Supplemental Material

JOURNAL OF CHILDREN'S ORTHOPAEDICS

Supplement to: Congenital pseudarthrosis of the tibia: biological and biomechanical considerations to achieve union and prevent refracture

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The Paley cross-union (X-UNION) surgical protocol

Evolution of the Paley X-UNION protocol

The author's first published report on congenital pseudarthrosis of the tibia (CPT) was in 1992.[1] Long-term follow-up of those and subsequently treated patients were first reported in El-Rosasy's doctorate thesis and demonstrated a high refracture and retreatment rate.[2] The initial treatment was resection of the hamartoma and bone grafting of the CPT site with Ilizarov apparatus for fixation. The healing rate was nearly 100% but the refracture rate was over 50%. In some cases an intramedullary rod (flexible titanium or Rush rod) was added to the Ilizarov-bone grafting treatment. The refracture rate dropped. Clearly the Ilizarov fixation method was excellent at obtaining union but failed to maintain union. The intramedullary (IM) rod was excellent at maintaining union and decreasing refracture. This was also the conclusion of the multicenter European Paediatric Orthopaedic Society (EPOS) study by Grill et al.[3] The efficacy of the IM rod was also increased by rodding both bones in the leg rather than just one.

The El-Rosasy study[4] also identified two other factors that significantly helped decrease refracture: 1) increasing the cross sectional area of union; and 2) eliminating angulation at the CPT site. After the El-Rosasy study the author combined all of these principles as well as added a periosteal graft. This was reported in a two-centre study by Thabet et al[5] (2008). Treatment involved resection of the hamartoma, iliac crest bone and periosteal grafting of the CPT site, and rodding of the tibia and fibula. Union was achieved in 100%. Refracture occurred in 8/20 (40%).

Obtaining union was easier and faster with this method but did not prevent refracture. We know from the principles and practice of treatment of adult nonunions, that successful treatment of atrophic nonunion, involves optimizing the mechanical and biologic environment of the pseudarthrosis site. Mechanical modalities include: correction of angular deformity, stability of fixation using external and/or internal fixation, increased bone width at the nonunion site, and reinforcing the bone strength with intramedullary fixation of the tibia and fibula. Biologic modalities include resection of surrounding or interposing fibrous tissue, resection of any hypovascular bone, and autogenous bone grafting. In Thabet et al[5] treatment of CPT had done all of these, with the addition of periosteal grafting. The bone graft was harvested with decancellousization of the ilium in many of these cases. The use of these mechanical and biologic modalities, lead to reliable CPT union in 100% of cases. It did not, however, prevent refracture despite the presence of tibial and fibular intramedullary rods.

What more could be done on both the biological and mechanical aspects of CPT to prevent refracture? Mechanically, union of the tibia was already achieved, the inside of the bone was already reinforced with an intramedullary rod in the tibia and fibula, and the union site width, was augmented by the bone graft. When reviewing the cases that did not refracture, in the Thabet et al[5] study, a cross-union to the fibula had developed in many of them. This stimulated the idea that we could increase the cross-sectional area of bone at the CPT site by intentionally creating a bony bridge between the tibia and fibula.

To achieve a cross-union, requires a large amount of autograft to be placed in between the tibia and fibula. This amount of autograft is not available from the anterior or posterior iliac crest of most young children. The only parts of the pelvis that contains such ample autogenous cancellous bone are the supra-acetabular region and the posterior iliac crest. To get to these regions surgically through one approach, one would need to split the iliac crest past the narrow bottle-neck of the thinnest part of the iliac wing. The author developed the method to do this in 2002 and was using it with the periosteal grafting cases previously discussed. At that time, it was only for bone grafting circumferentially around the tibia and fibula separately. As noted, this unintentionally generated a cross-union in several of those patients. By splitting the two tables of the ilium all the way to the sciatic

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notch, sacro-iliac joint, triradiate and acetabular dome, one can harvest bone from both the anterior and posterior aspects of the ilium, hollowing out the entire ilium down to the subchondral bone of the hip and triradiate cartilage, around the Gothic arch of the sciatic notch, to the cancellous bone of the posterior iliac crest as far as the subchondral bone of the sacro-iliac joint and posterior iliac spines. This is called decancellousization of the ilium. This method can yield as much as 20 cc of bone in a 12-month-old infant and up to 70 cc in a seven-year-old. With this large amount of cancellous bone graft, creating an intentional cross-union between the tibia and fibula is consistently possible as long as the bone graft is not resorbed. To prevent resorption zolidronic acid (ZA) is given two weeks prior to surgery. This allows the ZA to be taken up by the cancellous bone of the ilium. This makes the cancellous bone of the ilium non-resorbable. Finally, to boost the osteogenic response of this cancellous bone BMP2 can be used. The author started using BMP2 in 2002 and combined treatment with ZA with BMP2 in 2004. The concept is to down-regulate the excessive osteolysis with ZA and up-regulate the osteogenesis with BMP. The rationale for this is well explained by the later publications of Cho et al[6] 2008 and Schindler et al[6,7] 2008 and was discussed earlier. Cho et al[6] showed that the soft-tissue hamartoma around the CPT had excessive numbers of osteoclasts leading to an abnormal degree of osteolysis. They also showed that the response to BMP was more limited in neurofibromatosis (NF). These publications supported the combined use of BMP2 with ZA in a synergistic fashion.

In 2007, Paley[8] first combined all of these techniques to create an intentional cross-union between the tibia and the fibula. The treatment protocol was: presurgical infusion of ZA; hamartoma resection around tibia and fibula; tibial and fibular rodding; decancellousization of the ilium to harvest a large cancellous bone graft; harvest a periosteal graft from the underside of the iliacus muscle; apply a three-layer graft composed of periosteum around the CPT; cancellous bone between and around the tibia and fibula; BMP2 posterior and anterior to the bone graft covered by soft tissues. The last step was application of the Ilizarov apparatus. The Ilizarov apparatus was used to compress the CPT site and to give rotational stability. The smooth non-locking telescopic rod only gives angular support but does not prevent the bone ends pulling apart or rotating to each other. More recently (since 2014) the author replaced the external fixator with an internal fixator (locking plate).[9]

Paley X-UNION protocol surgical technique

Step 1: Preoperative biphosphonate infusion: one to two weeks prior to the surgery the patient is given a ZA infusion intravenously (0.02 mg/kg) over 30 minutes. One hour later calcium gluconate 60 mg/kg is given intravenously over the course of one hour. The patient is given 2 gm elemental calcium for seven days and Vitamin D supplementation of 1000 International Units (IU) for 14 days.

Step 2: Prep and incision: the patient is placed supine, with a bump under the ipsilateral buttock, on a radiolucent table. The entire lower extremity and hemipelvis are prepped and draped free. The leg is exsanguinated and tourniquet applied. The pseudarthrosis site is approached through an anterior longitudinal incision (Supplemental Fig. S1a).

Step 3: Skin elevation: the lateral skin and subcutaneous tissues are elevated off of the anterior compartment fascia. The medial skin and subcutaneous tissues are elevated off of the subcutaneous border of the tibia to the fascia of the deep posterior compartment (Supplemental Fig. S1b).

Step 4: Anterior fasciotomy: the anterior compartment fascia is incised longitudinally. Extend this fasciotomy subcutaneously proximally and distally (Supplemental Fig. S1c).

Step 5: Anterior compartment muscle elevation: the anterior compartment muscles including the anterior tibial vessels and deep peroneal nerve are gently elevated off of the lateral border of the tibia and off of the interosseous membrane until the medial wall of the fibula is exposed (Supplemental Fig. S1d).

Step 6: Posterior fasciotomy: the deep posterior compartment fascia is incised longitudinally (Supplemental Fig. S1e).

Step 7: Posterior compartment muscle elevation: the deep posterior compartment muscles are gently dissected off of the posterior hamartoma/periosteum around the tibia and off of the interosseous membrane (Supplemental Fig. S 1f).

Step 8: Interosseous membrane excision: incise the interosseous membrane on its tibial and fibular borders from anterior to posterior avoiding injury to the peroneal vessels. Excise the membrane in the entire region planned for the cross-union. Dissect around the fibular hamartoma circumferentially. Be careful to avoid injury to the peroneal artery as it passes from posterior to the membrane through to anterior to the membrane at the distal end of the interosseous space (Supplemental Fig. S1g).

Step 9: Hamartoma incision: longitudinally incise the hamartoma of the tibia anteriorly for the entire length of planned resection. Make sure that the incision of this tissue exceeds the region of thickened periosteum (Supplemental Fig. S1h).

Step 10: Circumferential excision of hamartoma: radially incise the periosteum proximally and distally to resect the hamartoma from the tibia (Supplemental Fig. S1i).

Step 11a: Excise hamartoma from fibula but do not release syndesmosis if fibula at station (type 1, 2a, 3, 4a)

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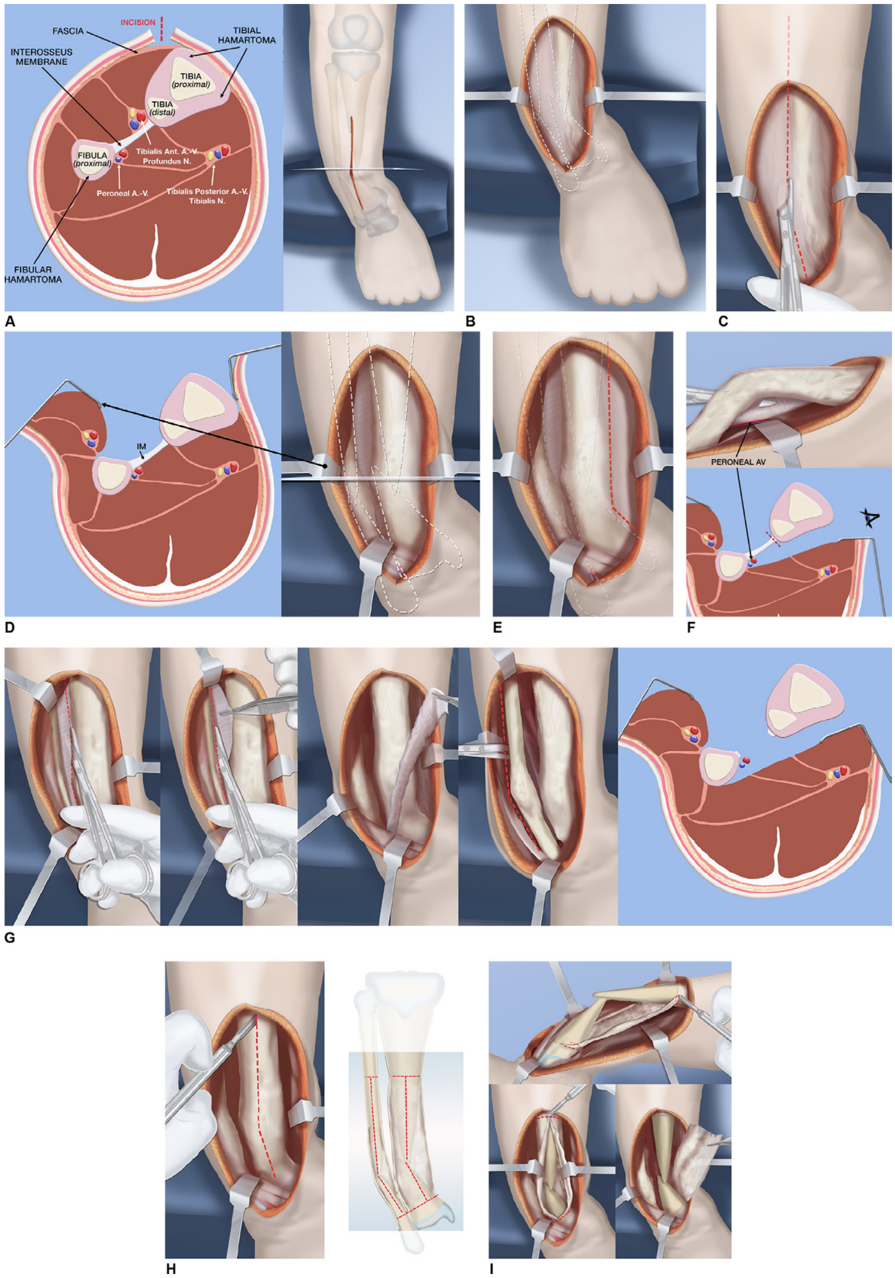
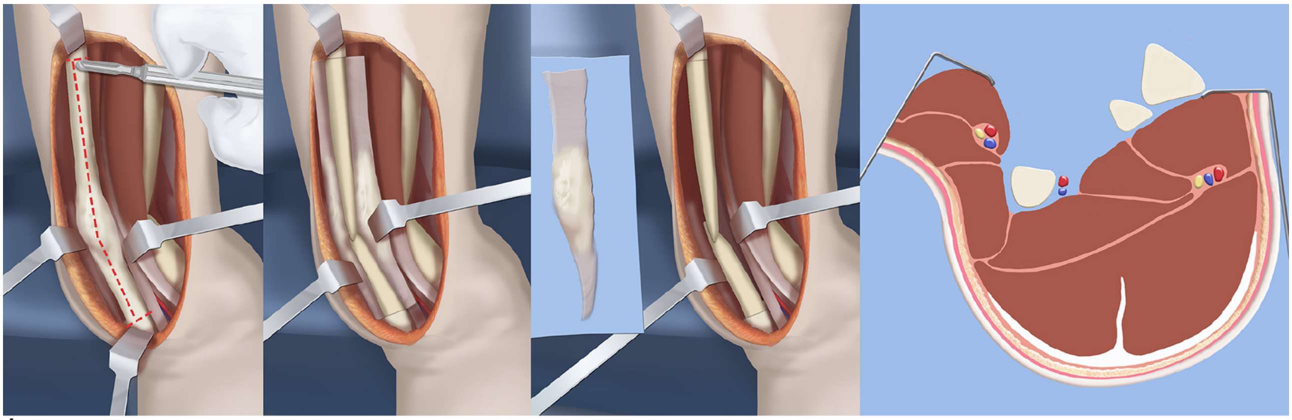


Supplemental Fig. S1 Reproduced with permission by the Paley Foundation: (**a**) incision (anterior view left and cross-section view right). Note the circumferential hamartoma surrounding the tibia and fibula on the cross section view. The anterior tibial and peroneal neurovascular bundles lie very close to this tissue and can be adherent to it; (**b**) anterior fascial exposure; (**c**) anterior compartment fasciotomy; (**d**) elevation of anterior compartment off of tibia, interosseous membrane and fibula; (**e**) deep posterior compartment exposure and fasciotomy; (**f**) deep posterior compartment elevation off of tibia, interosseous membrane and fibula (medial view above, cross-section view below); (**g**) excision of interosseous membrane sequence from left to right. Care is taken to avoid injury to the peroneal vessels, which lie posterolateral to the membrane. These vessels perforate the membrane distally to pass anterior to the membrane. Cross-section after excision of membrane is shown (right); (**h**) incision of tibial hamartoma (left). Dashed red lines show the anterior longitudinal incision for both the tibial and fibular hamartomas and the radial incision lines at the limits of resection (right); (**i**) the bone ends protrude through the anterior incision of the hamartoma (top). The proximal and distal radial cuts are made to circumferentially excise the hamartoma; (**j**) anterior and radial incision of fibular hamartoma with complete removal of this tumor (left to right). Cross-section view showing that both the tibial and fibular hamartomas have been excised circumferentially (right). The dissection does not extend to the distal tibiofibular syndesmosis since the fibula is not proximally migrated; (**k**) when the fibula is proximally migrated, the dissection should be extended distally to expose the tibio-fibular syndesmosis. The peroneal vessels run over this area (left) and should be mobilized and retracted out of the way (second to left). The anterior tibiofibular ligament is cut (middle) and then by separating the bones the posterior tibiofibular ligament is visualized (second to right) and cut (right). This frees up the fibula to be reduced to station when the fibula is proximally migrated.



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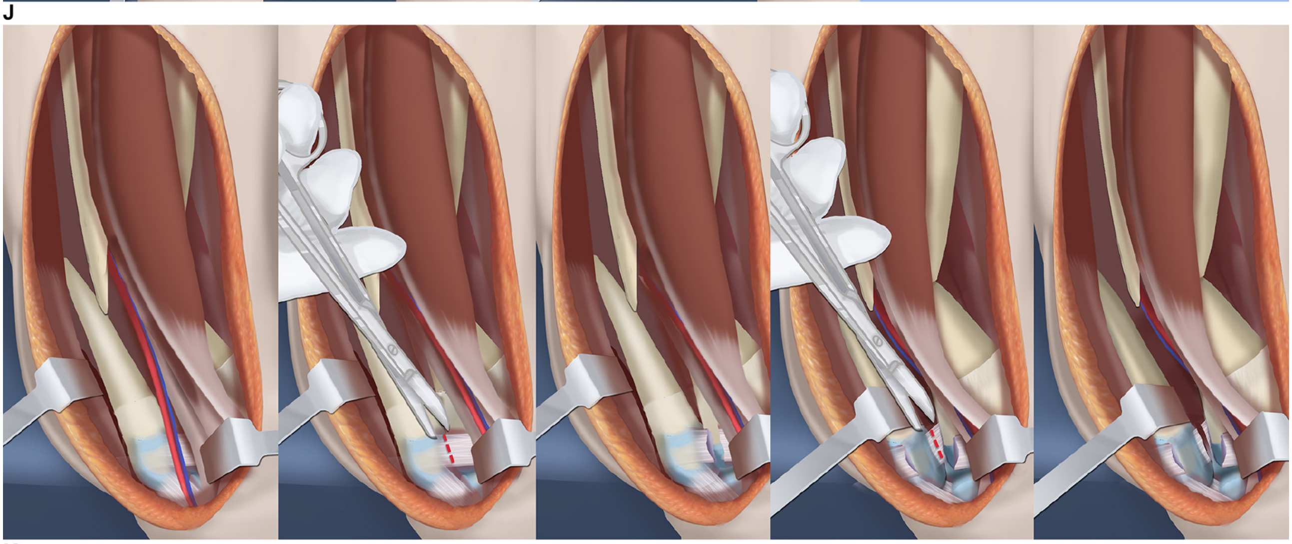

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Supplemental Fig. S1 Cont.

(distal fibular physis at level of ankle mortis): longitudinally and radially incise the periosteum of the fibula and excise the fibular hamartoma at the same levels as the tibial hamartoma excision (Supplemental Fig. S1j). Do not disrupt the syndesmotic ligaments.

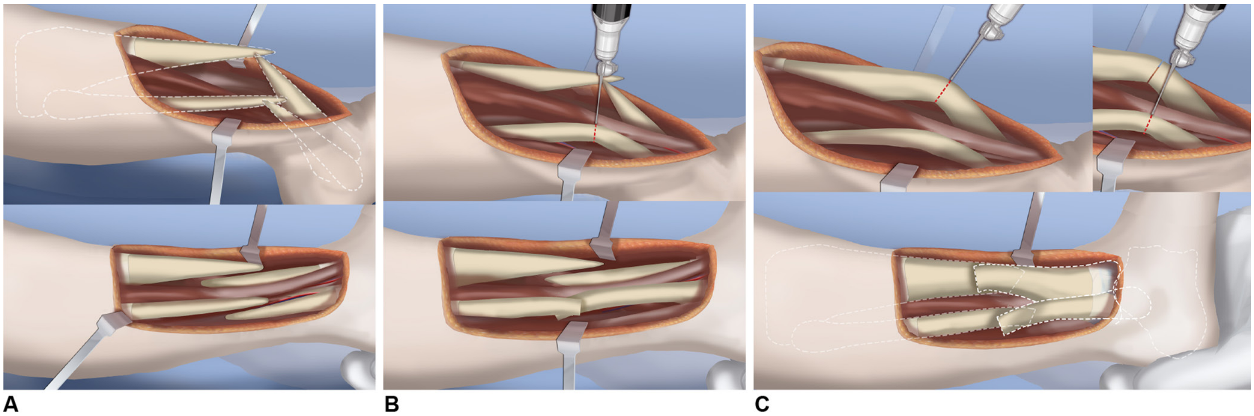
Step 11b: Also release syndesmotic ligaments if fibula proximally migrated/not at station (type 2b, 4b) (distal fibular physis proximal to level of ankle mortis): expose the anterior tibio-fibular syndesmotic ligament. If necessary, extend the skin incision distally, to allow for direct exposure of the tibio-fibular syndesmosis. Retract the peroneal artery and vein to see the ligament. Cut the anterior ligament and then separate the bones and under direct vision cut the posterior ligament (Supplemental Fig. S1k).

Step 12a: Bone end separation (type 4a: tibia and fibula CPT present): separate the tibia and fibula CPT bone ends from each other and allow the bone ends to overlap (shorten) as the deformity is straightened (Supplemental Fig. S2a).

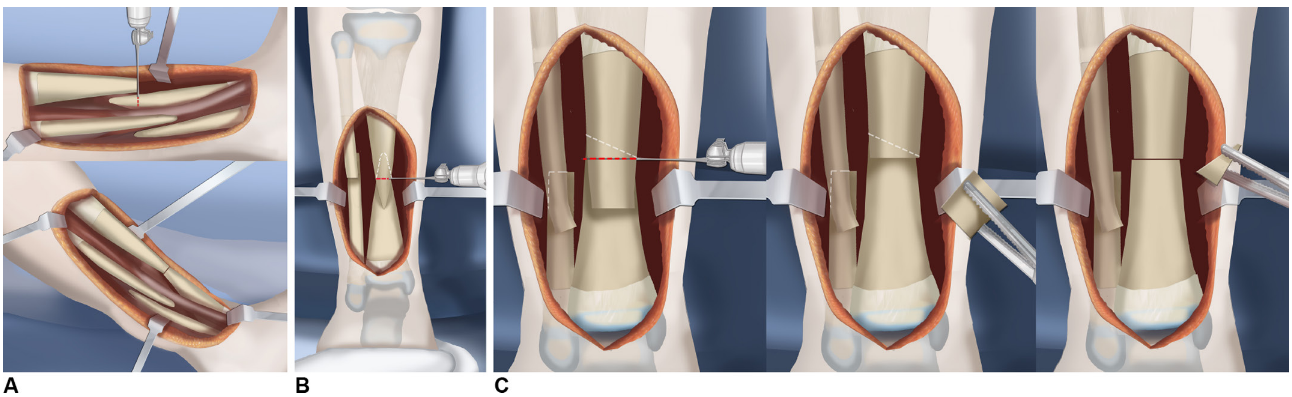
Step 12b: Bone end separation (type 3: tibia CPT present, fibula intact): osteotomize the fibula at the apex of its deformity. If the fibula has no angulation, osteotomize it at the level of the tibial CPT. Separate the fibular bone ends and allow the tibia and fibula bone ends to overlap, as the deformity is acutely corrected (Supplemental Fig. S2b).

Step 12c: Bone end separation (type 1: tibia and fibula intact): osteotomize the tibia at the apex of its deformity. Osteotomize the fibula at the apex of its deformity. If the fibula has no angulation, osteotomize it at the level of the tibial osteotomy. Separate the tibial and fibular bone ends and allow the tibia and fibula bone ends to overlap, as the deformity is acutely corrected (Supplemental Fig. S2c).

Step 13a: Square off tibial bone ends (type 4: tibia and fibula CPT present): mark the level where to cut the two ends of the tibia proximally and distally. Square them off using a saw and irrigation. Try and make the cuts so that the most sclerotic and atrophic parts of the tibia are resected. Try and minimize the resection. The intention is



Supplemental Fig. S2 Reproduced with permission by the Paley Foundation: (**a**) the anterolateral bowing of the congenital pseudarthrosis of the tibia (CPT) (top) can now be straightened by allowing the bone ends to shorten (overlap) relative to each other (bottom); (**b**) if the fibula is intact with a CPT of the tibia, the fibula must first be osteotomized (top) before the anterolateral bowing can be straightened (bottom) allowing for overlap of the bone ends; (**c**) if the tibia and fibula are both intact with anterolateral bowing, the tibia (top left) and fibula (top right) must first be osteotomized before the anterolateral bowing can be straightened (bottom) allowing for overlap of the bone ends.



Supplemental Fig. S3 Reproduced with permission by the Paley Foundation: (**a**) the tibial bone ends are cut at the level that will maximize the cross-sectional area of contact between the bone ends when the tibia is reduced. The fibula is not cut at this time; (**b**) in the case of an intact fibula: The tibial bone ends are cut at the level that will maximize the cross-sectional area of contact between the bone ends when the tibia is reduced. The fibula is not cut at this time; (**c**) in the case of an intact tibia and fibula: the tibial bone ends are cut at the level that will maximize the cross-sectional area of contact between the bone ends when the tibia is reduced. The fibula is not cut at this time. Since the distal tibia is shorter than the proximal tibia, the larger section is usually taken from the proximal end (left) and the minimal amount from the distal end (middle). At the end the bone ends are square so that they have good alignment (right).

not to resect the CPT. The cuts are simply to bring two ends of the tibia together in a straight alignment position with the minimum amount removed. The intention is to also make the tibial cuts at a level that is as wide as possible to maximize the bone to bone contact area (Supplemental Fig. S3a).

Step 13b: Square off tibial bone ends (type 3: tibia CPT present, fibula intact): follow the same instructions as in Step 13a (Supplemental Fig. S3b).

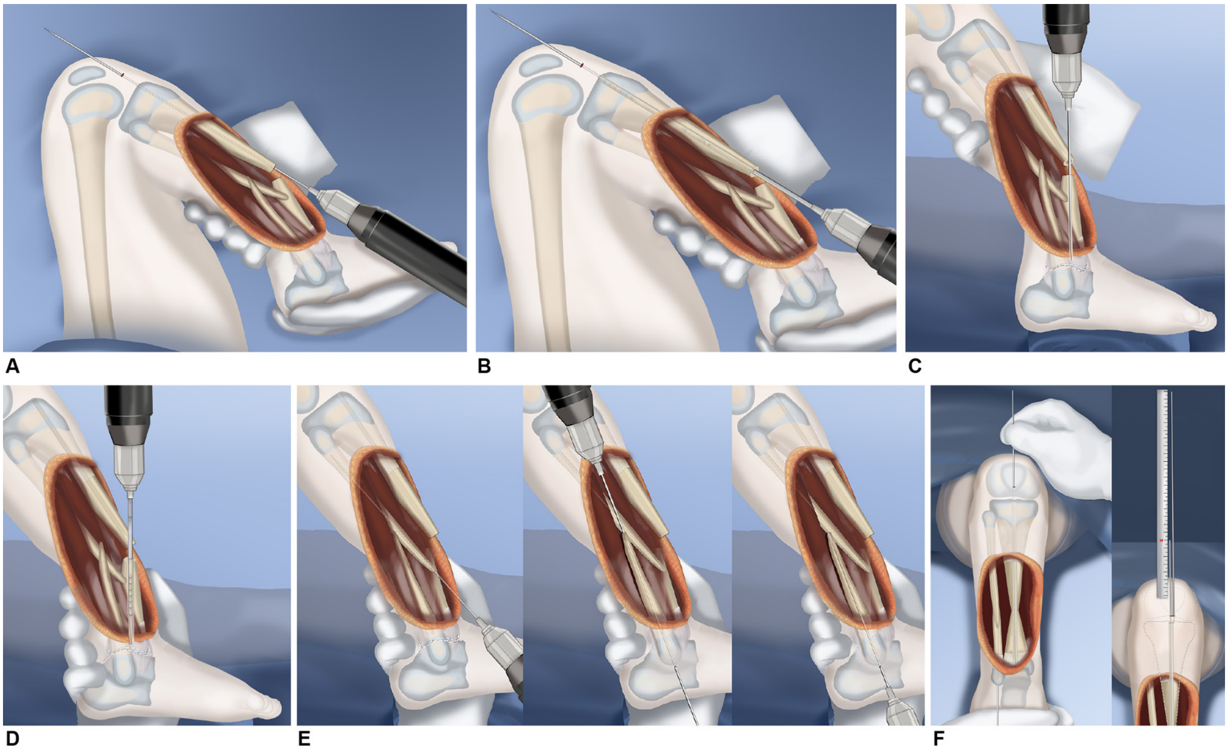
Step 13c: Square off tibial bone ends (tibia and fibula intact): follow the same instructions as in Step 13a (Supplemental Fig. S3c).

Step 14: Proximal guide wire: drill a retrograde proximal tibial guide wire. Make sure this guide wire enters at the distal medullary canal and exits through the proximal epiphysis in the anterior half of the tibia in the sagittal plane and midline in the frontal plane (Supplemental Fig. S4a).

Step 15: Proximal tibial reaming: drill the proximal tibia with a cannulated drill overtop the guide wire. Use the diameter of drill corresponding to the diameter of nail planned for fixation (Supplemental Fig. S4b).

Step 16: Distal guide wire: drill an antegrade distal tibial guide wire. Make sure this guide wire enters at the proximal medullary canal and ends in the mid distal epiphysis (Supplemental Fig. S4c).

Step 17: Distal tibial reaming: drill the distal tibia with a cannulated drill overtop the guide wire. Use the diameter



Supplemental Fig. S4 Reproduced with permission by the Paley Foundation: (**a**) a 1.8-mm guide wire is drilled retrograde through the proximal fragment to exit anteriorly in the tibia and distal to the patella. It is necessary to flex the knee maximally to pass this wire. Distally there may be no medullary canal. The wire should be drilled to recreate this canal; (**b**) a cannulated drill from the Fassier-Duval set is used to ream open the canal of the proximal tibia. These drill bits are slightly larger than the diameter of the corresponding nail. The smallest diameter nail is 3.2 mm; (**c**) a wire is drilled antegrade into the distal tibia. It is important to centre this wire in the distal tibial epiphysis as much as possible; (**d**) a cannulated drill from the Fassier-Duval set is used to ream open the distal medullary canal; (**e**) a small diameter wire (1 mm, 1.5 mm or 1.8 mm) is used to drill open the medullary canal of both ends of the fibula. If the fibular end is too thin or sclerotic, try drilling the wire from the epiphyseal end to exit the bone in the diaphysis. Even if the exit point is not the very tip of the fibula, the wire hole can be used later in an 'in – out – in' pathway. After completing this, back the wire out the distal end and leave it in the distal fragment; (**f**) insert a 1.8 mm Kirschner-wire down the tibia exiting out the upper end. This wire should span the entire length of the reduced tibia (left). Insert a second wire of the same length down the tibia to perch on the tibial plateau joint surface. Measure the difference in length between these two wires to determine the total length of the nail needed.

of drill corresponding to the diameter of nail planned for fixation (Supplemental Fig. S4d).

Step 18: Drill the fibula with a wire: use a wire to drill the two ends of the fibula from the level of the CPT or osteotomy. Distally it should come out the lateral malleolus. At the distal end leave the wire protruding out the lateral malleolus (Supplemental Fig. S4e).

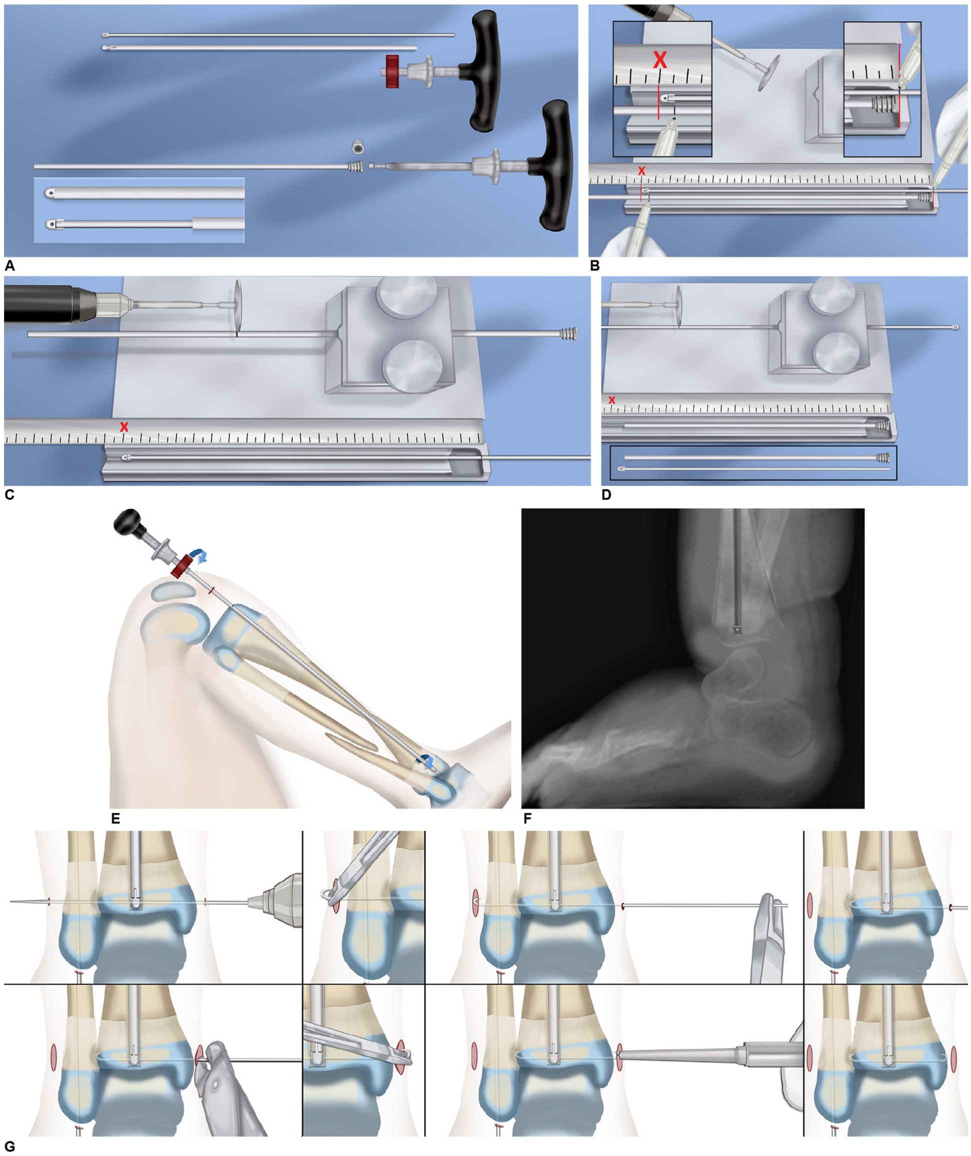
Step 19: Measure intramedullary length: measure the length of the reamed track of the intramedullary canal of the proximal and distal tibia combined by inserting a guide wire down the entire track. Measure the length of this wire protruding proximally by inserting a second equal length wire down to the proximal surface of the tibial epiphysis (Supplemental Fig. S4f).

Step 20: Fassier-Duval telescopic nail preparation: the Fassier-Duval telescopic nail is used. It consists of a male and female portion that fit together passively, by sliding the thinner male end into the larger diameter female part (lower picture). The male end has a hole at its end for locking into the distal epiphysis. The female part has a threaded head to screw into the proximal epiphysis. A T-handle with introducer is used to insert the male end (top upper picture), and a hexagonal driver is used for the female end (bottom lower picture) (Supplemental Fig. S5a).

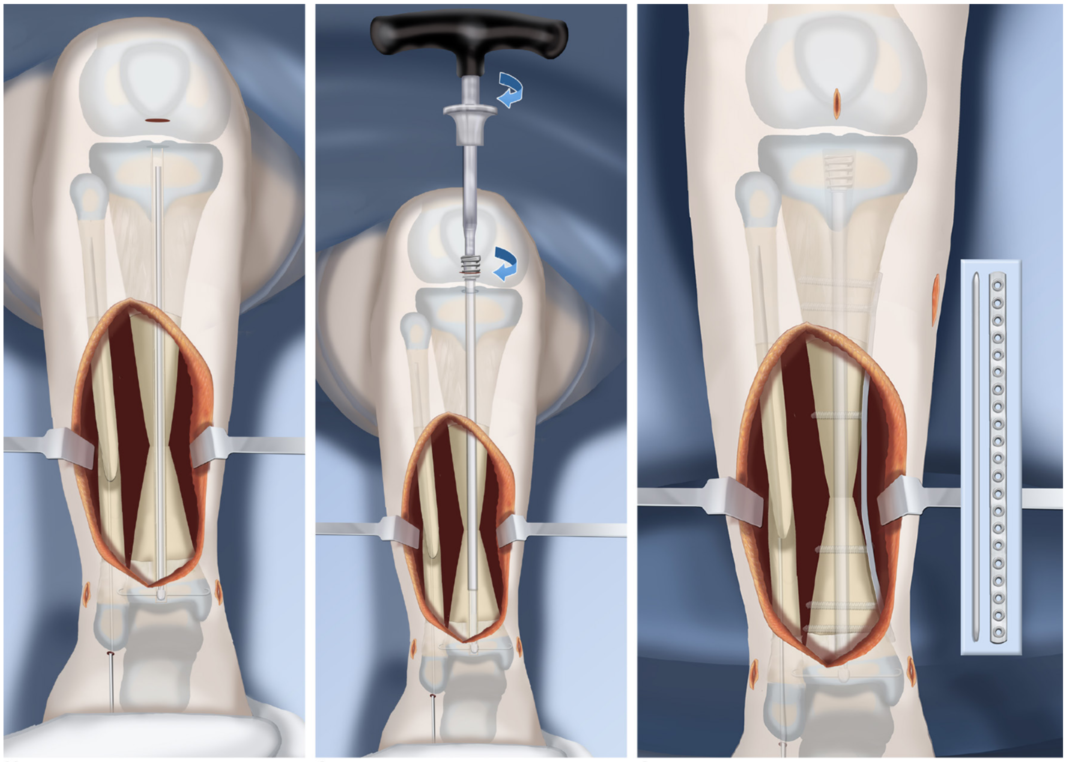
A special measuring jig is used to mark where to cut the male and female ends (Supplemental Fig. S5b)

A metal cutting saw is used to cut the female (Supplemental Fig. S5c) and male (Supplemental Fig. S5d) Fassier-Duval nails to the measured length while they are held in a special vice.

Step 21: Male component insertion: insert the precut male portion of the Fassier-Duval with the locking hole end into the epiphysis. Use the introducer to control the



Supplemental Fig. S5 Reproduced with permission by the Paley Foundation: (**a**) Fassier-Duval equipment, top: male rod with introducer. The introducer is slotted to control rotation of the male component that has a fin that fits in the slot at its distal end. Bottom: female end and its wrench. The female rod slides over the male rod. Note that the male rod has a locking hole at its end and not a screw end (the screw end is used for osteogenesis imperfect but not for congenital pseudarthrosis of the tibia (CPT)). This male end is the preferred type for CPT due to the small size of the distal tibial epiphysis; (**b**) the uncut male and female ends are measured relative to each other on a cutting jig, and relative to the total length of the nail 'x' measured determined in Supplemental Figure S4f. Both rods are marked; (**c**) the female end is fixed in a vice. A high-speed diamond tipped metal cutting wheel is used to cut the female end at the correct length; (**d**) a high-speed diamond wheel metal cutting wheel is used to cut the male end at the correct length (top). The male end is fixed in a vice. The customized male and female ends are measured to ensure they are the desired lengths (bottom); (**e**) the male introducer inserts the male rod through the canal of the tibia to the distal epiphysis. The hole in the nail is rotated so its opening is medial-lateral; (**f**) using the image intensifier the end of the nail is visualized and the image is magnified; (**g**) the hole in the rod is 1.6 mm diameter. A 1.5 mm Kirschner-wire is drilled from the medial side through the hole in the nail and out the lateral side (top left). The wire is then bent 180° (top left center) and pulled to the bone (top right center and top right). The wire is then cut short (bottom left) and bent 180° (bottom left center) and then impacted into the bone so it is completely flat under the thin skin on the medial side of the tibia (bottom right center). The wire is left flush to bone on the medial side (bottom right); h) the male rod is inside the tibia and is distally locked; i) the female rod is introduced overtop the male rod and advanced until its threaded head is screwed into the proximal tibial epiphysis; j) to control axial rotation and length, a small diameter locking plate is used on the medial side of the tibia. One screw is placed at either end and then the additional screws added. The screws pass either anterior or posterior to the rod. It is important that they do not impinge on the rod since both ends of the rod need to slide within the bone in response to growth from the physes at each end. The plate is shown in profile and face on. The author's preference is the EVOS plate (Smith & Nephew Orthopedics, Memphis, Tennessee) with its very small diameter screws. It is important not to use screws bigger than 2 mm or 2.7 mm in diameter.



Supplemental Fig. S5 Cont.

insertion. It is preferable to use what is known as the Paley modification or lengthening over nail (LON) version of the male Fassier-Duval nail. The screw ended male Fassier-Duval nail used for osteogenesis imperfect does not fix well into the small distal tibial epiphysis. Use the lockable male end, which uses a 1.5 mm wire for locking (Supplemental Fig. S5e).

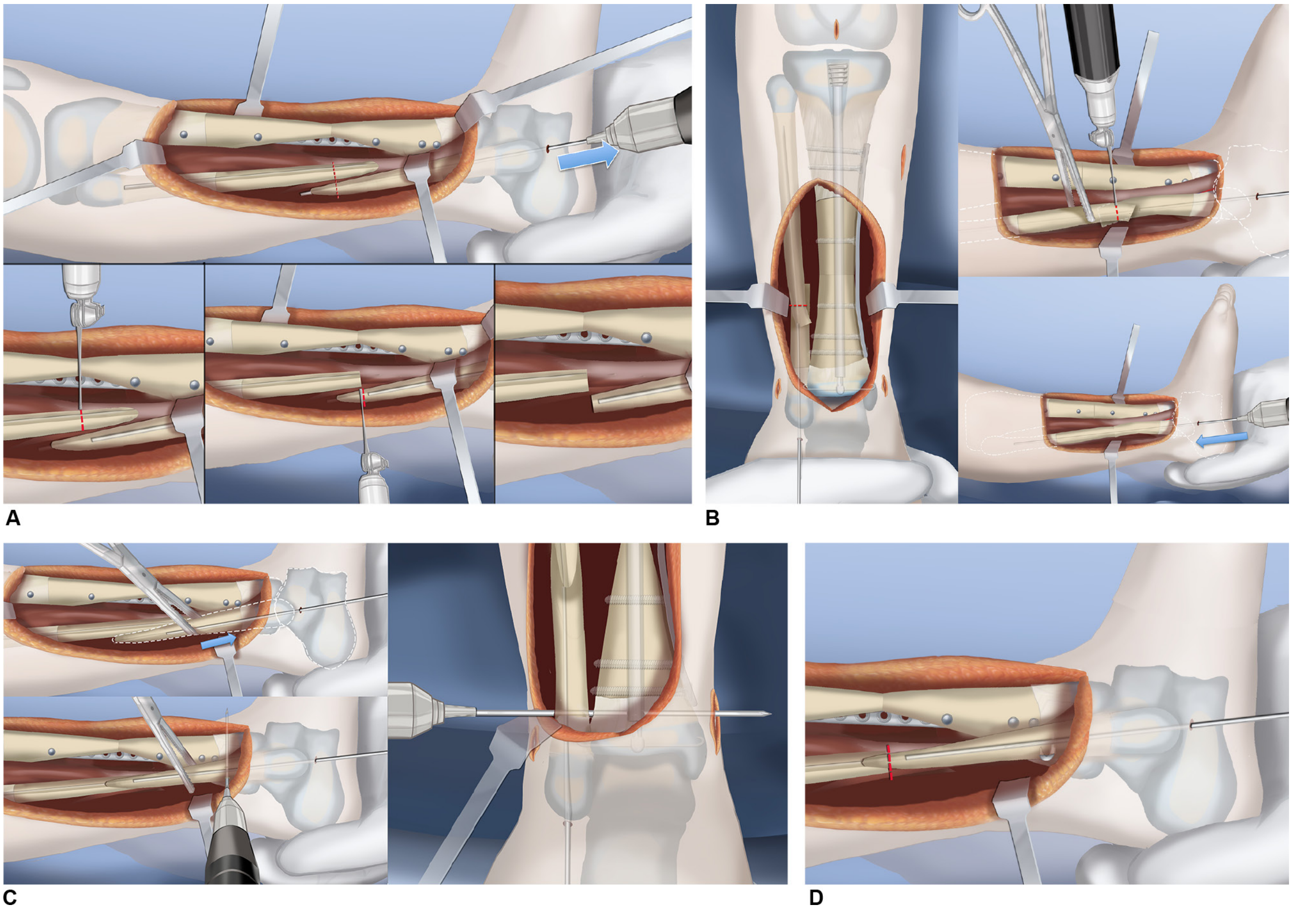
Step 22: Distal locking: using the handle of the introducer, rotate the male component so that the hole faces medial-lateral. Centre the hole on the image intensifier screen and magnify it maximally (Supplemental Fig. S5f). Now use a 1.5 mm Kirschner-wire and drill it from the medial side of the epiphysis centered on the hole. Drill it across the epiphysis and exit out the lateral side. Make a small incision on the lateral side and after bending the wire 180°, pull it back into the distal epiphysis. Do the same on the medial side. Cut the wire short and bend it using a small needle driver. Use a tamp (punch) to push the bent wire into the bone from medial to lateral so it is not prominent on the medial side (Supplemental Fig. S5g). Now remove the introducer (Supplemental Fig. S5h).

Step 23: Insert the female component: insert the precut female Fassier-Duval nail, capturing the male end in its canulation. Screw the female end into the upper tibial epiphysis (Supplemental Fig. S5i). Take care to use the fully threaded female end without the smooth extension.

Step 24: Locking plate: the locking plate should be a low profile thin long straight plate that extends from the distal to the proximal tibia. Lock it with two to three screws on either side of the CPT with compression of the pseudarthrosis (Supplemental Fig. S5j). Author's preferred plate is the EVOS (Smith & Nephew Orthopedics, Memphis, Tennessee) plate. This may require an additional medial incision for the proximal screws. Use manual compression to compress the bone ends. To compress the bone using the plate, first fix the two end screws. Then add the two more central screws and have these pull the straight plate down to bone. This deformation of the plate causes compression at the CPT site.

Step 25a: Cut the ends of the fibula when there is a CPT (if fibula at station-distal fibular physis opposite ankle joint line): the overlapping ends of the fibula can now be cut. It is best to err on cutting too little than too much. This way there is compression of the ends of the fibula when the two ends are brought together (Supplemental Fig. S6a).

Step 25b: Cut the ends of the fibula when the fibula was intact and was osteotomized (types 1, 2a, 3, 4a; fibula at station): the overlapping ends of the fibula can now be cut. It is best to err on cutting too little than too much. This way there is compression of the ends of the fibula when the two ends are brought together (Supplemental Fig. S6b).



Supplemental Fig. S6 Reproduced with permission by the Paley Foundation: (**a**) lateral view of the tibia with fibula at station, showing the screws of the plate posterior to the nail. The fibular bone ends are overlapping. The fibula is cut to maximize the diameter of contact. It is better to cut the bone 1 mm too long so that the bone ends are under compression when reduced; (**b**) if the fibula was osteotomized as in Supplemental Figures S3b or 3c, the ends of the fibula need to be cut square (left and top right) and then reduced and the wire advanced across the fibula to stabilize it (bottom right); (**c**) if the fibula has migrated proximally (top left) as in Supplemental Fig. S1k, the fibula should be acutely transported distally (bottom right). When it is restored to the correct station (distal fibular physis at level of ankle joint, the fibula should be transfixed with a wire to the tibia to hold it reduced at station (bottom left and right); (**d**) for the example in Supplemental Fig. S6c, the fibula can now be cut.

Step 25b: Cut the ends of the fibula after moving distal fibula to station (types 2b, 4b; distal fibula migrated proximally): move the distal fibula distally until its physis is at the level of the joint line. Transfix the fibula and tibia with a wire to hold the fibula at station (Supplemental Fig. S6c). Now cut the ends of the fibula relative to each other (Supplemental Fig. S6d).

Step 26: Fibular nailing: advance the wire that is protruding through the lateral malleolus into the proximal fibula up to the proximal fibular physis (Supplemental Fig. S7a). Cut and curl this wire and bury it into the lateral malleolus (Supplemental Fig. S7b).

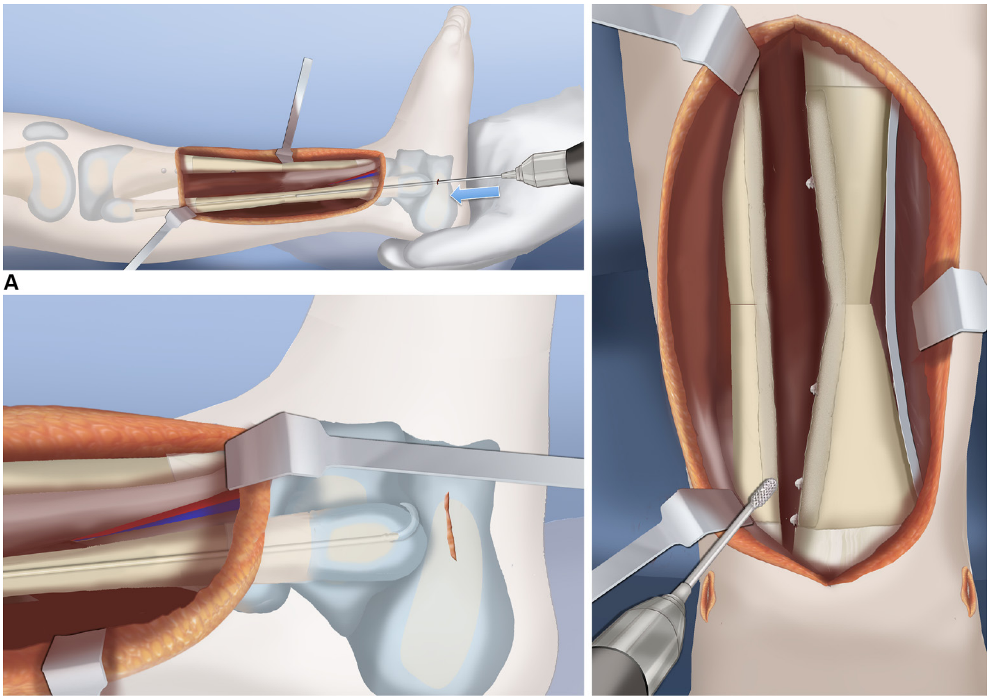
Step 27: Burr the opposing surfaces of the tibia and fibula: burr all the surfaces, which will be in contact with the bone graft. These are especially the opposing surfaces of the distal tibia and fibula (Supplemental Fig. S7c). Pack the wound with wet gauze and wrap the leg with an elastic bandage. Remove and/or deflate the tourniquet.

Step 28: Incision at the iliac crest: make a bikini incision at the iliac crest (Supplemental Fig. S8a).

Step 29: Expose the iliac apophysis: cut through the fascia over the sartorius and identify and decompress the lateral femoral cutaneous nerve. Expose the apophysis. Dissect the external oblique muscle off of the apophysis. Dissect and expose the apophysis from the superior to inferior iliac spine (Supplemental Fig. S8b).

Step 30: Split the apophysis: use a #15 blade, to cut through the apophysis from the inferior to superior spine and then across the iliac crest from anterior to posterior (Supplemental Fig. S8c).

Step 31: Periosteal graft harvest and meshing: peel the periosteum off of the medial side of the ilium. Incise the



Supplemental Fig. S7 Reproduced with permission by the Paley Foundation: (**a**) advance the fibular wire from distal to proximal into the predrilled hole in the proximal fibula; (**b**) cut, curl and bury the end of the wire. It is important to make sure this wire is not prominent since it will stay buried until skeletal maturity; (**c**) using a high-speed burr and irrigation, burr the facing sides of the tibia and fibula. This will help the cross-union to develop. The tourniquet is removed after this step.

periosteum on the undersurface of the iliacus muscle. Incise it in a rectangular pattern removing as large a piece as possible (Supplemental Fig. S8d). Dissect this periosteum from the muscle and then lay it on the plastic sheet used for meshing a skin graft. Mesh the periosteum in order to expand it (Supplemental Fig. S8e).

Step 32: Split iliac bone: use a thin osteotome to split the two cortices of the ilium. Start by splitting the cortices between the superior and inferior iliac spines (Supplemental Fig. S8f). Then go from anterior to posterior splitting the cortices to the same depth. The osteotome is advanced one cm at each place along the crest and then repeated one cm deeper (Supplemental Fig. S8g, h, i). This avoids splintering the crest especially posteriorly. It is very easy to perforate the cortices as one moves into the very thin region between the anterior and posterior crests. Advance the osteotome deeper at each level until one reaches the dome of the acetabulum and the triradiate cartilage anteriorly; the sciatic notch in the middle; the sacro-iliac joint posteriorly. When the split is deep enough, we start to lever the medial crest medially. Never lever the lateral crest laterally. This levering with the osteotome will gradually open the ilium like a book. It will hinge on the deepest cortex. Do not start the cancellous bone harvest until the two tables of the ilium are fully separated.

Step 33: Iliac bone decancellousization: curette the cancellous bone off of the two tables of the ilium. Do not harvest or break off segments of cortical bone. There is a large amount of cancellous bone in the supra-acetabular, triradiate, sacro-iliac notch and sacro-iliac/posterior iliac crest and spine regions. Gradually curette the areas until they are devoid of cancellous bone (Supplemental Fig. S8 j, k, l). Use the image intensifier to prevent penetrating or damaging the hip, triradiate or sacro-iliac regions. The harvested cancellous bone totals between 20 cc to 45 cc in most children even as young as 12 months of age (Supplemental Fig. S8m). In these youngest children both iliac crests can be harvested if the needed volume of bone is not procured from one side.

Step 34: Backfill iliac crest: backfill the iliac crest with a synthetic bone void filler such as a calcium phosphate or demineralized bone matrix (Supplemental Fig. S8n). Suture closed the iliac apophysis including securing the external oblique muscles (Supplemental Fig. S8o).

Step 36: Preparation of periosteum, BMP2 and bone graft: the periosteal graft was already harvested and should be stored in saline to prevent drying. At least 15 minutes prior to completing the bone graft harvest, a medium BMP2 (Infuse Implant, Medtronic, Memphis, Tennessee) with its four collagen sponges should be soaked

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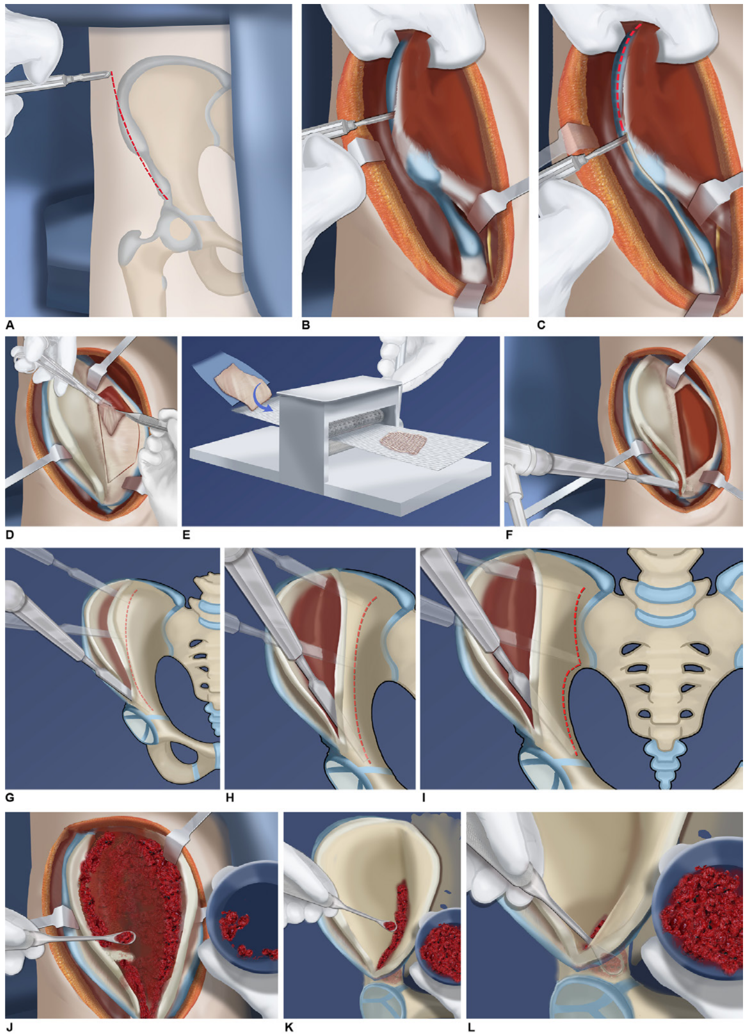
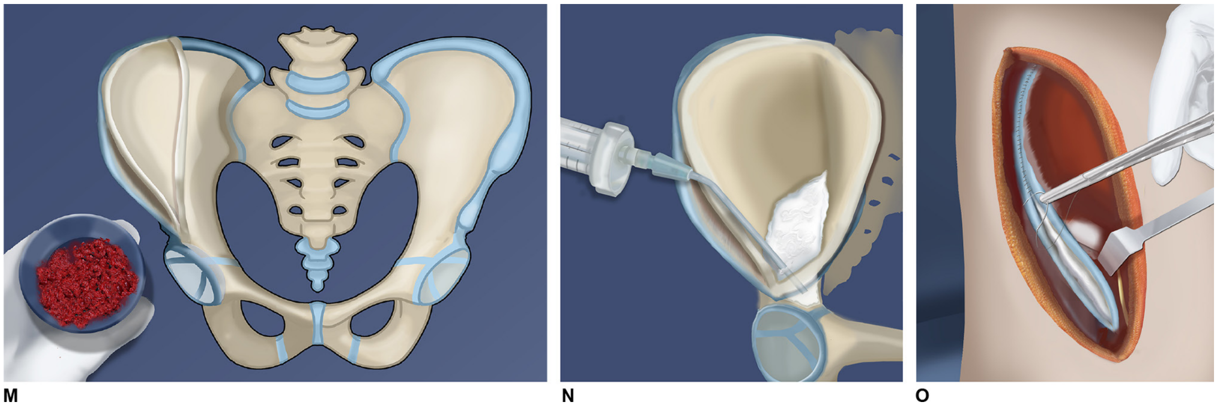


Supplemental Fig. S8 Reproduced with permission by the Paley Foundation: (**a**) make a long bikini incision along the ipsilateral iliac crest; (**b**) find and decompress the lateral femoral cutaneous nerve and protect it. Expose the cartilage of the iliac apophysis by raising some of the external oblique muscle from lateral to medial; (**c**) split the apophysis from the anterior inferior iliac spine (AIIS) to the anterior superior iliac spine (ASIS) to the crest of the tibia from anterior to posterior. Split the crest as far as possible all the way to the posterior iliac crest. This is much farther than normally done for a pelvic osteotomy; (**d**) reflect back the medial apophysis and periosteum off of the ilium. Incise the periosteum to excise a large rectangular window of periosteum from the undersurface of the iliacus muscle; (**e**) the periosteum immediately shrinks to one quarter its area. Use a skin graft mesher to mesh the periosteum, so that it can be enlarged again; (**f**) use a very thin, sharp osteotome to split the tables of the ilium. Start by separating the cortices between the AIIS and ASIS; (**g, h, i**) split the iliac tables apart by going from anterior to posterior and only penetrating a cm at a time. Do not advance too far at any one spot. Work back and forth from front to back penetrating ever deeper into the ilium and avoiding exiting the cortical bone. If accidentally the osteotome exits, try and find a way to reenter the bone at that level. It is possible to split even the thinnest part of the ilium with a thin sharp osteotome. Guide the osteotome with the image intensifier to avoid entering the hip or sacro-iliac joints or the triradiate cartilage. It is essential to reach these structures as well as the roof of the sciatic notch. Please note that the split goes all the way back to the posterior iliac crest. It is not difficult to reach there from this incision and from the front. It is important to follow the round contour of the iliac wing to avoid perforating the bone. This step is the hardest part of the entire procedure. Once the split is complete you can lever the medial wall away from the lateral wall using the osteotome. Do not do this prematurely and do not lever the lateral wall away from the medial; (**j**) the ilium will now open like a book exposing its entire cache of cancellous bone. Use straight and angled currettes to remove the spongiosa bone. Do not remove cortical bone; (**k**) currette out layer by layer of bone from superficial to deep. The entire posterior iliac crest cancellous bone can be retrieved by this approach; (**l**) the biggest trove of bone is above the dome of the acetabulum and extending to the triradiate cartilage. All of this bone should be removed without perforating these structures; (**m**) at the end there is no more cancellous bone in the ilium. This is why this is called decancellousization of the ilium. The cup should contain at least 15 mm to 20 cc of bone ranging up to 50 or more cc, depending on the age, sex and size of the patient; (**n**) the space created by the curettage can either be left to fill in on its own or can be back-filled with a bone void filler. The author uses a calcium phosphate cement that is injected and then hardens in place. It will slowly be replaced with normal cancellous bone; (**o**) suture closed the apophysis and also suture the external oblique muscle to avoid creating an abdominal hernia. Take care not to inadvertently catch the lateral femoral cutaneous nerve in the suture line.



Supplemental Fig. S8 Cont.

with BMP2 solution. This needs to sit for 15 minutes to allow the BMP2 protein to bond to the collagen sponge. The bed for the bone graft is dried and retracted prior to insertion of the periosteum and BMP. To minimize bone graft time out of the body (cold time) and maximize the viability of the bone graft, the bone graft harvest is timed to be ready for insertion only after all the bone graft bed has been fully prepared (Supplemental Fig. S9a).

Step 37: Insert BMP2 posteriorly: insert two of the BMP2 collagen sponges behind the tibia and fibula spanning the space between them. The BMP2 collagen sponges lie on the surface of the deep posterior muscles (Supplemental Fig. S9b).

Step 38: Periosteal grafting: apply the periosteal graft, cambium layer down, overtop the CPT site. Since the plate is on the medial side, the periosteum is mostly on the lateral, anterior and posterior sides. If possible, tuck the periosteum under the plate to anchor it and wrap it around the lateral and posterior sides (Supplemental Fig. S9c). The last two more central screws in the plate can be loosened and then retightened after tucking the periosteal graft under the plate.

Step 39: Insert the cancellous bone: insert the cancellous bone in layers between the tibia and fibula (Supplemental Fig. S9d). Pack it into this interosseous space and then layer some anterior to the tibia. Do not place any medially to avoid having the plate become covered by bone (Supplemental Fig. S9e). Check the location of the bone graft with the image intensifier to ensure that it covers the areas above and below the CPT of the tibia and fibula as desired.

Step 40: Insert BMP anteriorly: insert two of the BMP sponges anterior to the tibia and fibula spanning the space between them and overlying the cancellous bone graft (Supplemental Fig. S9f).

Step 41: Anterior muscle placement: allow the anterior compartment muscles to lie over the BMP2 sponges and tibia. This forms an important layer overtop the bone, bringing new blood supply to this healing area. The cancellous bone is sandwiched between two layers of BMP2 sponges, which are sandwiched between the anterior and posterior muscles (Supplemental Fig. S9g).

Step 42: Leg incisions closure: insert a hemovac drain between the muscle and subcutaneous tissues to help manage the deadspace and prevent hematoma (Supplemental Fig. S9h). Close the skin in layers and apply sterile dressings. Also close all the smaller leg incisions.

Step 43: Iliac incision closure: close this incision in multiple layers. Take care to avoid entrapping the lateral femoral cutaneous nerve with the fascial closure. A drain is not needed.

Step 44: Final radiographs: obtain an anteroposterior and lateral radiograph of the tibia (Supplemental Fig. S10).

Postoperative management: the leg is elevated for the first few days and ice is used for edema management. No splinting is needed initially since the internal fixation is sufficiently stable. A week after surgery a long leg cast is applied. The leg is kept non-weight-bearing for the first six weeks to protect the hip. After six weeks the patient is allowed to fully weight-bear on the leg with the cast. The weight-bearing restrictions for the first six weeks are for protection of the hip rather than the leg. The cast is removed after 12 weeks and radiographs are repeated. The cross-union is usually well established by 12 weeks. The cast is removed and an ankle-foot-orthotic (AFO) with articulated ankle is made. The patient is allowed to resume all activities with the brace. The brace does not need to be worn in the pool on in bed. No activity restrictions are placed.

Three months after the surgery a second ZA infusion should be done. The upper screws of the plate should be removed six months after surgery to help dynamize the tibia. The Fassier-Duval rod should be changed as needed to a larger diameter rod as the patient grows. Since the length of the bone doubles from age three years in girls and four years old in boys until skeletal maturity, the

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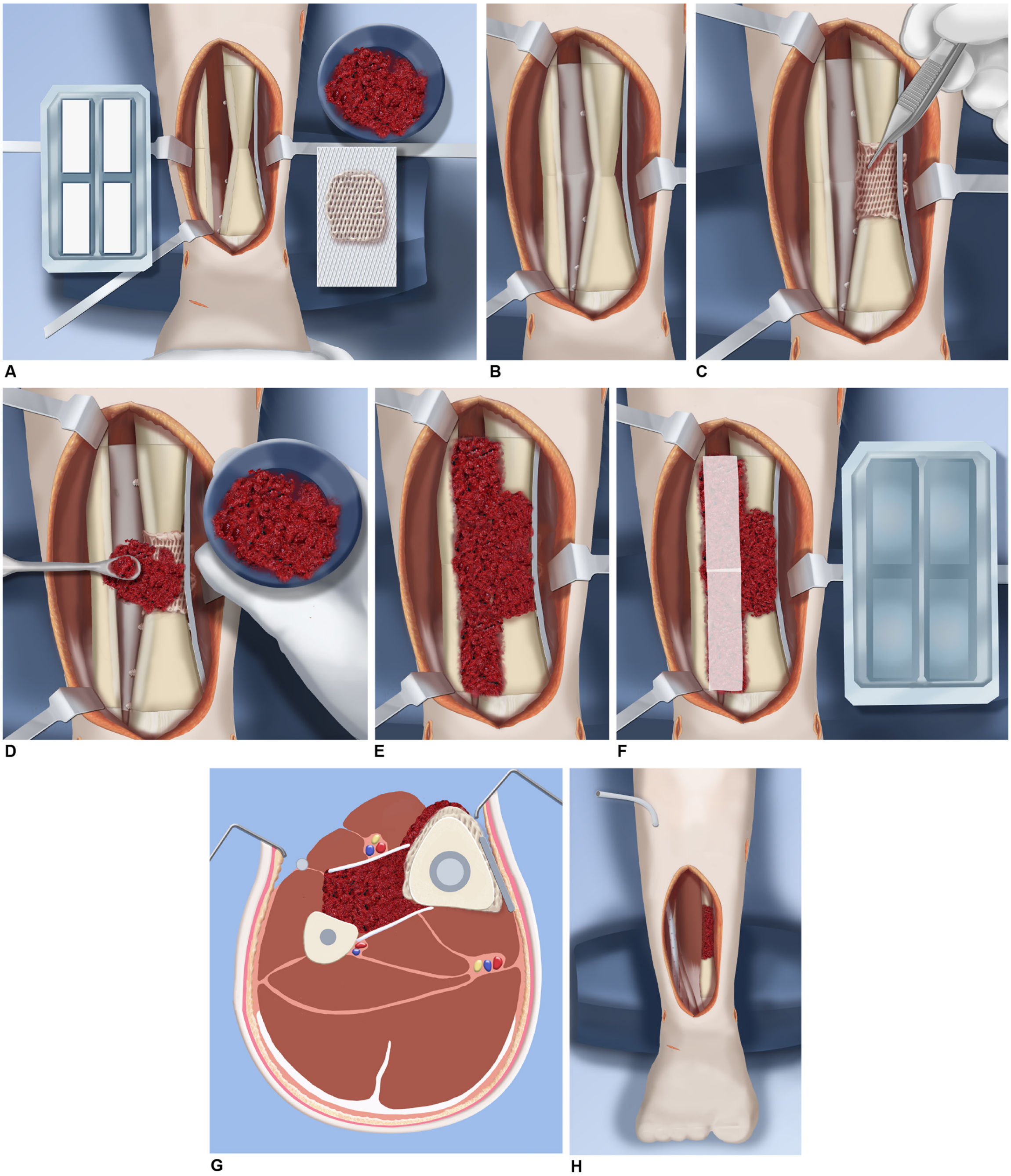
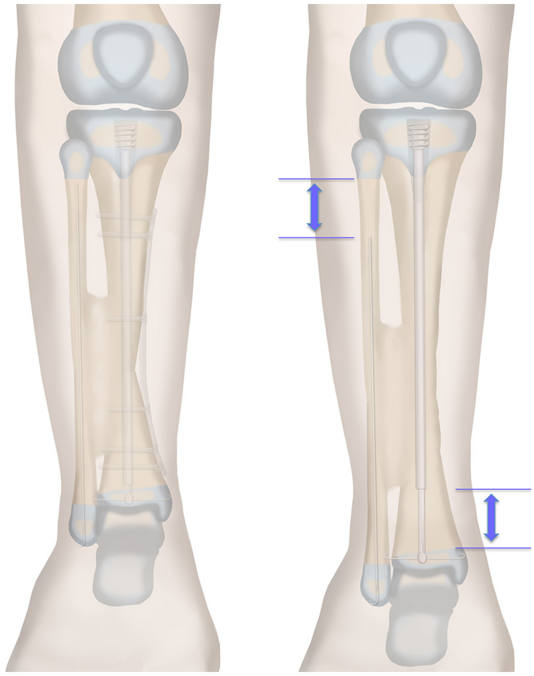


Supplemental Fig. S9 Reproduced with permission by the Paley Foundation: (**a**) 20 minutes prior to completion of the bone graft harvest, add the bone morphogenic protein (BMP) to the collagen sponges. Approximately four sponges (medium size Infuse implant, Medtronic, Memphis, Tennessee) should be prepared. The periosteum and bone graft are also ready and the surgical site should be irrigated and dried; (**b**) apply two BMP collagen sponges posterior to the tibia and fibula on the surface of the posterior muscles; (**c**) apply the periosteal graft cambium layer down on the congenital pseudarthrosis of the tibia (CPT) site. Tuck it under the plate to anchor it and wrap it around the lateral and posterior tibia; (**d**) pack layer by layer of the cancellous bone graft between the tibia and fibula; (**e**) extend the bone graft to the entire length of the area where the hamartoma was removed. There should be enough bone graft to create a long cross-union. Lay some of the graft anteriorly over the CPT site; (**f**) apply two more BMP collagen sponges anterior to the bone graft; (**g**) cross-section showing the BMP sandwich with the bone graft in the middle. Note that the BMP should be in contact with muscle so that it can recruit and induce cells from the muscle to form bone. The anterior and posterior tibial muscles lie on the surface of the anterior and posterior BMP sponges respectively. Note the location of the periosteal graft. It is critical that an adequate fasciotomy was performed since the anterior muscles have been displaced anteriorly; (**h**) the wound is seen prior to closure with the anterior muscles overlying most of the bone. A hemovac drain should be placed between the muscle and the subcutaneous tissue to help close the deadspace. It should not be in contact with the BMP sponges. The wound is closed in layers at this time.



Supplemental Fig. S10 Reproduced with permission by the Paley Foundation: by three months after surgery a robust cross-union exists between the tibia and fibula (left) The congenital pseudarthrosis of the tibia (CPT) of the tibia and fibula are usually healed by this time. Growth can proceed with telescoping of the Fassier-Duval nail (right). The plate does not impede bone growth. A second dose of zolidronic acid is given at three months after surgery.

telescopic rod has to be changed, at least once before maturity. Hemiepiphysiodesis is also performed if a valgus ankle or knee is present. The presence of the rod does not impede the use of a hemiepiphysiodesis screw plate device.

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