Surgical Treatment of the Femur in Osteogenesis Imperfecta with Intramedullary Elongating Rods

Otto Wörsdörfer, Ulrich Vetter, Rolf Brenner

Unfallchirurgisch-Orthopädische Klinik, Klinikum Fulda, Germany

Osteogenesis imperfecta is a hereditary disease of the connective tissue based on a defective synthesis of the collagen type I. Frequent fractures, a tendency of formation of a pseudarthrosis, progressive deformation and growth retardation are the clinical symptoms regarding the skeletal system [7]. The clinical classification of Sillence and Rimoin [5] allows a more precise description and prognosis of the various types of the disease (Table 1).

<table>
<thead>
<tr>
<th>Typ</th>
<th>Genetic</th>
<th>Fractures</th>
<th>Deformities</th>
<th>Sclerae</th>
<th>Clinical</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>AD</td>
<td>+</td>
<td>+/--</td>
<td>Blue</td>
<td>Mild</td>
</tr>
<tr>
<td>II</td>
<td>AR/AD</td>
<td>+++</td>
<td>++++</td>
<td>Blue</td>
<td>Lethal</td>
</tr>
<tr>
<td>III</td>
<td>AR/AD</td>
<td>+</td>
<td>+++</td>
<td>White-light blue</td>
<td>Severe</td>
</tr>
<tr>
<td>IV</td>
<td>AD</td>
<td>+ Variable</td>
<td>+/-</td>
<td>White</td>
<td>Variable</td>
</tr>
</tbody>
</table>

Mutation

| Subtype A: Dentinogenesis imperfecta absent.  
| Subtype B: Dentinogenesis imperfecta present. |

Table 1. Sillence classification of osteogenesis imperfecta [5] (AD = autosomal dominant; AR = autosomal recessive).

Surgical Principles

Deformation of the femur or fractures of the femur with displacement are corrected with segmental osteotomies and are stabilized with elongating rods according to Bailey-Dubow [1] following reaming the medullary cavity. The rods, which elongate during growth, are anchored transarticularly through the intercondylar notch of the distal femur.

Advantages

After implantation of nonelongating intramedullary rods [6], deformities and fractures often occur during longitudinal growth below the implant [2–4]. Frequent changes of rods, on average every two years, with increasing complications, are the consequence.

The elongating intramedullary rod is exchanged every four to five years, depending upon the age of the child.

Disadvantages compared to a nonelongating rod do not exist [4].
Remain ing Problems

No rotational stability of the rod after segmental osteotomy. Frequent mal-union in external rotation.

In spite of the implanted rod, fractures due to rotational forces are possible.

Indications

Frequent fractures.

Non-unions.

Moderate to severe deformities with functional impairment of the limbs.

Contraindications

Poor general condition; cardiopulmonary disease.

Lack of possibilities of anchoring the rod due to disorganization of the bone structure (extensive epiphyseal cysts, hyperplastic callus).

Diaphyseal diameter less than 3.5 mm (children under one year).

Patient Information

Higher risk of infection caused by a reduction of the immune status (prophylaxis with antibiotics).

Higher anaesthetic risk (malignant hyperthermia).

Rotational mal-union due to initial rotational instability.

Proximal migration of the rod.

Non-union.

Shorting of the leg, lack of extension of the knee.

Growth arrest of the distal femur.

Necessity of rod exchange after a few years.

Pre-operative Work-up

Relevant radiographs of the femur including the knee in two planes with a radiopaque ruler in order to measure length and diameter of the rod.

Planning of the osteotomies and the size of the bone wedges.

Calculation of loss of length due to bone removal.

Radiological determination of the rod length.

Fig. 1 Bailey-Dubow elongating rods.

Pre-operative preparation of the rod: here one must consider, that the two parts should slide into each other without any friction.

Pre-operative antibiotic prophylaxis.

Surgical Instruments

- Bailey-Dubow elongating rods\(^1\) with various diameters (3.2 mm, 4 mm, 4.7 mm, 5.5 mm, 6.4 mm (Figure 1).
- Drill bits of various diameters between 3 mm and 6.5 mm.

For intraoperative rod shortening:

- High speed cutter with adapter for a diamond saw and grinding tools\(^2\) in order to fit the elongating rods peroperatively.
- Standard orthopaedic instruments.

Positioning and Anaesthesia

- Endotracheal anaesthesia.
- Radiolucent table.
- Stable lateral position (Figure 2).
- The chest and back are supported with towel rolls, fixation with adhesive tape.
- The hip and knee joint of the other leg are bent at 90°.

\(^1\) Zimmer Chirurgie GmbH, D-6074 Rödermark.

\(^2\) Synthes, D-7801 Freiburg-Umkirch, D-4630 Bochum 1.

A-5035 Salzburg; Stratec medical, CH-4437 Waldenburg.