CASE 8.4
Cerebral Palsy | Level 3

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1. Identify subjective and objective findings indicative of spasticity or other complications associated with cerebral palsy.

SUBJECTIVE FINDINGS: The patient's chief complaint of pain, as defined by his mother, is a frequent complication of spasticity. Discomfort with movement, stiffness, and decreased mobility are symptomatic of spasticity. The patient's mother also reported a history of generalized spasticity with an early diagnosis of cerebral palsy. Spasticity is the most common motor abnormality associated with *cerebral palsy*, a term used to describe a range of developmental disorders of movement and posture. By definition, cerebral palsy must be ascribed to an injury or other disturbance in the developing fetal or infant brain. The majority of cases are acquired prenatally in contrast to this patient whose cerebral palsy is etiologically related to meningitis that occurred at 3 weeks of age. The noted intellectual disability, drooling, and constipation are consistent with additional common complications of cerebral palsy.

OBJECTIVE FINDINGS: The patient's pain has been evaluated using the **F**ace **L**egs **A**ctivity **C**ry and **C**onsolability (FLACC) behavioral pain scale. To assess a patient using the FLACC scale, the clinician observes the patient's behaviors in each of the five categories. Each category is scored from 0 to 2 using a rubric. A total score is then calculated between 0 and 10, with 10 indicating the highest assessment of pain. The patient's spasticity is generalized as it is found in all four extremities. The Modified Ashworth Scale rating of 3 indicates severe spasticity with difficult passive movement due to excessive increase in muscle tone. The inability to ambulate as noted by the rating of 5 in the Gross Motor Function Classification System (GMFCS) is also consistent with severe disease. The presence of contractures indicates prolonged spasticity. Truncal hypotonia and increased deep tendon reflexes are signs of abnormal neurologic control. Cognitive impairment is indicated by the developmental history findings of communication consisting of babbling and use of a communication board for five to six simple words. Excessive drooling with pooled saliva in the mouth are signs of impaired oral-motor function. The poor dentition is most likely related to excessive drool. The patient's gastroesophageal reflux disease (GERD) may contribute to the poor dentition. Impaired oral-motor function is a factor in the patient's failure to thrive. The patient's height and weight are at the 5th percentile on growth charts, and he requires enteral nutritional support. The osteopenia seen on x-ray is a signal for low bone mineral

density that is related to the patient's nonambulatory condition and nutritional deficits. The decreased bowel sounds and palpable stool found on abdominal exam and the gas-diluted loops of bowel seen on x-ray are signs of constipation.

2. Evaluate the suitability of intrathecal baclofen therapy to treat the patient's increased spasticity.

It is important to prioritize addressing the patient's chief complaint of pain. A pain scale, such as the FLACC behavioral assessment tool, can be particularly helpful in the evaluation of level of pain in nonverbal patients. A revised version of the FLACC scale is available that includes behaviors associated with pain in children with cognitive impairment in the scoring rubric. Using the FLACC behavioral scale, this patient has been assessed as experiencing significant pain and discomfort. A thorough evaluation should be undertaken to identify the source of pain and to treat the underlying cause. Symptomatic pharmacologic treatment should also be provided. Because the patient is already experiencing problems with sedation and constipation, treatment with acetaminophen or a nonsteroidal anti-inflammatory drug would initially be preferred over an opioid. Although the patient has had only minimal relief with ibuprofen, intravenous ketorolac is a reasonable initial choice for symptomatic relief with careful monitoring for gastrointestinal and renal adverse reactions.

Hip dislocation or subluxation and presence of fracture have been eliminated as possible pain sources in the patient. The increased spasticity should be addressed as a possible cause of the pain. In addition to relieving pain, the goals of optimizing therapy for spasticity include maximizing the patient's motor function, decreasing the contractures he is experiencing, and easing the caregiving needs for his family. Pharmacologic treatment options include a change in oral pharmacotherapy, botulinum toxin treatment, or intrathecal baclofen. The patient has already experienced sedation with oral therapy. Clonazepam was discontinued due to this adverse reaction, and his mother notes undesirable sedation with oral baclofen after a recent increase in dose. The antinociceptive effects of tizanidine could be beneficial in the patient, but its use may be limited by a high incidence of somnolence and uncertainty in optimal dosing. The patient's current slight elevation in liver function tests and positive family history for hepatic disease reduce the viability of treating with oral dantrolene. Botulinum toxin therapy is indicated for localized spasticity while the patient has generalized disease affecting all extremities and major joints. If a particular muscle group can be identified as a source for pain or the increased stiffness, botulinum toxin treatment in those muscles would be beneficial. Based on his poorly controlled spasticity that is affecting quality of life and the limitations of other treatment options, the patient is a candidate for intrathecal baclofen therapy. In addition, efficacy of intrathecal baclofen is best supported in patients with severe spasticity (GMFCS levels IV or V). Performed in a monitored setting, a bolus-screening dose of intrathecal baclofen (25 mcg) can be used to confirm if the patient will likely respond to the continuous infusion given through the implantable pump. The duration of effect of the bolus dose is typically 8 hours and using the Modified Ashworth or similar scale at 2-hour intervals provides a measurable way to assess the patient's response. If a positive effect is found, it would also be important to determine the parents' abilities to manage this therapy and willingness to follow up as recommended prior to initiating continuous infusion treatment through the implantable pump.

3. Develop a plan to appropriately counsel the patient's parents on intrathecal baclofen therapy.

Education of the patient's parents will be a key determinant for the success of intrathecal baclofen therapy. They should clearly understand the goals and expectations of treatment, the steps in initiating therapy (screening, preoperative assessment, surgery, postoperative care, and follow-up), adjustments needed to other medications, potential adverse reactions of baclofen and how to manage them. The parents will need in-depth training on the implantable pump and documentation of the