

Successful Pediatric Epidural Steroid Injection for Radicular Pain in Congenital Myasthenic Syndrome: Considerations for the Pain Physician

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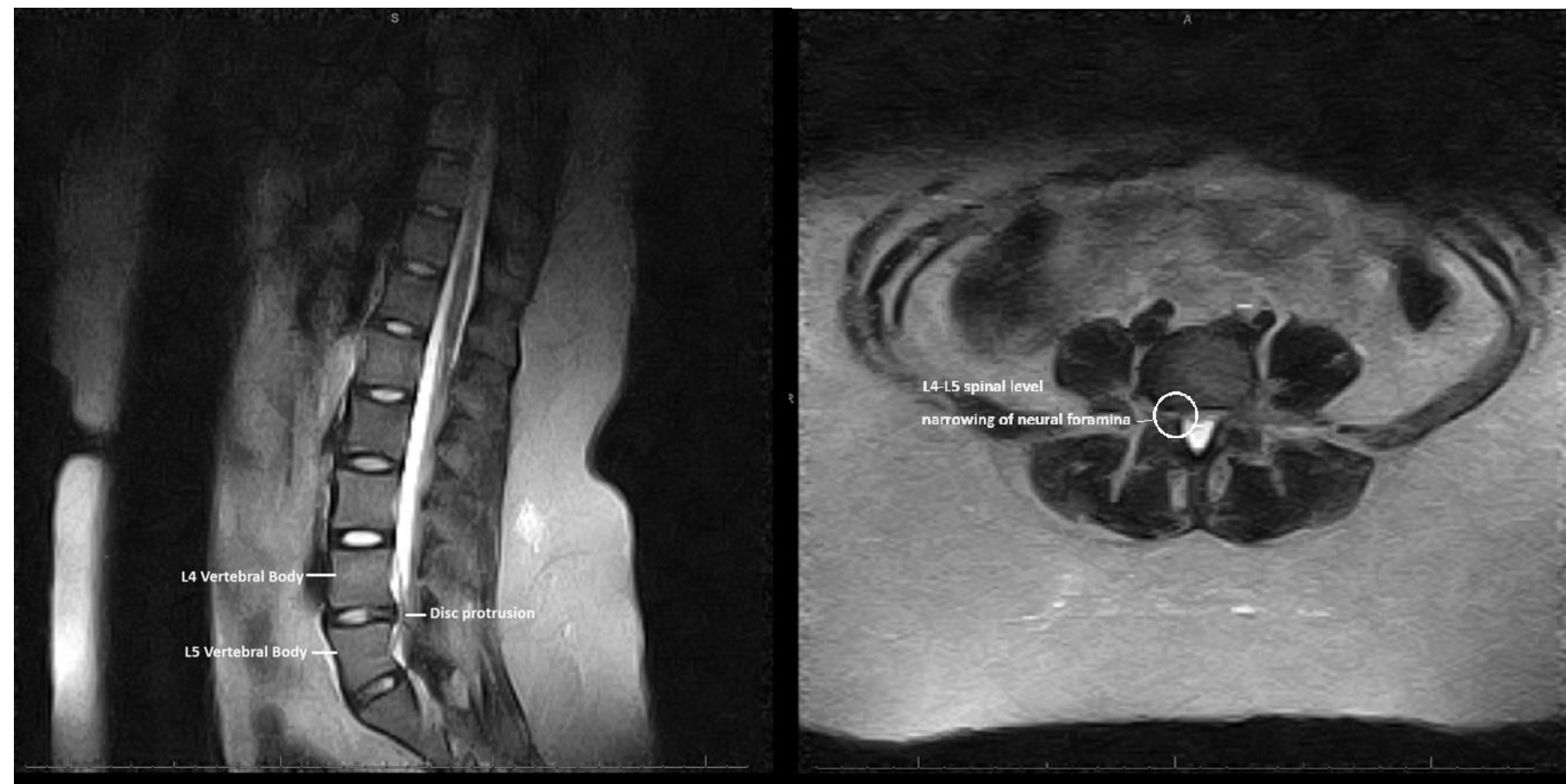
INTRODUCTION

Congenital myasthenic syndromes (CMS) are:

- inherited disorders of impaired neuromuscular transmission
- onset at birth-to-early childhood
- fatigable weakness of ocular or cranial muscles
- decremental electromyography (EMG) response

Childhood presentations can be complicated by neuropathic pain

Epidural steroid injections (ESI) involve the administration of medicines associated with increased risk of muscle weakness in CMS



Lumbar MRI: Sagittal (Left) and Axial (Right)

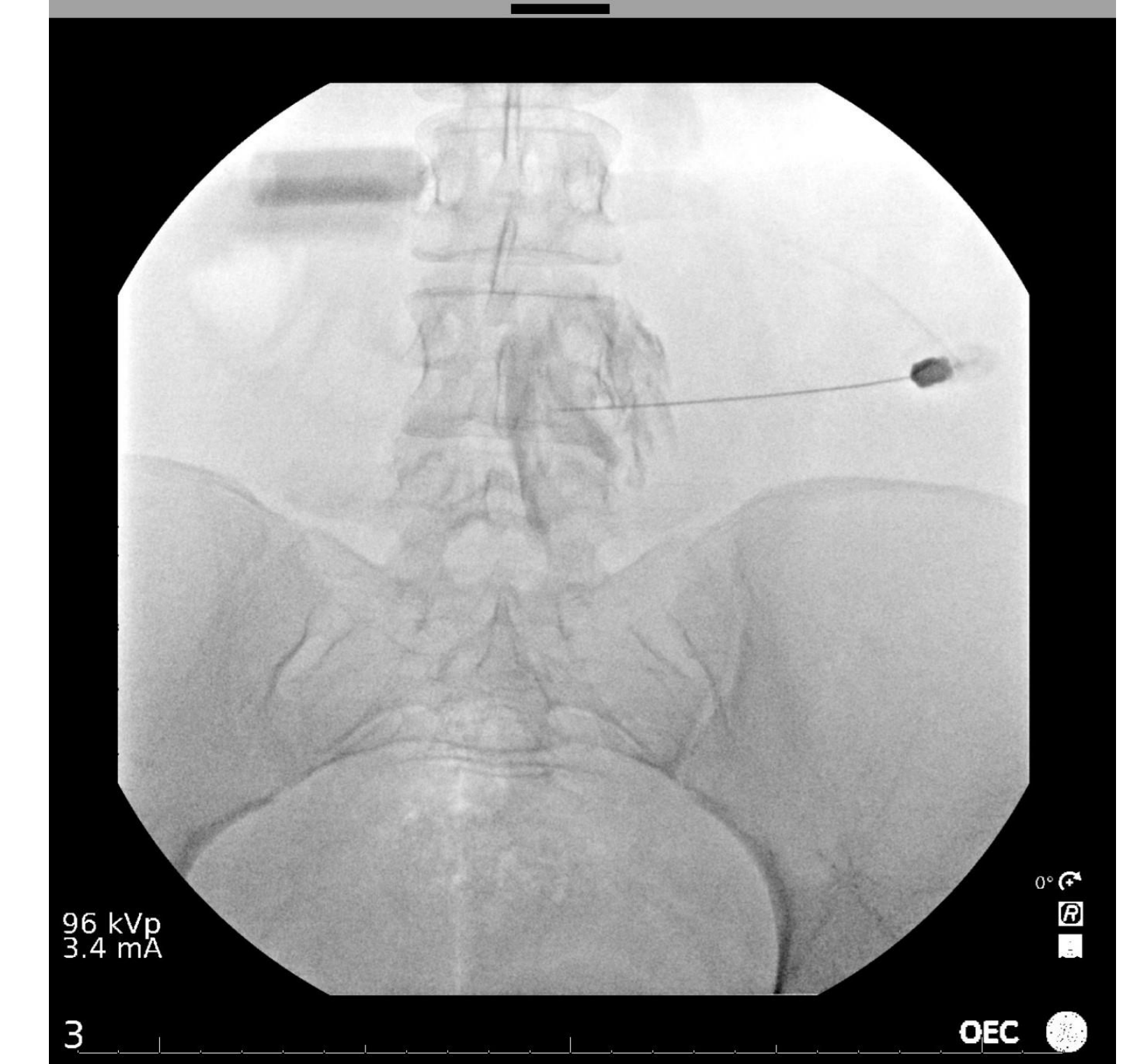
CASE PRESENTATION

- Patient: adolescent female with CMS (cholinergic receptor nicotinic epsilon [CHRNE] mutation, ptosis, ophthalmoplegia, fatigability, and lumbar disc disease)
- Her regimen of pyridostigmine 120mg 3 times daily PRN and 180mg nightly has helped her achieve remission
- Clinical course complicated by radiculopathy radiating from lower back to the right thigh; MRI with spinal canal stenosis due to broad based disc protrusion at the L4-5 level as above
- **Plan for right L4-L5 transforaminal epidural steroid injection**
- Meds to be administered intraprocedure: 10mg dexamethasone, 10mg of ropivacaine 0.2%, 630mg of iohexol
- 24-hour observation to monitor for respiratory and bulbar function

We performed a successful pediatric Epidural Steroid Injection in Congenital Myasthenic Syndrome

— while avoiding myasthenic exacerbation

— with careful endocrine consultation and overnight observation



L4-L5 Right ESI on Fluoroscopy

RESULTS

At 2-week follow up, patient had

- 100% pain reduction
- negative right slump test
- no subjective weakness to suggest CMS exacerbation

DISCUSSION

Various measures can be taken in the perioperative setting to allow for safe ESIs in this population. These measures include the avoidance, or minimizing dosages, of ester-type local anesthetics, neuromuscular blockers, corticosteroids, and iodinated contrast agents; planned observation for CMS exacerbation; and optimization of medical management with endocrinology's assistance

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Key Considerations for Pain Injection in Patient with Congenital Myasthenic Syndrome	
Pre-Op	Multidisciplinary Discussion with Endocrinology, Anesthesiology, and Family Continue Stable Administration of Acetylcholinesterase Inhibitors Coordinate for In-Hospital Admission for Respiratory Observation
Intra-Op	Consider Imaging without Using Iodinated Contrast Agent or Using Ultrasound Avoid Co-Administration of Local Anesthetic Agent with Paralytic Agent Minimize Dose of Corticosteroid Agent Minimize Dose of Iodinated Contrast Agent Minimize Dose of Local Anesthetic Agent
Post-Op	Mandatory Close Monitoring, Particularly of Respiratory and Bulbar Function