

Pharmacological Treatment of Spasticity in Children and Adolescents with Cerebral Palsy

Case Presentation

An 8-year-old girl is referred to the neurology clinic by her primary care provider. She is brought by her parents because of excessive tightness in her legs that interferes with diaper change and hygiene. This patient was born to a 23-year-old, gravida 1 mother after a 32-week gestation. Her birth weight was 4 lbs 2 oz, and she had an Apgar score of 7 and 9. She stayed in the NICU for 4 weeks, requiring assisted ventilation. She requires a wheelchair for mobility. She has no other medical problems. She has not had prior surgeries, has no known drug allergies, and is currently on no medications. There is no family history of neurological disease. Review of systems reveals she has limited vision, mild back pain, and incontinence but no difficulties with breathing, swallowing, or joint pain. She has no difficulties with constipation and no changes in urination. Her mood, behavior, and sleep are not an issue. She is in second grade and gets physical, occupational, and speech therapy at school.

On physical examination vitals are BP 106/68, HR 60 and regular, and RR 12. Her weight is in the 25th percentile, and her height is in the 50th percentile. Her head circumference is in the 50th percentile. She is in no acute distress. She is sitting in a wheelchair with a Gross Motor Function Classification System (GMFCS) level IV. Cardiac auscultation reveals normal heart sounds; lung sounds are clear. She is alert and follows simple commands. Her receptive language and expressive language are normal for her age, although there is mild dysarthria. Her memory is intact for three simple words in 5 minutes. She is oriented to self, parents, and location. Cranial nerve testing reveals a normal fundoscopic examination and difficulty with visual fixation; eye movements are full, and facial sensation is symmetric. Her mouth is open, and she is able to close it on command. Hearing is intact. Palate/tongue and uvula are midline, and she has a normal gag reflex. Sternocleidomastoid is 5/5 bilaterally; her tongue has normal movements without evidence of atrophy or fasciculations. Motor strength is 5-/5 throughout the upper extremities; legs are 3/5 bilaterally, with decreased bulk, and marked lower-extremity spasticity affecting the hip adductors, hamstrings, and ankle plantar flexors. There is fixed joint flexion contractures in both hips (10°), both knees (110° popliteal angle), and both ankles (80°). Hip abduction is 45° bilaterally with a catch and release at 15° . There is a negative Galeazzi sign. Sensory function is intact to light touch and pinprick throughout. Reflexes are 2/4 in the arms and 3/4 at the knees, and she has 10 beats of clonus in the ankles bilaterally. Coordination is mildly impaired in rapid alternating movements. Finger-nose-finger is normal. Coordination testing is limited in the legs by spasticity and weakness. She is able to stand and take a few steps with lots of assistance. When she stands, there is marked hip hyperadduction (scissoring). She can do independent transfers slowly with someone in attendance.

You reviewed the outside MRI films of the brain and agree the study is consistent with periventricular leukomalacia.

You diagnose the patient with cerebral palsy and, specifically, spastic diplegia with significant adductor spasticity due to periventricular leukomalacia. The patient's parents are interested in medical management of the spasticity to improve diaper change and perineal hygiene. You discuss with them the various medication options, to include botulinum toxin type A, and, possibly, diazepam and tizanidine. When they ask about the evidence to support the treatments, you notify them that there is current evidence that:

Questions

1. For localized/segmental spasticity that warrants treatment, _____ should be offered as an effective and generally safe treatment (Level A).¹
- A. Botulinum toxin type A (BoNT-A)
 - B. Dantrolene
 - C. Continuous intrathecal baclofen (ITB)
 - D. Oral baclofen
 - E. Tizanidine

The correct answer is A. For localized/segmental spasticity that warrants treatment, BoNT-A should be offered as an effective and generally safe treatment (Level A).¹

2. For generalized spasticity that warrants treatment, diazepam should be considered for _____, with caution regarding _____.
- A. Long-term treatment, toxicity
 - B. Long-term treatment, addiction
 - C. Short-term treatment, addiction
 - D. Short-term treatment, toxicity
 - E. Spasticity only in adults, addiction

The correct answer is D. For generalized spasticity that warrants treatment, diazepam should be considered for short-term treatment (Level B), with caution regarding toxicity.¹

3. True or False: There are insufficient data to support or refute the use of phenol, alcohol, or botulinum toxin type B (BoNT-B) (Level U).¹

The correct answer is True.

After discussing the options, the parents would like to proceed with BoNT-A injections today.

ICD-9-CM Coding Discussion

The diagnostic terminology in the record includes spastic diplegia, adductor spasticity, cerebral palsy and periventricular leukomalacia. Adductor spasticity would here be considered a manifestation of cerebral palsy, and spastic diplegia a form of cerebral

palsy. In ICD-9-CM,² “plegia” and “paresis” are interchangeable, and “plegia” is more often the listed descriptor. ICD-9-CM² does not make this diagnosis easy to find in the index. This is a case where the more general term is needed, and even then, the more “general” part of the term. The index entry is found as follows:

Palsy,

- cerebral (congenital) (infantile) (spastic) **343.9**
- athetoid **333.71**
- diplegic **343.0**
 - late effect - see Late effect(s) (of) cerebrovascular disease
- hemiplegic **343.1**
- monoplegic **343.3**
- noncongenital or noninfantile **437.8**
 - late effect - see Late effect(s) (of) cerebrovascular disease
- paraplegic **343.0**
- quadriplegic **343.2**
- spastic, not congenital or infantile **344.89**
- syphilitic **094.89**
 - congenital **090.49**
- tetraplegic **343.2**

And in the Tabular volume of ICD-9-CM² the code is:

- 343** Infantile cerebral palsy
 - 343.0** Diplegic
 - Congenital diplegia
 - Congenital paraplegia

The highest level of specificity for coding here is periventricular leukomalacia, making this the primary code. The cerebral palsy should also be listed as it is not specifically implied by the primary code. The submitted codes for this case would then be:

- 779.7 Periventricular leukomalacia**
- 343.0 Infantile cerebral palsy, diplegic**

Evaluation and Management Coding Discussion

The correct E&M code for the initial patient visit is a level 5 outpatient consultation code, code 99245. The history elicited and the examination is comprehensive and the medical decision making is high complexity based on the number of diagnoses and treatment options which is extensive and the amount of data reviewed or ordered is extensive.

Botulinum Toxin Injections

- 64612** Chemodenervation of muscle(s); muscle(s) innervated by facial nerve (eg, for blepharospasm, hemifacial spasm)

- 64613** neck muscles (e.g., for spasmodic torticollis, spasmodic dysphonia)
- 64614** extremity(s) and/or trunk muscle(s) (eg, for dystonia, cerebral palsy, multiple sclerosis)

Neurophysiological Guidance of Botulinum Toxin Injections

- +95873** Electrical stimulation for guidance in conjunction with chemodenervation (List separately in addition to code for primary procedure)
- +95874** Needle electromyography for guidance in conjunction with chemodenervation (List separately in addition to code for primary procedure)

(Do not report 95874 in conjunction with 95873)

Proper Use of the Botulinum Toxin Injection Codes

From a review of the Medicare Fee Schedule, CMS allows two units of codes 64612, 64613 and 64614 to be submitted if bilateral procedures are performed. According to the 2009 Medicare Physician Fee Schedule, bilateral procedures are payable at 150% of the allowed amount for a unilateral procedure. There is variability in how different insurance carriers handle these codes. Some may allow two units of each of the codes to be submitted if bilateral injections were done. Others may allow only one unit of each of the codes even for bilateral injections. The individual provider will need to determine what is the proper billing procedure for these codes in his or her locality. The official AMA CPT® stance is that these codes are billable once per day, so it is in your best interest to make sure you are following each individual payer's billing guidelines when billing for these services.

Can CPT® codes 64612-64614 be billed together? Yes, they can be reported together as they pertain to different anatomical regions. Can CPT® codes 64612-64614 be billed more than once on a date of service? Only if the procedure is performed bilaterally and the payer allows coding for bilateral procedures.

These codes do not include guidance by EMG or electrical stimulation, if done at the time of the injection, or the drug itself (see below).

How to Code for the Medication Used

In addition to coding for the procedure, physicians should also code for the drug itself. There are different Healthcare Common Procedure Coding System (HCPCS) supply codes³ for the two types of botulinum toxin currently in clinical use:

- J0585** Botulinum toxin type A, per unit
- J0587** Botulinum toxin type B, per 100 units

Botulinum toxin type A is supplied in single-dose vials of 100 units. To receive proper reimbursement, it's important to correctly code the amount used for each patient following individual payer instructions.

Similar considerations apply to botulinum toxin type B, although the units differ.

The Global Period for Chemodenervation Procedures

There is a 10-day global period for chemodenervation procedures. This means that payment for these procedures includes any related services performed one day pre-operatively, on the day of the procedure and 10 days post-operatively. Any E/M service related to the chemodenervation procedure that is performed within the first 10 days post-operatively won't be paid separately. If E/M services rendered in the postoperative period are unrelated to the reason for chemodenervation, then modifier 24 needs to be appended to the E/M code in order for it to be paid, and you must use a different diagnosis code on that claim. A separately identifiable E/M service on the date of injection should be billed with modifier 25, or, if it was a decision for surgery, modifier 57.

Payment Adjustment Rule for Multiple Chemodenervation Procedures

In the Medicare fee schedule, standard payment adjustment rules for multiple procedures apply to chemodenervation codes. If procedure is reported on the same day as another procedure with an indicator of 1, 2, or 3, rank the procedures by fee schedule amount and apply the appropriate reduction to this code (100%, 50%, 50%, 50%, 50% and by report). Base the payment on the lower of (a) the actual charge, or (b) the fee schedule amount reduced by the appropriate percentage.

About the Codes for Neurophysiological Guidance of Botulinum Toxin Injections

Prior to the chemodenervation procedure, it is sometimes necessary to perform a more precise localization for needle placement before the chemical is injected. Therefore the physician may perform electrical stimulation or needle EMG to achieve this localization.

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Patient Safety Tip

It is important to consider the weight of the child when treating with botulinum toxin. It is advisable to initiate treatments at the lower range of the therapeutic spectrum, with gradual incremental increases.

1. Delgado MR, Hirtz D, Aisen M, et al. Practice Parameter: Pharmacologic treatment of spasticity in children and adolescents with cerebral palsy (an evidence-based review): Report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurological Society. *Neurology*® 2010;74:336-343.
2. Centers for Disease Control and Prevention. International classification of diseases, ninth revision, clinical modification (ICD-9-CM). www.cdc.gov/nchs/icd/icd9cm.htm.

3. For current Level II HCPCS codes visit:
<http://www.cms.gov/Medicare/Coding/HCPCSReleaseCodeSets/Alpha-Numeric-HCPCS.html>

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