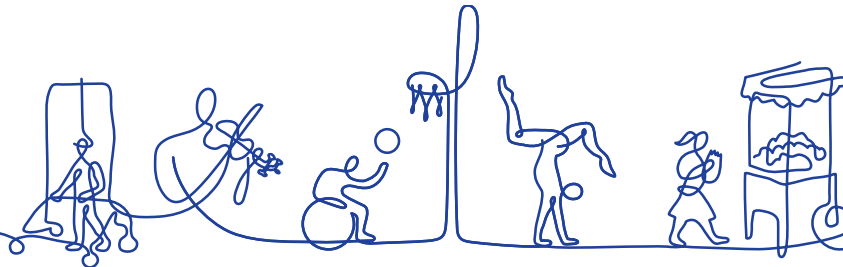


Orthopedic, Holistic and Surgical Approaches to Spine Deformity in Spina Bifida

Karl Rathjen, M.D.



UTSouthwestern
Medical Center



I have no disclosures

When you have the urge to operate on a patient with spina bifida ...

When you have the urge to operate on a patient with spina bifida ...

...lie down until the urge passes







Multidisciplinary Team

Neurosurgeon

Closure
Hydrocephalus

Urological surgeon

Incontinence
Retention
Diverticula
Pyelonephritis
Renal failure

Orthopedic surgeon

Scoliosis
Lordosis
Hip dislocation
Knee contracture
Foot deformities
Paralysis/ambulation

Parents



Social worker

Psychologist

Occupational / Physical Therapist

Orthotist

Neurologist

Pediatrician

Experienced Team

Neurosurgeon

Closure
Hydrocephalus

Urological surgeon

Incontinence
Retention
Diverticula
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Orthopedic surgeon

Scoliosis
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Parents



Social worker

Psychologist

Occupational / Physical Therapist

Orthotist

Neurologist

Pediatrician

Team Captain / Coach / CEO / General Manager



Great clinician



Team builder



Academic



Educator

Colleagues, Residents,
APPS, Nurses, Therapists,
Caregivers

Team Captain / Coach / CEO / General Manager





Important Facts to Remember

- Complex chronic medical condition effecting multiple organ systems
 - (Think brittle type one diabetic)
- Condition we “manage”, not “cure”
- Goal is to improve the function, limit “patient time”, maximize “child time”
 - Simple math: “2 vs 4”

Spine Deformities

- Scoliosis
 - Neuromuscular (long sweeping curve, pelvic obliquity, almost always non-ambulators)
 - “Idiopathic like” – ambulators, only thoracic
 - May have abnormally formed vertebrae (congenital scoliosis)
- Kyphosis – often a congenital component
- Lordosis – usually due to hip flexion contracture



Treatment Options

- Observation
 - Seating modifications (“Easier to change the environment than the child”)
- Bracing
 - Probably does not stop progression
 - Pressure Sores
 - ? “Buy time”
- Surgery



Who gets spine deformity?

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CLINICAL AND RADIOGRAPHIC PREDICTORS OF SCOLIOSIS IN PATIENTS WITH MYELOMENINGOCELE

BY JAYESH TRIVEDI, FRCS(ORTH), MCh(ORTH), JEFFREY D. THOMSON, MD,
COMMANDER JOSEPH B. SLAKEY, MC, USNR, JOHN V. BANTA, MD, AND PETER W. JONES, MSC, PHD, CSTAT

Investigation performed at the Connecticut Children's Medical Center, Hartford, Connecticut

- The higher the last intact laminar arch, higher risk of scoliosis
 - 93% of patients with thoracic last intact laminar arch have scoliosis
 - 44% L1-L3
 - 11% L4-5
 - 0% sacral

Natural History?

Quality of life and functional disability in skeletally mature patients with myelomeningocele-related spinal deformity

Marcin Sibinski^a, Marek Synder^a, Zoe C.J. Higgs^c, Jolanta Kujawa^b and Andrzej Grzegorzewski^a

- JPO B 2013
- 19 Adults, scoliosis > 20, No surgery
- Increasing cobb angle → lower QOL in Quality of Life in Spina Bifida Questionnaire
- “More severe scoliosis affects quality of life and is related to the degree of pelvic obliquity and age of the patients”
- No impact on physical function, self perception or self motivation

Natural History?

Journal of Pediatric Rehabilitation Medicine: An Interdisciplinary Approach 11 (2018) 243–254
DOI 10.3233/PRM-179451
IOS Press

Impact of neurological level and spinal curvature on pulmonary function in adults with spina bifida

Theresa M. Crytzer^{a,b,c,*}, Yu-Ting Cheng^b, Mary Jo Bryner^d, Robert Wilson III^d,
Frank C. Sciurba^{d,1} and Brad E. Dicianno^{a,b,c,e,f,1}

- 29 patients mean age 30
- 90% of thoracic level patients have restrictive pulmonary disease
 - Weakness of abdominal muscles and other accessory muscles for respiration
- Scoliosis independently negatively influenced lung function
- Interestingly severe is over 45 degrees... no mean or range

Surgical Outcomes?

Clin Orthop Relat Res (2022) 480:773-787
DOI 10.1097/CORR.0000000000002087

Clinical Orthopaedics
and Related Research®
A Publication of The Association of Bone and Joint Surgeons

Systematic Review and Meta-analysis

What is the Role of Scoliosis Surgery in Adolescents and Adults with Myelomeningocele? A Systematic Review

Viachaslau Bradko MD¹ , Heidi Castillo MD¹, Ellen Fremion MD^{2,3}, Michael Conklin MD⁴,
Benny Dahl MD, PhD^{5,6}, Jonathan Castillo MD, MPH¹ 

- Lit search for patients older than 15yo
- Overall complication rate for scoliosis surgery: 40-58%

- No clear functional or quality of life benefits following scoli surgery
- . Although studies regarding scoliosis surgery reported success in correcting coronal deformity and stopping curve progression, they found no clear benefits of treatment in terms of HRQoL
- Scoliosis itself seems to be associated with secondary impairments like incontinence, limited ambulation, pressure sores, especially in patients with hydrocephalus and higher level lesions
- “The level of neurologic dysfunction, hydrocephalus, and brainstem dysfunction are greater determinants of quality of life.”

Surgical Outcomes?

■ **SPINE** 2014

The long-term outcome of patients treated operatively and non-operatively for scoliosis deformity secondary to spina bifida



A. Khoshbin, L. Vivas, P. W. Law, D. Stephens, A. M. Davis, A. Howard, J. G. Jarvis, J. G. Wright

- 45 patients with scoliosis >50 degrees
- 75% treated operatively, mean age at follow up 27 op and 25 non-op
- Those who had surgery had bigger curve magnitude on average (88 vs 65)
- Op and non-op patients had similar pelvic obliquity in the end
- No difference in health related QOL for op vs nonop patients
- 32% infection, 17% pseudo rate
- A total of five patients who underwent fusion could walk at 'baseline' but were unable to at follow-up
 - Advancing age also linked to decrease in community ambulation... what's the real cause?

Surgical Outcomes?

JPO 2005

The Relationship Between Function, Self-perception, and Spinal Deformity

Implications for Treatment of Scoliosis in Children With Spina Bifida

Eugene K. Wai, MD, MSc,† Nancy L. Young, BScPT, PhD,†‡§¶
Brian M. Feldman, MD, MSc, FRCPC,†‡§¶ Elizabeth M. Badley, BSc, PhD,†
and James G. Wright, MD, MPH, FRCSC*†¶*

Of all aspects of spinal deformity, only coronal imbalance was significantly related to only one aspect of physical function (ie, sitting imbalance). No aspect of spinal deformity was related to self-perception. In conclusion, surgeons should be clear in their indications for surgery and recognize that **in the short term the potential benefit of surgery may be, at best, to improve only sitting balance.**

Surgical Indications

- Intractable skin problems
- Sitting imbalance
- Concern it will be problematic as an adult
- Seating problems
- Cosmesis
- Pain – Caution !!



Pre-operative Assessment

- Nutrition
- Shunt
- Pulmonary and Renal Function
- Peer and Parent support
- Psychology
- Plastic Surgery?
- Halo gravity traction?



Post-operative needs

- Pulmonary Toilet
- Nutrition
- Seating assessment

High rates of Peri-operative complications

- Multifactorial
 - Poor nutrition
 - Absent protective pain perception
 - Poor tissue perfusion
 - Poor skin overlying involved area of spine
 - Neurogenic bowel and bladder

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?

“Three Hurdles” to Surgical Success

- Blood loss
- ICU Stay / Pulmonary Function
- Wound Healing
- Seating mapping / reassess ADLs



Scoliosis – Operative Management

- Goal – Head and shoulders balanced over pelvis
- Anterior release / fusion and PSF to pelvis
 - Decreased risk for pseudarthrosis
 - Improved correction
- Posterior only fusion
 - 40 year review showed lower infection rate w posterior only
- Halo- gravity traction ?



Scoliosis – Operative Management

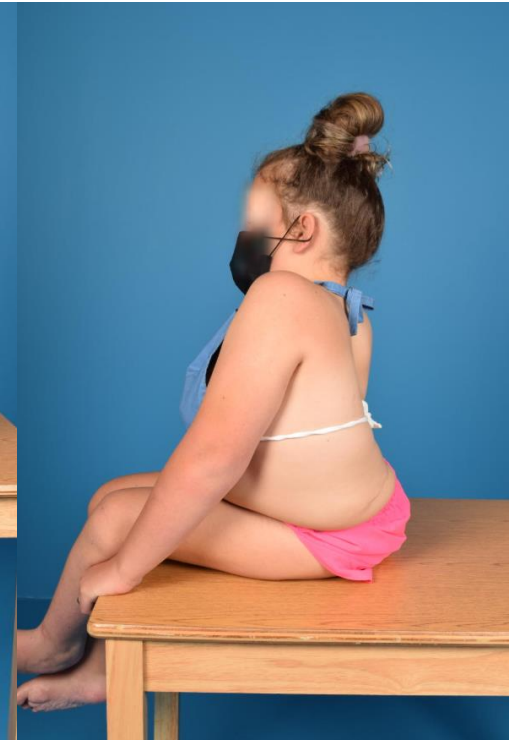
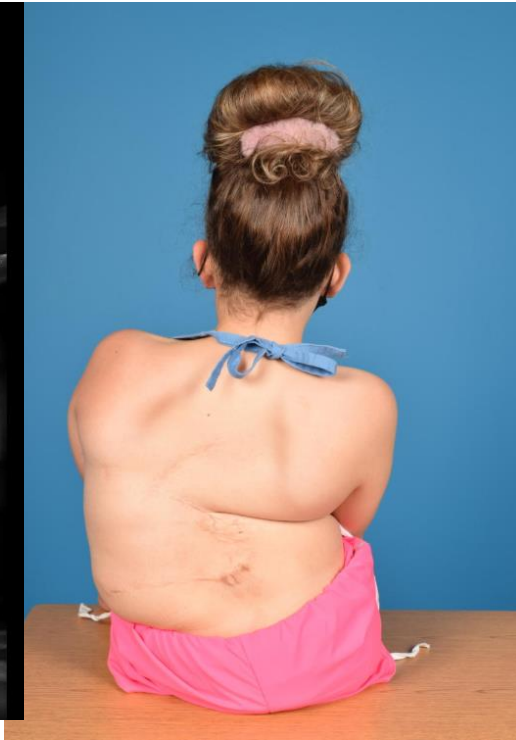
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 - 40 year review showed lower infection rate w posterior only
- Halo- gravity traction ?
- Tissue expanders
- Model ?



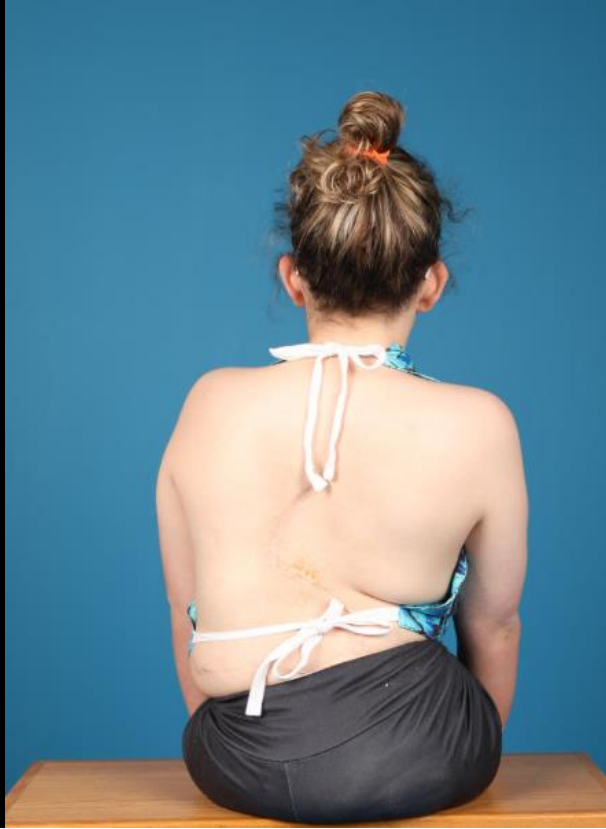
Scoliosis – Operative Management

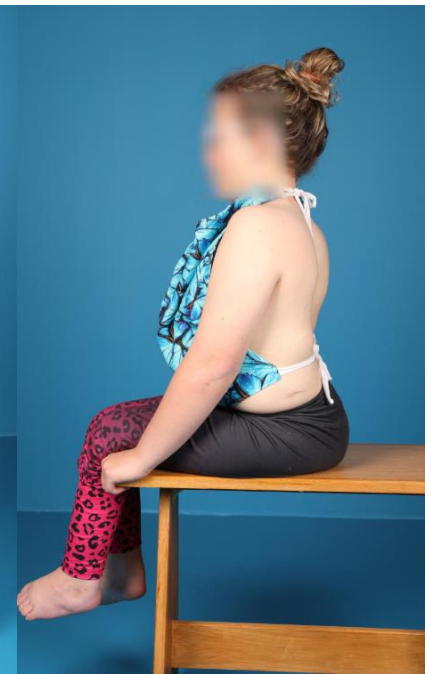
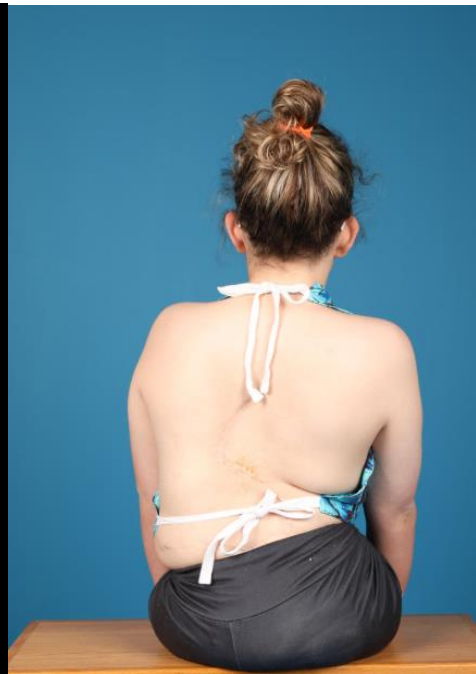
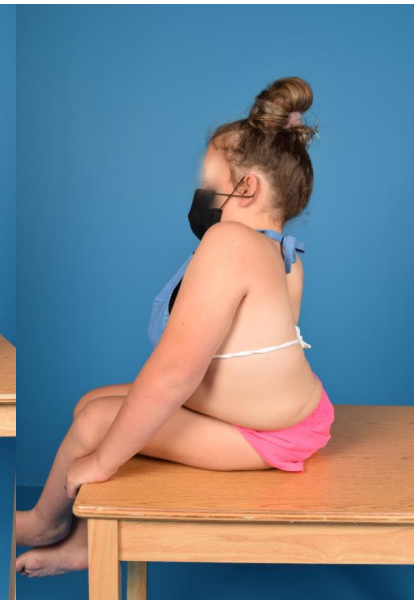
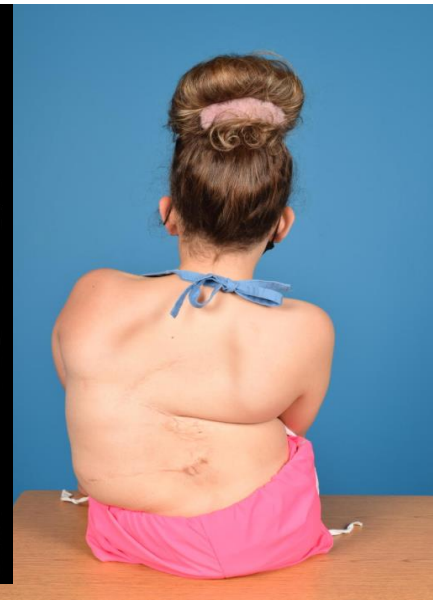
- Pre-op Considerations
 - Difficult catheterization: Admit night before
- Post-operative considerations
 - Incisional wound vac
 - Drain

MP, 11yo, 2010

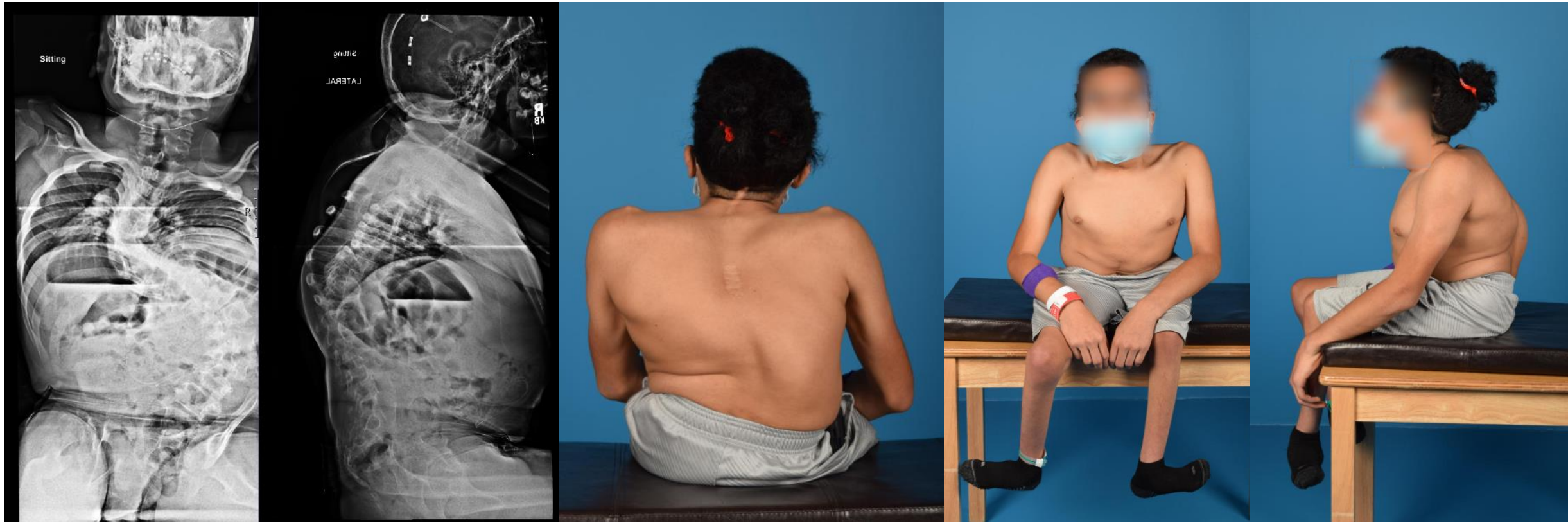








LM, 13yo, 2022

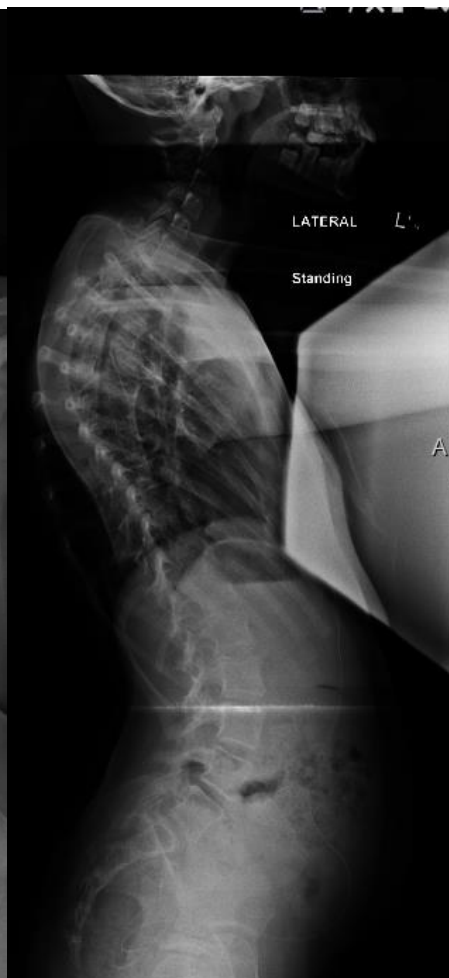
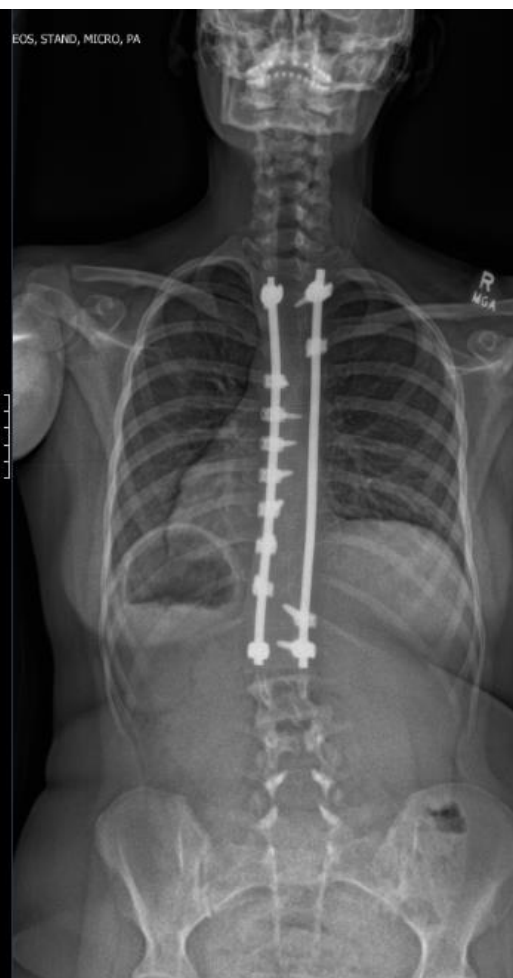






HW, 10yo, 2007





Kyphosis

- Less common than scoli (10% of myelo pts)
- More common in thoracic level
- Sharp kyphotic deformity = **gibbus**
 - Can cause skin breakdown
- Wheelchair modifications important!

Gibbus →



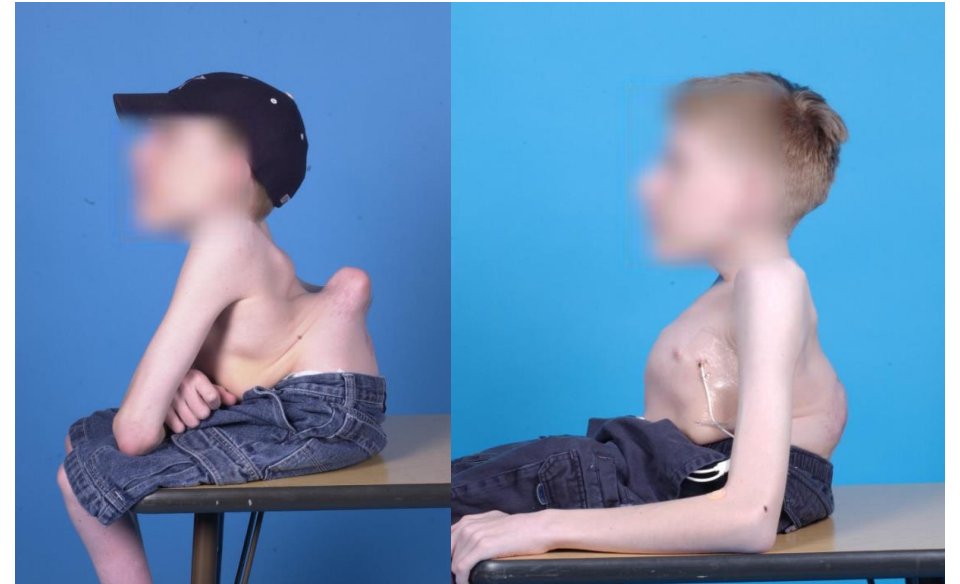
Surgical Treatment

- Kyphectomy - Gibbus resection and posterior fusion - all done “from the back”
- Historically, controversial due to high blood loss / complication / mortality

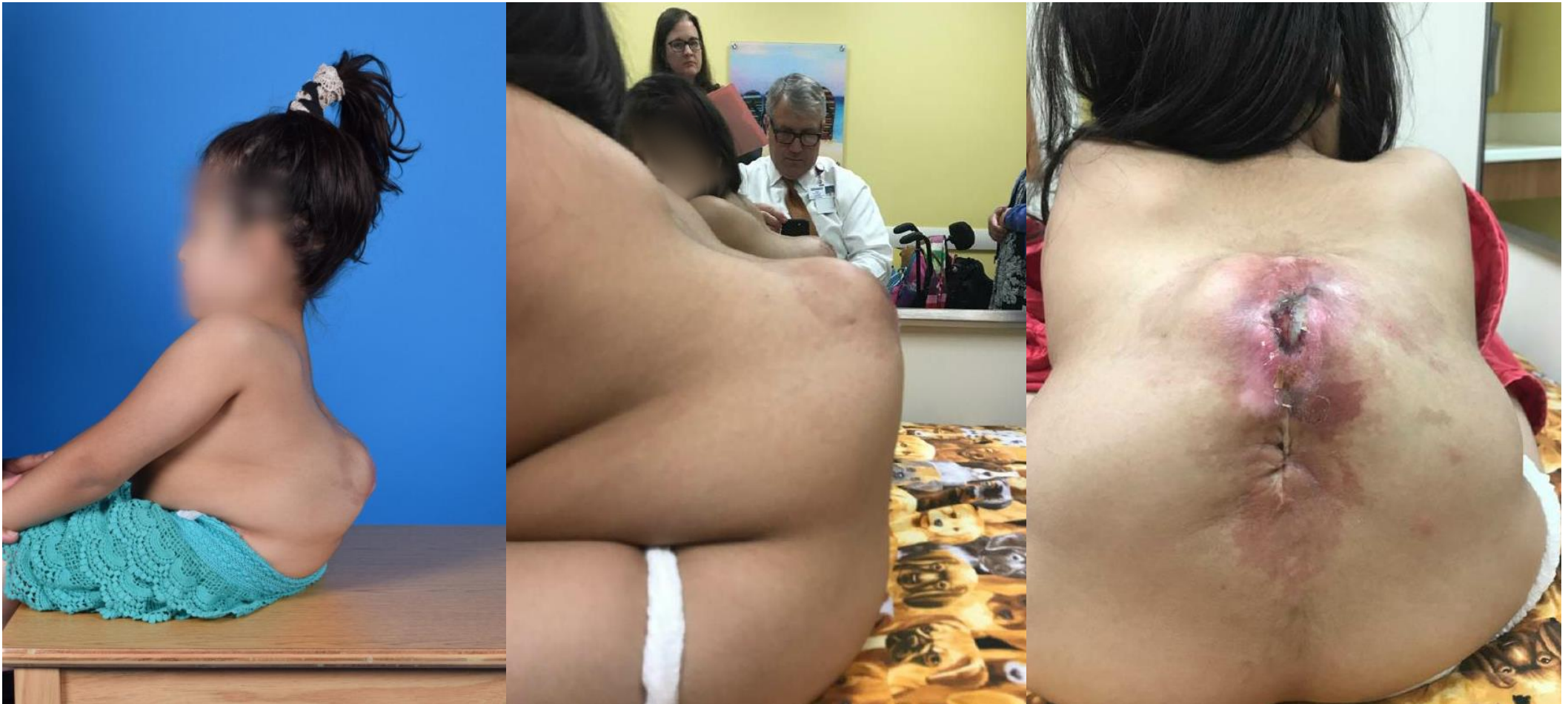


Surgical Indications

- Intractable skin problems
- Sitting imbalance
- Concern it will be problematic as an adult
- Seating problems



HR, 2014/2019 (11yo)









Kyphosis – operative management

Clin Orthop Relat Res (2011) 469:1279–1285
DOI 10.1007/s11999-010-1650-8

SYMPOSIUM: MYELOMENINGOCELE

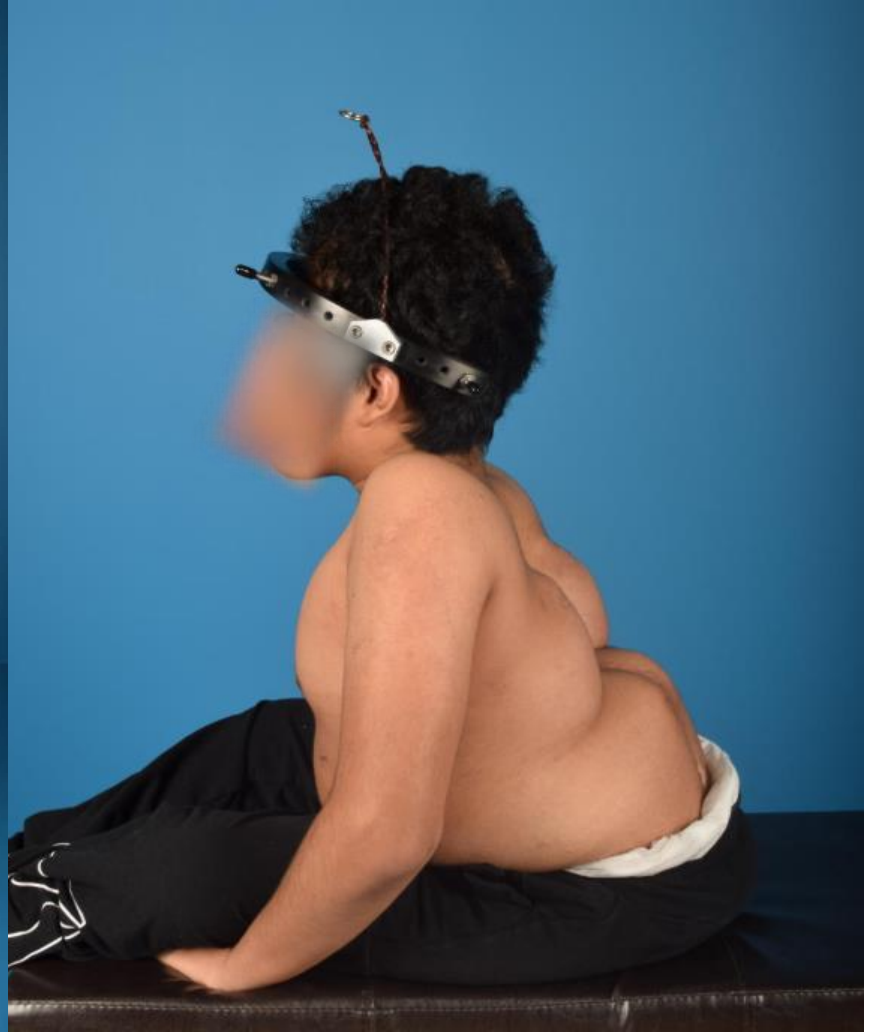
Kyphectomy Improves Sitting and Skin Problems in Patients with Myelomeningocele

Sumeet Garg MD, Matthew Oetgen MD,
Karl Rathjen MD, B. Stephens Richards MD

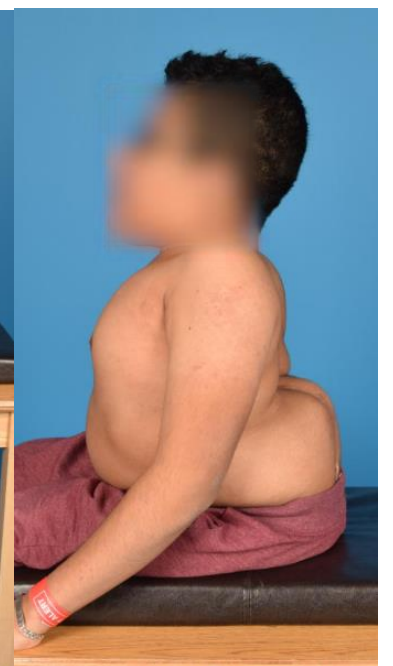
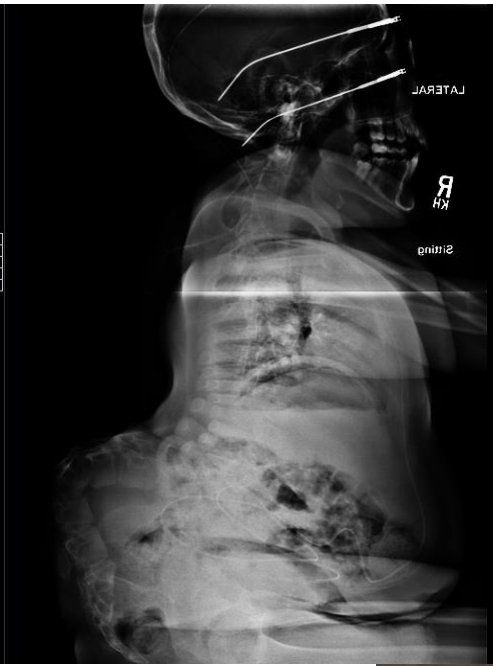
- Resolution of skin problems, improved seating balance in 17/18 pts
- 13/18 w at least one complication
- 7/18 pts w complications requiring re-operation

EE, 12, 2022









17 yo w progressive lordosis after PSF 10 yrs prior, 2009

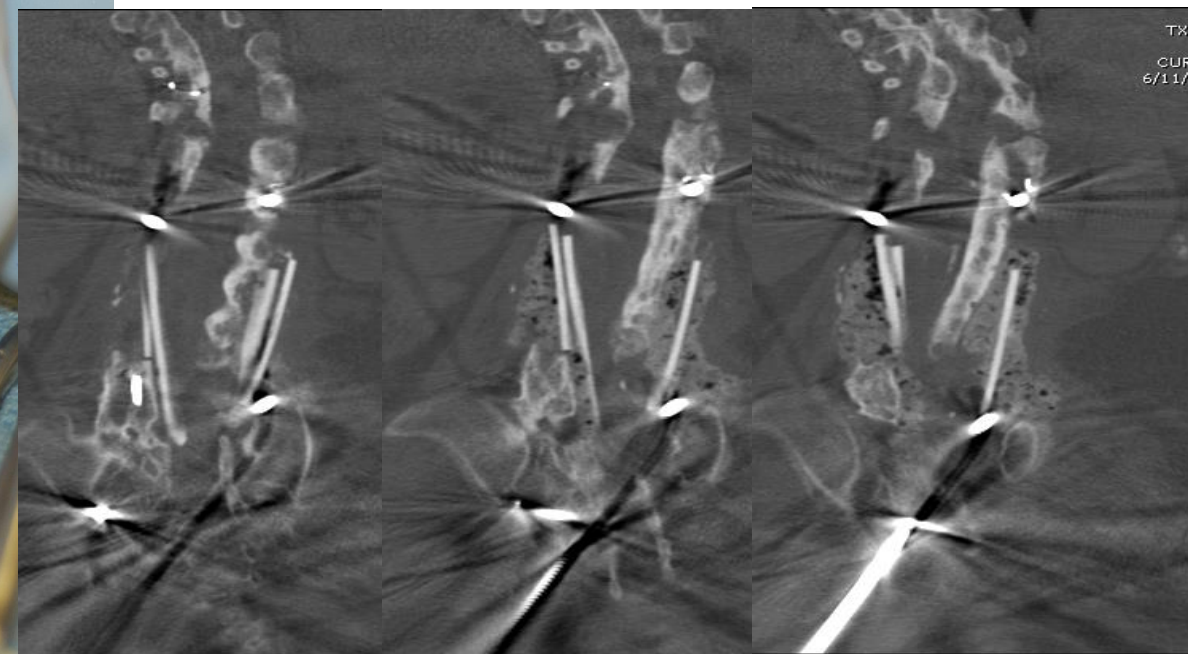
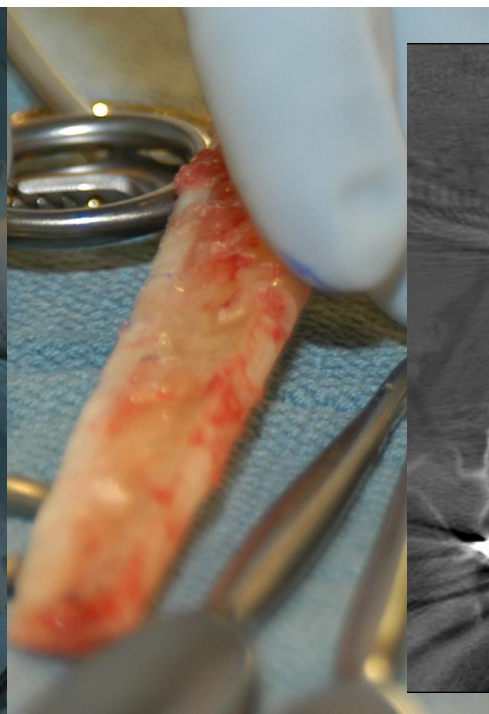


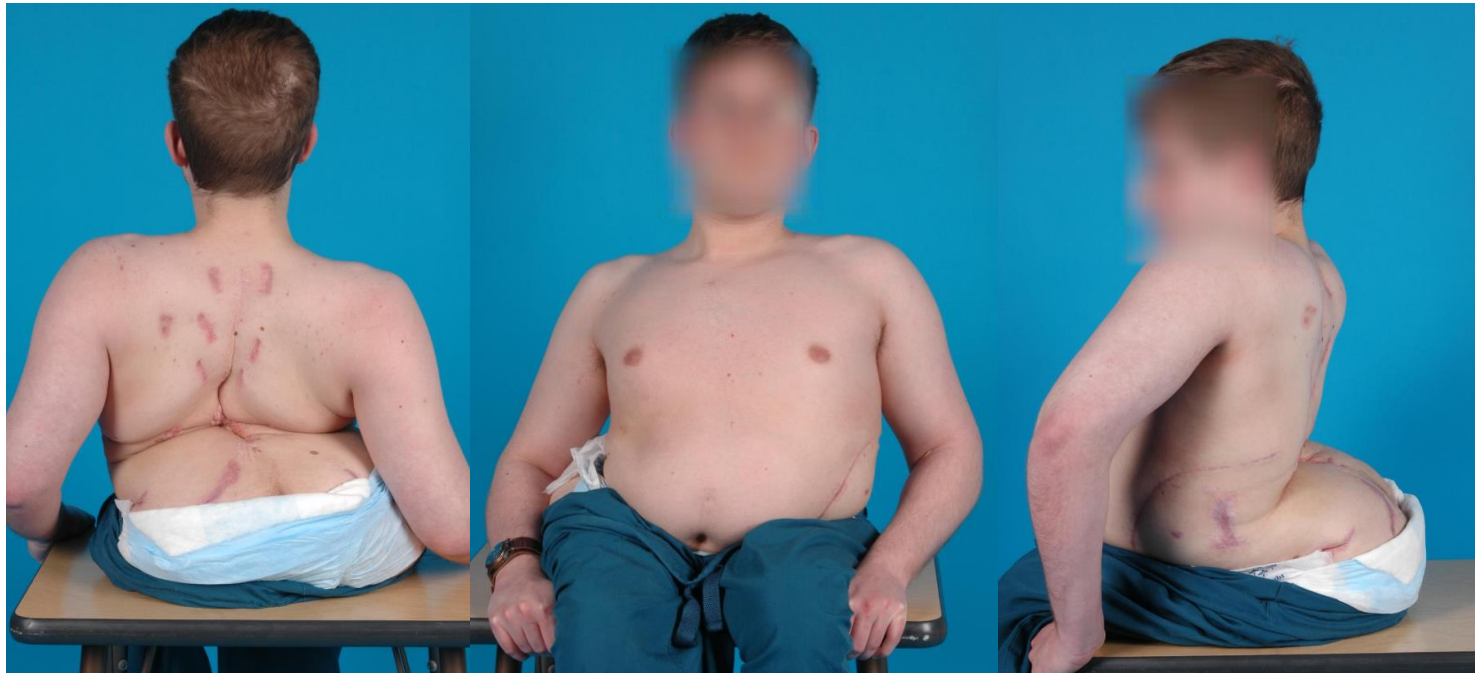


L5 Vertebrectomy, Spinopelvic Ilizarov frame









What's on the Horizon ?



Medical / Surgical Changes

- Improved wound / nutrition management
- “Improved” Research:
 - Functional outcome studies
 - Better natural history studies
 - Multicenter studies

Changes in “Priorities”

- Interventions aimed at improving global function (Independence)
- Emphasis on physical fitness
- Improved care (access) for adults w/ spina bifida

Thank You



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