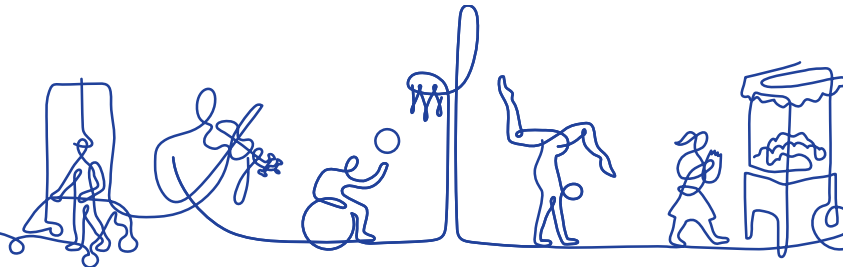


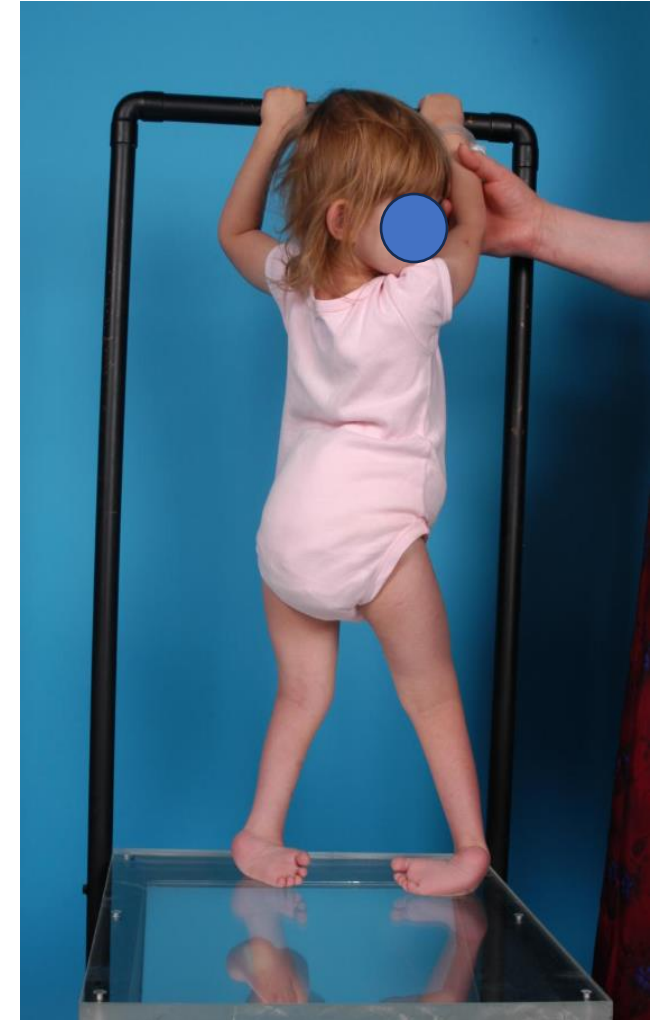
Orthopedic, Holistic, and Surgical Approaches to Lower Extremity Issues in Myelomeningocele: Lifespan Perspectives

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Will she walk ?



Parental Expectation

- Not walking is “worst disability”
- This misconception should not be surprising
 - First two years of life almost all “comparisons” are gross motor milestones
 - Admittedly the “thief of joy”, but every parent is “guilty”!
- Opportunity to educate:
 - Complex chronic medical condition effecting multiple organ systems
 - Condition we “manage”, not “cure”
 - Goal is to improve the function, limit “patient time”, maximize “child time”

Multidisciplinary Team

Developmental Pediatrician

Neurosurgeon

Closure
Hydrocephalus

Orthopedic surgeon

Paralysis/ambulation
Foot deformities
Knee contracture
Hip dislocation
Scoliosis
Lordosis
Wounds



Urological surgeon

Incontinence
Retention
Diverticula
Pyelonephritis
Renal failure

Parental *Education*

- Factors Effecting Independence
 - Communication
 - Behavior
 - Education
 - Mobility ??

Education and employment as young adults living with spina bifida transition to adulthood in the USA: A study of the National Spina Bifida Patient Registry

Tiebin Liu¹  | Lijing Ouyang¹ | William O. Walker^{2,3} | John S. Wiener⁴ |
Jason Woodward⁵ | Jonathan Castillo⁶ | Hadley M. Wood⁷ | Stacy T. Tanaka⁸ |
Richard Adams⁹ | Kathryn A. Smith^{10,11} | Joseph O'Neil¹² | Tonya R. Williams¹ |
Elisabeth A. Ward^{1,13} | Robin M. Bowman¹⁴ | Catharine Riley¹

- In this main analysis, any employment was independently and significantly associated with higher education level, sacral lower extremity functional level, being continent of bowel, a history of non-shunt surgery since last visit, and having private health insurance.
- In the subsample of visits with any employment full-time employment was independently and significantly associated with older age, higher education level, non-myelomeningocele spina bifida type, being a community ambulator, and without skin breakdown since last visit.

Factors Effecting Independence

- Communication
- Behavior
- Education
- Bowel Continence
- Private Insurance
- Mobility ??

Ambulation

- Types of ambulation
 - *Community ambulators* – walk and live in community with little to no restriction in walking, may need crutches and/or braces, can navigate ramps/stairs/public transportation
 - *Household ambulators*– walk only indoors on level floors, need assistive device, but can transfer themselves, need wheelchair for some indoor things (school), and for most community activities
 - *Exercise ambulators*– walk only during PT, use a wheelchair for everything else
 - *Non- ambulators* – use a wheelchair exclusively, but may be able to help transfer themselves

Ambulation

Factors affecting the potential for ambulation

- Level of involvement:
 - Mid-Lumbar and below → community or household ambulators
 - High Lumbar and above → exercise and non-ambulators
 - Toddler (2-4y) with 4 or 5? 5 voluntary quad strength can be at least a household ambulator in third decade of life
- Functionally it is a progressively deteriorating disease
 - De-tethering can result in further neurologic impairment
 - Age, size (especially weight / obesity) result in progressive functional impairment
 - Most “peak” at 8-10 years (strength and coordination improved before adolescent growth spurt).

Lower Extremity Issues

- Hip dislocation / dysplasia
- Knee contractures
 - Flexion
 - Extension
- Feet
 - Clubfoot
 - Equinovarus
 - Calcaneus
- Rotational differences
 - In-toeing
 - Out-toeing
- Pressure sores

Infants (0-18 mos)

- Hips
 - No treatment or imaging
- Knees
 - Cast contractures
 - Consider operative treatment (soft tissue release/ reconstruction) of non-responders that have quad strength
- Feet
 - Cast clubfeet and Vertical talus
- Rotational abnormalities
 - Start educating mid-lumbar and below!
- Pressure sores - uncommon unless brace related

JPO 2019

Hip Status and Long-term Functional Outcomes in Spina Bifida

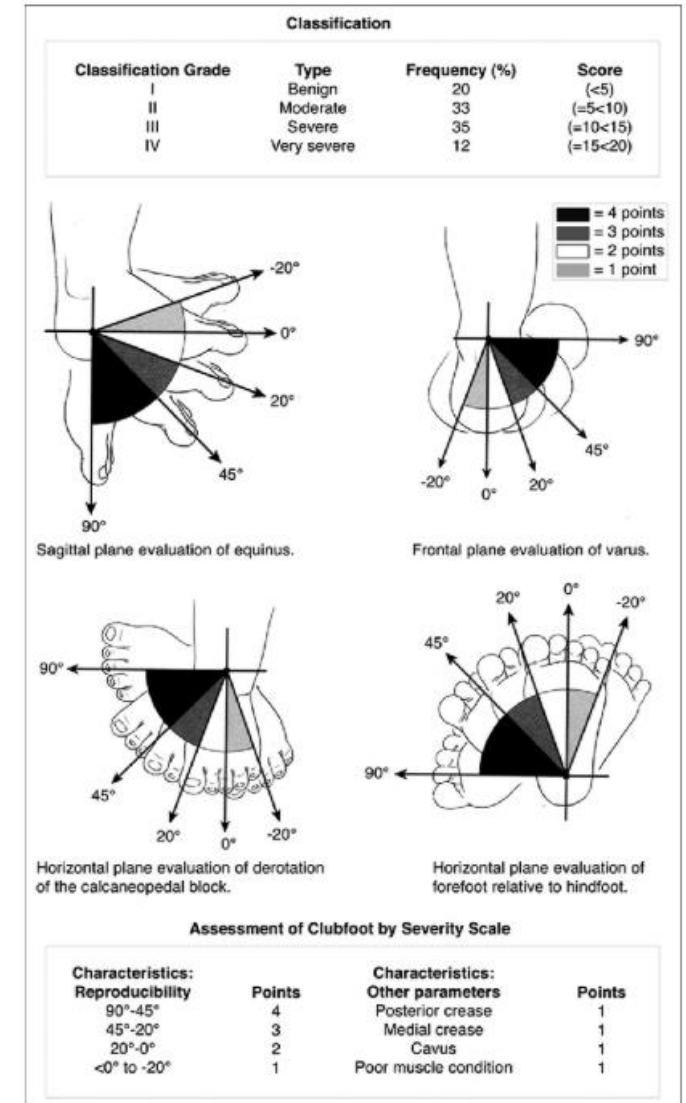
Rachel M. Thompson, MD, Joanna Foley, APN,† Luciano Dias, MD,‡
and Vineeta T. Swaroop, MD†*

- 31 patients from adult myelo clinic, mean 31y
- When level was controlled for, no difference in VR 12 or PROMIS scores
- No association of LLD and decreased quality of life
- **Worse outcomes reflective of neurological level and hip range of motion**
 - Decreased extension and abduction associated with worse VR12 mental scores, no other correlation with PROs
 - PROMIS: bilaterally located better in LE function scores than uni, but bilateral dislocated better than bilateral reduced for pain interference



Dimeglio Score

- 0-20 Pt score
- < 12 can treat when pulling to a stand
- > 12 – Ponseti cast, expect recurrence in those >16



Early Results of the Ponseti Method for the Treatment of Clubfoot Associated with Myelomeningocele

By David J. Gerlach, MD, Christina A. Gurnett, MD, PhD, Noppachart Limpaphayom, MD, Farhang Alaei, MD, Zhongli Zhang, MD, Kristina Porter, RN, BSN, Melissa Kirchhofer, MS, Matthew D. Smyth, MD, and Matthew B. Dobbs, MD

Investigation performed at the Department of Orthopaedic Surgery, Washington University School of Medicine, and St. Louis Shriners Hospital for Children, St. Louis, Missouri

- 39% of clubfeet in myelomeningocele patients were grade IV
 - 11% of idiopathic group ($p=0.014$)
- Initial correction achieved in 96.4% of myelomeningocele clubfeet
 - 100% in idiopathic group (not significant)
- Relapse in 68% of myelo clubfeet
 - 26% of idiopathic group ($p=0.001$)
- Recasting $\pm$ repeat Achilles tenotomy in 79% of relapses for myelo
 - 89% of idiopathic group (not significant)
- Successful treatment without extensive releases in 86%
 - 97% of idiopathic group (not significant)



Midterm Results of the Ponseti Method for Treatment of Clubfoot in Patients With Spina Bifida

Cameron Arkin, BA, Stephanie Ihnow, MD,† Luciano Dias, MD,‡ and Vineeta T. Swaroop, MD**

- 26 feet, 17 patients
- All got initial good correction with Ponseti
 - Did TAL at age 105 days
 - 100% recurrence with perc TAL, 18% with open tendon excision
 - 10/17 patients with skin irritation or breakdown
- 57% had recurrence
 - 10 posterior release, 4 posterior-medial-lateral release, tendon transfer, mean age 1.5 years

Congenital Vertical Talus

- 10% of patients
- Rocker bottom deformity
- Treatment
 - Reverse ponseti → open reduction TN joint/dorsolateral release + posterior release



Talus



Toddlers (2-6y)

- Hips
 - Begin radiographic surveillance of (strong) ambulators
- Knees
 - Maintenance
- Feet
 - Maintenance
- Rotational abnormalities
 - Frustrating time for mid-lumbar
 - Weak rotational control of hip
 - Like non-myelo children with time they get more strength and coordination. Brace options!
- Pressure sores - Educate. Treat when necessary.

Bracing

- In the first 4 yrs of life we can get every child “upright and mobile
- Start when pull to a stand or show an interest in exploring their environment
- Patients without a strong quad will need KAFO, if the hip is weak (usually is in rotation) will need extension to the trunk (HKFO, RGO)
- Very unusual for a patient braced above the knee to be anything but an exercise ambulator (too inefficient)
- Energy needed to walk may impair academic performance (Franks, 1991)



Bracing Rotational deformity

- AFO with straps, cables, lateral uprights
- Pick your poison:
 - Frustration from tripping with internal rotation in AFOs
 - Decreased independence / speed w/ pelvic extension



Latency (6-12)

- Hip dislocation / dysplasia
 - Surgically address progressive radiographic dysplasia in strong ambulators
- Knee contractures
 - Maintenance
- Feet
 - Consider surgical correction in flexible varus or valgus deformities
- Rotational differences
 - Consider surgical correction in severe deformities (supramalleolar osteotomy)
 - Clubfoot with “medial spin”
- Pressure sores - Educate. Treat when necessary.



Casting

- Smooth/even cast padding
- Avoid pressure points
- Extend casts past toes, toes visible
- Watch for sores on dorsum of toes if WB cast
- Plaster against the skin
- Essential to elevate operative extremity post-operatively



Adolescents

- Hip dislocation / dysplasia
 - Surgically address progressive radiographic dysplasia in strong ambulators
- Knee contractures
 - Maintennace
- Feet
 - Wait until near skeletal maturity to surgically address deformities – unless soft tissue wounds force your hand !
- Rotational differences
 - Avoid surgical treatment !
- Pressure sores - Educate. Treat when necessary.

Triple Arthrodesis

- Retrospective chart review (1962-2007)
- 33 feet in 27 patients
- Level
 - Sacral: 10
 - Lower lumbar: 6
 - Middle lumbar: 10
 - Upper lumbar/thoracic: 1
- 23/33 feet had no complications after initial postoperative healing
- 10/33 feet had either minor or major complications during follow-up



**Adults go to the orthopaedist like all adults:
When they have a problem!**

(Usually deformity, wound or pain)

Adult Consequences of Spina Bifida

A Cohort Study

James W. Roach MD, Barbara F. Short RN,
Hanna M. Saltzman

- 221 pts seen at TSRH from 1941 – 1964, 149 contacted for f/u
- “Many of the orthopedic surgeries performed for these children did not improve their function, and in several cases, seemed detrimental”
- “Our study also suggests orthopedic surgery should be very selective and focused on achieving a straight spine with a level pelvis, supple joints, and plantigrade feet”

Pressure Sores

- Loss of protective sensation
- Poor skin perfusion
- Neurogenic bowel and bladder
- Prone to development of pressure sores
 - Deformity / braces
 - Crawling / sitting
 - Iatrogenic (casting)
- Prevention/Treatment
 - **Education**
 - Skin checks / Perineal care

Peripheral Circulation in Patients With Myelodysplasia

Edward C. Sun, MD, Yi-Meng Yen, MD, PhD, Tze Ip, MD, and Norman Y. Otsuka, MD, FACS

- 41 myelo patients w/ age matched controls
- Myelo have lower ABIs, but similar TcO2 with control group (lower but not statistically significant)
- No difference between levels
- Maybe related to elevated homocysteine levels?

Pressure Sore Treatment

- Staged and coordinated care
 - Wound care nurse
 - Orthopaedic surgeon
 - Plastic surgeon
- Dry skin / relieve pressure – inpatient stay sometimes facilitates



Thank You



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