# *Transition times.*
# *Supporting the child and adolescent's transitions in Pediatric condition*

2nd S.I.P.Ped. International Conference
November 7 – 9, 2019
Palermo, Sicily ,Villa Magnisi, Ordine dei Medici Chirurghi e Odontoiatri

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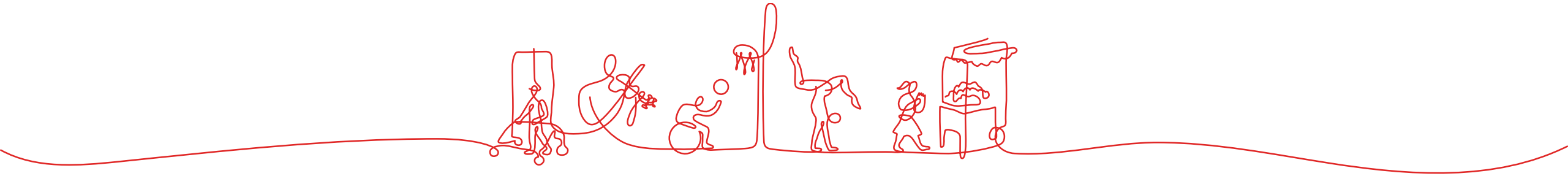

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# Beyond Broken Bones: Psychological Treatment of Pediatric Orthopedic Conditions

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UT Southwestern Medical Center

# Buongiorno da Dallas, Texas!


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Psychology Faculty

# Disclosures

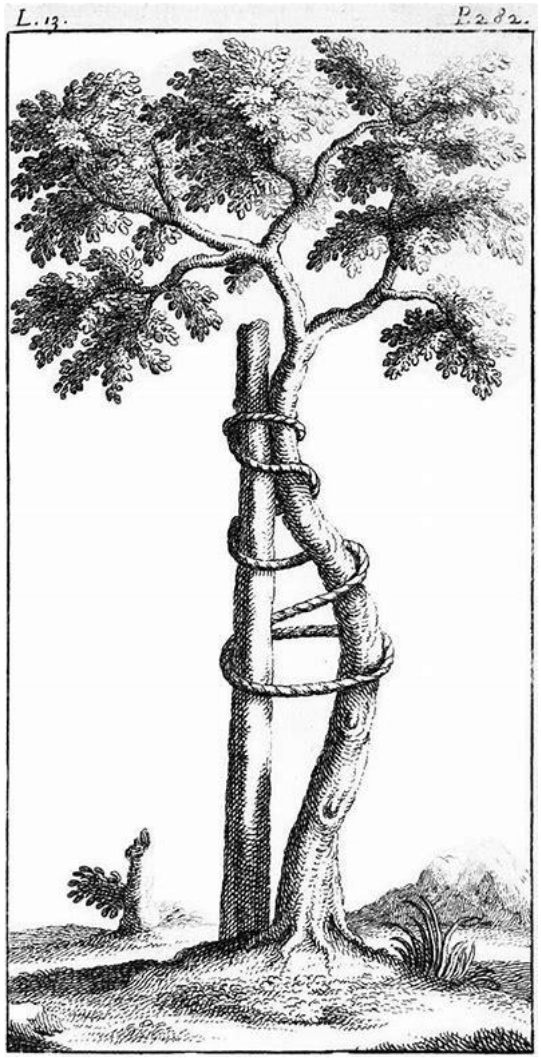
- **Nothing to Disclose**

# Objectives

- Recognize the role and importance of pediatric psychology in the treatment of congenital orthopedic conditions.
- Discuss opportunities for pediatric psychology in multi-disciplinary research studies and in improving post-surgical outcomes.
- Review the unique aspects of treating children and adolescents with orthopedic impairments by highlighting several case examples.
- Describe the difficulties associated with transitioning pediatric orthopedic patients to adult providers.



# Pediatric Orthopedics: A Brief History




**ORTHOS (Straight) and PAIDIOS (Child)= Orthopedics**

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# Documentation of Pediatric Orthopedic Conditions in Art

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# Documentation of Pediatric Orthopedic Conditions in Literature








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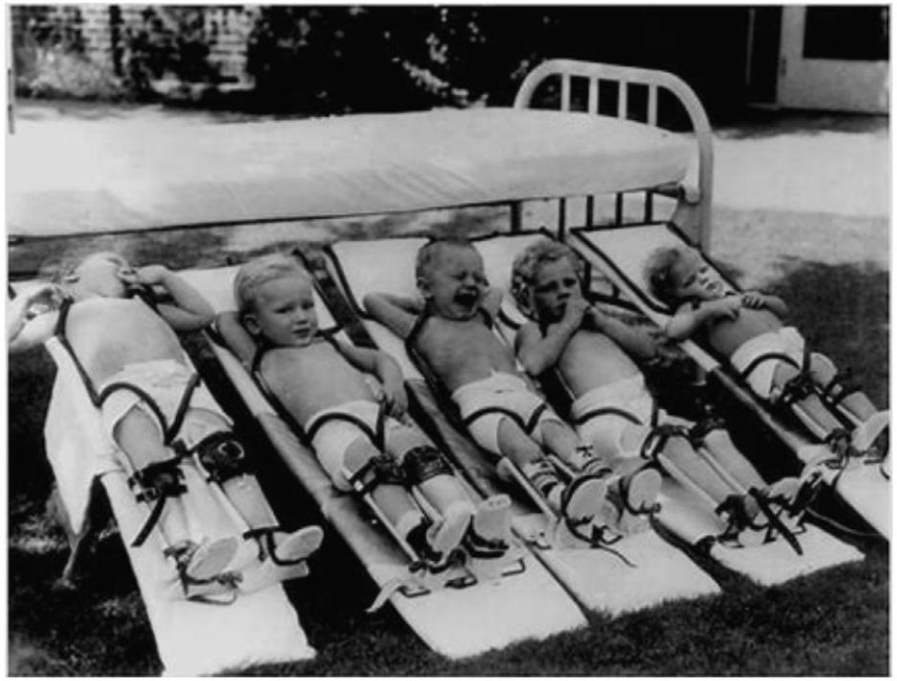
# Historic Treatment of Crippled Children

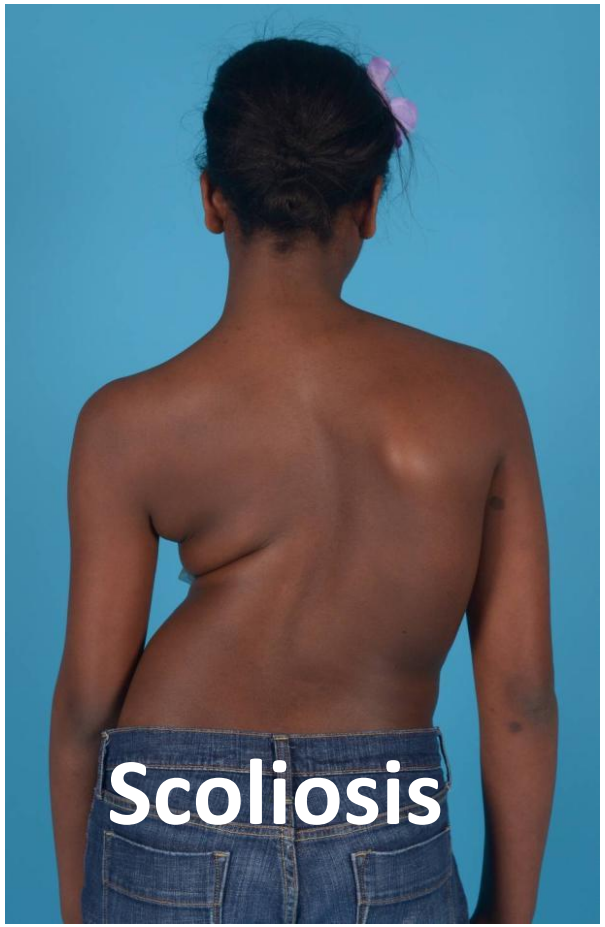


# Prevalence Rates of Chronic Illness in the US

- **Diabetes**—7 in 100,000 children age 4 and younger are diagnosed;
  15 in 100,000 children between the ages of 5 and 9 are diagnosed; and
  22 in 100,000 children between the ages of 10 and 14 are diagnosed
  with Type 1 Diabetes each year.
- **Sickle Cell Disease**—1 in every 300-500 African American newborns diagnosed each year.
- **Juvenile Rheumatoid Arthritis**—14 per 100,000 children diagnosed each year.
- **Cerebral Palsy**—1 to 3 per 1000 children (9000 each year)
- **Orthopedic Conditions (*Sample*)**
  - **Scoliosis**--3 million new cases of the condition are diagnosed in the United States each year, with a majority of them identified as idiopathic scoliosis — a type of scoliosis that presents in children between 10 to 12 years old.
  - **Clubfoot**—1 out of every 1000 children are diagnosed at birth each year
  - **Proximal Focal Femur Deficiency-**-1 in every 200,000 children are affected (varies in severity)

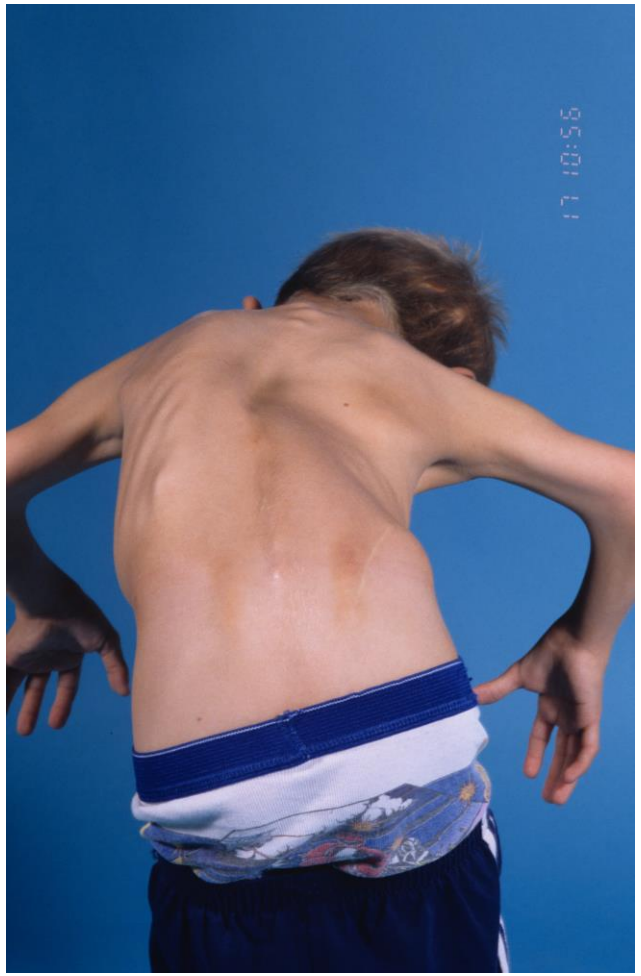
# What Do These Disorders Have in Common?

- Angelman Syndrome
- Conradi Hunermann Syndrome
- Duchenne Muscular Dystrophy
- Klippel-Feil Sydrome
- Larsen Syndrome
- Noonan Syndrome
- Marfan Syndrome
- Neurofibrometosis
- Prader-Willi Syndrome
- Rett Syndrome
- Spinal Muscular Atrophy

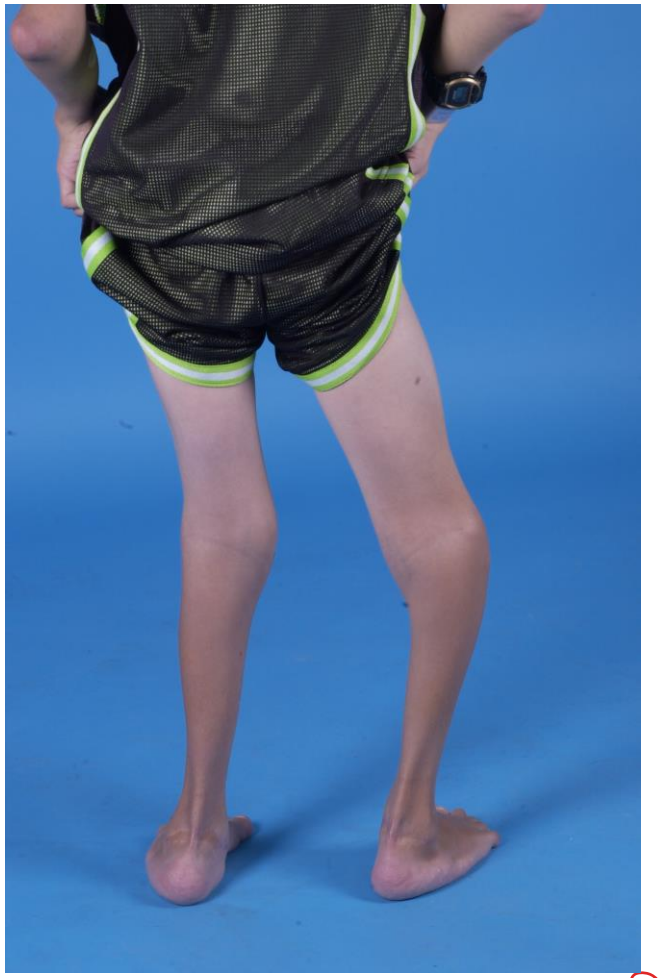



rarediseases.org

# Orthopedic Treatment ≠ Always One Surgery

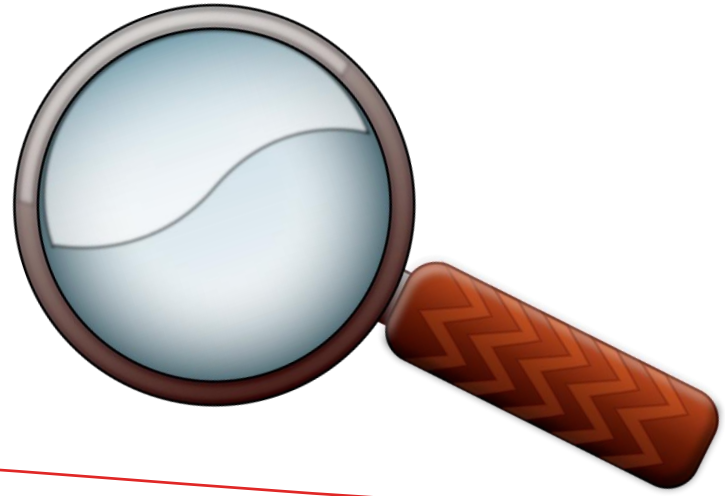


- Many children with orthopedic problems also have other medical diagnoses.
- Some children will need multiple orthopedic surgeries in addition to treatment for their other conditions.
- Many children with multiple diagnoses have a history of hospitalizations at several different institutions.
- The child and his/her family are often overwhelmed and exhausted due to the number of medical appointments and the schedule for ongoing treatment.



# Where is the Research on Orthopedic Conditions in the Pediatric Psychology Literature?

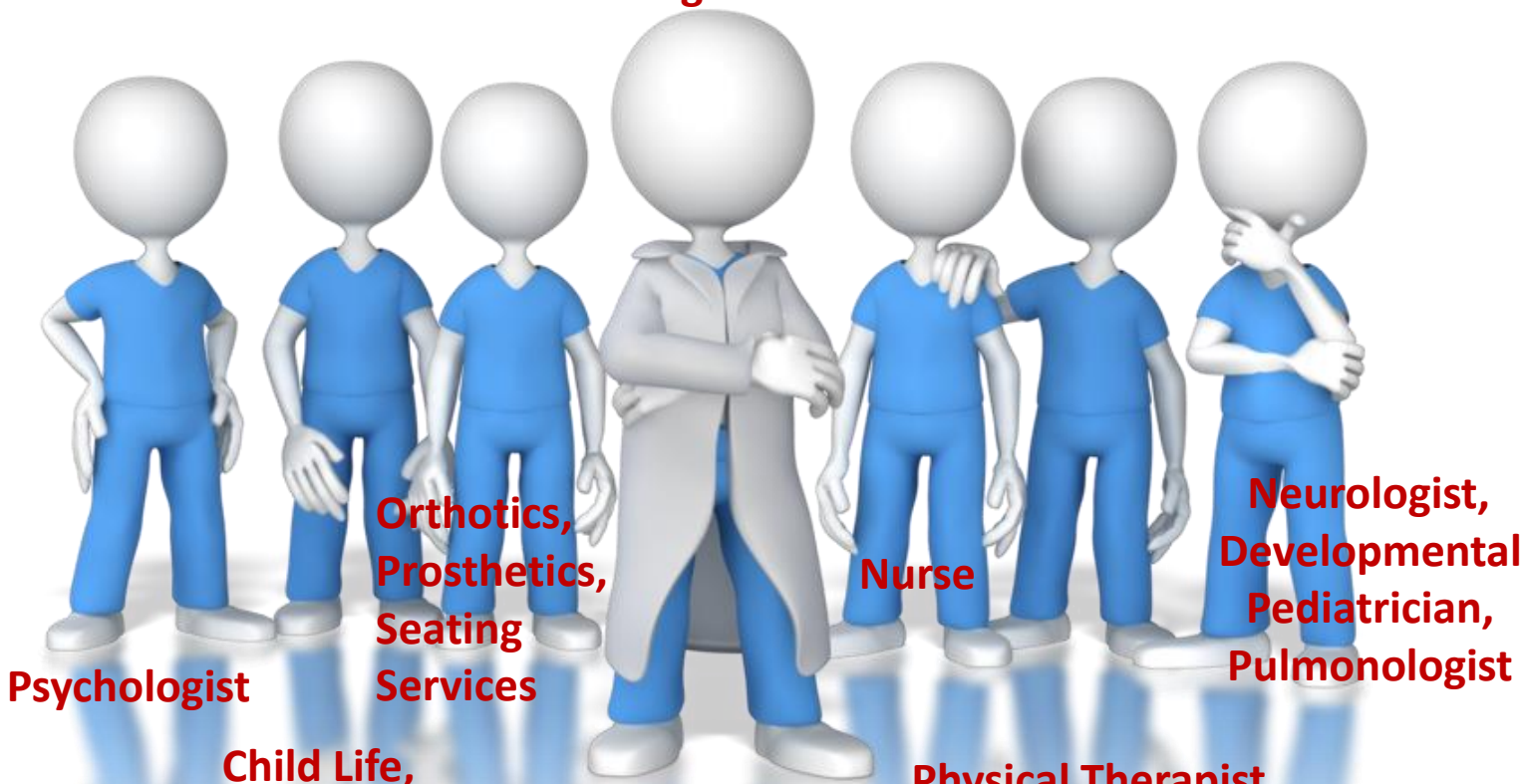
- # of articles in JPP in the last 10 years? 55 addressed orthopedic issues (mostly TBI/Injury)
- # of articles that address scoliosis? 20 articles throughout all of the years
- # of articles that reference psychological constructs in the Journal of Pediatric Orthopedics? 7 in 2019 alone!



# Psychologists Are Needed in Pediatric Orthopedics

# The "TEAM" Needed to Work With Complex Pediatric Orthopedic Patients

And, the child may have a host of other providers in different subspecialties… Oncology, Metabolic Clinic, Rheumatology, etc. in multiple locations.



# Children with Orthopedic Impairments Often Have Visible Differences

# Psychological Issues Faced by Pediatric Orthopedic Patients

- Anxiety and Depression (fear of surgery, treatment, and recovery)
- Behavioral Issues
- Self-esteem (conscientious about their appearance and functional differences)
- Body image
- Self-efficacy
- Pain Management Issues
- Functional Disability
- Peer Relationship Issues
- School Issues
- Family Issues
- Compliance with Treatment (bracewear, non-weight bearing, etc.)
- Medical Decision Making/Health Related Quality of Life



# Common Reasons for Referral to Psychology

- Poor coping skills
- School refusal
- Pain management concerns
- Pre-surgical preparation
- Medical decision making
- Best Practices Advisory (BPA)—*responses to a depression screener given in clinic*
- Peer related concerns
- Family concerns
- Noncompliance with treatment
- Issues related to cognitive, behavioral, or academic functioning
- Concerns with activities of daily living and/or adaptive skills, etc.

# Role of the Pediatric Psychologist

**Psychologist is an integrated member of the treatment team (in Ambulatory Care Clinic and on the Inpatient Unit) who:**

- **Provides Consult-Liaison Services  (integrated in clinics)**
- **Outpatient therapy and assessment**
- **Pre-surgical evaluation and preparation**
- **Inpatient care**
- **Participates in Training (Postdoctoral Fellows/Interns/Practicum students in psychology as well as consults with medical students, Orthopedic Residents, and Orthopedic Fellows)**
- **Conducts Multi-disciplinary Research**

# Presurgical Evaluation

- Assess relevant psychosocial factors that may impact disease etiology, course, and/or treatment.
- Look at family structure, organization, & support; cognitive, behavioral, & social functioning; and history of mental health symptoms and intervention.
- Determine risk for depression/self-harm.
- Assess knowledge of diagnosis; knowledge of operative plan; knowledge of post-operative expectations.
- Review adherence history.
- Evaluate pain symptoms and knowledge of pain management strategies.

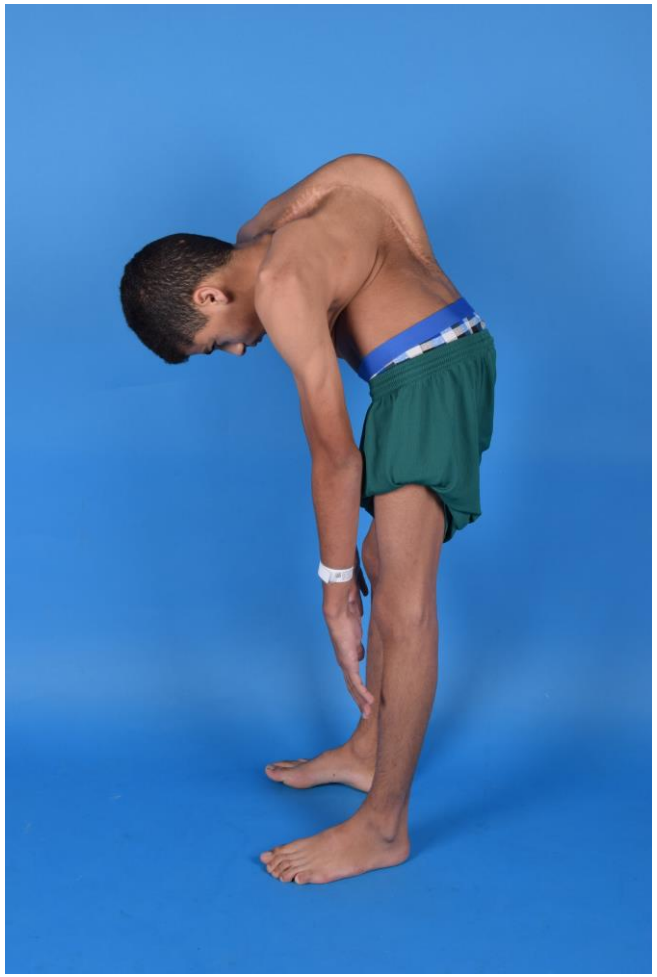
# Inpatient Treatment

- Participate in ongoing patient education
- Track relevant psychosocial factors that may impact disease etiology, course, and/or treatment
- Provide ongoing intervention throughout stay:
  - Pain management
  - Anxiety management
  - Mood management
  - Behavior management
- Provide referrals to community providers upon discharge

# Case Examples

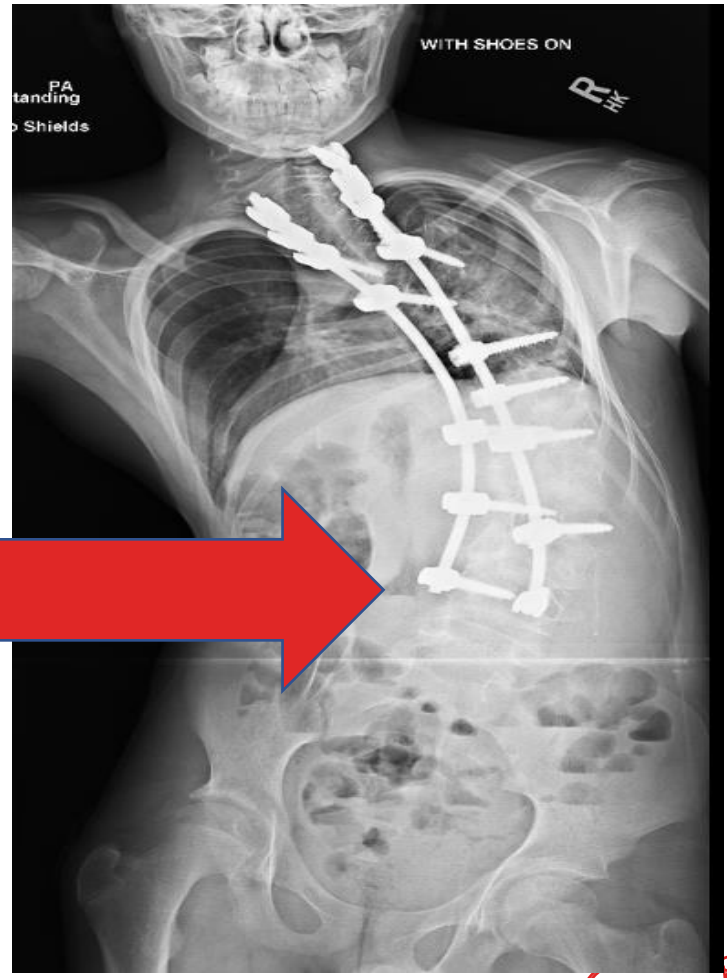


# Spine Deformity:  Kyphoscoliosis



- 14 year, 11 month old Middle Eastern male from Palestine who came to the US for evaluation and treatment of neuromuscular scoliosis.
- History of being bullied and treated like an outcast in his community because of his physical appearance.
- Patient was upset after surgery that his deformity was not 100% corrected (expectations were inappropriate).
- Anxiety over returning home, feared that he would continue to be socially rejected.

# Timing of Surgical Intervention Matters!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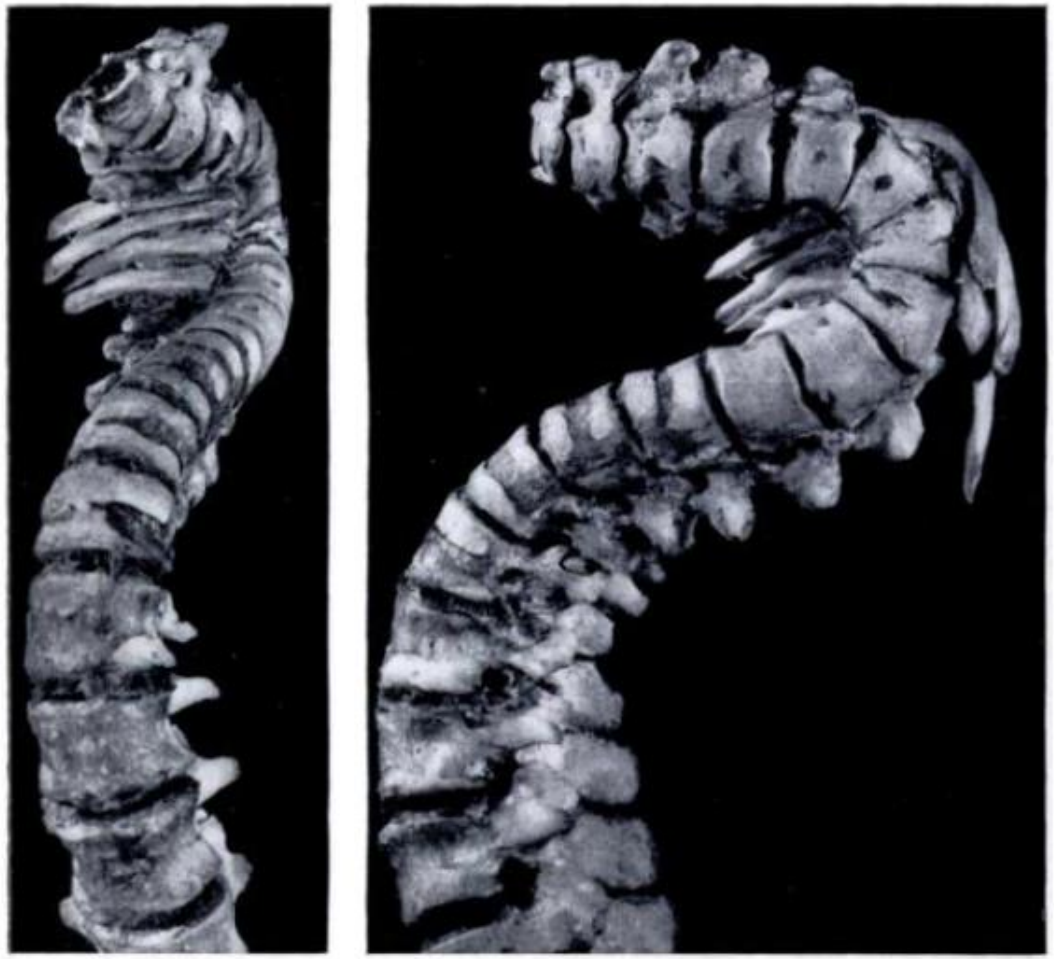
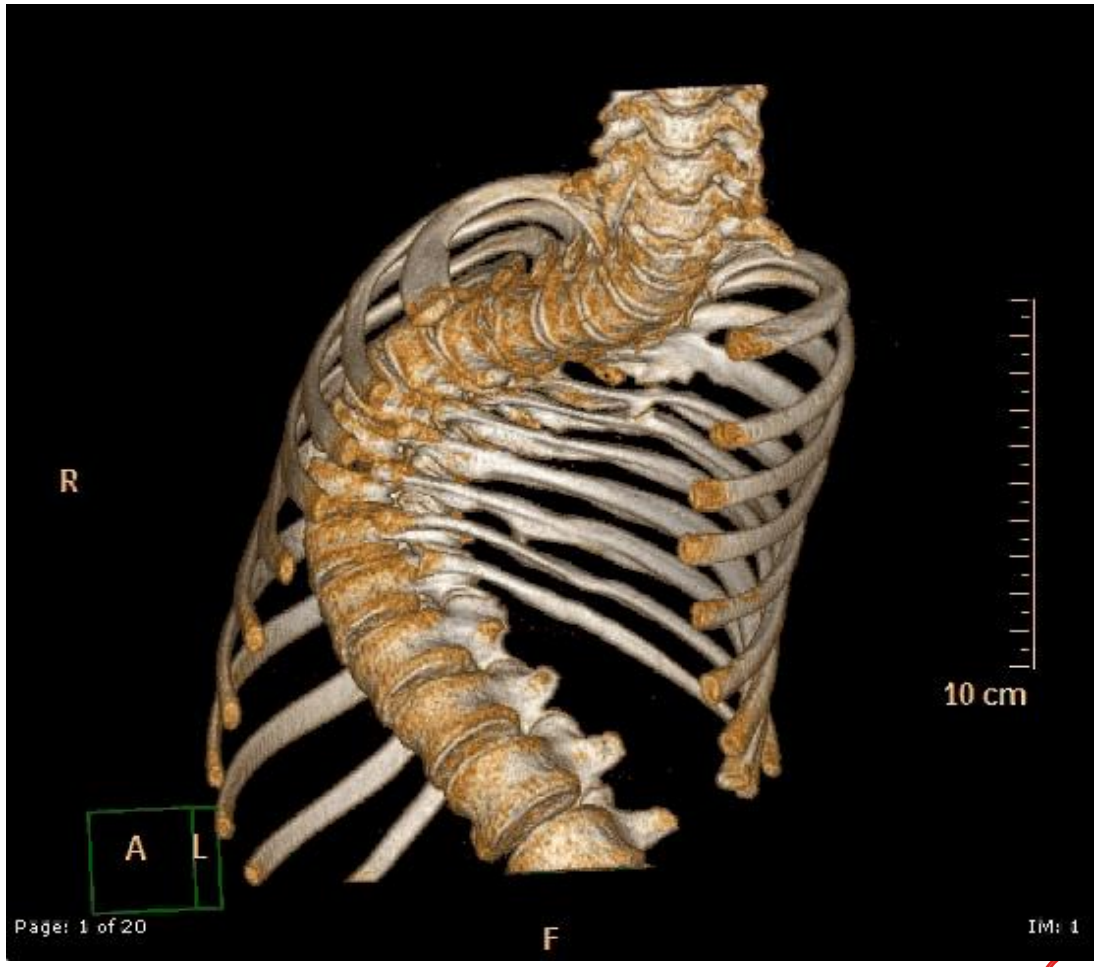
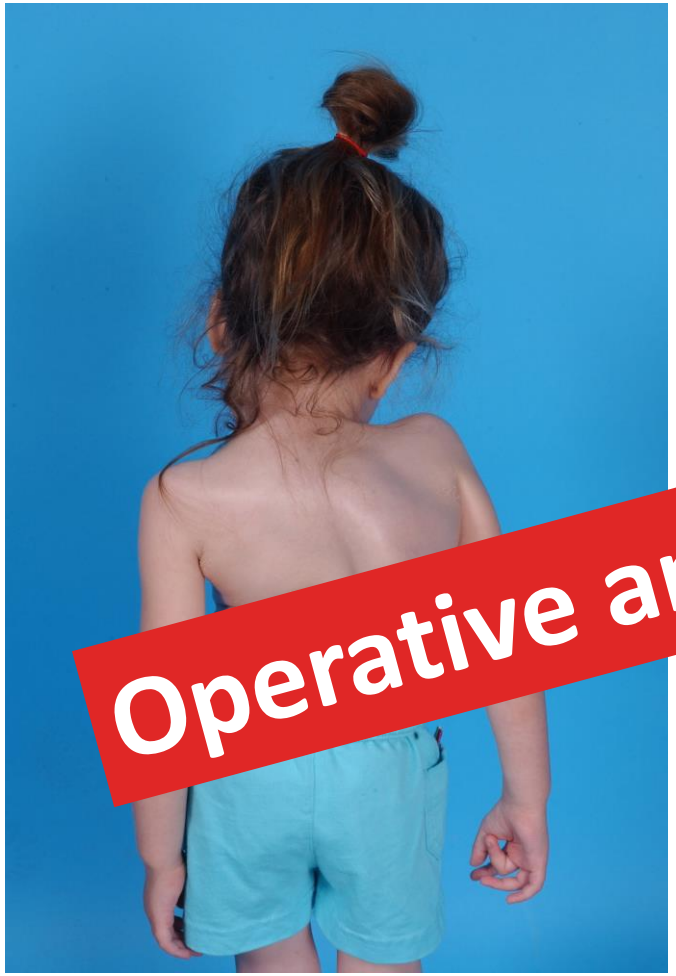
# Spine Deformity of Early Childhood Is Potentially Fatal (surgery is not just for cosmetic reasons)





# Early Onset Scoliosis (lengthy course of treatment)



**Operative and Non-operative Treatment**

Matsumoto, Williams, Corona, et al. (2014)

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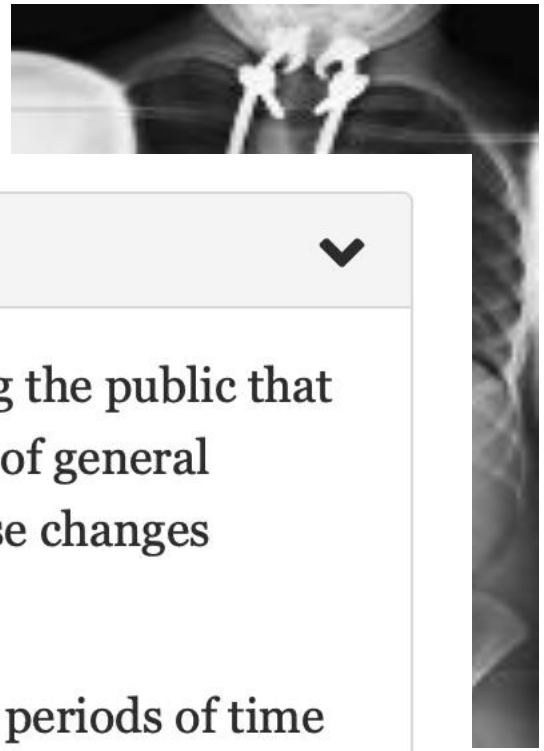
# Sample EOS Treatment Schedule



- 5/2006 Cast (Anesthesia #1)

**Safety Announcement**

**[ 4-27-2017 ]** The U.S. Food and Drug Administration (FDA) is notifying the public that we have approved previously announced label changes regarding the use of general anesthetic and sedation medicines in children younger than 3 years. These changes include:

- A new Warning stating that exposure to these medicines for lengthy periods of time or over multiple surgeries or procedures may negatively affect brain development in children younger than 3 years.

- 2/2013 Lengthening and revision proximal anchors (Anesthesia #13)
- 10/2013 Lengthening (Anesthesia #14)
- 2/2015 Lengthening **(Anesthesia #15)**

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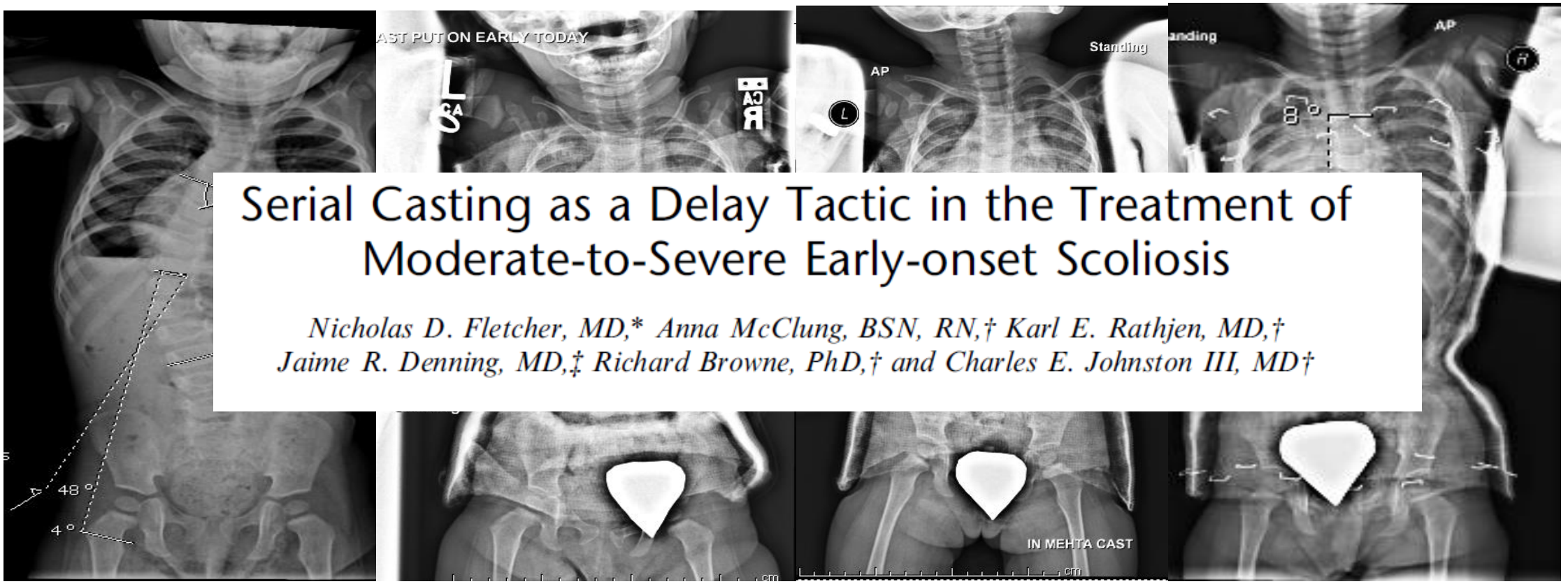
# Mehta Cast Series




Serial Casting as a Delay Tactic in the Treatment of Moderate-to-Severe Early-onset Scoliosis

*Nicholas D. Fletcher, MD,* Anna McClung, BSN, RN,† Karl E. Rathjen, MD,† Jaime R. Denning, MD,‡ Richard Browne, PhD,† and Charles E. Johnston III, MD†*

# Mehta Cast

- Made of synthetic materials
- Cannot get cast wet (need to protect it with plastic bags)—no bath, no pool!
- Parents often struggle with not being able to hold their child without cast.

Ramo and Colleagues (2015) looked at 34 patients and found that on average,
Age at 1st cast: 20.5 months
Total time in cast: 11.6 months
Number of casts: 5

# Growing Rods


## Psychosocial Effects of Repetitive Surgeries in Children With Early-Onset Scoliosis: Are We Putting Them at Risk?

*Hiroko Matsumoto, MA,* Brendan A. Williams, BA,* Jacqueline Corona, MD,† Jonathan S. Comer, PhD,‡ Prudence W. Fisher, PhD,§ Yuval Neria, PhD,§ Benjamin D. Roye, MD, MPH,*‖ David P. Roye1, MD,*‖ and Michael G. Vitale, MD, MPH*‖*

## Psychological Dysfunction in Children Who Require Repetitive Surgery for Early Onset Scoliosis

*John M. Flynn, MD,* Hiroko Matsumoto, MA,† Frances Torres, PhD,‡ Norman Ramirez, MD,* and Michael G. Vitale, MD, MPH§*

# Before and After Growing Rods

**Age 7**

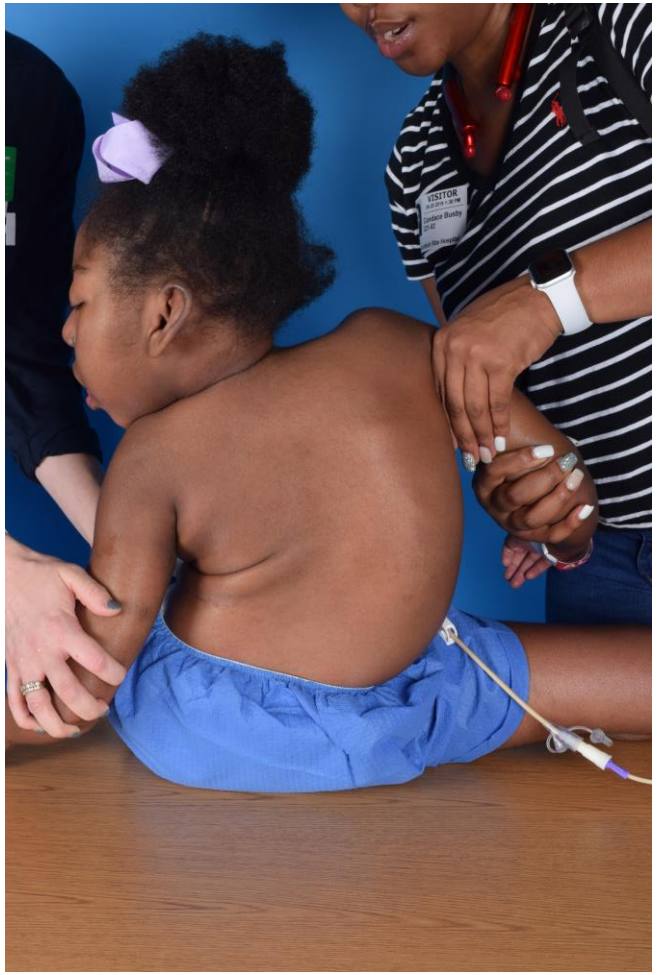
**Age 8**

**Age 9**

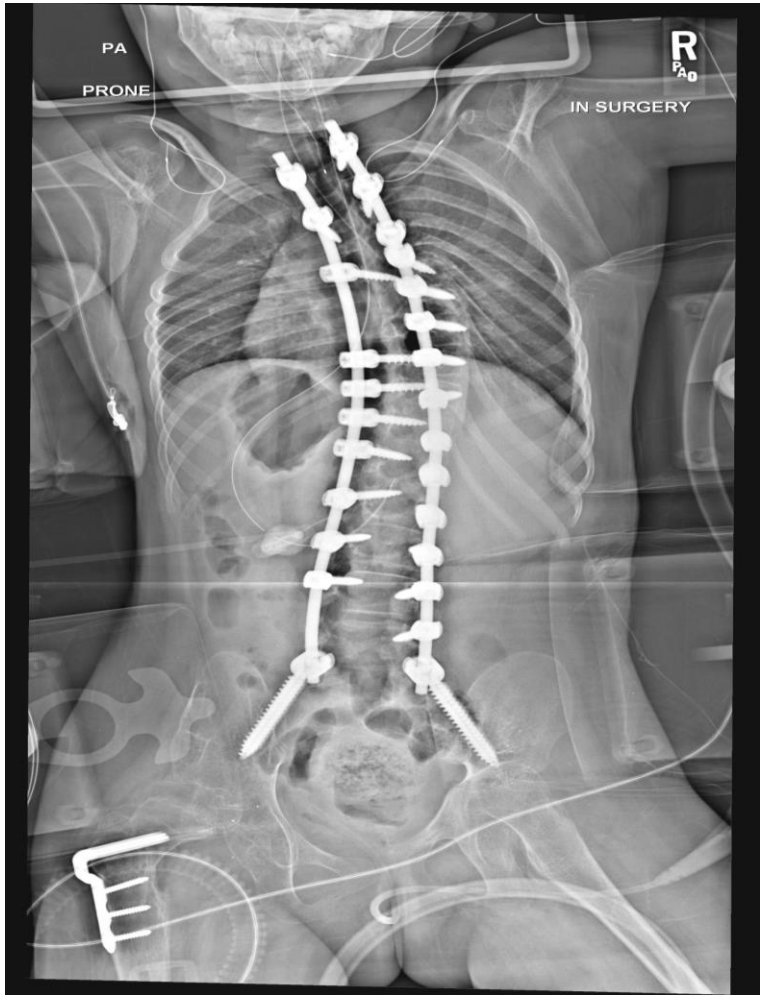
**Age 10**

# Neuromuscular Scoliosis

- Families decide to pursue surgery due to medical concerns related to the progression of their child's spinal deformity.
- Families are usually interested in improving their child's quality of life.
- Families do not view the surgery as "optional" but rather as "life saving."
- To assist these families with medical decision making, a specialty clinic was developed to allow families to meet providers on one day which allows providers to consult and participate in a staffing.

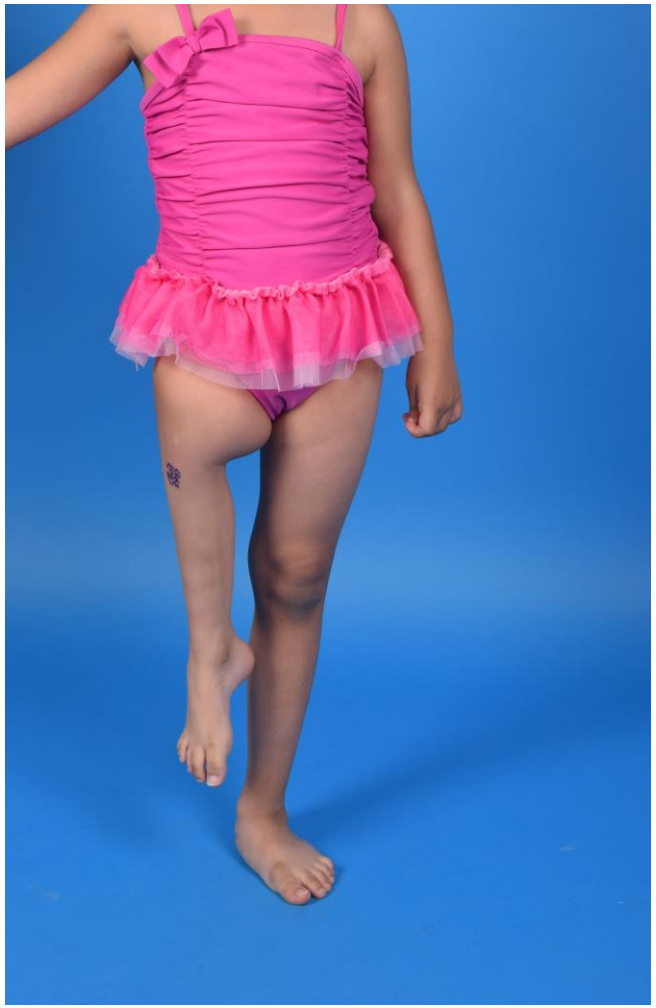


# Benefits of Spine Surgery in Neuromuscular Scoliosis



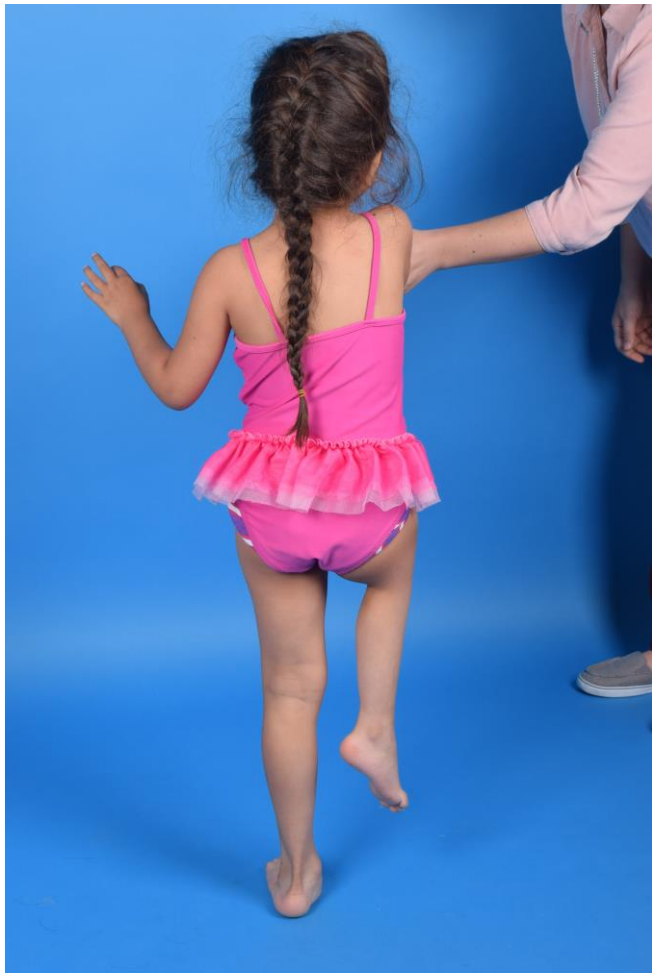

- Occurs in children with underlying neuromuscular disease.
- More common and severe in non-ambulatory patients (those whose neuromuscular disease prevents them from walking).
- Curve is almost always progressive.
- Progression of the curve can lead to a reduction of lung space, limiting lung function, and increasing the risk of pneumonia.
- Surgery stabilizes the curve and stops its progression.
- Regains or maintains the ability to sit upright.
- Improves/preserves lung function.

# Proximal Femoral Focal Deficiency



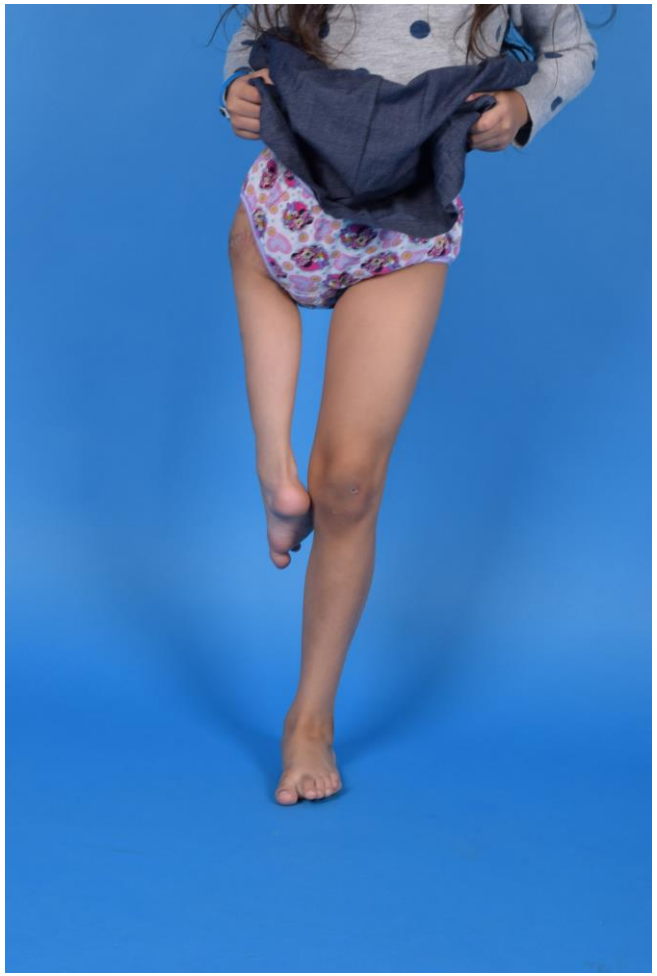
- 5 year, 1-month old Hispanic female from Honduras was brought to the United States for evaluation and treatment of a proximal femoral focal deficiency of her right lower extremity.
- Referred for a psychological consultation prior to surgery to confirm that she understood how the appearance of her limb would change and that she would continue to wear a prosthesis.
- Addressed previous history of bullying, self-acceptance, anxiety over medical procedures, and fear of being “stared at” by strangers.

# Before and After Rotationplasty: What is Different?



- Prior to Rotationplasty, patient used an equinous prosthesis. It is like having an AFO on a pylon and a prosthetic foot (rather than having an extremely large shoe lift).
- Patient could not wear pants (due to having her foot plantigrade on a stand), she could not bend her leg when sitting at a desk (had to keep her leg straight), and her difference was always visible.
- She did not attend school prior to her Rotationplasty due to bullying.

# What is a Rotationplasty?

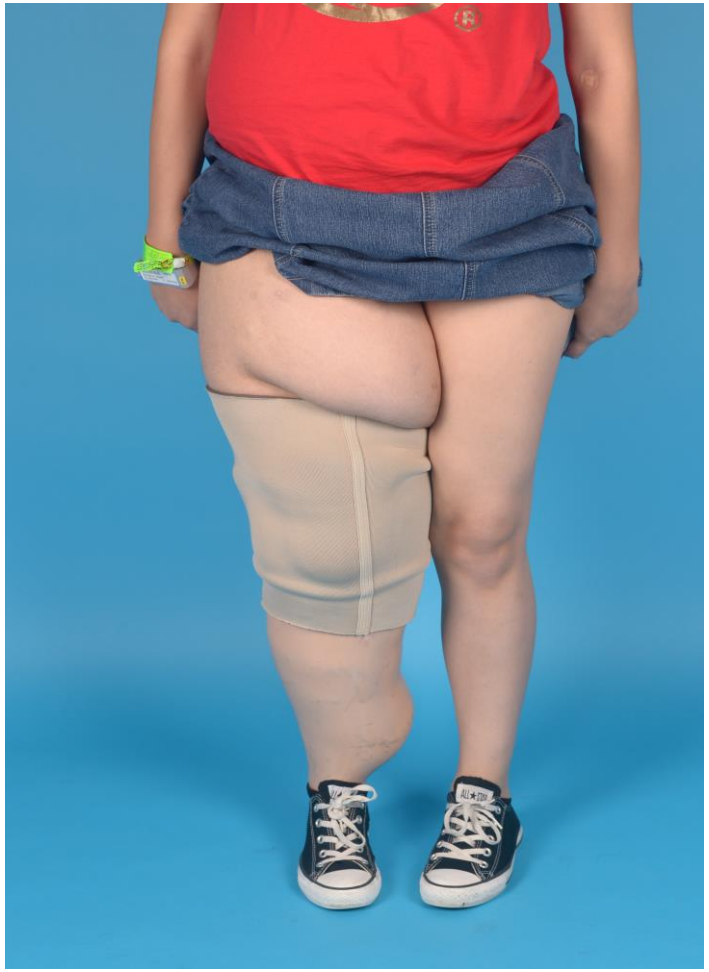


- The child's limb is surgically cut, rotated 180 degrees and reattached.  This allows the child's ankle to assume the function of a knee (improved function with a biologic knee instead of a mechanical one).
- The child then will use a below-the-knee prosthetic.
- She can bend her knee, sit in a chair (not having her leg straight), and ride a bicycle.

# CLOVES Syndrome

- 12 year, 9-month old Asian female with a history of CLOVES syndrome who was recently adopted from China (came to the US on January 19, 2018). She was referred for a psychological consultation to assess her coping skills and understanding of her diagnosis/treatment.
- CLOVES Syndrome stands for Congenital, Lipomatous, Overgrowth, Vascular Malformations, Epidermal Nevi and Spinal/Skeletal Anomalies and/or Scoliosis (Gene mutation (PIK3CA).
- Due to the overgrowth syndrome, she also has a lower limb length difference.

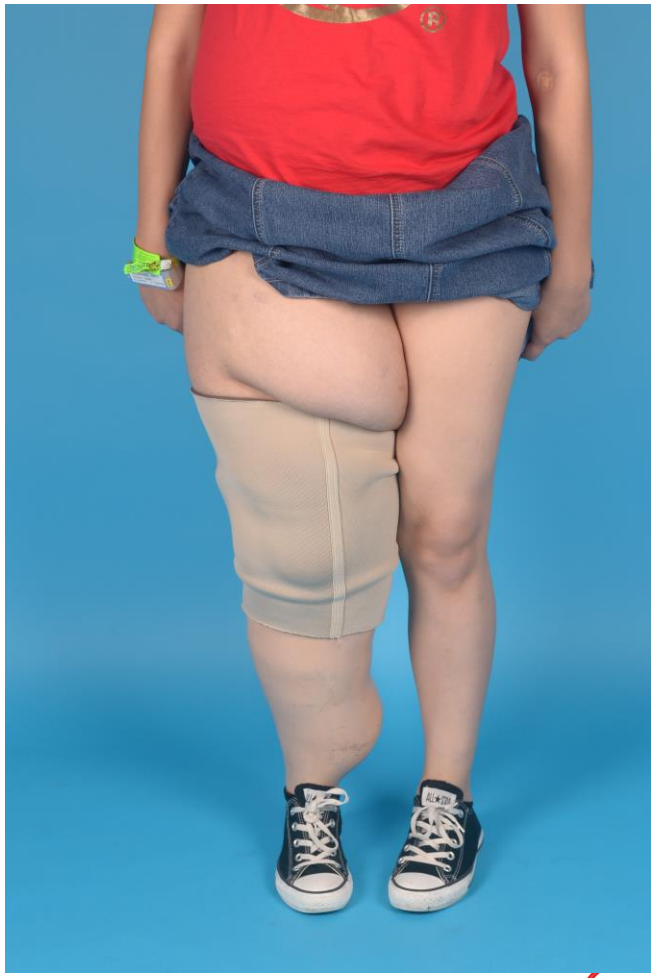
# Medical Decision Making:  Amputation



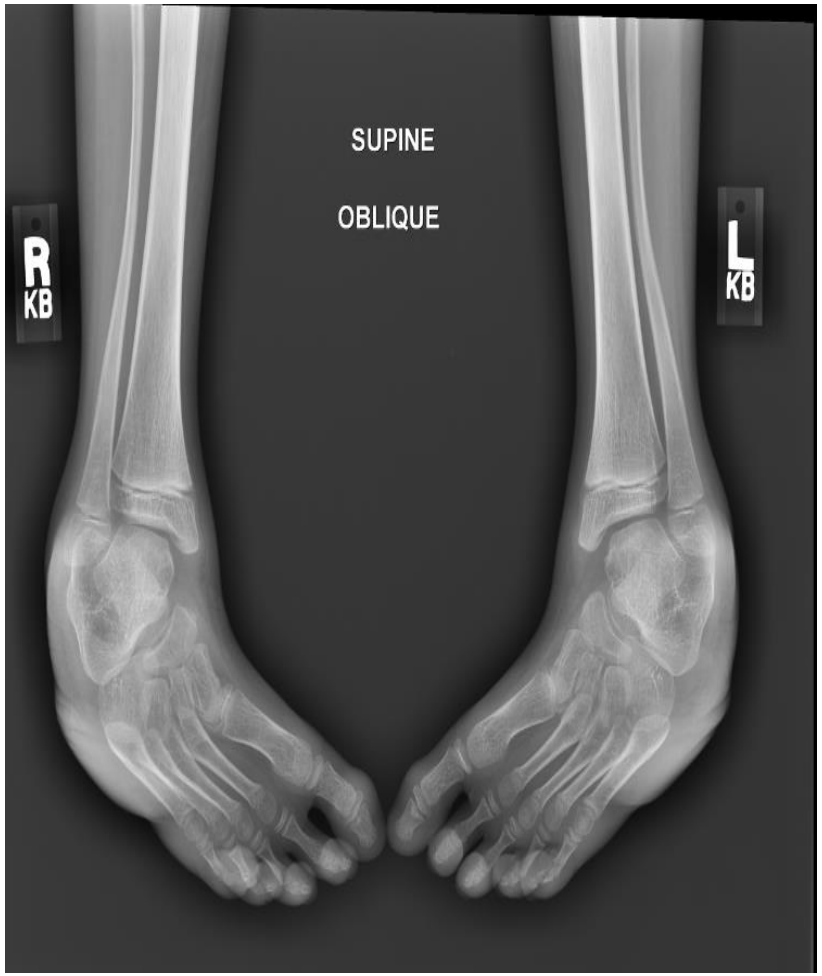
- Focus on function (and ability to engage in activities of daily living).
- Psychoeducational information was provided about possible treatment options.
- Patient and family met with a peer model who had undergone the same surgery several years before.
- Peer model and her mother discussed the advantages and disadvantages of having an amputation.
- Patient allowed to ask questions.

# Which Leg is More Functional?

- Psychologist assisted patient and family with medical decision making (patient recently had an amputation of her left foot).
- Focus was on improving function.
- Use of peer model helped patient to see what her leg would actually look like and how an amputation would impact her daily life.
- Discussion focused on activities of daily living and how the peer model's life changed after having an amputation.



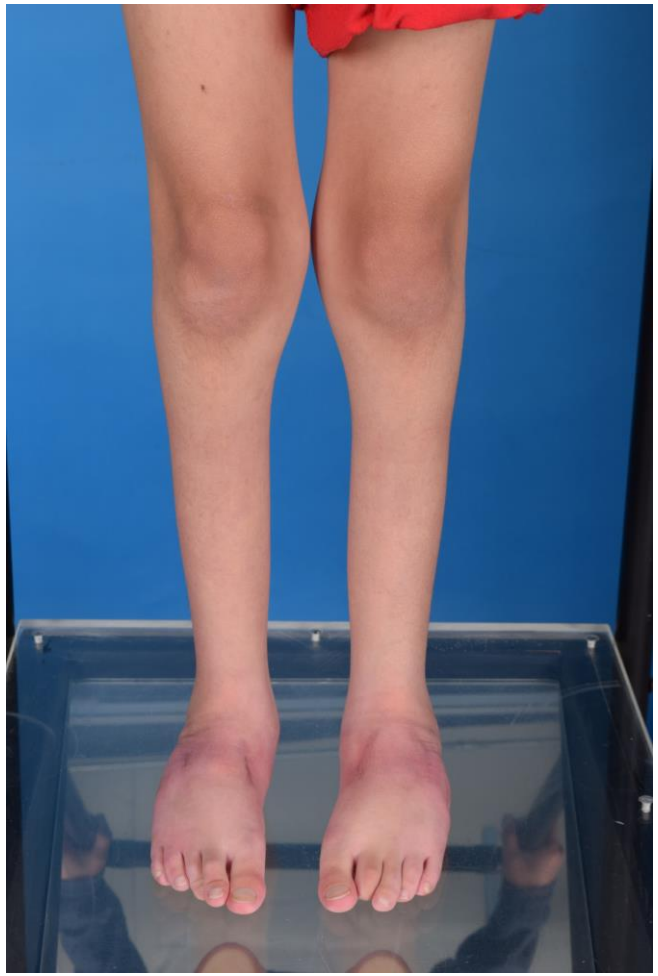
# Clubfoot




- Clubfoot is a congenital deformity that occurs in 1 in 10,0000 birth.
- It is more common in boys.
- Treatment consists of casting (Ponseti Method) to move the foot into an improved position.
- If non-operative treatments are not successful or achieve incomplete correction, surgery is often recommended.

# Serial Casting and Surgery to Correct Deformity

- 12 year, 9-month old Asian male who lived in a Buddhist monastery in Bhutan when a tourist noticed his uncorrected clubfeet.
- After arriving in the US, he underwent Ponseti casting for a month before undergoing a bilateral open Achilles tendon lengthening, plantar fascial release, and tibialis anterior transfer.
- He was in his casts for a total of 6 weeks before transitioning to AFOs for a short period of time.



# Orthopedic Surgery Provides Patients with Visible Changes

# Research to Improve Clinical Practice

- **Involvement in multi-disciplinary research allows us to treat the "whole" child.**
- **Improves patient outcomes.**
- **Increases patient and parent satisfaction.**
- **Opportunities to collaborate across providers and institutions to improve patient care.**

# Examples of Current Research

- **Pain Catastrophizing as a Predictor of Post-surgical Pain in AIS Patients**

- **Cognitive and Behavioral Functioning in Early Onset Scoliosis:  A Longitudinal Study**

- **Complex Care and Challenging Decisions: Parent Perceptions Surrounding Corrective Spinal Surgery in Pediatric Patients with Neuromuscular Scoliosis**

# Differences Between Medical Homes

## Pediatric

Access to a team of medical providers

Access to ancillary and support services (more likely to be integrated)

Availability of resources

## Adult

Often only see their physician during a visit

# Barriers to Transitioning to Adult Services

- **Pediatric Orthopedic Surgeons have a difficult time finding providers who have specialized knowledge about community resources for young adults with congenital disorders or pediatric-onset chronic diseases.**
- **Limited availability of adult specialty clinicians who feel qualified to work with this population.**
- **Lack of mental health and supportive services.**
- **Adult providers are not familiar with caring for patients who are reliant on caregivers.**
- **Loss of insurance coverage (some things no longer covered after they leave a pediatric setting) and cost of care.**
- **Families feel “lost” when navigating the adult system.**
- **Lack of available resources (without integrated care, often not shared by provider).**

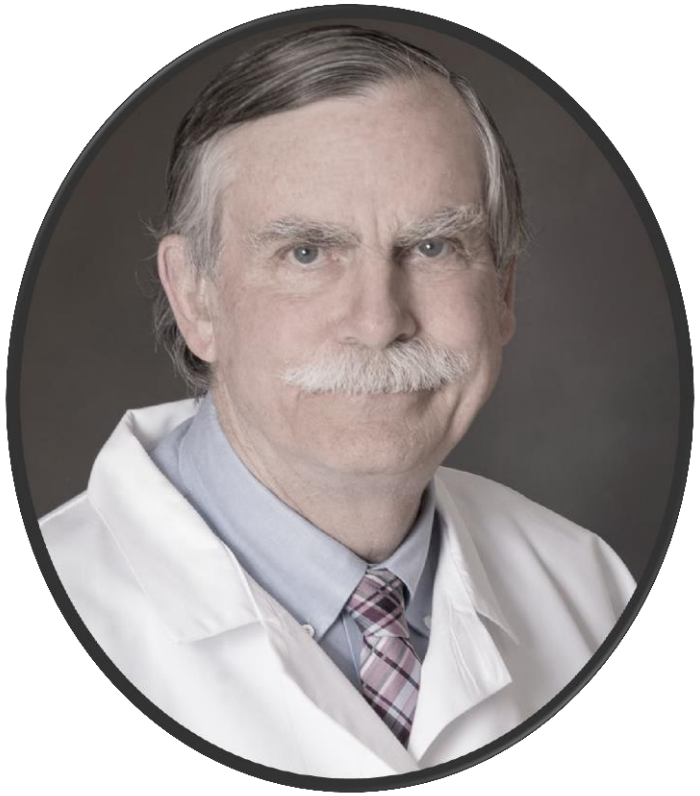
White & Cooley (2018)

# Future Directions

- **Increase the role of Pediatric Psychology in the assessment, treatment, and research conducted in Pediatric Orthopedics.**

- **Propose that pediatric psychologists from multiple institutions collaborate to develop programming to meet the needs of pediatric orthopedic patients.**

# Pediatric Orthopedic Surgeons Who Treat the "Whole" Child



Charles E. Johnston, MD

Thank you!

Amy L. McIntosh, MD

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# Grazie S.I.P.Ped. International Conference!

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