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

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Evaluation

• Causes of LLD
  – Congenital
  – Traumatic Growth arrest
  – Overgrowth
  – Neural inhibition

Total LLD (including foot) = (d2 - d1) + lift

Foot height difference = Total LLD - [(F1 - F2) + (T1 - T2)]
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
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
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
Ellis Van Crevald Syndrome
hemiepiphysiodesis

guided growth to Correct angular deformity
Age 16, short stature, residual deformity
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
Bilateral Humerus Lengthening 7 cm
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
Limb lengthening in children with Russell–Silver syndrome: a comparison to other etiologies


Journal of Children’s Orthopaedics
Official Journal of the European Paediatric Orthopaedic Society (EPOS)
ISSN 1863-2521
Volume 7
Number 2
J Child Orthop (2013) 7:151–156
DOI 10.1007/s11832-012-0474-3
Russell Silver Syndrome

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

Plan: 2.6 cm in femur and tibia
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

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<tr>
<th></th>
<th>SRS</th>
<th>Control</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>10.4</td>
<td>13</td>
<td>0.036</td>
</tr>
<tr>
<td>Lengthening (cm)</td>
<td>3.3</td>
<td>3.9</td>
<td>0.507</td>
</tr>
<tr>
<td>Follow up (months)</td>
<td>32 (16-38)</td>
<td>58 (12-130)</td>
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# Bone Healing Index

days of bone healing per cm of lengthening

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<th></th>
<th>SRS</th>
<th>Control</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bone Healing Index (BHI), days/cm</td>
<td>29</td>
<td>43</td>
<td>0.028</td>
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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
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
Precice® Nail

- Telescopic, magnet-operated device
Trochanteric Entry
Good for adolescent < 18 yrs.
12 year old male with congenital LLD
These were first 24 patients (August 2012-July 2013)

29% of patients who underwent limb lengthening surgery during that time period
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{\text{Distraction prescribed} - \text{Lengthening measured}}{\text{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 predictable 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
Age 16
LLD = 36 mm
LLD = 1.5 inches
Equal leg lengths
2 months
Minimal pain
No frame

MAGNETIC INTERNAL LENGTHENING NAIL
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
Thank You

www.hss.edu/limblengthening

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