Case 5: 10-Year-Old Boy with CPT and Failed External Fixation

*Presenter: John E. Herzenberg, MD **(JEH)**; Philip K. McClure, MD **(PM)***
*Expert Panel: Christopher Iobst, MD **(CI)**; Kenneth Noonan, MD, MHCDS **(KN)**; Hitesh Shah, MBBS, MS **(HS)***
*Moderator: Jennifer C. Laine, MD **(JL)***

Brief History: A 9+6 year-old boy presented to clinic with neurofibromatosis type 1 and congenital pseudarthrosis of his left tibia and fibula. He walked with an ankle foot orthosis (AFO) and a walker. Pain was a level 5 of 10 if he was being active. He was able to swim.

Past Medical and Surgical History: He had a history of a distal left fibular fracture at 4 years of age that did not heal despite casting. At age 7, he had a left tibia fracture that also did not heal with immobilization. At age 8, he was treated at an outside hospital with a circular external fixator for 10 months but remained unhealed (Figure 1). That treatment was complicated by local pin infections and pain.

Physical Exam: He had an obvious flail nonunion with an anterolateral angulation of the left tibia/fibula and a clinical leg length discrepancy of about 5 cm. Femurs were equal in length. He was neurologically intact, and both DP and PT pulses were present. The child was withdrawn, made no eye contact, and refused to answer any questions. The mother explained that he was traumatized by the prior treatment, and as a result, he did not interact in medical settings.

Radiographs: Demonstrated a 60 degree anterolateral tibial bow; a separate, proximal metadiaphyseal anterior tibial bow; and an atrophic nonunion with a distal bone fragment spike and distal osteopenia. Bone-age radiographs was 12.5 years and limb length discrepancy (LLD) was 5.5 cm. (Figure 2)

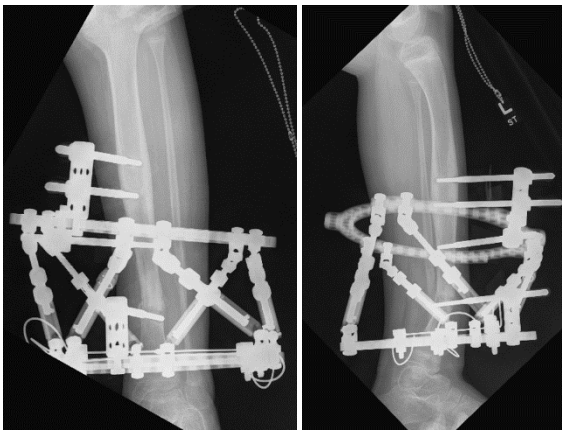

Figure 1. *Previous treatment. Anteroposterior (AP) and lateral views of the tibia with circular external fixator applied at an outside hospital for 10 months when the patient was 8 years old. Note external fixation in the nonunion site and absence of bone graft.*

Figure 2. *Radiographs at presentation. Standing radiograph of the lower extremities and lateral left tibia showing a nonunion of the left tibia and fibula, and about a 5.5 cm leg length discrepancy.*

Decision Point #1 – Expert Panel

JL: *Dr. Iobst, Dr. Noonan, and Professor Shah, what are the key problems and concerns for this patient? CI, KN, and HS all agree:*

- *Nonunion and deformity of the tibia and fibula*
- *History of external fixation with some questionable pin loosening at the level of the pseudarthrosis with concern for possible indolent infection/septic nonunion*
- *5 cm LLD*
- *Poor quality bone*
- *Calcaneal deformity*
- *Deep psychological impact to the patient*

Concerns include poor bone quality, high chances of recurrence of infection, difficulty to hold the proximal fragment and distal fragments. Overall, pain, infection, and psychological impact beyond nonunion of tibia and fibula are key.

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

KN: *I think that he has three potential options and would require extensive counseling and shared decision-making:*

1) Work up for infection and treatment if indicated (biopsy of pseudarthrosis and blood work). This could be followed by vascularized fibular transfer and open reduction internal fixation. Correction of LLD via combinations of contralateral distal femoral and proximal tibia growth arrest, ipsilateral femoral lengthening with Precice® nail (NuVasive, San Diego, CA).

2) Work up for infection and treatment if indicated (biopsy of pseudarthrosis and blood work). Osteotomy of proximal tibia and intramedullary (IM) fixation of the tibia/fibula with resection of pseudarthrosis and tibia-fibula cross-union with iliac crest bone graft (ICBG) and BMP-2 sponges. Would likely plate the proximal osteotomy site and pseudarthrosis site. Later correction of LLD via combinations of contralateral distal femoral and proximal tibia growth arrest, ipsilateral femoral lengthening with Precice® nail.

3) Amputation and prosthetic fitting

HS: *I would need to find out the source of pain, rule out infection, rule out deep psychological impact, consultation of clinical psychology. I would obtain complete blood count, ESR and CRP, bone scan, and CT scan to rule out infection.*

If there is no infection and the patient is ready for surgery, I would recommend: excision of pseudarthrosis, removal of hamartoma and diseased periosteum, intramedullary fixation with external fixation with Ilizarov, with cortico-cancellous grafting with the cross-union between tibia fibula.

The intramedullary fixation will prevent the possibility of re-fracture. External fixation is recommended to provide rotation stability and correct limb length. The cortical graft will prevent early resorption of bone graft. Cross-union will provide stability and enhance the chances of the union.

CI: *I would recommend:*

- *Debridement of the nonunion sites with removal of all abnormal periosteum*
- *Acute shortening of tibia/fibula*
- *SLIM™ nail (Pega Medical, Laval, Canada) or Fassier-Duval (FD) rod in tibia with Ilizarov wire in fibula*
- *Bone grafting (autogenous) to attempt to create cross-union between tibia and fibula*
- *Medical management with bisphosphonates (i.e., zoledronic acid)*

While treatment was recommended, the child was not emotionally ready for a surgical intervention at this time, so the family continued with the AFO and walker.

Follow-up visits at age 10 and 11+4 were made but the child still refused surgery. After intervention with a pediatrician/psychiatrist, the child was willing to consider surgery on condition that no external fixator would be used. At age 12, he underwent zoledronate infusion, followed one month later by the planned surgery.

Decision Point #2–Expert Panel

JL: *Dr. Iobst, Dr. Noonan and Professor Shah, would your approach change now that the patient is 12 years-old and refusing external fixation?*

HS: *The approach is the same even though the child is 12 years old. If the child does not want an external fixator, and infection is ruled out, we can use internal locking compression plate to provide rotational stability. Limb lengthening must be planned later to correct 5 cm shortening.*

CI*: There are now more options regarding internal fixation since he is closer to skeletal maturity and already has an LLD. Could potentially use a standard tibial nail for fixation of the nonunion instead of the FD rod or SLIM™ nail.*

KN: *Not really, I would not choose external fixation at this time anyway. I am glad that he has not presented with any signs or symptoms of infection.*

JL: *Any new concerns?*

CI*: I would stick with the same plan as outlined before (except the zoledronate infusion has already been given in this scenario.)*

KN: *He likely has a significant LLD that will require lengthening—a contralateral growth arrest will not recoup LLD due to his advanced skeletal age.*

HS: *With deep psychological impact, the child is delaying surgery for years. Staged surgery should be planned. The first surgery should be planned to achieve the union, and later surgery should be recommended to equalize limb length. The concerns are to explain two-staged surgeries.*

Treatment for Case 5

Through a 10 cm incision over the apex of the deformity, an extra-hamartomatous plane of dissection was especially tedious due to the fact that this was a revision case. The deep peroneal nerve and anterior tibial artery/vein were identified and retracted. We dissected circumferentially around the hamartoma surrounding the tibia and fibula. We then used sharp dissection and osteotomes to open up and mobilize the nonunions.

The hamartoma was then resected and the nonunions were dissected and mobilized. The two segments were then delivered into the wound and squared off with a small oscillating saw cooled with saline, perpendicular to the long axis of the bones. The fragments were allowed to overlap in the aligned position, with no tension on the posterior soft tissues, in order to gauge how much bone to resect at the nonunions. The fibula was reamed antegrade and retrograde with a 1.8 mm wire. The tibia was reamed antegrade and retrograde with a 1.8 mm wire followed by a 4.8 mm cannulated drill bit. In order to pass an antegrade nail, we had to perform an extension osteotomy of the upper tibia as there was a preexisting procurvatum deformity at the meta-diaphyseal junction. The tibial periosteum at this level was normal. We used a small oscillating saw to remove a small anterior based wedge and stabilized it with a small 4-hole plate at the upper end of the tibia. We then reduced the distal osteotomy and applied a 4.8 mm diameter SLIM™ nail, inserted antegrade in the tibia from a 2 cm parapatellar incision. Reaming was done with the tourniquet down to minimize heat. We then applied a small locking compression plate to the anterior medial surface of the distal tibia to control rotation. The fibula was fixed with a 1.8 mm intramedullary wire. A burr was used to roughen up the surfaces of the tibia and fibula for better incorporation of bone graft.

The right iliac crest was harvested as a tricortical cancellous bone graft, which was split with an oscillating saw into two cortical slabs. The volume of cancellous bone harvested was 45 cc.

The cortical bone plates were placed anteriorly and posteriorly between the tibia and fibula, after the method of Choi, with additional cancellous bone and BMP sandwiched between the plates. The surgical wound was closed over a medium suction drain, followed by a layered closure and splinting (Figures 3-6).

Figure 3. *Retrograde reaming showing the need for a proximal tibia osteotomy to straighten the procurvatum of the upper tibia.*

Figure 4. *Plate fixation is needed to control rotation after insertion of intramedullary rod.*

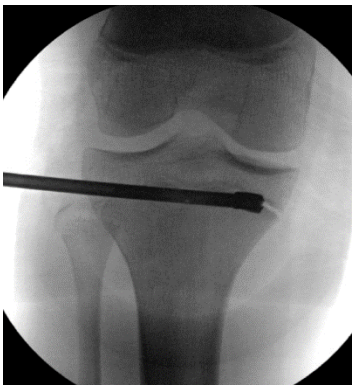

Figure 5. *Contralateral proximal tibial epiphysiodesis.*

Figure 6. *AP and lateral radiographs of left tibia immediately postop.*

The child was discharged on postop day 2. Subsequent visits at 1 week, 6 weeks, and 9 weeks were done, where gradual consolidation of the bone graft was noted. He underwent a second zoledronate infusion at 3 months postop.

By 3 months, he was observed to be fully healed at both osteotomies, with a 5 cm LLD, and allowed full weight-bearing (Figure 7).

Figure 7. *Three months postop showing progressive healing of the bone graft and synostosis.*

He continued to remodel the bone graft and synostosis (Figure 8).

Figure 8. *Seventeen months postop showing mature remodeling of the bone graft and synostosis.*

He underwent a second procedure 19 months after the open reduction internal fixation to remove the plates from the tibia (Figure 9). He remains fully active, using a hinged AFO and no other external support.

Figure 9. *Nineteen months after the ORIF procedure, the proximal and distal plates are removed.*

Presenter Commentary

JL: *Dr. Herzenberg, thank you for presenting this complex case which demonstrates many of the challenges orthopaedic surgeons face with this condition, including pseudarthrosis, deformity, leg length discrepancy, previous failed attempt at union, and the mental health toll this condition takes on its patients and families.*

In hindsight, if you could treat this patient again from the beginning, would you do anything differently?

JEH: *We would have preferred to receive him earlier in the course of his disease. The initial surgical treatment done at the outside hospital was inadequate and led to psychological trauma and delay in getting satisfactory treatment.*

We use a multimodal approach including bisphosphonate infusion pre-op and at 3-4 months postop, hamartoma resection, transverse osteotomy of the pencil tip ends for internal mechanical stability, autogenous iliac crest bone grafting with a cortical sandwich-style cross-union between tibial and fibula, BMP and cancellous autograft filling the sandwich, and intramedullary stabilization of tibia/fibula. We have also used meshed periosteal grafting from the iliac crest but find that this is less important than the other items listed.

Other authors have recommended the Fassier Duval telescopic rod for fixation of the tibia in CPT. We have observed many complications from this technique, including failure to slide, difficulty to remove the device when attempting to exchange, and bending of the smaller, male end of the device. Additionally, during the index surgery, which is quite complex and lengthy, fiddling with the telescopic mechanism and adding locking wires adds an additional level of unnecessary complexity. Therefore, we no longer recommend the Fassier Duval rod for the initial treatment of CPT. Instead, we use its cousin, the SLIM™ nail. As the tibia grows, the SLIM™ nail can be exchanged as needed. We find that this strategy is easier and less prone to complications, albeit at the cost of perhaps doing more exchanges than would be required if a Fassier Duval rod was used.