Physical Therapy & Myelodysplasia (Spina Bifida)
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Introduction

• Group of congenital defects of spinal cord, may include the brain
• Functional outcome varies with affected part of central nervous system
• May have genetic component
• Functional needs change over time and depend on secondary impairments
• Not equivalent to spinal cord injury in children
History

• Death prior to 1970
• Increased survival
  • Early closure of lesion
  • Shunt
  • Bladder program
  • Equipment
  • Delivery by c-section
• Fetal surgery: in utero repair
Fetal Surgery to correct defect

Newborn showing surgical site
Facts

• Etiology
  • Genetic and environmental factors

• Diagnoses
  • Test alpha-fetoprotein level at 15-20 weeks
  • Ultrasound for confirmation, high resolution US can predict motor level

• Incidence
  • 0.4-0.9/1000 U.S. births
  • Lower in African-Americans, high in Hispanic and highest for those of Celtic origin
Prevention: Folic Acid

• Prevents 50-70% neural tube defects
• 1996 folic acid added to grain products
• Recommendation for women: 0.4 mg/day
Pathoembryology

• Lesion occurs 22-28 days gestation
• Lesion related to 2 abnormal processes of nervous system formation:
  • Neurulation: folding of ectoderm on each side of notochord to form a tube, C-1 to S-2
  • Canalization: development of spinal cord distal to S-2

Overview of early Neural Tube Formation

neural plate  neural groove  neural tube

Layers

Ectoderm
Mesoderm (containing notocord)
Endoderm

Images - Lance Davidson
Types of Myelodysplasia: Meningocele

- No neural elements
- Usually no paralysis
Types of Myelodysplasia: Myelomeningocele

- Neural elements
  - Complete & corrected
  - Malformed & disconnected

- Impairment
  - Paralysis
  - Sensory loss
  - Mobility
  - Bowel & bladder
Impairments: Hydrocephalus

- Occurs in 80%
- Excessive cerebro-spinal fluid in ventricles
- Enlarged head if untreated
- Arnold-chiari malformation
- May require shunt
  - Signs of shunt failure: headache, fever, speech and vision problems, spasticity, change in performance
Hydrocephalus: Cognition and Perception

• Visual perceptual problems
• Decreased reading and math scores
• Cocktail Party Syndrome
  • Good vocabulary
  • Language with inappropriate meaning
  • Child may not understand as much as you think they do
Impairments: Bowel & Bladder

• Sphincter at sacral level
• Bladder Program
  • Goal: infection-free social continence preserving renal function
  • Clean intermittent catheterization
  • Prevent vesicouretral reflux and UTI
• Bowel Program
  • Prevent constipation
Secondary Impairments

- Contractures
- Obesity
- Fractures
- Pressure sores
- Latex allergy
- Tethered cord
- Seizures
- Hip dislocation
- Neuromuscular scoliosis
Physical Therapy: Assessment

- Strength: position young child against gravity, older child use traditional test
- Range of Motion
- Tone: flaccid in non-innervated muscles
- Sensation
- Skin
- Developmental Testing
Physical Therapy: Treatment

- **AGE**
  - Newborn and Infant
  - Toddler and Preschool
  - School Age

- **Level of Function**
  - Thoracic Level T 12 and above
  - High Lumbar Level L1-3
  - Low Lumbar Level L4-5
  - Sacral Level S1-2
PT Treatment: Newborn and Infant

- Seating and Positioning
- Range of Motion
- Splinting
- Education
- Ongoing Assessment of Strength
PT Treatment: Toddler and Preschool

- Developmental Milestones
- Positioning
- Stretching and strengthening
- Mobility and Activities of Daily Living (ADL)
- Environmental Exploration
- Prevent Secondary Impairments
PT Treatment: School Age

- Positioning
- Stretching and strengthening
- Independence in ADL’s
- Independent mobility
- Environmental Adaptations
- Recreational Activities
- Prevent secondary impairments
PT Treatment: Thoracic Level T12 & above

- Impairments
  - No active muscles in legs and possibly trunk
  - Contractures: external rotation
  - Clubfeet
  - Kyphoscoliosis
  - Pressure sores

- Treatment
  - Positioning
  - Function: rolling, sitting, commando crawl, wheelchair
  - Skin Protection
PT Treatment: High Lumbar Level L1-3

- Impairments
  - Hip flexion contractures
  - Hyperextended trunk posture
  - Risk dislocated hips, windswept hips
  - No active hip extension

- Treatment
  - Function: prone, quadruped pull to kneel, sit hands free
  - Ambulate with Reciprocating Gait Orthosis (RGO) or Hip-Knee-Ankle-Foot Orthosis (HKAFO) and walking aids
PT Treatment: Low Lumbar Level L4-5

- Impairments
  - Hip flexion contractures
  - Hyperextended trunk to promote standing balance

- Motor
  - Knee extension, some dorsiflexion and abduction

- Treatment
  - Function: crawling
  - Ambulation: household and community using ground reaction force ankle-foot orthosis or Knee-Ankle-Foot Orthosis (KAFO) with or without crutches
PT Treatment:
Sacral Level S1-2

• Impairments
  • Foot contractures
  • Pressure sores on feet

• Motor
  • Hip extensors, plantarflexors

• Treatment
  • Ambulation: 12-14 months-independent by 5 years, walker to crutches to no device, teach falling, Supramalleolar Orthosis (SMAFO) bracing
References