Case 1: 5-Year-Old Amish Boy with Impending Tibial Fracture

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Expert Panel: In-Ho Choi MD, PhD (IC); Charles Johnston, MD (CJ); Hitesh Shah MBBS, MS (HS)
Moderator: Jennifer C. Laine MD (JL)

Brief History: This is a 5-year-old Amish male with an underlying diagnosis of neurofibromatosis. He has a known right fibular pseudarthrosis and dysplastic but intact right tibia. He has been protecting the leg in a clamshell ankle foot orthosis (AFO) brace since age 2 but recently started having more pain in the right leg. He can ambulate independently but limps and cannot walk long distances anymore. He has not had any prior surgery. Leg length discrepancy is 26 mm (left > right).

Decision Point #1—Expert Panel

JL: Professors Choi, Shah, and Johnston, at this point, what are the key problems that should be considered for this patient?

IC: An impending fracture at the junctional area of the dysplastic mid-diaphysis has a strong tendency to convert to an atrophic-type CPT, particularly when marked anterior bowing of the tibia is seen with established fibular pseudarthrosis. This means that prompt surgical intervention is needed at this point of time without too much delay.

HS and CJ:
- Impending fracture of the lower third tibia leading to a non-functional extremity with a limb length discrepancy
- Secondary deformity of the ankle due to pseudarthrosis of fibula which produces valgus deformity which will eventually produce its own separate problems

JL: What are your treatment concerns specific for this patient?

IC: The treatment goals of CPT are osteosynthesis, correction of any angular deformity, stabilization of the ankle mortise, and lower limb-length equalization. Each of these goals is difficult to accomplish regardless of the surgical technique utilized. Potential issues and concerns in this particular case are as follows:
- How to deal with dysplastic segment of the tibia.
- How to manage established fibular pseudarthrosis with its distal segment being already proximally migrated to some extent.
- How to deal with 26 mm of leg-length shortening. Can we achieve successful osteosynthesis distally and length-gain proximally at the same time if residual shortening is deemed greater than 3 cm after surgical realignment?
**HC and CJ:** The sagittal plane is another concern for fixation with IM nailing.

**JL:** If this were your patient, what would you recommend and why?

**CJ:** This boy needs a functional limb for ambulation, activities of daily living—especially for life on the farm. I would recommend:

- **Step 1.** Apply frame to correct anterior bow without open surgery. Center of rotation of angulation (CORA) should be at the apex of bow. Expect that the pseudoarthrosis/impending fracture can be re-aligned closed, gaining length in the process. May induce some consolidation.
- **Step 2.** Once alignment and length gained:
  - **a.** Remove frame and observe/brace. Give time to allow pin sites to heal to minimize chance of intramedullary sepsis before open reconstruction below.
  - **b.** Resect bone: the amount of “diseased” bone may be relatively little and limited to the immediate bone ends at the pseudoarthrosis site. For the tibia IM fixation, we like antegrade flexible nail from lateral proximal tibial metaphysis and retrograde flexible nail from medial malleolus up to proximal tibial epiphysis and may provide better internal splinting than a single Fassier-Duval rod, for example. For the fibula, shorten and use thin rod/wire. Then attempt cross-union between tibia and fibula using iliac crest bone graft (ICBG) and BMP-soaked sponge.

**IC:** I would recommend a similar approach with a modification of the Ilizarov method coupled with intramedullary nailing. The Ilizarov method is certainly valuable for the treatment of CPT because it can address not only pseudoarthrosis but also all complex deformities associated with this condition. The basic biological considerations are the same as other methods, e.g., Charnley-Williams method of intramedullary fixation, microvascular transfer of a fibular graft, and the Masquelet technique—meticulous excision of hamartoma tissue, pseudoarthrosis resection, biological bone bridging of the defect by stable fixation, and the correction of any angular deformity.

In this particular case, I will use my modified Ilizarov technique, named “4-in-1 osteosynthesis,” in which all four proximal and distal tibial and fibular segments are put into one healing mass in combination with IM nailing. This is primarily considered for bony union with a large cross-sectional area and ankle stabilization.

A “4-in-1 osteosynthesis” seems to have several advantages as it (a) maximizes the cross-sectional area of healing at the pseudoarthrosis level, (b) facilitates bony healing owing to autogenous bone grafting over a wide area, (c) provides ankle stabilization and prevents proximal migration of the distal segment of the fibula, which causes ankle valgus, and (d) preserves ankle mobility. As a result, refracture risk will be markedly reduced.

**HS:** I agree with the prior opinions. The recommendations are for the union of the distal tibia and fibula and to prevent the progression of the deformity. The cortical bone graft resists the osteoclastic resorption.

**Treatment for Case 1**

At this point, the problem list included diagnosis of neurofibromatosis, right fibular pseudoarthrosis, impending right tibial fracture with anterolateral bowing of the right tibia, and leg length discrepancy of 26 mm.

The family decided to have reconstructive surgery. The plan was to excise the diseased tibial bone and surrounding periostem and transport the healthy proximal bone to the healthy distal bone. The patient received a zoledronic acid infusion 2 weeks prior to surgery.

The fibular pseudoarthrosis and abnormal periostem at the site were resected. The fibula was shortened and fixed with a 1.8 mm Ilizarov wire. A 5 cm section of the dysplastic diaphyseal tibia and surrounding periostem were excised (Figure 2). A 4-ring circular external fixator (three rings on the tibia and one on the foot) was applied to the tibia and foot using only Ilizarov wires. A proximal tibial osteotomy just below the tibial tubercle was performed. An intramedullary rod (Ilizarov wire) was placed in the tibia to guide the transport (Figure 3).

He was started on a rate and rhythm of 1 mm per day in four 0.25-mm increments on postoperative day 8. The rate and rhythm were adjusted on a weekly basis based
on the appearance of the regenerate bone between 0.5 mm per day and 1 mm per day (Figure 4). At 3 months after the index surgery, he was taken back to the operating room for bone grafting of the tibial docking site (Figure 5). Autograft was harvested from the left hemipelvis and packed around the fibula, the tibia, and the space in between the bones. The tibial wire was removed. No new wires were added and none of the initial wires needed to be removed at this procedure.

He continued compressing the docking site at 0.25 mm per day for the next 8 weeks. He was allowed to be weight-bearing as tolerated. A second zoledronic acid infusion was performed at 4 months after the index surgery.

At 7 months after the index surgery, he was walking independently with a walker. It was felt that he was ready for frame removal. He was placed in a short leg walking cast and then transitioned to an anterior clamshell AFO 6 weeks after frame removal (Figure 6).

At most recent follow-up 2 years after surgery, he is fully ambulatory without pain. Standing radiograph demonstrates residual leg length discrepancy of 14 mm and mild right knee valgus (Figure 7).

**Presenter Commentary**

**JL:** Dr. Iobst, thank you for sharing this complex case involving circular external fixation. Do you have any specific tips that you would like to share as you reflect on this case?

**CI:**
- I was taught a while ago only to use wires in the external fixator frame construct for patients with CPT. I did not use any half pins in the frame for this patient. The wire holes became enlarged over time (see Figure 6). Imagine what would have happened if a half pin had been used. The residual hole would have been even more at risk for fracture after removal.
- There was not a lot of access to space to pack bone graft between the two rings when approaching the end of transport.

**JL:** If you could treat this patient over again, would you do anything differently?
CI:
- I aggressively excised the entire portion of bone that I felt was dysplastic. Perhaps I could have removed less which would have made the transport distance smaller (or allowed for an acute shortening). It is hard to know if the dysplastic bone would have healed as well or not.
- Using a temporary wire in the tibia was theoretically helpful for guiding the transport, but its plantar exit made it impossible for the patient to bear weight. I would probably have avoided this wire and used a SLIM™ rod (Pega Medical, Laval, Canada) instead. I am not sure if the SLIM™ rod was available at the time of surgery. That is definitely something I would have done differently.

Figure 5a and 5b. Anteroposterior and lateral views after bone grafting the docking site, the fibular pseudarthrosis site, and the interosseous space.

Figure 6a and 6b. Anteroposterior and lateral views 6 weeks after fixator removal.

Figure 7. Standing AP bilateral lower extremity radiograph demonstrating improvement of the leg length discrepancy with well-healed regenerate bone and docking sites.