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Osteogenesis imperfecta: surgical treatment options with emphasis on today's orthopedic approach

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The treatment of osteogenesis imperfecta should be multidisciplinary and personalized. The best option for surgical treatment is implantation of an intramedullary telescopic nail, in many specialized hospitals the preferable method is Fassier-Duval telescopic nail. The main advantages of the Fassier-Duval intramedullary nail are fewer surgical scars, reduced blood loss, decreased time of operation, less pain, and better postoperative mobility. Orthopedic surgeons who are dealing with osteogenesis imperfecta have to answer two main questions, i.e. when to start with surgical procedures and which type of intramedullary nailing to use. There are a few things that are necessary for a satisfactory outcome of the surgical procedure: precise preoperative planning, all sizes of Fassier-Duval nail available in the operating room during the surgery, and intraoperative radiologic C-arm guidance.

Key words: osteogenesis imperfecta, Fassier-Duval, personalized and multidisciplinary approach

INTRODUCTION

Comprehensive treatment of osteogenesis imperfecta (OI) should be multidisciplinary and personalized. Multidisciplinary approach is necessary because multiple organs and regions are involved in patients with OI, and OI affects not only physical but also social and emotional development of the child. The medical team can include orthopedic surgeon, pediatrician, physical medicine and rehabilitation specialist, neurologist, radiologist, psychologist and social workers. There are many different types and subtypes of OI; it is impossible to have a universal approach and algorithm of treatment, which will be sufficient and successful for all patients. Therefore, it is very important to consider each patient separately and try to find the best treatment option for each patient individually. The leading signs of OI in a typical patient are multiple fractures of long bones and consecutive deformities and limitation of ambulation. The mainstay of modern orthopedic treatment is fixation of fracture and deformity correction to restore anatomical axis of lower extremities with intramedullary fixation. The purpose of this review is to present our hospital experience in the treatment of lower extremity long bones in patients with the most severe types of OI (types III and IV).

SURGICAL TREATMENT

The indications for surgical intervention are multiple fractures and severe angular deformities in children who are attempting to stand. Stabilization of the fracture and realignment of the long bone do not only decrease pain and reduce the risk of the fracture but also improve children's development, function and ability to stand and walk (1).

During history, there were many surgical methods aiming to fix and stabilize a fracture or to correct a bone deformity. In 1959, *Sofield and Millar* presented a new technique for surgical treatment of deformities and fractures in patients with OI (2). Their technique involved multiple osteotomies, realignment and internal fixation with solid intramedullary rod. In 1963, *Dubow and Bailey* proposed a principle of telescopic rod: each extremity of the rod is fixed in the proximal and distal epiphysis of the long bone (3). During growth,

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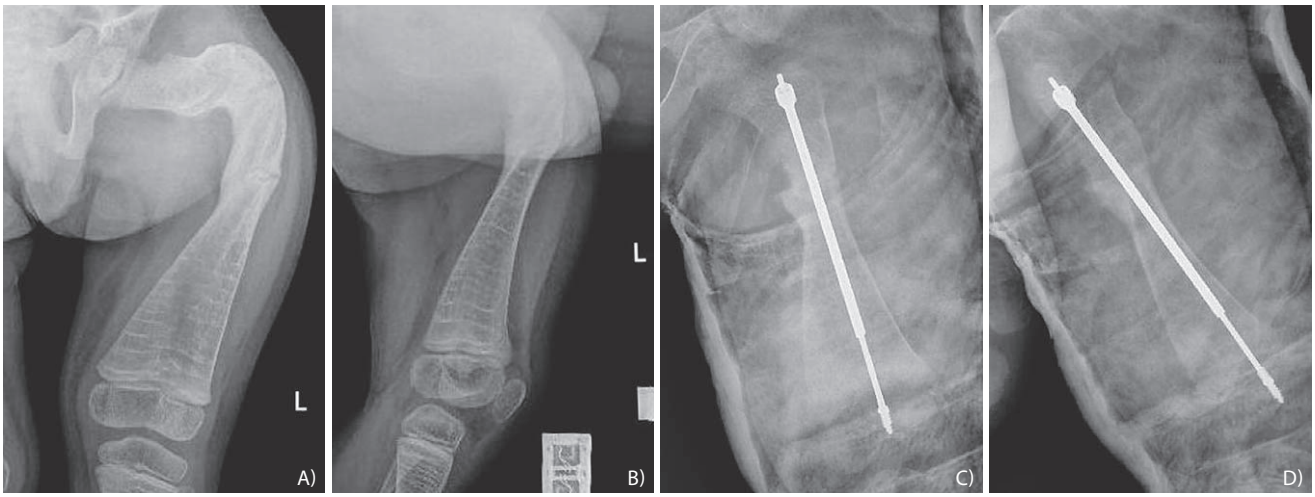

FIGURE 1. Preoperative radiograph of the left femur, anteroposterior view (A); preoperative radiograph of the left femur, lateral view (B); postoperative radiograph of the left femur, anteroposterior view (C); postoperative radiograph of the left femur, lateral view (D).

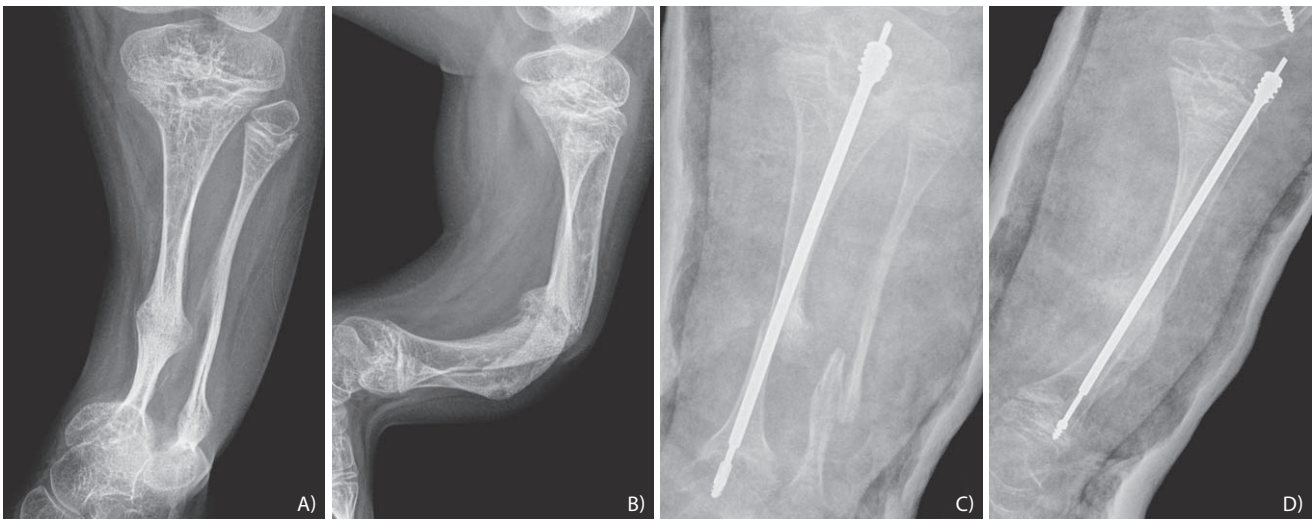

FIGURE 2. Preoperative radiograph of the left tibia, anteroposterior view (A); preoperative radiograph of the left tibia, lateral view (B); postoperative radiograph of the left tibia, anteroposterior view (C); postoperative radiograph of the left tibia, lateral view (D).

the 'male' rod is pulled out of the 'female' rod, which elongates like a radio antenna. As an option between solid and telescopic nail, sliding rods have been proposed (also called dual Rush rodding). The surgery consists of inserting two rods, one from the top down and one from the bottom up. During growth, the rods slide away from each other. In 1986, Sheffield rod was presented as a modification of Bailey-Dubow rod (4, 5). Sheffield rod was manufactured from stainless steel and its T-shape ends were permanently fixed to the body of the rod (in Bailey-Dubow rod, T-shape ends were free from the rod and before implantation they should have been screwed). This entire construct has a disadvantage of opening the knee or ankle joint on the distal side of the telescoping nail.

Today, there is a strong consensus that the best option for surgical treatment is implantation of an intramedullary telescopic nail. The Fassier-Duval (FD) telescopic nail is a preferable method in many specialized hospitals dealing with OI. The advantages of the FD intramedullary nail are fewer surgical scars, reduced blood loss, decreased time of operation, less pain, and better postoperative mobility (6). There is no need for knee arthrotomy in femoral surgery, or for ankle arthrotomy in tibia surgery because rodding is done through the greater trochanter or through the cortex of the proximal tibia (7). The major advantage of this technique is that multiple bones can be treated in the same surgical procedures, which reduces the time required for rehabilitation (1). In some cases, non-telescoping rods (Kirschner wires) or flexible non-telescoping nails (Rush, Nancy) can be used.

The key question is when it would be possible to start with intramedullary nailing of long bones. As soon as the child starts to walk, OI bones will break. The minimal diameter

size of FD telescoping nail is 3.2 mm. Consequently, the age of the patient and the size of the bone are limiting factors for intramedullary nailing. Generally, surgery is rarely performed before the age of 24 months. Repeated fractures (two or more) on the same bone in the period of 12 months are an indication for intramedullary stabilization. The child's ability to walk is limited by varus femur deformity and anterior curvature of the tibia, which are usual deformities in patients with OI. Femur varus deformity is better tolerated than anterior curvature of the tibia. Long bone deformity of more than 20 degrees will eventually lead to fracture. This information should be given to the patient's parents to warn them of the possible need for surgical intervention. Long-standing deformity of lower extremity bones will lead to alteration of bone morphology and the intramedullary canal will be narrowed and the bone will assume a rib shape. These facts impose the question of which type of intramedullary nail to use. The lateral profile radiograph of lower extremity may give false impression of enough space in the intramedullary canal. The anteroposterior radiograph of lower extremity will show that the intramedullary space is more narrowed than anticipated. The surgeon should have all these details in mind when planning surgery. This means that it is necessary to have FD rods of all sizes available in the operating room. Without intraoperative radiological C-arm guidance, FD telescoping surgery is not possible (8-11) (Figure 1 A-D and Figure 2 A-D).

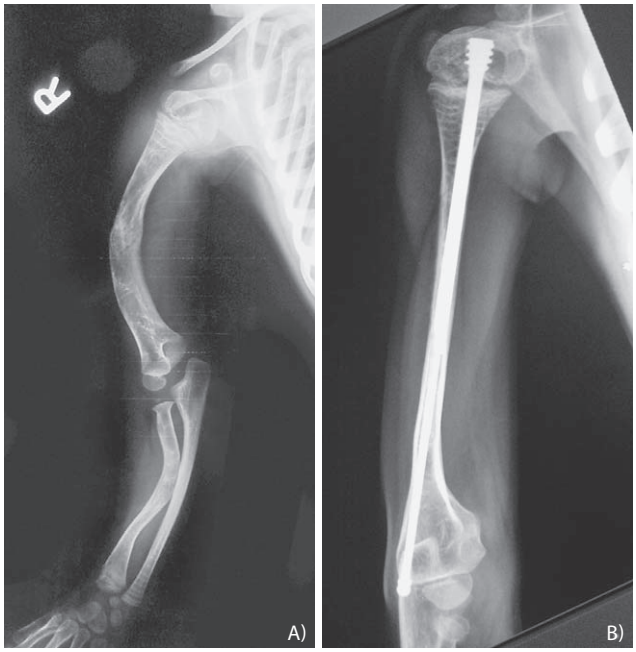

FIGURE 3. Preoperative radiograph of the right humerus (A); postoperative radiograph of the right humerus (B).

Recently, ever more patients are surgically treated for upper extremity deformities. With the administration of bisphosphonates and advances in lower extremity surgery outcomes, OI patients are using upper extremities more actively, placing more demands on function. Historically, there was one upper extremity surgery *per* ten lower extremity surgeries. This has been changed and upper extremity surgeries are nowadays more frequent. Also, FD rod is used in humerus, and solid rods are used for both bones in the forearm. Initial results are very promising (12-14) (Figure 3 A-B and Figure 4 A-D).

In the period from May 2016 to August 2017, ten children underwent surgery for OI at St. Catherine Speciality Hospi-

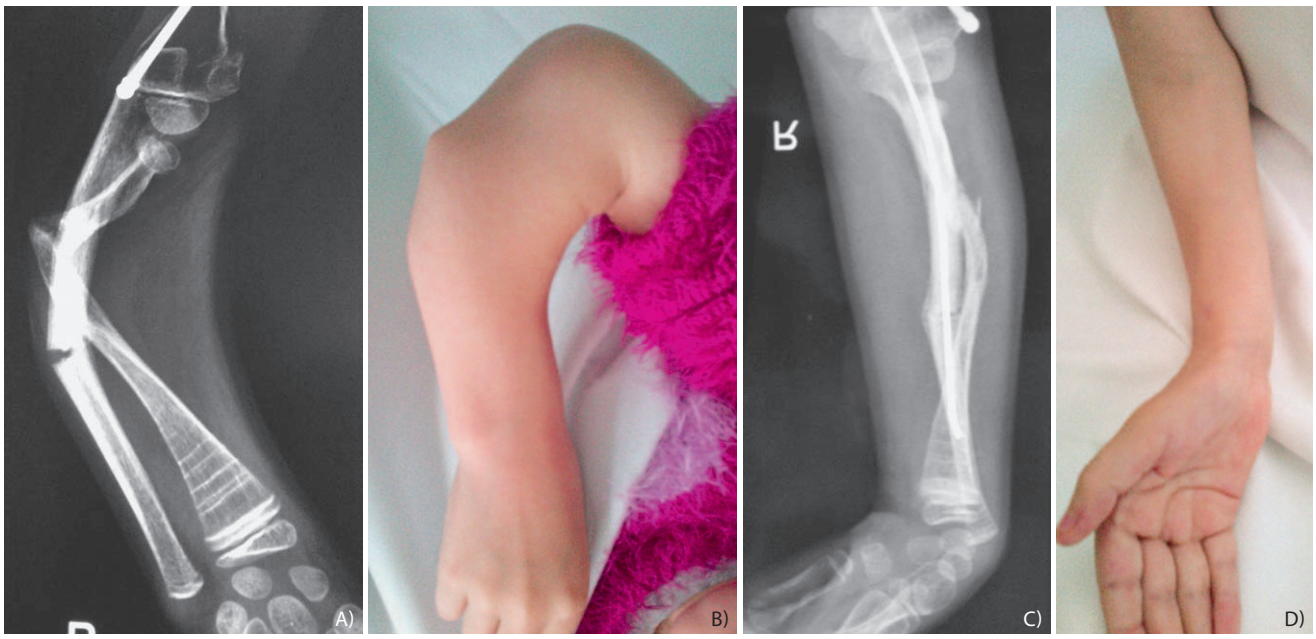

FIGURE 4. Preoperative radiograph of the right forearm (A); photograph of the right forearm before the surgery (B); postoperative radiograph of the right forearm (C); photograph of the right forearm after the surgery (D).

tal. The total number of operated bones was 16 (12 femurs and 3 tibias, and one tibial revision after surgery elsewhere); in two cases, we operated two bones in a single anesthesia. Of the total of 16 bones (segments), in 14 cases we did FD rodding, and in one patient FD rodding and augmentation with two Kirschner wires and cerclage wires. In only one patient, FD rodding was not possible due to the extremely small size of the proximal part of the femur; in this case, we performed osteotomy and fixation with two Kirschner wires and cerclage wires.

Preoperative planning of the surgical intervention is important for successful outcome. In our patients, most of the deformities were very severe with long bone shaft bowing by more than 60 degrees. In addition, bowing was simultaneous in two planes (or 3-D deformity). Consequently, preoperative planning included one or two osteotomies. Sometimes, triangular wedge shape excision of the bone was not enough. In order to overcome short soft tissue (tendon and muscles) on the concave side of angulated bone, we added some bone shortening intraoperatively, i.e. trapezoid wedge shape bone excision. Also, aponeurotic muscular release was necessary in some patients.

Very important for a good functional outcome of surgical therapy is personalized physical therapy. The main goal of physical therapy is to restore function of the extremities. It can be performed before and/or after surgery, and should be careful and individualized. It is particularly important in young children to improve weight bearing, prevent fractures, and increase strength and mobility during fracture healing. Physical and occupational therapy include muscle strengthening, prevention of joint contracture, prevention of malalignment, and improvement in everyday personal activities (15). There are many details in planning surgery in patients with OI, and the best approach is to have multidisciplinary consultation to discuss each individual patient before scheduling him/her for surgery.

CONCLUSION

The best option for surgical treatment of OI is implantation of an intramedullary telescopic nail. In many hospitals dealing with OI, the FD telescopic nail is the most frequently used implant. It is very important to know that a new generation of intramedullary nail is under way. It is expected that the new generation of the nail will additionally improve surgical treatment of OI and make it less technically demanding.

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SAŽETAK

Osteogenesis imperfecta: mogućnosti kirurškog liječenja s naglaskom na današnji ortopedski pristup

Darko Antičević, Željko Jeleč

Liječenje bolesti osteogenesis imperfecta treba biti multidisciplinsko i personalizirano. Najbolji način kirurškog liječenja je ugradnja intramedularnog teleskopskog čavla, pri čemu većina specijaliziranih bolnica prednost daje metodi Fassier-Duvalova teleskopskog čavla. Glavne prednosti Fassier-Duvalova intramedularnog čavla su manje kirurških ožiljaka, manji gubitak krvi, skraćeno vrijeme operacije, manja bolnost i bolja poslijeoperacijska mobilnost. Ortopedski kirurzi koji se bave liječenjem osteogenesis imperfecta moraju odgovoriti na dva glavna pitanja: kada započeti kirurške zahvate i koji tip intramedularnog čavla primijeniti. Nekoliko je stvari neophodno ispuniti da bi ishod kirurškog zahvata bio zadovoljavajući: precizno prijeoperacijsko planiranje, dostupnost svih veličina Fassier-Duvalova čavla u operacijskoj sali za vrijeme operacije te intraoperacijsko radiološko vođenje pomoću C-ruke.

Ključne riječi: *osteogenesis imperfecta*, Fassier-Duval, personalizirani i multidisciplinski pristup