Supporting Children with Neuromuscular Spinal Deformity Needing Scoliosis Surgery: a Team Approach

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OUTLINE:
- Introduction
- Prevalence
- Risk Factors
- Natural History and impact on function
- Treatment
- Outcomes
- Complications
- Case Presentation

Introduction:
Cerebral palsy is a disorder of movement and posture secondary to a static encephalopathy affecting the immature brain.

Incidence: 2-3/1000 live births

Causes: Pre, peri and postnatal causes

Co-morbidities:
It may be associated with a number of comorbidities depending on the extent of the brain lesion.

- CNS – seizure disorder, abnormal tone, poor cognition, learning disabilities, attention & communication issues
- Vision, Hearing & dental - impairment
- RS – Reactive / restrictive pulmonary disease, sleep apnea
- GI – GERD, dysphagia, G tube, constipation, drooling
- Nutrition – poor
- GU – Incontinence, UTI, voiding dysfunction
- MSK – contractures, deformities, decreased ambulation, osteoporosis
- Psychiatry – behavioral issues, sleep
- Pain – MSK, GI causes, dental, GU
Surgery in children with CP: national utilization rates

Children with cerebral palsy may need a number of inpatient admissions for surgery. The top five surgeries they have during their lifetime are as follows:

1. Gastrostomy
2. Musculoskeletal soft tissue surgery
3. Fundoplication
4. Scoliosis surgery
5. Bony hip surgery

Costs and Complications of Hospitalizations in children with CP

As compared to normally developing children, inpatient admissions in children with cerebral palsy are more likely to have:

• Longer Hospital stays
• Higher total charges
• More associated diagnosis
• More procedures
• Higher complication rates
• More likely to be transferred

CP Neuromuscular Scoliosis:

Overall prevalence: 25%

Risk factors: GMFCS 4 &5, increasing age, Spastic diplegia and quadriplegia. Prevalence is around 50% in children with GMFCS 4& 5 level function by the age of 18 years.

Natural History of untreated NM scoliosis:

• Slow Progression initially
• Once curve is > 50 degrees will rapidly progress with pelvic obliquity
• Difficulty with sitting and loss of upper extremity function
• Increased risk of ischial decubiti
• Increased risk of costo-pelvic impingement and pain
• Progression of restrictive lung disease

Treatment of NM scoliosis – nonsurgical options:

• Better tone management
• Maintaining ambulation / standing
• Proper seating and positioning, bracing

**Treatment of NM scoliosis – surgical options:**

*a) Indications for surgery:*

• Progressive curve > 50
• Loss of sitting balance
• Ischial decubiti
• Costo-pelvic impingement
• Deteriorating Pulmonary function

*b) Goals of surgery:*

• Prevent progression
• Correct safely to restore spinal balance
• Facilitate seating & positioning
• Pain relief & reduce complications (decubiti, pulmonary)
• Improve quality of life

*c) Details of surgery:*

• Posterior spinal fusion
• Posterior spinal instrumentation from T2 to pelvis
• Severe, rigid curves (> 100 degrees)
  – Anterior spinal release ??
  – Anterior spinal fusion ??
• Young children
  – Fusionless spine surgery

**Outcomes of surgery:**

• HRQL improved significantly 71 %
• CPCHILD score increased from 45 to 58
• Change in Pain was the most significant factor affecting QOL changes
• Parent satisfaction 92%
• Sitting balance improved 93%
• Cosmesis improved 94%
• Functional improvement limited 8 – 40%

**Complications: 30% Overall**
1) Pulmonary: 22 (30%) - pneumonia, pneumothorax, atelectasis, pleural effusion, prolonged mechanical ventilation and longer stay in intensive care unit (ICU).

2) Hardware related: 12.5 % (implant perforation and penetration, revision of implant for infection and skin irritation, implant breakage, loosening or cut-out of implant.

3) Infections: 10.9 %
   a. Prevalence: 3 – 15 % vs 0.5% in AIS
   b. Risk factors: ASA score >= 2, Cerebral palsy, GMFCS 4/5, VP shunt, bladder / bowel incontinence, malnutrition, obesity, hypothermia, prolonged operative time, blood loss and transfusions, number of levels fused > 10, fixation to pelvis, implant prominence, implant type – unit rods Vs contoured rods, implant material – stainless steel Vs Titanium, postop drains reduces risk,
   c. Steps to prevent SSI: see above

4) GI: paralytic Ileus (5 % vs 3%), pancreatitis,

5) Wound healing:

6) Bleeding:

7) Neurologic: 3 % (0.5%)

8) UTI: 5 % vs 0.7 %

9) Central lines placed in 6 % vs 1.3 %

10) Pseudarthrosis: 11 %

Review of Literature:

- Does PSF improve quality of life of CP children with NM scoliosis?
- Complication rates of PSF for NM scoliosis in CP children.
- What measures optimize outcomes of PSF for NMS in CP children?

TEAM Approach to children with childhood disabilities:

- Team: ‘‘a small group of people with complementary skills who are committed to a common purpose, performance goals, and approach for which they are mutually accountable.’’

Steps to Optimize care in CP children having surgery for NMScoliosis.

1) Preoperative:
   a. Detailed multisystem Evaluation – pulmonary, GI, GU, CNS, dental, CVS
   b. Care pathway with a dedicated care coordinator: Matched case comparison study -reduces LOS, PICU stay and complication rates
   c. Preoperative chlorhexidine prep night before and morning of surgery
   d. Preoperative urine cultures and treatment if positive
   e. Nutritional evaluation & supplementation
   f. Preoperative patient education sheet, shared decision making
   g. Preoperative nasal decolonization
   h. Alteration of seizure medications,
i. Optimizing bone health
j. Treating constipation
k. Optimizing dental health
l. Understand communication issues – CFCS classification, assistive technology

2) **Intraoperative:**
   a. Maintaining optimal pre & intraoperative body temperature (> 36C)
   b. Total intravenous Anesthesia (TIVA)
   c. Multimodal neural monitoring – SSEPS, MEPS
   d. Anti fibrinolytics- TXA
   e. Hair clipping preferred to shaving
   f. IV antibiotics – (ancef + gram –ve cover, Vancomycin powder in bone graft) agent, timing, dosing, redosing (every 4 hours)
   g. High efficiency teams – select surgeons, anesthetiologists, scrub techs, select circulating nurses
   h. Two surgeons – Shrader MW et al matched case comparison study showed decreased surgical time, less EBL, LOS, fewer complications
   i. Cell saver
   j. Reduce traffic in OR / limited access
   k. Minimising infections – Irrigation
   l. Impervious dressings
   m. Closing lower ½ of incision with a nonabsorbale stitch

3) **Postoperative:**
   a. Hospitalist co-management: Reduces variation in outcomes, improves quality but with increased initial costs.
   b. Postoperative IV antibiotics till removal of catheters & drains (day 3)
   c. Reducing postop dressing changes

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