Spina Bifida: The Indications for Surgery

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Spina Bifida

A spectrum of conditions involving malformation of the vertebrae, nerve roots or spinal cord.
Spina Bifida

- Myelomeningocele is the commonest form
- Often complicated by:
  - Hydrocephalus
  - Hydromyelia
Presentation

• Myelomeningocele
  – CNS complications
  – Deformities
    • Hip dislocation
    • Knee contractures
    • Club feet

• Childhood motor problems / deformity
  – Tethered cord syndrome
Contractures

• Intra-uterine positioning of flaccid joints
  – conservative Rx

• Muscle paralysis / imbalance
  – poor response to conservative Rx
  – mixture of upper and lower motor neurone lesion
Neurosurgery

• Primary closure - selection
• Later in life - to *halt* deterioration
  – Tethered cord
  – Diastematomyelia (bony spur)
Orthopaedic Surgery

• To correct muscle imbalance
• To correct deformity
• Procedures derive from polio surgery
• Rarely operate in first year
Assessment

• Is walking a realistic aim?
• Key is L4 function; quadriceps
• Walking is feasible if:
  – Functioning quads and medial hamstrings
  – Good sitting posture and balance
  – Good upper limbs
Features to Assess

- Muscle and sensation charts
- Sitting balance
- Upper limbs
- Spinal curvatures
- Hip ROM and stability
- Knees
- Tibiae; rotational deformity
- Ankles
- Feet
Grading

- Three grades
- By upper limit of the lesion; not clear-cut
- Determines which muscles are working and which are unopposed
- Prognostic value; chances of walking
- Risk to hips
Grade 1

- Lesion up to T12
- Flaccid paraplegia
- No deforming forces on hip joints
- Upper limbs and trunk usually affected
- Chairbound
Grade 2

- Up to L4
- Deficient hip abductors and extensors
- Hip dislocation likely *but*
- Variable L4 (quadriceps) preserved
- Prospects for walking good
- AFO and crutches
Grade 3

- Below L5
- Paralysis below knee
- Hip is safe
- Foot and ankle problems; just AFO or callipers
Aims of Treatment

• Walking *if* realistic
  – ? reduce dislocated hips

• Comfortable, stable posture
  – Walking (+/- orthoses)
  – Sitting

• Prevent pressure points - trophic ulcers
  – Plantigrade feet
Principles of Treatment

• Start conservative treatment immediately
• Delay surgery until 1 - 4 years of age
• Balance muscles and correct deformity
• Individualise the surgery
• Multiple procedures at one session
• Avoid NWB
Indications for surgery by region

- Hip
- Knee
- Ankle
- Foot
Hip - features

- Contracture +/- dislocation
- Flexion / adduction deformity
- Risk of dislocation greatest at L3-4
- +/- pelvic obliquity
- Reduction is *not obligatory*; ? just address the contracture
Hip subluxation & dislocation

- With >50% of myelomeningoceles
- Distinguish from a typical DDH
- High lesions give “teratologic” dislocation
- Paralytic common at L3-L4 level
- Difficult to prove benefit of reduction
Hip - treatment

• Early stretching and splintage
• Consider surgery at 24-30 months
• To consider;
  – Unilateral or bilateral dislocation
  – Quadriceps function
  – Response of flexion contracture to stretch
  – Upper body function (crutches)
Hip- surgery

- Weak contracture:
- Strong contracture:
- Bilateral dislocation - correct contracture *if* good arms and trunk:
- Unilateral dislocation in potential walker:
- iliopsoas tenotomy
- anterior hip release
- ? varus derotation osteotomy
- Open reduction, release and balance
Hip- balancing

- Sharrard Procedure + adductor tenotomy
  - Strong quads, hip flexors and adductors
  - Weak abductors
  - Pure LMN lesion; no spasticity
- Transfer external oblique to greater trochanter
Sharrard Procedure
Hip- containing the head

• If >20-30 degrees for concentricity,
  – Varus femoral osteotomy
• If acetabular dysplasia is severe,
  – Chiari osteotomy
Pelvic Obliquity

- Hip subluxation, ischial ulcers
- Try splinting / positioning hip
- If unsuccessful;
  - Tenotomy to release hip contracture
  - Then correct scoliosis
  - If >20 degrees obliquity remains, iliac osteotomies (up to 40 degrees achievable)
Knee:
Grade 1; Flexion contracture (common)

- Usually stretching and splintage
- Aim for KAFO if there is good function above waist
- Accept up to 20 degrees
Knee:
Surgery for flexion contracture

- Hamstring tenotomy
- Posterior capsulotomy
- Supracondylar femoral osteotomy
- Only near skeletal maturity
- Also for valgus deformity
Knee:

Grade 2; Extension contracture (rare)

- Postural fixed flexion at birth
- Frequent stretching
- Gives a stable extended knee
- Recurvatum usually iatrogenic
- Rarely, serial casts or extensor release
Knee:
Grade 3; Delayed flexion contracture (uncommon)

- After 10 yoa
- Resistant to orthoses
- Lengthen flexors (rather than tenotomy)
Ankle Deformity

- Commonly valgus; pressure ulcers
- Difficult to control in orthosis
- Pathology:
  - Fibular shortening
  - Valgus distal tibia
  - Valgus talar tilt
  - Lateral shift of os calcis
Ankle
Treatment

• Surgery:
  – Achilles tenodesis
  – Calcaneal osteotomy
  – If >10-15 degrees, add lower tibial hemi-epiphysiosodesis or supra malleolar osteotomy

• Subtalar fusions unpredictable
Achilles tenodesis
Foot: Pathology

- Often rigid
- Unopposed tibialis anterior +/- peronei
- Talus and os calcis rotated
- Subluxed calcaneocuboid and talonavicular joints
- ? tibial torsion
- ? pes cavus
- ? hammer / claw toes; callosities
Foot- general approach

• Aim for:
  – prevention of high pressure points
  – plantigrade foot
  – mobile, braceable foot (total contact AFO)

• Operate when child is starting to stand

• Radical releases +/- bony procedures

• Prefer osteotomies to fusions

• No two cases the same; tailor the Rx
Hindfoot deformities

Cavovarus

- Cavus causes the varus
- Assess hindfoot suppleness by Coleman’s test
- Mobile hindfoot; plantar release
- Stiff hindfoot; add closing wedge osteotomy of base of first metatarsal
Hindfoot deformities

Supination

Due to unopposed tibialis anterior

- Mobile with no gastrocnemius
- Mobile with active gastrocn.
- Fixed deformity

- Tenotomise tibialis anterior
- Transfer of tibialis anterior to dorsum
- Osteotomise base of 1st MT and 1st cuneiform
Hindfoot deformities

*Calcaneovalgus*

Due to dorsiflexors and evertors

- Bulky heel- ulcers
- Splinting / serial casts
- Tenotomise dorsiflexors and peronei longus & brevis
- Medial displacement os calcis osteotomy
Hindfoot deformities

*Vertical talus*

- Requires surgical treatment +/- prior splintage
- Radical posteromedial-lateral release
  - Achilles lengthening
  - tib post lengthening
  - subtalar release
  - lateral talocalcaneal ligament and capsule
Hindfoot deformities

*Isolated Equinus*

- Occasionally isolated and symptomatic
- Sequentially;
  - Serial cast
  - Excise segment of Achilles
  - Radical posterior release
  - Osteotomy or talectomy
Hindfoot deformities

*Equinovarus club foot*

- Starting to walk
  - Radical posteromedial-lateral release
  - Tenotomise tibialis anterior
- Older child
  - Shorten lateral column- os calcis wedge osteotomy
  - Add tallectomy in severe cases
Mid & Forefoot Deformities

- Claw/hammer toes
  - flexor tenotomy
- Pes cavus
  - Plantar fascia release +/- 1st MT closing wedge osteotomy
- Claw/hammer toes with pes cavus
  - tenotomy plus metatarsal suspension
Jones Procedure: metatarsal suspension
Summary

• Realistic aims for adulthood
• Anticipate problems
• Detailed assessment - understand pathology
• Individualise treatment - wise matching of the right patient to the right procedure(s)
Thank You