

Clinical case

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## Polysegmental distraction osteosynthesis in combination with a telescopic transphyseal rod for Ollier's disease

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### Abstract

**Introduction** The use of combined techniques for long bone lengthening in patients with Ollier disease is of interest due to issues of both external fixation time reduction and improvement in the strength properties of the lengthened bone. The uniqueness of the presented clinical case is simultaneous femur and tibia lengthening over a titanium telescopic rod in the femur and tibia combined with external osteosynthesis and osteotomies.

The **purpose** of this study is to demonstrate the stages and results of polysegmental bone lengthening in a child with Ollier disease using a technique combining the Ilizarov fixator and a titanium telescopic rod.

**Material and methods** This clinical case is a five-year-old patient with Ollier disease without comorbidities, but with a significant discrepancy in the length of the lower limbs, impaired weight-bearing ability, and impaired gait biomechanics. Surgical treatment included multisegmental femur and tibia lengthening over a titanium telescopic rod inserted retrograde into the femur and antegrade into the tibia, simultaneous osteotomies and placement of a wire-and-halfpin external fixator.

**Results** Exceptional data were obtained confirming the possibility of the titanium telescopic rod extension during distraction by more than 20 % without locking or loss of fixation of the threaded parts in the epiphyses. The elongation amounted to 11.4 cm (femur: 5.8 cm; tibia: 5.6 cm). The overall external fixation index was 12.02 days/cm. At a 12-month follow-up after dismantling the Ilizarov apparatus, growth of the elongated bones and telescoping of the femoral rod continued.

**Discussion** This combined technique is consistent with the current concept of extended telescopic reinforcement in children with genetic diseases. The divergence of the titanium rod components both during the distraction period and during the first year after completion of limb lengthening demonstrates the feasibility of this combination and the reliability of using a telescopic titanium rod. This study demonstrates for the first time that rapid divergence does not predispose to locking of titanium rods and continues with growth.

**Conclusion** This case demonstrates the effectiveness of using a combination of telescopic intramedullary titanium rods and external fixation for simultaneous lengthening of two adjacent segments in a child with Ollier disease. The findings on further natural growth of the lengthened segments along with telescoping of the rod are unique.

**Keywords:** telescopic rod, Ollier disease, limb lengthening

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## INTRODUCTION

Multiple enchondromatosis, or Ollier disease, is a rare pathology of the human skeleton (one case per 100,000), characterized by multiple enchondromas or cartilaginous masses in the metaphyseal and diaphyseal regions of long and/or short bones of the extremities [1–4]. Treatment of diseases accompanied by low bone strength properties (Ollier disease, osteogenesis imperfecta, polyostotic fibrous dysplasia, metabolic osteopathies) is symptomatic and aimed at eliminating and preventing significant bone deformities. Intramedullary telescopic rodding is a key element of osteosynthesis [5–10]. Reinforcement technologies simultaneously used with external fixation have been described in foreign and domestic literature [8–12]. Lengthening over a rigid rod using an external fixator or its analogues eliminates the risk of fractures after removal of the external fixator and significantly shortens the rehabilitation period [13–16]. In pediatric orthopedics, telescopic rods inserted transphyseally for deformity correction and fixed in the proximal and distal epiphyses provide bone reinforcement throughout the body growth period [6, 16–18]. The uniqueness of the clinical case presented is due to the combination of simultaneous femur and tibia lengthening over a titanium telescopic rod implanted in the femur and tibia simultaneously with external osteosynthesis and osteotomies.

The **purpose** of this study is to demonstrate the stages and results of polysegmental bone lengthening in a child with Ollier disease using a technique combining the Ilizarov fixator and a titanium telescopic rod.

## MATERIAL AND METHODS

A patient with radiographically and genetically confirmed Ollier disease and no concomitant somatic diseases applied for surgical treatment at the age of five. Prior to his presentation to the Ilizarov Center, the patient had not undergone any surgical treatment. Shortening of the femur and tibia had increased as the child grew and was 18 cm at hospitalization.

Chondromatous lesions were located in the proximal and distal femur and tibia, affecting the epimetaphyseal zones. They were thickened bone sections with typical mineralization defects and alternating areas of mineralization and radiolucent inclusions (Fig. 1). Functional limitations included decreased walking ability, the need for assistance, and the use of shoes with significant compensation for the shortening of the left lower limb.

For studying the child's treatment outcomes, we identified key points of his examination: admission, surgery, day 10 and day 30 of distraction phase, the end of distraction, achievement of union, the period immediately after removal of the external fixator (EF), six months after EF removal, and 12 months after EF removal. At each of those points, we performed radiography in the frontal and lateral views, clinical examination of the child, and measurement of the lengthening amount (distraction gaps). We also studied the formation of distraction regenerates, assessed the position of the ends of the telescopic rod, and compared the lengths of separation of bone fragments and rod components.



**Fig. 1** Lower limb radiographs at admission

For telescopic reinforcement, a titanium intramedullary telescopic rod was used (RU No. RZN 2017/5875 dated July 10, 2017, included in the set of implants and instruments for pediatric orthopedics "Orthokid", TU 9437-001-73747729-2014).

### ***Stages of the surgical lengthening over a telescopic rod***

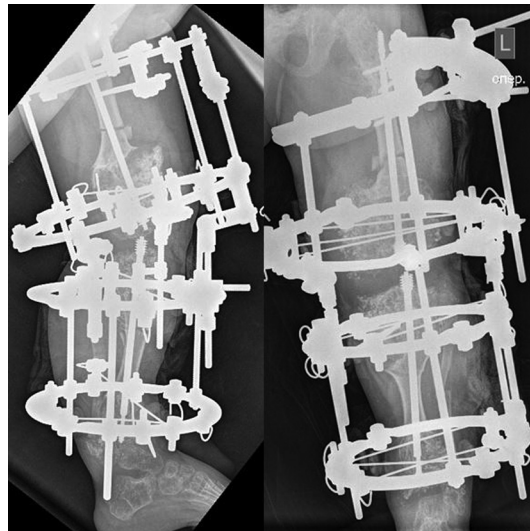
Initially, retrograde transphyseal femoral rodding was performed through a medial parapatellar approach. A guide wire was drilled under the guidance of an image intensifier through the apex of the intercondylar fossa along the medullary canal to the level of the antecurvatum deformity in the lower third of the femur through a 1-cm skin incision. Next, an osteotomy was performed at the apex of the deformity with a chisel. The guide wire was then advanced to the proximal femur. The canal was drilled along the wire using a 5.7-mm cannulated drill from the kit. Next, the guide wire was removed, and the inner portion of the telescopic rod was inserted into the growth plate of the greater trochanter using a T-handle. Subsequently, the outer (hollow) portion of the 5.5-mm diameter telescopic rod was inserted, with the threaded portion immersed strictly into the distal epiphysis. The length of the outer portion was determined intraoperatively and shortened accordingly. The final step in the femur involved screwing the inner portion of the rod into the greater trochanter so that the entire threaded portion was proximal to its growth plate. The protruding portion of the inner element was cut as short as possible. The osteotomy wound was irrigated and sutured layer by layer.

From the same parapatellar approach, a guide wire was inserted antegrade under radiographic control, entering through the intercondylar space along the medullary canal to the level of the deformity in the upper third of the tibia. An osteotomy at the apex of the deformity was performed through a 1-cm approach. An osteotomy of the fibula was also performed through a 1-cm skin incision in the lower third of the tibia. Next, a cannulated 5.8-mm drill was used to drill the canal down to the distal metaphysis, and the inner portion of the telescopic rod was advanced to a level directly above the distal growth plate. Next, the correspondingly shortened outer portion, 5.5 mm in diameter, was inserted, and the threaded portion was screwed into the proximal tibial epiphysis using a cannulated screwdriver, that did not extend beyond the growth plate line. The final step in placing the tibial telescopic rod was to screw the threaded portion into the distal epiphysis so that the entire thread was distal to the growth plate line. Cutting off the protruding portion of the internal component was performed immediately before irrigation and layer-by-layer suturing of the parapatellar approach. Radiographic checks of the entire limb confirmed the correct placement of the telescopic rods (Fig. 2).

The wire-and-halfpin variant of the external fixator consisted of two tibial rings, a ring over the lower third of the femur, and a segmental short arch at the level of its upper third. The distal femoral ring and the distal tibia ring had only 1.8 mm wires at the metaphyseal level. The proximal femoral arch used only 4.5-mm halfpins screwed into the proximal metaphysis, and the proximal tibia ring had a combination of wires and a half-pin, also located at the metaphyseal level (Fig. 3). At the knee joint, hinges were used to connect circular supports.



**Fig. 2** Radiograph taken during surgery after osteotomies, introduction of intramedullary rods and before placement of the external fixator



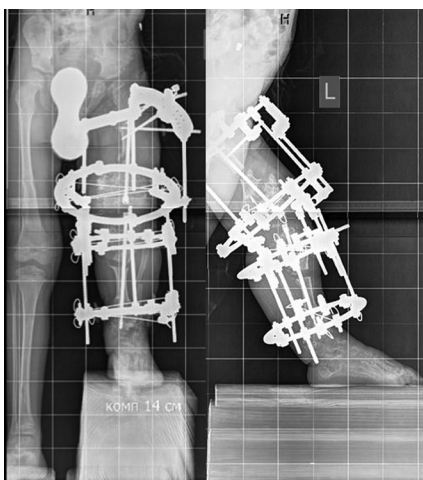
**Fig. 3** Radiographs taken after completion of combined osteosynthesis

### RESULTS

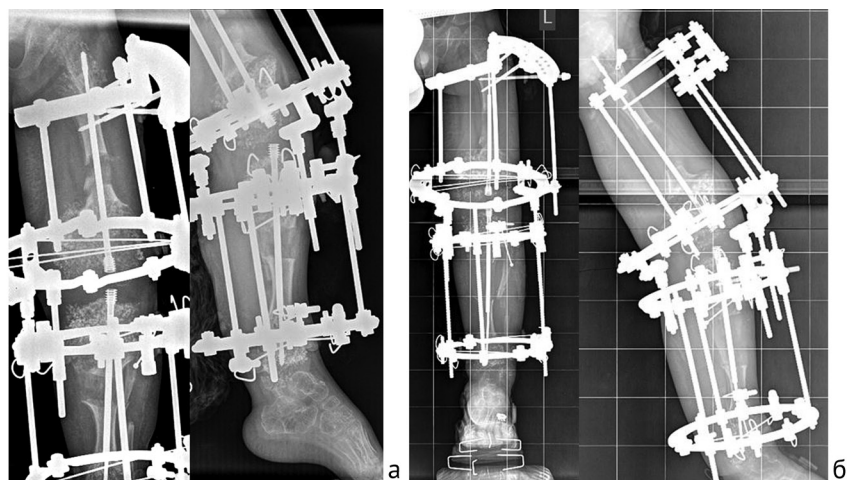
The immediate postoperative period was uneventful. The patient was able to stand upright on the second postoperative day, being assisted. Three days after surgery, distraction was initiated in the femur and tibia. The initial distraction rate was 1 mm per day.

A 10-day follow-up image revealed no separation of the fibula fragments, which was considered premature consolidation and necessitated a repeat osteotomy and insertion of an additional wire to secure the proximal fibula fragment (Fig. 4). Subsequent radiographs showed normal separation of the bone fragments (Fig. 5). The structure of the distraction regenerate was represented by longitudinal bands with a high degree of mineralization, and the periosteal component of developing bone callus was clearly visible. Extension of the telescopic rod parts occurred without any locking and, consequently, without loss of fixation of the threaded parts in the epiphyses and apophysis of the greater trochanter.

At the end of the second month of lengthening, given the cumulative relative lengthening of more than a third of the lower limb's length, distraction was stopped. Throughout the distraction period, the patient maintained the ability to walk with crutches and functional weight-bearing.



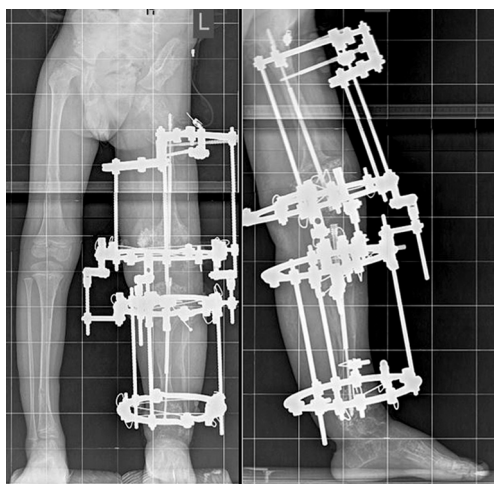
**Fig. 4** Radiographs on the 10<sup>th</sup> day of lengthening



**Fig. 5** Radiographs during the distraction period: (a) 30 days of distraction, (b) 60 days of distraction

The patient spent the fixation period at his home. His limb condition was monitored via correspondence, with radiographs taken every 30 days. Bone union was confirmed radiographically after 30 days of fixation (Fig. 6). However, due to the remote distance from the Ilizarov Center, the patient was only able to attend it for removal of the fixator after a long enough period.

The fixator was removed under general anesthesia, leaving the titanium telescopic rods *in situ* (Fig. 7). Post-removal radiographs showed the correct position of the telescopic rods, adequate extension, and maintenance of fixation of the threaded parts in the epiphyses and apophysis of the greater trochanter. A plaster cast was applied for 10 days. Weight-bearing was allowed on the first postoperative day.



**Fig. 6** Radiographs of the lower limb, 30 days of fixation period



**Fig. 7** Radiographs after external fixator removal

Follow-up checks were performed at six and 12 months after removal of the external fixator. The elongated segments continued growing; there was no recurrence of deformities, and correct telescoping of the titanium rods were noted without locking or protrusion into the knee or ankle joints. Bone callus remodeling showed all the characteristics of a healthy bone tissue remodeling without the inclusion of chondromatous foci in its structure (Fig. 8).



**Fig. 8** Radiographs taken 6 (a) and 12 (b) months after external fixator removal

The resultant quantitative treatment parameters were:

- Femur lengthening by 5.8 cm, tibia lengthening by 5.6 cm, which totally amounted to 24.5 % of the initial limb length,
- The total external fixation index was 12.02 days/cm,
- Considering the treatment outcome, as well as the complication that occurred, which required an unplanned intervention in the period before dismantling the Ilizarov apparatus, but did not lead to a deterioration in the achieved result, the lengthening outcome can be classified as corresponding to grade IIa according to the Lascombes classification [19].

#### DISCUSSION

Lengthening of long bones in children suffering from diseases associated with low bone strength (Ollier disease, osteogenesis imperfecta, polyostotic fibrous dysplasia, metabolic osteopathies) is rarely used in surgical practice. This is due to the difficulty of performing external osteosynthesis, which must continue for a long time what may be impossible in the conditions of impaired bone strength [20–22].

On the other hand, telescopic transphyseal reinforcement is the method of choice for mechanical strengthening of long bones in such conditions [7, 15, 16]. Difficulties with the use of telescopic rods include cases of non-extension, locking, and loss of fixation of threaded parts in the epiphyses [22, 23].

The feasibility of combining intramedullary reinforcement and external fixation for deformity correction in children with osteogenesis imperfecta has been demonstrated: this combination provided both mechanical and functional advantages [5, 24, 25].

Further development of this approach was envisaged in the use of this combined technique not only for deformity correction but also for limb lengthening.

This clinical case demonstrated the feasibility of this approach without compromising the formation of the distraction regenerate and its maturation time in big lengthening magnitude (polysegmental distraction osteosynthesis). Furthermore, the clinical case demonstrated the safety of using a titanium telescopic rod as an internal fixator without the risk of deformity and/or locking during accelerated telescoping, even during distraction. However, some doubts about the rod's quality had been raised, based on the nature of the alloy used.

The success of such surgeries depends on both the precision of the procedure itself (correct placement of the telescopic rod, minimizing blood loss and trauma by using a single approach to implant two telescopic rods, and restoring acceptable anatomical relationships in the operated segments) and ensuring stable external fixation providing weight-bearing of the lower limb to ensure early axial loading. This approach enabled to significantly shorten the time of external fixation required for lengthening two segments, prevented complications such as bone fragment displacement in the postoperative period, and long-term deformity recurrence during natural growth.

#### CONCLUSION

This clinical case demonstrated the feasibility of lengthening long bones in patients with Ollier disease using an external fixator along with simultaneous placement of a transphyseal titanium telescopic rod. The study of this lengthening procedure showed for the first time that "accelerated" extension of the rod components during distraction does not lead to sliding locking and, consequently, to the loss of the required fixation of the threaded components in the epiphyses. Furthermore, no delay in the formation and maturation of the distraction regenerate was observed. One year

after dismantling the Ilizarov apparatus, the growth of the elongated segments continued and divergence of the telescopic rod components was observed. It reflects the preserved function of prolonged limb reinforcement both after reconstructive interventions and in the presence of transphyseal reinforcement.

**Conflict of interests** The authors declare no conflict of interest.

**Source of funding** None.

**Ethical statement** This study was approved by the Ethics Committee of the Ilizarov Center (protocol dated November 29, 2024, No. 1(76)).

**Informed consent** The patient's authorized representatives provided written informed consent for surgical intervention and processing of anonymous data.

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