Context

- Flaccid, progressive, proximal, LE
- Phenotype varies
- Promising strategies, alter the natural history
  - Gene therapy (AAV9)
  - SMN2 splicing modulation
- Challenging convention
- Positioning, comfort, ease of care
- Limited upright mobility (stander)

Maximize ambulation

<table>
<thead>
<tr>
<th>SMA type</th>
<th>Achieved function</th>
<th>Life expectancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ia (more severe)</td>
<td>No sitting or turning</td>
<td>&lt;2.5 years</td>
</tr>
<tr>
<td>Ib (less severe)</td>
<td>No sitting or turning</td>
<td>2.5–20 years*</td>
</tr>
<tr>
<td>II (intermediate)</td>
<td>Sitting ability</td>
<td>2.5–30 years*</td>
</tr>
<tr>
<td>IIIa (mild; retarded motor)</td>
<td>Walking ability</td>
<td>Fourth to fifth decade</td>
</tr>
<tr>
<td>IIIb (mild; normal motor)</td>
<td>Walking ability</td>
<td>Fourth to sixth decade</td>
</tr>
<tr>
<td>IV (adult)</td>
<td>Walking ability</td>
<td>Normal</td>
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</tbody>
</table>

Decision Making
CRAWLING PATTERNS IN NEGLECTED POLIOMYELITIS IN THE SOLOMON ISLANDS

A. B. Cross, Solihull, England

THE JOURNAL OF BONE AND JOINT SURGERY

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SQUATTING GAIT

INFANT-LIKE CRAWL

BUTTOCK PIVOTING
• What we see versus what pt/family sees
• Establish realistic, functional goals
• Proactive versus reactive
Contractures

• Why?
  Denervation/weakness, immobilization/positioning

• What?
  Contractile Tissue (↑ connective tissue)
  Connective tissue (lose extensibility)
  Joint (fibrofatty connective tissue)

Farmer SE, James M. Disability and Rehabilitation 2001;23:549.
• 143 pt, Type II, 5 yrs fu
• Hip: 45° FC, ↓ rotation, add, abd
• Knee: 37° FC
• Ankle: Equinus (13° in 31%)

• 44 pt

• Type II
  - Flexion contracture
    • 48% hip, 89% knee, 52% ankle

• Type III
  - Flexion contracture
    • 18% hip (45° )
    • 29% knee (27° )
    • 53% ankle (15° )

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>No. of patients</th>
<th>No contractures (*) % patients</th>
<th>Mild contractures (*) % patients</th>
<th>Severe contractures (*) % patients</th>
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</thead>
<tbody>
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<td>1</td>
<td>10</td>
<td>80%</td>
<td>20%</td>
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<td>2</td>
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<td>60%</td>
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<td>9</td>
<td>33%</td>
<td>67%</td>
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<td>10</td>
<td>0</td>
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<td>8%</td>
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<tr>
<td>12</td>
<td>5</td>
<td>0</td>
<td>20%</td>
<td>80%</td>
</tr>
</tbody>
</table>

Strategy

- Multidisciplinary
- Multimodality
- Relentless/sustainable
Nonoperative: Proactive vs. Reactive

- Positioning
- Physical therapy
- Serial casting/wedging
- Orthoses
  - Static, Dynamic
  - When and how long
- Practicality and sustainability
- Shared responsibility
Reactive: Achieve and maintain range of motion

- Operative
  - Soft tissue
    - Muscle lengthening or release
  - Osteotomy
- Early mobilization
- Risk of losing function
- Deterioration/recurrence over time
Hip Flexion Contracture
Flexion + Abduction
Knee Flexion Contracture

- Stretch casting
- Soft tissue lengthening/release
  - Hamstrings
  - Posterior capsule
- “Guided growth”
- Extension osteotomy
Guided Growth???

- Indications evolving
- 1°/month
Distal Femoral Extension Osteotomy
Hip Dysplasia
Hip: What we think we know

- Subluxation and dislocation very common, II > III
- Pain/functional consequences rare
- Pathologic anatomy?
- Reactive rather than proactive

Evans
Pathologic Anatomy?

- Osseous
  - Proximal femur
  - Acetabulum
- Soft tissues?
Relationship between hips and spine
Relationship between hips and spine
Treatment

- Age
- Unilateral versus Bilateral?
- Subluxation or Dislocation
- Ambulatory?
- Symptoms?
What is the pain generator?

- Soft tissue contracture
- Bony (impingement or arthritis)
- Local pressure from asymmetric seating position
Subtrochanteric Resection

Valgus/Pelvic Support Osteotomy