HIRSCHSPRUNG'S DISEASE: PRE AND POSTOPERATIVE CARE IN NEONATES - GCNC

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[1] which prevents peristaltic activity through the bowel[2].
- 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. A contrast enema is sometimes performed.
- 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[5] 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[1]
- 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.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
Date Effective:	1st February 2017	**Review Period:** 3 years
Team Leader:	Nurse Educator	**Area/Dept**: GCNC

Date of Publishing: 12 January 2017 10:49 AM Date of Printing:

K:\CHW P&P\ePolicy\Jan 17\Hirshprungs Pre and Post Op Mgmt GCNC.docx

CHANGE SUMMARY

Outline a summary of changes to the revised document.

- References updated
- Information revised by surgical team and neonataologists

READ ACKNOWLEDGEMENT

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

TABLE OF CONTENTS

Background **4**
Diagnosis **4**
Rectal examination *4*
Abdominal X-ray *4*
Rectal suction biopsy *4*
Contrast enema *5*
Pre-operative Management **5**
Insertion of an intragastric tube *5*
Commencement of intravenous fluids (IVF) *5*
Regular rectal washouts *5*
Antibiotics *6*
Surgical Intervention **7**
The Soave Procedure 7
Post-op care **8**
Monitoring for infection *8*
Pain Management *8*
Hydration *9*
Aspiration of gastric tube *9*
Commencement of Enteral Feeds *9*
Observation for Abdominal Distention *9*
Respiratory Support *10*
Blood tests *10*
Care of rectal tube *10*
Stoma care *10*
Related Documents **11**
GCNC documents *11*
SCHN documents *11*
References **12**

Background

Hirschsprung's disease is a congenital malformation which is characterised by a lack of ganglion cells in the intestine[1] which prevents normal peristaltic activity through the bowel[2]. The symptoms of Hirschsprung's disease in neonates include; abdominal distension, vomiting and failure to pass meconium within 48 hours of birth[3].

Hirschsprung's Disease occurs 1 in every 5,000 neonates and is the most common cause of large bowel obstruction[2] in the neonatal period.

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[4].

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 nursery.
- When a gloved finger is inserted into the rectum, an explosive release of gas and meconium may occur[1].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. The X-ray can be performed either in the nursery or in the medical imaging department.

Rectal suction biopsy

- The demonstration of aganglionosis on rectal biopsy establishes the definitive diagnosis of Hirschsprung's disease[1].
- The biopsy is carried out by the surgical team in the treatment room of the nursery. 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.

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[4].
- 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[7]
- 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[3].
- 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.

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[5] and keep the neonate comfortable. In order to achieve these goals, the following is required:

Insertion of an intragastric tube

- An intra-gastric tube (IGT) is inserted (refer to Enteral Feeding - Neonates practice guideline) and placed on free drainage.
- The IGT 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 10-20mlg/kg), the registrar is informed and gastric replacement may be commenced with 0.9% Sodium Chloride.

Commencement of intravenous fluids (IVF)

- Initially the infant is made nil by mouth (NBM) and intravenous maintenance fluid of N/4 Saline + 10% Dextrose is commenced[3].
- It may be necessary for electrolytes or extra dextrose 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[3].

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[8].
- Normal saline is used which relieves the obstruction, decompresses the bowel and ensures adequate elimination[5].
- The rectal washout is performed in the treatment room of the nursery 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 a small pot
- Rectal Tube- Sizing will be advised by surgical team but usually size 10fr or 12 fr
- 50ml Tumi Syringe
- Disposable kidney dish
- Lubricating Jelly
- Wipes and New Nappy.

Antibiotics

- Infants with clinical confirmation of enterocolitis are commenced on broad spectrum antibiotics[3] (i.e. Metronidazole)
- Enterocolitis is a serious condition which may occur with prolonged abdominal distension and constipation[8]. Clinical indications of enterocolitis include; fever, abdominal distension, spontaneous diarrhoea and vomiting[4].

Enteral Feeds

- Once the bowel has been decompressed, enteral feeds can be commenced[3].
- 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 *Enteral Feeding - Neonates* practice guideline).

- Once feeds have been commenced the IGT should no longer be on free drainage and can be aspirated 4th hourly, or as specified by the surgical team or Neonatologist.
- The residual volume of feeds needs to be documented in the input section of the fluid balance chart.

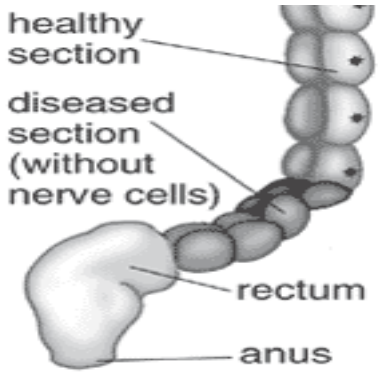
Surgical Intervention

The aim of surgery is to move normal ganglionated bowel down to the anus[1]. 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[1]. The repair involves the removal of aganglionic bowel and joining of normal bowel to the anus[1]. 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.

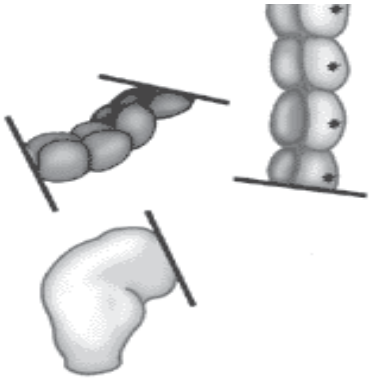
The Soave procedure involves a single stage operation done either entirely from the anus or from both the abdomen and the anus. Commonly, the aganglionic rectum and colon is identified by biopsies performed laparoscopically. The colon is mobilized laparoscopically and removed through the anus. Dissection 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

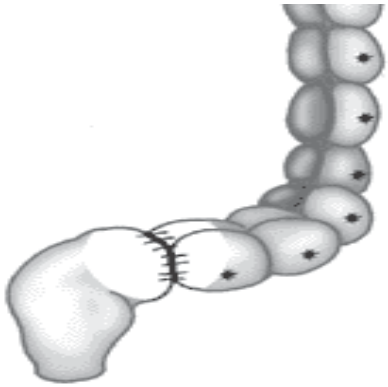
Step 1: Diseased bowel preventing peristalsis.



Step 2: Diseased bowel is dissected.



Step 3: The healthy bowel is anastomosed to the anus



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[1].
- 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[3].

Pain Management

- The Pain Assessment Tool (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 nursery 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 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, tolerance is evaluated by aspirating the gastric tube 4th hourly at a minimum.
- All gastric losses and residuals must be entered into the input/output section of the electronic medical record.

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 via an intragastric tube and once the infant is stable and showing signs of sucking oral feeds will be introduced.
- The feeds will be graded up as tolerated by the infant (refer to *Enteral Feeding - Neonates* practice guideline.
- When feeds have been commenced, the IGT is capped and aspirated 4th hourly to determine the infant's tolerance to feeds.

Observation for Abdominal Distention

- The abdomen needs to be closely observed for distension.
- 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 and the registrar and then reviewed by the surgical team. An abdominal X-ray is useful to assist in interpreting the change in clinical status.

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

Respiratory Support

- Occasionally the infant will return from theatre intubated and ventilated. On return from theatre the position of the endotracheal tube (ETT) is confirmed on chest X-ray and with measurement using a tape measure. The ETT should be positioned in the mid-trachea between the thoracic inlet and carina (T1- T2).
- If the ETT is not positioned correctly it is re-taped in an appropriate position specified by the registrar. Re-taping is performed by two staff, one being an accredited RN.
- It is advisable that the ETT be suctioned upon return to the nursery to ensure patency of the tube and clear any secretions present within the tube. An in-line suction catheter should be connected when the patient is back from OT.
- To make certain that the infant is being ventilated adequately the chest is auscultated for equal clear breath sounds.
- 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 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 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 and 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. (BMJ)
- 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 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 CHW *Ostomy Nursing Care* procedure for further information on stoma care.

Related Documents

GCNC documents

Pain Management in Newborn Infants Practice Guideline:
http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2006-0028.pdf

Enteral Feeding – Neonates Practice Guideline:
http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2007-0002.pdf

Respiratory Support in the NICU Practice Guideline:
http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2007-0005.pdf

SCHN documents

Sucrose: Management of Short Duration Procedural Pain in Infants Practice Guideline:
http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2006-8241.pdf

Ostomy Nursing Care Procedure:
http://chw.schn.health.nsw.gov.au/o/documents/policies/procedures/2006-8041.pdf

References

1. Kenner, C. and Lott, J.W. (eds) 2007 *Comprehensive neonatal care: An interdisciplinary approach.* 4th edn, WB Saunders, Philadelphia pp. 118-120, 204.
2. Levene, M.I., Tudehope, D.I. and Sinha, S. 2008. *Essential Neonatal Medicine.* 4th edn, Blackwell Publishing, Massachusetts pp. 251-252.
3. Barr, P. 2013 *Newborn Intensive Care – A handbook for staff. 4th* Ed, The Children's Hospital at Westmead, p.97.
4. Georgeson K. 2010. 'Hirschsprungs Disease' in *Ashcroft's Pediatric Surgery 5th Ed.*, Holcomb III and Murphy Eds. Saunders Imprint of Elsiever, Philadelphia, pp456-467.
5. London, M.L., Ladewig, P.W., Ball, J.W., and Bindler, R.C. 2003 *Maternal-Newborn & Child Nursing: Family-centered care*. Pearson Education, New Jersey, p.157.
6. Ryckman, F.C. and Balisteri, W.F 2002 The neonatal gastrointestinal tract: Part two – upper gastrointestinal disorders. In Fanaroff, A.A and Martin, R.J, editors. *Neonatal-perinatal medicine: diseases of the fetus and infant,* ed 7. St Louis, Mosby. P.87
7. Weeson D. E, Singer J.I and Hoppin A.G 2016 *Congenital aganglionic megacolon (Hirschsprung's disease*) UpToDate. P1-29
8. Bischoff, A. and A. Pena (2016, 19/10/2016). "Hirschsprung's Disease." BMJ Best Practice. 2016, from http://bestpractice.bmj.com.acs.hcn.com.au/best-practice/monograph-pdf/750.pdf.