Spasticity Management in EOS: What works and what doesn’t

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Disclosures

• Consultant and Teaching Arrangements: Depuy; Biedermann Motech; Stryker

• Royalty agreements: Stryker

• Institutional research support: Childress Institute for Pediatric Trauma
Overview

• Spasticity basics/primer
• Discuss the current treatments available
• Answer some specific questions regarding children with spinal deformity and spasticity
Definition

• Velocity dependent resistance to movement

• Altered skeletal muscle performance marked by:
  – Paralysis
  – Increased tendon reflex / hyperreflexia
  – Hypertonia

• Other types of hypertonicity
  – Dystonia
  – Rigidity
  – Athetosis-Dyskinesia
Etiology

• Damage to the CNS → decreases the net inhibition of peripheral nerves
  – Cerebral palsy
  – Multiple sclerosis
  – Spinal cord injury
  – Acquired brain injury
When do we treat spasticity? (Tone management)

• How severe is it?
• What are your goals?
  – Functional
  – Ease of care
  – Positioning
  – Pain
  – Contractures/joint deformities
State of the Art

• Therapy based:
  – PT/OT: ROM, Electrical stimulation, FES
  – Splinting/casting (static, dynamic)
  – Oral medications
    • Benzos (most commonly valium)
    • Baclofen
    • Dantrium
    • Tizanidine
    • Neurontin
    • Trazadone
State of the Art

• Intervention
  – Injected medication
    • Botulism toxin (Type A and type B)
    • phenol
  – Surgery
    • Intrathecal baclofen pump insertion
    • Selective dorsal rhizotomy
    • Orthopaedic surgery
Injections

• Phenol
  – Need compounding pharmacy
  – Cheap, but more time consuming
  – Immediate effect

• Botox
  – Most effective in smaller muscles
  – Peak effect is 7-10 days post-injection
ITB

- Programmable pump/reservoir
- Candidates for therapy typically have severe spasticity and have responded favorably to a trial dose.
- Carries higher complication profile than SDR
- Reversible
SDR

- Commonly used in ambulatory patients
- Fewer complications
- Permanent
SDR vs. ITB

• SDR and ITB have historically been used to achieve different goals.

• SDR has traditionally been favored in ambulant children with moderate CP (GMFCS level II or III) to improve their gait.

• ITB has historically been reserved for children with 4-limb involvement who are nonambulatory, usually with the goal of reducing spasticity as a way to improve comfort and decrease the caregiver burden.

• However, there are an increasing number of reports of profound benefits in GMFCS level IV/V following SDR.
Questions in 2019

• All things being equal, is SDR or ITB better?
• Does surgical treatment of spasticity in the skeletally immature patient INCREASE curve progression?
• Does type of treatment matter in patients at risk for curve progression?
Selective dorsal rhizotomy for spasticity not associated with cerebral palsy: reconsideration of surgical inclusion criteria

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Children with spastic diplegia from cerebral palsy (CP) experience measurable improvement in their spasticity and motor function following selective dorsal rhizotomy (SDR). The role of this operation in the treatment of other spasticity causes is less well defined. A literature review was undertaken to survey outcomes from SDRs performed outside the CP population. Multiple sclerosis was the most common diagnosis found, accounting for 74 of 145 patients described. Selective dorsal rhizotomies have also been reported in patients with traumatic brain and spinal cord injuries, ischemic and hemorrhagic stroke, neurodegenerative disease, hypoxic encephalopathy, and other causes of spasticity. Outcomes from surgery are generally described as favorable, although postoperative assessments and follow-up times are not standardized across reports. Long-term outcomes are sparsely reported. Larger numbers of patients and more detailed outcomes data have the potential to form a basis for expanding the inclusion criteria for SDR.

(10.3171/2013.8.FOCUS13294)

KEY WORDS • selective dorsal rhizotomy • spasticity • cerebral palsy • hypertonia

SELECTIVE dorsal rhizotomy (SDR) is a standard treatment option for spastic paraparesis associated with cerebral palsy (CP) in selected patients. However, the diagnosis of CP encompasses a broad variety of underlying pathologies that appear early in life secondary to significant sensory loss and ataxia.¹¹ The technique of isolating dorsal lumbar nerve fascicles and dividing only those demonstrating some type of abnormal physiological response on electrical stimulation was developed later.¹²,²⁵ This strategy resulted in a significant reduction in adverse
Intrathecal baclofen versus selective dorsal rhizotomy for children with cerebral palsy who are nonambulant: a systematic review

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OBJECTIVE Cerebral palsy (CP) is the most common childhood physical disability. Historically, children with hypertonia who are nonambulatory (Gross Motor Function Classification System [GMFCS] level IV or V) were considered candidates for intrathecal baclofen (ITB) therapy to facilitate care and mitigate discomfort. Selective dorsal rhizotomy (SDR) was often reserved for ambulant children to improve gait. Recently, case series have suggested SDR as an alternative to ITB in selected children functioning at GMFCS level IV/V. The objective for this study was to systematically review the evidence for ITB and SDR in GMFCS level IV or V children.

METHODS Medline, Embase, Web of Science, and Cochrane databases were systematically searched. Articles were screened using the following inclusion criteria: 1) peer-reviewed articles reporting outcomes after SDR or ITB; 2) outcomes reported using a quantifiable scale or standardized outcome measure; 3) patients were < 18 years old at the time of operation; 4) patients had a diagnosis of CP; 5) patients were GMFCS level IV/V or results were reported based on GMFCS status and included some GMFCS level IV/V patients; 6) article and/or abstract in English; and 7) primary indication for surgery was hypertonia. Included studies were assessed with the Risk of Bias in Non-Randomized Studies - of Interventions (ROBINS-I) tool.

RESULTS Twenty-seven studies met inclusion criteria. The most commonly reported outcomes were spasticity (on the Mean Ashworth Scale) and gross motor function (using the Gross Motor Function Measure), although other outcomes including frequency of orthopedic procedures and complications were also reported. There is evidence from case series...
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• Collection of case series (no comparative studies) that provided evidence that both treatments can lower spasticity and improve gross motor function in nonambulatory patients

• Rates of PSF
  – 0% to 28% in the SDR articles
  – 17%–23% in the ITB articles

• Complication rates are higher in the ITB group
Intrathecal baclofen therapy for treatment of spasticity in infants and small children under 6 years of age

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Received: 9 April 2019/Accepted: 5 August 2019
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Abstract
Purpose The aim of this study is to prove the efficacy and safety of intrathecal baclofen therapy in infants and children below 6 years of age by retrospective analysis of our pediatric cohort of 135 primary pump implantations.
Methods Between 2007 and 2018, 17 patients with pump implantations were below 6 years of age. Data were acquired retrospectively with a follow-up of 12 months to 11 years regarding complications.
Results The youngest infant was 11 months at implantation with a bodyweight of 6, 4 kg, and 63 cm length. Surgical complications were comparable to published literature and mainly involved the catheter (2 catheter dislocations and 1 catheter transection). Catheter function was not affected by the age of the patients.

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- 17 patients
- 11 months (6.4kg)

Fig. 1 Smallest infant implanted at 11 months of age, 6, 4-kg body weight, 63-cm length, using radiopaque Indura 1P catheter by Medtronic. a At 6 months, although already indicated for ITB therapy, there is not enough soft tissue space for implantation in the “pump triangle”. b X-ray post-surgery at 11 months. C: 4 years post-surgery
Rapid Progression of Scoliosis Following Insertion of Intrathecal Baclofen Pump

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Abstract: Placement of an intrathecal baclofen pump is a common treatment of spasticity in cerebral palsy patients. Though effective, the hardware is prone to malfunction, and multiple revisions are often necessary. Additional complications have also been described, including infection and drug toxicity or withdrawal. The authors report another complication of intrathecal baclofen therapy: accelerated progression of scoliosis after pump insertion. A retrospective chart review was performed on four patients who showed accelerated scoliotic progression after intrathecal baclofen pump insertion for treatment of spasticity. Cobb angles were measured from preoperative and postoperative radiographs to determine the rate of scoliotic degeneration both before and after pump insertion. Although there may not be a direct correlation between intrathecal baclofen and scoliosis, the authors consider the issue important enough to warrant discussion with any patient considering intrathecal baclofen.

Key Words: Cerebral palsy, spasticity, IRI, Cobb angle, post-membrane defect, dysmetria, and seizures. In addition, pump hardware itself is prone to malfunction: infection is common, and pump failure occurs in 5% to 10% of patients.

We report the occurrence of another complication associated with intrathecal baclofen therapy: rapid progression of scoliosis after pump insertion.

METHODS

Under Institutional Review Board approval at the University of Wisconsin Hospital and Clinics, a retrospective chart review was performed on four patients who exhibited rapid scoliotic deterioration requiring spinal fusion after insertion of an intrathecal baclofen pump for treatment of spasticity secondary to cerebral palsy (Table 1). Information on patient age and gender, dates of pump placement and spinal fusion, baclofen dose, dosing schedule, and dosing modifi-
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- 4 patients with rapid progression of their curve following ITB treatment
Do baclofen pumps influence the development of scoliosis in children?

Clinical article

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Object. Intrathecal baclofen is an effective treatment for spasticity in patients with cerebral palsy. There has been increasing concern, however, that intrathecal baclofen may accelerate the development of scoliosis in this population. To this end, the authors reviewed their population of pediatric patients with baclofen pumps to assess the incidence of scoliosis.

Methods. This was a retrospective chart and radiology review of all pediatric patients with baclofen pumps. Cobb angles were measured preoperatively and on follow-up images.

Results. Of 38 patients identified, 32 had adequate data available for inclusion in the study (16 with cerebral palsy, 7 with dystonic cerebral palsy, 4 with head injury, and 5 with other diagnoses). The mean age at pump insertion was 10.6 years and the mean follow-up period was 31 months (range 1–118 months). The mean annual Cobb angle
Intrathecal baclofen pumps do not accelerate progression of scoliosis in quadriplegic spastic cerebral palsy

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Received: 20 January 2016 / Revised: 28 April 2016 / Accepted: 29 April 2016 / Published online: 6 May 2016 © Springer-Verlag Berlin Heidelberg 2016

Abstract
Purpose To compare scoliosis progression in quadriplegic spastic cerebral palsy with and without intrathecal baclofen (ITB) pumps.
Methods A retrospective matched cohort study was conducted. Patients with quadriplegic spastic cerebral palsy, GMFCS level 5, treated with ITB pumps with follow-up >1 year were matched to comparable cases by age and baseline Cobb angle without ITB pumps. Annual and peak coronal curve progression, pelvic obliquity progression and

Conclusions Patients with quadriplegic spastic cerebral palsy with and without ITB pumps showed significant curve progression over time. ITB pumps do not appear to alter the natural history of curve progression in this population.

Keywords Scoliosis · Intrathecal baclofen · Cerebral palsy · Spastic quadriplegia

Introduction
Of the 9 patients without scoliosis in the ITB group, all developed scoliosis. 5/6 in the non-ITB group. 5 in ITB group and 9 in non-ITB group went on to require spinal fusion.

### Table 1 Baseline data for the two matched groups

<table>
<thead>
<tr>
<th></th>
<th>ITB pump (n = 25)</th>
<th>Non-ITB pump (n = 25)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>9.4 (4.2)</td>
<td>9.2 (4.0)</td>
<td>0.16</td>
</tr>
<tr>
<td>Sex</td>
<td>16 M 9 F</td>
<td>11 M 14 F</td>
<td>0.26</td>
</tr>
<tr>
<td>Baseline coronal Cobb angle</td>
<td>25.6° (22.4)</td>
<td>29.7° (21.9)</td>
<td>0.06</td>
</tr>
<tr>
<td>Scoliosis at enrollment</td>
<td>16/25</td>
<td>19/25</td>
<td>0.54</td>
</tr>
<tr>
<td>Pelvic tilt</td>
<td>3.2° (6.7)</td>
<td>7.1° (9.0)</td>
<td>0.04*</td>
</tr>
<tr>
<td>Pelvic tilt at enrollment</td>
<td>10/25</td>
<td>6/25</td>
<td>0.36</td>
</tr>
<tr>
<td>Risser grade at enrollment</td>
<td>0.9 (1.4)</td>
<td>1.0 (1.5)</td>
<td>0.70</td>
</tr>
<tr>
<td>Baseline hip instability</td>
<td>10/25</td>
<td>4/25</td>
<td>0.11</td>
</tr>
<tr>
<td>Follow-up (range)</td>
<td>4.3 (1.0–7.8)</td>
<td>3.5 (1.0–7.5)</td>
<td>0.16</td>
</tr>
</tbody>
</table>

Value given as mean (SD) unless otherwise stated
* indicates statistical significance

- Scoliosis defined as Cobb angle >10°
- Pelvic tilt vs horizontal >5°
- Hip instability defined as radiographic evidence of hip subluxation/dislocation/prior surgery for hip dislocation
Rushton, et. al.

- Of the 9 patients without scoliosis in the ITB group, all developed scoliosis. 5/6 in the non-ITB group.
- 5 in ITB group and 9 in nonITB group went on to require spinal fusion
Risk factors for progressive neuromuscular scoliosis requiring posterior spinal fusion after selective dorsal rhizotomy

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OBJECTIVE Selective dorsal rhizotomy (SDR) via limited laminectomy is an effective treatment of lower-extremity spasticity in the pediatric population. Children with spasticity are also at risk for neuromuscular scoliosis; however, specific risk factors for progressive spinal deformity requiring posterior spinal fusion (PSF) after SDR are unknown. The authors' goal was to identify potential risk factors.

METHODS The authors performed a retrospective cohort study of patients who underwent SDR via limited laminectomy between 2003 and 2014 and who had at least 1 year of follow-up. They analyzed demographic, clinical, and radiographic variables to elucidate risk factors for progressive neuromuscular scoliosis. The primary outcome was need for PSF.

RESULTS One hundred thirty-four patients underwent SDR and had at least 12 months of follow-up (mean 65 months); 48 patients (36%) had detailed pre- and postoperative radiographic data available. The mean age at surgery was 10 years (SD 5.1 years). Eighty-four patients (63%) were ambulatory before SDR, 109 (82%) underwent a single-level lami-
Ravindra, et al.

• Nonambulatory status and preoperative Cobb angle > 30° were associated with a need for PSF after SDR (univariate analysis)

• No variables were independently associated with PSF after SDR in the multivariable analysis.

• Did not identify any variables that clearly modify the risk of progressive neuromuscular scoliosis in patients undergoing SDR via limited laminectomy.
Spasticity treatment in 2019

• The indications are expanding for SDR
• Surgical treatment of spasticity is safe for the skeletally immature patient
• Collaboration is a GREAT thing
Thank you!