## 51. CASE REPORT: ADULT HIRSCHSPRUNG DISEASE

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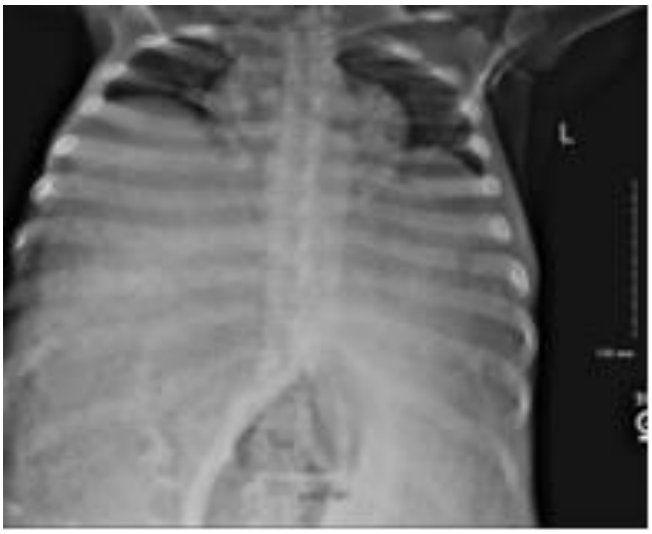
**BACKGROUND**: Hirschsprung's disease (HD) remains the most common condition to cause low functional intestinal obstruction in children. It is caused by the failed migration of colonic ganglion cells during gestation. The aganglionic sections of the colon are subsequently unable to relax, halting digestive consumption within the gastrointestinal tract resulting in prolonged constipation symptoms. Although it is primarily diagnosed in infants and children, there are cases of diagnosis not being made until adulthood. **CASE**: A 19-year-old African American male with a history of asthma, chronic constipation, and medication noncompliance that presented to a local free-standing emergency department with constipation and severe abdominal distension. Per the patient's mother, AB was also previously evaluated by a gastroenterologist at a children's hospital and no findings that they are aware of were discovered. His vitals on presentation were temperature 36.8°C, heart rate 98, blood pressure 130/99, respiratory rate 32, oxygen saturation of 95% on room air. On physical exam he had diminished breath sounds at his expected lung bases and his abdomen measured 112 cm at its largest circumference. At the emergency department a KUB was obtained and showed functional obstruction diffuse fecal loading throughout the colon. AB was subsequently admitted to the hospital. He was started on oral polyethylene glycol, dulcolax, and senokot with minimal response. A pulsated irrigation evacuation (PIE) procedure was performed on 2/10 with significant stool debulking, but his abdominal circumference remained unchanged. He was started on polyethylene glycol via NG tube and underwent manual disimpaction on 2/11 with large volume stool evacuation. His team of physicians decided that he needed a colon biopsy to confirm the suspicion of Hirschsprung's. After discussion with the patient, he deferred colonoscopy and biopsy until outpatient. After continued improvement in his stool output and abdominal circumference to roughly 65 cm, he was transitioned to oral polyethylene glycol and ultimately discharged on polyethylene glycol with follow up appointment scheduled to establish care with a primary care provider and a local gastroenterologist. The patient underwent an outpatient colon/rectum biopsy approximately one month after discharge. The biopsy was sent to pathology and was reported as follows: "Sections from the rectal biopsy specimen show well-oriented fragments of anorectal mucosa consistent with segments of squamous mucosa with adjacent colonic type columnar mucosa. Findings are consistent with sampling at the anal transitional zone. There is no significant inflammatory infiltrate. No ganglion cells are identified. Occasional lymphoid aggregates are noted. These findings are consistent with Hirschsprung's disease". **CONCLUSION**: Hirschsprung's induced constipation can lead to severe consequences and should be promptly treated. Since there are no guidelines for severe constipation in HD patients, we recommend a stepwise approach starting with osmotic laxatives and stool softeners in conjunction with manual disimpaction. For refractory cases we recommend PIE, in areas where this procedure is available. Although this stepwise approach to severe constipation avoidance of life-threatening complications, definitive treatment is still colectomy once the bowel is evacuated; if the patient is agreeable to an invasive procedure.

**Figures:** Radiograph of the patient read as the following. the lungs are hypo-aerated. There is severe diffuse fecal loading throughout the colon. There is mild bowel distention in the mid-abdomen, likely from functional obstruction. No obvious free air or pneumatosis.



**Key Words**: Hirschsprung's disease, chronic constipation, slow transit time, acute severe constipation, adult Hirschsprung's disease.