

Congenital Pseudarthrosis of the Tibia



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Disclosures

- K2M Consultant
- Medtronic Consultant
- POSNA President and BOD member
- AAP Immediate Past Chair and Section on Orthopaedics Executive Committee
- Project Perfect World Board of Directors.
- Miracle Feet Medical Advisory Board.



Learning Objectives

- Learn about underlying pathogenesis of CPT
- All options involve complete resection of the hamartoma and diseased periosteum
- All procedures try to create new local vascularized tissue.
- Understand the 3 common treatment options (local bone graft, free fibula transfer, ilizarov) that include IM tibia stabilization

What is Congenital Pseudarthrosis of Tibia?

- 1/150,000 births
- Spectrum from bowing to frank pseudarthrosis
- NF1 1/4000 AD, 1987 clinical criteria
- 4% of NF1 have CPT. 50% of CPT is in NF1
- Prognosis worse: fibula involved, more severe form, younger age, near ankle.



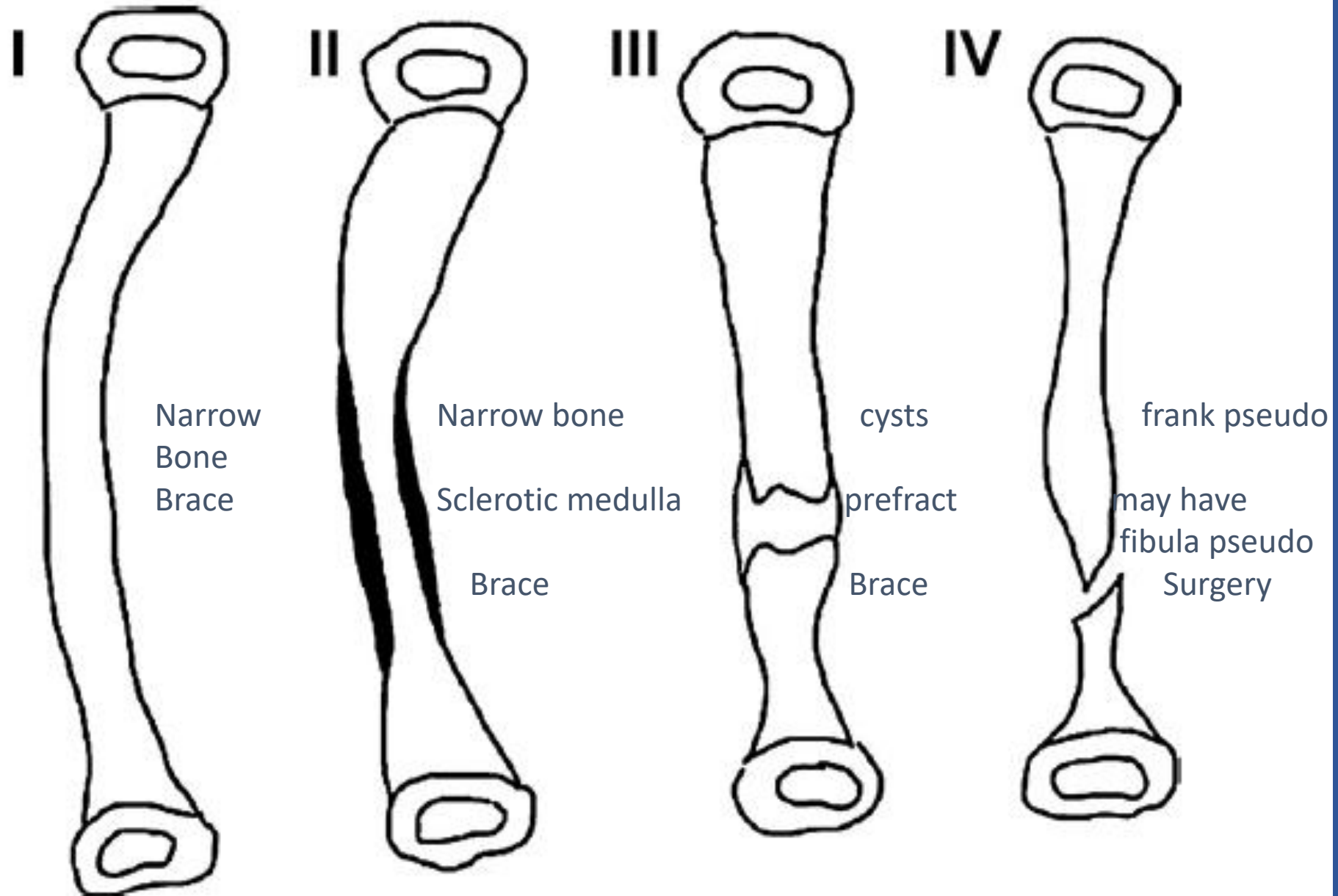
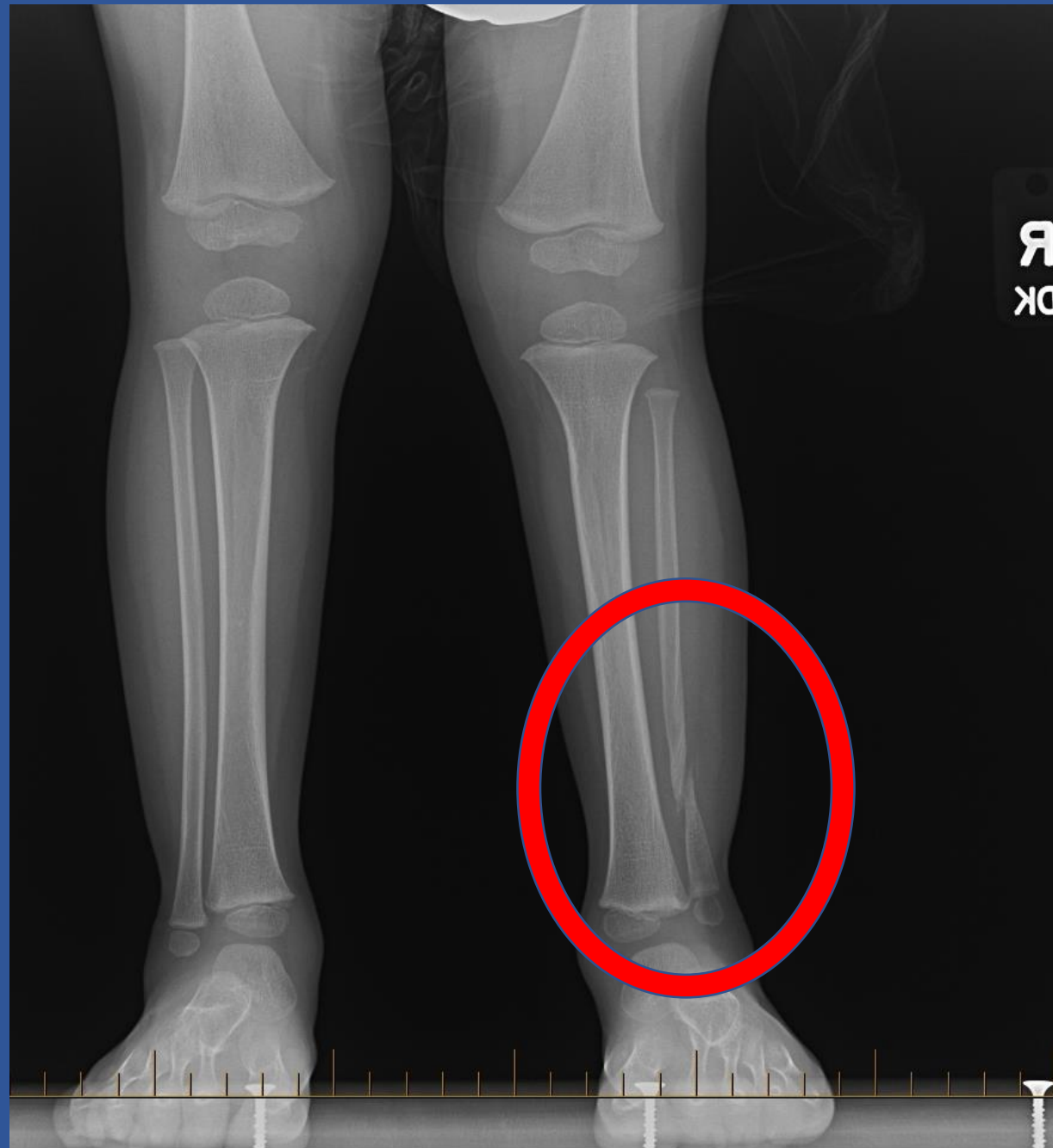


Figure 3 4 stage Crawford classification.

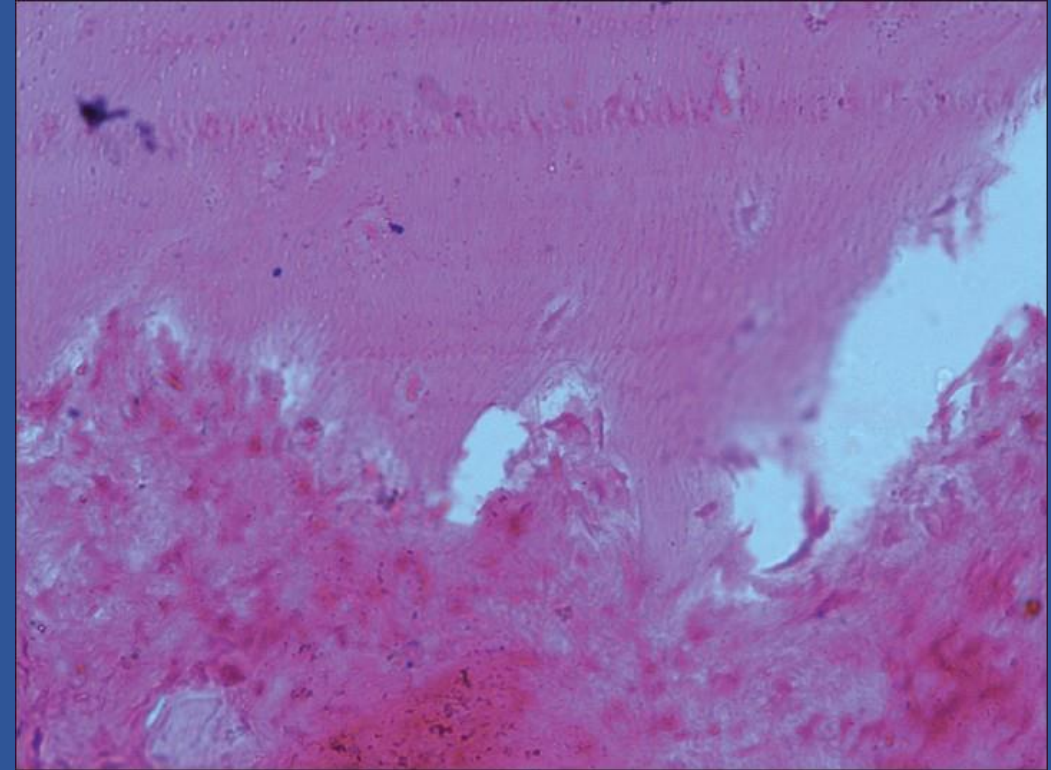
Can also
involve fibula



3yo boy NF1

Pathogenesis

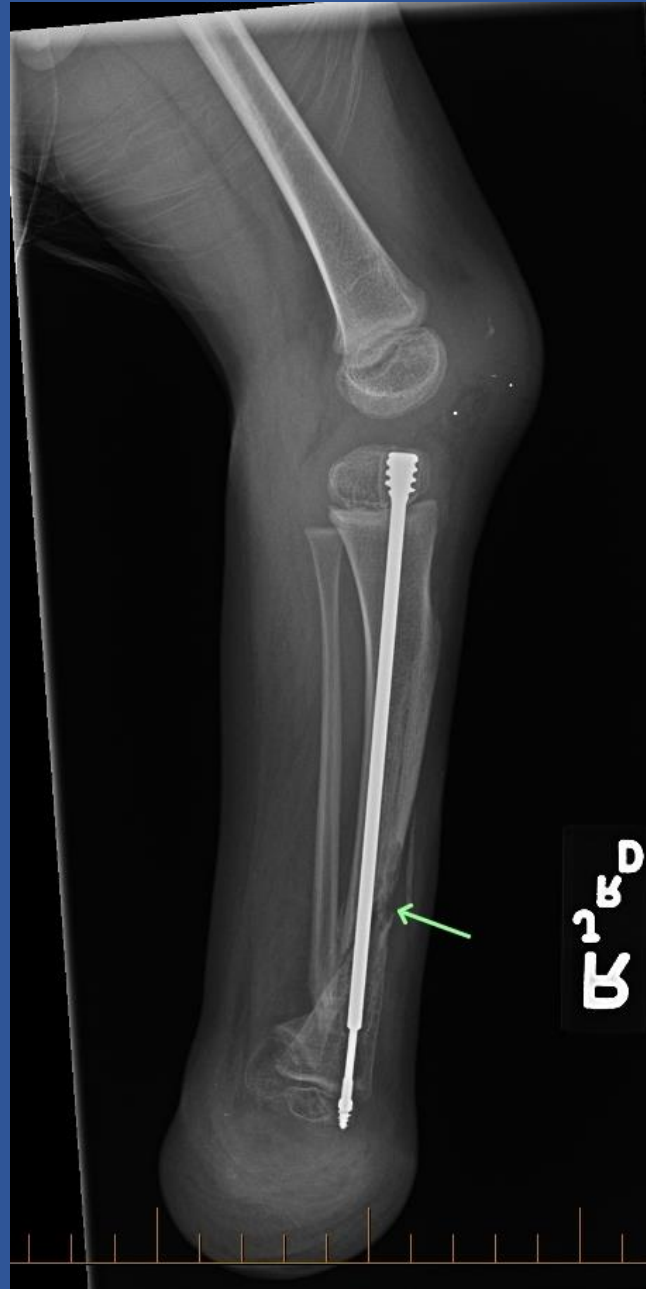
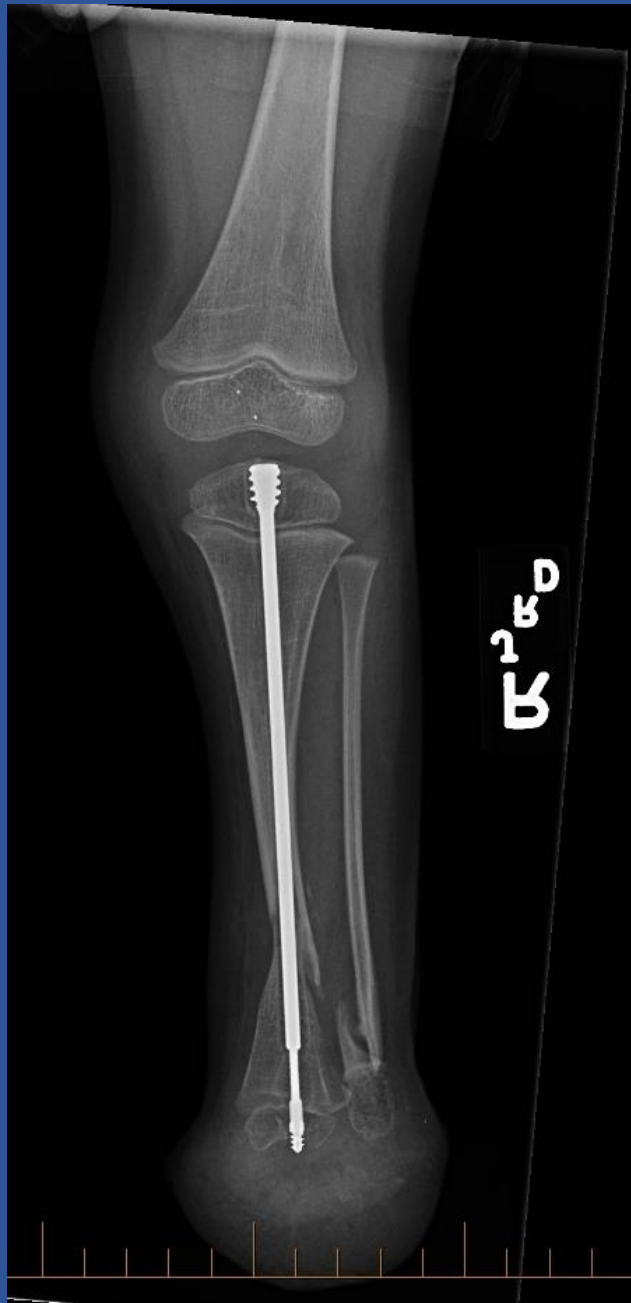
- Decreased osteogenic capacity and local vascularization
- Fibrous hamartoma and thickened periosteum
- Creates a constricting ring, decreased vascularization
- In NF1 codes for neurofibromin allows for active form RAS-GTP
- Defective osteoblasts, increase osteoclasts.



Treatment- the problem



9 mo with NF1 and
Crawford IV CPT



4 yo after numerous attempts with ICBG, BMP, Williams rod through hindfoot resulting in Syme amputation, but still not united



8 yo failed 15
surgeries. BKA is
"final" solution

Standard Treatment Options

- Nailing through ankle and bone grafting. 24-80% union
- Vascularized Fibula- contralateral or ipsilateral. 70% union, esp if older
- Ilizarov- 50-90% union. EPOS had best results with resection, compression, include foot. 50% need more surgery to treat refracture or pin infection. 25% valgus- recommend telescoping nail.



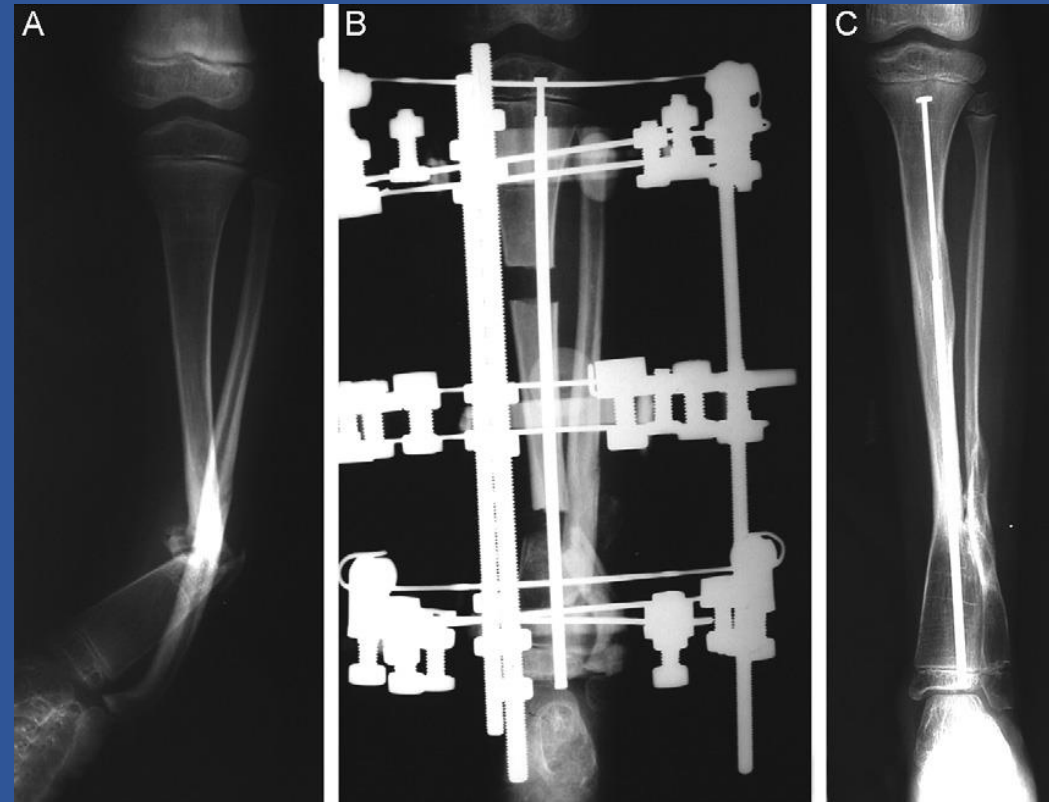
Standard Treatment Options

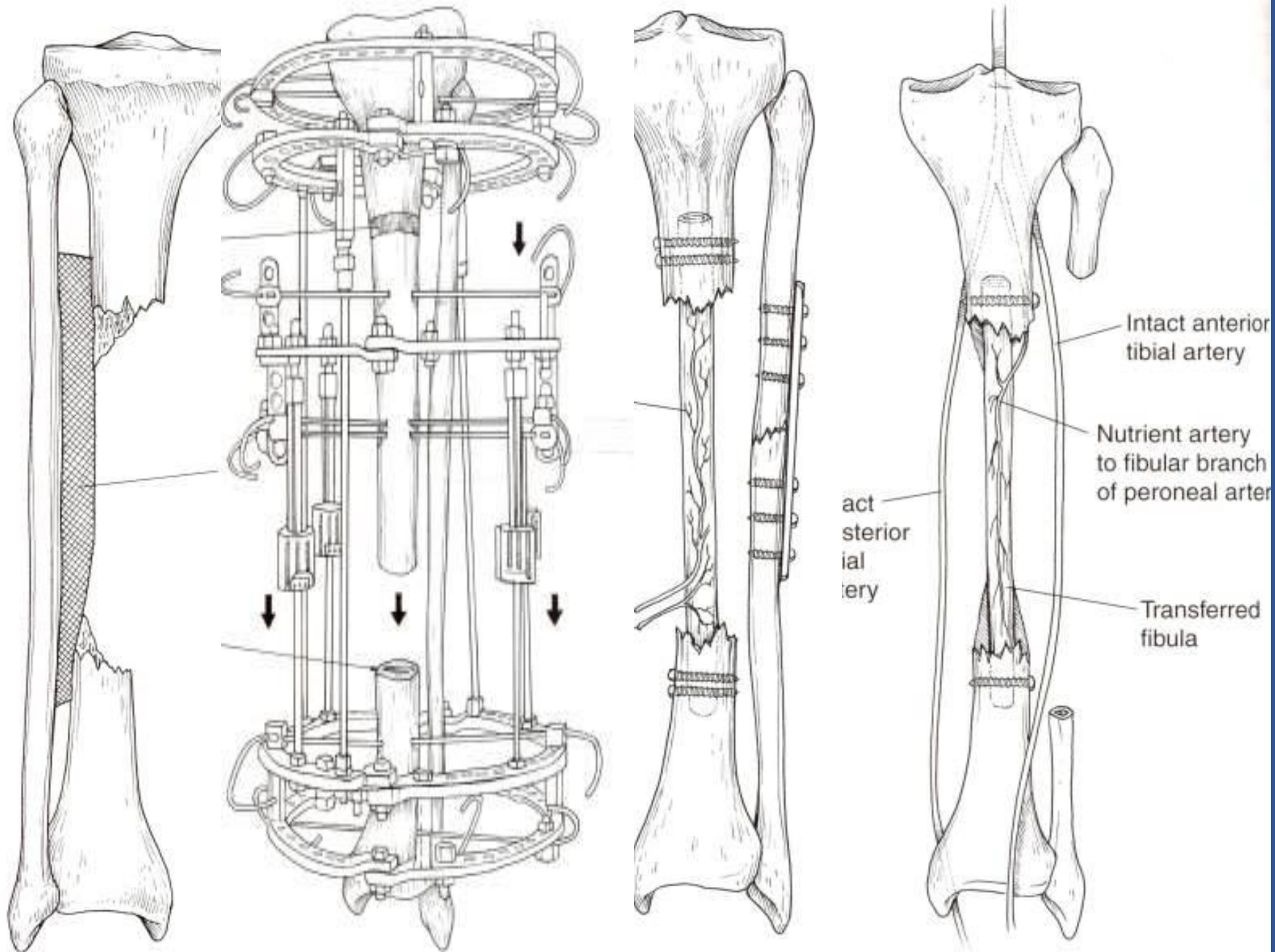
- Nailing through ankle and bone grafting. 24-80% union
- Vascularized Fibula- contralateral or ipsilateral. 70% union, esp if older. Harder to use nail. Treat 3-5 years
- Ilizarov- 50-90% union. EPOS had best results with resection, compression, include foot. 50% need more surgery to treat refracture or pin infection. 25% valgus- recommend telescoping nail.



Standard Treatment Options

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Newer techniques

- RhBMP-2 and RhBMP-7. 2 acts earlier (mesenchymous cells) than 7 (osteoblasts). Reports of infections, heterotopic ossification (Woo 2013)
- Complete resection of hamartoma and diseased periostium, cement spacer, induced pseudosynovial membrane to create a biological chamber for ICBG. Can be used <3 years old and up to 30-50% of the tibia. (AC Masquelet 1991)
- Periosteal graft from pelvis (Paley 2008)
- Electrostimulation
- Autologous and Bone marrow Cells

- 12 yo boy train coupling injury
- PT nerve visually intact
- Underwent emergent successful vascular reconstruction



- Damage control with external fixator
- But left with 8 cm tibia bone and soft tissue defect.



- latissimus free flap with 8 cm PMMA spacer
- Deformity adjusted with the fixator
- 6 weeks later (2) posterior ICBG inserted into defect. Don't pack too tight
- 6 months later, healed with no deformity, fully back to sports.



Summary

- Always consider the diagnosis of NF1
- Pay attention to the numerous technical details (Johnston CE, Birch JG 2008)
- Must remove the abnormal tissue. Like tumor surgery.
- Easier to treat after age 3 years, but severity determines results.
- Needs biology addressed, preferred with patient's own vascular tissue- via transport, free fibula or vascular chamber.
- Deformity correction and stable intramedullary internal fixation.
- Fibular pseudarthrosis and contribution to deformity
- Long term bracing to prevent late deformity which leads to fracture
- Follow to maturity

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