Chirurgia (2013) 108: 120-125
No. 1, January - February
Copyright© Celsius

Use of Minimally Invasive (Percutaneous) Fassier-Duval Telescopic Rod on an 8 Year Old Patient with Lobstein Disease

R. Bălănescu[1,2], Al. Ulici[1,2], D. Rosca[2], L. Topor[1,2], M. Barbu[2]

[1]"Carol Davila" University of Medicine and Pharmacy Bucharest, Romania
[2]"Grigore Alexandrescu" Clinical Emergency Hospital for Children Bucharest, Romania

Rezumat

Utilizarea pe cale minim invazivă (percutană) a unei tije telescopice Fassier Duval la un copil în vârstă de 8 ani cu maladie Lobstein

Maladia Lobstein este o suferință severă caracterizată în primul rând printr-o fragilitate osoasă care generează apariția de multiple fracturi în special la nivelul oaselor lungi. Fracturile multiple la nivelul aceluiași os generează în timp deformări osoase importante cu impotență funcțională consecutivă. Tratamentul maladiei Lobstein trebuie să se refere atât la creșterea rezistenței osoase și prevenirea apariției fracturilor, precum și la tratarea fracturilor proaspăt produse. În ceea ce privește alegerea materialului de osteosinteză la copil sunt numeroase constrângeri generate de dimensiunea redusă a oaselor, prezența cartilajului de creștere și pierderea aportului sangvin periostal. Actual pentru tratamentul pe termen lung al fracturilor oaselor lungi se folosesc tijele telescopice Fassier Duval care pot fi menținute pe toată durata creșterii. În acest articol prezentăm un pacient în vârstă de 8 ani cu o fractură de femur survenită pe fondul maladiei Lobstein care a fost tratată inițial prin introducerea unei broșe fixe centromedulare. Ulterior a fost montată o tijă telescopică pe cale minim invazivă printr-o tehnică îmbunătățită în serviciul nostru. Considerăm că atunci când nu exista posibilitatea aplicării tijei telescopice de la început, stabilizarea fracturii trebuie făcută într-o asemenea manieră încât să permită convertirea cât mai puțin invaziv a montajului.

Cuvinte cheie: Lobstein, fragilitate osoasă, Fassier Duval, tijă telescopică

Abstract

Lobstein disease is a severe disease characterized primarily by bone fragility which causes numerous fractures, especially in long bones. Multiple fracture of a bone will cause in time a deformity with consecutive functional impotence. The treatment for Lobstein disease in children must take into account both increasing bone strength and fracture prevention and treatment of possible fractures. Regarding the choice of synthesis material, in children, there are many constraints related to the small size of the bones, presence of growth plate and the risk of periosteal blood supply infringement. Currently, for the long-term fixation of long bone fractures in children, telescopic Fassier Duval rods are used, allowing reinforcement throughout growth. In this article we present an 8 years old patient, with a femoral fracture and Lobstein disease, who underwent initial intramedullar fixation with a Kirschner pin which was subsequently replaced by a Fassier Duval telescopic rod. Mounting the telescopic rod was performed minimally invasively through a technique improved in our service. We believe that this method can be applied in all cases where other types of fracture stabilization was used, and to ensure straightness of the medullar canal.

Key words: Lobstein, bone fragility, Fassier Duval, telescopic rod

Corresponding author: Alexandru Ulici, MD
Iancu de Hunedoara Bvd., no. 30-32
Sector 1, Bucharest, Romania
E-mail: alexandruulici@me.com

Introduction

Lobstein disease is the most common constitutional bone fragility in children, the result of connective tissue damage due to a biochemical defect of genes A1 and A2, responsible for producing collagen. Collagen damage, according to most authors, is both quantitatively and qualitatively. Only in type I is 50% of the normal amount of collagen produced.

Ekman made the first scientific description of osteogenesis imperfecta in 1788. Following this there have been several classifications of the disease (Looser, Seedorf, Shapiro) but the most used is the classification proposed by Sillence in 1979 initially divided into 4 main types of osteogenesis imperfecta: Type I - mild affection, lifetime blue sclerae, autosomal dominant, Type II - lethal, Type III - severe disease, most often sclerae are normal, the transmission is either autosomal dominant or recessive, Type IV - disease ranging from mild to severe (1,2,3). Sillence's initial classification has been enriched in the last decade with three more types. In 2000 and 2002 Glorieux introduced type V and type VI, and Ward introduced in 2002 type VII (1,2,3).

In Type I Osteogenesis Imperfecta bone fragility is reduced, and fractures occur later at a free interval after birth. Sclerae are blue and retain this aspect for life. Skin biopsy may reveal quantitative changes of collagen (1,3).

Type II is the most severe form of osteogenesis imperfecta, and according to some authors it is not compatible with life. Fractures are present at birth, and the skull looks membranous. Sclerae are blue (1,3).

Type III is characterized by bone fragility and severe deformity of long bones. Fractures can occur at birth or later, from minor injury or even in the absence of any trauma. The time of fracture is an important indicator of prognosis: according to how early they occur, the prognosis is more reserved. Time of consolidation of these fractures is similar to that of fractures occurring in previously healthy bones, non-union occurring rarely. Multiple fractures installed in the same bone lead to its deformity, deformation will favour the occurrence of new fractures. Sclerae are normal in this type (1,2,3).

Type IV is characterized by a low fragility, fractures occurring after the first years of life. Sclerae colour is normal. Skin biopsy shows a qualitative change of collagen (1,2,3).

Type V is characterized by hypertrophic callus developed after a fracture and the occurrence of hyperdense metaphyseal bands. Sclerae are of normal appearance and imperfect dentinogenesis is absent. Transmission is autosomal dominant (1,3,4).

Type VI is characterized by frequent fractures, deformities of long bones, vertebral compression. Sclerae are of normal appearance. Paraclinical investigations may show an increase in alkaline phosphatase. Glorieux mentions that histology is dominated by osteoid tissue, demineralised bone matrix in the absence of hypocalcemia (1,5).

Type VII is autosomal recessive and characterized by a shorter lower limb with consecutive coxa vara installed (1,6,7).

Drug treatment in osteogenesis imperfecta is symptomatic and not curative. Since the early '90 bisphosphonates have been widely accepted in the treatment of osteogenesis imperfecta, resulting in a decrease of fracture rates and increased vertebral bone density (8,9).

Surgical treatment has three goals: treating displaced fractures after more or less intense trauma, correcting deformities after vicious consolidation and prevention of new fractures (10,11,12,13).

Sofield and Miller, in 1959, describe a technique for bone realignment by multiple osteotomies folowed by intramedullary fixation. Initially for intramedullary fixation rods or fixed pins were used (Kuntscher, Rush, 3 mm Kirschner pins). They were not however able to grow with the bone, so they became ineffective very quickly and in some cases even favoured the occurrence of new fractures at the migration site (1,14).

This inconvenience was first removed in 1963 when Bailey and Dubow introduced telescopic rods into current practice. These rods were elongated in parallel with bone growth, allowing them to be maintained a longer period of time (1,12, 13,15,16,17). Although their use has been a net improvement in the treatment of osteogenesis imperfecta, a number of disadvantages determined that these rods be abandoned by most authors. The main disadvantages were represented by detaching of the "T" part and the need for intraarticular articular approach for intramedullary mounting. To remove the inconvenience of the "T" piece, the Sheffield rod has this piece firmly attached to the main component (16,17,18,19).

As an alternative to the telescopic system, Luhmann proposes to use two Rush rods mounted opposite one another to slip one against the other along with the growth of the bone segment (1,11,12,). After the development of titanium elastic rods (offsprings of the rods described by Metaizeau) Rush rods were replaced with these. The arch ensures an additional bone stability, and their introduction is by extraarticular approach (1,11,13).

Currently, the telescopic intramedullary rods described by Fassier and Duval are produced by Pega Medical in Canada (20,21). The major advantage of these rods is that they do not require articular approach for their fitting, with a screw-like fixing method for both male and female component. Indication for Fassier Duval rods is reinforcement of lower limb in children who have significant growth potential (20,21).

Case report

The studied case is of a boy 8 years of age, who presents in our service for further treatment after a fracture of the left femur (Fig. 1). The patient was diagnosed three years ago (at age 5) with Lobstein disease (osteogenesis imperfecta) type IV following a femur fracture that was treated orthopaedically.

Three months before the patient suffered a transverse fracture of the left femur which was treated through open and internal reduction with an intramedullary 3 mm Kirschner pin (Fig. 2). Skin biopsy performed during surgery confirmed the connective tissue fault and maintained the type



Figure 1. *Fracture of left femur*



Figure 2. *Centromedular K-wire*

IV osteogenesis imperfecta classification (fracture occurred late after birth, sclerae were normal, imperfect dentinogenesis absent, qualitative changes of collagen).

At admission the patient had a limp with shortened support time on the left lower limb, postoperative scar healed, and the synthesis material was present in the supra trochanteric region. During examination, absence of limb length discrepancy, absence of pelvic imbalance or any clinical deformity of the spine were determined.

Left knee mobility is normal: flexion 140 degrees, extension 0 degrees.

Passive mobility of the hip can not be appreciated because of significant pain in the gluteal region, pain triggered mainly by external rotation (20 degrees of external rotation triggers discomfort) and abduction of the thigh on the pelvis.

Radiological examination highlights the consolidation of the left femur fracture and the tendency of the synthesis to migrate (Fig. 3, 4).

Paraclinical investigations showed a discrete increase in alkaline phosphatase, erythrocyte sedimentation rate 30 mm / h, normal serum calcium.

Considering net patient discomfort, in conjunction with radiological image and the need to keep the syntesis material for a long time we decided patient admission and replacing the existing Kirschner pin with a telescopic Fassier Duval type rod(considering the growth potential remaining).

Treatment

Radiological appearance, showing that both in AP and lateral projection the bone is straight, led us to attempt a minimally invasive percutaneous insertion of the Fassier Duval rod. To achieve this goal it was necessary to modify the original technique and adapt it to the present conditions. All calculations and preoperative measurements

Figure 3. *Migration of the K-wire (AP image)*





Figure 4. *Migration of the K-wire (lateral view)*

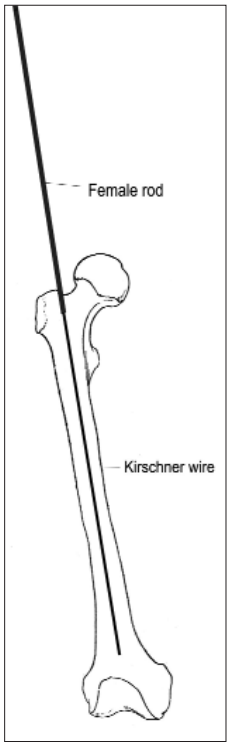


Figure 5. *Using K-wire for guide*

were made under the premise that no corrective osteotomy will be required.

Choosing the rod

The original technique described by Pega Medical for the telescopic intramedullary Fassier Duval rod system recommends that the length of the rod be calculated preoperatively after a preliminary sketch to take into account any necessary corrective osteotomies. Male rod length is measured from the greater trochanter to the distal end of the femur adding an extra 1-1.5 cm which will have to exceed the greater trochanter. Female rod is measured from the greater trochanter (considering the fact that proximal threads of the rod must be fully included in the greater trochanter to avoid any stall post-operatively) to the male rod thread, minus mandatory 7 mm. All measurements are made taking into account the radiological image multiplication factor of 1.3.

Considering the fact that in our patient the preoperative plan excludes corrective osteotomies, the telescopic rod choice was made on the AP radiogram, maintaining the multiplication factor of 1.3. Choosing the distal thread of the male rod was done by measuring the distal femur metaphysis and we decided to use the long threaded rod model (even more because we wanted to avoid using the distal stabilization pin).

Rod thickness was determined by measuring the medullary canal in the narrowest point, both on AP and lateral radiographic incidence. Chosen thickness was 4 mm.

Operative field

To obtain an optimal surgical field we must take into account three objectives: easy approach to the trochanteric region, Rx view of the entire bone segment (femur) and easily raising the thigh to obtain at least two radiological projections. To achieve these goals we placed the patient in a lateral semi-decubitus support with support under the hip and we used a hip dressing. The patient was placed on a radiolucent operating table that allowed the introduction of an image intensifier under the thigh.

Conductor introduction

Given that the patient had been operated previously we decided to use the existing Kirschner pin as a guide to preserve the safest intramedullar path. The problem was that the existing pin thickness (3 mm) did not allow the use of the original kit reamers supplied. Under these circumstances we decided to extract the pin and replace it with a much thinner (1.5 mm) pin. To avoid the risk of losing the entering point at the greater trochanter and be forced to turn minimally invasive surgery in invasive surgery, we extracted the Kirschner pin only partially and introduced the female's rod first 2 cm. Then we extracted the thick pin and introduced a new thin guide pin in the metaphyseal region of the distal femur (Fig. 5)

Reaming

Reaming was performed with a 4.25 mm reamer introduced from the proximal to the distal end on the new guide pin to the distal metaphyseal region of the femur. We recommend that distal reaming be performed partially progressing in the metaphysis to allow a greater thread stability of the male component of the telescopic rod.

The male component insertion:

- *After proper reaming of the medullary canal the reamer and guide pin are both extracted. The male component of the system is mounted in his conductor and is*



Figure 6. *Distal part of fassier Duval rod*

inserted through the greater trochanter. This maneuver should be performed under X-ray control and by putting gentle pressure so as not to create a false path. The male component will be threaded distally until it exceeds the growth plate. Rx control is required to ensure that the articular cartilage is not exceeded.

Preparation insertion of female component:

- *For safe installation, in this case we opted for cutting the female rod intraoperatively. On image intensifier we measured the necessary length respecting the rule to cut the rod with 7 mm shorter than the starting point of the thread of the male rod. Particular caution was taken to the intratrohanteric portion of the proximal thread. When measuring the female rod we kept in mind that the proximal threads must be fully included in bone and not only in the cartilage region of the greater trochanter to avoid any subsequent stall. Using the male component as a guide, we inserted the female component progressively checking that the distal thread do not goes beyond the articular cartilage (Fig. 6).*

Fitting control

At the end of surgery, before cutting the surplus of the male rod we performed a Rx control on film to check the following: male rod thread must not penetrate the knee joint and female rod thread should be fully included in the bone structure of the greater trochanter.

Postoperative conduct

Immediately postoperatively the patient was immobilized in a plaster (pelvis-thigh-calf-foot) for 14 days especially for the analgesic effect, considering the fact that no osteotomy was performed on the femur. After 14 days we removed the plaster and we authorized walking with a support device for two weeks. Four weeks after surgery the patient had resumed walking without support.

Discussion

The minimally invasive technique without osteotomy of the femur, different from the original minimally invasive technique (20,21), in which femoral corrective osteotomy is required, performed through percutaneous approach, is possible only in situations where previously there was a fracture that was treated with an intramedullary rod (Rush or Kirschner) which ensures the rectitude of the medullary canal.

In all other cases it is absolutely necessary to perform an osteotomy of the femur and to apply the original technique. So when using elastic rods or if the femur is not fractured, its physiological or acquired curvature requires performing



Figure 7. *Titanium elastic nail used for osteosinthesis*



Figure 8. *Final result*

one or more osteotomies and retrograde reaming of the proximal fragment (*Fig. 7*).

Conclusion

The minimally invasive technique of anterograde reaming from the greater trochanter to the distal epyphisis can be used to solve a previously treated fracture of the femur when an rigid intramedullary nail was used (*Fig. 8*)

To avoid loosing access to the medullary canal we recommend that the initial rod, especially if it is a Kirschner wire, to be used as a first guide for the male component.

If conditions do not allow the use of the telescopic rod as a first intention, a femur fracture in this type of patient should be treated with an intramedullary rigid fixation of maximum 3 mm. In this way, at a later moment, a telescopic rod might be mounted easily through a minimally invasive using the existing wire as a guide.

The use of the image intensifier, much reduced by this method, remains necessary to verify correct placement of the distal and proximal part of the fitting.

Postoperative immobilization in a cast has analgesic purposes only, walking (when it is possible) is authorized at a maximum period of 14 days after surgery.

Reference

1. Burnei G, Vlad C, Georgescu I, Gavriliu TS, Dan D. Osteogenesis imperfecta: diagnosis and treatment. J Am Acad Orthop Surg. 2008;16(6):356-66.
2. Glorieux FH. Osteogenesis imperfecta. Best Pract Res Clin Rheumatol. 2008;22(1):85-100.
3. Sillence D, Senn A, Danks D. Genetic heterogennity in osteogenesis imperfecta. J Med Genet. 1979;16(2):101-16.
4. Glorieux FH, Rauch F, Plotkin H, Ward L, Travers R, Raughley P, et al. Type V osteogenesis imperfecta: a new form of brittle bone disease. J Bone Miner Res. 2000;15(9):1650-8.
5. Glorieux FH, Ward LM, Rauch F, Lalic L, Roughley PJ, Travers R. Osteogenesis imperfecta type VI: a form of brittle bone disease with a mineralisation defect. J Bone Miner Res. 2002;17(1):30-8.
6. Ward LM, Rauch F, Travers R, Chobot G, Azouz EM, Lalic L, et al. Osteogenesis imperfecta type VII: an autosomal recesive form of brittle bone disease. Bone. 2002;31(1):12-8.
7. Lalenda M, Morisette J, Ward LM, Rauch F, Lalic L, Roughley PJ, Glorieux FH. Osteogenesis imperfecta type VII maps to the short arm of chromosome 3. Bone. 2002;31(1):19-25.
8. Ward LM, Rauch F, Whyte MP, D'Astous J, Gates PE, Grogan D, et al. Alendronate for treatment of pediatric osteogenesis imperfecta: a randomized placebo-controlled study. J Clin Endocrinal Metab. 2011;96(2):355-64.
9. Ruck J, Dohan-Oliel N, Montpetit K, Rauch F, Fassier F. Fassier Duval femoral rodding in children with osteogenesis imperfecta receiving bisphosphonates: functional outcomes at one year. J Child Orthop. 2011;5(3):217-24.
10. Esposito P, Plotkin H. Surgical treatment of osteogenesis imperfecta: current concepts. Curr Opin Pediatr. 2008;20(1): 52-7.
11. To M, Gupto V, Chow W. Surgical management of long bone pseudarthrosis with severe length discrepancy in osteogenesis imperfecta. J Pediatr Orthop B. 2013;22(1):63-9.
12. El-Adl G, Khalil MA, Enan A, Mostafa MF, El-Lakkany MR. Telescoping versus nontelescoping rods in the treatment of osteogenesis imperfecta. Acta Orthop Belg. 2009;75(2) :200-8.
13. Sułko J, Radło W. Osteogenesis imperfecta – operative treatment on lower extremities in children with osteogenesis imperfecta. Chir Narzadow Ruchu Ortop Pol. 2005;70(3):189-93. Polish
14. Abulsaad M, Abdelrahman A. Modified Sofield-Millar operation: less invasive surgery of lower limbs in osteogenesis imperfecta. Int Orthop. 2009;33(2):527-32.
15. Karbowski A, Schwitalle M, Bremmer R, Lehmann H, Pontz B, Worsdorfer O. Experience with Bailey-Dubow rodding in children with osteogenesis imperfecta. Eur J Pediatr Surg. 2000; 10(2):119-24.
16. Oznur A, Tokgozoglu AM, Alpaslan AM. Complications in the use of the Bailey-Dubow extensible nail. Clin Orthop Relat Res. 1999;(366):286-7.
17. Janus GJ, Vonpaemel LA, Engelbert RH, Prujis HE. Complications of Bailey-Dubow elongating nail in osteogenesis imperfecta: 34 children with 110 nails. J Pediatr Orthop B. 1999; 8(3):203-7.
18. Nicolaou N, Bowe JD, Wilkinson JM, Fernandes JA, Bell MJ. Use of the Sheffield telescopic intramedullary rod system for the management of osteogenesis imperfecta: clinical outcomes at an average follow-up of nineteen years. J Bone Joint Surg Am. 2011;93(21):1994-2000.
19. Luhmann SJ, Sheridon JJ, Capelli AM, Schoenecker PL. Management of lower extremity deformities in osteogenesis imperfecta with extensible intramedullary rod technique: a 20-year experience. J Pediatr Orthop. 1998;18(1):88-94.
20. Sandell LJ. Multiple hereditary exostosis, EXT genes, and skeletal development. J Bone Joint Surg Am. 2009;91 Suppl 4:58-62.
21. Birke O, Davies N, Latimer M, Little DG, Bellemore M. Experience with Fassier-Duval telescopic rod: first 24 consecutive cases with minimum of 1 year follow-up. J Pediatr Orthop. 2011;31(4):458-64.