

64th ANNUAL DIAGNOSTIC SLIDE SESSION 2023

CASE 2023 [#5]

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Clinical History:

This 3 1/2-year-old girl had a history of Hirschsprung Disease, diagnosed soon after birth (born preterm at 26 gestational weeks). She had repeated enterocolitis alternating with chronic constipation, treated by her parents at home with over-the-counter enema preparations. She had undergone a “pull-through” procedure as well as placement of an intraosseus line for treatment of dehydration (6 and 2 months prior to death, respectively). The latter procedure was complicated by osteomyelitis, detected and treated 2 weeks prior to death. Four days prior to death, the child had continued vomiting and poor fluid intake. Her parents administered saline enemas the night prior to death. After the parents found her lethargic, she was brought into hospital and died despite resuscitation efforts.

Autopsy findings: Hirschsprung Disease:
- Status post “pull-through” procedure (intact)
- Dilated large colon filled with liquid stool
- Full-length, full-thickness colitis

Material submitted: 1. Gross image of mid-pons
2. H&E- and LFB/PAS-stained sections of pons

Points for discussion: 1. Diagnosis
2. Pathogenesis