

Hirschsprung's disease: nursing management and evidence-based care



**ASSIGN
BUSTER**

Hirschsprung's disease also known as congenital megacolon or aganglionic megacolon, is a rare birth defect of the colon in which the colon is missing vital nerve cells that are required to initiate peristalsis. It occurs in approximately 1 in 5, 000 live births and is predominantly found in male children. While Hirschsprung's disease is thought to be a genetic condition, recent studies have found another possible cause may be linked to perinatal selective serotonin reuptake inhibitor use with a 95% increase being found in women whom took a minimum of two SSRI prescriptions during the first trimester.

Nursing management

There is currently no education or preventative measures that can be taken to prevent a child from developing or getting Hirschsprung's disease. However, one of the most common complications associated with Hirschsprung's disease is enterocolitis and is responsible for more than half of the deaths associated with Hirschsprung's disease. The best way to prevent Hirschsprung-Associated Enterocolitis is early diagnosis and treatment. Delayed diagnosis within the prenatal period, increases the likelihood of a child developing Hirschsprung-Associated Enterocolitis by approximately 18-50%. (1) Due to this a nurse must be aware of the signs and symptoms of Hirschsprung-Associated Enterocolitis which include fever, abdominal distention, explosive watery diarrhea, foul-smelling stools, pain, rectal bleeding, and poor appetite and can occur prior to or after treatment for Hirschsprung's disease. Infants and young children require more frequent assessments as they have a higher risk of developing enterocolitis due to their weakened immune systems. Preoperative preventative measures

typically include prophylactic routine rectal washout treatments and/or probiotics such as lactobacillus, bifidobacterium, and saccharomyces. (1) Therapy required during a flare-up of enterocolitis include broad-spectrum antibiotics and intravenous fluid resuscitation.

During a washout procedure, the infant should be placed on their back with legs in a frog-like position. Calming measures such as swaddling of the arms, comfort, and play therapy should be used at this time. Sucrose may also be given prior to and during the procedure if needed.

Nursing care for Hirschsprung's disease is primarily depends on the age of the child and treatment required. During the neonatal period, diagnosis support is the most immediate concern. A nurse may help a parent adjust to the new diagnosis, assist with parent-child bonding, prepare and support the parents during the medical-surgical decision making process, and teach the parents about how to perform post-operative care.

A nurse is also responsible for preparing the child for surgery. This includes making sure the patient is stable enough for surgery and if not initiating systematic treatments which may include; enemas, and/or " low fiber, high calorie, high-protein diets". (2) Saline enemas for bowel emptying may also be required for older children as well as prophylactic antibiotic therapy.

When the child is to undergo an emergency operation, the nurse should monitor the patient's vital signs for signs of shock, fluid and electrolytes, and for signs of bowel perforation. A child should also be prepared psychologically for the procedure in age appropriate detail.

Immediate postoperative care includes monitoring of vital signs, maintenance of fluid and electrolyte balance, and pain management. During the postoperative period, the nurse should include the child and their parents in care. This includes how to monitor for signs of wound infection and irregular bowel movement passage. For children with a new colostomy, the postoperative period is the best time to teach both an older child and their parents how to correctly perform colostomy and skin care.

Evidenced-Based Care

The golden standard diagnostic method for Hirschsprung's disease is a full thickness bowel biopsy as they are the only known method for confirming the absence of ganglion cells within the colon and/or rectum. However, this procedure carries many associated risks such as sepsis, bleeding, perforation, and anesthesia related problems thus rectal suction biopsies are a commonly used alternative. A rectal suction biopsy, like the full thickness bowel biopsy, extracts a small tissue sample from the rectum which is then studied for the presence or absence of ganglionic nerve cells. In neonates, however, this may be difficult due to their immature ganglion cells not having a sufficient amount of cytoplasm. This can cause them to resemble lymphocytes, stromal cells, or endothelial cells thus making it difficult to identify the ganglion cells. As a result, acetylcholinesterase histochemistry (AChE) is used to assist in obtaining an accurate diagnosis. However, this test can be difficult to interpret which in turn, can lead to a false positive or negative result.

In search of a another diagnostic method, Dr. I. Barshack conducted a study using calretinin immunohistochemical staining and found that while a ganglionic colon with hirschsprung's disease and normal colon expressed calretinin, an aganglionic hirschsprung's colon did not. This has concluded that the calretinin immunohistochemical staining test can be useful in diagnosing aganglionic hirschsprung's disease. A study conducted to compare the calretinin immunohistochemical staining and the acetylcholinesterase histochemistry (AChE) showed a large difference between the two tests but no difference in sensitivity. The acetylcholinesterase histochemistry (AChE) test showed a 93. 5% sensitivity, 100% specificity, and 97. 8% accuracy whereas the calretinin immunohistochemical staining showed a 90. 5% sensitivity, 85. 2% accuracy and a 100% sensitivity. (International Journal of Surgical Pathology) This concluded that combined both methods could be extremely useful in diagnosing hirschsprung's disease.

In cases where an immediate diagnosis is needed however, the above diagnostic methods are not feasible as they can take up to 24 hours to complete. Due to this, the Modified Rapid AChE method was introduced. The Modified Rapid AChE Method was derived from utilizing the Kobayashi rapid AChE method which took only 10 minutes to complete and was performed by utilizing H₂O₂ as a way to speed up the copper ferrocyanide oxidation reaction. However, when performed it showed a weakly stained fibers thus leading to an inconclusive diagnosis. In taking from Kobayashi's method scientists were able to modify the stains used and make them stronger and more permanent which resulted in accurate diagnoses Of the 114 cases

studied using the Modified Rapid AChE Method, no false-positives or negatives occurred as compared to the acetylcholinesterase histochemistry (AChE) and calretinin immunohistochemical staining. This method can be highly advantageous when in need of an immediate diagnosis especially within the intraoperative setting. The only disadvantage found when using this method is the use of potentially toxic chemicals however, these hazards can be avoided if a proper chemical hood is utilized.

Currently, a variety of surgical procedures exist to treat children with Hirschsprung's Disease. The most common type of procedure performed is the laparoscopic pull through procedure, mainly the transanal endorectal pull through procedure, the Soave, the Duhamel, and the Swenson. However, many of these procedures are associated with frequent complications and often require a reoperation. For this reason, a new technique called the Laparoscopic-Assisted Heart Shaped Anastomosis was developed in hopes it would decrease the incidence of postoperative constipation and soiling which occurs approximately 10-20% of the time when the laparoscopic pull through method is used. During the Laparoscopic-Assisted Heart Shaped Anastomosis procedure almost all of the internal sphincter is left in place, as opposed to nearly half during a laparoscopic pull through procedure, and the left transverse colon and splenic fixtures are mobilized, allowing the colon to be pulled through without tension. As a result, postoperative complication rates dropped from over 25% for early postoperative complications to 7.8% and from about 40% for late complications to 11.4%. Currently, this procedure is being widely used in a variety of Chinese medical centers as it is simply to

perform and learn, eliminates the need for a colostomy or urethral catheter, and can be performed with minimal pelvic cavity mobilization.

In conclusion, while Hirschsprung's disease is curable, if left untreated it can result in death. However, many of the procedures used to treat Hirschsprung's disease often lead to postoperative complications. Long-term these complications can lead to a poor quality of life and severely affect the child psychological health. While other treatments are currently being studied, such as neuronal cell transplantation, it may still be a few years until these treatments are brought into practice. Until then, it is important that healthcare providers remain vigilant to the signs and symptoms related to these complications and intervene when appropriate. Supportive care may also be beneficial for these children, especially when self-esteem is affected.

Reference Page

1. Frykman, P. K., & Short, S. S. (2012). Hirschsprung-associated enterocolitis: Prevention and therapy. *Seminars in Pediatric Surgery*, 21 (4), 328-335. doi: 10. 1053/j. sempedsurg. 2012. 07. 007
2. The Royal Children's Hospital Melbourne. (n. d.). Retrieved April 14, 2019, from https://www.rch.org.au/rchcpg/hospital_clinical_guideline_index/Bowel_Washout_Rectal/
3. Perry, S. E., Hockenberry, M. J., Lowdermilk, D. L., Wilson, D., Cashion, K., Rodgers, C. C., & Alden, K. R. (2018). Chapter 41: Health Problems of Children. In *Maternal Child Nursing Care* (6th ed., pp. 1183-1185). St. Louis, MO: Elsevier.

4. Jeong, H., Jung, H. R., Hwang, I., Kwon, S. Y., Choe, M., Kang, Y. N., . . . Kim, S. P. (2018). Diagnostic Accuracy of Combined Acetylcholinesterase Histochemistry and Calretinin Immunohistochemistry of Rectal Biopsy Specimens in Hirschsprung's Disease. *International Journal of Surgical Pathology*, *26* (6), 507-513. doi: 10. 1177/1066896918761235
5. Naguib, M. M., Robinson, H., Shoffeitt, C., Howe, H., Metry, D., & Shehata, B. M. (2016). Modified Rapid AChE Method (MRAM) for Hirschsprung Disease Diagnosis: A Journey from Meier-Ruge Until Now. *Fetal and Pediatric Pathology*, *35* (6), 399-409. doi: 10. 1080/15513815. 2016. 1214200
6. Jiao, C., Yu, D., Li, D., Wang, G., & Feng, J. (2018). A Long-Term Follow-Up of a New Surgery Method: Laparoscopy-Assisted Heart-Shaped Anastomosis for Hirschsprung's Disease. *Journal of Laparoendoscopic & Advanced Surgical Techniques*, *28* (4), 471-475. doi: 10. 1089/lap. 2017. 0275
7. Nielsen, S. W., Ljungdalh, P. M., Nielsen, J., Nørgård, B. M., & Qvist, N. (2017). Maternal use of selective serotonin reuptake inhibitors during pregnancy is associated with Hirschsprung's disease in newborns – a nationwide cohort study. *Orphanet Journal of Rare Diseases*, *12* (1). doi: 10. 1186/s13023-017-0667-4