Original Articles

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Results of using transphyseal elastic intramedullary nailing in patients with severe types of osteogenesis imperfecta

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Introduction The work analyzes the results of using elastic opposite-directional transphyseal reinforcement for treatment of severe orthopedic complications in patients with osteogenesis imperfecta as well as the ways and their effectiveness to overcome the shortcomings of this technique. **Material and methods** The series included 24 patients. Among them, four patients had osteogenesis imperfecta of Type III, 19 were of Type IV, and one patient had Type VIII. The patients' mean age was 14.4±2.8 years (range: 2-46 years). A total of 52 reconstructive surgical interventions were performed. Elastic intramedullary reinforcement was used in 83 segments. The combined osteosynthesis technique (the Ilizarov fixator and/or subperiosteal reinforcement) was used in 27 cases. **Results** Correction of deformities was achieved in all the cases. Consolidation at the osteotomy level was obtained after 26.2±7.8 days on the average (from 3 to 12 weeks postoperatively). The period of follow-ups was from 6 months to 4 years. Twenty-four complications were observed in 8 patients (33.3 % of cases). Twenty three additional interventions due to the problems of correction were performed in 20 patients (unplanned surgeries were necessary in 83.3 % of cases). The ability to stand in the vertical position and bear weight on the lower limbs with the use of auxiliary supports or without them was achieved in 22 cases out of 24 (91.7 %). Patients' ambulation ability was evaluated with the Gillette scale and improved in 21 cases out of 24 (87.5 %). In all the cases, patient's care facilitated, self-service capabilities improved, and patients' social activity increased. A more comfortable position for sitting was achieved in 22 patients. In 100 % of the cases, pain reduced or disappeared by doing exercises or during walking, including exercise therapy or hygiene procedures. **Conclusion** The technique of transphyseal reinforcement using elastic titanium nails is indicated for deformity correction in children with severe types of osteogenesis imperfecta. Lower limb deformity correction and increased bone strength contributed to patients' motor activity and improved their quality of life. Complications were not rare after surgical treatment but their timely correction enabled to retain the achieved anatomical and functional results. The use of a minimum fixation, the Ilizarov apparatus following corrective osteotomies and intramedullary reinforcement, assisted in achieving patient's early verticalization and full weight-bearing on the operated limb as well as helped avoid a number of complications. Osseointegration of intramedullary implants with bioactive coating prevented their migration in the long-term period. Telescopic constructs of such nails should not be used in children under the age of 10 in order to avoid their locking in the medullary canal due to osseointegration.

Keywords Osteogenesis imperfecta, surgical deformity correction, transphyseal elastic intramedullary reinforcement, osseointegration

Osteogenesis imperfecta (OI) is a group of genetic diseases that is characterized by the fragility of bone tissue, frequent fractures, skeletal deformities and osteopenia [1-4]. In the majority of cases, OI is caused by a dominant mutation of the genes that are responsible for collagen type I synthesis [2, 3]. Its incidence ranges from 1/10000 [4] to 1/20000 newborns [5]. Sillence's clinical and radiographic classification [1] is the most used one for the cases in which a genetic typing of the anomaly was not conducted. This classification was widened later. Types V through VIII were added that are characterized by a radiographic OI manifestations but have a recessive type of disease inheritance [3, 4, 6].

The main goal of orthopaedic treatment of limb deformities and fractures in the OI patients is maintenance of their motor activity, autonomy, abilities to acquire and develop movements. The treatment methods should exclude prolonged immobilization that results in secondary decrease in bone mineral density on the background of osteopenia that this disease features [7-18].

Indications to surgical interventions are lower limb angular deformities greater than 10° to 15° and torsion in association with functional limitations, nonunion, bone defects, varus of the proximal femur (neck-to-shaft angle of 95° degrees or lower), inability of autonomous or passive vertical positioning or walking due to frequent fractures [4, 6, 19-23].

The use of telescopic intramedullary constructs is the main ways of providing osteosynthesis in correction of pediatric orthopedic limb pathology [20, 24-27]. Transphyseal sliding flexible intramedullary nailing remains one of the ways of telescopic osteosynthesis [25, 28-30]. It is used in the bones that have a small external diameter or when the bone marrow canal either is obliterated or its reaming is not able to provide enough widening for telescopic rod introduction [21, 25, 28-30]. Its shortcomings are

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those of the telescopic systems in general: migration of elastic nails, secondary torsional bone fragment displacements, and inability to load the limb. The latter drawback results in secondary osteoporosis and, therefore, is a precondition for nail migration and bone fractures [25, 29, 30].

Our paper presents the results of our own study on the use of elastic counter-directional bone reinforcement for management of orthopaedic complications in severe OI types as well as on the ways and their efficiency in overcoming the shortcomings of this technique.

MATERIAL AND METHODS

Between 2012 and 2015, we studied 29 patients who suffered OI and passed surgical treatment at our institution. For assessments of the outcomes, we chose the cases that corresponded to the inclusion criteria: the method of intramedullary reinforcement with curved inside the bone elastic nails that are introduced in opposite direction to each other, either alone or in combination with other osteosynthesis methods for correction of deformities, limb discrepancy, femur and/or tibia pseudarthrosis or defects. Patients had severe OI types (III, IV, and VIII). Period of follow-up was not shorter than 6 months. We selected 24 patients according to the criteria mentioned.

Mean patients' age was 14.4±2.8 years (range: 2-46 years). There were 16 patients younger than 18 years old among them. Only 11 patients took biophophonates in this group. Four patients were of OI type III, 16 had type IV, and one was of type VIII. Seven patients had a laboratory confirmation of OI diagnosis. In the remaining cases, OI was diagnosed only by clinical and radiographic examination. Seven individuals were previously treated for deformities that were corrected using osteosynthesis with angular stability plates (5 cases), or osteosynthesis with either diafixing wires or a rigid intramedullary rod (2 cases).

In all the cases, deformities in the lower limbs were greater than 30 degrees and biomechanical axis deviation was more than 20 mm from the knee joint centre They had a history of multiple fractures before admission. Femoral nonunion was present in 6 cases and one patient had tibial nonunion after a corrective osteotomy. Osteosynthesis material was present in several cases: plates in 5 cases and intrumedullary nails in 2 cases.

Fourteen patients had Level 2 of motor activity according to Gillette scale (Gillette Functional Assessment Questionnaire [31]. Such patients are able to do several steps with somebody's assistance but cannot bear weight on the limbs. Five individuals had Level 1 and were unable to walk. Five were of Level 3 and were able to walk during the rehabilitation session only but needed somebody's assistance in other circumstances. Thus, none of the patients of this series could walk independently in the everyday life before surgery.

Lower limb deformities caused inconvenience in a sitting position. Twenty-two patients out of 24 had pain and/or fear of fractures by doing passive or active movements with the limbs.

A total of 52 reconstructive surgeries were performed in these patients. Elastic intramedullary nailing was used in 83 segments. Details of reconstruction variants with the use of elastic intramedullary nailing are given in **Table 1**.

In the majority of interventions (40), titanium elastic nails were used that were from 1.5 to 4 mm in diameter chosen in regard to the bone marrow canal. Titanium intramedullary nails with bioactive coating and from 1.8 to 2 mm in diameter were used in the other 12 operations. In children with open growth zones, intramedullary nails were introduced transphyseally in the direction towards each other in order to achieve their telescopic divergence as bone segments grow. Titanium nickelide meshes TN-10 (filament thickness of 150 mcm, mesh diameter of 2 to 2.5 mm) 60 × 80 mm in size were used for subperiosteal reinforcement as they were found efficient for thickening of the cortical layer. The mesh covered the bone in a circular manner at the osteotomy level and extended for 6 to 8 cm to the adjacent bone areas. The periosteum was separated preliminary and sutured to the mesh to maximally cover the bone circumference.

Types of surgical interventions are given in **Figure 1**.

Deformity correction planned, consolidation at the osteotomy level, deformity recurrence, frequency of migration or non-divergence of the intramedullary elements, necessity of unplanned interventions were analyzed. Moreover, patients' motor activity according to Gillette questionnaire, pain relief or its change by limb movements, and improvement of patients' care and self-service ability were assessed.

The quantitative findings obtained were statistically processed using Microsoft Excel 2016. The statistic study also included descriptive statistics: mean values (**M**) and standard deviation (**δ**).

Table 1

Surgical interventions

Type of operation	Number of operations
Acute deformity correction at one, two or three levels, elastic intramedullary nailing	21 (including subperiosteal reinforcement with a titanium mesh – 3)
Acute deformity correction at one, two or three levels, elastic intramedullary, minimal fixation osteosynthesis with the Ilizarov apparatus	17 (including subperiosteal reinforcement with a titanium mesh – 3)
Gradual deformity correction or lengthening with transosseous osteosynthesis in combination with elastic intramedullar nailing	8 (including subperiosteal reinforcement with a titanium mesh – 4)
Preventive transphyseal elastic nailing	4
Coxa vara correction and femoral neck pseudarthrosis management according to Fassier	2
Total	52

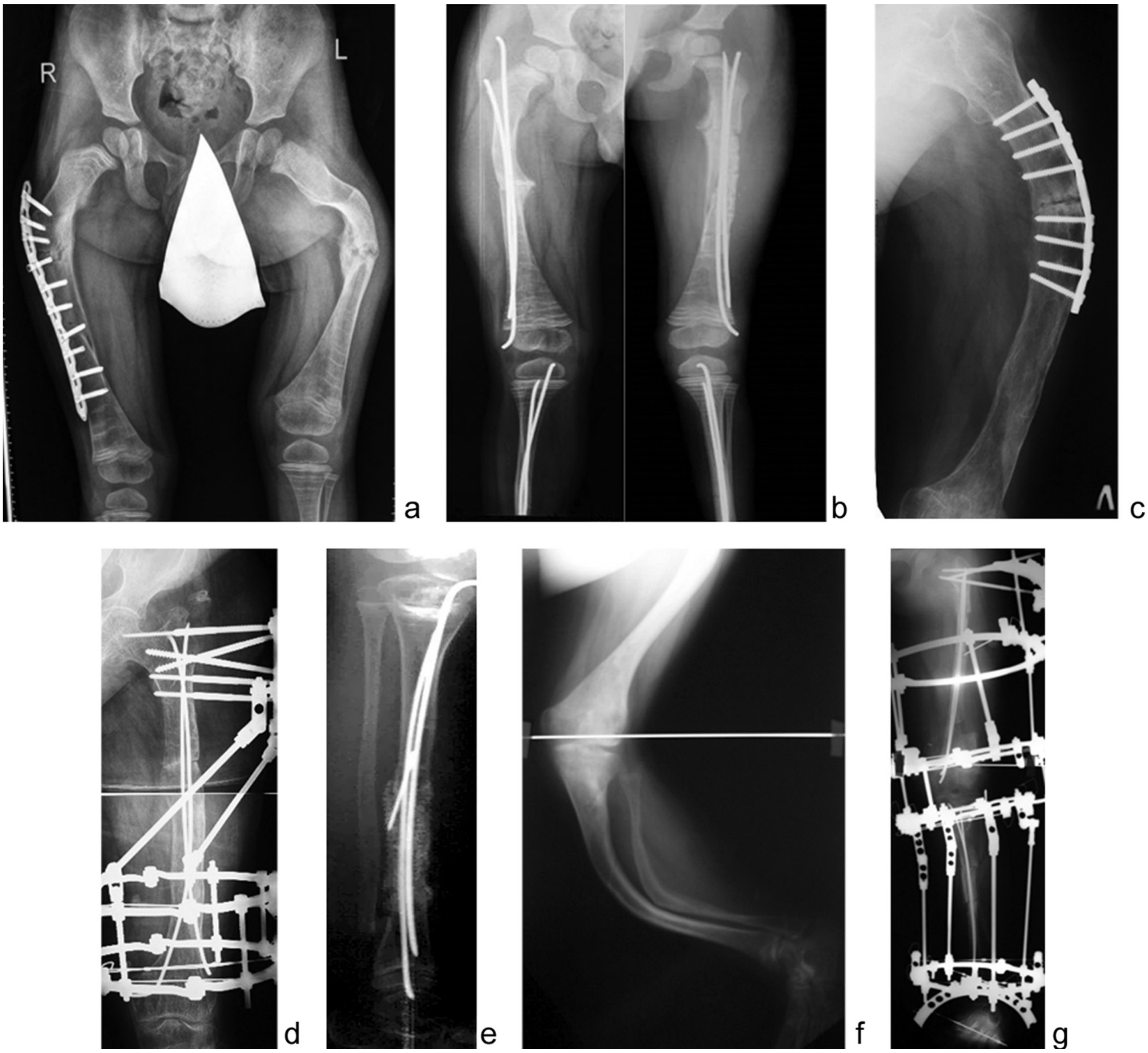

Fig. 1 Radiographs of segments as examples of the intervention types: Before deformity correction (**a**). After acute correction and transphyseal elastic intramedullary nailing (**b**). Before deformity correction in the femur (**c**). After acute correction and transphyseal elastic intramedullary nailing in combination with the Ilizarov osteosynthesis (**d**). Intamedullary and subperiosteal reinforcement (**e**). Before deformity correction (**f**). After gradual correction of the deformity with the Ilizarov apparatus in combination with intamedullary and subperiosteal reinforcement (**g**)

RESULTS

Deformity correction was achieved in all the cases. Osteotomy consolidated after a mean period of 26.2 ± 7.8 postoperative days (from 3 to 12 weeks). A clear continuous periosteal response was seen at the osteotomy level. Manifestations of radiographic bone union did not differ between the patients with isolated intramedullary nailing in combination with either subperiosteal reinforcement and/or osteosynthesis with the Ilizarov apparatus.

In cases of deformity correction combined with the transphyseal intramedullary nailing, patients could securely bear full weight load on the operated limb and walk with additional support means starting from week 5 or 6 after surgery that is by the end of immobilization with the plaster cast or by the moment of transition to a detachable orthosis (**Fig. 2**).

We revealed secondary torsion in 8 segments in the early period after the use of isolated intramedullary nailing and corrective osteotomies that needed additional correction (**Fig. 3**).

The use of transphyseal intramedullary nailing for deformity correction enabled the patients to stand on the operated limbs fully bearing weight on them and start independent walking with additional support means (crutches or walkers) from postoperative days 3 to 7 when they were inpatient (**Fig. 4**). External fixation in its minimal variant could prevent secondary torsion of bone fragments in all the cases. The external fixator was used until primary bone callus was formed, by weeks 3 or 4, that was sufficient to prevent bone fragment rotation in the conditions of intramedullary osteosynthesis that continued.

The external fixation in the cases when deformity correction was combined with the resection of pseudarthrosis area continued from 3 to 12 weeks (31.1 ± 12.7 days). During this period mild compressive efforts were maintained between the Ilizarov apparatus supports. Vertical positioning and weight bearing on the operated limbs was an obligatory condition (**Fig. 5**). Manifestation of bone callus was the criterion to terminate the osteosynthesis with the Ilizarov apparatus.

The patients were followed up after the reconstructive treatment (range: 6 months – 4 years). Twenty-four complications were observed in 8 patients (33.3 % of cases). They were angulation recurrence in the segments operated (deformity angle more than 10°) in four patients, secondary torsion in 6 segments of 4 patients (isolated intramedullary reinforcement), migration of intramedullary nails in 5 cases, non-divergence of biocoated intramedullary nails due to a continued longitudinal segment growing and possible osteointegration in 4 patients under 10 years of age, 3 cases of non-displaced fractures of the reinforced segments, delayed consolidation in two patients who were 39 and 46 years old. Correction of these problems required 17 unplanned operations. Moreover, preventive introduction of additional intramedullary elements was necessary in three cases due to intramedullary nails locking even in the absence of new deformities. Nails were changed in three more cases as these children continued growing. Thus, twenty-three additional interventions due to the problems of correction were performed in 20 patients, or unplanned surgeries were necessary in 83.3 % of cases. This study only enumerates the problems encountered without their detailed analysis which will be done in a separate paper. However, we should state that there were 3.13 operations per patient in this series during the follow-up period.

As for functional outcomes, the ability to stand in the vertical position and bear weight on the lower limbs with the use of auxiliary supports or without them was achieved in 22 cases out of 24 (91.7 %). Patients' ambulation ability was evaluated with the Gillette scale. Level 1 was one case (not able to make a single step), three patients were of Level 3 (able to walk during the rehabilitation session but needed somebody's assistance in other circumstances), five were of Level 4 (able to walk slowly at home but ambulation was not a preferable type of motion at home), Level 5 in 10 cases (able to walk more than 4.5 m up to 15 m at home or school and ambulation is the main way in home conditions), and five were patients of Levels 7 and 8 (able to walk independently on an even surface and to overcome steps and unevenness with a minimal help or under a supervision). Totally, motor activity improved in 21 cases out of 24 (87.5 %). In all the cases, patient's care facilitated, self-service capabilities improved, and patients' social activity increased. A more comfortable position for sitting was achieved in 22 patients. In 100 % of cases, pain reduced or disappeared by doing exercises or during walking, including exercise therapy or hygiene procedures.

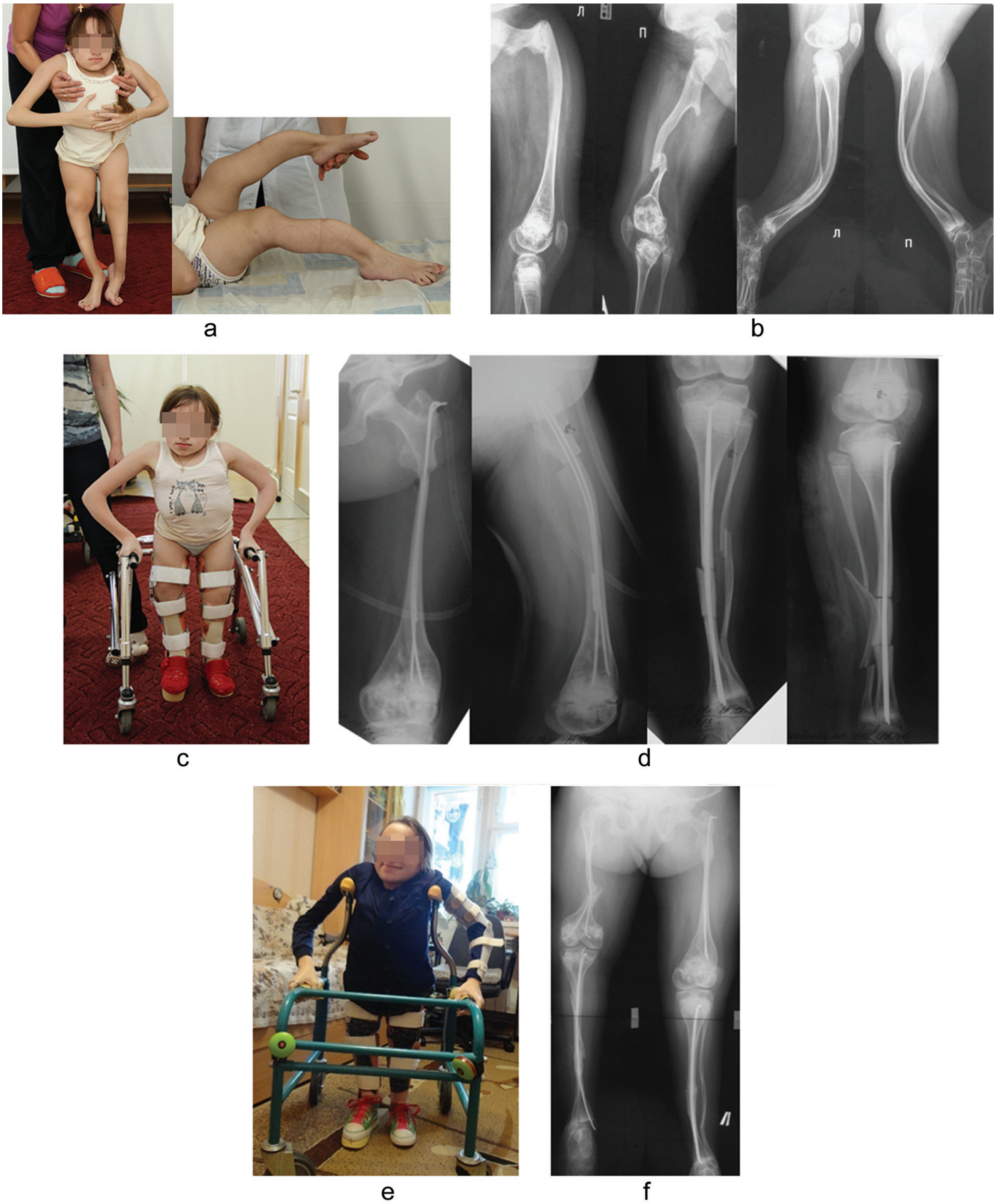

Fig. 2 Patients V, OI type III: Before treatment; loading and passive vertical positioning impossible (**a**). Preoperative X-rays of the limbs (**b**). Patient in the vertical position with walkers and orthotic appliances, fully loading the limbs (3 months after treatment) (**c**). X-rays of the femur and tibias upon consolidation and plaster cast removal (**d**). 2 years after treatment, independent ambulation at home wearing orthotic faixators (**e**). Axial X-rays 2 years after treatment, correct limb axis and absent implant migration (**f**)

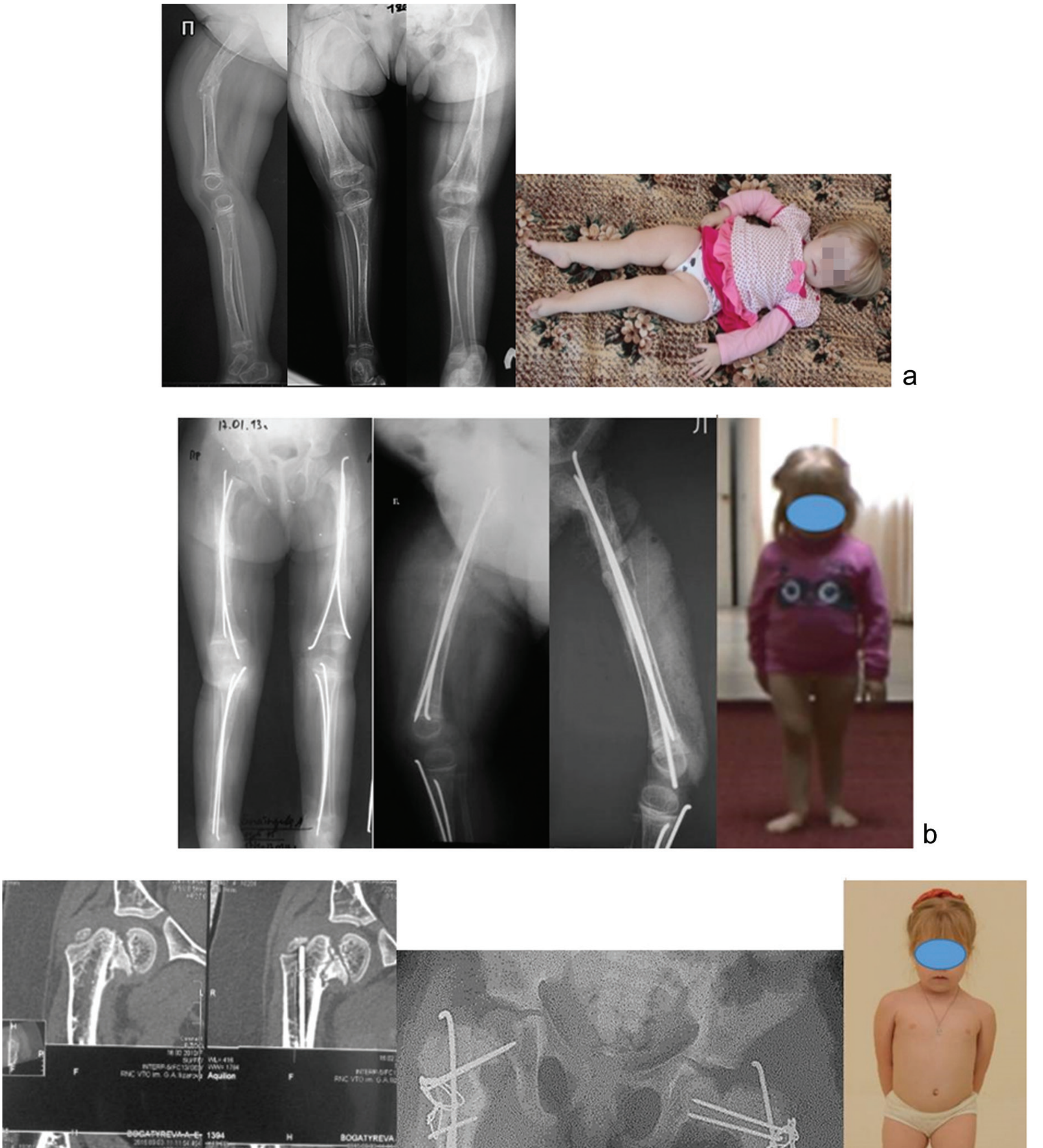

Fig. 3 Patient B, OI type IV: Preoperative lower limb X-rays and a photo. Patient is unable to stand or walk (**a**). Postoperative X-rays of lower limbs after deformity correction and intramedullary nailing. Femoral neck retroversion is clearly seen in the lateral views that explains her standing in the position of external orientation of the feet and knee joints (**b**). Stage of varus deformity and femoral neck pseudarthrosis correction for elimination of torsion. Orientation of her feet is correct when the patient stands independently (**c**)

Fig. 4 Patient K, OI type IV: Patient's photo and lower limb X- rays before treatment. Patient was not able to stand due to frequent fractures, had external torsion and a shorter right femur, varus deformity in the left femur (**a**). Photo and lower limb X- rays after the first surgery: corrective osteotomy of the right femur, transphyseal intramedullary osteosynthesis with elastic nails in the opposite direction to each other and minimal Ilizarov apparatus fixation, and preventive transphyseal reinforcement of the tibia with the nails in the opposite direction allowed axial loading from the 4th postoperative days (**b**). Photo and X-rays after the second intervention that followed the first one 3 weeks later: corrective osteotomy of the left femur in combination with a 2-cm resection for equalization of leg length, transphyseal intramedullary osteosynthesis with elastic nails in the opposite direction to each other and minimal Ilizarov apparatus fixation, preventive transphyseal reinforcement of the tibia with the nails in the opposite direction and removal of the Ilizarov apparatus from the right limb that allowed a full axial loading on the operated limb (**c**). Photo and an X-ray of the lower limbs 6 months after the treatment. The axes of the limbs were correct, intramedullary nails did not migrate, and the secondary torsion was absent. The patient is able to stand and walk independently (**d**)

Fig. 5 Photo and X-rays of patient D., OI type VIII: X-rays of lower limb segments. Reinforcement surgeries had been previously done at a foreign hospital. Defect-diastasis due to right femur fragments divergence, proximal migration of the Rush rod and valgus antecurvatum are seen (**a**). X-rays after mild resection (1-2 mm) of the ends of the femoral fragments and transphyseal intramedullary reinforcement with the nails in the opposite direction, minimal Ilizarov apparatus osteosynthesis, corrective osteotomies of the tibia, transphyseal counter-directional intramedullary and subperiosteal nailing. The patient was able to stand up on day 7 post-surgery and could exercise full weight-bearing on the operated limb (**b**). The apparatus was removed 8 weeks after achieving bone union. Vertical positioning and full loading on the lower limbs was possible with the use of walkers and orthosis (**c**)

DISCUSSION

The main tasks of surgical orthopaedic treatment by correction of limb deformities and fractures in severe OI types are maintenance of their motor activity, autonomy, ability to acquire and develop movements, or namely, improvement of their quality of life in regard to their OI type. This task is a part of a multi-disciplinary approach to their treatment that solves the same tasks [14-18].

The main osteosynthesis means used in pediatric OI patients are intramedullary telescopic constructs: transphyseal intramedullary nails [25, 28-30], expandable Bailey-Dubow rods [24], and telescopic Fassier-Duval rods [28, 33, 34].

Transphyseal elastic intramedullary nailing features a number of positive features such as possibility to apply it in small sizes of the bone marrow canal or after its reaming in cases of obliteration. This type of osteosynthesis is possible to use with a subperiosteal location of thin nails at the diaphysis level and transosseous transphyseal nails at the metaphyseal level when it is a necessity in children under one year of age [21, 25, 28-30]. Boutard et Laville reported on the success of this technique in 14 patients with severe OI types in a mean age of 4 years (minimal age of 5 days) with the rate of operations per patient equal to 2.5 (range: 1-5) [25]. The study did not report the problems of nail divergence but their change into ones of a larger length and diameter as children continued to grow was necessary in 75 % of cases. Frequency of severe complications in that series was 25 % and those were: fractures (mostly with an oblique and spiral fracture line in the diaphysis that passed between the nails), implant migration, nonunion, bone shortening due to impaction [25, 29, 30]. The Fassier-Duval telescopic rod is considered to be a non-rigid telescopic construct that is commonly used for long bone deformity correction in children and in preventive osteosynthesis [21, 33, 34]. However, the complication rate by its application is 35 % [33]. There are general problems associated with the use of telescopic intrameduallary systems [25, 35, 36] such as necessity to change systems as a child grows, telescopic system parts divergence, migration of nails or system parts seen in 10.5-23.7 % cases, deformation of telescopic system rods (up to 18.8 %), non-union or separation of bone fragments (7.2 %), non-expansion of nail parts (2.1 %), nail break (6.9 %), fractures at the osteotomy level in the conditions of telescopic intramedullary osteosynthesis with the Fassier-Duval rod (20-25 %). It is important to note that the independent use of intramedullary constructs foresees a period of strict additional immobilization for 4 to 6 weeks when limb loading is not allowed in order to prevent secondary deformities. This period may cause secondary postoperative osteoporosis [25, 28, 37-39]. Moreover, straight telescopic rods and elastic intramedullary nails do not completely prevent secondary torsion of bone fragments in the early postoperative period. Thus, femoral neck retroversion develops that clinically is associated with a marked lateral rotation of the entire limb [7, 21, 22, 24].

In accordance with the data of the published literature, angulation was corrected in all the cases of our series. Functional abilities of patients and their quality of life improved considerably. We did not observe serious problems with bone consolidation. There were only two adult patients with delayed union in the area of a wedge corrective resection at the level of pseudarthrosis and plating osteosynthesis that continued more than three months.

Total rate of complications that was 33.3 % is similar to the one observed with the use of the Fassier-Duval telescopic rod [33]. The mean number of operation per patient coincides with the data of using elastic transphyseal reinforcement [25].

We should note also that the use of the Ilizarov apparatus osteosynthesis in its minimal volume as additional reinforcement during the first 3 to 4 weeks after the operation showed the benefits of this technology over the standard use of intramedullary constructs and plaster immobilization. This approach enabled patients to acquire the vertical position fully bearing weight on the operated limb almost from the first postoperative days. It completely excluded the risk of secondary osteoporosis, bone fragments divergence, and nonunion. There was not any case of oblique or spiral fractures at the diaphyseal bone level in the cases of titanium nickelide mesh application for subperiosteal reinforcement.

The use of elastic intramedullary nails with bioactive coating [40, 41] was justified to prevent implant migration at long term. However, all the cases of intramedullary implants non-divergence were observed with the use of biocoated nails in children under 10 years of age. The obvious explanation of the phenomenon is osteointegration of nails in the bone marrow canal. Our own experience and literature data [25] show that the non-divergence problem due to child's growth does not arise if the regular titanium nails are properly fixed to the epiphyses. Therefore, we believe that the use of telescopic intramedullary nailing in OI is feasible only if children are older than 10 years of age when the period of their intensive growth terminates.

CONCLUSION

The technique of transphyseal elastic reinforcement with the titanium nails can be indicated to correct deformities in children that suffer from severe OI types. Lower limb deformity elimination and a reinforced bone provide an increase in patients' mobility and improve their quality of life. Orthopaedic complications are not rare occasions after operative treatment in this group of patients but their timely correction enables to retain anatomical and functional outcomes achieved. The use of osteosynthsis with the Ilizarov apparatus in its minimal variant for 3 to 4 weeks after corrective osteotomies and intramedullary reinforcement provides an early verticalization of patients and complete weight bearing on the operated limb as well as help avoid several complications such as secondary osteoporosis and secondary rotation of bone fragments. The use of intramedullary implants with bioactive coating prevents migration of nails at long term. However, nails should not be used as telescopic constructs in children younger than 10 years old in order to avoid their locking in the bone marrow canal due to their osteointegration.

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REFERENCES

1. Sillence D., Senn A., Danks D.M. Genetic heterogeneity in osteogenesis imperfect. *J. Med. Genet*. 1979. Vol. 16, N 2. pp. 101-116
2. Cheung M.S., Glorieux F.H. Osteogenesis imperfecta: update on presentation and management. *Rev. Endocr. Metab. Disord*. 2008. Vol. 9, N 2. pp. 153-160
3. van Dijk F.S., Pals G., van Rijn R.R., Nikkels P.G., Cobben J.M. Classification of Osteogenesis Imperfecta revisited, *Eur. J. Med. Genet*. 2010. Vol. 53, N 1. pp. 1-5
4. Glorieux F.H. Osteogenesis imperfecta. *Best Pract. Res. Clin. Rheumatol*. 2008. Vol. 22, N 1. pp. 85-100
5. Michelle C., Patel V., Amirfeyz R. et al. Osteogenesis imperfecta. *Curr. Orthop*. 2007. Vol. 21. pp. 236-241
6. Rauch F., Glorieux F.H. Osteogenesis imperfecta. *Lancet*. 2004. N 363 (9418). pp. 1377-1385
7. Engelbert R.H., Uiterwaal C.S., Gulmans V.A., Pruijs H., Helders P.J. Osteogenesis imperfecta in childhood: prognosis for walking. *J. Pediatr*. 2000. Vol. 137, N 3. pp. 397-402
8. Binder H. Rehabilitation of infants with osteogenesis imperfect. *Connect. Tissue Res*. 1995. Vol. 31, N 4. pp. S37-S39
9. Land C.. Rauch F., Montpetit K., Ruck-Gibis J., Glorieux F.H. Effect of intravenous pamidronate therapy on functional abilities and level of ambulation in children with osteogenesis imperfect, *J. Pediatr*. 2006. Vol. 148, N 4. pp. 456-460
10. Zionts L.E., Nash J.P., Rude R., Ross T., Stott N.S. Bone mineral density in children with mild osteogenesis imperfecta. *J. Bone Joint Surg. Br*. 1995. Vol. 77, N 1. pp. 143-147
11. Ruck-Gibis J., Plotkin H., Hanley J., Wood-Dauphinee S. Reliability of the gross motor function measure for children with osteogenesis imperfect. *Pediatr. Phys. Ther*. 2001. Vol. 13, N 1. pp. 10-17
12. Russell D.J., Rosenbaum P.L., Cadman D.T., Gowland C., Hardy S., Jarvis S. The gross motor function measure: a means to evaluate the effects of physical therapy. *Dev. Med. Child. Neurol.* 1989. Vol. 31, N 3. pp. 341-352
13. Haley S.M., Coster W.J., Ludlow L.H., Haltiwanger J.T., Andrellos P.J. Pediatric Evaluation of Disability Inventory (PEDI): Development, Standardization and Administration Manual. Boston, MA: New England Medical Center Hospitals, Inc. and PEDI Research Group. 1992
14. Porat S., Heller E., Seidman D.S., Meyer S. Functional results of operation in osteogenesis imperfecta: elongating rods and nonelongating rods. *J. Pediatr. Orthop*. 1991. Vol. 11, N 2. pp. 200-203
15. Engelbert R.H., Helders P.J., Keessen W., Pruijs H.E., Gooskens R.H. Intramedullary rodding in type III osteogenesis imperfecta. Effects on neuromotor development in 10 children. *Acta Orthop. Scand*. 1995. Vol. 66, N 4. pp. 361-364
16. Dogba M.J., Rauch F., Wong T., Ruck J., Glorieux F.H., Bedos C. From pediatric to adult care: strategic evaluation of a transition program for patients with osteogenesis imperfect. *BMC Health Serv. Res*. 2014. Vol. 14. p. 489. doi: 10.1186/s12913-014-0489-1
17. Montpetit K., Dahan-Oliel N., Ruck-Gibis J., Fassier F., Rauch F., Glorieux F. Activities and participation in young adults with osteogenesis imperfect. *J. Pediatr. Rehabil. Med*. 2011. Vol. 4, N 1. pp. 13-22
18. Montpetit K., Palomo T., Glorieux F.H., Fassier F., Rauch F. Multidisciplinary Treatment of Severe Osteogenesis Imperfecta: Functional Outcomes at Skeletal Maturity. *Arch. Phys. Med. Rehabil*. 2015. Vol. 96, N 10. pp. 1834-1839
19. Volkov M.V. *Vrozhdennye zabolevaniia kostnoi tkani u detei* [Congenital diseases of bone tissue in children]. M.: Meditsina, 1985, 487 p.
20. Cole W.G. Early surgical management of severe forms of osteogenesis imperfect. *Am. J. Med. Genet*. 1993. Vol. 45, N 2. pp. 270-274
21. Zeitlin L., Fassier F., Glorieux F.H. Modern approach to children with osteogenesis imperfect. *J. Pediatr. Orthop*. 2003. Vol. 12, N 2. pp. 77-87
22. Fassier F., Sardar Z., Aarabi M., Odent T., Haque T., Hamdy R. Results and complications of a surgical technique for correction of coxa vara in children with osteopenic bones. *J. Pediatr. Orthop*. 2008. Vol. 28, N 8. pp. 799-805
23. Aarabi M., Rauch F., Hamdy R.C., Fassier F. High prevalence of coxa vara in patients with severe osteogenesis imperfect. *J. Pediatr. Orthop*. 2006. Vol. 26, N 1. pp. 24-28

24.Bailey R.W., Dubow H.I. Evolution of the concept of an extensible nail accomodating to normal longitudinal bone growth: clinical considerations and implications. *Clin. Orthop. Relat. Res*. 1981. N 159. pp. 157-170
25.Boutaud B., Laville J.M. Elastic sliding central medullary nailing with osteogenesis imperfecta. Fourteen cases at eight years follow-up. *Rev. Chir. Orthop. Reparatrice Appar. Mot.* 2004. Vol. 90, N 4. pp. 304-311
26.McHale K.A., Tenuta J.J., Tosi L.L., McKay D.W. Percutaneous intramedullary fixation of long bone deformity in sever osteogenesis imperfect. *Clin. Orthop. Relat. Res*. 1994. N 305. pp. 242-248.
27.Stockley I., Bell M.J., Sharrard W.J. The role of expanding intramedullary rods in osteogenesis imperfecta. *J. Bone Joint Surg. Br.* 1989. Vol. 71, N 3. pp. 422-427
28.Esposito P., Plotkin H. Surgical treatment of osteogenesis imperfecta: current concepts. *Curr Opin. Pediatr*. 2008. Vol. 20, N 1. pp. 52-57
29.Lascombes P. Flexible intramedullary nailing in children. The Nancy University Manual. Berlin, Heidelberg: Springer-Verlag, 2010
30.Metaizeau J.P. Sliding centro-medullary nailing. Application to the treatment of severe forms of osteogenesis imperfect. *Chir. Pediatr*. 1987. Vol. 28, N 4-5. pp. 240-243
31.Novacheck T.F., Stout J.L., Tervo R. Reliability and validity of the Gillette Functional Assessment Questionnaire as an outcome measure in children with walking disabilities. *J. Pediatr. Orthop*. 2000. Vol. 20, N 1. pp. 75-81
32.Popkov D.A., Kononovich N.A., Mingazov E.R., Shutov R.B., Barbier D. Intramedullary Elastic Transphyseal Tibial Osteosynthesis and Its Effect on Segmental Growth. *Vestn. Ross. Akad. Med. Nauk*. 2015. N 4. pp. 441-449
33.Ruck J., Dahan-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. Vol. 5, N 3. pp. 217-224
34.Fassier F., Glorieux F. Osteogeneis imperfecta in the child. In: Cahiers d'enseignement de la SOFCOT. Paris:Expansion Scientifique Française, 1999. pp. 235-252
35.Zionts L.E., Ebramzadeh E., Stott N.S. Complications in the use of the Bailey-Dubow extensible wire. *Clin. Orthop. Relat. Res*. 1998. N 348. pp. 186-195
36.Larson T., Brighton B., Esposito P. et al. High reoperation rate and failed expansion in lower extremity expandable rods in osteogenesis imperfecta. In: Proceedings of the Annual Meeting of the Pediatric Orthopaedic Society of North America (POSNA). Waikoloa, Hawaii, 2010
37.Bilsel N., Beyzadeoglu T., Kafadar A. Application of Bailey-Dubow rods in the treatment of Osteogenesis Imperfecta. *Eur. J. Orthop. Surg. Traumatol.* 2000. Vol. 10, N 3. pp 183-187
38.Sułko J., Radło W. Operative management of long-bone of the upper limb in children with osteogenesis imperfect. *Chir. Narzadow Ruchu Ortop. Pol.* 2005. Vol. 70, N 3. pp. 195-199
39.Zeitlin L., Fassier F., Glorieux F.H. Modern approach to children with osteogeneis imperfecta. *J. Pediatr. Orthop.* 2003. Vol. 12, N 2. pp. 77-87
40.Popkov A. Bioactive implants in reconstructive othopedics. In: Combined stimulating methods in reconstructive surgery in pediatric orthopedics. Ed. D. Popkov. New York: Nova Science Publishers Inc., 2015. pp. 81-108
41.Popkov A., Aranovich A., Popkov D. Results of deformity correction in children with X-linked hereditary hypophosphatemic rickets by external fixation or combined technique. *Int. Orthop*. 2015. Vol. 39, N 12. pp. 2423-2431

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