



Shriners Hospitals
for Children®— Northern California

FAQ from the 12/7/2020 Webinar

Virtual Physician CME Program

Hirschsprung Disease – What Every Pediatric Provider Needs to Know

December 7, 2020

1 Diagnostics & Work Up

Q1: If I have a patient diagnosed with Hirschsprung Disease who has no current reported issues, should h/she still see a colorectal provider?

A1: Yes, we recommend that any child diagnosed with Hirschsprung Disease be followed by a colorectal surgeon until they reach adulthood. Having a multidisciplinary team follow them even if they are not presenting symptoms is important to anticipate and address long-term issues that children with Hirschsprung Disease can have. If a patient has moved and has not identified a local specialist for colorectal care, we strongly advise and help families select a specialist and establish a care plan.

Q2: At what point would you refer a patient with idiopathic constipation?

A2: History is very important when considering referring to specialty care for idiopathic constipation. Any patient who has struggled with constipation since birth, has failure to thrive, abdominal distention or blood/mucous in their stool should be referred. Additionally, school aged children with encopresis and those with medically refractory idiopathic constipation should be referred.

Q3: How accurate are the screening tools when diagnosing HD Disease?

A3: A combination of dedicated colorectal history, physical exam including rectal exam, an abdominal x-ray, and a contrast study have high sensitivity for Hirschsprung Disease. However, none of these tests are perfect. While anorectal manometry can be diagnostic in an older infant or child, if the patient has a very distended rectum then we may not be able to rule out Hirschsprung Disease with that procedure and the patient

may need a rectal biopsy. Rectal biopsy is the gold standard test and will definitively diagnose Hirschsprung Disease.

Q4: Describe the frequency of late diagnosis, misdiagnosis and impact/implications

A4: Fortunately late and/or misdiagnosis is rare but implications can be severe. Outcome from a late diagnosis is dependent on the length of their diseased colon and time it takes to identify a correct diagnosis.

2 Surgery

Q5: How long would you expect a child with Hirschsprung Disease to have an ostomy?

A5: The length of time a child with Hirschsprung Disease depends on why the ostomy was created in the first place. Some infants may have an ostomy for less than a year; some children may need an ostomy for several years. Some children may never have an ostomy.

Q6: Of the pediatric patients referred to you who have had previous surgeries, what percent require a correction and why?

A6: We are a referral center for patients with complex colorectal disorders. Approximately 20% of patients need some sort of surgical intervention at Shriners. Nationally, the percentage is low single digits.

3 Case Scenarios, Rectal Irrigations, HAEC, Post-Surgery, Motility Testing

Q7: Which kids with Hirschsprung Disease would most benefit from motility testing?

A7: Children with Hirschsprung Disease who are struggling with obstructive symptoms, soiling, or repeated episodes of enterocolitis may benefit from motility testing. We will first evaluate for anatomic and pathological causes of these problems. If a child is still having symptoms after these areas are evaluated, then motility testing may help guide treatment options.

Q8: Are there dietary restrictions for kids with Hirschsprung Disease?

A8: Dietary recommendations are highly individualized to the specific child depending on their unique case. We are fortunate to have a registered dietician as part of our multidisciplinary team who can help guide and individualize care.

Q9: What kind of ongoing care throughout the patient's life is needed to minimize the risk and complications of Hirschsprung Disease?

A9: Children with Hirschsprung Disease, even those who have never had problems stooling after their initial surgeries, require long-term follow-up with a specialist experienced in treating Hirschsprung Disease. Children may develop issues with obstruction, soiling, or enterocolitis later in childhood. These problems are often not appreciated until a child starts to potty train. In addition, we are learning more and more from adolescents and adults about the long-term impact of this disease throughout life with a specialist experienced in treating Hirschsprung Disease.

Q10: How do I get a formal consult?

A10: Please fax your referral to 916-453-2191, email it to referrals.ncal@shrinenet.org, or call 916-453-2191. You can find our referral form and more information at <http://shrinerschildren.org/referrals>.