Elongation of Long Bones for Short Stature in Patients with Hypophosphatemic Rickets

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The orthopedic manifestations of hypophosphatemic rickets are short stature in childhood and osteomalacia in adulthood, coxa vara with anterior and lateral bowing of the femurs, genu valgum or varum, and tibial torsion. Medical treatment is based on high doses of vitamin D3 and phosphate. Appropriate treatment does not always prevent skeletal deformation, hence the necessity of surgical orthopedic treatment. Surgical treatment may be problematic, however, due to disturbed bone metabolism that could cause delayed union or even non-union at the osteotomy site [1].

We present our experience with elongation in two siblings, with a relatively long-term follow-up. The history of five affected family members revealed mostly favorable results: four required corrective osteotomies, and the outcome was poor in one. To the best of our knowledge, there is no previous report on bone elongation for short stature in hypophosphatemic rickets.

Patients and Methods

Patient 1

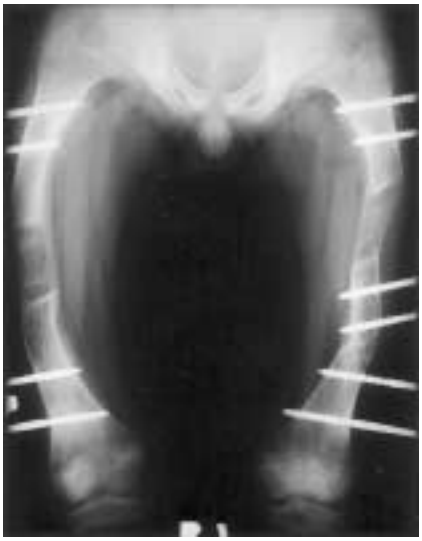
M.G., 23 years old, underwent her first surgical procedure of bilateral supramalleolar derotation valgus osteotomy to treat bilateral tibia vara at 3 years of age. Postoperative follow-up was uneventful and the osteotomies healed well. Physical development was normal, except for short stature and genu varum.

At age 14 she was 130 cm in height, with mild genu varum and normal range of movements in the lower extremities. She was offered an elongation procedure since no significant further growth was expected and her cooperation probability was high. At this time, the authors were already markedly experienced with elongation for short stature in patients with achondroplasia [2,3]. The patient underwent bilateral femoral osteotomies that were fixed with a Wagner external fixator. Gradual distraction achieved total lengthening of 10 cm. The external fixators were removed simultaneously after 13 months. One month thereafter, she broke her left femur during physiotherapy and was treated conservatively with a spica plaster cast for 6 weeks. Currently, 10 years after surgery, her height is 147 cm and her body proportions are normal. She works as an accountant and is extremely satisfied.

Patient 2

S.G., 18 years old, the brother of M.G., underwent bilateral femoral elongation at age 12. Preoperatively he had a windswept deformity with right genu valgum and left genu varum, necessitating double femoral osteotomies on the left side to correct femoral alignment. This "shishlik" (skewer-like) bone was fixed by a modified Wagner external fixator with three sets of double pins [Figure].

The postoperative course was markedly affected by his lack of cooperation. His tolerance of pain was low and this and his general attitude hampered physiotherapy. Marked limitation in range of knee motion, as well as his mental condition mandated cessation of elongation after distraction of 5 cm. The osteotomies and elongation site healed well and the external fixators were removed after 11 months. Gradually, his preoperative windswept deformity recurred. Three years later his right femur was operated on to correct axis deviation. He underwent double femoral osteotomy, which was fixed with retrograde intramedullary nailing introduced from the knee. The postoperative period was without any complications and again the osteotomy healed. He had undergone a similar corrective osteotomy of the left femur a year later and this osteotomy also healed well.



X-ray of S.G. 7 months post-surgery for elongation of both femurs and correction of axis deviation on the left. Wagner external fixator was used for the right femur, and the modified Wagner with three sets of Schantz screws and double osteotomy were used on the left. Good bone has formed at the elongation sites.

Comment

Since 1974 we have used the Callotasis principal with Wagner's external fixator. The elongation rate was 1 mm per day (0.25 cm four times a day). However, occasionally it was necessary to stop the distraction for a few days because of a decreasing range of movement in the knee joints. The external fixator was left *in situ* until radiographic union was established, with reformation of the intramedullary canal. The healing index

of about 30–40 days per cm was in accordance with other reports (about 1 month of healing for each centimeter of elongation) [4].

In our experience, the common indications for bilateral elongation of long bones are congenital short stature and achondroplasia. No report was found on elongation in metabolic bone disease in general, and on hypophosphatemic rickets specifically.

Theoretically, there are certain reasons to hesitate before elongating bones in metabolic bone diseases. Firstly, disturbance in bone metabolism may cause delayed or even non-union of fractures, osteotomies and other bone operation. This is even more problematic since in elongation, especially by callotasis, the osteogenic ability is already overused [1,5]. Second, the quality of the new bone formation could be low, due to disturbance of bone metabolism and to osteogenesis, thus increasing the risk of refracture and deformities [1].

A significant number of complications occurred in our patients. The sister's femur was refractured during physiotherapy 1 month after the external fixator was removed. Her brother had joint contractures, pin tract infection, severe mal-union, and did not achieve the planned length. This can be partially attributed to his treatment non-compliance, secondary to his immature personality of which we were unaware despite preoperative psychological evaluation.

We had an opportunity to examine these children's mother and through her we obtained information regarding other family members. The mother, who was 43 years old at the time of examination, was 145 cm in height with proportional limb length and normal range of movements in her upper and lower extremities. All other five affected family members had undergone corrective femoral osteotomies in childhood and were treated with a spica plaster cast. According to the mother, the grandmother and great-grandmother were satisfied with the results and had a normal life. Her sister had pain after surgery which gradually subsided, and her brother is confined to a wheelchair. Needless to say, all were significantly short.

To summarize, we have reported on two patients suffering from hypophosphatemic rickets who had bilateral femoral elongation procedure using a Wagner external fixator. The final results were good in the sister and poor in her brother. A history of an additional five affected family members revealed good results with corrective osteotomies, with the exception of one poor result. From our experience, although metabolic bone disease is not a common indication for elongation and could be problematic, it is not contraindicated. The key to success, as in other cases of elongation, is good patient selection and compliance, and an excellent surgical technique.

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