AMSER Case of the Month
November 2023

3-year-old female with unsteady gait

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Patient Presentation

HPI: 3 yo F presents with unsteady gait and refusal to walk long distances. Mother denies incontinence or other symptoms. History of seizures controlled with levetiracetam. Pt is a recent immigrant to the United States.

Developmental History:
• Speech delay, meeting all other milestones through 3 years old

Physical Exam:
• Erythematous patch with hypertrichosis on the lumbar spine
• Spontaneous extremity movement with normal muscle tone
• No other abnormalities

Labs:
• CBC and CMP within normal limits
What Imaging Should We Order?
### ACR Appropriateness Criteria

**Variant 4:** Child. Chronic progressive ataxia. Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tbody>
<tr>
<td>MRI head without and with IV contrast</td>
<td>Usually Appropriate</td>
<td>O</td>
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<tr>
<td>MRI head without IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>MRI complete spine without and with IV contrast</td>
<td>May Be Appropriate</td>
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<td>May Be Appropriate</td>
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<td>CT head without IV contrast</td>
<td>May Be Appropriate</td>
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<tr>
<td>MR spectroscopy head without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>O</td>
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<tr>
<td>MRA head and neck without IV contrast</td>
<td>Usually Not Appropriate</td>
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Lumbar spine MRI ordered
Findings (unlabeled)
Findings (labeled)

- Hypoplasia and incomplete fusion of the posterior vertebral elements of L3-L5
- Low-lying tethered cord at L4
- Type II diastematomyelia L1-L4 with intervening fibrous septum
- Small intradural fatty mass on the L posterior hemicord at L3
- Single dural sac without evidence of hydromyelia
Final Dx:

Type II Diastematomyelia
Diastematomyelia (Split cord malformation)

- **Definition**: A rare form of closed spinal dysraphism in which a sagittal cleft caused by abnormalities in vertebral body development and posterior element fusion causes a partial or complete division of the spinal cord into two hemicords.

- **Clinical features**:
  - Tethered cord syndrome
    - Motor weakness, sensory loss, reflex changes
    - Urinary incontinence, urinary tract infections
    - Foot deformities, scoliosis, kyphosis
  - Dermatologic lesions
    - hemangiomas, hypertrichosis, hyper/hypopigmentation
  - Spinal lipomas
  - Low back pain
# Imaging features and classification

<table>
<thead>
<tr>
<th>Type I</th>
<th>Type II</th>
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<tbody>
<tr>
<td>• Duplicated dural sac</td>
<td>• Single dural sac</td>
</tr>
<tr>
<td>• Midline osseous/cartilaginous spur</td>
<td>• No spur/septum</td>
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<tr>
<td>• Hydromyelia is common</td>
<td>• Hydromyelia possible</td>
</tr>
<tr>
<td>• Dermal manifestations</td>
<td>• Spina bifida possible</td>
</tr>
<tr>
<td>• Vertebral abnormalities</td>
<td>• Other vertebral abnormalities are less common</td>
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<tr>
<td>• Usually symptomatic with tethered cord syndrome and scoliosis</td>
<td>• Less symptomatic or asymptomatic</td>
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<td>• Cord may be incompletely divided</td>
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Case Discussion

• Some of the findings characteristic of a Type I Diastematomyelia are present, such as a midline septum, tethered cord, and cutaneous findings. However, the single dural sac containing both hemicords and absence of hydromyelia is suggestive of a **Type II Diastematomyelia**.

• Due to this patient's presentation with a symptomatic tethered cord, surgical release of the spinal cord is necessary to prevent progressive neurological decline.

• Surgical management is not always indicated if the patient does not present with a tethered cord or neurological symptoms. Even after surgical release, tethering may reoccur and subsequent revisions may be necessary.
References:


• Khoury C. Closed spinal dysraphism: Pathogenesis and types. UpToDate, Post, TW (Ed), UpToDate, Waltham, MA, 2021

• Khoury C. Closed spinal dysraphism: Clinical manifestations, diagnosis, and management. UpToDate, Post, TW (Ed), UpToDate, Waltham, MA, 2022

