.</td>
<td>14 Girl</td>
<td>TSF® + ESIN Femur</td>
<td>4.1 cm</td>
<td>10.7</td>
<td>Valgus 10°</td>
<td>25.6</td>
</tr>
<tr>
<td>Ro.</td>
<td>14 Boy</td>
<td>TSF® + ESIN Femur</td>
<td>5.1 cm</td>
<td>13.1</td>
<td>Varus 5°</td>
<td>28.2</td>
</tr>
<tr>
<td>W.</td>
<td>7 Girl</td>
<td>TSF® + ESIN Femur</td>
<td>6 cm</td>
<td>25</td>
<td>Varus 10°</td>
<td>20.5</td>
</tr>
<tr>
<td>Re.</td>
<td>12 Girl</td>
<td>TSF® + ESIN Femur</td>
<td>7.5</td>
<td>20.1</td>
<td>Varus 21°</td>
<td>12.3 (total)</td>
</tr>
<tr>
<td>B.</td>
<td>21 Male</td>
<td>Ilizarov + ESIN Tibia</td>
<td>5.0 cm</td>
<td>11.9</td>
<td>Varus 42°</td>
<td>10.9 (total)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ilizarov + ESIN Femur</td>
<td>8.0 cm</td>
<td>24.2</td>
<td>Recurvatum 8</td>
<td></td>
</tr>
</tbody>
</table>
Table 2  Healing index reported in both groups.

<table>
<thead>
<tr>
<th>Lengthening method segment</th>
<th>Group</th>
<th>Healing index day/cm</th>
</tr>
</thead>
<tbody>
<tr>
<td>Monosegmental/femur</td>
<td>I (EF + ESIN)</td>
<td>23.3 ± 4.3*</td>
</tr>
<tr>
<td></td>
<td>II (EF)</td>
<td>31.6 ± 5.3</td>
</tr>
<tr>
<td>Monosegmental/tibia</td>
<td>I (EF + ESIN)</td>
<td>22.4</td>
</tr>
<tr>
<td></td>
<td>II (EF)</td>
<td>35.7 ± 6.1</td>
</tr>
<tr>
<td>Polyssegmental/femur and tibia</td>
<td>I (EF + ESIN)</td>
<td>11.6 ± 0.99</td>
</tr>
<tr>
<td></td>
<td>II (EF)</td>
<td>19.9 ± 3.7</td>
</tr>
</tbody>
</table>

* The difference is significant: \(p = 0.0231\) (Student \( t \) test) and \(p = 0.0359\) (Wilcoxon Rank-Sum test).

Discussion

In the management of lower limb-length discrepancy in Ollier’s disease, such difference may be of more than 30 cm [16]. Such value is hardly predictable at the end of the lengthening phase since discrepancies and growth disorders tend to intensify unlike congenital disorders. Therefore, no forward-looking analysis can be performed.

In the light of various series published on limb lengthening in Ollier’s disease [4,8,17], it is well admitted that lengthening is not more complex than in other cases of limb discrepancy. Actually, growth disorder involves bone tissue only while soft tissues have a normal aspect: therefore, this explains the relative lengthening procedure easiness compared with that performed in the management of congenital aetiologies [4,17]. Enchondromal lesions do not induce more specific complications than normal bone, wire and pin fixation appears sufficient [4,8,17,18]. Bone regeneration is normal even if lengthening was performed at the enchondromal lesions [4]. This neo-ossification is independent from the enchondromatosis since it involves the periosteum and not the physis [7]. However, the improvement in the lengthening conditions is still under debate. A longer external fixation treatment period is often required in the femur with a reported HI between 35-day/cm and 45-day/cm [3,6,7]. Watanabe has reported differences between lengthening procedures when performed at the enchondromal lesions with a HI of 39.7-day/cm and lengthening performed in the healthy bone with a HI of 30.8-day/cm [8]. During four femorotibial homolateral simultaneously performed lengthenings procedures with the Ilizarov technique, Curran has reported a femoral HI of 28-day/cm and a tibial HI of 29-day/cm [17]. Stanitski et al. have reported a femoral lengthening of 9 cm for a 7.5-month treatment period [5].

The use of external fixation in bone lengthening has proved useful but also reports complications such as wire and pin tract infections, joint stiffness and patient discomfort. Rybka and Richter have demonstrated an increased incidence of pin-tract infections and hemorrhagic complications at the end of the fixation period [19]. Fractures occurring after removal of the external fixator have also been reported [4]. In our group II, among the 37 patients treated with external fixation only, three pathologic fractures at the enchondromas and three deformities at the lengthening site were observed after removal of the external fixator. On the other hand, in patients from group I treated with ESIN, no secondary fracture was noted during follow-up.

This is why we advocate the use of fixator-assisted ESIN in the management of limb lengthening [20,21]. However, this concept is different from the association of two rigid implants when combining monolateral fixator [22] with rigid intramedullary nailing [23,24].

Choice between the Ilizarov fixator and the TSF® is made according to the surgeon’s preferences and availability of the device. When correction is scheduled using the TSF® fixator, it facilitates limb lengthening and concomitant three-plane correction of associated deformities [13]. If some series report a higher HI with a mean value of 48.4-day/cm [15], other series do not find significant differences between the TSF® and the Ilizarov fixators [10].

According to our experience based on seven cases of limb lengthening performed on different bone segments, the combination of external fixation with ESIN in patients suffering from multiple enchondromatosis, demonstrates unquestionable advantages. When comparing our results obtained with and without associated ESIN, the HI is significantly reduced in group I that is in patients with fixator-associated ESIN.
The combination of both osteosynthesis devices decreases the maturation period of the bone regenerate and accelerates bone healing. As noted in an experimental surgical study conducted in dogs, intramedullary nailing induces a neo-ossification of endosteal callus type occurring along the nail length [25], which can be clearly observed induces a neo-ossification of endosteal callus type occurring along the nail length [25], which can be clearly observed. Moreover, gradual sliding of the intramedullary nails through the regenerated bone area during the lengthening process does stimulate new bone formation; however, these data should be confirmed by further investigations.

Curved intramedullary nails do not inhibit the endosteal regenerate formation. Furthermore, they provide additional mechanical stability at the lengthening site, thus preventing the risks of bone fragment translation relative to one another [9]. In order to improve this stability, the curved nails should be introduced so that their curved section is situated close to the osteotomy site, opposite their intramedullary entry hole. Therefore, the lengthening process helps the nail slide toward the metaphyseal region and displaces the curved section at the level of the regenerated bone area at the end of the lengthening process.

Strict respect of the operative technique will determine the success of the procedure as demonstrated in our group I. Percutaneous osteotomy associated with external fixation and ESIN will enhance periosteal preservation at the lengthening site and intramedullary bone vascularity. Biological lengthening rate, early joint mobilization and splint support after removal of the external fixator are part of the operative technique. These requirements should be respected to ensure the success of the ESIN and significant decrease of the HI [9,20].

Conclusion

The ESIN is a mini-invasive intramedullary osteosynthesis method initially described in the management of fractures in children. It demonstrates a major advantage when associated with the Ilizarov external fixator for limb lengthening in the treatment of Ollier’s disease: it consists in reducing the bone regenerate healing time (significant decrease in HI) which allows early removal of the external fixator. The external fixation treatment period is then reduced by about 8 days for each monosegmental lengthening centimetre.

Conflicts of interest

None.

References