Osteogenesis imperfecta

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Abstract. — Several factors have modified the surgical treatment of osteogenesis imperfecta (OI). The use of bisphosphonates means that rodding prior to walking age is not necessary. Surgery is performed at a younger age, as patients develop faster and the indications for rodding now include previously wheelchair-bound patients. More active patients are also more at risk of fractures, particularly of their upper extremities. However, the possible long-term effects of bisphosphonates on bones remain to be observed. The evolution of orthopaedic surgery towards minimally invasive techniques has led to percutaneous osteotomies and rodding, reducing soft tissue trauma (scarring, blood loss, pain) and allowing faster rehabilitation. The development of a new concept of telescopic rod for femoral rodding has now eliminated the need for knee arthroplasties. The multidisciplinary approach to this disease allows better care of the OI patient.

Keywords: osteogenesis imperfecta, bisphosphonate, telescopic rodding, percutaneous osteotomy, coxa vara.

Introduction

When discussing the surgical treatment of osteogenesis imperfecta (OI) in 2003, the necessity of a "team approach" to such a disease must first be emphasised. Surgery in itself is very limited without good metabolic preparation. The use of bisphosphonates (since 1992 in our experience) has modified the orthopaedic management of OI (Glorieux FH, Bishop NJ, Plotkin H, et al. Cyclic administration of pamidronate in children with severe osteogenesis imperfecta. N Engl J Med 1998; 339: 947-52). The following algorithm (Fig 1) summarises the different changes in the management of OI in the era of bisphosphonates. Finally, good co-ordination and communication between the orthopaedic surgeon, physiotherapist, occupational therapist and orthotist is the key to successful postoperative care. The ultimate goal of the treatment of OI children is to maximise function so as to provide better independence (mobility and self-care) at adult age, allowing social integration.

1. The effects of the use of bisphosphonates in the orthopaedic management of OI are illustrated for babies (1) and older children (2). The long-term effects (on bone remodelling) remain to be described (3).

TREATMENT OF FRACTURES

While most fractures in OI can be treated with (light) immobilisation, surgical treatment is sometimes indicated.

What not to do

Osteogenesis imperfecta bone is fragile; therefore, the fixation of screws in such bone is poor. Furthermore, a plate inserted...
on OI bone creates a stress riser, and fracture of the bone above or below the plate is most likely to occur (fig 2). For the same reasons, skeletal traction is not recommended, as the traction pin may cut through the bone and risk damaging the nearby growth plate. The use of skin traction is advocated for femoral fractures. The process of fracture healing in OI does not seem altered; therefore, there is no need for prolonged periods of immobilisation. On the contrary, a spica cast left in place for many weeks is accompanied by bone loss in the lower extremities, which in addition to natural osteopenia will be a cause of refracture when the cast is removed. Patients should be immobilised for short periods only, and then mobilised with protection.

What to do

Intramedullary rodding of the long bones is the treatment of choice for long bone fractures (see below).

Fracture-avulsion of the olecranon apophysis or the tubial tubercle tuberosity is particular in OI children. The olecranon fracture can be simply treated with a tension band (care should be taken not to tighten the wire too much, as it might cut the bone) [22]. Absorbable wires and sutures can be used. Fixation of the tubial tubercle is classically carried out with screws. If the availability of screws is limited, a temporary cerclage of the patellar tendon can be performed to protect the fixation, as in the treatment of patella alta in cerebral palsy patients [14]. The cerclage wire is removed at 6 weeks to allow normal contraction of the quadriceps.

General principles for long bone rodding in OI

INDICATIONS

As a baseline, mild types of OI with straight bones and a low fracture rate do not need intramedullary rodding. Patients with significant long bone deformities (> 20°) and repetitive fractures benefit from intramedullary (IM) rodding, as straightening of the extremities allows for a better function while the metallic “tutor” reduces the fracture rate.

With the use of bisphosphonates, the motor development of OI children is faster and they start standing at a younger age than in the past. As soon as the child tries to stand, if the legs are deformed there is a high risk of fracture, and this is a good time to correct deformities and to rod the legs.

Rodding prior to walking age in very fragile type III osteogenesis imperfecta patients (to reduce the fracture rate) is rarely indicated, as with the administration of bisphosphonates a rapid improvement of bone strength is possible. Rodding babies carries a high risk of complications and therefore should be avoided. Upper extremity rodding is performed at a much later age, when functional impairment has been identified.

The two main problems encountered (besides bone fragility) are the size of bones and growth. As regards the size of the bone, most “classic” IM rods are designed for adult bones and cannot be used in OI children (sometimes in adolescents). Therefore, small nails are preferably used (diameter from 2 to 6 mm). As regards growth, as the bone gets longer with growth the nail becomes relatively short and the bone deforms or breaks at the rod end(s). To prevent this problem, two thin rods can be inserted in opposite directions (Métazieux [15]; Sofield and Millar [23]) or a telescopic rod can be used (Dubow and Bailey [1, 2]; Fassier–Duval [8]).

PRE-OPERATIVE EVALUATION

Choice of type of rod: solid or telescopic

Solid rods are used in adolescents and patients with limited growth potential (“popcorn” physis): Rush rods, Williams rods [26], K-wires, or elastic rods. Telescopic rods give better results in femurs than in tibiae. The original Dubow–Bailey rod has been modified by several authors to reduce mechanical complications: the Sheffield rod [29] has a T-piece permanently fixed to the rod ends; Finidori modified the T-piece [28] to prevent its migration; Karpowski modified the male sleeve [30]; and Ozmur described a locking device [31]. The Fassier–Duval rod is another type of telescopic rod inserted only anterogradely, avoiding joint damage, and screwed at both ends of the bone [6].

Rod diameter

The shape of the OI bone (“scoliotic” bone, deformed in three dimensions) and the elliptical canal [15] makes this choice difficult. A larger rod will give more stability and protection against fractures, but causes bone loss (“disappearing bone disease”) around the rod. Therefore, thinner rods are recommended (acting as an internal tutor, and not “replacing” the bone). There is no precise rule on how to choose the rod diameter. Leaving 2 mm of bone on each side of the rod in any treatment plan is a safe measure, taking into account X-ray magnification. This choice is not a problem when using solid rods, as Rush rods or Kirschner (K)-wires are available in very small diameters. The smallest diameter for telescopic rods is 3.2 mm, which limits their use in very young children.

Rod length

Due to the three-dimensional deformity, an accurate assessment of the length of long bones in OI is difficult. Preoperative templating with the choice of osteotomy sites and the amount of bone to be resected has to be carried out, taking into account X-ray magnification and the fact that the greater trochanter is incompletely ossified. It also varies with the type of implant used. Finidori [26] recommends taking the distance between the two epiphyses and removing 1 or 2 cm.

Anaesthesia consultation

In most OI cases, it is better to refer the patient for a pre-operative anaesthesia consultation. There is a risk of fracture when manipulating OI children, intubation is often difficult, and these children have an elevated basal metabolism leading to intraoperative hyperthermia (non-malignant type). Bleeding tendency as well as neurological compression (basioccipital invagination) are other risk factors that have to be discussed with the parents before surgery.

PATIENT POSITIONING

A radiolucent table is used to allow image intensifier guidance during the surgical procedure. For lower extremity rodding, the patient is positioned over the edge of the table with a radiolucent bag (saline bag, for example) under the buttock (semi-lateral position). This position allows easier manipulation of the rod out of the buttock.
Osteogenesis imperfecta (retrograde or anterograde rodding). For upper extremity rodding, the arm is so short that it is often easier to leave the patient on the table with the arm parallel to the trunk rather than using a "hand table".

INTRA-OPERATIVE RECOMMENDATIONS
If a tourniquet is to be used, elevation of the limb is sufficient prior to inflating it. The use of Esmarch bands is dangerous (risk of fracture). Bone-holding clamps should not be used, as fragile bones might be crushed.

Soft tissues in the concavity of the long bone deformity are under tension after the bone has been straightened: at the time of reduction, one should be sure that the patient's muscles have been paralysed for easier manipulation. Too much tension leads to joint contracture or bone collapse; therefore, do not hesitate to shorten the bone to reduce soft tissue tension. Despite some bone shortening, the straightened leg is always longer postoperatively compared to preoperatively.

POSTOPERATIVE MANAGEMENT
In OI, the bone healing process is not slower compared to that of normal bone. Prolonged immobilisation is therefore detrimental to the bone as it produces osteoparenia. In our experience, most of the patients have only 3 weeks of postoperative immobilisation (femurs and tibiae) followed by bracing and progressive rehabilitation, as after 3 weeks rotation is stabilised. The spica cast is of particular concern, as it prevents the patient from sitting. Two long leg casts with a unit bar ("A" frame) are an alternative to prevent malrotation in OI children. This allows the sitting position (good for the spine and makes travel easier for the child). Light or ultralight postoperative immobilisation is preferred. Posterior back slabs are very useful. Fibreglass is also a material of choice, as it is both light and resistant. Unfortunately, it is more difficult to mould compared to plaster of Paris. Light thermoplastic braces are very useful for the treatment of OI patients. To avoid unnecessary delay, the braces can be moulded in the operating room, at the end of surgery, prior to applying the cast. They are delivered and fitted when the cast is removed after a few weeks. Patients who have never walked before surgery are prescribed a long-leg brace with "drop-lock knees" locked in extension for walking, and rigid AFOs (ankle-foot orthosis). As soon as the child shows good quadriceps strength, the knees are unlocked and the thigh part of the brace can be removed. The next step is to switch from rigid AFOs to hinged AFOs, allowing calf muscle development. Of high importance, the AFOs are bi-valved with an anterior panel to resist the progressive tibial bowing which is always anterior or anteromedial.

Femoral rodding

"SHISH KEBAB" TECHNIQUE
The original Sofield and Millar technique has been well described in many papers [10]. It has been widely used in the last 30 years, and its principles are still applicable. The trend of surgical techniques toward "minimally invasive surgery" [12, 30] and a better understanding of the physiology of bone healing have changed the approach toward rodding long bones in OI. Removing the shaft of the femur off the wound after proximal and distal osteotomies must no longer be done, as it suppresses all vascularisation to the bone. More "conservative" surgical options are now available.

BAILEY-DUBOW NAILING
The use of a telescopic rod has proven to be effective, and although the rate of complications is quite similar between solid rods and telescopic rods [23], the need for revision surgery due to growth is reduced with telescopic rods. Over the years, the surgical technique has been modified to reduce soft tissue trauma and bone devascularisation. This procedure can now be performed with "open" or "closed" performance osteotomies.

Open osteotomy technique
A posterolateral approach to the femur is made at the apex of the deformity(ies). The femur is exposed subperiosteally and the osteotomy carried out. The intramedullary canal of the proximal femur is prepared and increasing sizes of drill bits are used if the diameter of the canal is too small for the rod size. The proximal sleeve with its drill end [11] is passed retrograde through the medullary canal and the tip of the greater trochanter. A small incision in the buttock allows the rod to exit, and the drill-end is replaced with the "T" end.

The distal femur is prepared using the same technique and a second osteotomy is performed if necessary (a third is rarely indicated). Care must be taken not to elevate the periosteum circumferentially on the intercalary fragment(s) to avoid devascularisation of the bone segment(s). A third incision (parapatellar) exposes the intercondylar notch. A vertical incision is made in the non-weight-bearing zone of the distal femur and the male rod is introduced and pushed up to the osteotomy site. Reduction of the osteotomy and alignment of the two rods allows the smooth sliding of the obturator rod into the proximal sleeve (female rod). The female rod is pushed down so that the "T" part lies against the greater trochanter, under the gluteus muscles (to avoid possible migration of the rod in the soft tissues). Some authors advocate embedding the "T" part in the greater trochanter to avoid migration. The male "T" end is pushed under the articular cartilage into the epiphysis and turned 90° to prevent backing up in the joint. If several osteotomies are performed, it is easier to reduce the most proximal one(s) over a K-wire (male rod size) and push the female rod down to the most distal osteotomy site. Then the reduction of the last osteotomy is carried out with the obturator rod sliding into the sleeve as described above.

Percutaneous technique
A small incision is made over the greater trochanter and, under C-arm guidance, a K-wire (male size) is pushed to the apex of the deformity (fig 3A). A percutaneous osteotomy of the femur is made through the quadriceps muscle using a 3- to 5-mm wide osteotome (fig 3B). The osteotomy is performed in the convexity of the deformed bone, and manual osteoclasis allows straightening of the bone. The K-wire is pushed more distally (fig 3C). The sleeve rod is pushed, over the K-wire, into the bone. The obturator rod is introduced using the same technique as in the "open osteotomy technique" (fig 3D). The vertical part of the obturator in the sleeve is carried out under C-arm guidance; a second osteotomy may be required (fig 3E). It may seem hazardous, but the very small diameter of OI bones makes this step quite easy. The proximal and distal fixations of the rod is similar to the "open osteotomy technique" (fig 3F). This technique has also been used for Sheffield rod exchange [14].

ELASTIC RODS (METAIZEAUX TECHNIQUE)
Largely used in paediatric trauma for femoral, forearm, tibial and humeral fractures, this technique has also been described for OI patients [15]. Here, the rodding is bipolar, one rod is introduced from the knee (same approach as the Dubow-Bailey rod) up into the femur and the femoral neck, while the second rod is introduced from the greater trochanter down into the lateral condyle. The surgery can be performed through the skin under C-arm guidance or after osteotomy for axial correction. The advantage is that the sliding of the rods due to migration in opposite directions during growth maintains the "tutor" effect. Great care must be taken during surgery not to perforate the thin and fragile cortices of the OI bone.

FASSIER-DUVAL TELESCOPIC IM SYSTEM
This concept was developed from the observation of many mechanical complications with the Dubow-Bailey rods, and the evolution of surgery toward limited exposure [6]. The principles are as follows: the telescopic rod is introduced from the greater trochanter down (eliminating the need for a knee arthroscopy), and the two components are screwed at both bone ends (fig 4). Its long-term effects on the growth of the greater trochanter remain to be observed. As in Dubow-Bailey rodding, two different techniques are used, as described below.

Open osteotomy technique
The first step is the same as for the Dubow-Bailey technique: subperiosteal exposure of the femur through a posterolateral incision. Reaming of the proximal femur is carried out up to 3.5 mm (for a 3.2-mm rod) or 4.5 mm (for a 4-mm rod) (fig 4B) using cannulated drill bits. A K-wire, male size, is passed retrogradely into
the proximal femur, and exits in the buttock through a small incision. The male driver is pushed down over the K-wire to the osteotomy site (fig 4C).

If the distal femur does not require a second osteotomy, it is reamed as well, and the K-wire is removed from the male driver and replaced with the male nail (locked into the sleeve of the driver) (fig 5).

After reduction of the osteotomy, the male nail is driven distally. The position of the distal fixation has to be carefully assessed: the rod must be as seen on both the anteroposterior (AP) and lateral views (fig 6). It is better to perform a second osteotomy than to accept a suboptimal position of fixation. The threads must be totally within the epiphysis, not left across the physis (risk of growth disturbance). The male driver is removed, using a K-wire for counter-pressure on the nail while pulling on the driver (fig 7A). The female rod is pushed over the male (fig 7B), and screwed into the greater trochanter (fig 7C). The threads must be in the bone, not in the non-ossified part of the greater trochanter. C-arm control of the position of the two rods is made before the excess male rod that remains is cut just above the greater trochanter (5 to 10 mm). Because the tip of the male slides progressively into the sleeve during growth, it is recommended to check the smoothness of the male nail after it has been cut (with the special template tool) to ensure that sliding will occur (fig 7D).

**Percutaneous osteotomy technique**

A K-wire is inserted from the greater trochanter down to the apex of the deformity (fig 8A). Over the K-wire, progressive reaming is carried out according to rod size. The cannulated drill bit and the K-wire are replaced with the male nail in the driver, which is pushed to the apex of the deformity. A percutaneous osteotomy is performed (fig 8B) through the quadriceps muscle and by progressive osteolysis, the bone is straightened out. The nail is pushed through the osteotomy into the distal femur (fig 8C) and screwed in the distal femoral epiphysis as in the open osteotomy technique (be sure to check the position of the distal fixation very carefully). The driver is removed while ensuring not to back up the male nail. The female nail is pushed onto the male nail and screwed into the greater trochanter. The male nail is cut (care should be taken with the cut end, see "open technique"). If a second osteotomy is necessary, the same technique applies (fig 8D). The final aspect is shown in figure 8E.

The limitation of this technique is the size of the medullary canal. If the diaphysis distal to the osteotomy is too thin and has to be reamed up to 3.5 mm, it is safer to use the "open technique". It is also possible, but more difficult, to perform a percutaneous osteotomy, push the K-wire beyond the osteotomy site, and ream the distal femur. Unfortunately, due to the size of the implant, it is necessary to remove the K-wire and the reamer in order to insert the nail (Unlike adult femoral nails, there is no "exchange" device maintaining the reduction of the osteotomy). Therefore, while the surgeon removes the K-wire from the osteotomised femur and replaces it with the male nail on the driver, an assistant maintains the femur in
longitudinal traction to avoid displacement of the osteotomy(ies). When the two components of the rod have been introduced, C-arm control of the distal fixation (AP and lateral) is carried out. The AP assessment of the proximal fixation might be difficult due to a hip flexion contracture secondary to the correction of the anterior bow of the femur. This often requires tilting the C-arm so that the X-ray beam is perpendicular to the proximal femur. Beware of misleading images: hip flexion contracture progressively resolves in the first few postoperative months with physiotherapy.

**Tibial rodding**

**CLASSIC ANTEROGRADE NAILING WITH A SOLID ROD**

The apex of the tibial bow (usually anteromedial) is located under C-arm guidance and the bone exposed by subperiosteal dissection through a direct anterior incision. The osteotomy (closing wedge) is performed with an oscillating saw (care should be taken not to burn bone ends) or with an osteotome (with or without predrilling). Following the tibial osteotomy, a fibular osteotomy is necessary in order to straighten the leg. Simple osteolysis of the fibula is sufficient in young children (always carried out in valgus to prevent stretching the common peroneal nerve), and avoids unnecessary scars, bleeding, and prolonged surgery. The size of the canal is assessed and the diameter of the rod chosen. If necessary, reaming with progressive sizes of drill bits can be carried out. When a second osteotomy is needed (rarely a third), the distal fragment is reamed to the level of the second osteotomy before the osteotomy is performed (reaming a free bone fragment will make it “spring”, and all periosteal attachments will be stripped; the intermediate bone fragment becomes a bone graft with devascularised bone and risk of delayed healing).

Then two options are possible: a second osteotomy is carried out following the same principles, and the distal tibia is reamed to the size of the nail. The nail is introduced proximally through the prespatial area of the tibia (as in adult tibial nailing). The nail is pushed down through the proximal tibial growth plate. Multiple attempts at rodding must be avoided, as growth disturbance may occur. Leaving a smooth intramedullary rod through the growth plate and perpendicular to it has been proven not to prevent growth (there are no reports of such a complication in the literature). During this manoeuvre, the knee is flexed and the nail must remain anterior in the tibia. Perforation of the posterior cortex is frequent, because the latter is extremely thin. To avoid this problem, the rod can be bent anteriorly a few degrees in its last 3-5 cm (Rush rods are pre-bent by the manufacturer). The bent rod passes more easily in the proximal third of the tibia. When the rod exits at the osteotomy site, the bones are aligned and the nail is pushed distally. The same thing occurs if a second osteotomy is performed.

The other method for tibia rodding when a second osteotomy is necessary consists of reaming the fragment down to the second osteotomy level and stabilising the first osteotomy site with the rod prior to carrying
out the second osteotomy. This technique has the advantage of better preserving the soft tissue attachments on the intermediate tibia segment. It is also technically easier, as at any time during the operation the surgeon has only two bone segments to deal with. The choice of rod type is left to the surgeon’s preference: Rush rods have a smooth “hook-end” that is easy to embed in the proximal tibia epiphysis, but the length has to be predetermined. K-wires can be cut intraoperatively to the appropriate length, but the proximal end needs to be bent and sometimes remains prominent, causing mechanical irritation. When the rod is inserted, the mobility of the ankle joint must be checked: correction of severe tibial bow often leads to equinus of the ankle (posterior structures in the concavity of the deformity are too short). If this is the case, percutaneous Achilles tendon lengthening can be carried out, but it is better to short the tibia by removing a slice of bone at one osteotomy site. This second option does not weaken the calf muscles. At the completion of surgery, the periosteum is tightly sutured to reduce rotation at the osteotomy site(s).

**DUBOW-BAILEY RODDING**

This is a bipolar rodding technique, requiring a proximal tibial approach (the same as for classic anterograde rodding). The distal approach to the ankle joint can be medial (through the deltoid ligament and the capsule) as originally described, or lateral [10], cutting all three components of the lateral collateral ligament. The talus is subluxated to access the middle of the tibial plafond which is perforated to introduce the obturator (or the sleeve for the Finidori method), the end of which is pushed inside the distal tibial epiphysis after completion of the rodding. Variants of this technique include medial malleolar osteotomy in older children [20] and rodding through the medial malleolus [10]. In the latter technique, pre-bending of the obturator rod is necessary to avoid valgus deformity of the ankle. Telescopic rodding of the tibia has a higher complication rate compared to femoral rodding. The surgical trauma to the ankle ligaments and the permanent damage to most of the weight-bearing surface of the ankle joint limits its indications.

It is very simple and less harmful to anatomical structures to change an outgrown solid tibial rod whenever necessary. The “ultimate” telescopic tibial nail which does not require ankle arthroscopy remains to be designed.
ELASTIC RODDING (MÉTAIZEAU)

This technique involves bifocal rodding: one rod is introduced through the medial malleolus and driven up into the medial tibial plateau. The lateral (proximal) rod is inserted from the prespiral surface of the tibia and fixed distally in the lateral portion of the tibial epiphysis.

Results of lower extremity rodding

The complication rate with telescopic (Dubow–Bailey) rods has been reported in the literature to be between 39% and 72%, and the reoperation rate below 40% (most mechanical complications with the telescopic rod, such as loosening of the "T" part, do not always need surgery). These results can be compared with the 50% complication rate and 50% reoperation rate for non-telescopic rods, which need to be replaced when outgrown. Nevertheless, many authors have tried to improve the design of the telescopic rods to reduce the mechanical complications. The Fassier-Duval rod has now been used for 2 years with a complication rate of 15% and a reoperation rate of 7%.

Coxa vara

Varus deformity of the proximal femur is common in OI. True coxa vara must be differentiated from "false" coxa vara, which is due to a misleading radiological projection of a proximal femur that is deformed in three dimensions. In the "false" coxa vara, the neck-shaft angle of the proximal femur is normal, as it appears after regular rodding with c-arm guidance. The true coxa vara neck-shaft angle is below 110°, which is a self-aggravating condition, and its treatment in OI is difficult: the use of hip plates (even of paediatric size) is not recommended (poor screw fixation, risk of fracture below the plate) in very young children. Furthermore, IM rods with femoral neck support do not exist in paediatric size; the mechanical resistance of the rod would be jeopardised. In 1978, Wagner described a technique of osteotomy fixation using K-wires. Widman reported a series of coxa vara corrections with this technique, but not in OI patients. When the coxa vara is severe in OI, Findorff recommends performing an osteotomy distal to the lesser trochanter, with fixation by a rod introduced through the lateral cortex of the proximal femoral metaphysis and exiting at the base of the femoral neck.

At the Shriners Hospital for Children in Montreal, we use an original combination of these two techniques as follows. The approach to the proximal femur is posterolateral, with the patient in a semi-lateral position. The greater trochanter is exposed and two smooth K-wires of appropriate size are inserted from the greater trochanter into the femoral head. If there is already a nail in the femur, one K-wire is placed anterior to it; the other is posterior, both converging in the epiphysis. If there is no rod in the femur, sufficient space between the K-wires should be kept to allow the passage of the nail. A transverse osteotomy is carried out below the lesser trochanter (after the IM rod has been removed if necessary) and the K-wires are used as a "joystick" to bring the proximal femur into valgus as much as necessary. Using a drill bit the size of the nail, the lateral cortex of the proximal femur is perforated and the drill bit pushed in the direction of the base of the femoral neck. The angle between the drill bit and the K-wires indicates the amount of correction. The nail is introduced (retrograde for a classic Dubow–Bailey, or anterograde for a Rush rod or Fassier-Duval rodding). The medial proximal part of the distal fragment can be cut (wedge) to allow better contact between the diaphysis and the proximal metaphysis. This cut also prevents rotation of the diaphysis. The nail is pushed into the distal femoral segment. The K-wires are bent to become parallel to the diaphysis, cut to an appropriate length and secured to the diaphysis with two c-cure wires.

Upper extremity rodding

Upper limb deformities in OI patients have been relatively rarely studied. Only five papers have mentioned surgery for upper extremity deformities in OI since the article by Root in 1981. It was believed that considerable deformity had to be present in the upper limbs before surgery could be indicated. Our experience has proven that self-care and mobility are often limited in patients with type III osteogenesis imperfecta.

HUMERAL RODDING

Most of the growth of the humerus comes from the proximal humeral growth plate; therefore, solid rods (Rush, K-wires) introduced anterograde provide a very timely limited protection of the bone, which bows
below the rod. Telescopic rodding with the Dubow–Bailey rod has a high complication rate and is not widely used. The two techniques described below offer better protection for the humerus during growth.

### Elastic rods (Métaizeau)

This technique involves bifocal rodding: the proximal rod is inserted from the lesser trochanter down, through a limited lateral transadductor approach, splitting the muscle fibres along their axis. The rod has to be precontoured in an "S" form, so that the distal fixation point is in the medial column of the supracondylar area. The distal rod is introduced through the lateral condyle (direct percutaneous approach) and pushed up into the humeral head. As most of humeral deformities affect the distal third of the bone, the approach for any corrective osteotomy is anterolateral between the brachial anterior and brachio-radialis muscles. Careful dissection and protection of the radial nerve is carried out prior to the osteotomy. Reoperation through scar tissue carries a potential risk of injury to the radial nerve. It is recommended to use magnifying glasses. A posterior approach through the triceps (with or without disinsertion of the triceps) is an alternative. The weight of the upper extremity creates distraction at the osteotomy site during the postoperative period when the patient is sitting or standing. It is wise to add a cerclage wire to maintain the contact between bone ends. This cerclage also limits rotation at the osteotomy site.

### Fassier–Duval rodding

Anterograde nailing is performed through the deltoid muscle and the rotator cuff, just anterior to the acromion. As for the femur, the rodding can be done after an open osteotomy (distal third) or a percutaneous osteotomy (only in the proximal two-thirds). The male is screwed distally into the lateral condyle; the female is embedded in the head of the humerus. The excess of the obturator (male) nail is cut flush to the hollow part (female) to prevent interference with the rotator cuff muscles. A smooth range of motion of the shoulder must be obtained before closure of the wound. On X-rays, the proximal end of the female nail may appear outside the ossified portion of the humeral head while inside the head (the cartilaginous, non-osseified part).

### Postoperative management

Thin and cylindrical rods do not prevent rotation; therefore, complete immobilisation of the shoulder is mandatory for 3–4 weeks post-surgery. Immobilisation can be carried out with a brachioradial back slab of plaster of Paris or fiberglass, incorporated with a Vietnam sling, a Stevenson or a Dujarrier type of immobilizer.

### FOREARM RODDING

The size of the forearm bones does not allow the use of large rods. Even the smallest elastic rod may be too big for the bone. K-wires are the material of choice for IM rodding. The radius is approached distally through a radial approach. The superficial branch of the radial nerve is identified and protected. The rod is introduced through the styloid process under C-arm guidance, and progressively pushed up into the radial head. Osteotomies are usually performed through a Henry approach. The K-wire is bent and cut, and its top is embedded in the radial styloid to prevent protrusion through the tendons. The ulna is rodded from the olecranon down. In osteotomies performed by a direct ulnar approach, here also the tip of the K-wire has to be embedded in the olecranon to avoid bursitis and pain caused by a protruberant rod.

### Results

Functional outcome studies on upper limb surgery in OI are rare.[1] Nevertheless, with the use of bisphosphonates, more children with OI (particularly Type III) want to stand and walk; and as a consequence of the increased use of their upper limbs, the incidence of fracture is higher. These patients cannot spend many weeks not using an arm, and should benefit from rodding surgery. More outcome studies are needed to assess the long-term functional changes that can be achieved with surgery.

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### References


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