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Combination of flexible intramedullary nailing and Ilizarov frame for salvage of femur and humerus nonunion in a girl with osteogenesis imperfecta

E.R. Mingazov[1], P. Foster[2], A.V. Popkov[1], D.A. Popkov[1]✉

[1] Ilizarov National Medical Research Centre for Traumatology and Orthopedics, Kurgan, Russian Federation
[2] Leeds Teaching Hospitals NHS Trust, Leeds, UK

Corresponding author: Dmitry Popkov, dpopkov@mail.ru

Abstract

Background Fixation of pathological long bones with telescopic intramedullary rods is well known to be a technically challenging procedure even in specialist centres, with a high complication rate due to rod migration, hardware failure, nonunion or malunion. However there is very little guidance in the literature regarding salvage treatment options when failure occurs. **Aim** We demonstrate a surgical technique that can be used for salvage treatment of both femoral and humeral complex nonunions following Fassier-Duval (FD) rodding in a child with osteogenesis imperfecta (OI). **Case description** A 13 year-old girl with OI type VIII presented sequentially with nonunion and deformity of the femur then the humerus following previous FD rods in those segments. The femur was also complicated with metallosis between the steel rod and an overlying titanium plate. Both segments were treated with pseudarthrosis debridement, removal of metalwork and stabilisation with hydroxyapatite (HA)-coated flexible intramedullary nails, with temporary Ilizarov frame to provide enough longitudinal and rotational stability to allow immediate weight-bearing. The femur Ilizarov frame was removed after 64 days, and the femur remained straight and fully healed at 2.5 years. The frame time for the humerus was 40 days, complete union was achieved and upper limb function restored and maintained at 9 months. **Discussion** The transphyseal telescopic rod is the traditional implant of choice in terms of treating fractures and stabilising osteotomies for deformity in OI. However, it does not provide enough torsional or longitudinal stability by itself to allow early weight-bearing which is detrimental to bone healing in this vulnerable patient group. The incidence of delayed union or nonunion at osteotomy site in telescopic rod application is not negligible: up to 14.5-51.5 %. Although the technique we have shown in this case may not be applied to all complex OI patients, we believe that the combination of flexible intramedullary nails and Ilizarov frame provides a favourable environment for bone healing in complex or revision cases. As a secondary learning point the initial revision surgery to the left femur demonstrated the perils of using a steel rod and a titanium plate in a biologically active environment which in this case lead to metallosis and lysis. **Conclusion** We found the technique of HA-coated flexible intramedullary nails combined with the Ilizarov frame effective in the salvage of failed telescopic rods in both femur and humerus and feel this technique can be used as a salvage option in similar cases worldwide. This case also demonstrates the perils of using different metals in combined internal fixation.

Keywords: osteogenesis imperfecta type VIII, flexible intramedullary nailing, external fixation, telescopic rodding

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BACKGROUND

Osteogenesis imperfecta (OI) is a phenotypically and genetically heterogeneous group of inherited bone dysplasias characterized by frequent fractures, bone deformities, low bone mineral density and osteopenia [1-4]. Although rare, each patient often requires multiple operations on several limbs throughout childhood and beyond, in order to maintain autonomy, self-care, physical activity and the ability to acquire and develop motor skills including independent ambulation.

The workhorse implant for the treating surgeon in cases of OI is the telescopic rod which strengthens and protects the full length of the long bones, where one component (male) slides inside the other (female) with growth. As well as allowing fractures and osteotomies to heal the rods prevent deformity and fracture recurrence [5, 6]. Models include the Fassier-Duval rod [7–9], the Sheffield rod [10], dual interlocking telescopic rod [11], and the titanium telescopic rod [12, 13].

Whatever the exact implant design telescopic rodding carries a steep learning curve, and even in the most experienced hands has a high complication rate, partly related to the lack of rotational and longitudinal stability of the implant [6, 11, 14, 15]. The technique is known to be even more difficult and prone to these complications in the humerus, including secondary displacement, malunion, nonunion, hardware failure and migration over time and bone growth [16-19]. This means that revision surgery in these cases is not at all unusual, and is even more challenging than primary cases requiring individual solutions as well as classic techniques [9, 18, 20-22].

Aim This article presents a case report of operative treatment of a 13-year-old girl affected with osteogenesis imperfecta type VIII, who was consecutively operated on with a combined technique with hydroxyapatite-coated flexible intramedullary nailing (HA-FIN) and reduced external frame for salvage of femoral pseudarthrosis complicated with metallosis and pseudarthrosis of humerus after Fassier-Duval (FD) rod failure in both segments.

CASE DESCRIPTION

The initial treatment of this child with regards to the right femur was described in a published report [18]. Two years later the patient represented to us with a symptomatic nonunion in the other (left) femur, having been treated with primary FD rod elsewhere in 2013, and then attempted revision with exchange FD rod followed by additional titanium plate 4 months before presentation.

On review, the girl was unable to walk or stand, on a background of short stature with rhizomelia, white sclerae, myopia and partial hearing loss. Pain was localized to the midshaft of the left femur and knee, in the area of the mobile nonunion site. There was a leg length discrepancy of 1.5 cm and 25 degree loss of extension in the left knee. There was no evidence of infection including on blood tests. Clinical alignment of the right lower limb remained excellent without any deformity rebound or implant migration, with satisfactory X-rays. Views of left femur showed a clear nonunion with a 4 mm gap, with FD rod in place as well as lysis around the supplementary plate and screws.

Left femur surgery

The existing FD rod was removed as well as open debridement of the pseudarthrosis and overlying plate and screws. Extensive metallosis was noted around the soft tissues and periosteum around the plate. The canal was reamed to remove any bone fragments from the screw tracts and the previous apex anterior deformity corrected. Two 2.5 mm HA-coated titanium flexible intramedullary nails (Orthopediatrics nails modified by Metis Ltd, Tomsk, Russia) were inserted in anterograde fashion. The Ilizarov apparatus (Experimental Plant at the Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, Kurgan, Russia) consisted of a short proximal arch with three half-pins and a distal ring with three wires. This was a static construct which did not require any postoperative adjustment or correction.

Standing and progressive weight-bearing commenced on day 3 and the patient was discharged home on day 8 with a physical therapy programme including early knee movement. Postoperatively, no problems or obstacles were encountered such as pin-site infection or loosening. X-rays at 60 days showed union and the fixator was removed at day 64 under general anaesthetic. The lower limb was protected in a split cast for a period of two weeks and then weight-bearing was recommenced. Bisphosphonate therapy was resumed at 4 months. Independent standing and pain-free walking was restored and maintained at 2 years, with full union and alignment of the femur demonstrated on X-rays with remodelling and restitution of the intramedullary canal.

Right humerus presentation and surgery

Shortly after this episode there was a similar presentation of the right arm in the same child. The right humerus had been previously treated in another institution with FD rod. A painful and mobile non-union of the distal humerus was noted, with functional impairment. X-rays revealed an established nonunion as well as broken male component of the FD rod. There was no evidence of infection including on blood tests.

A proximal approach was used to remove both the female component of the FD rod as well as the proximal part of the broken male component. The nonunion site was debrided back to viable bone and the distal part of the male component removed. In similar principles to the femur surgery, the humerus was stabilised with two HA-coated flexible intramedullary nails and a reduced Ilizarov frame, consisting of 4 half-pins fixed over a short Ilizarov plate connected with threaded rods. This provided excellent bone contact, rotational stability and allowed free elbow movement. Neither casting nor bracing was required or any postoperative adjustments to the Ilizarov apparatus.

There were no problems or obstacles postoperatively. The humerus healed and the fixator was removed at 40 days, and the arm was left free to move. Bisphosphonate therapy was recommenced at 6 months. At 9 months there was pain-free restoration of upper limb function, as well as radiological union and remodelling of the canal.

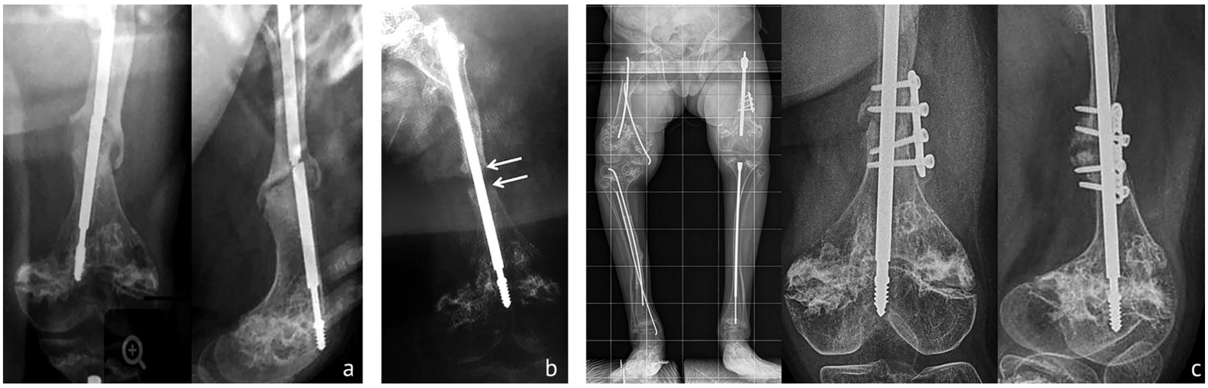

Fig. 1 Preoperative radiographs: a – pseudarthrosis of the left femur and hardware fracture; b – fractured rod has been replaced with another Fassier-Duval rod, note interfragmentary diastasis (fleshs) as result of longitudinal instability; c – standing radiograph of lower limbs, augmented radiographs demonstrate loosening of the plate and bicortical screws, nonunion

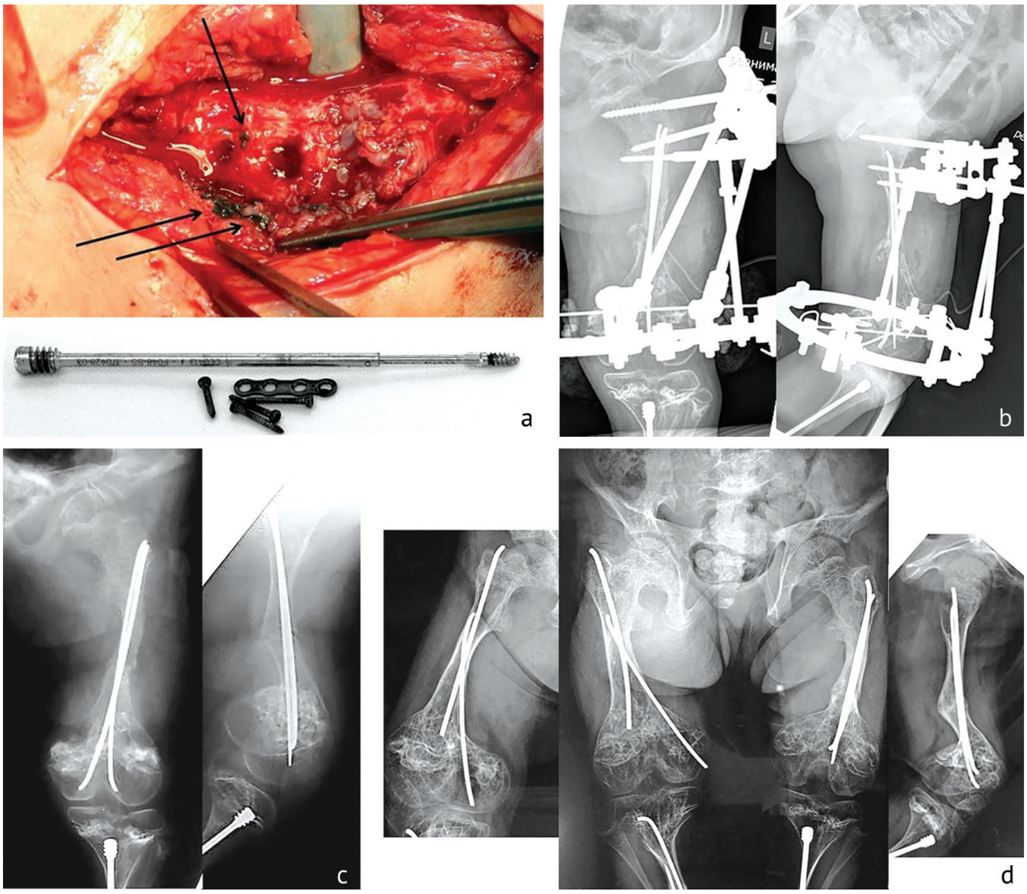

Fig. 2 Treatment of left femur pseudarthrosis: a – intraoperative view: nonunion site extensive metallosis of multiple charred appearance inclusions on the intraoperative view, removed hardware; b – postoperative X-rays; c – day 64 after surgery, radiographs at frame removal; d – 2 years after frame removal from left femur

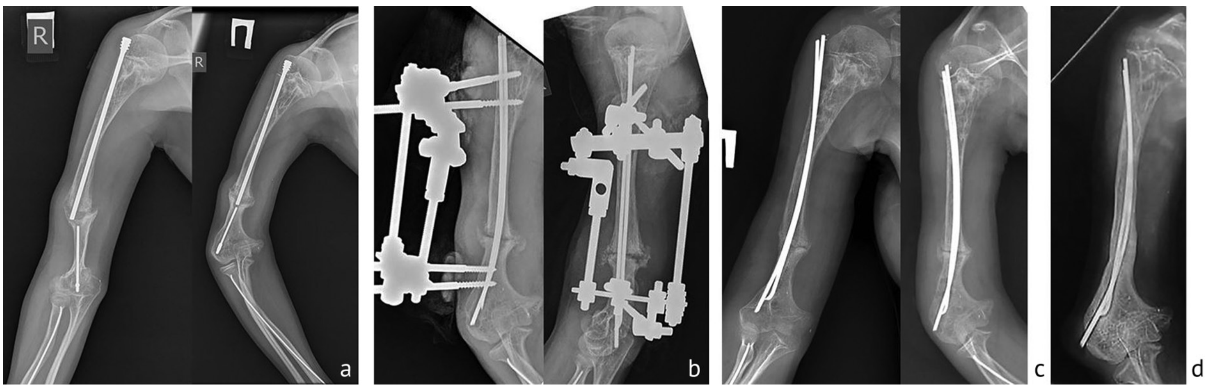

Fig. 3 Treatment of right humerus pseudarthrosis: a – preoperative radiographs, pseudarthrosis and broken Fassier-Duval rod; b – postoperative radiographs; c – bone union achieved, frame removed on day 40; d – 9 months after frame removal from right humerus

Fig. 4 Functional outcome

DISCUSSION

The transphyseal telescopic rod is the traditional implant of choice in terms of treating fractures and stabilising osteotomies for deformity in OI [5-8]. However, it does not provide enough torsional or longitudinal stability to allow early weight-bearing [6. 8, 10, 11, 16] which is detrimental to bone healing in this vulnerable patient group [23, 24] as well as the overall rehabilitation [22].

The incidence of delayed union or nonunion at osteotomy site in telescopic rod application is not negligible: up to 14.5 % [16]. Munns et al observed delayed bone union in 103 cases out of 200 (51.5 %) operated on with the Fassier-Duval rod [17]. Shin et al reported 5 cases of persisting cortical gap for a series of 59 rods [11]. The case presented in our article does not match to reported data. Nonunion and rod fracture occurred in two segments of three within period of 3 and 5 years. The peer-reviewed literature reports that for a five-year period, the telescopic rods did not required surgery in 63 % for the femur and 64 % for tibia according to Cox et al [25]. Shin et al used Dual Interlocking telescopic rod (D-ITR) and reported 75 % survival rate for 5.3 years follow-up, which is thebest result among all known telescopic rods [11]. On the other hand, the rate of revision surgery also depends on the severity of OI and it reaches 67.86 % in OI type III and 31.82 % in OI type IV [26]. We presume that there could be features of bone union in patients with type VIII of OI (recessive inheritance pattern). There is no relevant literature for OI type VIII specifically.

Even in "classic" OI the literature suggests that corrective osteotomy of the humerus with FD rod carries a high complication rate [19, 20]. Grossman et al reported revision rate of 34.3 % within 35 months including unscheduled surgery for nonunion and malunion in 8.6 % of cases [19].

Although the technique we have shown in this case may not be applied to all complex OI patients (such as the very young) we believe that the combination of HA-coated flexible intramedullary nails and Ilizarov frame provides a favourable environment for bone healing in complex or revision cases [28, 29]. There is an enhanced mechanical stability (both particularly rotational and longitudinal) compared to telescopic rods alone, as well as a favourable osteoconductive environment.

As a secondary learning point, the initial revision surgery to the left femur demonstrated the perils of using a steel rod and a titanium plate in a biologically active environment which in this case lead to metallosis and lysis [30, 31]. The technique of combined telescopic rodding and plate fixation per se is supported by the literature, but the metals used should be similar, also locking unicortical screws rather than plates with bicortical conventional screws [15, 27].

CONCLUSION

We found a high rate of nonunion and pseudarthrosis associated with hardware fracture in a patient with rare type VIII of osteogenesis imperfecta initially treated with FD rods in each segment. A combination of pseudarthrosis site resection, elastic intramedullary nailing with osteoinductive coating and external frame applied for a short period provides early function including full weight-bearing and enables bone union and further favorable bone remodeling in long-term follow-up. The surgeon should also be aware about the risks of use of dissimilar metals in combined osteosynthesis resulting in osteolysis and early loosening of implants.

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Information about the authors:

1. Eduard R. Mingazov – Candidate of Medical Sciences, orthopedic surgeon, ed.dobryak@gmail.com, Scopus: 56897223100;
2. Patrick Foster – MD, PhD, pediatric orthopedis surgeon, patrick.foster@nhs.net, Scopus: 7201517406;
3. Arnold V. Popkov – Doctor of Medical Sciences, Professor, orthopedic surgeon, apopkov.46@mail.ru, https://orcid.org/0000-0001-5791-1989;
4. Dmitry A. Popkov – Doctor of Medical Sciences, Professor, orthopedic surgeon, dpopkov@mail.ru; https://orcid.org/0000-0002-8996-867X.

Contribution of the authors:

Mingazov E.R. – Development, research activities; Application of formal methods for the analysis or synthesis of research data.
Popkov A.V. – Responsibility for managing and coordinating the planning and conduct of research activities
Foster P. – Preparation and writing of the initial draft (draft) of the work.
Popkov D.A. – Gathering data/evidence, Preparing and writing an initial draft of the paper, Applying formal methods to synthesize study data. Conducting surgical treatment.