Case 2: 5-Year-Old Honduran Girl with Established Pseudarthrosis

*Presenter: Charles Johnston, MD **(CJ)***
*Expert Panel: Benjamin Joseph, MBBS, MS, MCh **(BJ)**; Joachim Horn, MD, PhD **(JH)***
*Moderator: Jennifer C. Laine MD **(JL)***

Brief History: A 5-year-old girl from Honduras presented for treatment of a deformed right lower extremity on which she had never been able to bear weight (Figure 1).

Past Medical History: She was known to have multiple café-au-lait spots but was otherwise healthy and had normal growth and development in spite of being unable to walk on her right leg. A "fracture" had occurred prior to walking age, with no treatment being available locally. She had undergone an unknown surgery in San Pedro Sula at age 2 but still had never been able to bear weight. We were informed that the patient lived in a marshland-like area of the Miskito Coast, where there was always wet ground or free-standing water such that most inhabitants dispensed with shoes. In this area, prosthetic limbs—especially feet—would likely not last long due to immersion, and prosthetic services are not available.

Physical examination: Exam was notable for 5 cm shortening and painless mobility of the pseudarthrosis site (Figure 2). The foot was smaller in size, but all motor function was intact. There were no other stigmata of Neurofibromatosis Type 1 (NF-1) beside the café-au-lait lesions.

Decision Point #1—Expert Panel

***JL:** Dr. Joseph and Dr. Horn, at this point, what are the key problems that need to be addressed for this patient?*

***BJ:** I like to list the problems for these complicated patients:*

- *Pseudarthrosis of tibia and fibula (achieving union is difficult)*

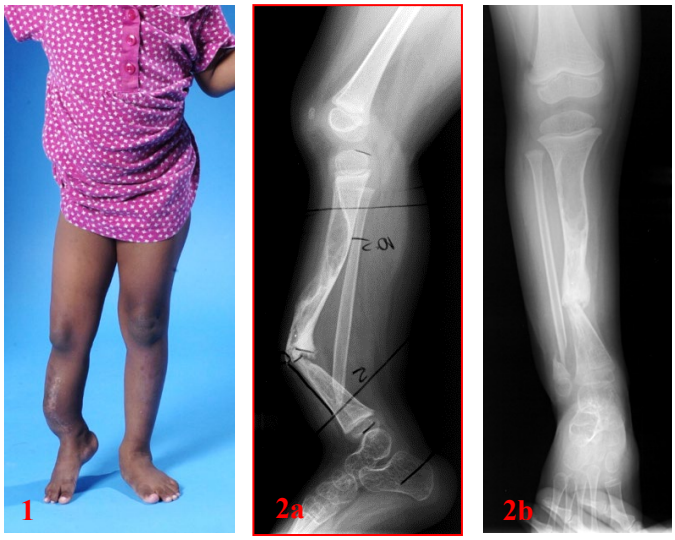

***Figure 1.** Clinical appearance, age 5. **Figure 2a, 2b.** Initial radiographs. Note large fibrous lesion in proximal tibia above the pseudarthrosis.*

- *Lesion extending into the proximal tibia from the pseudarthrosis*
- *Shortening (primarily in the tibia)*
- *High risk for re-fracture once union is obtained*
- *Propensity for ankle valgus*

***JH:** I agree. In addition, I am concerned about the recurvatum deformity in the proximal tibial diaphysis, the distal fibular pseudarthrosiss, and proximal migration.*

***JL:** What are your concerns for this particular patient?*

***JH:** Besides the tibial pseudarthrosis, the biggest concerns are the recurvatum deformity in the proximal tibial diaphysis, the large fibrous lesion in the proximal half of the tibia, the extensive sclerosis of the tibial bone in the diaphyseal area, the apparent leg length discrepancy*

(LLD). The living conditions in a marshland-like area of the Miskito Coast would be a concern.

BJ: *Union may not be achieved after one round of surgery, there is a high risk of re-fracture following union, and there is a high chance of ankle valgus developing.*

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

BJ:

Recommendation	**Reason**
Surgical intervention without further delay	Shortening is likely to increase as delay in obtaining union occurs
Excision of pseudarthrosis including the hamartomatous tissue	Essential in order to obtain union
Autogenous cortical bone grafting	Cortical bone tends to resist resorption
Attempt to obtain cross-union of the tibia and fibula in addition to union of tibia and fibula (four-in-one union)	Risk of re-fracture is minimized if four-in-one union is obtained
Intramedullary fixation placed centrally in the long axis of the tibia	The rod shares the load
Retain intramedullary (IM) rod until skeletal maturity	Minimizes risk of re-fracture
Clamshell orthosis until skeletal maturity	Minimizes the risk of re-fracture

JH: *I would recommend surgery to achieve union at the pseudarthrosis. The method of choice would be the X-union protocol as described by Paley. Treatment and surgery would include all steps as described by Paley. However, the method has to be modified for this particular patient. At the pseudarthrosis site, bone ends must be osteotomized with minimal bone resection.*

The recurvatum deformity in the proximal diaphyseal area and the large fibrous lesion represent challenges. The recurvatum deformity in the proximal shaft requires an osteotomy at the apex of the deformity in order to allow for IM rodding with a Fassier-Duval (FD) nail. After the osteotomy, the intermediate fragment at the midshaft (pseudarthrosis distal to fragment, osteotomy proximal to fragment) should under no circumstances lose all of its soft tissue attachment. To achieve this, circumferential excision of the periosteum at the pseudarthrosis site should not be extended proximally to the level of the osteotomy.

Furthermore, the osteotomy should not be "complete." Only the anterior part of the tibia, where the fibrous lesion is, should be osteotomized, and the posterior cortex should stay intact. The recurvatum deformity is then corrected by hinging at the posterior cortex (anterior opening wedge). During reaming for the FD rod, care must be taken to preserve the soft tissue attachment at the osteotomy site. Fibrous tissue should be removed and samples from the fibrous lesion should be sent to pathology examination. Reaming through the sclerotic diaphyseal area might be challenging—a 2.5 mm drill might be used to establish a canal before using FD reamers.

Another concern affecting the surgical technique is the fixation method. Intramedullary fixation with an FD rod should be used. However, the proximal extent of the fibrous lesion jeopardizes sufficient and stable fixation with an additional plate. I would therefore instead prefer fixation with an Ilizarov external fixator in addition to the FD rod. Two Ilizarov wires could be placed in the proximal fragment avoiding the area with the fibrous lesion and at the same time respecting the proximal tibial growth plate.

When cancellous bone is inserted between the tibia and fibula, care should be taken to harvest a large enough amount of bone so that the bone grafting covers the pseudarthrosis area, the osteotomy site, and the proximal area of the fibrous lesion.

Another concern is the relative shortening of the fibula. Depending on the degree of valgus instability, this might

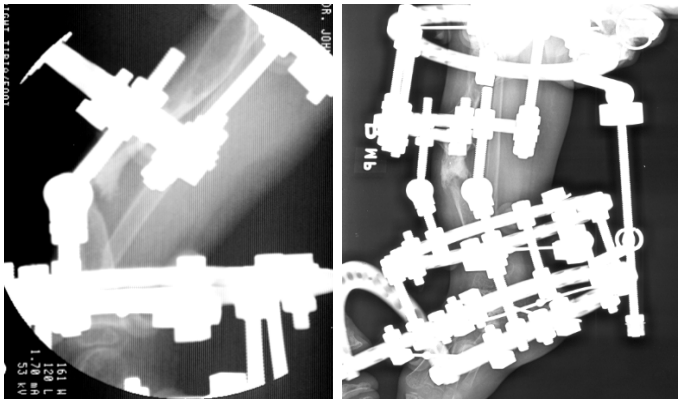

Figure 3a (left). *Date of surgery, frame application.* ***Figure 3b (right).*** *Ten days later, gradual correction of anterior bow.*

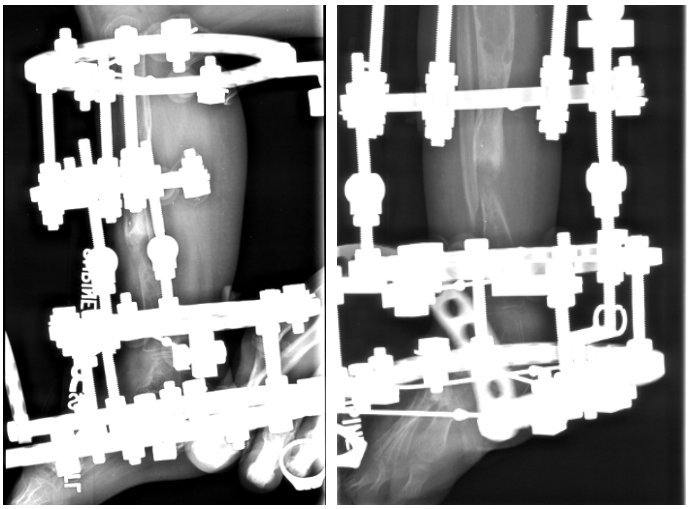

Figure 3c, 3d. *Lateral and anteroposterior (AP) radiographs after 22 days of gradual correction.*

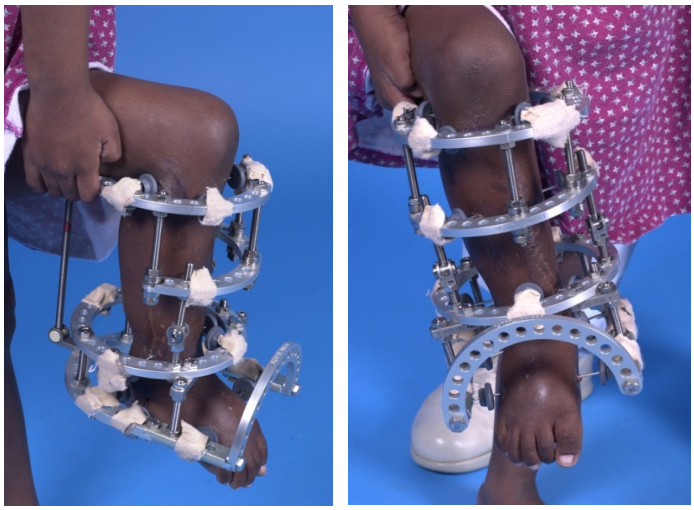

Figure 4. *Angular correction completed after slightly more than 3 weeks.*

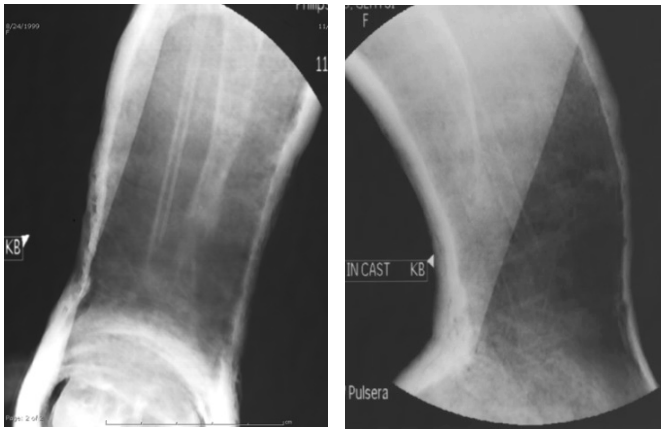

Figure 5. *Frame removal and long leg cast application for one month.*

be addressed (optional) by releasing the anterior and posterior tibio-fibular ligaments and slight shortening of the tibia relative to the fibula.

Treatment for Case 2

An external ring fixator was applied to gain gradual deformity correction with maximum length. No surgery was performed at the pseudarthrosis site. Proximally, one ring captured the segment with multiple wires through the non-ossified lesion with a floating half ring to locate the hinges at the pseudarthrosis level. Distally, the tibia segment and foot were captured using one circular ring and a foot stirrup with calcaneal and metatarsal wires. Hinges were placed over the anterior edge of the distal tibial fragment to produce slight distraction on the pseudarthrosis site as angular correction proceeded.

After slightly more than 3 weeks, angular correction was completed, the frame was removed, and the extremity was casted for 1 month to allow the pin sites to "heal" (Figures 3–5).

The definitive procedure then included resection of the hamartomatous tissue in the pseudarthrosis site and intralesional removal of fibrous tissue from the proximal lesion, which was later filled with autogenous iliac crest bone graft (ICBG). Fibular osteotomy with slight shortening was also performed, and IM fixation was achieved in the fibula. In the tibia, two titanium rods were utilized: one antegrade from the lateral proximal metaphysis, the second retrograde from the medial malleolus. ICBG was packed around the tibial site and then wrapped with BMP-soaked sponges (Figure 6).

Figure 6. *Day of surgery: resection of hamartomatous tissue, intralesional curettage of proximal metatphyseal defect, fibular osteotomy, IM rodding both bones, addition of ICBG to both pseudarthrosis site and proximal lesion.*

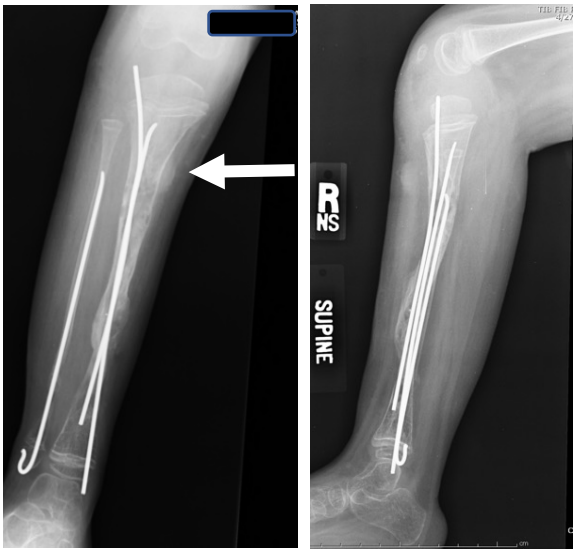

Figure 7. *At 18 weeks, the lateral rod tip has backed out proximally and is palpable, and there is a sinus medially (arrow) with purulence expressed when the rod tip was pushed back in with an impactor.*

Postoperatively, the patient was casted for 8 weeks. She developed good callus at the pseudarthrosis site and was transitioned to a clam shell fracture brace (ankle foot orthosis (AFO) with anterior shell for circumferential support) and allowed weight-bearing as tolerated. At 18 weeks postop (Figure 7), she re-presented with a small draining sinus on the medial side of her leg at a wire scar, and we also noted prominence of a rod tip proximo-laterally (Figure 7).

Decision Point #2—Expert Panel

JL: *Dr. Joseph and Dr. Horn, at this point, what are the key problems that need to be addressed for this patient?*

JH: *The primary concern at this point is, of course, the infection and the required treatment to solve this complication. In addition to the infection, other concerns are:*

- *Migration of the IM rod*
- *The fact that titanium implants are inserted across the peripheral part of the physis in the distal tibia and most likely in the proximal physis as well. Titanium provides a high degree of biocompatibility, and because of that, the risk of formation of a physeal bar might be greater than for steel implants crossing the physis (usually a "pseudomembrane" forms around steel)*
- *The relative shortening of the fibula*

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

BJ*:*

Recommendation	**Why**
Culture	To decide appropriate antibiotic
Remove the nail under the sinus and irrigate the track	To facilitate eradication of infection
Avoid weight-bearing till infection settles	To give rest and facilitate healing

JH: *I would recommend the following treatment steps:*

- *Removal of the titanium rods in the tibia*
- *Surgical debridement of the infection site, including the entire medullary canal (assuming deep infection affecting the medullary canal). For this purpose, surgical approach at the proximal shaft area where the fibrous lesion was located. If the canal is not accessible, a hole should be made (drill + chisel) big enough to allow reaming of the medullary canal in proximal and distal directions with flexible, very thin reamers. Injury of the proximal and distal tibial physis should be avoided. If standard reamers are too big, one might just use flexible blunt wires or whatever fits into the canal. A hole (5 mm) in the distal tibia, just above the physis, should be established to allow for irrigation of the medullary canal*

with several liters of fluid, where the irrigation fluid might be brought in at the proximal hole and flow out through the hole in the distal tibia. After debridement and irrigation, antibiotic-loaded bead chains might be placed in the medullary cavity for 2 weeks. If the earlier pseudarthrosis site is still healed and stable, a posterior splint can be used during the whole course of the treatment for the infection (6 weeks).

- *If there is any doubt concerning the pseudarthrosis site, an Ilizarov frame should be applied. Two olive wires very proximally and two wires very distally might be sufficient. Only touch-toe weight bearing allowed.*
- *When the patient is free of infection, at least 6 weeks after debridement and irrigation, an FD rod is inserted. In order to allow for insertion of the FD rod, a proximal osteotomy might be necessary. If there is any suspicion that the former pseudarthrosis area might have refractured during surgery, the Ilizarov should be kept in place in addition to the FD rod until healing is apparent. If refracture and no healing, X-UNION technique should be considered.*

Continued Treatment for Case 2

The patient returned to surgery where pus was expressed from the sinus tract. We therefore retrieved the lateral entry rod by pulling it proximally and followed the medial sinus tract to a discrete hole in the tibia, which was carefully enlarged and allowed the second (retrograde) rod to be grasped with a clamp and gently tapped distally until the rod tip could be retrieved at the medial malleolus and pulled out. Using a sterile brush with flexible handle, the intramedullary canal was sequentially "brushed" through the proximal fenestration, then irrigated with a small catheter, with egress from the medial malleolar rod hole. An intramedullary suction drain was then placed via the proximal fenestration and the leg splinted, although during the procedure, the stability of the callus and pseudarthrosis union to gentle stress was confirmed. Cultures grew MRSA, so intravenous vancomycin was administered for 21 days until the ESR and CRP were normalized.

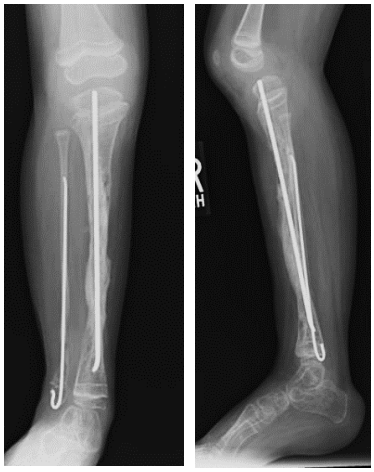

***Figure 8**. Three months following re-rodding and 6 weeks of antibiotic therapy. All wounds were healed, ESR was normal, and the patient returned home, weight-bearing as tolerated.*

***Figure 9.** Increasing valgus deformity age 8.*

The intramedullary fixation was then replaced using a single larger diameter titanium rod placed antegrade from an entry hole proximal to the tibial tubercle (Figure 8). Oral antibiotics were continued for three additional weeks. At 3 months after re-insertion of the IM rod, ESR was normal, and so the patient returned home to Honduras.

Eighteen months later, now age 8 years, she returned due to increasing valgus deformity (Figure 9). With the angular deformity, the apparent limb length difference was also notable. Although most of the estimated 6 cm discrepancy was in the right tibia, some overgrowth of the right (ipsilateral) femur was also noted.

Angular correction and length equalization were required. Although both could obviously be achieved with a frame, the patient flatly refused a second one.

Decision Point #3—Expert Panel

JL: *Dr. Joseph and Dr. Horn, at this point, what are the key problems that need to be addressed for this patient? What are your concerns for this patient?*

BJ and JH: *The main concerns are the amount of shortening and the severe valgus deformity taken into consideration her young age, the amount of remaining growth, and the possible formation of a physeal bar. Another concern is the patient's refusal to be treated by an external frame.*

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

BJ*:*

- *Guided growth of the proximal tibia*
- *3 cm shoe lift until timed contralateral epiphysiodesis to reduce the discrepancy to about 3 cm at skeletal maturity (because the patient will not tolerate an external fixator).*

JH: *First, I would attempt advanced imaging to see if there is a proximal tibial physeal bar.*

- *If no physeal bar is present, the angular deformity might be addressed by a medial hemiepiphysiodesis. If angular correction then is successful and the patient still refuses external fixation for lengthening, leg length discrepancy could be addressed by shoe augmentation and later proximal tibial (and possibly distal femur) epiphysiodesis. Epiphysiodesis might only allow reduction of LLD so that shoe augmentation still would be necessary.*
- *If a physeal bar is present, and assuming the patient still refuses an external fixator, removal of the bar according to the method as described by Langenskiold should be considered depending on the size of the bar. After the Langenskiold procedure, growth should be monitored. If the procedure is successful, some of the valgus deformity might correct spontaneously over time. Contralateral epiphysiodesis is an option to reduce LLD. Residual valgus deformity might be corrected by a medial closed wedge osteotomy and internal fixation close to the cessation of growth. Lengthening cannot be done if the patient refuses an external fixator, since the size of the medullary canal would not allow the use of internal lengthening devices.*

Continued Treatment for Case 2

Since she previous had intramedullary sepsis, which was still quiescent, we acquiesced in her decision to avoid external fixation and resorted to growth modulation and contralateral femoral epiphysiodesis, even though this would result in her knees being asymmetric in height due to the overgrowth already present in the ipsilateral femur.

Trans-physeal screws (PETS) were placed medially at both ends of the tibia, but by age 9, it was clear that the proximal tibial implant was ineffective (Figure 10a), and so was revised to an "8 plate" (Figure 10b).

Figure 10. *The proximal tibia epiphysis has grown off the trans-physeal screw (PETS) with minimal improvement in the proximal valgus (10a). An 8 plate was then substituted (10b).*

One year later, sufficient angular correction was achieved (Figure 11), indicating removal of the modulation implants. No rebound phenomenon occurred in follow-up.

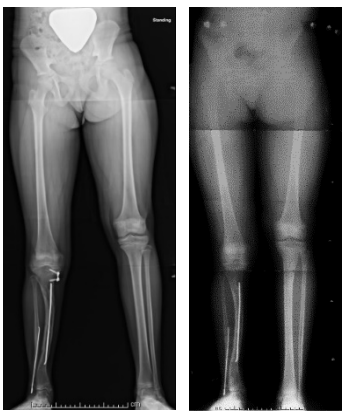

Figure 11. *Valgus correction at age 12 (11a left), which was maintained a year later after implant removal (11b right). Distal femoral epiphyseodesis on the contralateral side was also performed.*

Figure 12. *Most recent radiographs and clinical appearance, age 14.*

Functionally the patient was fully active without any external support, which she actually had discarded on her return to Honduras some 6 years earlier.

See Case 2 Outcome video of patient age 12.

She was able to run, albeit with a very modest cadence. At her last follow-up at skeletal maturity (age 14), her limb lengths were within 2 cm of being equal (Figure 12), and she was entering high school with no restrictions on activity.

Presenter Commentary

JL: *Dr. Johnston, thank you for sharing this challenging case, highlighting not only the orthopaedic complexity but the potential social complexity. Do you mind commenting on your initial decision-making?*

CJ: *I remember we thought she would have no access to prosthetics, so amputation as primary treatment was out. She had never walked on that leg, so primary amputation was the main consideration at first glance.*

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

CJ: *There is not much else I would have done differently considering what we had to work with. I might consider waiting longer after the frame came off to let the pin tracks mature and suppress with antibiotics longer, to hopefully prevent the infected IM rod. We could have done a fixator assisted nailing so that they would be done simultaneously, but I do not think we could have gotten complete acute correction due to the longstanding pseudarthrosis bow.*

JL: *Any additional pearls based on this case, or experience with other cases like it?*

CJ: *Unless there is a compelling patient or family philosophy pushing toward primary amputation, usually at least one attempt to gain union is appropriate. This is due to satisfactory long-term function of a CPT that gets truly healed on the first attempt with appropriate IM fixation remaining in place.*