Limb Lengthening in Children
RSS and other etiologies
Pediatric Endocrine Rounds, Weill Cornell Medical Center, January 22, 2013

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Causes of LLD

- Congenital
- Traumatic Growth arrest
- Tumor
- Overgrowth
- Neural inhibition
Hemiatrophy
Age 13
LLD 5 cm divided femur/tibia
Valgus deformity
Multiplier 1.12
PLLD = 5x1.12 = 5.6 cm
Plan: correct valgus
2.8 cm femur
2.8 cm tibia
Overlengthen by 6 mm
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
The image is manually calibrated.
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 4, valgus deformity
LLD = 4 cm mostly femur
M = 1.83
PLLD = 7.3 cm

Plan: lengthen femur 4 cm
Correct deformity
2nd lengthening in future
Age 8, congenital
LLD 5.5 cm, femur /tibia
M = 1.53
PLLD = 8.5 cm

Plan: 4.5 cm lengthening
Of femur
Second future
lengthening
tibia
Limb Lengthening in Children with Silver-Russell Syndrome: A Comparison to Other Etiologies

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Silver-Russell syndrome (SRS) - 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
Abnormalities of GH secretion have been reported in many SRS 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 SRS (reported average 3.1 cm) many patients will present for limb equalization surgery; however, epiphysiodesis is not a good option.
We asked whether pediatric patients with SRS (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 SRS
- 21 segments in 19 patients – Control
  Posttraumatic 8/7
  Congenital 9/8
  Tumor 4/4
## Methods

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

**days of bone healing per cm of lengthening**

<table>
<thead>
<tr>
<th>SRS</th>
<th>Control</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Bone Healing Index (BHI), days/cm</strong></td>
<td>29</td>
<td>43</td>
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</tbody>
</table>

RSS patients had significantly faster bone healing during limb lengthening.
Subgroup comparison of Bone healing Index

<table>
<thead>
<tr>
<th>Condition</th>
<th>SRS, 29</th>
<th>Trauma, 31.4</th>
<th>Congenital, 41.4</th>
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</thead>
<tbody>
<tr>
<td>Congenital</td>
<td>P=0.032</td>
<td>P=0.068</td>
<td>P=0.068</td>
</tr>
<tr>
<td>Tumor, 66</td>
<td>P=0.019</td>
<td>P=0.04</td>
<td>P=0.162</td>
</tr>
<tr>
<td>Trauma, 31.4</td>
<td>P=0.298</td>
<td></td>
<td>P=0.068</td>
</tr>
</tbody>
</table>
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
All SRS patients had good outcome, no significant problems

No premature consolidation on hGH

No hGH-related orthopedic complications (LCP, SCFE, scoliosis)
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
PLLĐ ~ 5.3 cm
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
Ellis Van Crevald Syndrome
hemiepiphysiodesis
Hypophosphatemic Rickets
Age 9
5 cm LLD
7 cm LLD
At age 15
Total Lengthening on R lower extremity
5+9+7=
21 cm
Relative contributions of various growth plates
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
12 y/o, sarcoma excision prox tibia
Reconstructed with free fibula.
Nonunion, varus deformity, growth arrest proximal tibia, flap-poor skin

R, L tibia 300mm
M=1.18
300 x 1.18 = 354
354-300 = 54 mm
54 mm x 60% = 33 mm
L ankle valgus from free fibula Donor site
Age 15.5
L ankle straight, R tibia healed.
LLD 3 cm

Plan: lengthen femur. Avoid tibia
Failed free fibula reconstruction of Osteosarcoma resection

LLD = 6 cm
Free fibula
Nonunion + deformity
LLD
Nonunion repair
Deformity correction

8 cm (2 cm overlengthened)
Fracture of fibula

8 cm

6 mos
CPT- congenital pseudoarthrosis of tibia
Ollier’s Disease
10 cm Bilateral tibial lengthening Age 13
Blount’s Disease