HIRSCHPRUNG'S DISEASE: PRE AND POST-OPERATIVE CARE IN NEONATES - GCNC - CHW

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- Hirschsprung's disease is a congenital malformation which is characterised by a lack of ganglion cells in the intestine[2] which prevents peristaltic activity through the bowel[3].
- Abdominal distension, bilious vomiting and/or enterocolitis and an X-ray that suggests distal bowel obstruction may suggest Hirschsprung's disease. Diagnosis is confirmed by rectal examination, and suction rectal biopsy.
- The main goals of pre-operative management are to prevent vomiting, reduce abdominal distension, prevent the development of secondary colitis, maintain nutrition and fluid and electrolyte balance[6] and keep the neonate comfortable. This is achieved through the insertion of a gastric tube, commencement of intravenous fluids, rectal washouts, the use of antibiotics and enteral feeding if tolerated.
- The most commonly used surgical procedure to treat Hirschsprung's Disease at the Children's Hospital at Westmead is the Soave procedure where normal intestine is "pulled through" to the anus[4]
- Long segment or complex disease may be initially managed with a stoma.
- Post-operative care involves monitoring for abdominal distension or infection, fluid intake, managing pain, stoma care (if present), ensuring hydration and commencement of oral feeds when appropriate.

This document reflects what is currently regarded as safe practice. However, as in any clinical situation, there may be factors which cannot be covered by a single set of guidelines. This document does not replace the need for the application of clinical judgement to each individual presentation.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
Date Effective:	1st June 2023	**Review Period**: 3 years
Team Leader:	Clinical Nurse Consultant	**Area/Dept**: GCNC - CHW

CHANGE SUMMARY

- Due for Mandatory review
- References updated
- Inclusion of risk of enterocolitis

READ ACKNOWLEDGEMENT

- All clinical staff working in Grace Centre for Newborn Care (CHW) should read and acknowledge they understand the contents of this document.

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Background

Hirschsprung's disease is a congenital malformation which is characterised by a lack of ganglion cells in the intestine[2] which prevents normal peristaltic activity through the bowel[4]. The symptoms of Hirschsprung's disease in neonates include; abdominal distension, vomiting and failure to pass meconium within 48 hours of birth[5].

Hirschsprung's Disease occurs 1 in every 5,000 neonates and is the most common cause of large bowel obstruction[4] in the neonatal period. A male to female ratio of 1:3 to 4:1; when the entire colon is involved, the sex ratio more nearly approaches 1:1. [13]

Ganglion cells are needed for peristalsis and its absence results in a functional bowel obstruction. Absence of ganglion cells in the distal part of the large intestine is usually from the rectosigmoid colon onwards although it can extend proximally into the small bowel as well. Hirschsprung's Disease occurs as an isolated phenotype however there are strong associations that support a genetic etiology[6].

Total colonic aganglionosis is a relatively uncommon form of Hirschsprung's disease. It occurs in approximately 2–13 % of Hirsphprung's cases and involves the entire colon which is aganglionic but may extend proximally into varying lengths of small bowel [7] As a result, it should be separated into Total colonic aganglionosis, defined as aganglionosis extending from the anus to at least the ileocaecal valve but no more than 50 cm small bowel proximal to the ileocaecal valve, and **total colonic and small bowel aganglionosis** which may involve very long segments of small bowel aganglionosis. [7] Definitive diagnosis of total colonic or total colonic and small bowel aganglionosis is determined intra-operatively at time of surgical repair.

Pathophysiology

The most accepted theory of the cause of Hirshprung's Disease is that there is a defect in the craniocaudal migration of neuroblasts originating from the neural crest, a process that begins at four weeks of gestation and ends at week 7 with the arrival of neural crest-derived cells at the distal end of the colon. Failure of the cells to reach the distal colon leaves that segment aganglionic and therefore nonfunctional, resulting in Hirshprung's Disease. Defects in the differentiation of neuroblasts into ganglion cells and ganglion cell destruction within the intestine may also contribute to the disorder.[12]

Genetics

Mutations in several genes have been identified in patients with Hirshprung's Disease. Hirshprung's Disease is a genetically complex disorder caused by variants in multiple rare genes with low penetrance and variable expression. Thus, individuals with multiple pathogenic variants have substantially increased risk compared with those with fewer pathogenic variants. For nonsyndromic forms, long-segment disease tends to be transmitted by autosomal dominant inheritance and short-segment disease often reflects autosomal recessive or multifactorial inheritance. [10]

Diagnosis

If it is suspected that an infant has Hirschsprung's disease a number of diagnostic tests may be carried out to confirm the diagnosis.

Rectal examination

- The rectal examination is performed by the surgical team in the NICU.
- When a gloved finger is inserted into the rectum, an explosive release of gas and meconium may occur[2].The nurse caring for the neonate provides containment and gives sucrose if appropriate (refer to the *sucrose administration* practice guideline).

Abdominal X-ray

- Signs of distal obstruction ie, decreased or absent air in the rectum and dilated bowel loops proximal to the aganglionic region will suggest Hirschsprung's disease.[4] The X-ray can be performed either in the NICU or in the medical imaging department.
- Refer to the Transfer of a neonate to operating theatre or other department for additional information

Rectal suction biopsy

- The demonstration of aganglionosis on rectal biopsy establishes the definitive diagnosis of Hirschsprung's disease[5].
- The biopsy is carried out by the surgical team. No anaesthesia is necessary and it is not expected to be painful for the child as the biopsy is taken from above the dentate line which lacks pain fibres.
- The nurse or parent provides comfort measures during the procedure (refer to Pain Management in Newborn Infants practice guidelines) and gives sucrose if appropriate (refer to the Sucrose Administration practice guideline).
- The nurse may also be required to obtain equipment for the biopsy as requested by the surgical team.
- The specimen is sent fresh to pathology.
- The result of the biopsy is ordinarily available within 24 - 36 hours depending on the timing of the submission of the specimen.
- Bowel washout should not be performed for at least 24hours following rectal biopsy. [4]

Contrast enema

- A contrast enema may be performed in order to demonstrate a transition zone, which is most commonly seen in the recto-sigmoid colon[2].
- The contrast enema is useful for pre-surgical planning because it will assist in determining the length of aganglionic segment. However, it is not always consistent with its true pathologic location[11]

If rectal examination or bowel washouts have occurred prior to the enema, the study may be hard to interpret or the transition zone may not be seen[4].

- A contrast enema is performed in the fluoroscopy facility in medical imaging. The nurse caring for the neonate will need to escort the infant to the department (refer to the Transfer of a Neonate to Operating Theatre and other Hospital Investigative Departments guideline).

Pre-operative Management

The main goals of pre-operative management are to prevent vomiting, reduce abdominal distension, prevent secondary colitis, maintain nutrition and fluid and electrolyte balance[6] and keep the neonate comfortable. In order to achieve these goals, the following is required:

Insertion of a gastric tube

- An gastric tube is inserted (refer to Feeding the high risk neonate practice guideline) and placed on free drainage.
- The gastric tube is aspirated 4th hourly at a minimum. More frequent aspiration may be requested by the surgical team or Neonatologist.
- The volume of aspirate is documented in the output section of the fluid balance chart.
- If gastric aspirates are large (greater than 10-20mL/kg), the registrar or nurse practitioner is informed and gastric replacement may be commenced with 0.9% Sodium Chloride.

Commencement of intravenous fluids (IV)

- Initially the infant is made nil by mouth (NBM) and intravenous maintenance fluid of 0.225% Sodium Chloride + 10% Glucose is commenced[4].
- It may be necessary for electrolytes or extra glucose to be added to the IV fluid based on the neonate's blood sugar level, plasma electrolytes, urine output and specific gravity and body weight [12].

Regular rectal washouts

- Regular rectal washouts may be necessary if clinically indicated and if requested by the surgical team. Washouts are ordinarily performed once daily but may be more frequent initially to achieve decompression.
- Rectal washouts are extremely valuable to prevent enterocolitis[10].
- Normal saline is used which relieves the obstruction, decompresses the bowel and ensures adequate elimination[5].
- The rectal washout is performed in the NICU by the surgical team.
- The nurse caring for the neonate is present during the washout to implement comfort measures (refer to *Pain Management in Newborn Infants* practice guidelines) and to administer sucrose if appropriate (refer to the *Sucrose Administration* practice guideline).

- In some cases the rectal catheter may be left insitu to aid the passage of meconium and intestinal gas and prevent the need to reinsert the catheter for each washout, thus reducing stress.
- Occasionally, daily washout may not be necessary and they may be performed less frequently; this will be decided by the surgical team involved in the neonates care.

Equipment required:

- Disposable gloves and gown
- Incontinence Sheets/ Bluey
- Normal Saline warmed in designated "bowel washout" jug
- Nelaton Catheter (Rectal Tube) - sizing will be advised by surgical team but usually size 10fr, 12 fr or 14fr
- 60mL Terumo Syringe catheter tip
- Disposable kidney dish
- Lubricating Jelly
- Wipes and New Nappy.

Risk of enterocolitis

Neonates with Hirchsprungs disease are at greater risk of enterocolitis, the mechanisms of which are not fully understood. Contributing factors include stasis and reduced production of protective mucin, which lead to bacterial overgrowth in the lumen of the bowel proximal to the involved segment, and possibly translocation of bacteria through the mucosa. Hirchsprungs disease associated enterocolitis is associated with intestinal dysbiosis, with reduced commensal bacteria (eg, bifidobacteria and *Lactobacillus*); in some cases, pathogenic bacteria (eg, *Clostridioides* [formerly *Clostridium*] *difficile*) are isolated, and rotavirus and *Staphylococcus aureus* may also have a role. The bowel wall in the affected area is subsequently invaded by colonic organisms, which can lead to pneumatosis intestinalis, intestinal perforation, peritonitis, systemic sepsis, shock, and death[13]

- Infants with clinical confirmation of enterocolitis are commenced on broad spectrum antibiotics[12]
- Enterocolitis is a serious condition which may occur with prolonged abdominal distension and constipation[2]. Clinical indications of enterocolitis include; fever, abdominal distension, spontaneous diarrhoea and vomiting[12].

Enteral Feeds

- Once the bowel has been decompressed, enteral feeds can be commenced[1].
- The surgical team and Neonatologist will liaise to determine the amount of enteral feed to be given.
- Feeds will be graded up as tolerated by the infant (refer to *Feeding the high risk neonate- GCNC practice guideline*)

- Once feeds have been commenced the gastric tube should no longer be on free drainage, or aspirated routinely, or as specified by the surgical team or Neonatologist.[9]

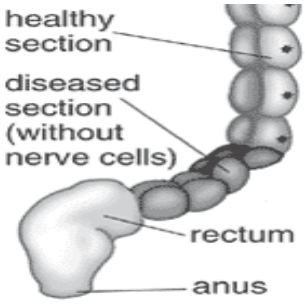
Surgical Intervention

The aim of surgery is to move normal ganglionated bowel down to the anus[6]. In the past this was achieved with a two-to three-stage pull through, with a stoma initially and then definitive repair between 6 and 12 months of age. The repair involves the removal of aganglionic bowel and joining of normal bowel to the anus[8]. Primary repair is now more common with an endorectal pull-through[1]. The most commonly used surgical procedure to treat Hirschsprung's Disease at the Children's Hospital at Westmead is the Soave procedure.

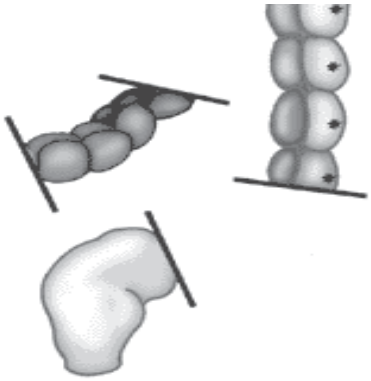
The Soave procedure involves a single stage operation done either entirely from the anus or from both the abdomen and the anus. Commonly, the aganglionic rectum and colon is identified by biopsies performed laparoscopically. The colon is mobilized laparoscopically and removed through the anus. Dissection from the anus involves an endorectal dissection which essentially is creating a plane between the mucosa and the muscularis of the rectum and pulling out a sleeve of mucosa. The peritoneal cavity is then entered and the mobilized bowel is pulled through from below, removed to the desired point and anastomosed to the anus[6]

The Soave Procedure

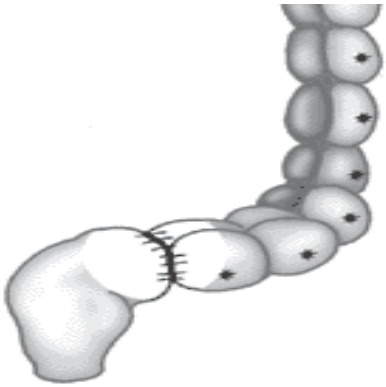
Step 1: Diseased bowel preventing peristalsis.



Step 2: Diseased bowel is dissected.



Step 3: The healthy bowel is anastomosed to the anus



Treatment of total colonic aganglionosis

Total colonic involvement has significantly higher morbidity and mortality than short segment Hirschsprung's disease and requires more a complex medical and operative management . Multiple procedures exist for the treatment of total colonic aganglionsis; however, there is no current consensus on a superior operative procedure. [8]

Treatment of Total colonic aganglionosis involves:

- Early decompression of the colon by an ostomy using the most distal ganglionated bowel in the neonatal period. [6]
- This is typically followed by a second reconstructive procedure after the child has had adequate time to grow and nutritional, fluid, and electrolyte abnormalities are corrected.[8]

Post-op care

Monitoring for infection

- The infant needs to be monitored for signs of infection such as poor feeding, irritability, temperature instability and lethargy[12].
- If infection is suspected timely intervention with broad spectrum antibiotics, following blood culture, is important since infants with Hirschsprung's Disease are at increased risk of Enterocolitis[5].

Pain Management

- The Modified Pain Assessment Tool (m-PAT) is used to measure the infant's pain every two hours in the initial post-operative period and every four hours after that. Refer to the *Pain Management in Newborn Infants* practice guideline for how to score infants and the action required for each score.
- In general infants are commenced on a narcotic infusion to prevent pain. The infusion can be weaned as tolerated. Intravenous paracetamol can be administered as an adjunctive analgesic within the first 24-48 hours. Once the infusion is ceased it is important to continue to monitor pain using the PAT and treat pain accordingly.
- Occasionally the infant may return to the intensive care with an epidural catheter inserted
- The pain team and/or anaesthetic registrar is responsible for the epidural infusion.
- The nurse is responsible for observing the insertion site and ensuring the dressing is intact on an hourly basis and completing the epidural section in the electronic medical record as well as any hard copy charts supplied by the pain team and/or anaesthetic registrar. The nurse should also be responsible for double checking the order of the epidural on the Epidural chart
- If any problems or concerns arise regarding the epidural catheter and infusion the pain team and/or anaesthetic registrar need to be contacted as soon as possible.
- For more information on the care of an infant with an epidural refer to the CHW *Pain Management* practice guideline .

Hydration

- Intravenous fluids are commenced post-operatively as ordered by the registrar, nurse practitioner or neonatologist caring for the infant.
- Hydration is ensured by monitoring urine output, urine specific gravity and blood electrolyte levels.
- Initially crystalloid fluids are used until feeds are commenced. If it is anticipated that the infant will not commence feeds or be likely to be tolerating full feeds within 3-5 days, Total Parental Nutrition (TPN) may be commenced until feeds are tolerated.

Aspiration of gastric tube

- The infant will continue to have a FG8 gastric tube in situ until feeds are commenced, at which stage the intra-gastric tube will be replaced with a FG6.
- While the infant is NBM the gastric tube is on free drainage and aspirated 4th hourly or more frequently if requested by the surgical team or neonatologist.
- Once the infant is having enteral feeds, the gastric tube is clamped and tolerance is evaluated by assessing for abdominal distension, vomiting or discomfort. If these symptoms are present, the nasogastric tube is aspirated to decompress the stomach. Attempts at recommencing enteral feeds should be made in conjunction with surgical team and neonatologist.
- Routine aspiration of gastric tubes is not based on evidence and can delay the attainment of full enteral feeds by up to 6 days[2]
- The use of gastric residuals to monitor feed tolerance has been deemed a time consuming practice that causes significant confusion in regards to feeding management, and could cause harm to the delicate gastric mucosa and also potentially lead to the loss of essential gastric enzymes[12]

Commencement of Enteral Feeds

- Enteral feeds will be commenced once the surgical team is satisfied that the infant has minimal clear gastric aspirates and the abdomen is less distended.
- The neonatologist will generally decide how much feed to commence once discussion with the surgical team and nurse caring for the infant has occurred.
- Feeds will generally be commenced orally however some infants may require intragastric feeds until they are showing signs of oral feed capability (refer to *Feeding the high risk neonate- GCNC practice guideline*)
- The feeds will be graded up as tolerated by the infant. When feeds have been commenced, the gastric tube is capped and aspirates are discontinued.

Observation for Abdominal Distention

- The abdomen needs to be closely observed for distension.[5]
- An accurate description of the abdomen needs to be documented in the assessment chart of the electronic medical record. Any changes in appearance need to be reported to

the nurse in-charge, nurse practitioner and/or the registrar and then reviewed by the surgical team. An abdominal X-ray is useful to assist in interpreting the change in clinical status.[5]

- If the infant is receiving enteral feeds and the abdomen becomes distended, feeds should be ceased and the infant made nil by mouth (NBM), gastric tube aspirated and reviewed by the surgical team.

Respiratory Support

- Occasionally the infant will return from theatre intubated and ventilated.
- For further information on how to care for an intubated and ventilated patient refer to the *Respiratory Support in the NICU* practice guideline .

Blood tests

- The registrar, nurse practitioner or neonatalogist may request post-operative blood tests, which can be collected via an arterial line (if available) or venipuncture.
- Post-op tests generally include electrolytes, urea and creatinine (EUC) and a full blood count (FBC).
- Other tests may be requested at the discretion of the registrar, nurse practitioner or neonatologist.
- Sucrose may be given if collection is by venipuncture (refer to the *Sucrose Administration* practice guideline).

Care of rectal tube

- The infant may occasionally return from theatre with a rectal tube insitu to aid the passage of meconium and to keep the rectum patent.
- The rectal tube is secured using tape and it is the responsibility of the nurse caring for the infant to ensure that the rectal tube is secure and taping adjusted if necessary.
- If it is suspected that the rectal tube is blocked, the nurse-in-charge, nurse practitioner and/or registrar must be informed. The surgical team needs to be contacted to review the infant.

Stoma care

- A stoma or ileostomy might be formed by the surgical team temporarily to provide protection for the future resection or repair. [11]
- In the event that a stoma is formed, the nurse caring for the infant is responsible for attending to stoma care and educating the parents as to how to care for the stoma and how to use stoma appliances.
- The stoma clinical nurse consultant may be contacted to advise on stoma care and education as well as answer any questions the parents or nursing staff may have.

- Ensure that the stoma is functioning adequately, and assess the stoma for perfusion, moistness and skin integrity[5]. The observation of the stoma must be noted in the clinical assessment chart in the electronic medical record.
- The registrar, nurse practitioner and nurse in charge must be informed if any bleeding or change in stoma perfusion is noted.
- Parents should be shown how to fit the stoma bag and care for the surrounding skin.
- Refer to the Ostomy Nursing Care procedure for further information on stoma care.

Related Documents

GCNC documents

- Pain Management in Newborn Infants Practice Guideline
- Feeding the high risk neonate Practice Guideline
- Respiratory Support in the NICU Practice Guideline
- Transfer of a neonate to operating theatre and other hospital investigative departments GCNC practice guideline

SCHN documents

- Sucrose: Management of Short Duration Procedural Pain in Infants Practice Guideline
- Ostomy Nursing Care Procedure
- Pain Management: Epidural infusions

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