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?

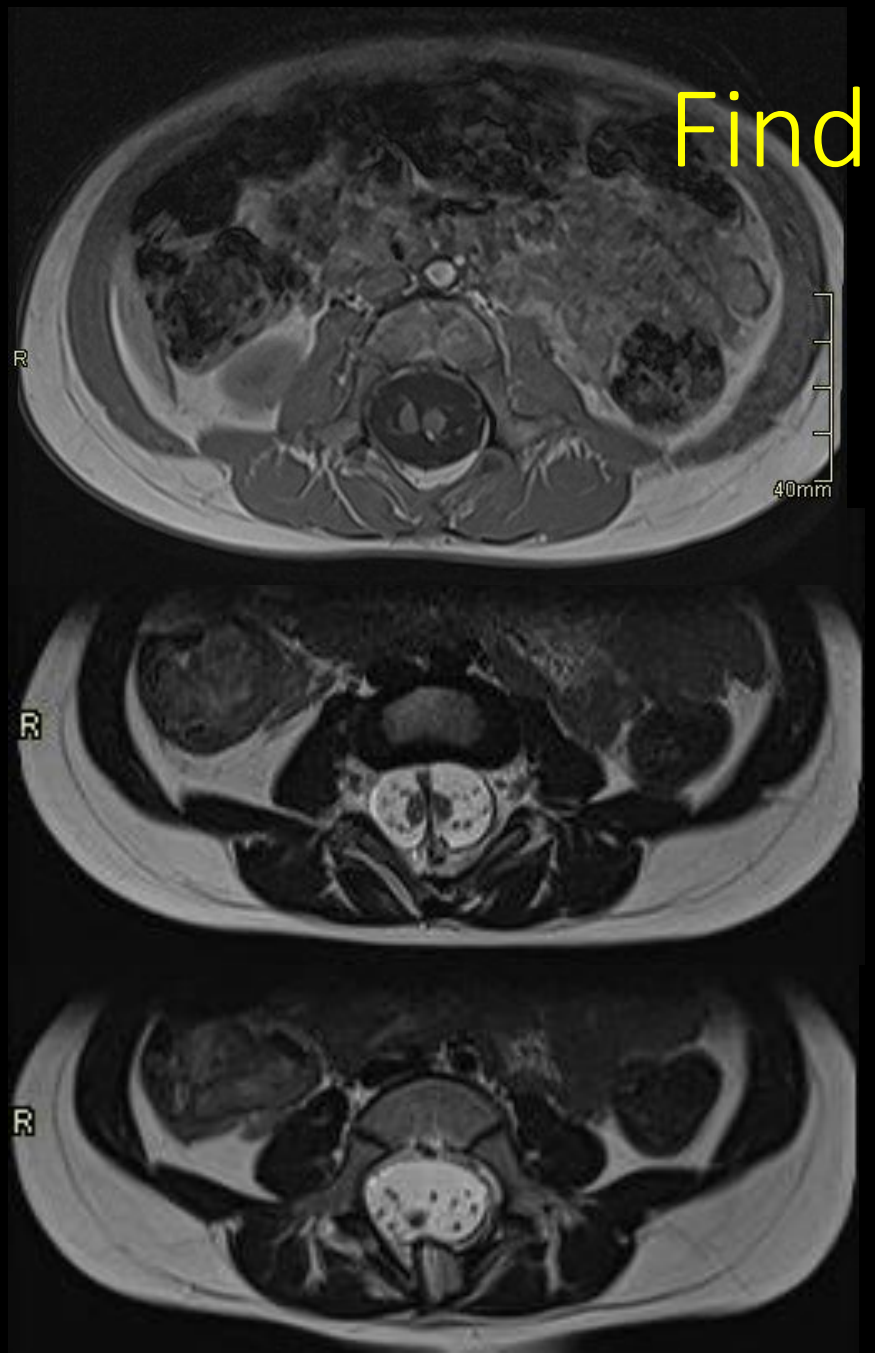
ACR Appropriateness Criteria

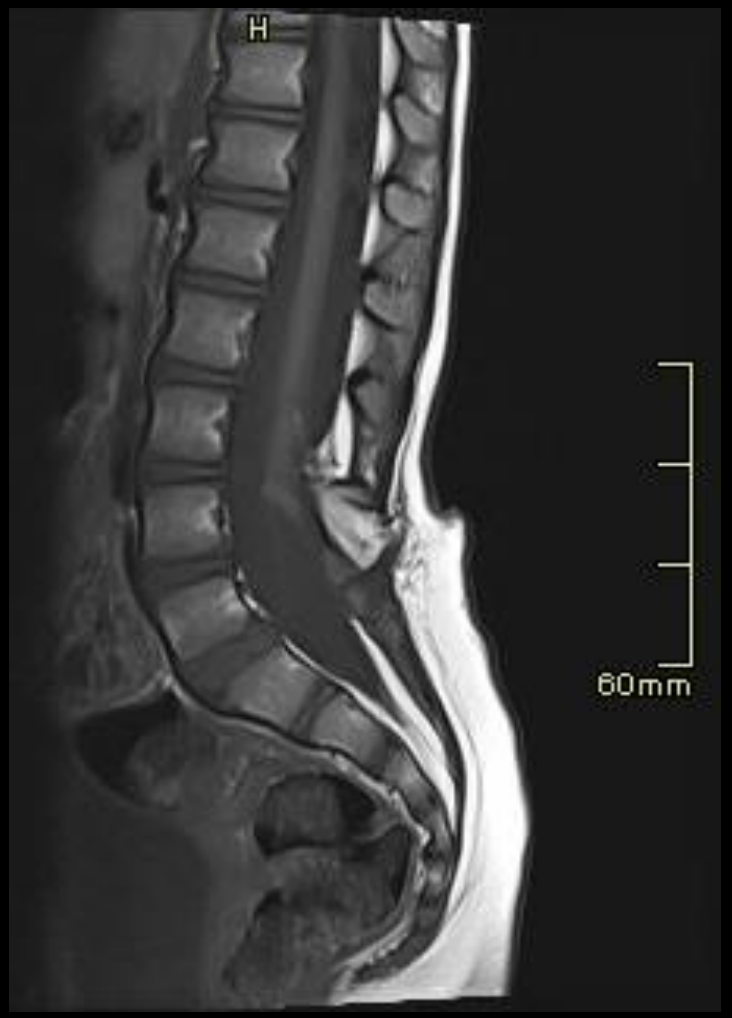
Variant 4: **Child. Chronic progressive ataxia. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head without and with IV contrast	Usually Appropriate	O
MRI head without IV contrast	Usually Appropriate	O
MRI complete spine without and with IV contrast	May Be Appropriate	O
MRI complete spine without IV contrast	May Be Appropriate	O
CT head without IV contrast	May Be Appropriate	☢☢☢
MR spectroscopy head without IV contrast	Usually Not Appropriate	O
MRA head and neck without IV contrast	Usually Not Appropriate	O

Lumbar spine MRI ordered

Findings (unlabeled)





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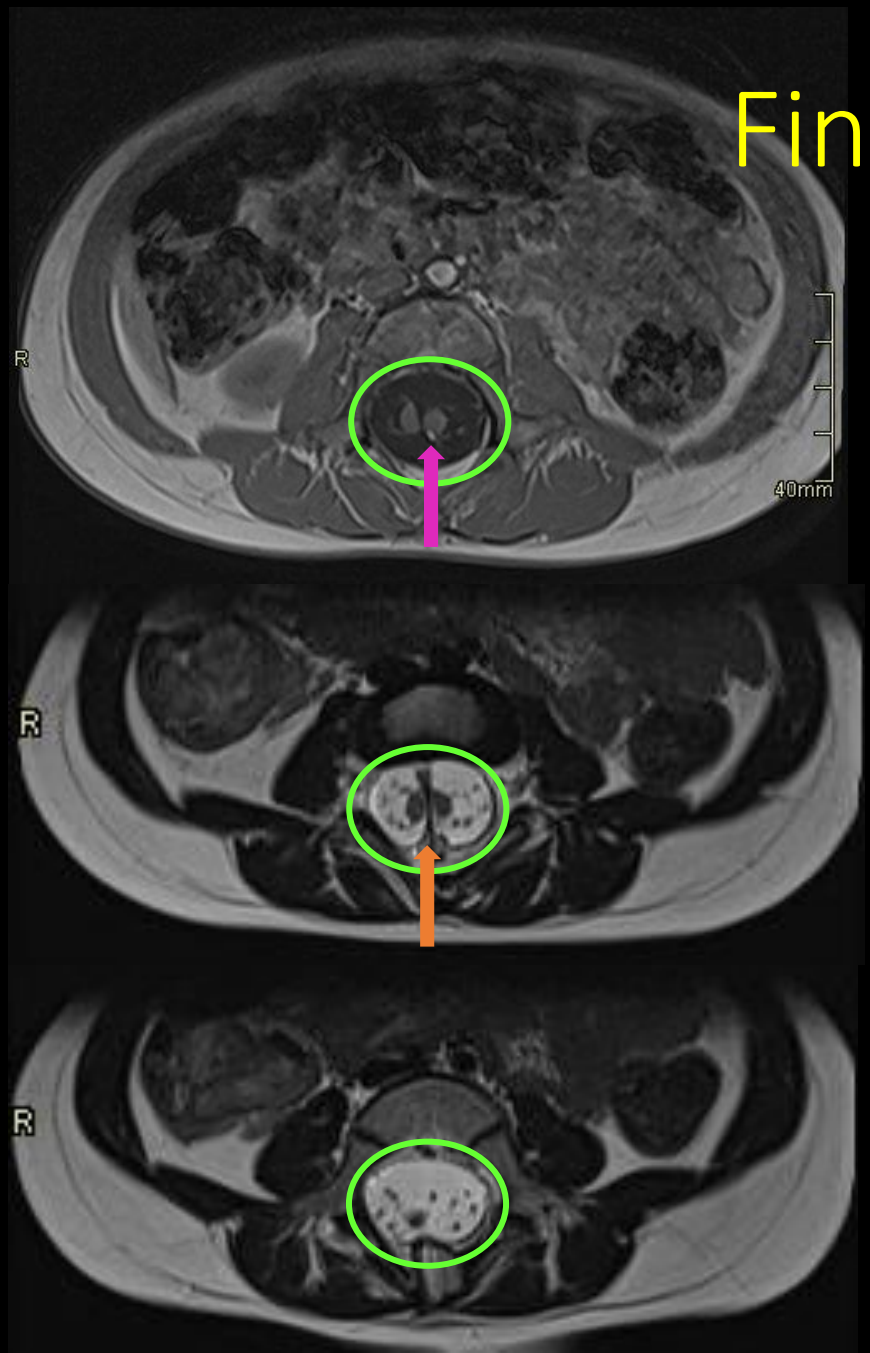
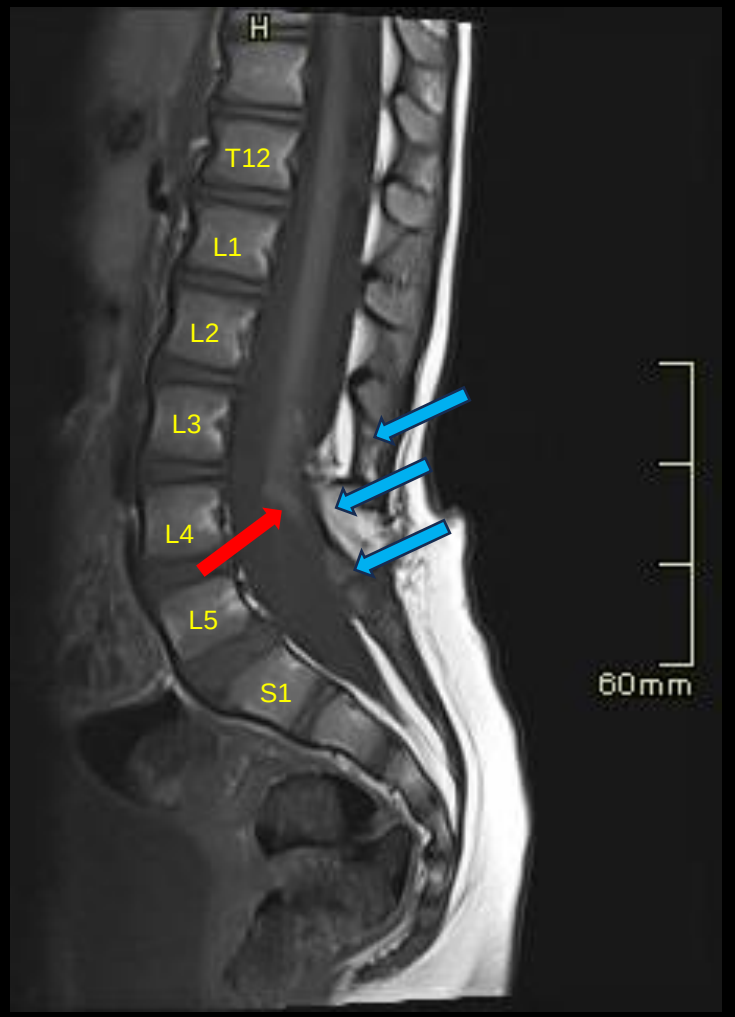


- Hypoplasia and incomplete fusion of the posterior vertebral elements of L3-L5
- Low-lying tethered cord at L4
- Type II diastematomyelia L1-L4 with intervening fibrous septum
- Small intradural fatty mass on the L posterior hemicord at L3
- Single dural sac without evidence of hydromyelia



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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

Type I

- Duplicated dural sac
- Midline osseous/cartilaginous spur
- Hydromyelia is common
- Dermal manifestations
- Vertebral abnormalities
- Usually symptomatic with tethered cord syndrome and scoliosis

Type II

- Single dural sac
- No spur/septum
- Hydromyelia possible
- Spina bifida possible
- Other vertebral abnormalities are less common
- Less symptomatic or asymptomatic
- Cord may be incompletely divided

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:

- Agarwalla, P. K., Dunn, I. F., Scott, R. M., & Smith, E. R. (2007). Tethered Cord Syndrome. Neurosurgery Clinics of North America, 18(3), 531-547. https://doi.org/10.1016/j.nec.2007.04.001
- 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
- Miller A, Guille J, Bowen J. Evaluation and Treatment of Diastematomyelia. The Journal of Bone & Joint Surgery. 1993;75(9):1308-17. doi:10.2106/00004623-199309000-00006 – Pubmed
- Sami H, Ross E, Walter M, Goli S. Split Spinal Cord (Diastematomyelia). Neurology. 2003;60(3):491. doi:10.1212/01.wnl.0000044730.82558.ac – Pubmed
- Tuite, G. F., Thompson, N. P., Austin, P. F., & Bauer, S. B. (2018). Evaluation and management of tethered cord syndrome in occult spinal dysraphism: Recommendations from the international children's continence society. Neurourology and Urodynamics, 37(3), 890-903. https://doi.org/10.1002/nau.23382