



**XVI CONGRESO  
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# Evaluation and Treatment of Children with Arthrogryposis

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# Arthrogryposis

- Group of non progressive conditions involving 3 or more joint contractures
- 1 in 3,000 – 4,000 births
- Etiology: A lesion affecting the neuromuscular pathway during the formation of the limbs in utero results in joint contracture

emedicine.medscape.com

<https://www.nationwidechildrens.org/conditions/arthrogryposis>

# Three Classifications

## Classic Amyoplasia

- Significant loss of muscle mass
- Symmetric joint contractures in the extremities – can range in severity
- Disorder of formation in the neuromuscular axis



## Distal Arthrogyryposis

- Preservation of muscle mass
- Affects the joints of the hands and feet
- Radial head dislocation
- Cause is genetic: 19 different types



## Syndromic arthrogyryposis

- Limbs, spine and viscera
- Includes 300 different conditions
- Underlying cause: lack of fetal movement



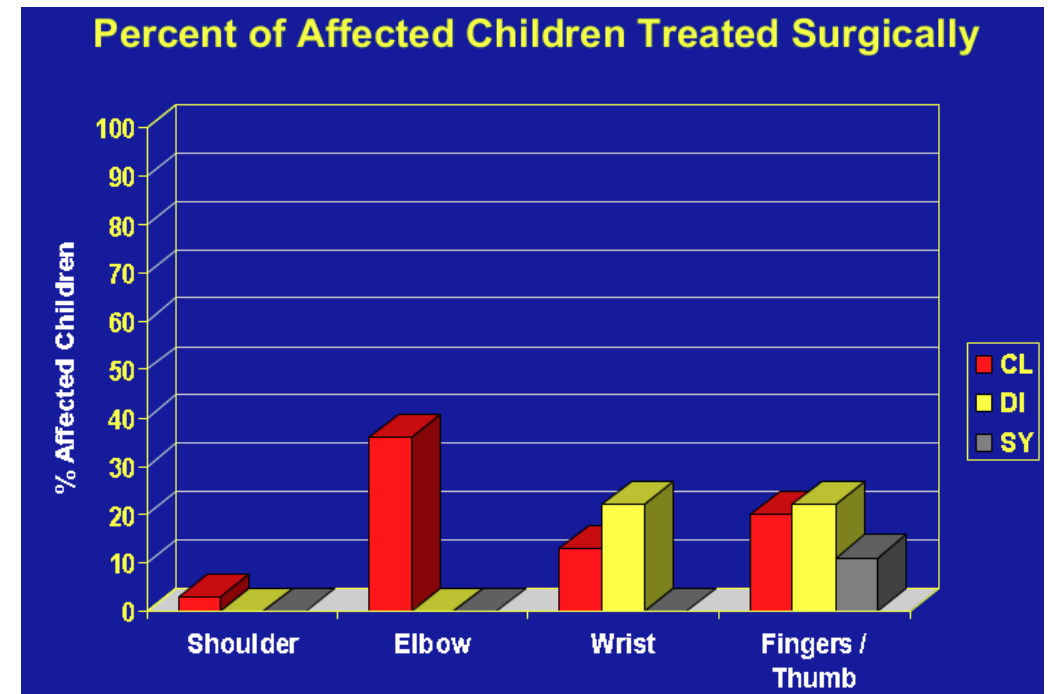
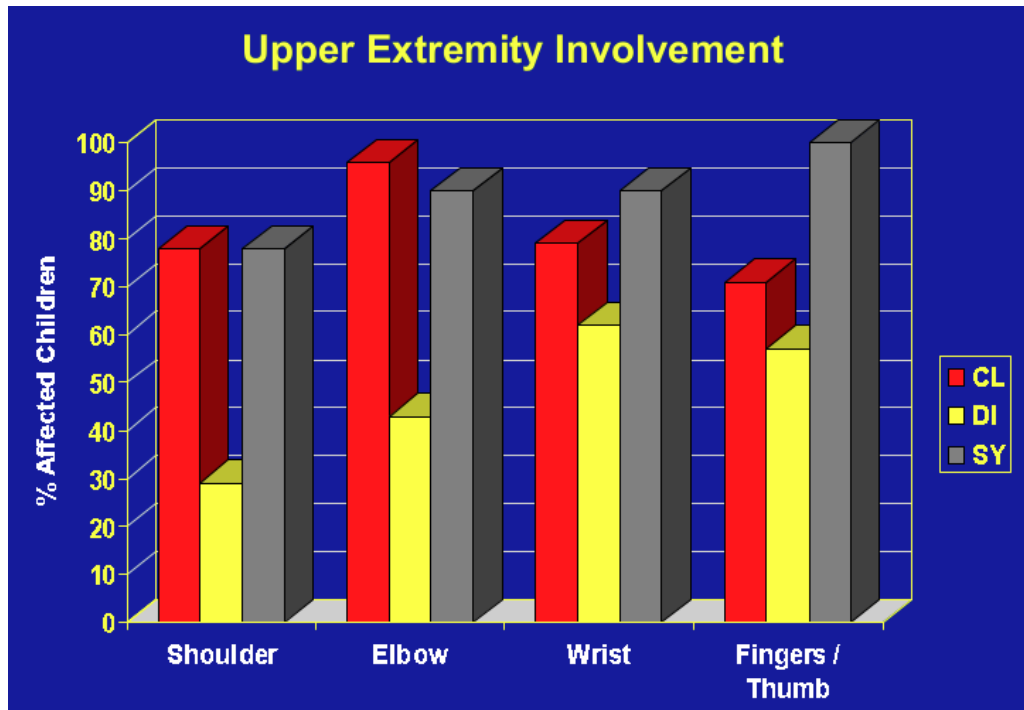
[shrinershospitalsforchildren.org/vanbosse](http://shrinershospitalsforchildren.org/vanbosse)

The underlying condition cannot be treated but the orthopedic manifestations can be by focusing on positioning the limb for function:  
58 children in this study



### Arthrogyrosis: A Review and Update

Michael Bamshad, Ann E. Van Heest and David Pleasure  
*J Bone Joint Surg Am.* 2009;91:40-46. doi:10.2106/JBJS.I.00281



# Treatment Requires Multidisciplinary Collaboration

- Surgeons, therapists, orthotists, nurses
- Educate the family about the diagnosis
- Major characteristics
- Functional Limitations
- Recommended Therapies



# Occupational Therapy Goals Given Their Structural Limitations

- Attain maximum upper body flexibility, strength and function so they can position their arms for optimal use
- Maximize participation in everyday activities



# Assessment: Upper Extremity A/PROM



**Shriners Hospitals**  
**for children**  
 Form 1025B Rev. 11/97  
 PHYSICAL OR OCCUPATIONAL  
 THERAPY RECORD

LEFT				UPPER EXTREMITY PASSIVE ROM				RIGHT			
				Date							
				Examiner's initials							
				Shoulder: Flexion	0-180°						
				Extension	0-60°						
				Abduction	0-180°						
				Ext. Rotation	0-70°						
				(shoulder 90°abd)							
				Int. Rotation	0-70°						
				(shoulder 90°abd)							
				Elbow: Flexion	0-150°						
				Extension	150-0°						
				Forearm: Supination	0-80°						
				Pronation	0-80°						
				Wrist: Flexion	0-80°						
				Extension	0-70°						
				Ulnar dev	0-30°						
				Radial dev	0-20°						
				Thumb: Abduction	0-70°						
				Fingertip to DPC	cm						
Splint: No Yes Type _____											
ADLs:											



# Assessment: Upper Extremity Strength and Movement Patterns Functional Video



## Assessment: Activities of Daily Living

- Interview based on age and function
- Identify areas of concern:
  - Self-feeding and toileting
  - Dressing, grooming, bathing
  - Mobility: ambulation, assistive devices

# Assessment: Pediatric Evaluation of Disability Inventory (PEDI)

- Functional assessment with developmental norms
- 6 months – 7.5 years
- Administration 45-60 minutes
- 197 functional skills
- Self care, mobility, social functioning
- Can use the self care scale only (eating, drinking, brushing teeth, washing, dressing, toileting)
- Isolate components of self care activities that are most difficult
- Raw and scaled scores

# PACS: Pediatric Activity Card Sort

## Child Centered Goals:

- Promotes goal setting by the child
- 100 pictures of different activities
- Yes / No cards to learn child's desire for participation
- Identify three goals they want to accomplish
- <https://caot.ca/client/product2/12/item.html#:~:text=This%20helpful%20assessment%20tool%20focuses,of%20occupational%20performance%20and%20engagement>
- <https://datkanx11.nl/links-english/>



# Treatment: PROM

Improve passive mobility in all affected joints



# What is known about conservative therapy's to improve PROM

## **Palmar 1985**

- 95 infants with arthrogryposis
- Intensive PROM, serial casting, and splinting
- Substantially increased patient function
- Passive wrist motion increased 50%

## **Smith 2002**

- Serial casting 17 wrists (12 amyoplasia, 5 distal)
- Average follow up 6 years
- Greatest gains after first casting session
- Average final correction of 33 degrees - wrist
- Distal group responded best – no recurrence
- Amyoplasia group – high rate of recurrence and less improvement

# Fit Early Custom Orthotics or Serial Casts



# Therapeutic Techniques to Maximize PROM

- Sing a nursery rhyme song during stretching
- Massage tight muscles held at end range
- Use of heat prior to stretching
- Joint mobilization and distraction
- Upper extremity weight bearing





# Strengthen Available Musculature

- Improved PROM can unmask active function
- Therapeutic techniques to maximize strength
  - antigravity movements
  
- Select the “just right” activity
  - motivate the child
  - ensure success



# Activities of Daily Living

Most children age 5 and older with amyoplasia were ambulatory and relatively independent in activities of daily living

Ambulatory	– 85%
Feeding	– 75%
Toileting	– 35%
Bathing	– 25%
Grooming	– 20%
Dressing	– 10%

Sells JM, Jaffe KM, Hall JG. Amyoplasia, the most common type of arthrogryposis: the potential for good outcome. *Pediatrics*. 1996 Feb;97(2):225-31. PMID: 8584382.

# Self-Feeding is a Top Priority

- Functional activity to work on elbow mobility
- 90 degrees passive elbow flexion

Arm push method



Table push method



# Self-Feeding Adaptive Devices

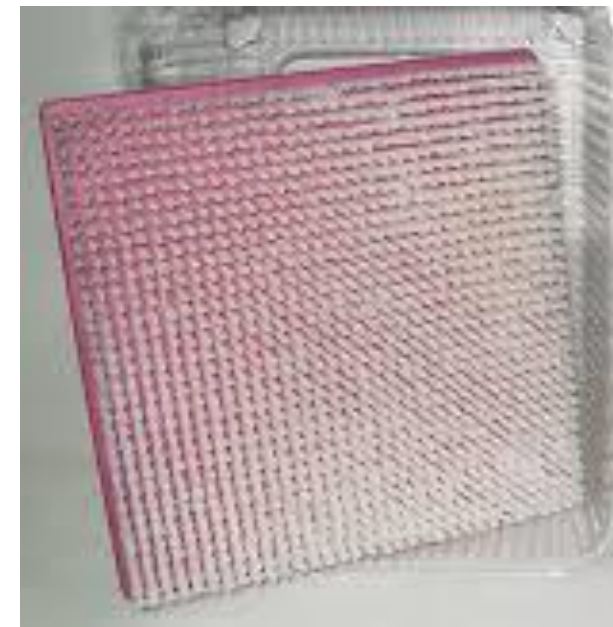


# Toileting

- Difficulties reaching when wiping
- Adaptive techniques (to improve arm reach)
- Extended handle bottom wiper
- Bidet



## Bathing: Equipment for Limited Reach



# Grooming: Extended Handle Devices



# Upper Body Dressing Tree



- Position the height of the hooks mid back level
- Use the hooks to support the shirt
- Work your body into the shirt



# Difficulties Reaching Feet



# School Skills

- Determine if any adaptive devices or adaptations may be needed
- Forearm rests, split key boards
- Tilted or lowered work surfaces



# Wheeled Mobility

- Manual versus power wheelchair
- Vehicle, home and car accessibility
- Need for custom seating



# Resources to learn more

- You Tube videos of persons with arthrogryposis with clips for inspiration or “what works for me”

Misha Dream Walker

Rexi James

Chris Hartwick

- Arthrogryposis Multiplex Congenital Support Group (AMCSI)

On-line resources: diagnosis, treatment, adaptive devices

- Facebook group

<https://www.facebook.com/athrogryposis>

- Pinterest

[83 Arthrogryposis ideas | adaptive equipment, encouragement, adaptive devices \(pinterest.com\)](#)



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- <https://www.facebook.com/arthrogryposis>
- [Living with AMC — AMC Support](#)
- [83 Arthrogryposis ideas | adaptive equipment, encouragement, adaptive devices \(pinterest.com\)](#)
- Ann Van Heest : Arthrogryposis Presentation 2017 University of Minnesota
- Mary Beck OTR/L: Treatment recommendations from the Paley Institute [Homepage - Paley Orthopedic & Spine Institute \(paleyinstitute.org\)](#)

Thank you for your attention!



