Leg Lengthening in RSS: Who, Why, & Where?

Magic Foundation Convention
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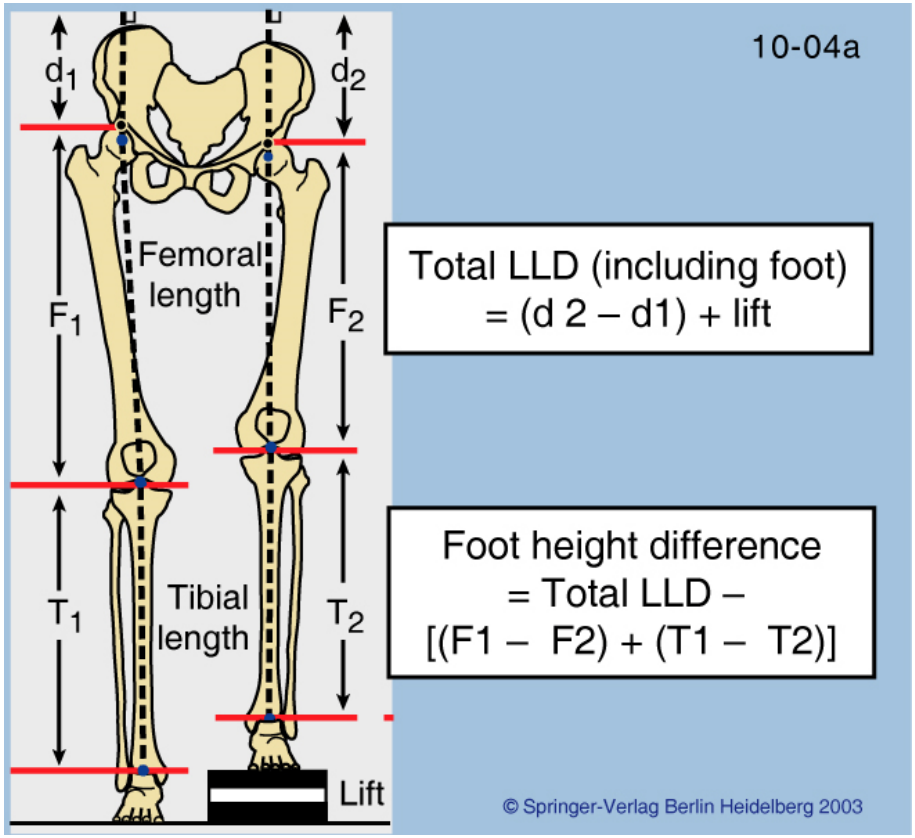
HOSPITAL FOR SPECIAL SURGERY

Evaluation

- Causes of LLD
  - Congenital
  - Traumatic Growth arrest
  - Overgrowth
  - Neural inhibition



Congenital LLD

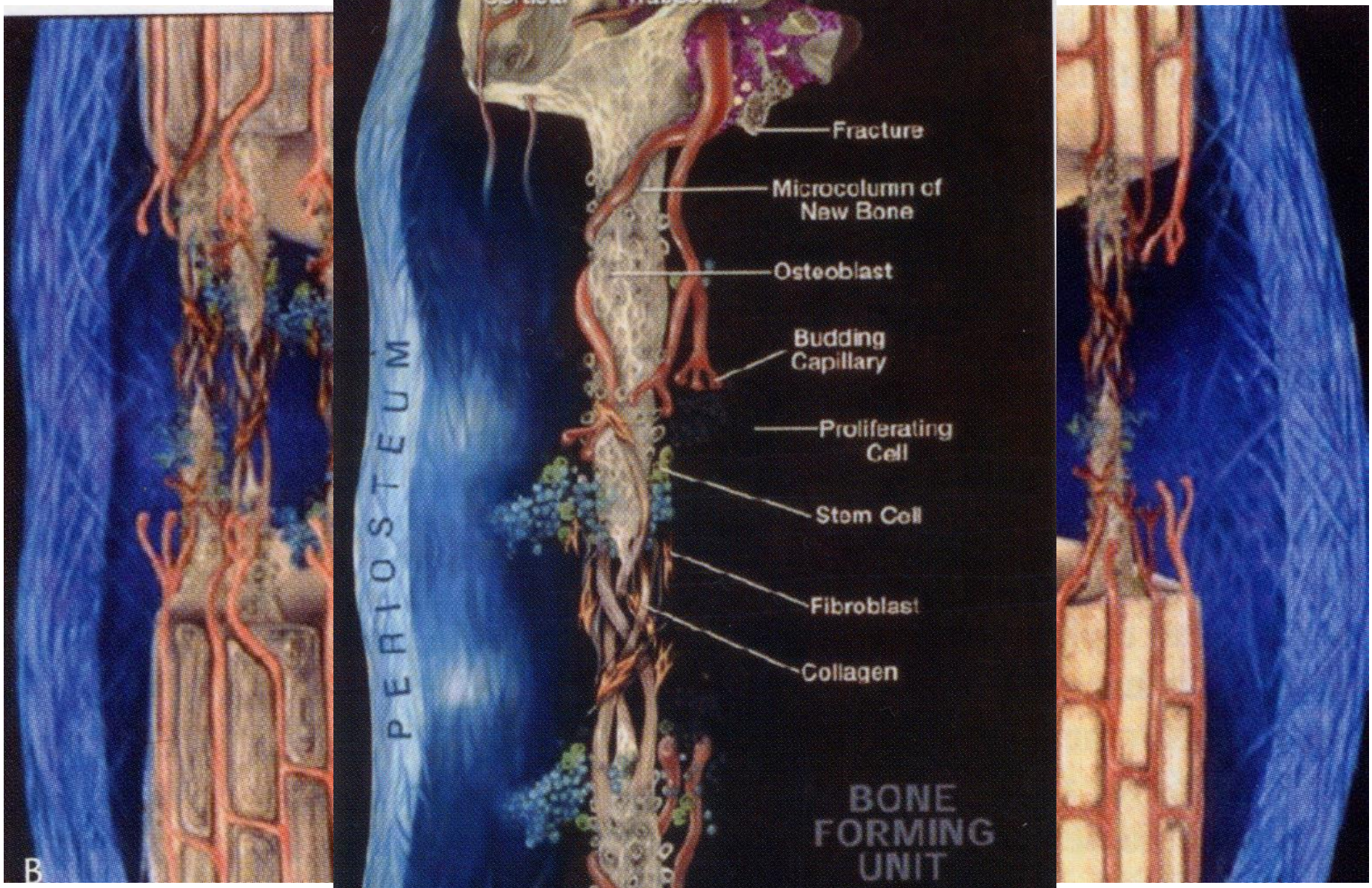
- Age at presentation
- Current LLD
- Distribution femur and tibia
- Bone Age
- Multiplier
- Compliance with shoe lift

- Treatment options
  - One stage lengthening
  - Two or more stages
  - Femur lengthening
  - Tibia lengthening

Growth Arrest

- Extent of damage
- Deformity
- Shortening
- Growth remaining
- Bone Age
- Multiplier

- Treatment options
  - Hemiepiphysiodesis to prevent worsening deformity
  - Close growth plate, correct deformity and lengthen leg
  - One or more lengthenings
  - Bar excision

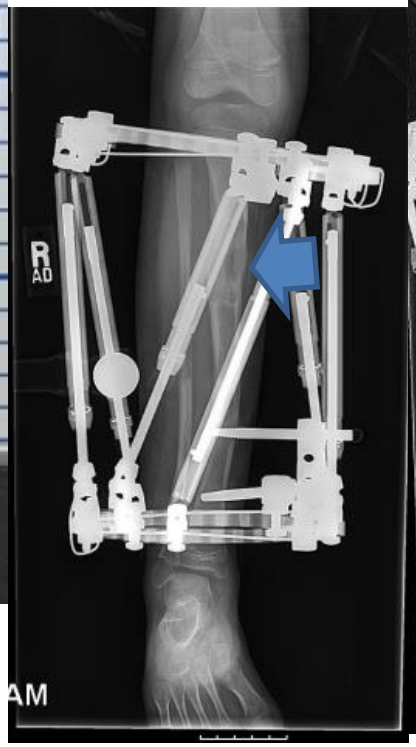

Cortical
Trabecular
Fracture
Microcolumn of New Bone
Osteoblast
Budding Capillary
Proliferating Cell
Stem Cell
Fibroblast
Collagen
PERIOSTEUM
BONE FORMING UNIT
DISTRACTION OSTEOGENESIS
A
B

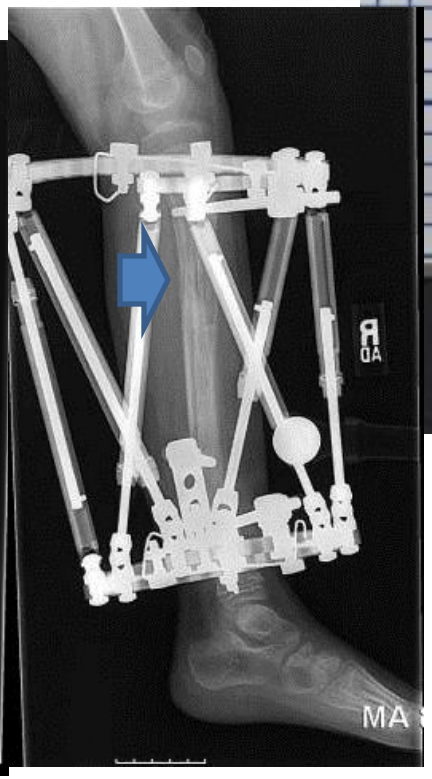
Outline

- Limb Lengthening examples
- RSS limb lengthening
- New Magnetic Internal Lengthening Nail
  - PRECICE NAIL
- Question & Answer

Posteromedial bow, age 6
LLD= 36 mm, all tibia
M= 1.68
PLLD= 6.1 cm

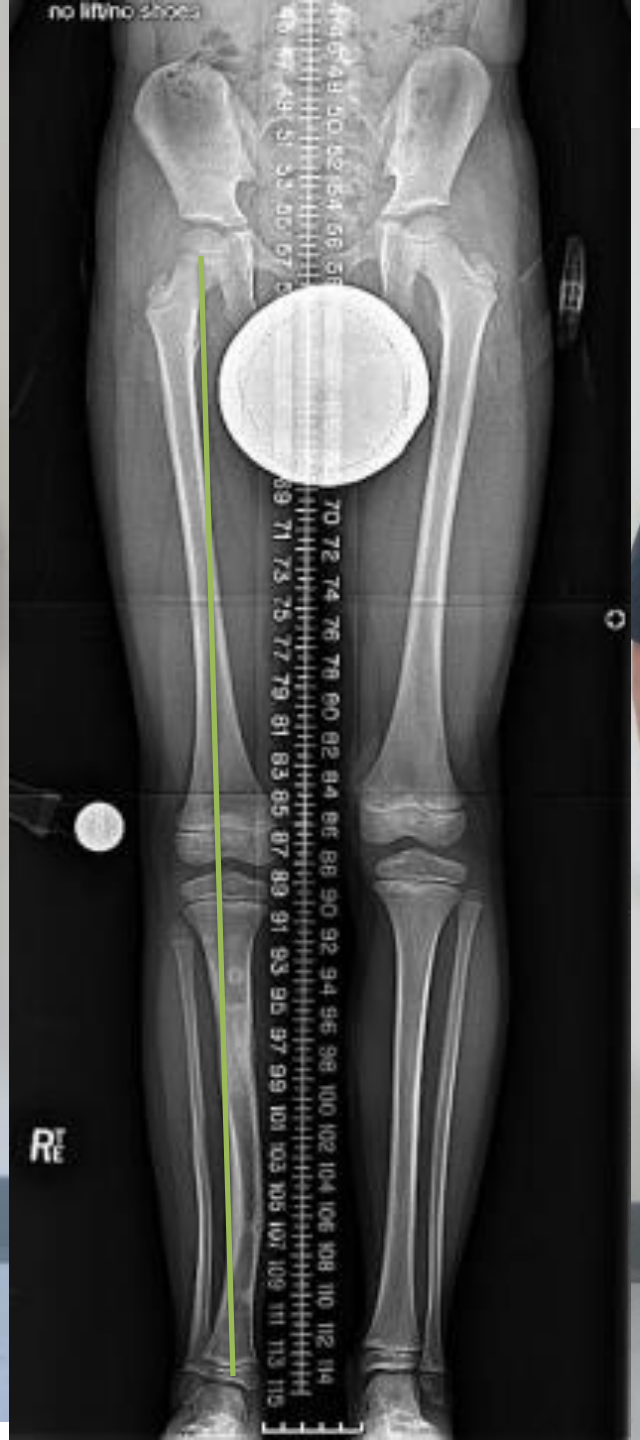
Plan: lengthen tibia 3.6 cm
Correct some prox tibia varus
Second lengthening in future





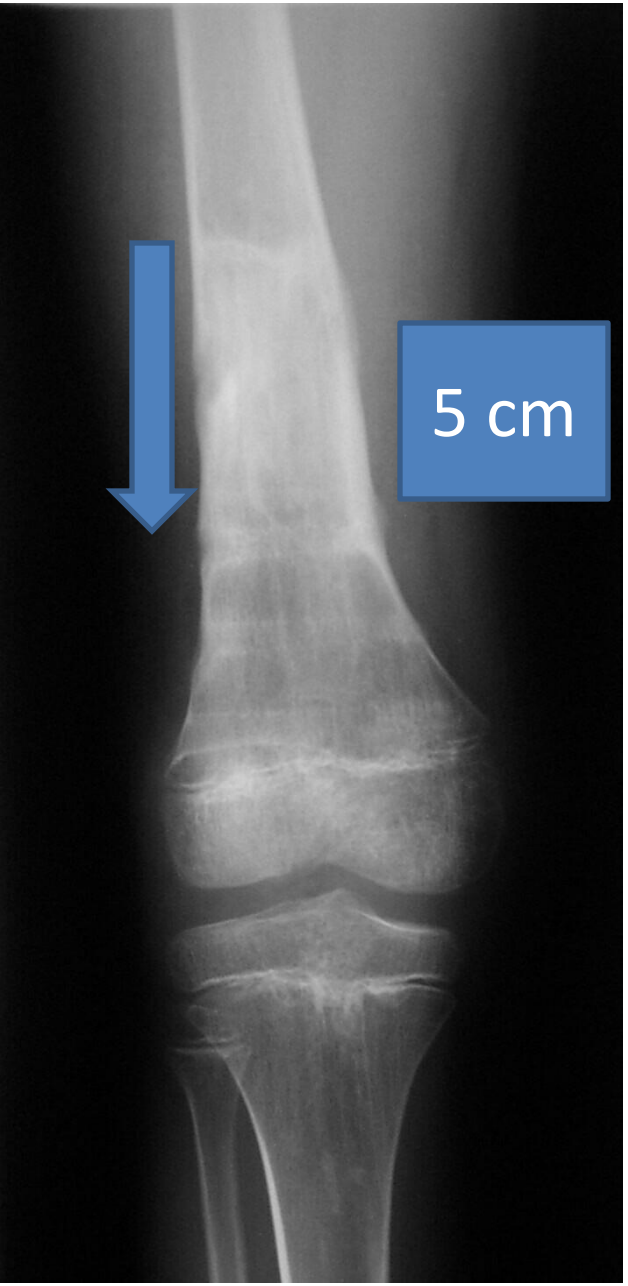
3 months
In frame

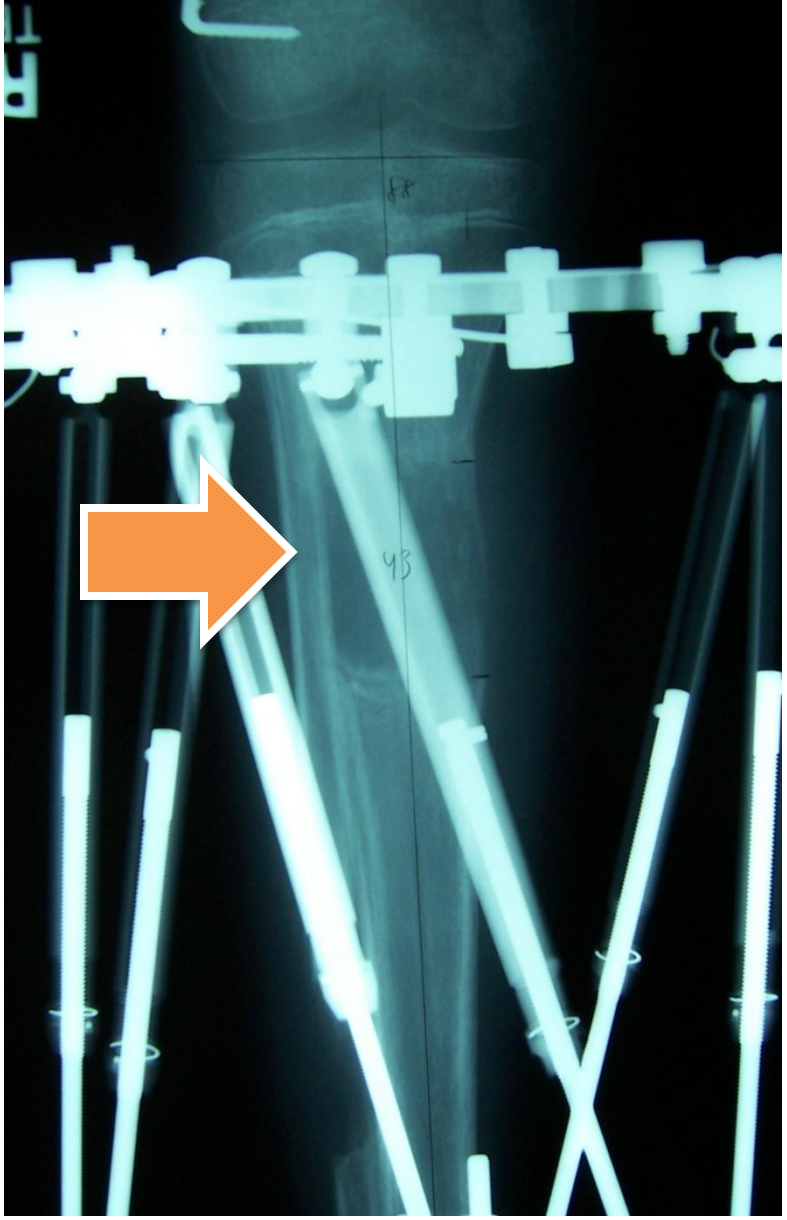

GILLS ROCK
FISH BOIL
DOOR CO.

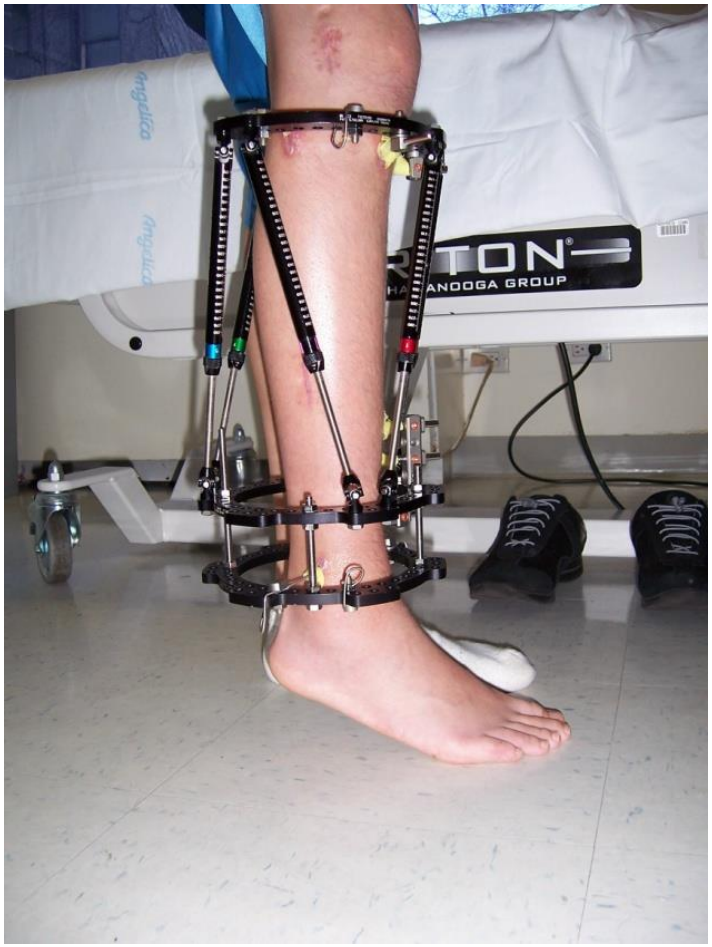

no lifting shoes


STANDING


SPONGEBOB
ATHLETICS


5 cm



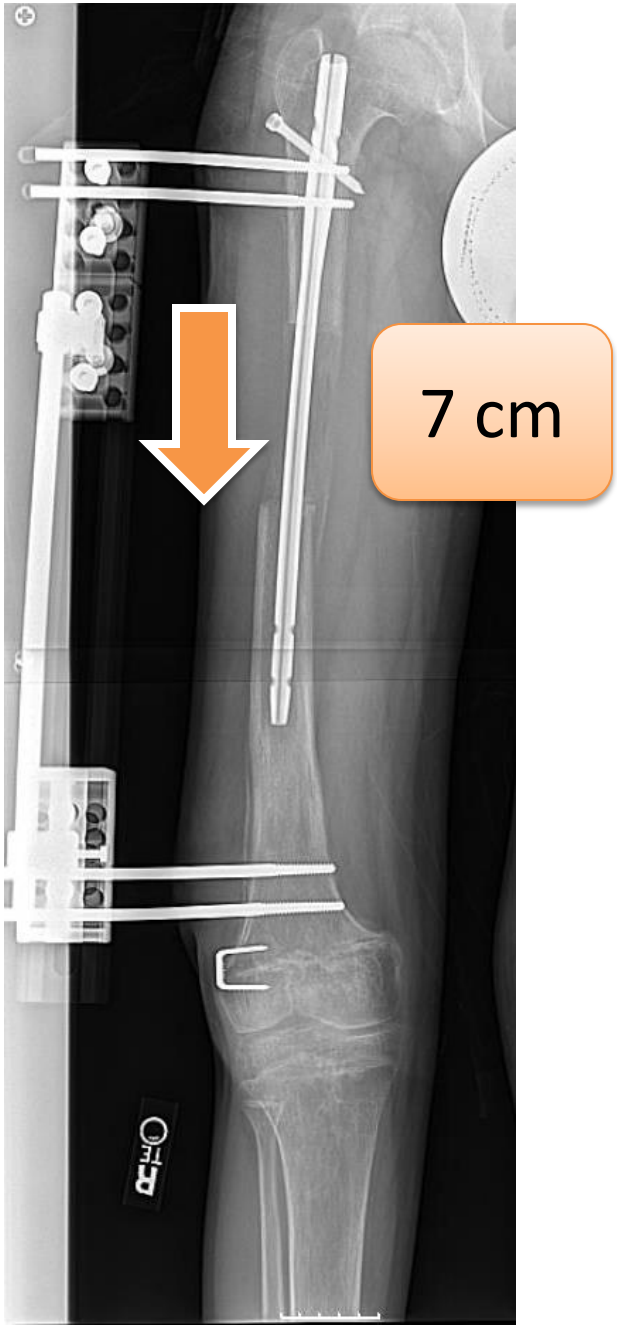

TON
ANOOGA GROUP

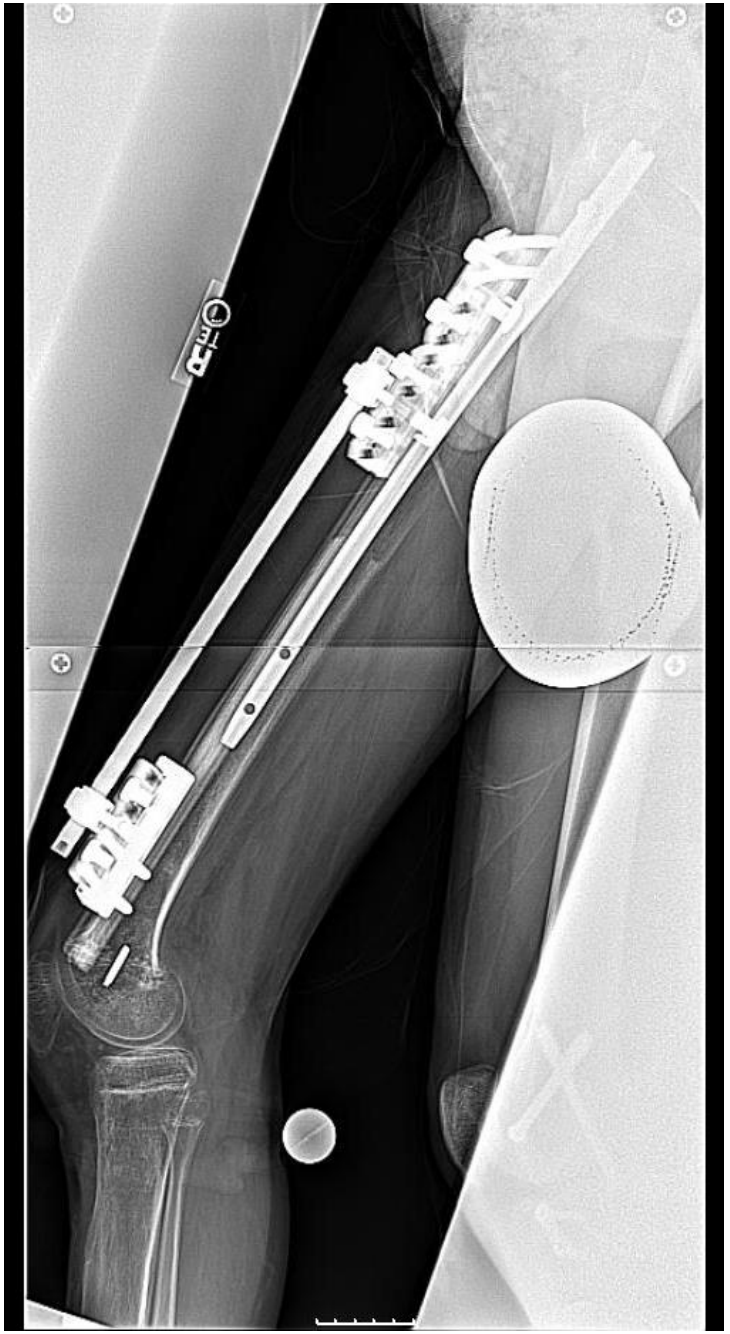

ARMY

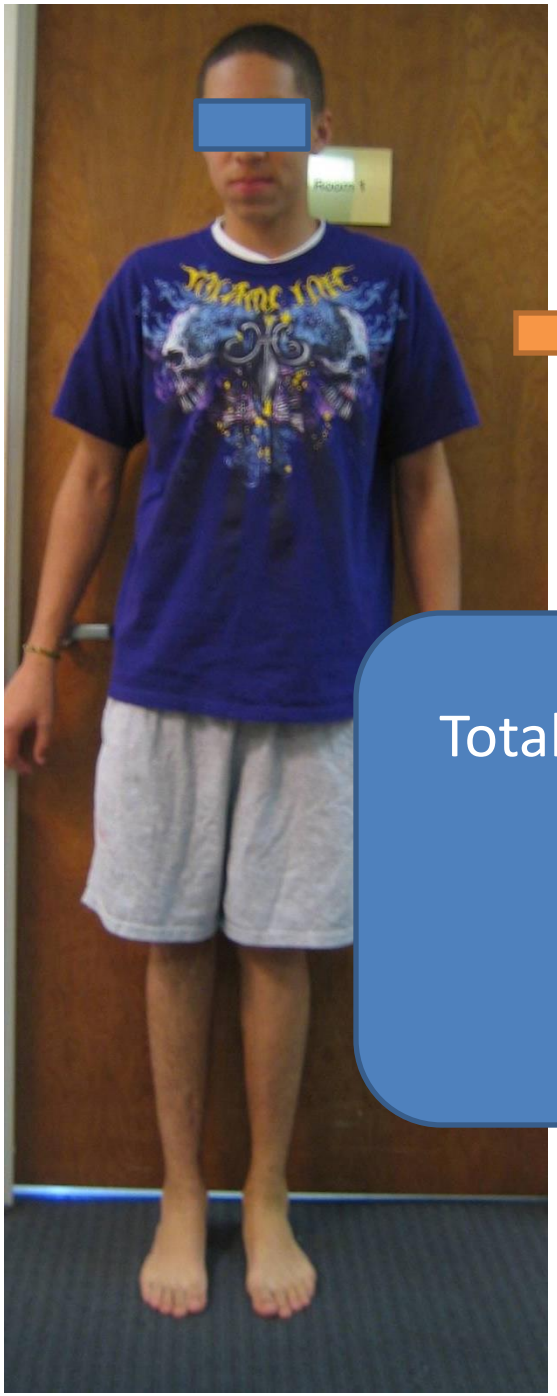

9 cm
SUPINE
R
AD

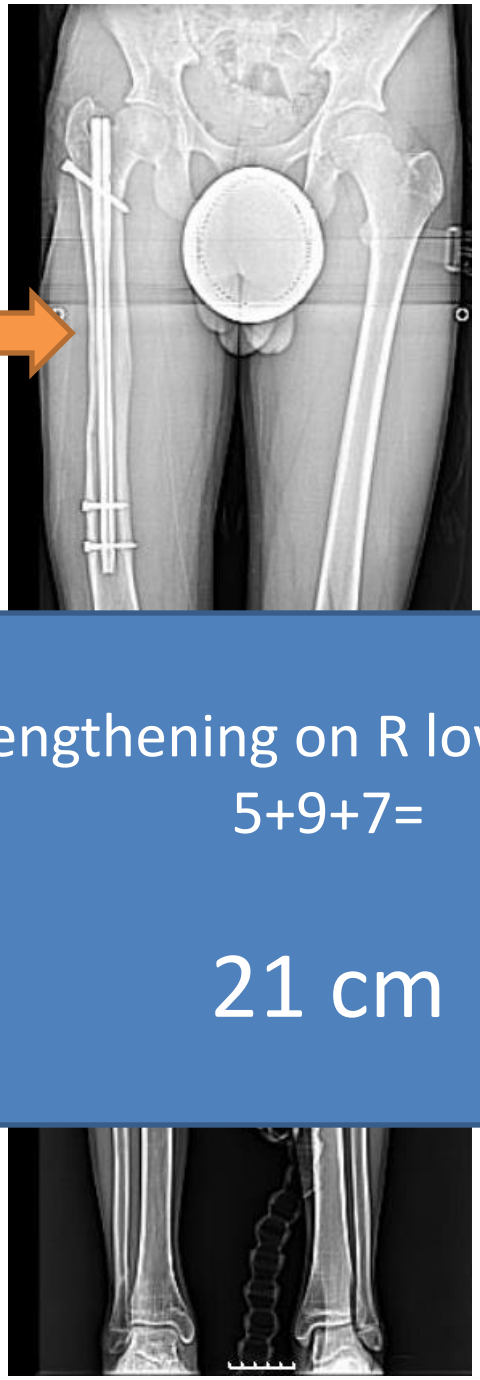

R
J01


NIKE


7 cm

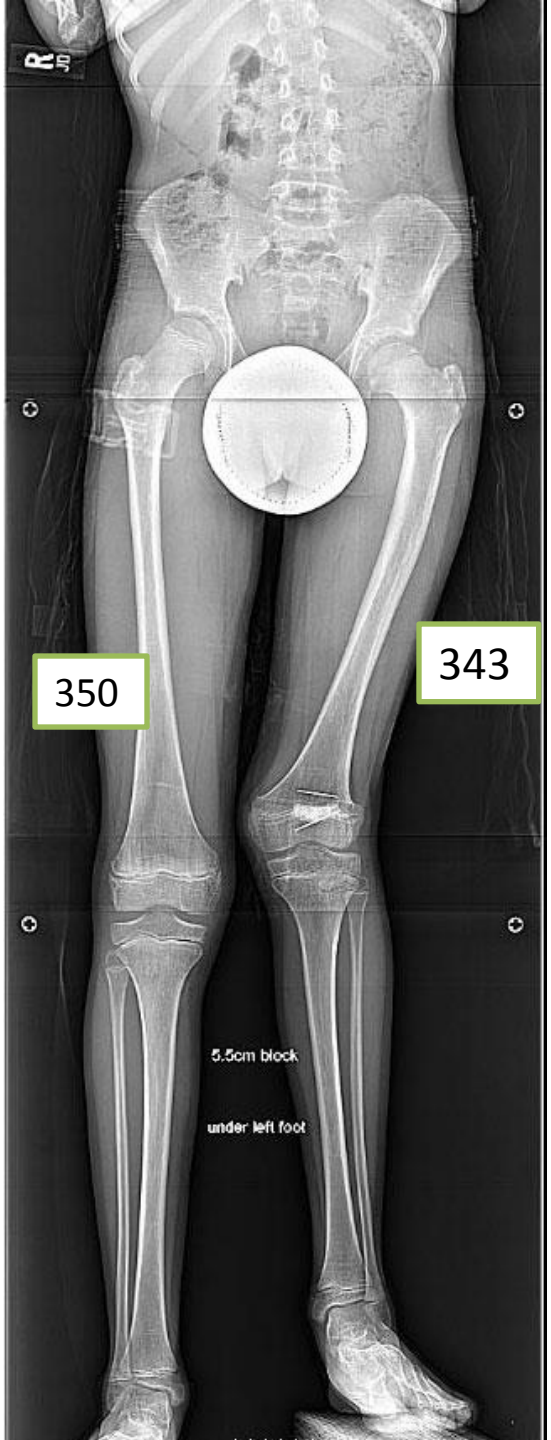






Total Lengthening on R lower extremity
5+9+7=

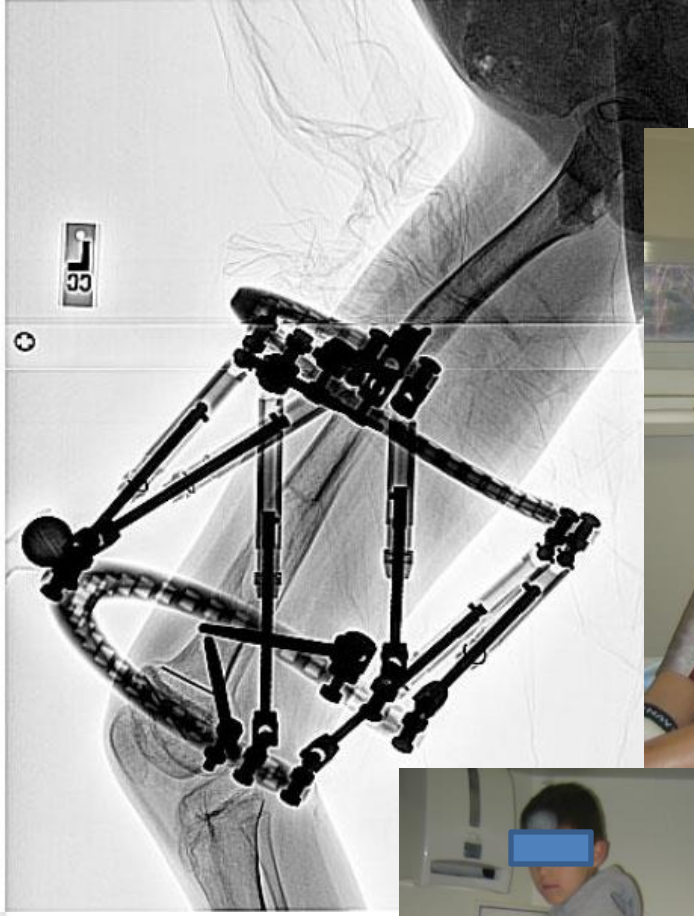
21 cm

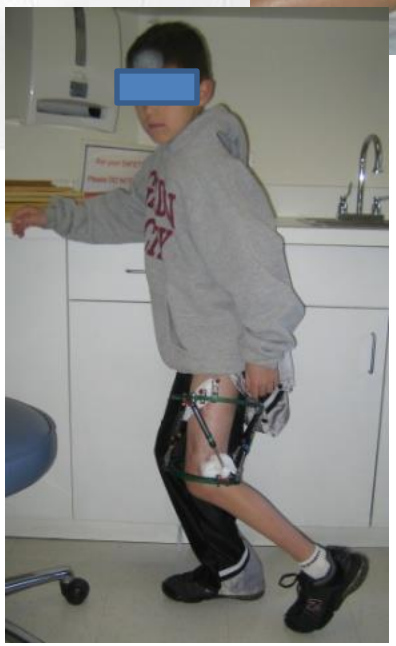


Age 8
Distal femur growth arrest
Proximal tibial also
LLD 7 cm
Valgus deformity

PLLD
M= 1.47
R femur= 350 x 1.47
R femur will be 515
515-350= 165 mm
165 x 70%= 11.5 cm

Plan: lengthen femur
7 cm, correct valgus,
Close growth plate.
Second lengthening of
about 5 cm. femur and / or
tibia



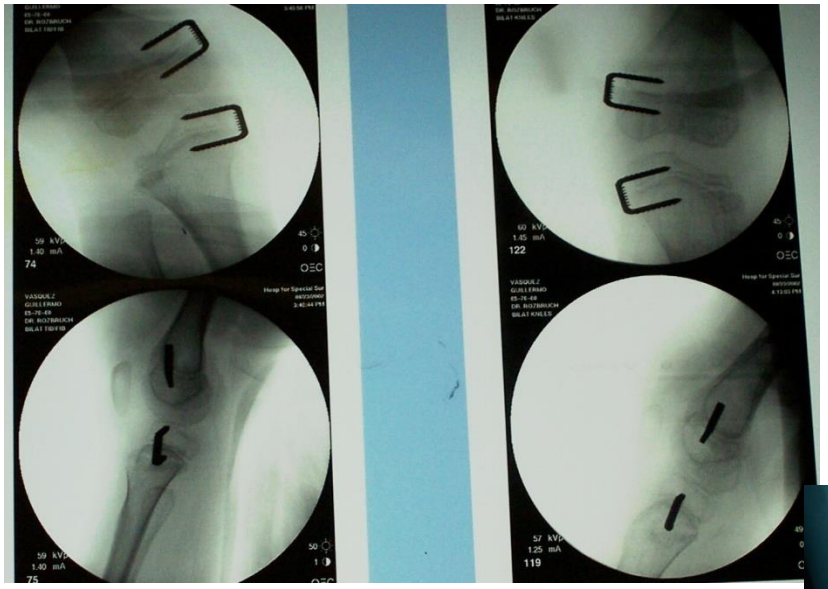



FOOTBALL IS
EVERYTHING
LACROSSE

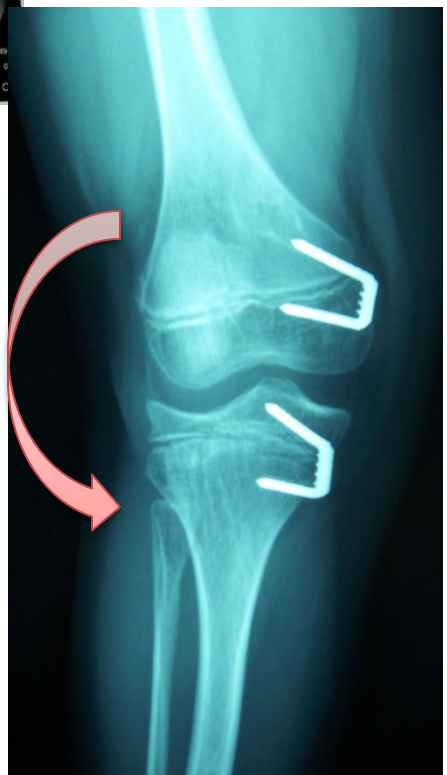

lift/no shoes

Ellis Van Crevald Syndrome

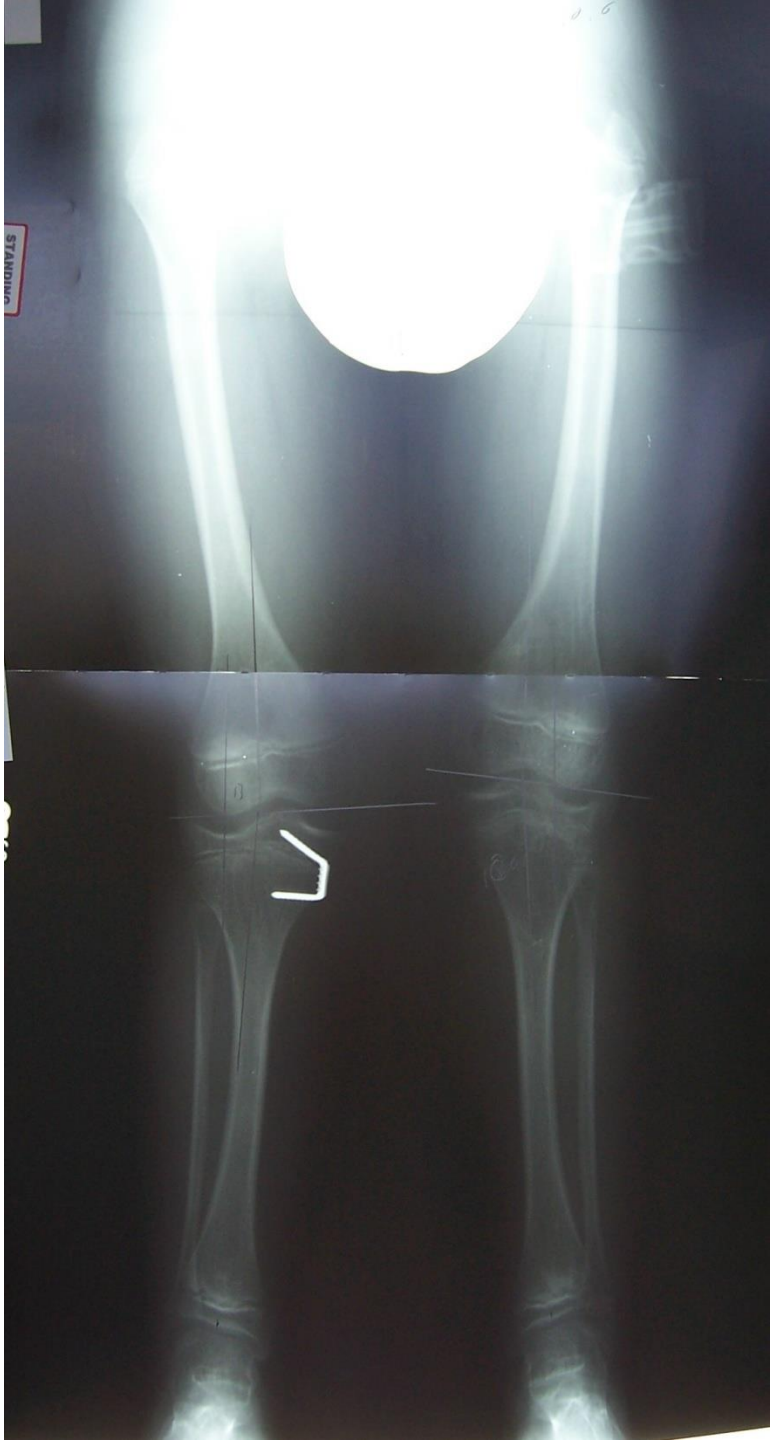
hemiepiphysiodesis



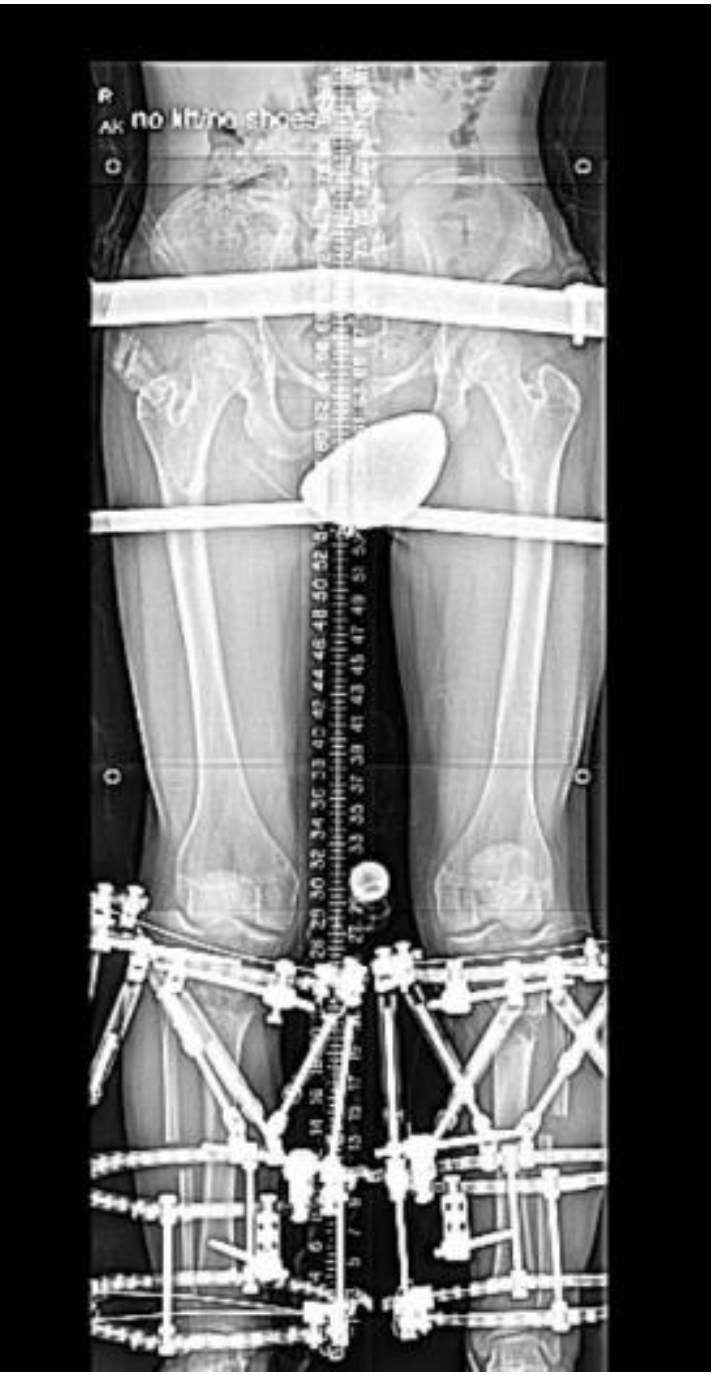
guided growth to
Correct angular deformity

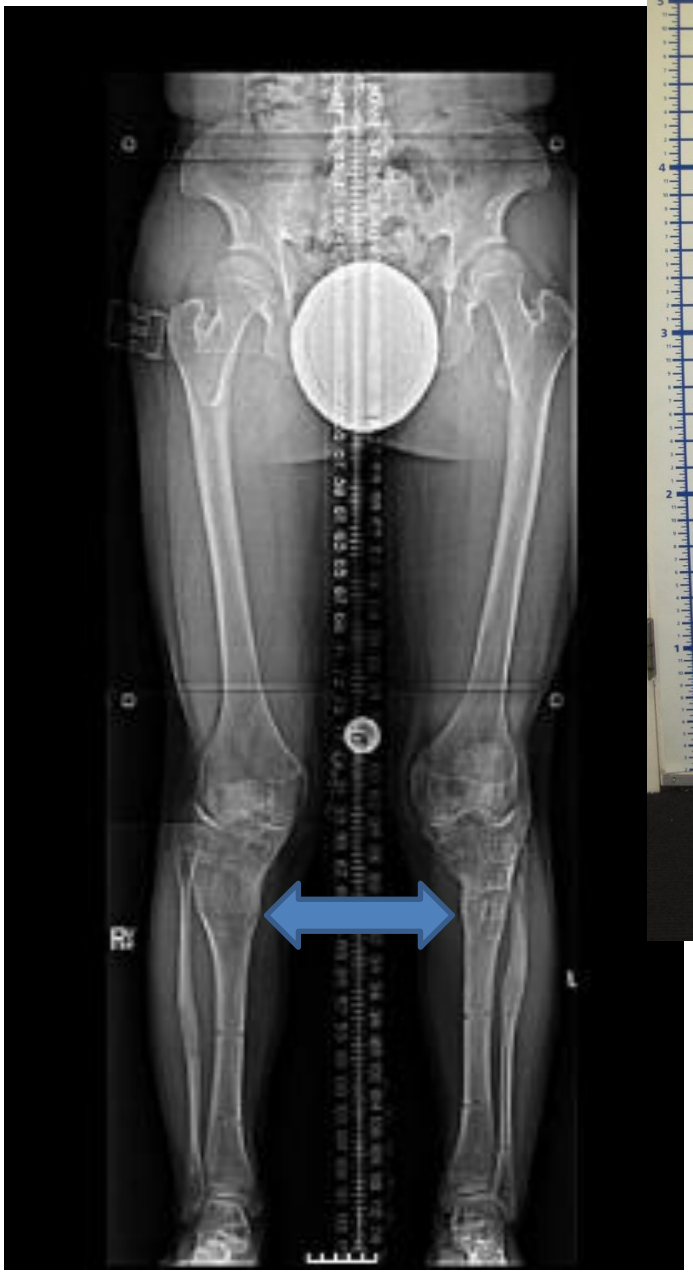



WEST COAST



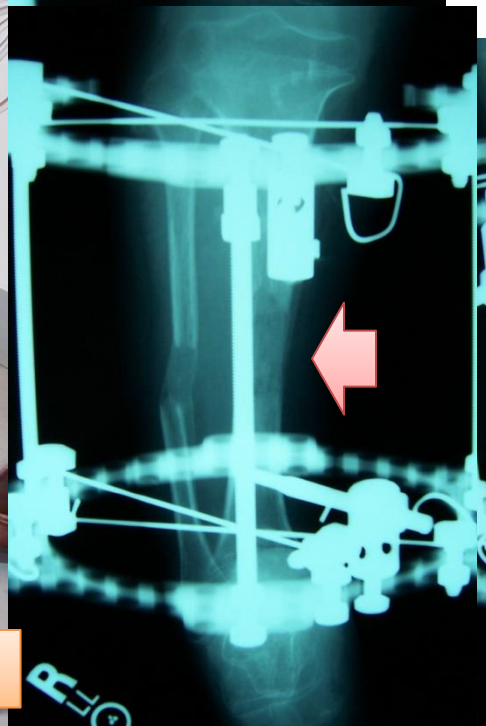
Age 16, short stature, residual deformty

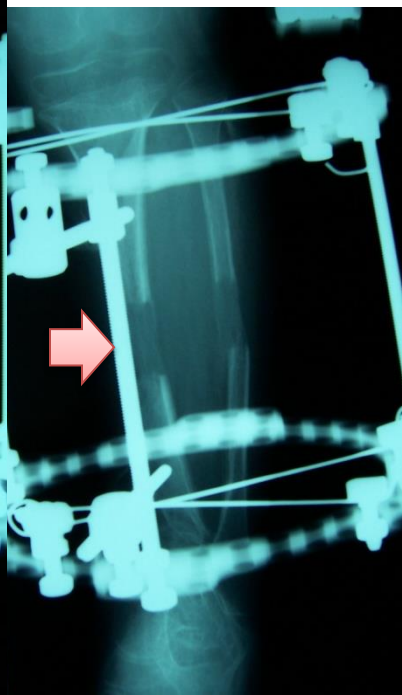

R
AK
no lifting shoes



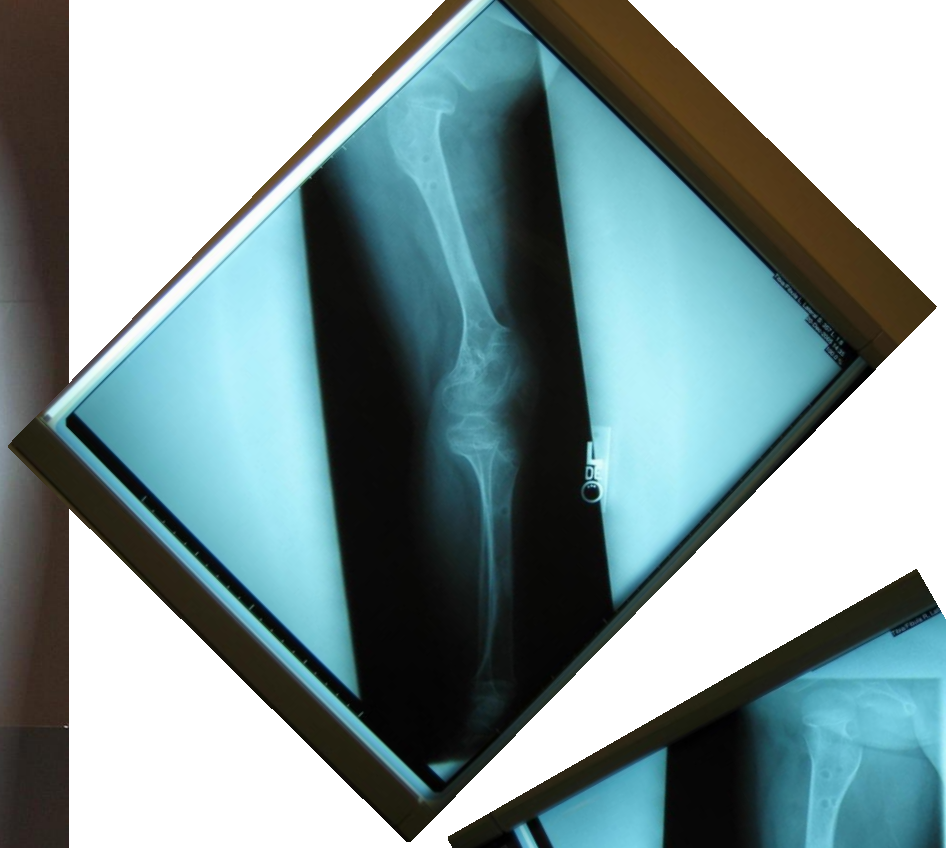

GUILLERMO
2 VIEWS

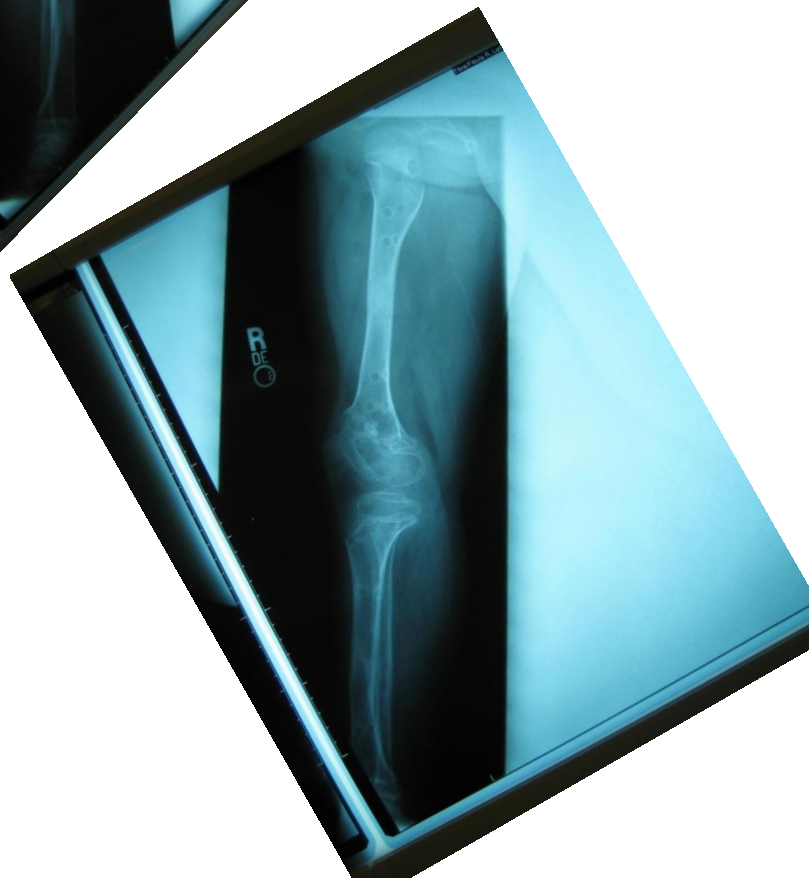
Achondroplasia
Predicted adult height 3’ 11”

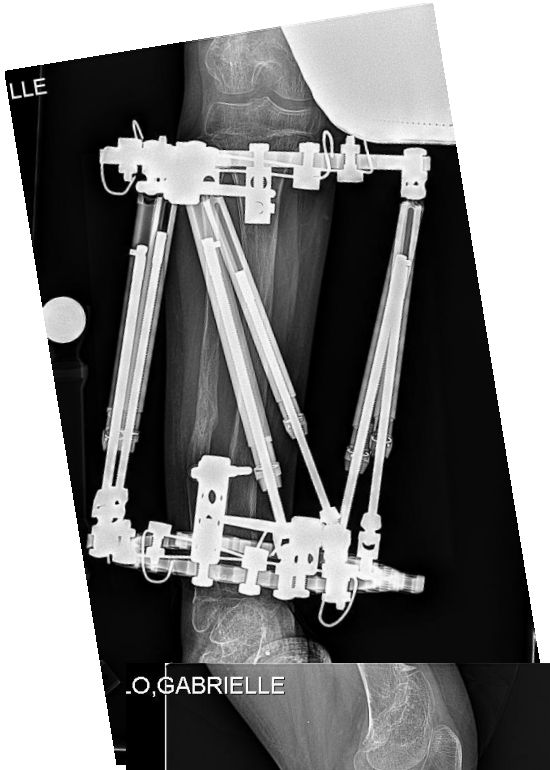


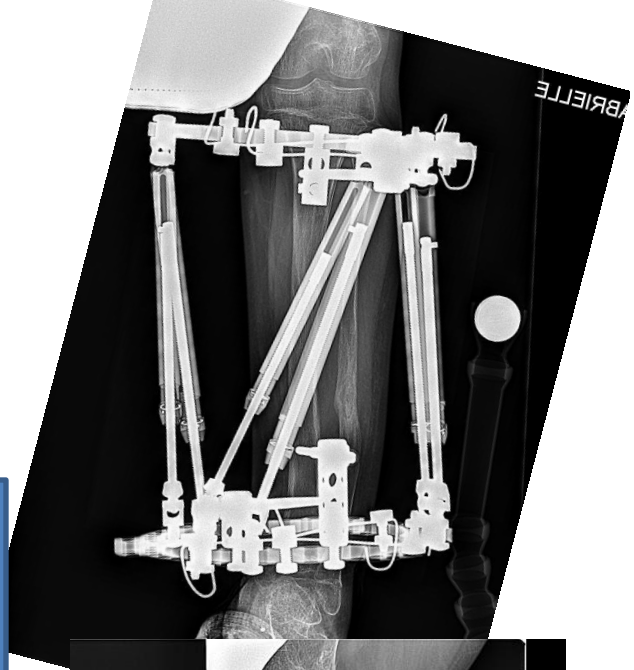


Age 7: B femur and tibia lengthening 5 cm

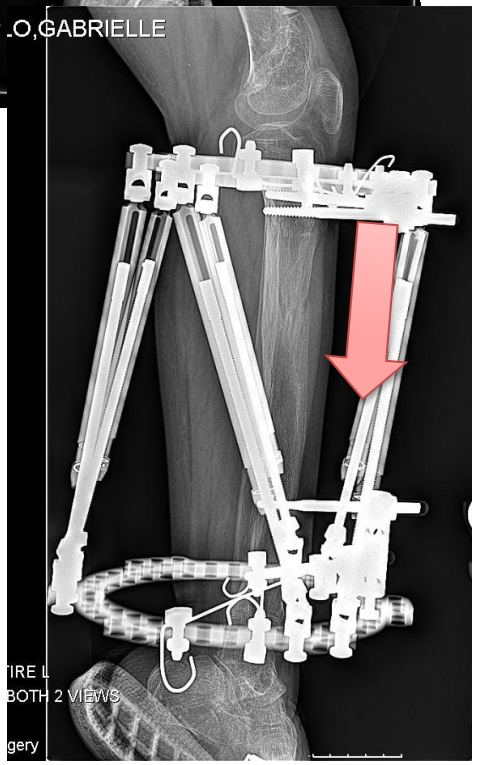


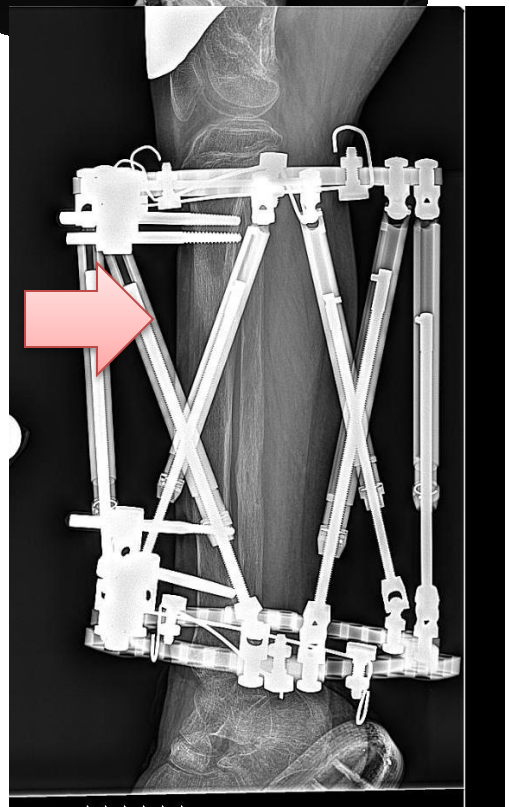






10 cm
Bilateral tibial lengthening
Age 13



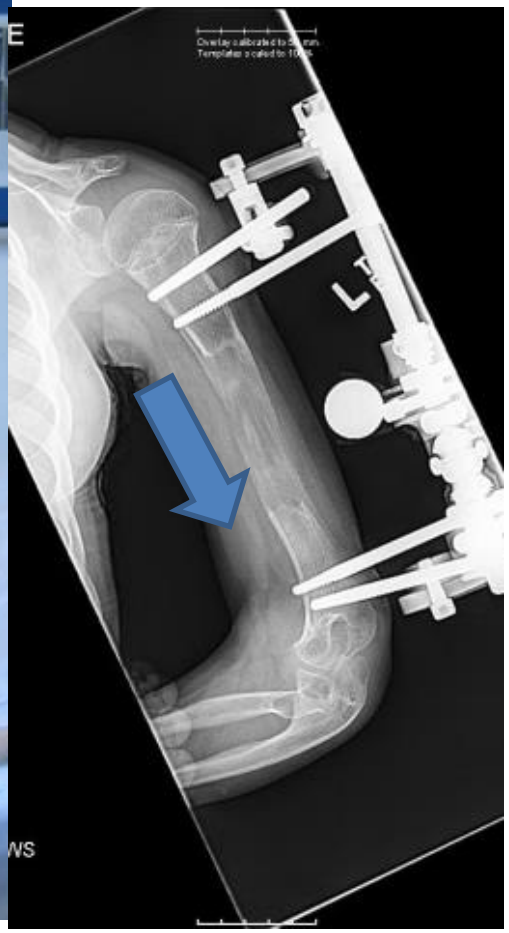


+15 cm in LE's
BUT Arms short


R
OB

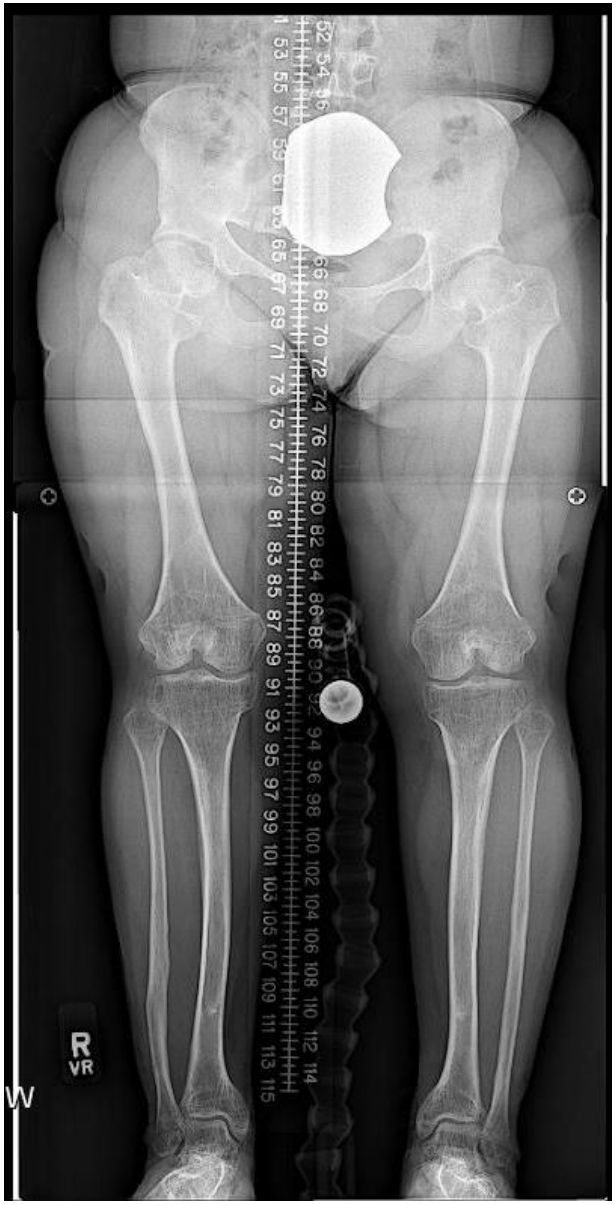

SMGH


Image manually calibrated.
OB



Bilateral Humerus Lengthening 7 cm

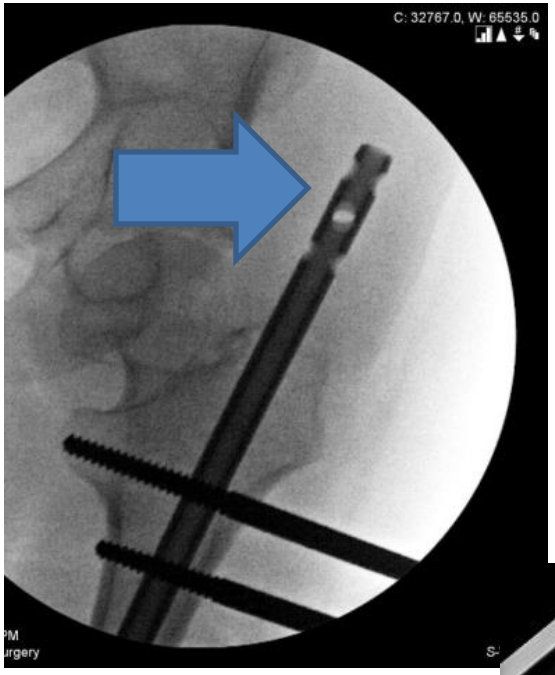

S.M.G.H.
S.M.G.H.
BRIELLE
L
ORE VIEWS



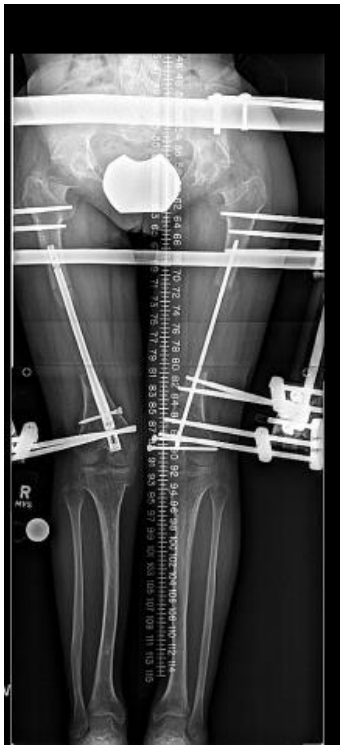
Age 16
Preop for B femur lengthening

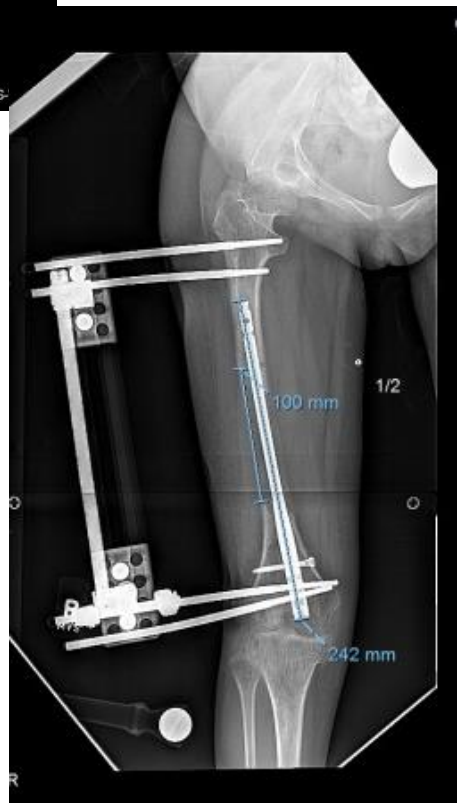
Goal 10 cm
Bone too short for
internal lengthening rod

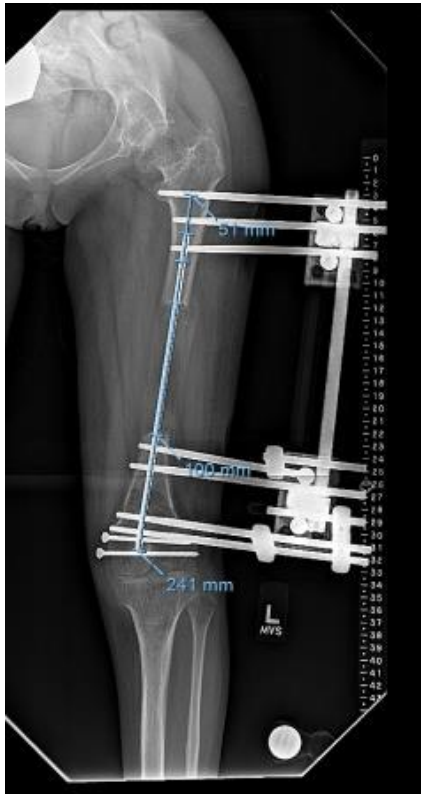
Plan: LON



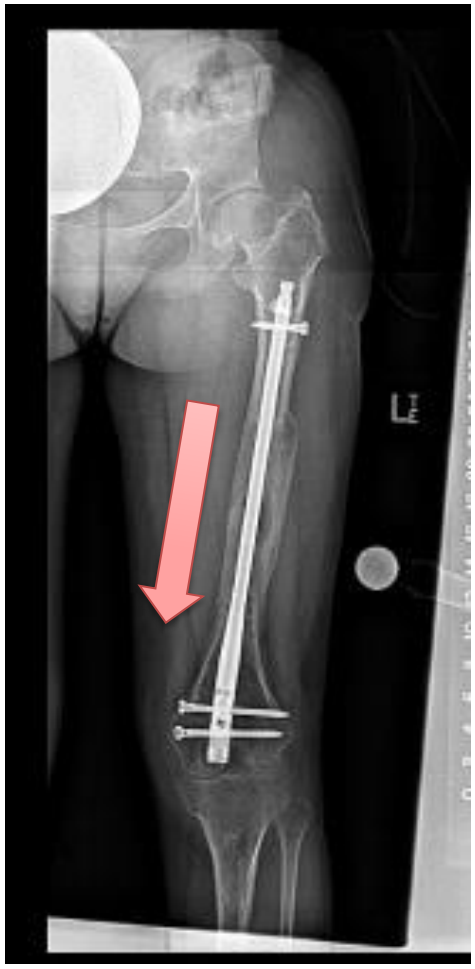
Lengthening over a nail



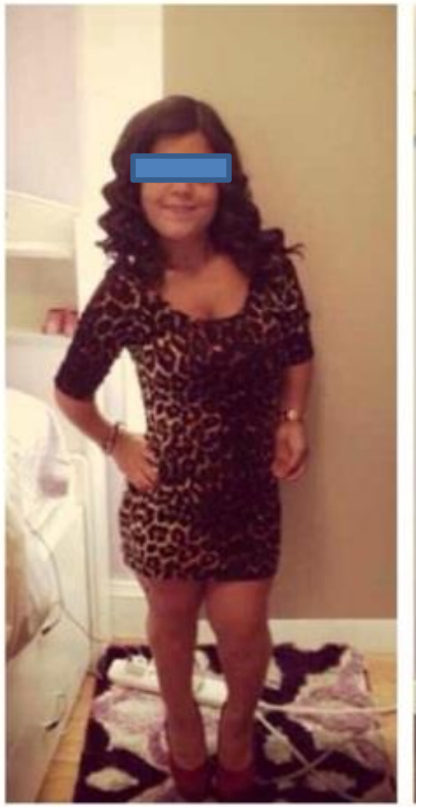









Retrograde tibial nails
Plus 10 cm



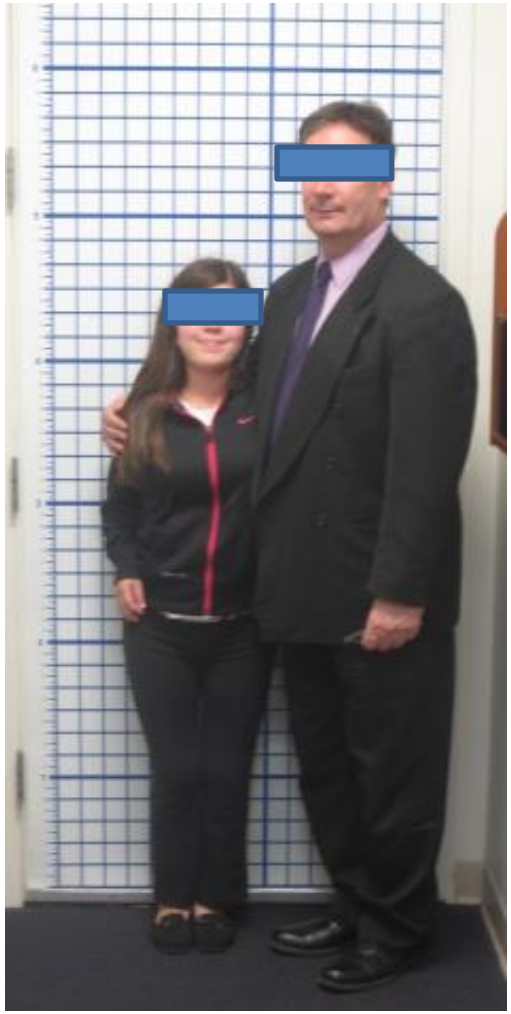
Now 4’ 9”
Age 7: 5cm
Age 13: 10 cm
Age 16: 10 cm
Total: 25 cm height

Age 14: 7 cm arms

Dressed for Prom

Age 7

Age 14



Age 16

Limb lengthening in children with Russell–Silver syndrome: a comparison to other etiologies

V. Goldman, T. H. McCoy, M. D. Harbison, A. T. Fragomen & S. R. Rozbruch

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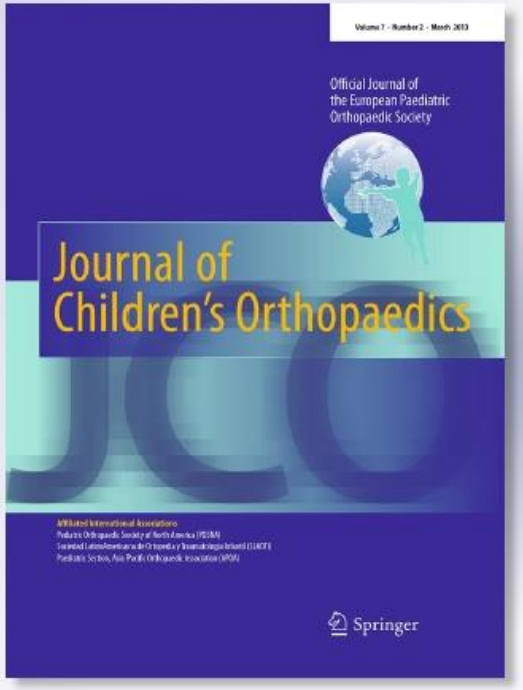
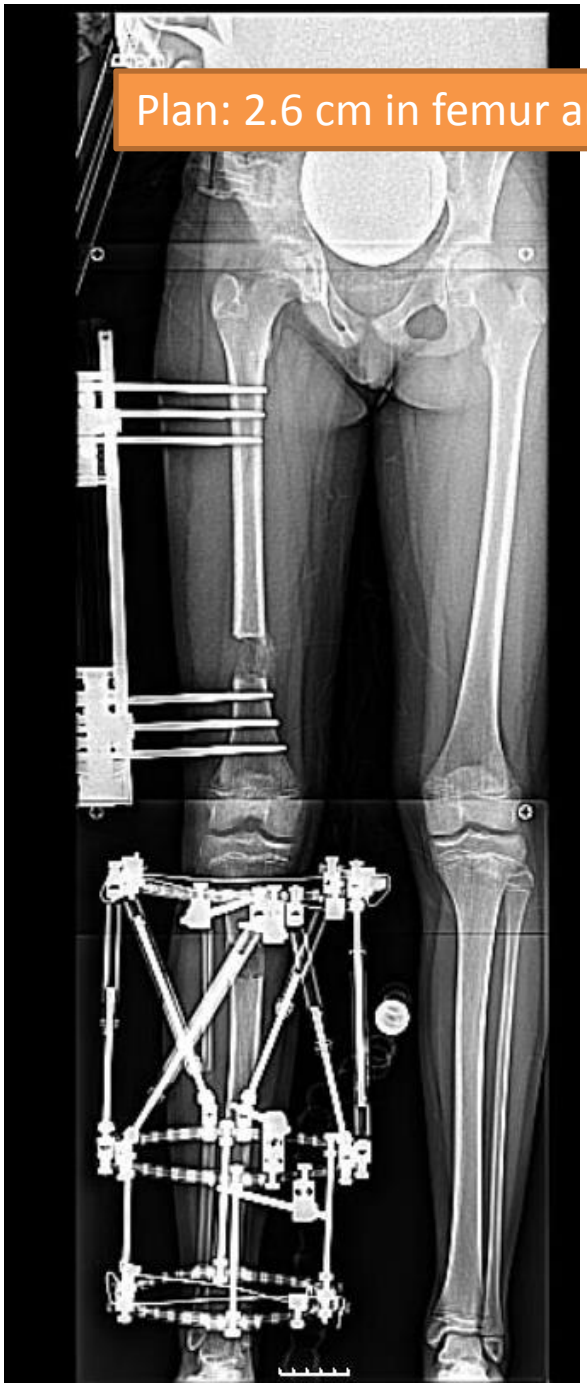
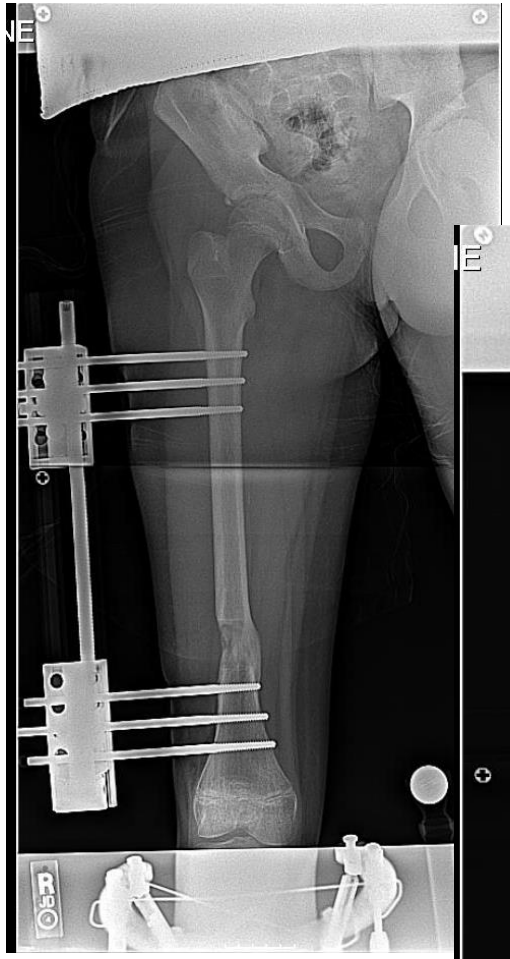


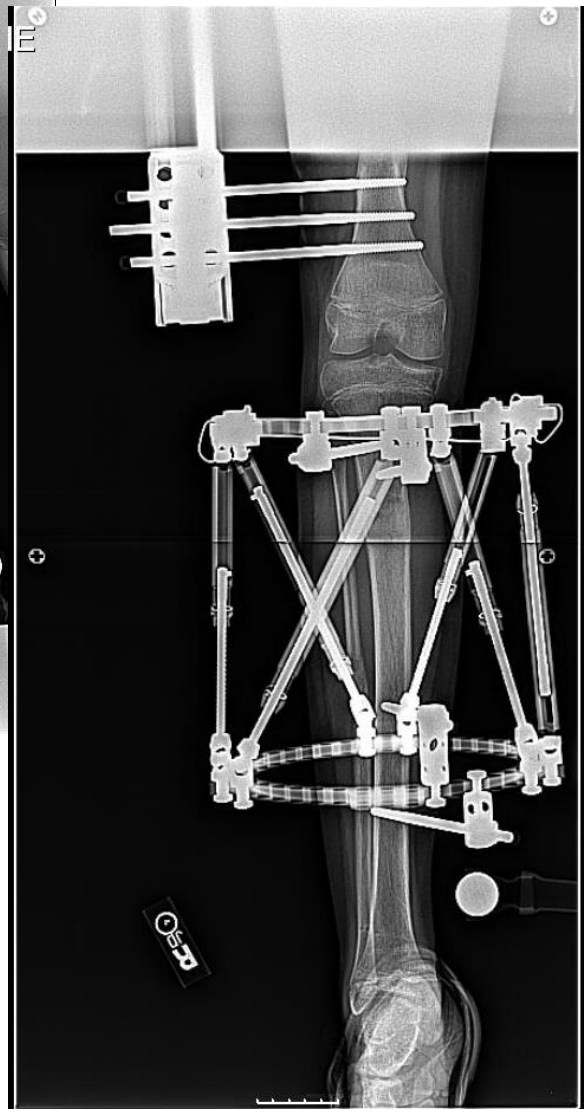
Springer

age 13
LLD 5 cm divided femur/tibia
M= 1.03
PLLD= 5.2 cm

Plan: 2.6 cm in femur and tibia








R
JD
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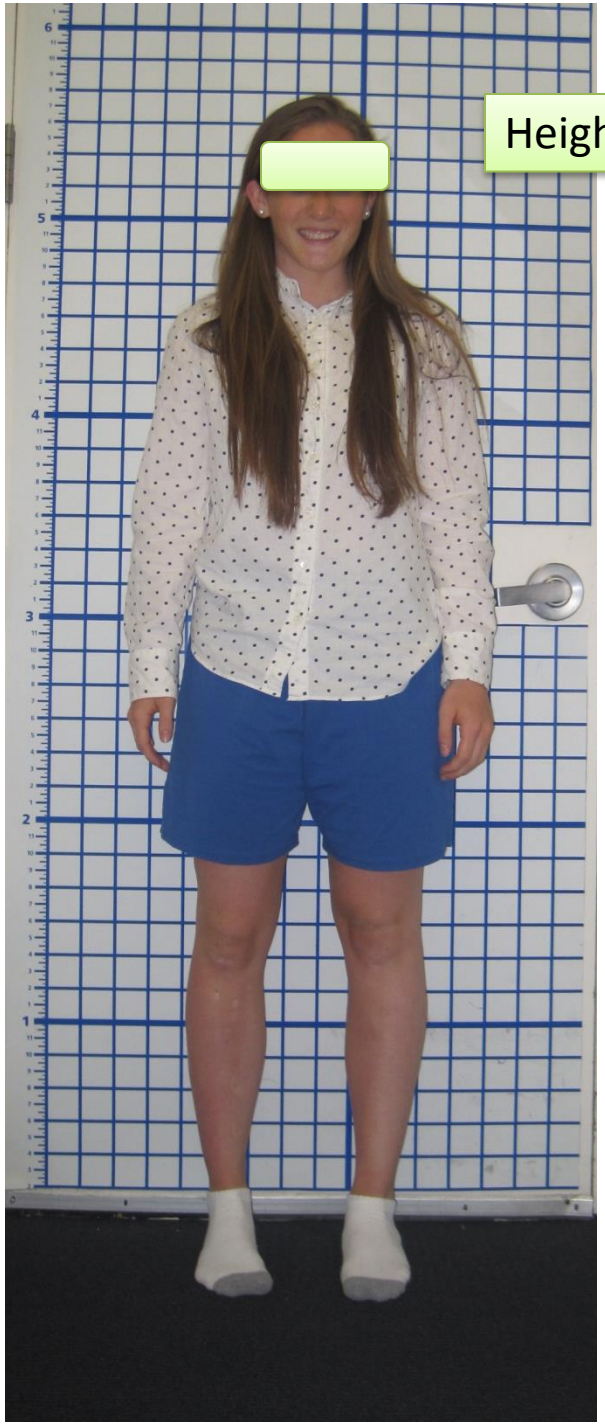


Height 5’ 7”

Introduction

- Russell Silver syndrome (RSS) - rare
- IUGR, difficulty feeding, postnatal growth retardation.
- LLD - more than 90% of patients.
- Bone healing following lengthening is a concern (inadequate caloric intake)
- No specific data published about SRS lengthening
- Short Stature treated with HGH
  - Avoid epiphysiodesis

Growth hormone (GH)

- Abnormalities of GH secretion have been reported in many RSS children
- Human GH treatment benefits - increased linear growth without concomitant increases in LLD (not limited for SRS patients)
- While hGH therapy increases total limb length it does not appear to induce limb specific catch-up growth or reduce the discrepancy between limbs.
- Given the frequency and severity of the LLD associated with RSS (reported average 3.1 cm) many patients will present for limb equalization surgery; however, epiphysiodesis is not a good option

Research question

- We asked whether pediatric patients with RSS (treated with hGH) will have uniformly good bone healing following leg lengthening.

Methods

- Retrospective comparison
- Study group - SRS patients with LLD - lengthening while on GH
- Control group – general pediatric lengthening patients (congenital, post-traumatic, tumor)

Methods

- 7 limb segments in 5 patients with RSS
- 21 segments in 19 patients – Control

Posttraumatic 8/7

Congenital 9/8

Tumor 4/4

Methods

	SRS	Control	P value
Age (years)	10.4	13	0.036
Lengthening (cm)	3.3	3.9	0.507
Follow up (months)	32 (16-38)	58 (12-130)	

Bone Healing Index

days of bone healing per cm of lengthening

	SRS	Control	P value
Bone Healing Index (BHI), days/cm	29	43	0.028

RSS patients had significantly faster bone healing during limb lengthening

Discussion

Function limiting LLD vs. concern about bone healing.

Scarcity of literature on SRS lengthening

hGH has known positive effect on fracture healing, not well documented for human limb lengthening

Recent animal studies also showed that GH improved muscle recovery during limb lengthening

Discussion

- All SRS patients had good outcome,

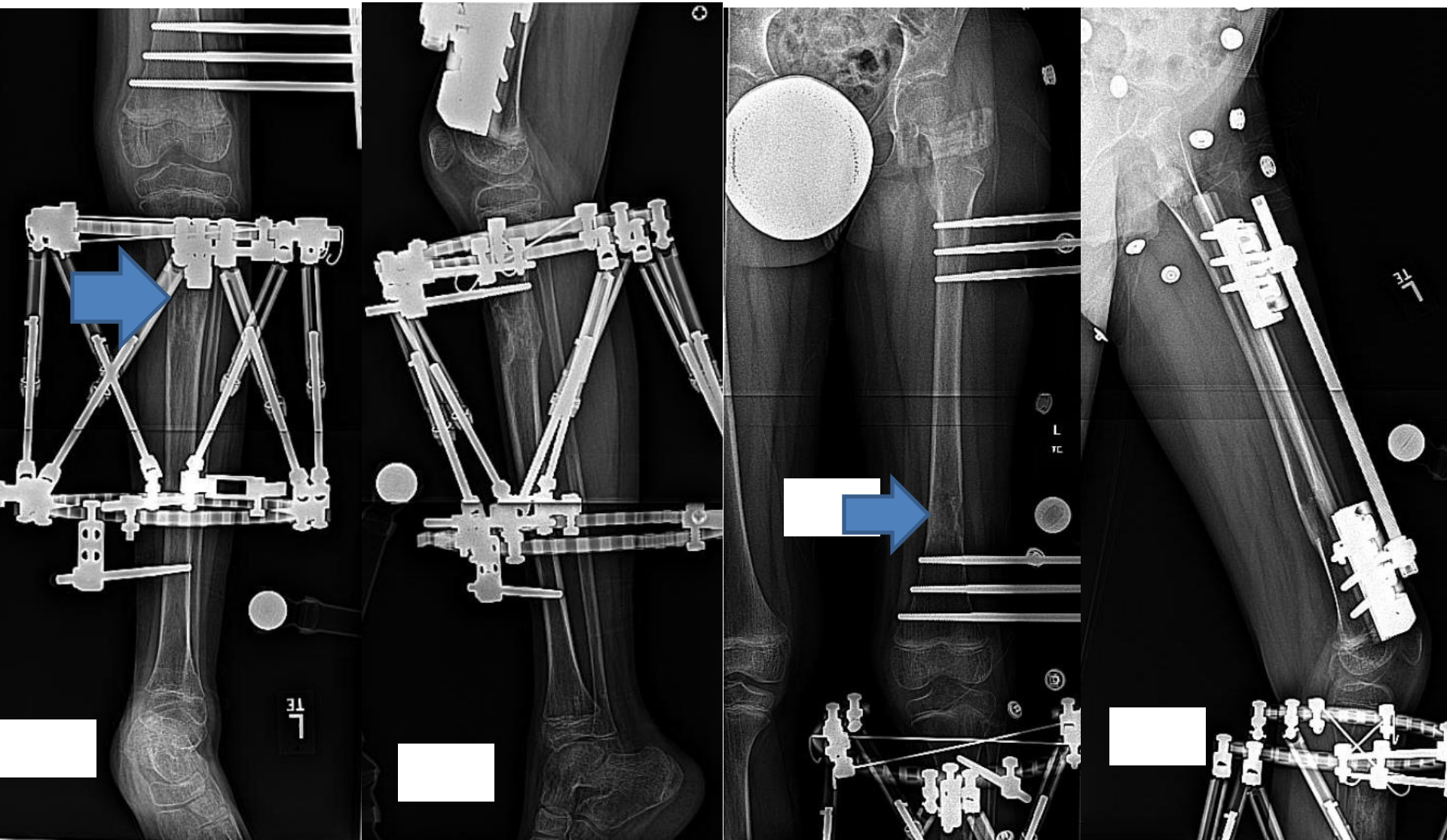
no significant problems

- No premature consolidation on hGH
- No hGH-related orthopedic complications (LCP, SCFE, scoliosis)

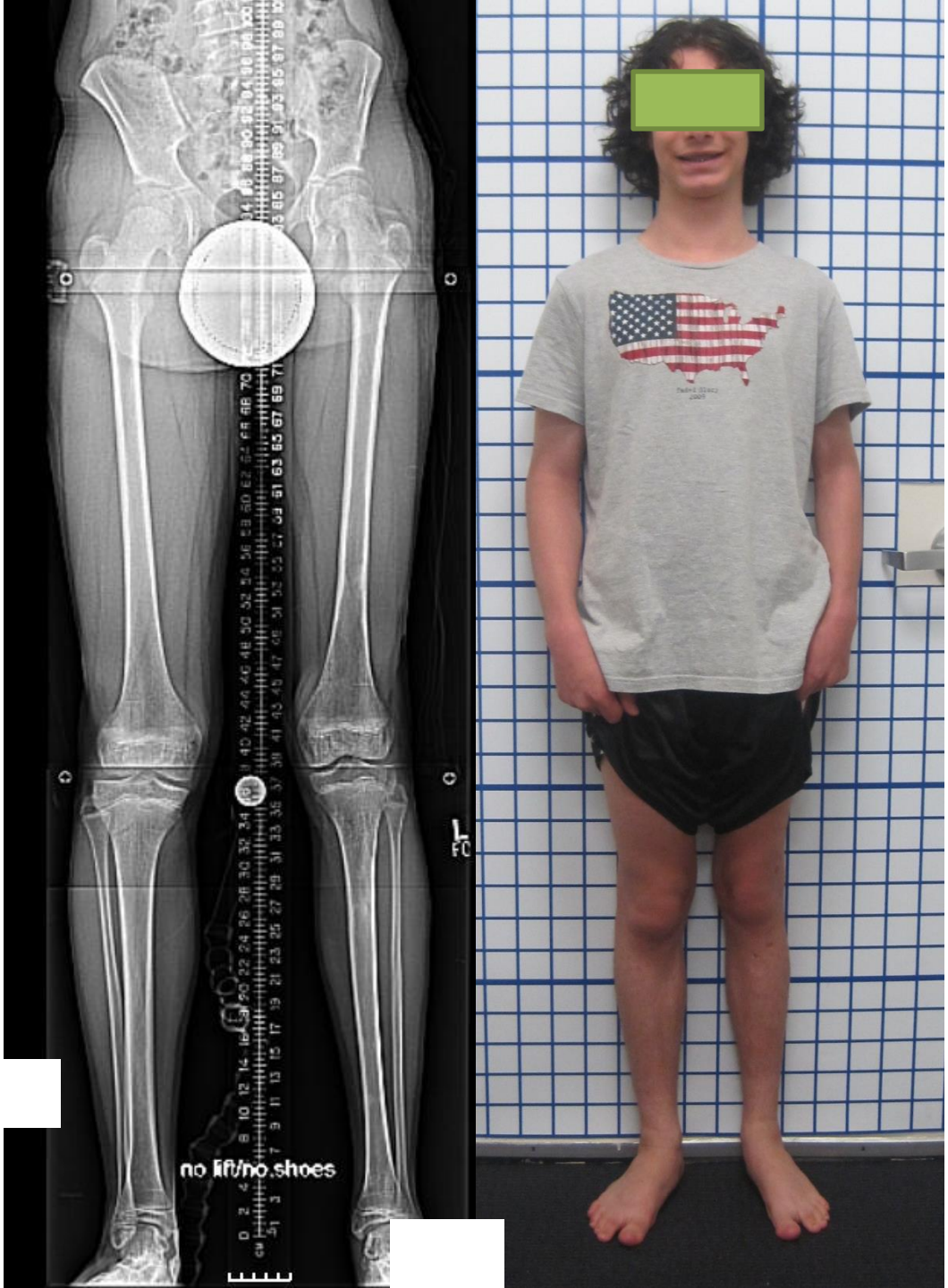
Conclusion

- SRS patients treated with hGH -uniformly good healing of bone regenerate
- SRS BHI is significantly shorter than in a general pediatric population.
- hGH may significantly improve regenerate formation and consolidation

Age 12 yo
LLD 39 D/45 ID mm
Δ F 27 mm
Δ T 18 mm
PLLD ~ 5.3 cm




new balance
621
new balance
621

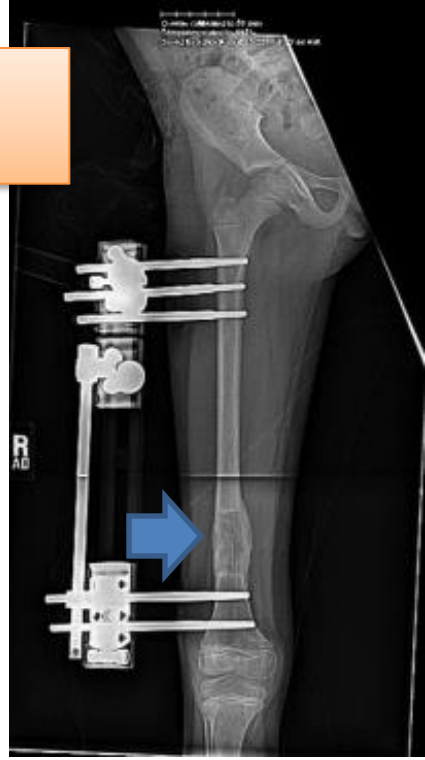

no lift/no shoes

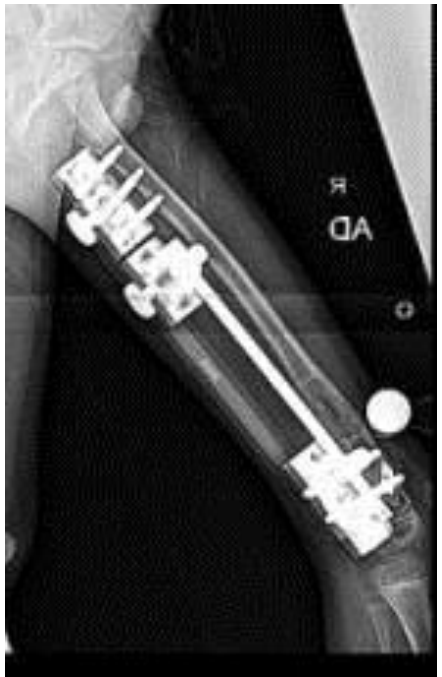
RSS, age 8
LLD 4 cm divided femur/tibia
M= 1.33
PLLD= 5.3

Puberty will be delayed and on HGH
PLLD will be greater (6-7 cm)
Lengthen tibia 4 cm to correct LLD
Lengthen femur in future

Age 6, congenital
LLD 4.2 cm mostly femur
M= 1.68
PLLD= 7 cm

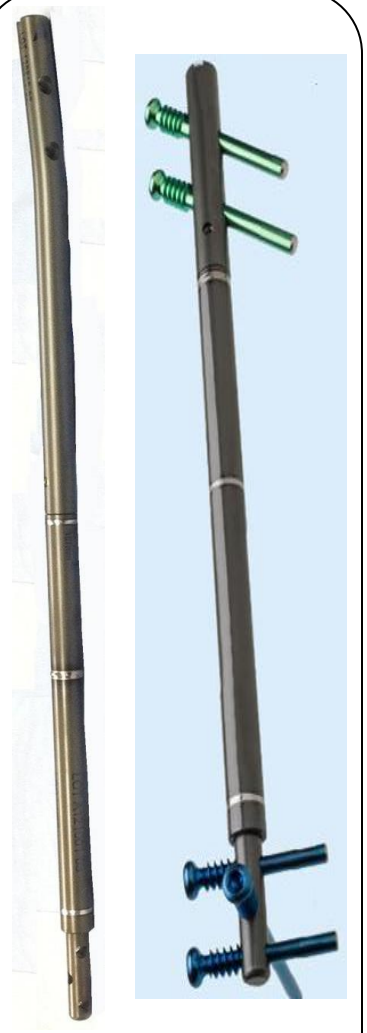
Plan: lengthen femur 4 cm
Lengthen tibia in future





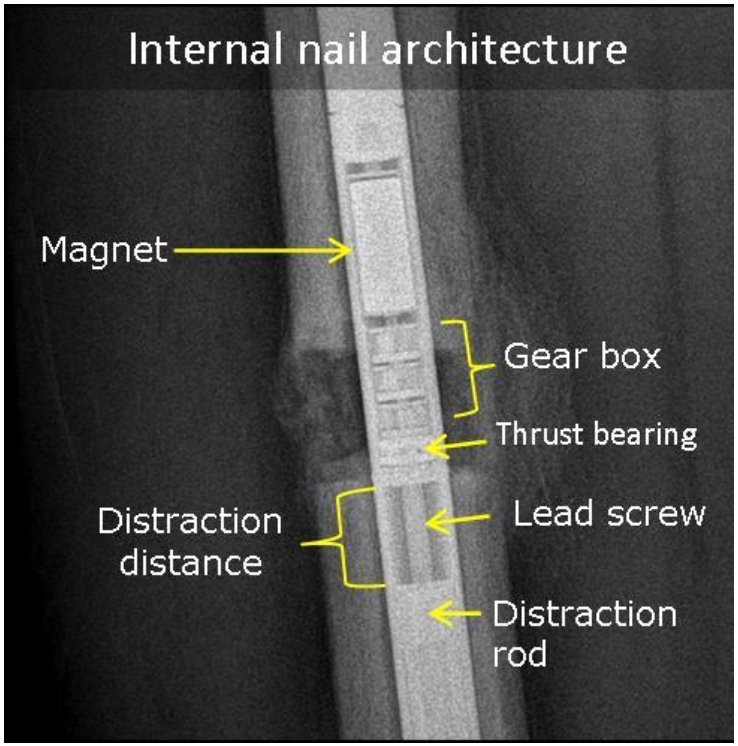

2 years later
With emerging
LLD
618 mm
629 mm

Precice® Nail



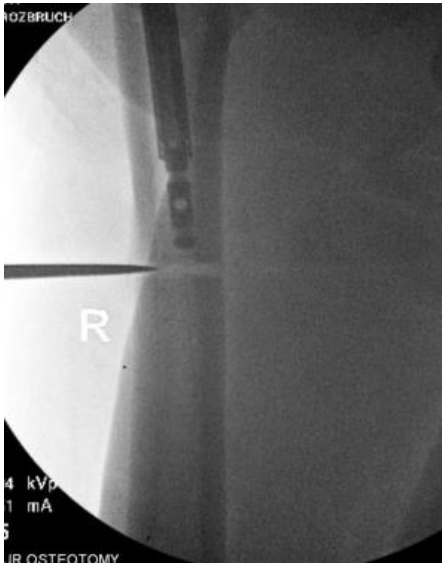
Precice® Nail

- Telescopic, magnet-operated device




STITCH
NK 1 VIEW
rgery


Glen
C: 32767.0
OEC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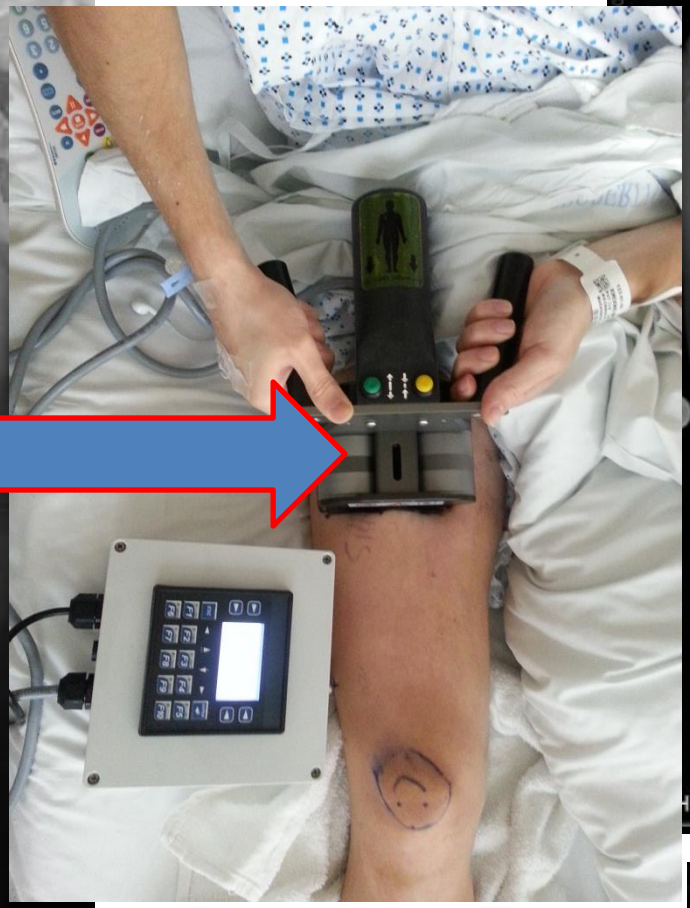
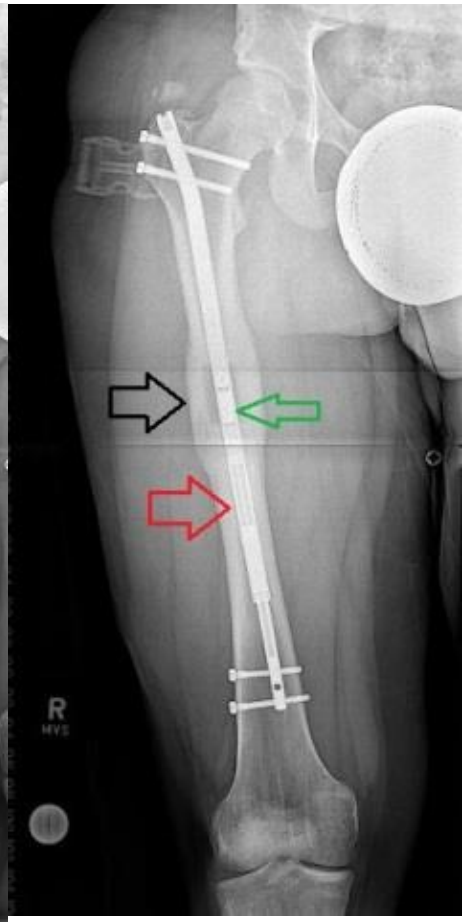
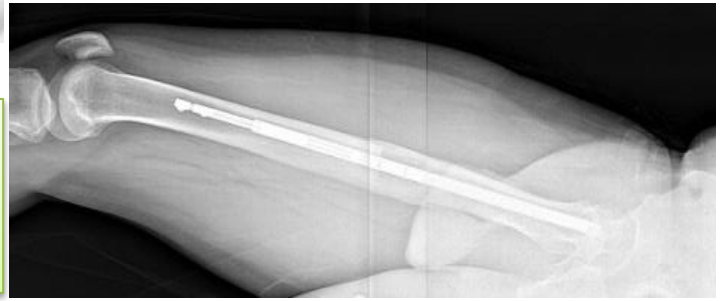
STEOTOMY
:49:49 AM
ecial Surgery

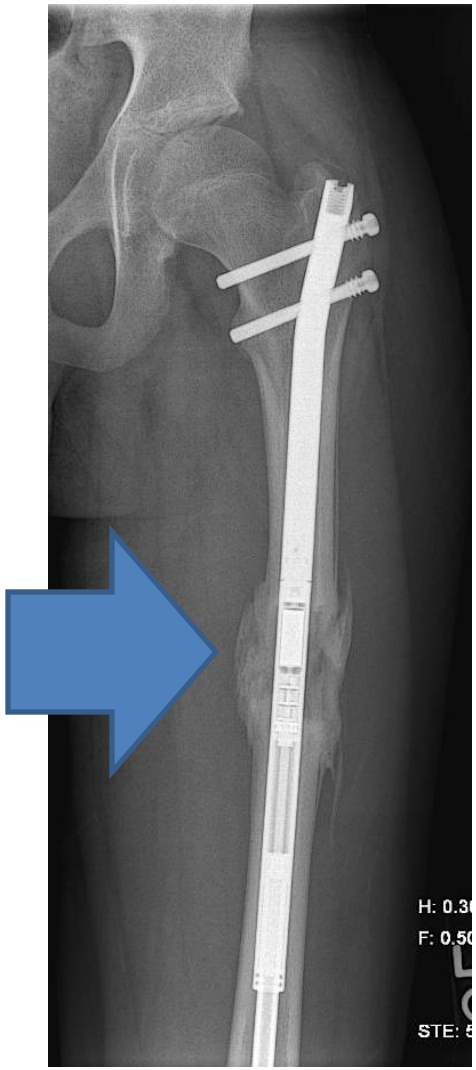




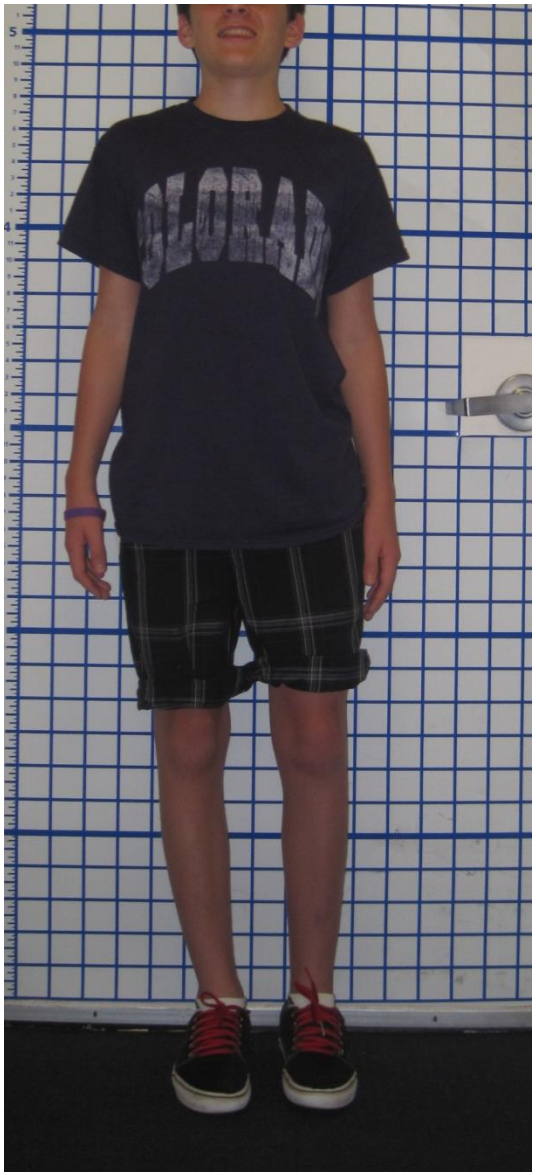


Trochanteric Entry
Good for adolescent
< 18 yrs.

12 year old male with congenital LLD




LEFT
H: 0.30
F: 0.50

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Clin Orthop Relat Res
DOI 10.1007/s11999-014-3575-0

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SYMPOSIUM: 2013 LIMB LENGTHENING AND RECONSTRUCTION SOCIETY

Precision of the PRECICE® Internal Bone Lengthening Nail

Yatin M. Kirane MBBS, DOrtho, PhD,
Austin T. Fragomen MD, S. Robert Rozbruch MD

Abstract

Background Previous designs of internal bone lengthening devices have been fraught with imprecise distraction, resulting in nerve injuries, joint contractures, nonunions, and other complications. Recently, a magnet-operated PRECICE® nail (Ellipse Technologies, Inc, Irvine, CA, USA) was approved by the FDA; however, its clinical efficacy is unknown.

Questions/purposes We evaluated this nail in terms of (1) accuracy and precision of distraction, (2) effects on bone alignment, (3) effects on adjacent-joint ROM, and (4) frequency of implant-related and non-implant-related complications.

Methods We reviewed medical and radiographic records of 24 patients who underwent femoral and/or tibial lengthening procedures using the PRECICE® nail from August 2012 to July 2013 for conditions of varied etiology, the most common being congenital limb length discrepancy, posttraumatic growth arrest, and fracture malunion. This group represented 29% of patients (24 of 82) who underwent a limb lengthening procedure for a similar diagnosis during the review period. At each postoperative visit, the accuracy and precision of distraction, bone alignment, joint ROM, and any complications were recorded by the senior surgeon (SRR). Accuracy reflected how close the measured lengthening was to the prescribed distraction at each postoperative visit, while precision reflected how close the repeated measurements were to each other over the course of total lengthening period. No patients were lost to followup. Minimum followup from surgery was 3 weeks (mean, 14 weeks; range, 3–29 weeks).

Results Mean total lengthening was 35 mm (range, 14–65 mm), with an accuracy of 96% and precision of 86%. All patients achieved target lengthening with minimal unintentional effects on bone alignment. The knee and ankle ROM were minimally affected. Of the complications requiring return to the operating room for an additional surgical procedure, there was one (4%) implant failure caused by a nonfunctional distraction mechanism and six (24%) non-implant-related complications, including premature consolidation in one patient (4%), delayed bone healing in two (8%), delayed equinus contracture in two (8%), and toe clawing in one (4%).

Conclusions We conclude that this internal lengthening nail is a valid option to achieve accurate and precise limb lengthening to treat a variety of conditions with limb shortening or length discrepancy. Randomized, larger-sample, long-term studies are required to further confirm

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All ICMJE Conflict of Interest Forms for authors and *Clinical Orthopaedics and Related Research®* editors and board members are on file with the publication and can be viewed on request.
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Each author certifies that his or her institution approved the human protocol for this investigation, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.

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- These were first 24 patients (August 2012-July 2013)
- 29% of patients who underwent limb lengthening surgery during that time period

Kirane Y, Fragomen AT, Rozbruch SR: Precision of the Precice Internal Lengthening Nail, Clin Orthop Relat Res. 2014 Dec;472(12):3869-78

Primary Outcome Variables

I. Accuracy of Lengthening

– Distraction distance & accuracy measured using a calibrated digital radiology system (PACS, OnePacs LLC, New York, NY)

$$A)\ \%\ Error = \frac{Distraction\ prescribed\ -\ Lengthening\ measured}{Distraction\ prescribed} \times 100$$

$$B)\ Accuracy\ of\ distraction\ =\ 100 - \%\ Error$$

II. Change in bone alignment

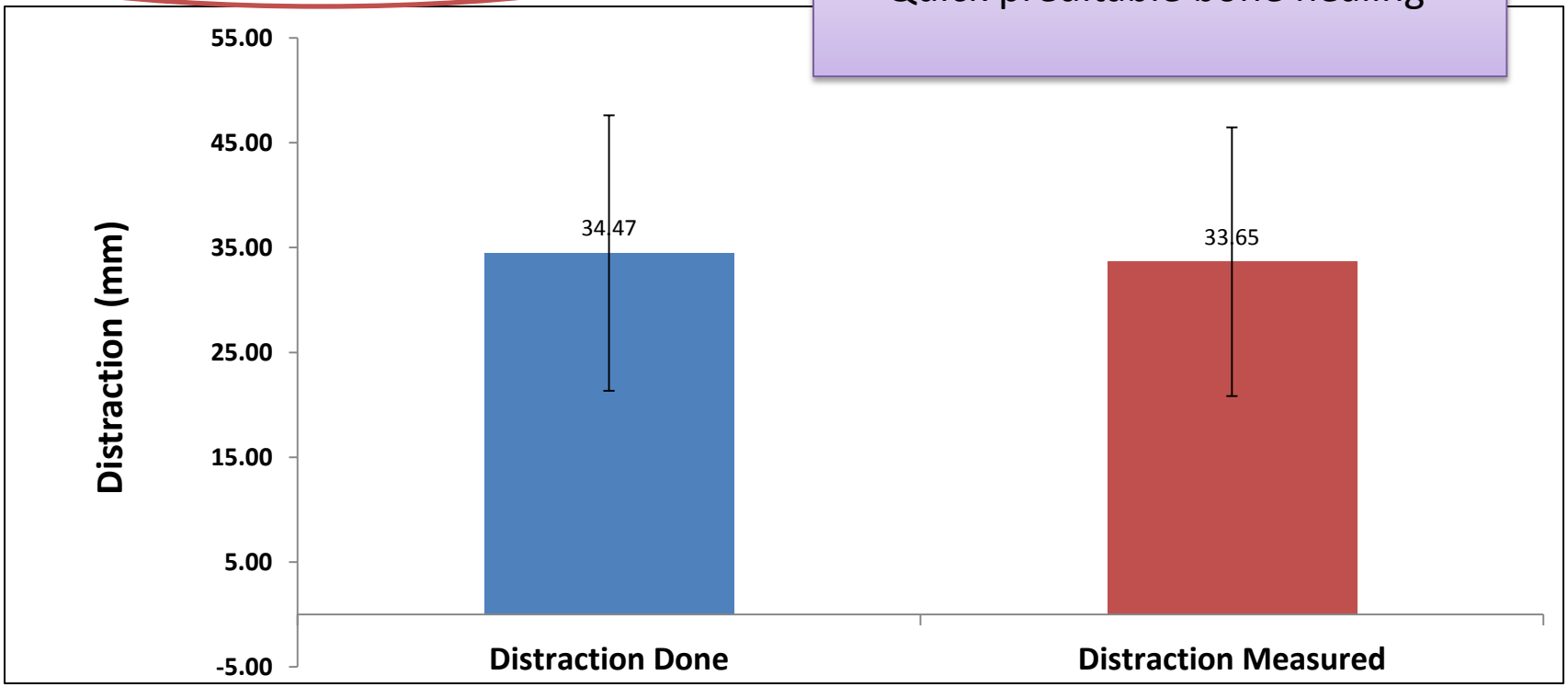
III. Effect on adjacent joint ROM

I. Accuracy of Lengthening

At 19 weeks follow-up (range, 1-42 weeks):

- Average lengthening was **35 mm** (range, 14mm-65mm)
- Accuracy was **96%**

- Minimal Pain
- Minimal temporary joint stiffness
- Quick preditable bone healing



Age 8, congenital
LLD 5.5 cm , femur /tibia
PLLD= 9.5 cm

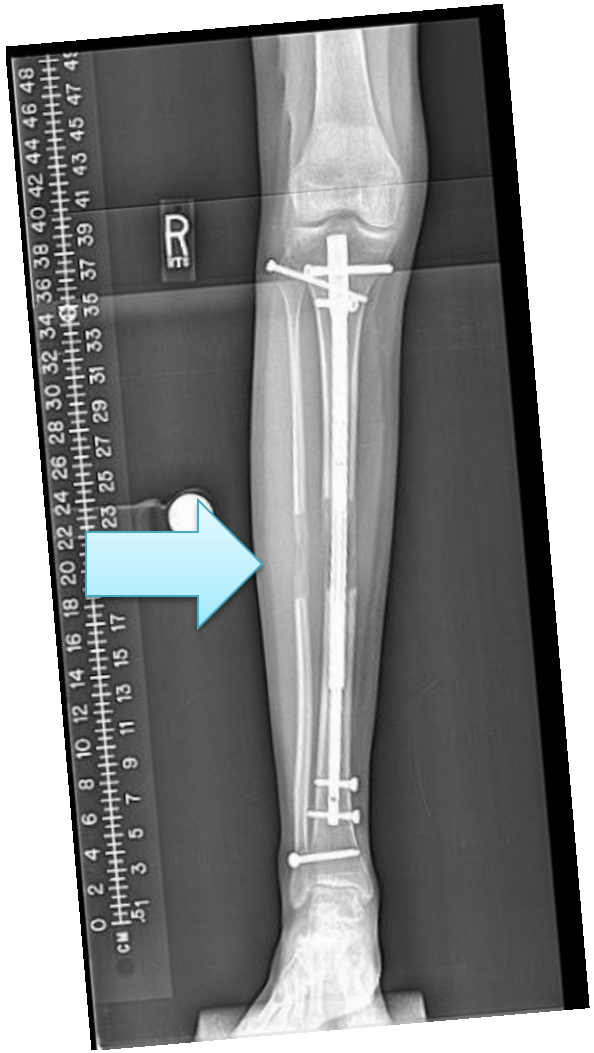
Plan: 4.5 cm lengthening
Of femur
Second future lengthening
tibia

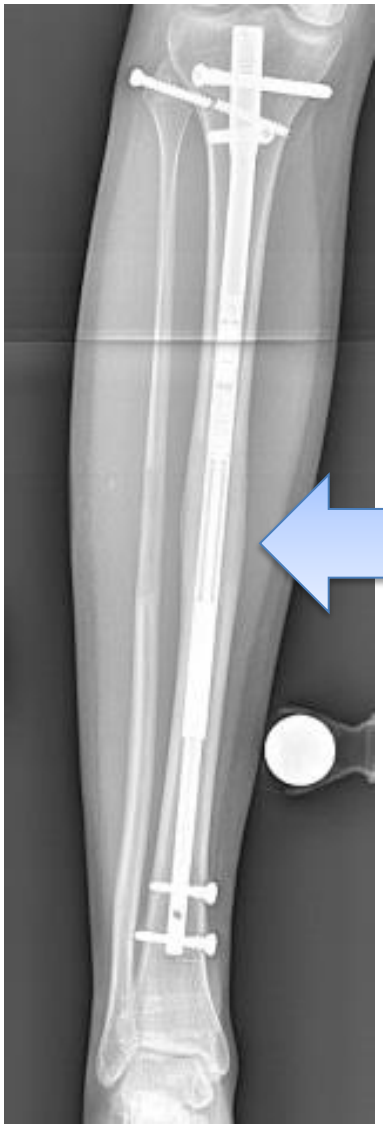
Age 15, LLD now 5 cm

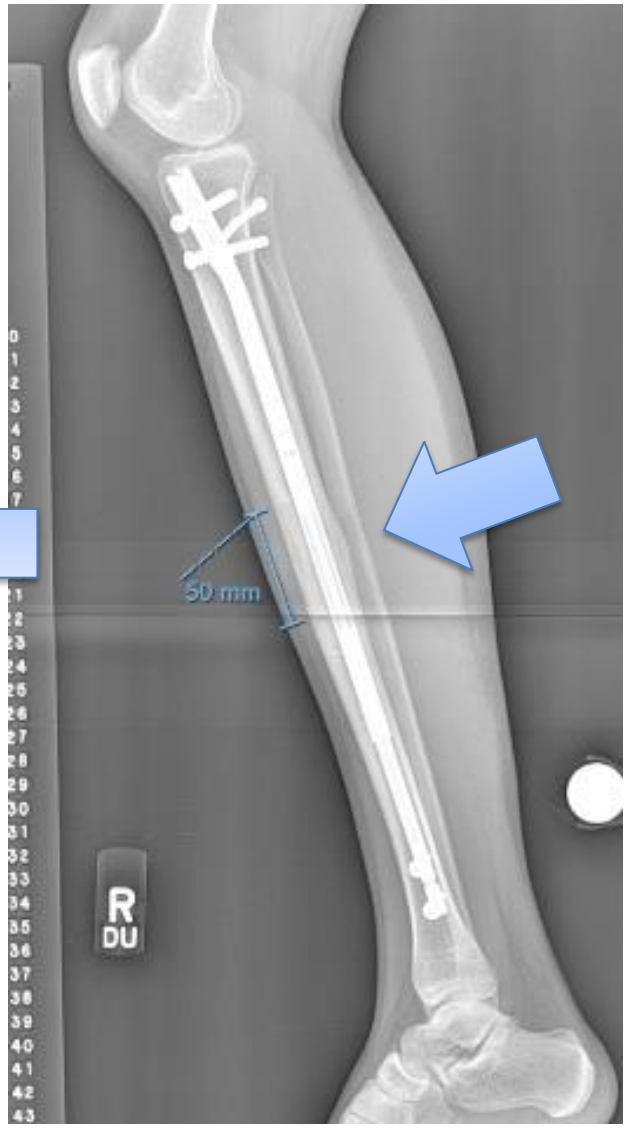
Growth plate is
Now closed.

Use of an IM nail
Is safe

5 cm lengthening with Precice





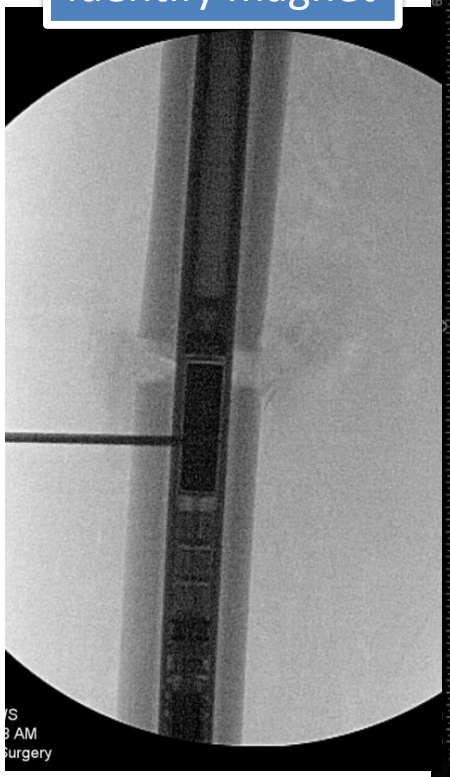

50 mm
R
DU

Age 16
LLD = 36 mm
LLD= 1.5 inches

MAGNETIC INTERNAL LENGTHENING NAIL

Equal leg lengths
2 months
Minimal pain
No frame

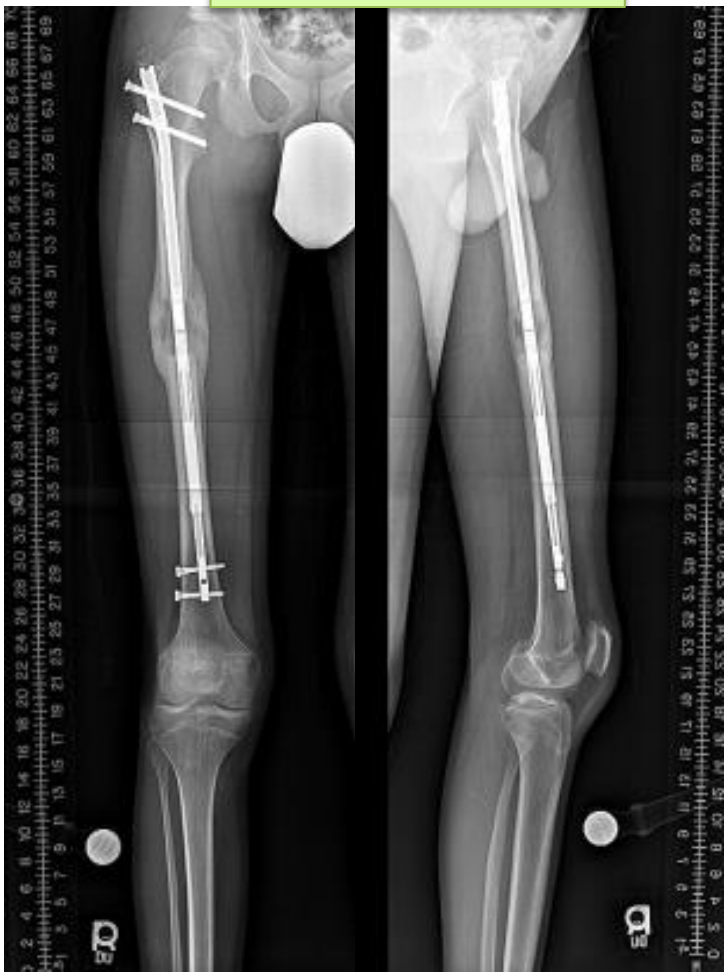
Surgery
Cut bone
Identify magnet



5 weeks
Out to length

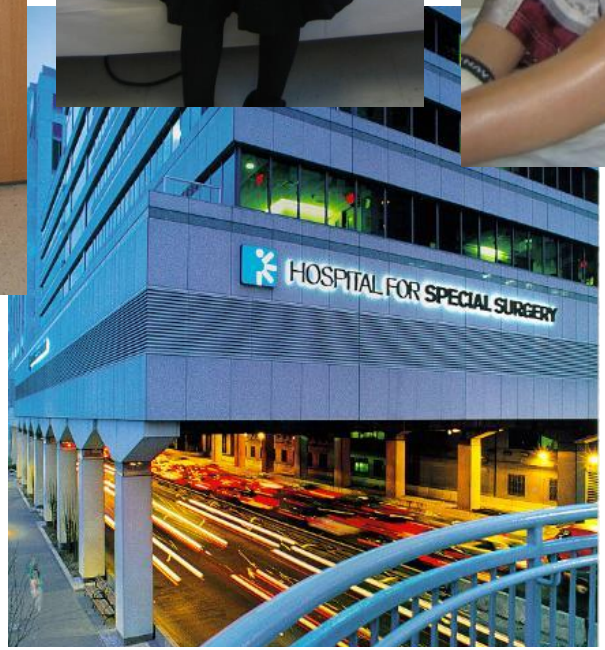
2 months
Bone consolidation progressing

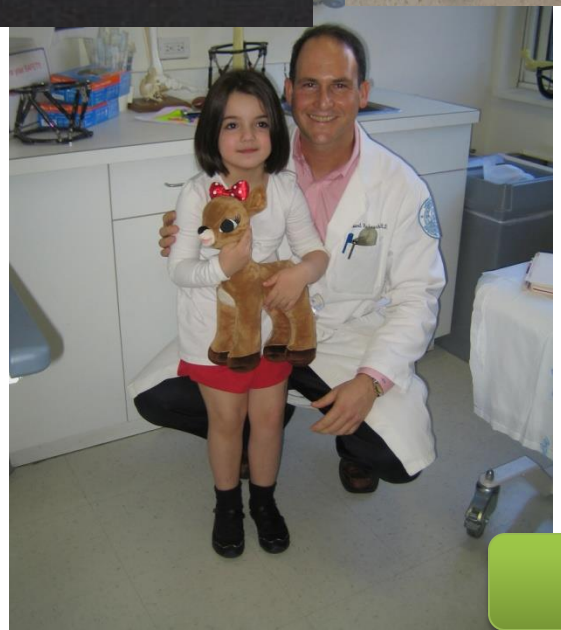
3 months
All healed
Full weight bearing



RSS Limb Lengthening

- LLD divided between femur and tibia
- Presenting as teenager
  - One lengthening femur and tibia
- Presenting as child
  - 2 lengthenings
- Use Internal lengthening nail
  - After age 10 in femur
  - After growth plate closure in tibia
- HGH enhances bone healing
- LLD prediction needs modification for longer growth period





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Thank You

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