Pediatric Upper Extremity Limb Deficiencies: Identification and Interventions

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  • Children’s Healthcare of Atlanta Hand and Upper Extremity Program,
    Atlanta, GA

Disclaimers:

• We are not hand therapists
• Many of the kids we see have hands, but not all of them.
• We see children with orthopedic conditions which impact on fine
  motor skills and independence in ADL’s.

Objectives

The presenters will:
• Describe the importance of hands for the infant from a sensory motor
  aspect
• Identify the standardized tools to use to help with quantifying physical
  limitations and to track change in children with limb differences.
• Describe range of motion and strength issues which impact on age
  appropriate exploration and skill with upper body using arthrogryposis and
  skeletal dysplasia as examples.
• Describe types of upper extremity congenital and acquired amputations
  and limb deficiencies
• Provide opportunity to expand the tool box for treatment ideas for children
  with upper limb differences

Upper Extremity Development

• Arm buds develop at 24 days of gestation at C5 - C8
• 7 weeks gestation the limb is elongated and has sections that become hand,
  forearm, and upper arm.
• By 8 weeks gestation webbed fingers are present and the arms begin to rotate in.
• Localized movement of the upper body begin around 10 weeks.


Upper Extremity Development

• By 12 weeks gestation hands are fully formed but held in a semi-flexed position with thumbs in
  opposition.
• 14 weeks gestation fingers move synchronously.
• 15-16 weeks isolated movement of fingers begin.
• By 23 weeks gestation, fetuses can purposefully bring hands together.
Hands are Sensory Rich

They are smooth and hairless and densely innervated with sensory nerves.

By 12 weeks of gestation fetuses are exploring by reaching hand to face.

By 20 weeks gestation, if the fetus in not sucking fingers or thumb, it could indicate a problem. Some begin this skill as early as 10 weeks gestation.

Prenatal Diagnosis

Routine structural ultrasound at 18-22 wks
  - Presence of 4 limbs
  - Not standardized as to the detail of exam

Ultrasound detection rates:
  - 64% of all fetal anomalies
  - 25-29% of isolated limb anomalies

Why is fetal development so important?

- Insults occurring in the first trimester of life can impact on fetal growth
- Causes
  - Defective gene coding for development
  - Growth interference
    - Structural
      - Uterine wall
      - Septate Uterus
      - Bicornicate Uterus
      - Didelphic Uterus
    - Fibroids
    - Incomplete rupture of inner amniotic membrane
  - Maternal illness

Sometimes our babies do not come out as expected

- They may have orthopedic limitations but not cognitive deficits.
- We need to create opportunities for exploration to match cognitive ability.
- Remember babies come out hard wired to bring hand to mouth.
  - If they can not perform this task, they lose opportunity for play and exploration.
  - Self feeding becomes a challenge.
  - Self soothing may be challenging especially in times of teething.

Some skills are missed and some are done differently from a developmental standpoint when there are upper extremity differences.

Consider the movements needed for normal postural responses and skills we would typically test:

- ATNR
- STNR
- Prone propping
- Pull to sit

How do we measure these kids?

- No one testing tool is the correct tool for birth through adulthood.
- It is hard to tease out skill versus function with skill being the specific task and function including the compensatory strategies.
- It is important to consider where the plan of care is going to go and evaluate with that in mind.
- Make sure to include tools that ask the parent and the child, when appropriate, how it is going. Often you will find out more than you thought.
Early Developmental Tools

- AIMS
- Bailey
- Peabody Developmental Motor Test
- TIMP

Due to upper extremity issues, not all cognitively appropriate children will be able to score well on these standardized tests. Consider these tests as scores to qualify for service and to plan for care but do not make assumptions on cognition based on reaching, bimanual skills, and upper body weight bearing abilities.

Range of Motion

- When available, check motion shoulder, elbow, wrist, and hand.
- Do passively and actively
- When available, check for symmetry of motion and ability to bring hands together.
- Look functionally
  - Ability to grasp items in one hand
  - Ability to hold in 2 hands
  - Strategies used to hold an item
- Take photographs to document positions difficult to measure.

Muscle strength

- For the very young, it is important to try to tease out what muscles the child is using in the upper extremity.
- Although manual muscle testing on testing with a hand held dynamometer may not be reliable in the very young, the ability to palpate muscles to identify what is working gives help in identifying treatment plans.
- AMS: Active Movement Scale

Modify tests used for other realms

- PEDI: Pediatric Evaluation of Disability Inventory (6 months to 7.5 years)
- PEDI—CAT: Pediatric Evaluation of Disability Inventory Computer Adaptive Test* (birth – 20 years)
- ASK: Activities Scale for Kids (5-15 years)
- PROMIS: Patient Reported Outcomes Measurement Information System* (short form has a paper copy) 8-17 for child self report parent proxy for 5-17
- PODCI: Pediatric Outcomes Data Collection Instrument
- CHEQ: Children’s Hand Use Experience Questionnaire (6-18 years)*
- ABILHAND-Kids

Ask the Caregiver and the Child about Skills

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Ask the Caregiver and the Child about Skills

- These tests seem to tease out functional skills rather than specific movements.
- Do not look at quality of movement.
- Tools do not easily follow the lifespan for long term data collection.
- Many require computer access.
- For those that need computer access, the therapist then needs some place to put the data to make sure it is secure.

Hand Specific Tools

- Manual Abilities Classification System (MACS)
  - Designed for CP.
  - Looks at bimanual skills.
- HOUSE Thumb Classification System
  - Classification of spontaneous UE use with emphasis on thumb posture.
- Zancolli’s Classification System
  - Posture of wrist and hand.
- Bi-Manual Fine Motor Function (BMFM)
  - Fine motor looking and grasp, hold, and manipulate objects for each hand.
- Assisting Hand Assessment (AHA)
  - Useful for unilateral disability.

Where do physical therapists see children who have dysmorphic upper extremity/hand issues?

<table>
<thead>
<tr>
<th>Medically based</th>
<th>Educationally based</th>
</tr>
</thead>
</table>

Medically based

- NICU
  - Early medically based intervention prior to start of EI services.
  - For equipment related issues.
  - Episodically for specific skills not addressed in school environment.
  - Post-operatively.

Educationally based

- Birth to 3 programs are family centered and often are therapy rich.
- Preschool hand function skills impact management of personal items: coat, backpack, art projects. May impact ability to use some playground equipment.
- Unless hand function impacts on gross motor skills, by the time the child gets to school age, PT is not always seeing the child.

Many times in the medical model and the educational model

Occupational Therapy takes ownership for the upper extremity.

Always remember Physical Therapy has much to offer and is an important member of the team.
Let's look as some of examples

Come meet some specialty diagnoses

- Open your tool box
- Get out your duct tape
- Find some PVC pipe
- Play with some InstaMorph and Sugru
- Use some Velcro and some left over splint supplies

Leave here thinking you can adapt something.

Skeletal Dysplasia: a Big Umbrella

- Genetic disorder that impacts on growth of bone and connective tissue.
- Hypermobility of joints
- Shortened extremities
- More than 200 types
  - Achondroplasia
  - Osteogenesis Imperfecta
  - Multiple Epiphyseal Dysplasia
  - Diastrophic Dysplasia
  - And about 200 other types

What are the Potential Upper Limb Related Functional Limitations due to the Skeletal Dysplasia?

- Shortened or bowed limbs of the upper extremity will impact on reaching.
- Potential issues with bimanual skills.
- Potential issues with managing self care.
  - Feeding
  - Dressing
  - Bathing
  - Toileting
- Bone fragility and joint hypermobility may impact on ability to lift objects or sustain postures.
  - May not be able to use a standard pediatric assistive device for walking due to hand grip and wrist hypermobility issues.

Skeletal Dysplasia in the NICU

- Positioning and Handling
  - Emphasis on getting hands towards midline and face.
  - Holding splints for fractures, contractures, and instability as needed.
- Family and Staff Education
  - Maintain overall precautions based on diagnosis or presentation.
    - There is a bias for the skeletal dysplasia population to keep babies horizontal until they seek more upright positions naturally.
    - This helps prevent long term spinal complications with flattening of vertebrae.
    - It also presents with upper extremity challenges while waiting for the child to be ready to move.
    - Many types of skeletal dysplasia have unstable cervical spines at an early age.

After Hospital Discharge

- Medically services are based on medical necessity.
- May need some splinting for improving position.
- May need supportive device for fracture management.
- Emphasis on family training so they can carry out care at home.
- Medically based therapist may be the link between the orthopedist and the community based therapist.
Early Intervention Services

Birth to three programs are family centered. Often these children enter the system after the first birthday. Work to enhance play and participation at home.

Be aware that creeping on hands and knees may never be a feasible task due to arm length, upper extremity stability, and hand position.

Medically Based Expectations: The first three years

- Natural development of sitting skills.
- Opportunity for play in preferred positions.
- Use of ADL’s as therapeutic opportunities.
- Once ready to get up on feet, identify hand grip that works for the child, if an assistive device is needed.
- Support families through surgical interventions and medical changes.

Transition to Preschool

Maximizing independence in school based ADL

Enhancing fine motor skills

Independence in school based ADL

Management of toileting and hand washing is a quality of life issue in school.

Changing requirements for management of personal items increases complexity of task as the child ages.

Enhancing fine motor skills

Transition to School Age

- Focus on independence in school environment.
- Work towards reducing support of school aide when possible.
- May need to work on energy conservation techniques.
- Reduced writing and typing expectations.
- Work stations with tools set up for use.
- Emphasis on skills needed to attend college independently.
- Skills may change with surgical interventions not only of upper extremity but also of lower body and spine.
Surgical Interventions

- Upper extremity surgical interventions are not common in this population except for stabilization.
- May see rodding of upper extremity fractures to reduce bowing.
- With lower extremity interventions, may need to think creatively for an assistive device.

Medically based intervention

- The child who is ready for ADL.
- Work on flexibility and strength in order to do clothing management.
- Identify equipment needed to enhance independence.
- Think not only of bathing, dressing, grooming, feeding, and toileting but also access to environment.
- Encourage adaptations of environment to allow independence in function at home.

Tools to enhance long term independence

Individuals who have skeletal dysplasia often do not have aggressive surgical management of the upper extremity. They do have difficulty with reach that can impact on independence in function.

- Strategies to enhance independence for a lifetime should begin early and should track age appropriate expectations for skills.
- Sometimes things need to be adapted aggressively to allow independence in skills.

Arthrogryposis Multiplex Congenita

- Neuromuscular non progressive disorder characterized by joint contractures and associated muscle weakness.
- Amyoplasia is most common type which is not genetically linked.
- An insult that occurred during the first trimester of pregnancy.
- Other types of arthrogryposis may have genetic linkage.
  - Distal arthrogryposis
  - Multiple Pterygium Syndrome

Timing of insult impacts on upper extremity involvement
What are the Potential Upper Limb Related Functional Limitations due to the Arthrogryposis?

Stiff limbs of the upper extremity with poor hand alignment accompanied with antigravity strength deficits will impact on

- All fine motor skills.
- Potential issues with managing self care.
- May need adaptations to allow written communication.
- May not be able to use a standard pediatric assistive device for walking due to hand grip issues.
- May need creative interfaces with power mobility.

• May need adaptations to allow written communication.
  • All fine motor skills.
  • Potential issues with managing self care.
  • Feeding
  • Dressing
  • Bathing
  • Toileting

• Teaching caregivers gentle stretching of all extremities during times of care.
  • Upper body with feeding
  • Lower body with diaper changes
  • Total body with bathing
  • Soft lightweight splints to hold wrist in best alignment that is comfortable.
    • Craft foam
    • Vetfoam
  • Identify feasibility of hand to mouth for self soothing.
  • If eating by mouth, early strategies to help hold bottle or breast.

NICU

Other Hand Splint Ideas

After Hospital Discharge

First six months of arthrogryposis

Medically Based Expectations

The first 3 years

- Use early plasticity of muscles and joints to enhance alignment of upper body.
- Identify toys the child can manipulate to have enjoyable experiences with hand use.
- When using electronic devices, please use a stylus early. This early tool use will translate to better tool use in the future.
- Fabricate and adapt splints to meet growth and change and to enhance function.
- Procure equipment to enhance function but keep in mind, it is always best to do things without adaptations when possible.
- Explore self feeding options.
Early Intervention

- Birth to three program often starts around 3-4 months of age.
- Depending on upper body presentation, may never have ability to weight bear on arms in prone.
- Need to work on strategies to enhance mobility and reaching.
- Work with family to use manipulative toys rather than screen related toys.
- Use action songs whenever possible.
- Support family on self feeding opportunities.

Arm Lifting Devices

Surgical Timing

For the upper extremity, the child needs to be big enough to tolerate the amount of anesthesia necessary to perform the procedures.

A baby who had undergone serial casting to manage elbow extension contractures often will have a better result than one who has not. The use of the range while waiting for the child to be big enough.

Surgical Intervention: Toddler

- Elbow capsulotomy
- Triceps lengthening
- Carpal wedge osteotomy
- Opponensplasty for the adducted thumb

Post-operative Management

- Splinting to hold range.
- Active training to use new alignment.
- Look functionally first.
  - Play with manipulative toys
  - Work on self feeding
  - Go swimming

Transition to Preschool

• Most children with arthrogryposis are cognitively bright and should be mainstreamed starting in preschool.
• Upper body limitations due to strength and range of motion impact heavily on preschool tasks.
  • Playground skills
  • Art activities
  • Snack time
  • Hand washing
  • Management of personal items
• May need power mobility to be the center of fun on the playground.

Writing Adaptations

Scissors

Transition to School Age

Surgical Interventions

• If there is adequate muscle power and potential for mobility, create a hand that is useful with technology.
  • Align thumbs to be able to use hand held technology.
  • Align upper body to allow bimanual skills.
  • Create power to actively flex elbows
• All upper body surgical interventions at this age are active choices made by the child.
• Not everyone is a surgical candidate.

Medically based intervention

• The child who is ready for ADL
• Skills have to be revisited multiple times as developmentally the child is not always ready.
• Work on strength and range of motion to enhance access, even if the child is not ready to perform tasks.
• After surgical intervention, may need to relearn tasks.
Adaptive Equipment: Dressing

May need strategies to prep upper body in order to reach for bathing. Off-the-shelf items can help with independence.

Adaptive Equipment: Bathing

Arthrogryposis has very diverse presentation with goal of transitioning to adulthood with a level of independence for self care and employment.

For Adaptive Equipment Ideas Please Follow Nemours on Pinterest

- https://www.pinterest.com/nemours/osteogenesis-imperfecta/
- https://www.pinterest.com/nemours/skeletal-dysplasia/
- https://www.pinterest.com/nemours/arthrogryposis/

Thank you!

rdonohoe@Nemours.org
Congenital and Acquired Limb Deficiencies

- Children are not miniature adults
- "Thus one may define the child with limb difference as a skeletally immature individual with either an amputation or a congenital limb deficiency of one or more limbs"
- Tissues in children are quite different than those in adults - blood supply, healing potential and general tissue metabolism usually are maximized in children
- Childhood are always changing in their developmental capabilities - this is rarely true for their adult counterpart

Classification of Congenital Limb Deficiencies

Classification systems generally to describe limb deficiencies

- Frantz and O’Rahilly
- ISPO
- Functional
- IFSSH - International Federation for Societies for Surgery of the Hand. Specialized hand and upper extremity classification

Classification - Frantz and O’Rahilly

The International Terminology

- Name the Side (L, R, or B)
- Decide if the deficiency is longitudinal or transverse
- Transverse resembles a surgical amputation. No remaining distal part
- Small remnants of rudimentary fingers or toes ("nubbins") do not count
- Longitudinal is anything else

Classification – Functional and Reimbursement

Representative of transverse, acquired, or traumatic amputation levels

- Above Knee = Trans femoral
- Below Knee = Trans tibia
- Above Elbow = Trans humeral
- Below Elbow = Trans radial
Classification, where we are now

1973
International committee of the International Society for Prosthetics and Orthotics (ISPO) produced an International Terminology accepted by the World Health Organization and the International Standards Organization 1983
IFSSH classification system specific for upper limb deficiencies based on the ISPO classification

IFSSH Classification System
• Presented by Swanson in 1960-70s
• Morphologic characteristics and patterns of failure
• Not classified by causation

Surgical Principles: IFSSH Classification System
• Under-formation
  • Lengthening
  • Transplantation- allograft, autograft
  • Re-alignment with tendon transfers

Upper Limb Congenital Anomalies
• Limb deficiencies that can involve upper and lower limbs
• Associated orthopedic anomalies
• Most frequently involve surgeons, PT/OT and P&O

Type I: Failure of formation
• Transverse and longitudinal deficiencies
• Cleft hand/sybrachydactyly
• Bilateral club feet

Surgical Principles: IFSSH Classification System
• Over-separation, laxity
  • Reduction
  • Stabilization
  • Augmentation if possible- lengthening >2 cm length
  • Over-growth- decrease length and volume and increase function and appearance
  • Reduction
  • Debulking
  • Structural failure
  • Prevent progression of the deformity
Transverse Deficiency

- Sporadic, not associated with other anomalies
- Most common at the proximal forearm
- Possible vascular insufficiency
- Fit with prosthesis at 6 months of age
- Equal limb lengths
- Developmental goals
- Prosthesis may not be worn or recommended

Phocomelia

- Absent intervening segment
- Can affect all limbs
- Genetic, drug-induced, environmental exposures
  - Thalidomide, Accutane, Zica????
- Prostheses and adaptive devices
- Surgery can help re-orient limbs/digits to be more functional

Radial Longitudinal Deficiency

- 1:30,000 births
- Associated malformations
  - TAR, Holt-Oram, VACTERL, Fanconi anemia
- Therapy:
  - PROM
  - Splinting
  - Centralization
  - Progressive soft tissue distraction
  - Pollicization, creating a thumb
  - Therapy is a must post-op !!!
Ulnar Longitudinal Deficiency

• Very rare: 1/100,000

• 1/3 to 1/2 have other musculoskeletal differences

• Differ from RLD in that it does not involve other organ systems

• Can coexist with:
  • PFFD, fibula deficiency, phocomelia, scoliosis

• Lower extremity involvement can be syndromic:
  • Fibula-Ulna, Femur-Fibula-Ulna

Spectrum of involvement

• Hand, wrist, elbow

• 75% of thumbs are affected

• In plane of other digits

• Unstable elbow/forearm

Therapeutic Aims:

• Stretching/splinting for ROM

Surgical Aims:

• Improve hand/digital function

• 1/3 have syndactyly

• Realign with osteotomies

Longitudinal Deficiency, hands

• Carpal coalitions

• Ectodactyly

• Ulnar

• Radial

• Thumb-in-palm

• Narrowed thumb web

• Syndactyly

• Metacarpal profile

• Single palmar crease

Cleft Hand

• Typical vs atypical

• Typical:
  • Often bilateral, foot involvement, family history
  • V-shaped, central (longitudinal) deficiency
  • “A functional triumph, a social disaster” - Flatt
  • Can include EEC syndrome
  • Surgery around 1 year of age
Cleft Hand
Surgery
- Close cleft and open 1st web space
- Therapy mandatory for post-op management

Cleft Hand: Atypical
- Symbrachydactyly
  - Usually unilateral
  - U-shaped central defect
  - Affects ulnar hand first
- Adaptive devices/prostheses
- Surgery
  - Provide digital length/function
  - Transfer toe phalanges or entire toe to the hand
- Therapy for post-op management

Cleft Hand-Transverse Metacarpal- atypical presentation

Syndactyly
- Digital coronal plane angulation
  - Middle phalanx deformity
  - Usually the little finger
  - Often bilateral > 60 syndromes
- Treatment based upon extent of deformity
  - Significant static angle
  - Dynamic increase
  - Abnormal growth plate

Thumb-in-plane

Clinodactyly
- Digital coronal plane angulation
- Middle phalanx deformity
- Usually the little finger
- Often bilateral > 60 syndromes
- Treatment based upon extent of deformity
  - Significant static angle
  - Dynamic increase
  - Abnormal growth plate
Clinodactyly

- Physiolysis
- Osteotomy

Kirner’s Deformity

- Tip deformity
- Finger looks like it has previously been injured
- Treatment is rarely required

Amniotic Constriction Band

- Typically involves multiple limbs
- Can cause amputations
- Can have craniofacial anomalies
- Multiple theories of causation
  - Intrinsic
  - Extrinsic
  - Vascular

Goldfarb, 2009 JBJSAm.

Moran, 2013 JAAOS.

Acquired, medical causes

- Vascular insult
  - IV infiltrate, associated with NICU interventions
  - Blood clots
- Cancer - solid tumors
  - Amputations
  - Limb sparing
- Trauma
  - MVA
  - Lawn mower
  - Train
  - Land mines

Acquired

- Septicemia
- Purpura Fulminans

Kretz, 2004

Cheung & Upton, 2015
When it’s not just the arms! Considerations

- Function and ADLs
- Maximize sensory input for all developmental domains
- Treatment considerations: be aware of independent body part substitution and O&P adaptations
- Prosthesis and shoes should never impede function
- Emphasis on potential
- Foot function for ADLs

Associated Anomalies, Cornelia de Lange Syndrome

- Head to toe
- Cranial nerves
- Palate
- Eye
- Chest/rippers
- Petechia

- Anus
- Suckling
- CNS
- Scoliosis
- Renal Ultrasound
- Heart Echo
- CBC w/ platelets
- Genetics consult

One Upper and One Lower

- Treat like single limb deficiency
- Will they need walking support?
- Children may choose not to use upper extremity prosthesis
  - Adaptations and modifications

Bilateral Upper Extremity Deficiencies

Support function!!

- Loose fitting shoes that are easy to take on and off
- Adapt the environment first, ask the child. They find a way 😊
- ADL adaptations: dressing, toileting, food prep and eating, writing, play
- Adapted sports and recreation

Bilateral Upper Extremity and One Lower

- Uppers below elbow and lowers below knee
  - Certain ambulation
  - Prosthetic fitting options for UE

- Long above elbow and above knee
  - Ambulation with assisted devices

- Power mobility

Bilateral upper extremity and one lower

Lower Limb Function

- Ambulation
- Must function as hand
  - “Hand” function takes priority

PFFD

- Lower limb surgical options, not so sure
- Extension prosthesis to equalize limb lengths may restrict upper limb function
- Promote Hand function and independence

Bilateral upper and lower extremities: level dependent

- Uppers below elbow and lowers below knee
- Certain ambulation
- Prosthetic fitting options for UE

- Long above elbow and above knee
- Ambulation with assisted devices

- If short residual limbs
- Decrease chance of prosthetic wear
- Power mobility
Reflections

Learned from experience

- Every child is different
- Respect the family’s beliefs and values
- Advanced technology may not be best
- Never underestimate a child’s abilities
- Adapt
  - Every child has different needs and goals
  - Prepared to be amazed
  - Prioritize staging of prosthetic fitting

Prosthetic options

“No single prosthesis can address the multiple deficits associated with upper limb loss…”

Deb Latour, OTR/L Amputee Coalition Annual Conference, 2016

Prosthetic options

- Myoelectric, externally powered
- Body Powered
  - Activity specific
- Passive or non-articulating (avoid using the term cosmetic)
- No prosthesis
- 3-D printing prosthesis - future?

Outcome Specific – Goal Oriented

Self Adaptation

- Focus on therapy
- Age appropriate adaptations and modifications for ADLs and independence
- Transference of hand function to feet
- Hands used for task specific needs
- No prosthesis necessary
- Self esteem and societal issues
Challenges

• Midline in all planes
• Most basic of needs still unmet even with advanced technology
  • Personal Hygiene
  • Feeding
  • Sports
  • Recreation
  • The Arts

Successful Outcomes

• Options
• Choices
• Education
• Support
• Dedication
• Commitment
• Function
• Beliefs and values
• Training

Creative Solutions for age appropriate functions

Upper Limb, “to wear or not to wear”

• Children wear upper extremity prosthesis. Some do not. It is okay either way
• Not one correct answer. There is “no proof”
• Age of starting fitting
• Skills of the treating center
• Bias of the center
• Funding
• Location from center, accessibility

Unrealistic Expectations

5 articles relating to the question, “to wear or not to wear”

<table>
<thead>
<tr>
<th>Author</th>
<th>Journal</th>
<th>Level of evidence</th>
<th>Treatment</th>
<th>Follow up</th>
<th>Lost to study</th>
<th>Correlation between 2 gold standards</th>
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<tbody>
<tr>
<td>Wagner et al.</td>
<td>JPO 2007</td>
<td>no level reported</td>
<td>yes</td>
<td>no</td>
<td>173 subjects</td>
<td>Literature review</td>
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<td>James et al.</td>
<td>JBJS 2006</td>
<td>Level II</td>
<td>yes</td>
<td>no</td>
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<tr>
<td>Crandall et al.</td>
<td>JPO Orthopedics 2002</td>
<td>no level reported</td>
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<td>Davids et al.</td>
<td>JBJS 2006</td>
<td>Level III</td>
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<tr>
<td>Burger et al.</td>
<td>Disability Rehab 2003</td>
<td>no level reported</td>
<td>yes</td>
<td>no</td>
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Authors may refer to their journal for further information.
Other evidence, more positive

- 26 of 29, 90%, subjects reported using prosthesis. 26 averaged 9.2 hours and 14 of the 26 reported year round full time users. Subjects reported prosthetic use if provided with multiple prostheses.

| Study | Level | Therapy | No. of subjects | No. of users | No. of reported | No. of reported | Recommended measurement tools | Outcome Instruments
|-------|-------|---------|----------------|-------------|----------------|----------------|-------------------------------|-------------------|
| Manley et al. | Level V | No therapy | 73 | 46 | 26 | 14 | ACMC, DASH (adolescence) | ACMC, DASH (adolescence)
| Wright et al. | Level IV | Yes | 34 | 34 | 26 | 14 | ACMC, DASH (adolescence), DASH (adult), DASH (adolescence) | ACMC, DASH (adolescence), DASH (adult), DASH (adolescence)

**Pediatric Measurement Tools**

Wright et al. - Systematic review of outcome measurement tools used in the field of upper limb prosthetics, adult and pediatric

- Recommended pediatric measurement tools
  - Assessment for Capacity for Myoelectric Control, ACMC
  - Disability of Arm, Shoulder, and Hand Questionnaire, DASH (adolescence)
  - University of New Brunswick Test
  - Assisting Hand Assessment, AHA
  - Prosthetic Upper Extremity Functional Index, PUFI
  - ABILHAND-Kids


**Take home message for prosthesis use = FUNCTION**

Upper Limb Early Infant Fitting

- Equal arm length, postural balance and symmetry
- Propping in all positions, gross motor
- Oral motor
- Fine motor

**Prosthetic considerations**

- Socket design aids in proprioception during crawling
- Hand design for age appropriate function
- Stylized cosmetics

TRG Infant Alpha Hand, Greek Series
LIL' E-Z HAND
Fast Forward in Time, what we have learned

- Early referral with early fitting before 3 years of age
- Capturing child's capacity to use prosthesis vs. actual performance
- Insurance challenges:
  - Inaccurate information of optimal functional outcomes
  - Inaccurate interpretation of age-appropriate prosthetic use
- Requires practice and training

Early referral

- Pre-natal
- Familiarity with team
- Answer questions
- Alleviate fears and uncertainty
- Ongoing education
- Provide age-appropriate developmental activities
- Handling and positioning
- Periodic re-assessment
- Meet needs of child's rapidly changing function
- Problem solve with children and families
- Prepare for prosthetic intervention if appropriate

Therapist role: educator, advocate, liaison

- Not an emergency
- Ongoing education to parents and community resources, Early Intervention referral
- Communicate findings about function and development with the physicians
- Upper extremity involvement has greater impact than lower limb

Therapist role: Adapt and Modify

The therapist role is to support their patient and family to achieve the best possible outcomes

Adapt the environment not the child

Adapt and Modify

- Need was identified by pre-school teacher
- Toddler wanted to participate in music circle time but having challenges with grasp
- Child very familiar to clinic team
- Although greater function without prosthesis at this time, adapted device to hold drum stick was fabricated = success

Therapist Role: Support Fine and Gross Motor Development

- Prone positions, transitional movements, and play are very challenging for infants and toddlers with upper extremity limb loss.
- Bilateral > unilateral.
- Shorter the length of limb > challenge
- Tummy Time and upper extremity weight bearing are important
- Equal arm lengths
Child with multi limb involvement

- The more limbs involved, the greater impact on development
- Requires more resources
- Collaboration with community providers
  - Early Intervention Services
  - School system teachers and therapists
  - Provide input for developing:
    - IEP
    - 504 education plans
    - Seating and mobility
  - Community adapted sports and recreation
  - Transition planning
  - College resources

Post-op management

- Wound care
- Edema control
- Shaping
- Scar massage - prevent adhesions
- Splinting
- Range of motion - active and functional
- Fine motor manipulation and inter and intra digit prehension

Support Function, Balance, Midline

- Hygiene
- Independent ADLs
- Play
- Recreation

Feeding and Oral Motor Development

- Hand to mouth = sensory, oral motor, fine motor, and speech development
- Necessary for feeding

Promote Midline Function all planes

Reflections: Measuring success

"Just have to do things a little differently"