The Child with AMC: A Pediatric Hand Surgeon’s Approach

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History and physical exam

- Occupational therapist and surgeon together
International Classification of Functioning, Disability and Health

- Impairment
  - Motion, strength
- Activities
  - Taking care of yourself
- Participation
  - Involvement in a life situation (ability to do what you want to do)
The Hand and Arm in AMC

- Will he need his hands and arms for mobility (scooting, crawling, walking)?
The Hand and Arm in AMC

- Can she feed herself?
- Can she dress herself?
- Can she perform personal hygiene (toileting)?
The Hand and Arm in AMC

- Can she do activities that are important to her?
Most common form of arthrogryposis

Clinical diagnosis (no definitive test), not usually diagnosed before birth

Sporadic (not inherited)

Failure of development of the spinal cord in late 1st trimester of pregnancy (after arms and legs have formed)

Characteristics of infants

- Bright, assertive, engaged
- Usually elbows are extended (straight) and foot deformity (clubfeet) are present
- Deformities are usually symmetric
From Hall et al “Amyoplasia Revisited” (2014)
Long-term functional and mobility outcomes for individuals with arthrogryposis multiplex congenita

Hirmand Nouraei¹ | Bonita Sawatzky¹,² | Megan MacGillivray²,³ | Judith Hall⁴

- 177 adults with AMC from 15 countries
- 90% arm and leg involvement
- 75% of people with AMC live independently
- 3x more likely than general population to have a graduate degree
- Quality of life comparable to general population
- 50-60% had back or joint pain
The Hand and Arm in AMC
The Hand and Arm in AMC

- Consider the arm and hand
  - Flexibility (range of motion)
  - Power (strength)
- Shoulder
- Elbow
- Wrist
- Thumb
- Fingers
Consider the arm and hand
  - Flexibility (range of motion)
  - Power (strength)

Shoulder – internal rotation

Elbow - extension

Wrist - flexion

Thumb - flexion

Fingers – extension or flexion
The Hand and Arm in AMC

- Consider the arm and hand
  - Flexibility (range of motion)
  - Power (strength)
- Shoulder
- Elbow - extension
- Wrist
- Thumb
- Fingers
Elbow Extension Contracture

- Flexibility and power are separate issues
- Flexibility alone may be sufficient
- Flexibility is necessary before muscle transfer
Passive Elbow Stretching

- Babies are stretchy!
Elbow Extension Contracture

Algorithm: Flexibility

Elbow Extension Contracture

Vigorous passive stretching (< 1 year)

< 60° flexion: elbow release and triceps lengthening

60-90°: continue stretching, splinting

> 90° flexion: nighttime splint
Posterior capsulotomy & triceps lengthening

- Gains flexibility (average 40° flexion)
- Post-operative therapy and splinting is important
  - Alternating flexion and extension to avoid the opposite problem (flexion contracture)
Elbow Extension Contracture

Algorithm: **Power**

- Passive flexion maintained (≥ 90°)
- Weak or absent active flexion that limits activities
- Able to cooperate with therapy (> 3 years old)

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**Steindler flexorplasty**

- Strong finger/wrist flexors
- Stable wrist (in neutral or extension)

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**Long head of triceps transfer**

- Strong triceps
- Possibly contraindicated after elbow release and triceps lengthening

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Other arm or hand reconstruction, staged or at the same time, as indicated
The Hand and Arm in AMC

- Consider the arm and hand
  - Flexibility (range of motion)
  - Power (strength)
- Shoulder
- Elbow
- Wrist - flexion
- Thumb
- Fingers
Wrist flexion contracture

- Flexed, ulnar deviated
- Position weakens grip
- Goals of treatment:
  - Maintain motion
  - Reduce flexion contracture to improve grasp
  - Improve ability to perform self care
Treatment of wrist flexion contracture

- Consider the child’s ability to walk
  - Walking is often delayed in AMC due to hip/knee/leg problems
  - Children who aren’t walking yet may bear weight on flexed wrist
  - Changing wrist position may impair ability to “scoot”

- Hand surgeon & leg surgeon should plan treatment together
Treatment of wrist flexion contracture

- **Dorsal carpal wedge osteotomy**
  - Removes part of wrist bones to improve wrist position
  - Maintains motion at the main wrist joint
  - Includes a tendon transfer to help power wrist extension
Treatment of wrist flexion contracture

- Dorsal carpal wedge osteotomy
  - Removes part of wrist bones to improve wrist position
  - Maintains motion at the main wrist joint
Treatment of wrist flexion contracture: dorsal carpal wedge osteotomy

- Improves wrist extension about 40 degrees and maintains motion
- Improvement lasts at least 6 years
- Long term night splinting helps maintain improvement
- New position improves function
Treatment of wrist flexion contracture: dorsal carpal wedge osteotomy

- **Age**
  - Kids > age 7 may get a better result but…
  - Older kids may have more flexion deformity & changing the position of their wrist may interfere with their established grasp pattern

- **Fingers**
  - Grasp may not improve after wrist position is changed if fingers are very stiff

- **Wrist surgery** may be combined with other operations
The Hand and Arm in AMC

- Consider the arm and hand
  - Flexibility (range of motion)
  - Power (strength)
- Shoulder
- Elbow
- Wrist
- Thumb - flexion
- Fingers
The Hand and Arm in AMC

- Index finger dorsal rotation flap helps thumb extend, and opens first web space
- Easier to grasp larger objects
- For older kids, joint fusion (stiffening) can also be helpful
Index dorsal rotation flap to open thumb

Before surgery

Thumb

After surgery

After surgery
The Hand and Arm in AMC

- Consider the arm and hand
  - Flexibility (range of motion)
  - Power (strength)
- Shoulder
- Elbow
- Wrist
- Thumb
- Fingers - flexion
Camptodactyly (finger flexion contracture) release

- Release does not increase motion, just shifts the arc from flexion to extension
- Gets the finger out of the way, but decreases child’s ability to grip
The Hand and Arm in AMC

- Consider the arm and hand
  - Flexibility (range of motion)
  - Power (strength)
- Shoulder – internal rotation
- Elbow
- Wrist
- Thumb
- Fingers - flexion
External rotation osteotomy of the humerus

- Helps rotate hands away from the midline
- Enables 2 handed grasp of larger objects
- Can interfere with some effective grasp patterns
I’m pretty cute...even with my publication privacy blindfold on!