

Myelo/Lipomeningocele and Caudal Regression

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Myelomeningocele

- Etiology
- Pathology
- Natural History
- Exam
- Treatment

Etiology of MM

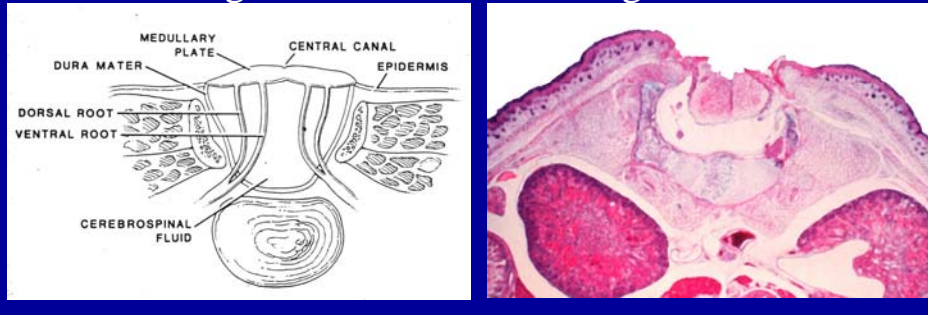
- Not known for certain - 2 possibilities:
 - 1: 1° disturbance of neural tube. normally starts in thoracic spine; progresses cranially and caudally
 - currently favored theory
 - explains brain pathology
 - 2: rupture of formed neural tube 2° ↓ CSF resorption

Etiology/epidemiology

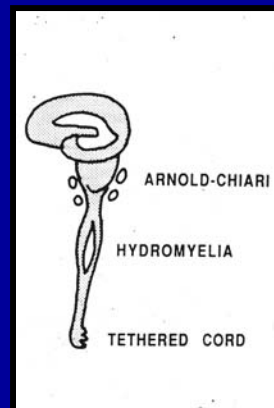
- Multifactorial inheritance
 - 3% risk in 1° family member
- Abnormal folate metabolism
 - periconceptual folate 0.4 mg
- Incidence declining
 - greater in certain countries
- Prenatal Dx:
 - alpha fp; ultrasound

Pathology - myelomeningocele

- Open neural plaque instead of closed cord
- Usually corresponds to bony, sensory level
- Posterior elements folded out
- Missing/denervated muscle, ligaments, skin



- Neurologic damage variable, asymmetrical
- Chiari II
- hydrocephalus
- Possible syrinx



Pathology - Lipomeningocele

- Large lipoma in neural elements
- Usually a distal problem (L4-S1)
- Hydrocephalus rare; IQ = OK
- Skin intact
- Almost all community ambulators
- May be associated with Exstrophy/caudal regression

Ethics

- Lorber's proposed contraindications to sac closure:
 - High lesion
 - Congenital hydrocephalus
 - Skull defects
 - Congenital kyphosis

– Didn't work in practice
- Now practice is to treat all aggressively

Natural History

- Untreated: 90% dead by 2 yrs:
 - hydrocephalus
 - meningitis
 - renal problems, later
- Treated: 95% survive to adulthood
 - 30% have IQ > 100
 - Decreased abstract reasoning, independence
 - May lose function due to tether, syrxinx
 - Pressure sores a major problem

Exam

- Baseline x-ray of whole spine, pelvis
- Observe posture for clues to level
- Stimulate above lesion for neuro response
 - Otherwise reflex activity may confuse
- Check back for gibbus, scoli, Sprengel
- Note ROM of all joints

Motor Levels

- Hip flexors: L1-3
 - adductors: L2-4
 - abductors: L4-S1
 - extensors: L5-S1
- Knee flexors: L4-S2
 - extensors: L2-4
- Ankle DF: L4-5
 - PF: S1





Rancho Functional Classes

- Dependent sitter
- Independent sitter
- PT walker
- Household walker
- Community walker

Goals

- Thoracic: balanced sitting
- Upper Lumbar: household walker in LLB
- Lower Lumbar: community walker with crutches/walker, AFOs
- Sacral: brace-free community ambulator

General Treatment Plan

- Close spine at birth
- Shunt 0-4 months
- Foot cast/surgery in first year
- Foot-knee-hip surg. prn to stand by 18 mos
- Avoid or at least postpone bony fusions
- ± Bladder augmentation later

Braced Standing-High Level MM

- Advantages
 - improved independence, socialization
 - ?bone density
- Disadvantages
 - effort
 - cost
 - may require surgical releases

Determinants of Function

- Motor level: strength, spasticity
- Coordination (Chiari)
- Obesity
- Motivation/family
- Contractures
- Not just walking

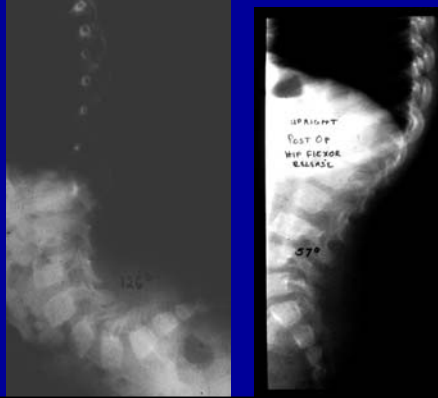
Bracing in MM

- Stepwise progression
- Parapodium: total support for early standing
- Reciprocating Gait Orthosis for high level
- HKAFO for Mid level
- AFO or FR-AFO



Hip Deformity in MM

- Flexion contracture in high level: correct if >45 in walker



Hip Deformity in MM

- Dislocation: greatest in mid-level
 - Does not impair function in childhood
 - Treat only if good walker
 - Otherwise risk of stiffness a bigger problem

Treatment of Hip Dislocation

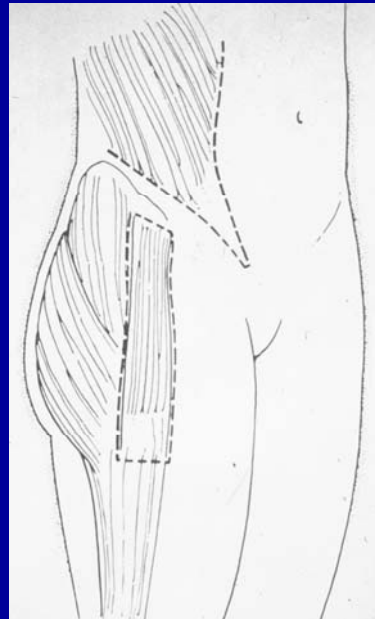
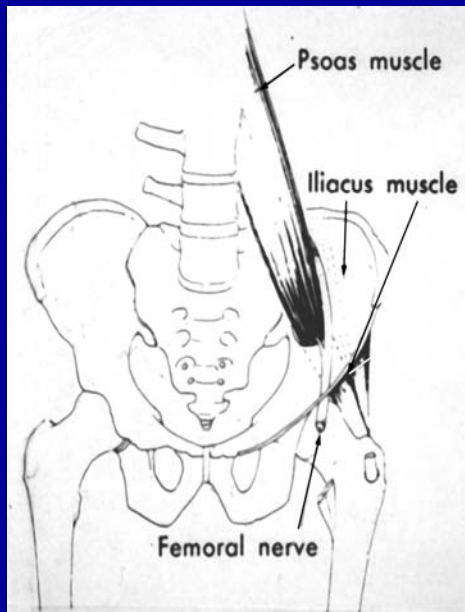
- Bracing in infancy?
 - Controversial
- Operative treatment
 - Less often indicated than in CP
 - Because results are less predictable
 - Hips can become stiff
 - Redislocation –due to tether
 - Dislocations less often painful
 - Dislocations do not impair function

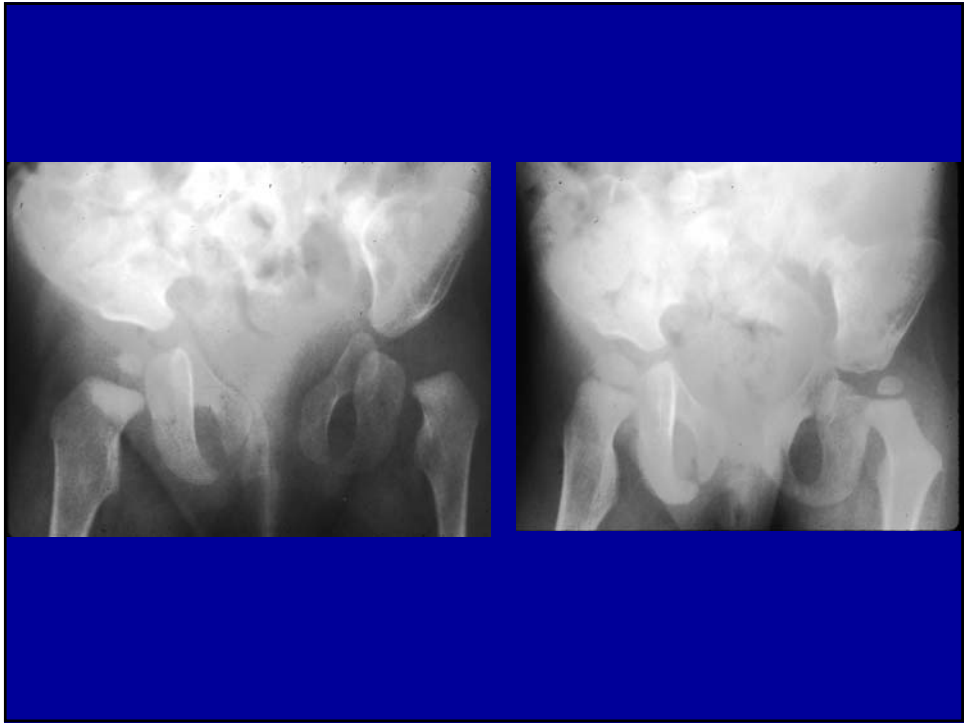
Indications for Operative Rx of Hip Dysplasia

- L5 level or lower
 - Able to walk without aids
- Occasional L4
- Best if hip is subluxating, not dislocated from birth

Operative Treatment of Hip Dysplasia

- Open reduction capsulorrhaphy, plus
 - muscle balancing (releases, transfers)
 - psoas (Sharrard)
 - ext oblique
 - adductors to ischium
 - femoral and iliac osteotomy



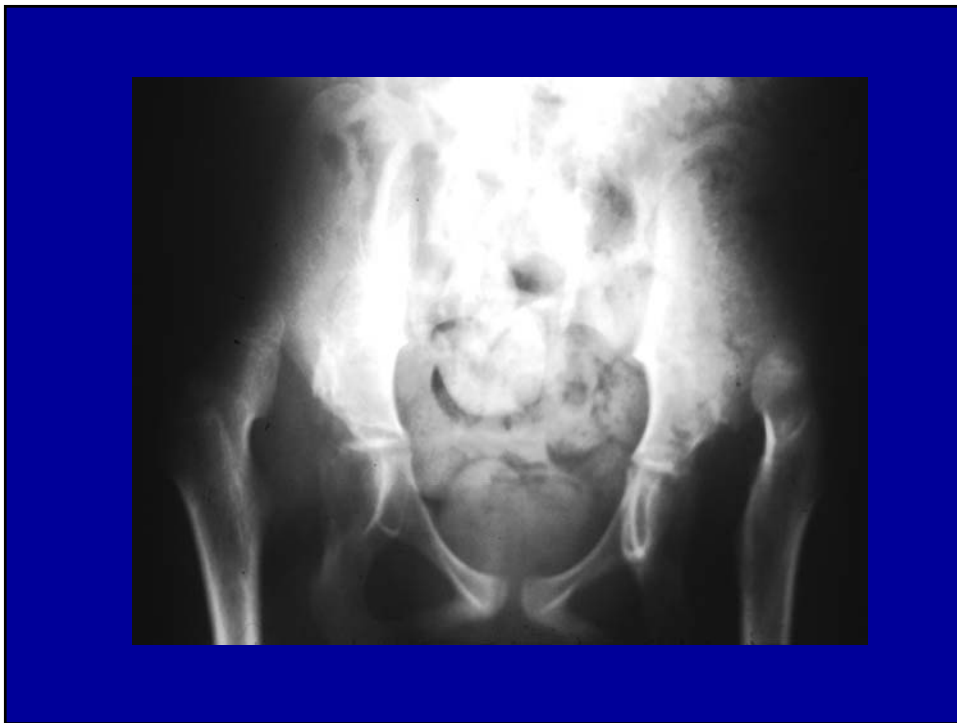


Knees

- Radical release or osteotomy for flexion contracture $> 45^\circ$ in walker
- Quad stretching, lengthening for contracture

Clubfeet

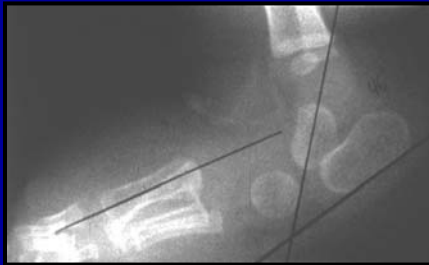
- Should be treated for shoe wear, self-image
- Ponseti cast successful in 30%
- Radical release
- Osteotomies/decancellation prn

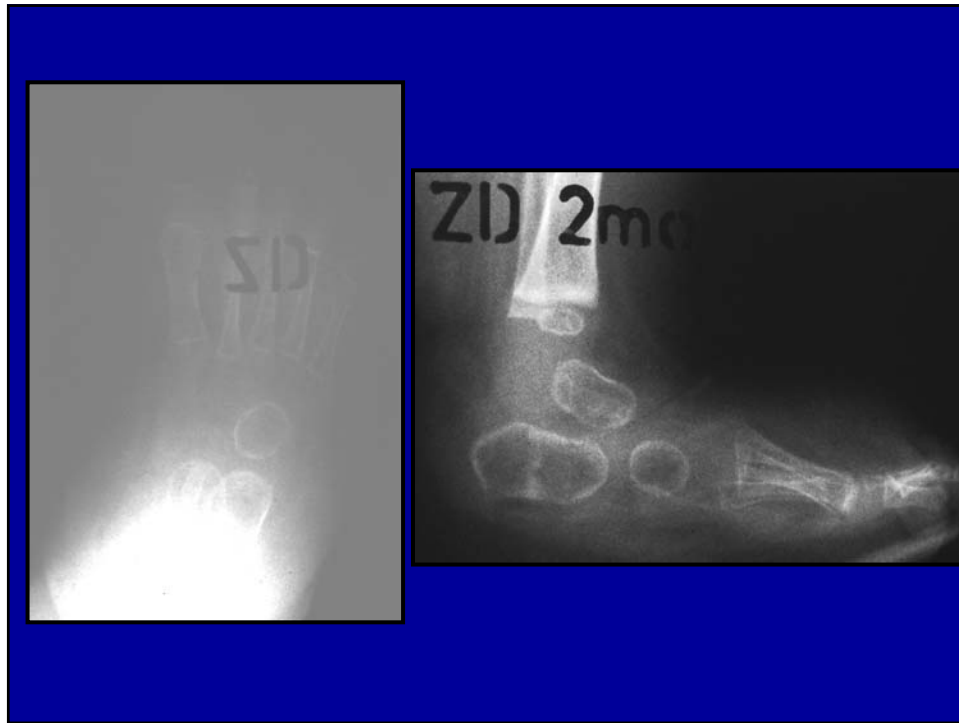




Other Foot Deformities

- Vertical talus
 - Treat if walking potential





...other MM foot deformities

- Calcaneus foot
 - Ant Tib transfer at 5 if walker
- Valgus foot
 - if mild, brace
 - callouses/sores in brace
 - plafond often tilted
 - epiphyseodesis, osteotomy



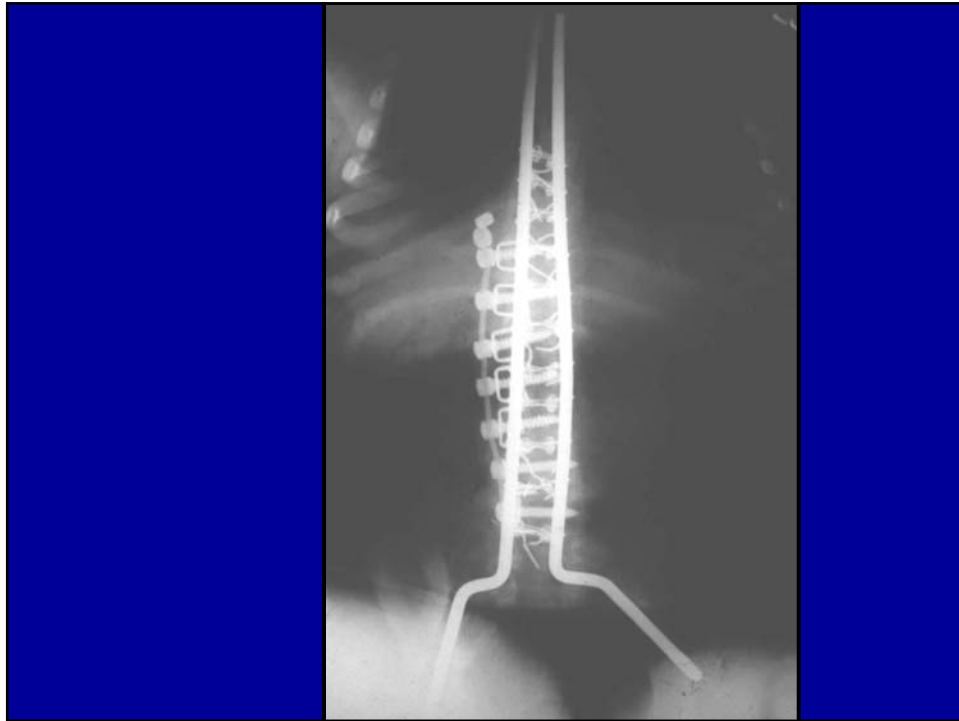
Rotational Deformities

- Due to subtle muscle imbalance
- Treatment
 - accept if minor
 - twister cables if young
 - osteotomies if needed
 - minor role of muscle transfers

Scoliosis

- Most in high level lesions
- Rule out tether if early, severe, preop
- Brace works better than in CP
- Surgery if functionally significant
- Beware long fusions
- High infection rate-- A+P recommended





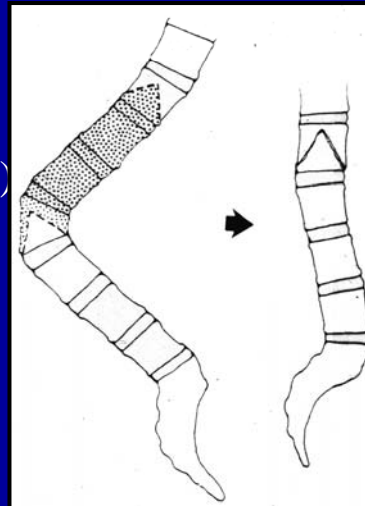
Kyphosis

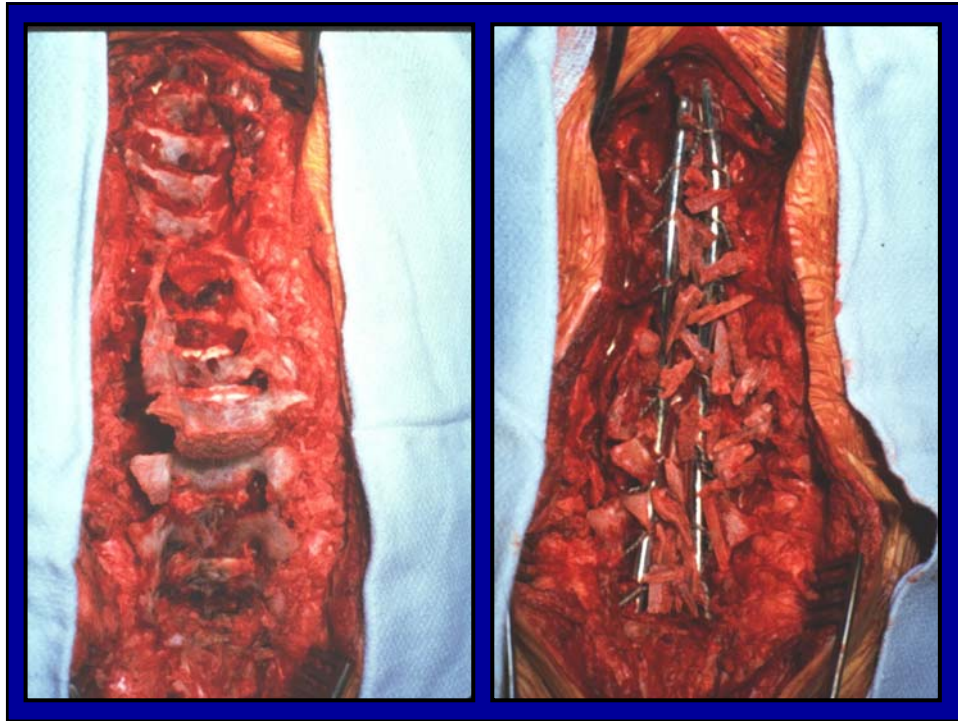
- Congenital gibbus
 - high level lesion
 - present at birth but worsens with time
 - Kyphectomy if desire improvement in pressure sores, posture
 - May have value in newborn closure
- Acquired kyphosis: due to extensor weakness. Usually no treatment



Kyphectomy

- Posterior approach only
- Resect upper horizontal limb
- Cantilever straight (McCarthy)
- Localized fusion
 - Allow longitudinal growth





Tethered cord

- Due to adhesions around defect & closure
- Appears with growth; \pm worse with activity
- Signs & Sx:
 - back pain
 - leg pain
 - new weakness
 - unexplained scoliosis

- Radiographic signs nonspecific; clinical Dx
- Surgery improves Sx, at least short-term



Latex allergy

- Occurs in 20-50%
- due to frequent challenges, esp. to mucous membranes
 - multiple daily catheterizations
 - multiple operations
- Now avoid in all MM

Fractures

- More common due to
 - osteopenia
 - surgery/cast
 - lack of sensation, protection
- Most common cause of warm, swollen limb with mild fever
- Often extensive periosteal reaction develops
- Prevention



Pressure Sores

- Insensate skin + bony prominences + stiffness
- Enforced postop bedrest may trigger



Lumbosacral Agenesis

- Absence of Sacrum \pm lumbar vertebrae
 - Many variations, levels
- Often Maternal diabetes (1% risk)
- Motor level & contractures correspond to last intact vertebral level
- Notochord induces vertebral and motor development





- Sensation usually preserved
 - Neural crest cells migrate later
- Renal, other anomalies may coexist
- Severe contractures, especially w. high level
 - webbing of hip, behind knee
 - neurovascular bundle under skin

Treatment of LS Agenesis

- Individualize
- Less treatment for more proximal defects
- Osteotomies \pm releases & bracing for mild contractures
- Disarticulation and bracing sometimes recommended for severe contractures
- Spine lengthening/stabilization
 - no proven functional benefit

References

- Guille JT: Lumbosacral Agenesis: A new Classification. JBJS 84A 32-37, 2002.
- Phillips WA et al: Orthopaedic Management of Lumbosacral agenesis. JBJS 1982: 1282-94
- Sarwark JF, Lubicky JP, eds: Caring for the Child with Spina Bifida. Rosemont IL: AAOS, 2002

- ...Thank You