

Constipation in an Infant:

The role of imaging in evaluating for
Hirschsprung disease

Amber A. Garza

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DII RAD 4001 elective

Reviewed By: M. Kumaravel MD



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The University of Texas
Health Science Center at Houston

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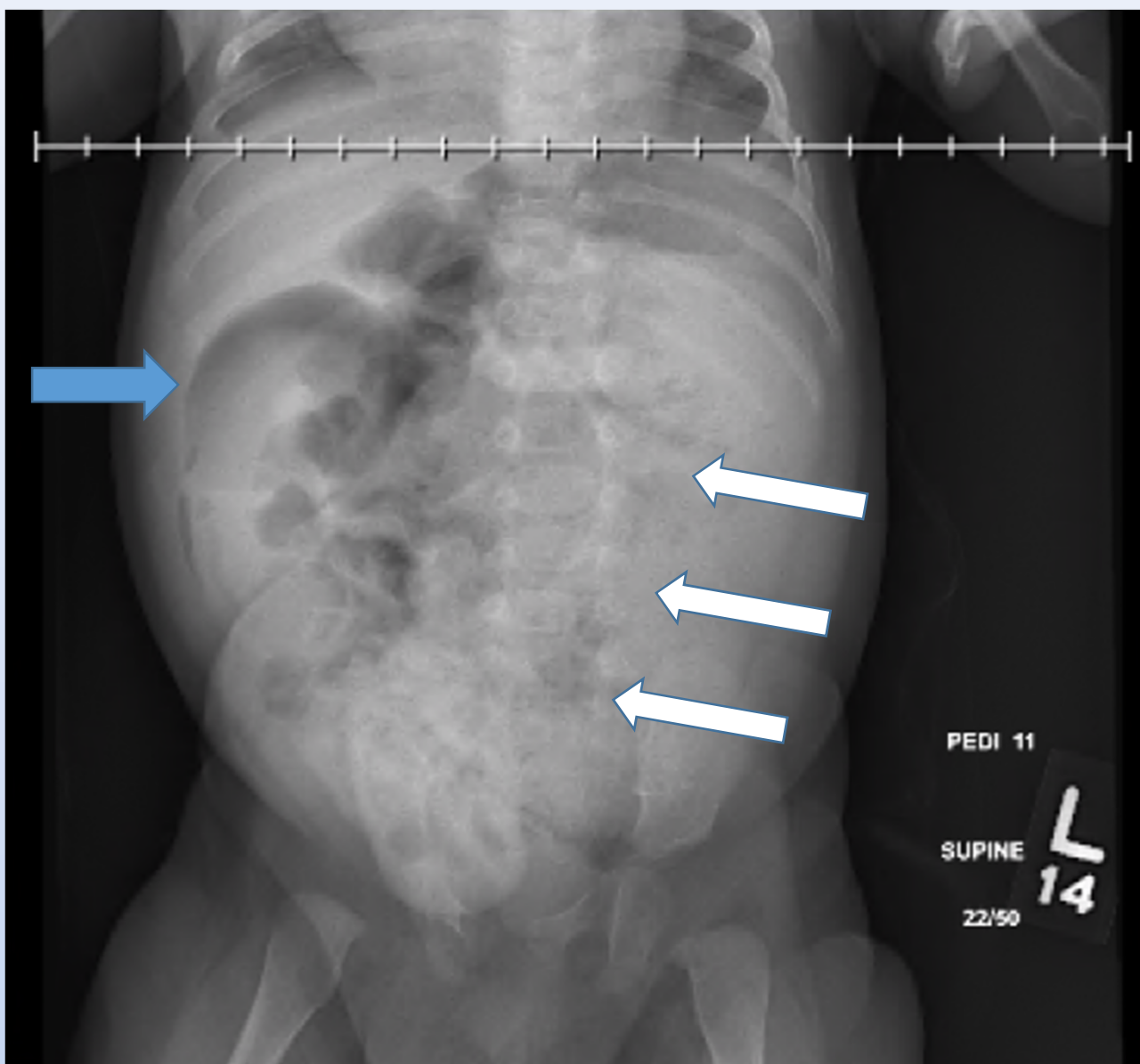
Medical School

History

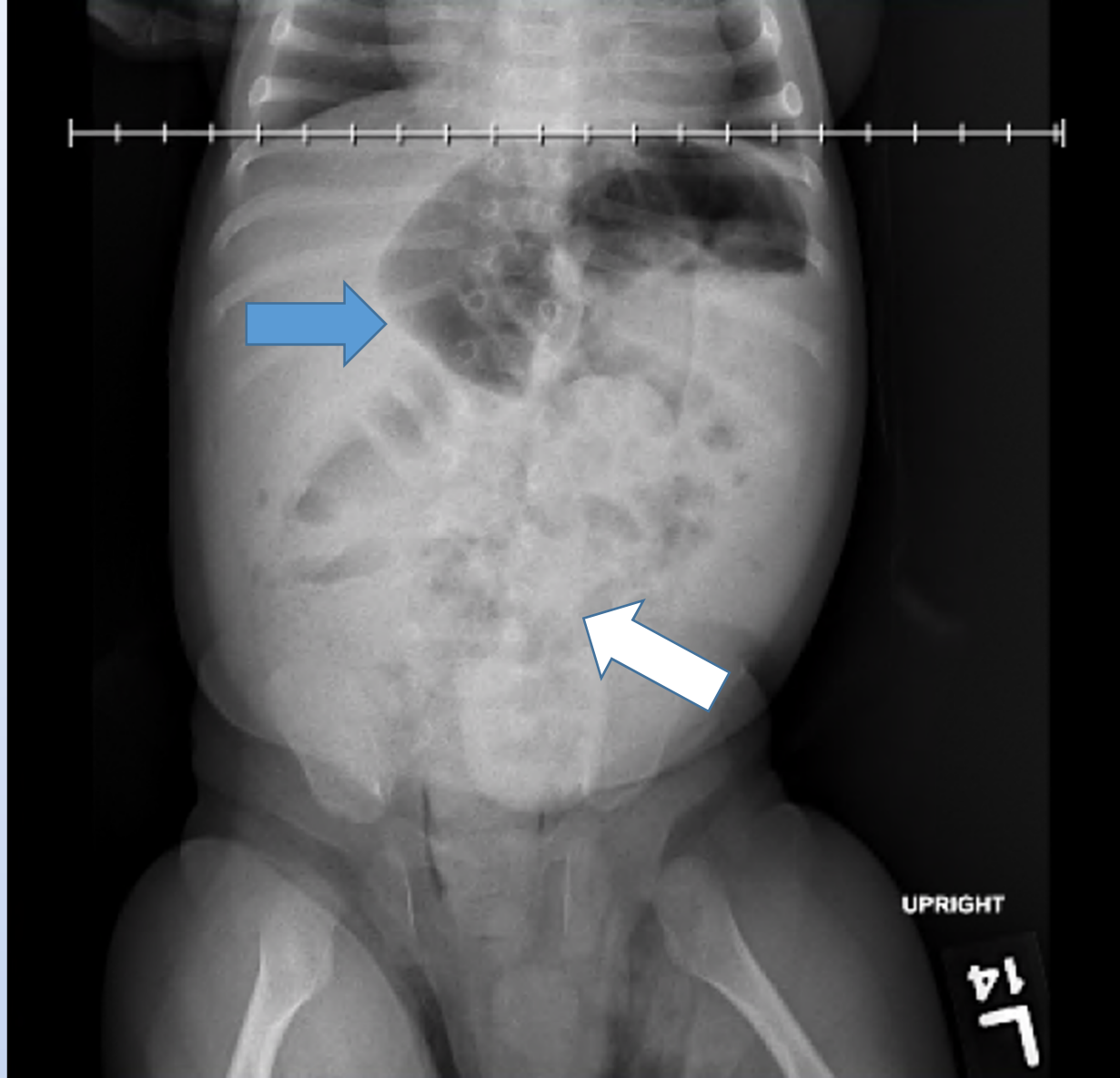
- 2 m.o. AA Male with worsening constipation x 2 weeks
- Intermittent constipation since birth
- Perinatal course: failure to pass meconium
- Abdominal exam normal except minimal distention

Differential Diagnosis

- Idiopathic constipation
- Hirschsprung disease
- Congenital hypothyroidism normal TSH and T4
- Cystic fibrosis normal initial newborn screen



Abdominal X-ray - supine



Abdominal X-ray - upright

Barium
Enema



Hirschsprung Disease

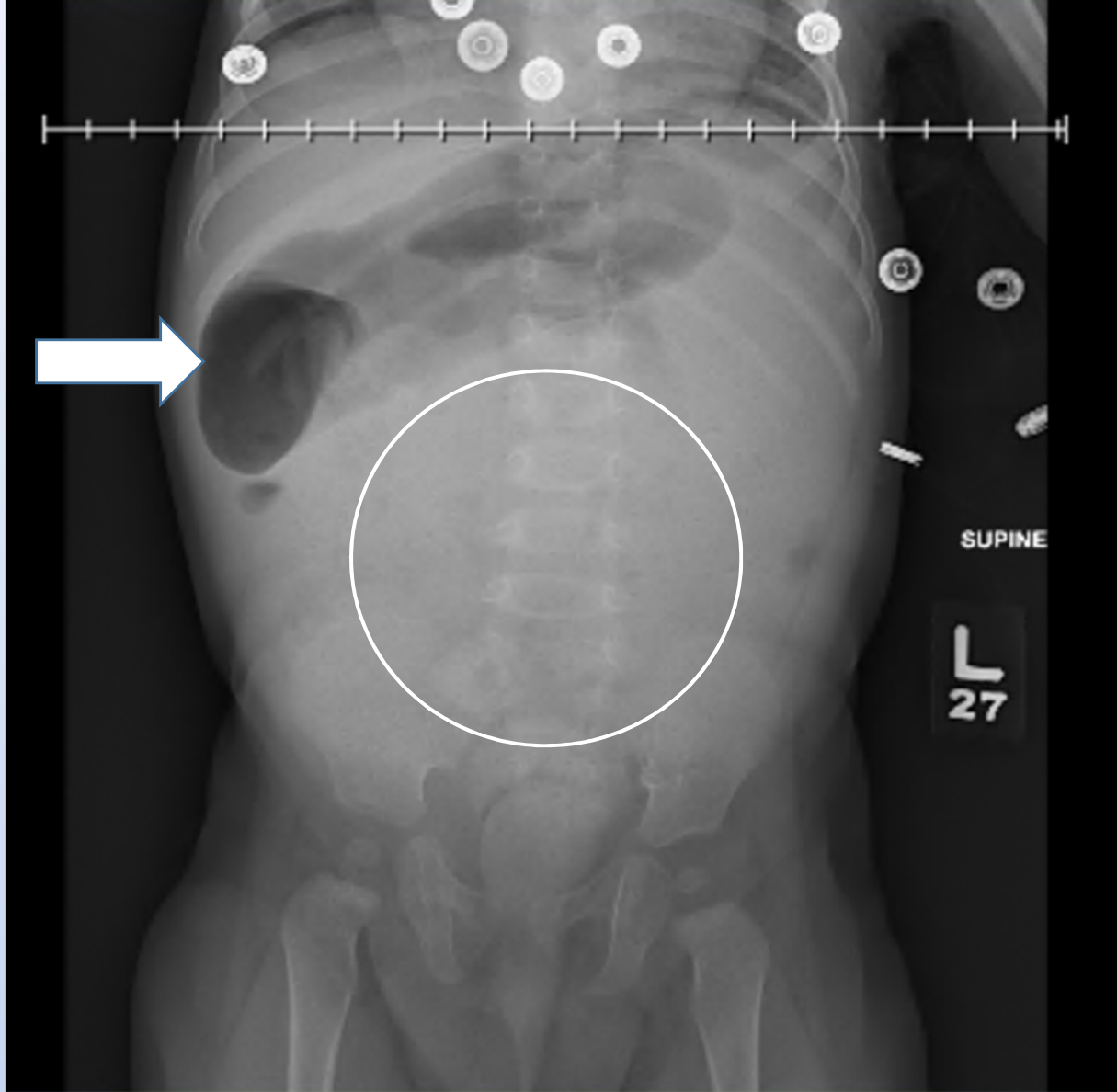
- Failure of cranio-caudal migration or neural crest cells resulting in absence of motor nerves in bowel segments
- ~ 80% of cases involve short-segment disease
- Typically diagnosed in neonatal period
- Initial patient evaluation with abdominal radiograph and contrast enema
- Gold standard for diagnosis is rectal biopsy

Findings

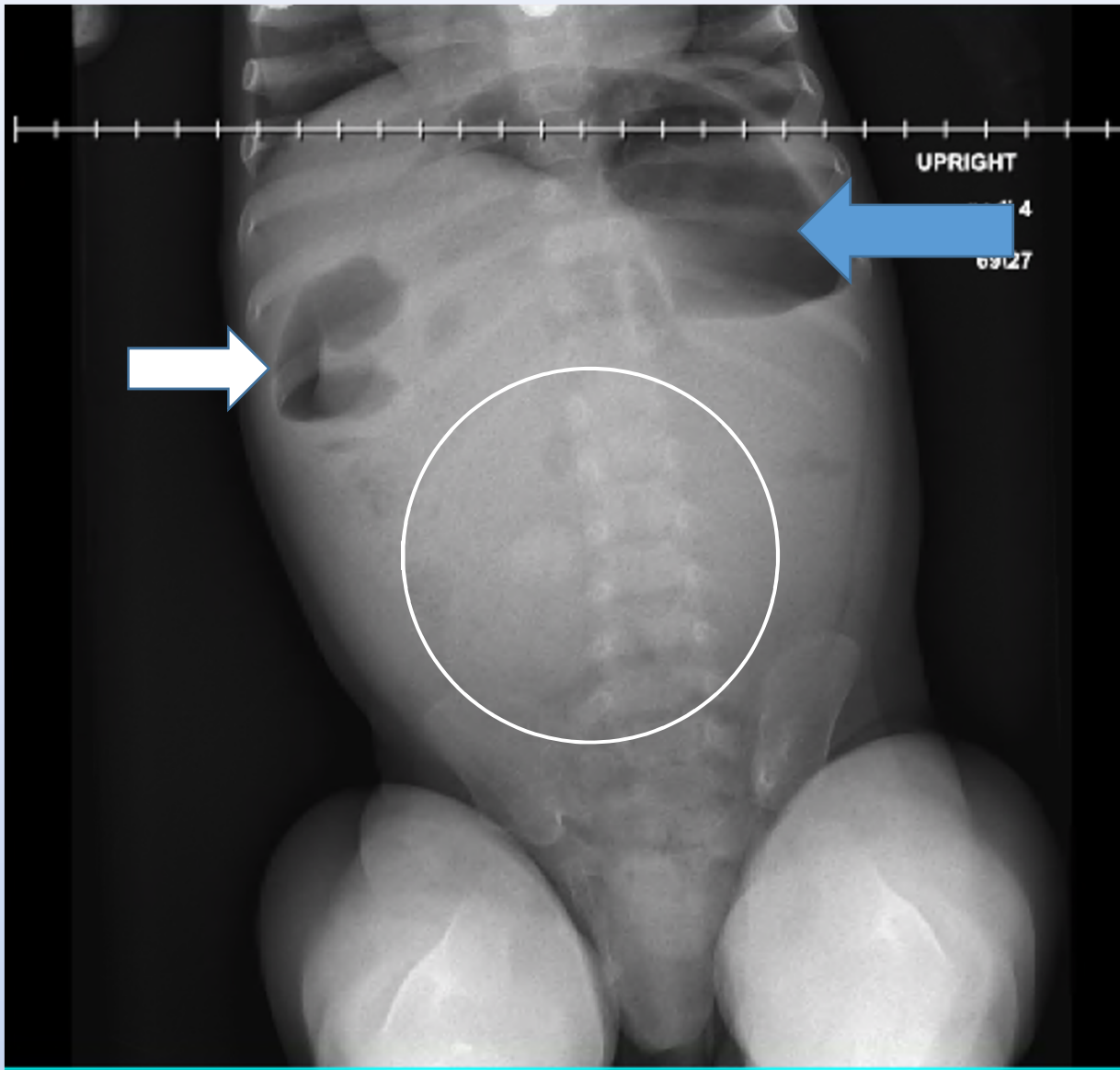
- Rectal suction biopsy – indeterminate results
- Parents declined repeat biopsy due to patient's clinical improvement after receiving laxative
- Patient discharged with outpatient follow-up

6 months later...

- Patient (now 8 m.o.) again presented to ED
- Severe constipation and abdominal distention x 4 days
- Non-bloody, non-bilious vomiting x 1 day



Abdominal X-ray - supine



Abdominal X-ray - upright

Final diagnosis

- Open rectal biopsy – absence of ganglion cells
 - **Diagnosis of Hirschsprung disease confirmed**
- Transmural sigmoid biopsy – ganglion cells present
- Patient underwent laparoscopic leveling colostomy

Were the Appropriate Images Ordered?

- ACR-SPR indications for abdominal radiograph include:
 - constipation
 - evaluation of congenital GI abnormalities
- ACR-SPR indications for contrast enema include:
 - Investigation of potential causes of constipation
 - Suspected congenital disease of the colon

References

- Badner, JA, *et al.* (1990). A genetic study of Hirschsprung disease. *American Journal of Human Genetics*, 46(3).
- Herring, W. (2016). *Learning radiology: recognizing the basics*. Philadelphia, PA: Elsevier.
- Lorijn, FD, *et al.* (2005). Diagnosis of Hirschsprungs disease: a prospective, comparative accuracy study of common tests. *The Journal of Pediatrics*, 146(6).
- Putnam, LR, *et al.* (2015). The utility of the contrast enema in neonates with suspected Hirschsprung disease. *Journal of Pediatric Surgery*, 50(6).
- The American College of Radiology – ACR-SPR practice parameters