

Local Guideline



John Hunter
Children's Hospital
CHILDREN, YOUNG PEOPLE AND FAMILIES



Health
Hunter New England
Local Health District

Omphalocele Management in the neonate

Sites where Local Guideline applies	Neonatal Intensive Care Unit, JHCH
Target audience:	NICU clinical staff, which provides care to neonatal patients with an omphalocele in Delivery Suite and NICU.
Description	Guideline for the management of infants with abdominal wall defects such as omphalocele
This Local Guideline applies to:	
1. Adults	No
2. Children up to 16 years	No
3. Neonates – less than 29 days	Yes
Keywords	Approval gained from the Children Young People and Families Network on (20/10/2014) Abdominal wall defect, Bowel, Exomphalos, Gastroschisis, Malrotation, Omphacocele, vac dressing
Replaces Existing Local Guideline and Procedure	Yes
Registration Number(s) and/or name and of Superseded Documents	5.8.6 Guideline for the Management of Abdominal Wall Defects
Related Legislation, Australian Standards, NSW Health Policy Directive, NSQHS Standard/EQUIP Criterion and/or other, HNE Health Documents, Professional Guidelines, Codes of Practice or Ethics:	
<ul style="list-style-type: none"> NSW Health Policy Directive 2007_079 Clinical Procedure Safety http://www0.health.nsw.gov.au/policies/pd/2014/pdf/PD2014_036.pdf NSW Health Policy PD 2005_406 Consent to Medical Treatment http://www.health.nsw.gov.au/policies/PD/2005/pdf/PD2005_406.pdf NSW Health Policy Directive PD 2007_036 Infection Control Policy http://www.health.nsw.gov.au/policies/pd/2007/pdf/PD2007_036.pdf NSW Health Policy Directive PD 2010_062 Antenatal Maternal Referral/Transfer: Known Congenital http://www0.health.nsw.gov.au/policies/pd/2010/pdf/PD2010_062.pdf 	
Prerequisites (if required)	N/A
Local Guideline Note	This document reflects what is currently regarded as safe and appropriate practice. The guideline section does not replace the need for the application of clinical judgment in respect to each individual patient but the procedure/s require mandatory compliance . If staff believes that the procedure/s should not apply in a particular clinical situation they must seek advice from their unit manager/delegate and document the variance in the patients' health record.
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This Local Guideline contains advice on therapeutics	No
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Note: Over time links in this document may cease working. Where this occurs please source the document in the PPG Directory at: <http://ppg.hne.health.nsw.gov.au/>

RISK STATEMENT

This local guideline has been developed to provide guidance to clinical staff in NICU when caring for an infant with an omphalocele. It ensures that the risks of harm to infants and staff are identified and managed.

Any unplanned event resulting in, or with the potential for injury, damage or other loss to infants/staff/family as a result of this management must be reported through the Incident Information management System and managed in accordance with the Ministry of Health Policy Directive: Incident management PD2007_061. This would include unintended injury that results in disability, death or prolonged hospital stay.

RISK CATEGORY: *Clinical Care & Patient Safety;*

OUTCOMES

1	Delivery at JHH will enable expert teams of surgeons and Neonatal clinicians to avoid trauma to the bowel and ensure heat, fluid and electrolyte loss is minimised at and immediately post-delivery.
2	Surgery will be performed following consultation with surgeons
3	Parents will be kept informed throughout admission of plan of care, investigations and prognosis
4	For infant to commence oral feeds as soon as possible

ABBREVIATIONS & GLOSSARY

Abbreviation/Word	Definition
ABG	Arterial blood gas
BSL	Blood sugar level
DDA	Dangerous Drugs of Addiction
EUC	Electrolytes, Urea and Creatinine
FBC	Full blood count
IV	Intravenous
JHH/JHCH	John Hunter Hospital/John Hunter Children’s Hospital
MO/NNP	Medical Officer/Nurse Practitioner
NICU	Neonatal Intensive Care Unit
PICC	Peripherally inserted central catheter
PIPP	Premature infant pain profile
PN	Parenteral Nutrition
PR	Per rectum

GUIDELINE

Omphalocele - Definition

An omphalocele occurs following a disruption to the development of embryonic folds in the 10th week of fetal life. A fault in the lateral body fold or the cranio-caudal fold that would normally converge at the umbilical ring and close the abdominal wall, leads to an abdominal defect of variable size at the umbilical ring. It generally occurs in the mid abdomen but may be seen in the upper or lower abdomen. It causes loops of bowel and sometimes liver and other abdominal organs to herniate through the defect at the umbilicus with an amnion sac covering the contents.¹ As many as 10-50% of infants presenting with an omphalocele will have associated anomalies such as cardiac defects, neurologic, genitourinary and skeletal and chromosomal anomalies, such as trisomy 12, 18 & 21 and Beckwith Wiedemann syndrome¹.



Intact Omphalocele



Small Omphalocele

<http://emedicine.medscape.com/article/975583-overview>¹³

Giant Omphalocele

An omphalocele that is >5cm in diameter is classified as 'giant'. The large size of the defect causes underdevelopment of the abdominal cavity and disproportion between the abdominal cavity and its contents. This makes immediate reduction impossible due to increased abdominal pressure.

Other features of a Giant omphalocele:

- Large, mostly centrally located abdominal wall defect however some occur in the upper abdomen/lower chest as well as the lower abdomen
- Liver located outside of the abdominal cavity, within the omphalocele sac
- Small, un-developed thoracic cavities which leads to restrictive lung disease and pulmonary hypoplasia¹³



Giant Omphalocele

<http://emedicine.medscape.com/article/975583-overview>¹³

Antenatal preparation and delivery of an infant with an Omphalocele

- Antenatally the mother is seen at least once by the neonatologist and surgeon, and is able to visit the NICU
- Ideally a management plan is formulated between 28 weeks and 32 weeks of gestation. This should be discussed with the parents and written in the case notes
- There are no significant differences in short term outcomes for infants with a prenatal diagnosis of abdominal wall defects who are delivered vaginally⁷. This should be the preferred mode of delivery unless contraindicated by obstetric requirements.
- The NSW consensus meeting⁸ on abdominal defects advise that no elective delivery should take place before 37 weeks gestation unless there is foetal distress
- Delivery of a neonate with omphalocele is very similar to one with gastroschisis.

Delivery room management of a neonate with an Omphalocele

- Follow the Flow Chart (Appendix 1) for an Omphalocele
- Ward reduction equipment box, metal frame and plastic wrap can be found on the shelf next to the DDA cupboard in Level 3 and should be kept ready in the NICU. Check that a silo is available in the equipment box if ward reduction has been planned. Spare silos available in theatres.
- The NICU will provide a MO/NP and nurse at delivery to provide resuscitation and care of the neonate with an omphalocele.
- Resuscitation is the initial priority for newborns with an omphalocele. It is preferable to avoid bag and mask ventilation to minimise gas in the stomach and bowel. If prolonged respiratory support required, particularly for moderate to large defects, intubate with an endotracheal tube. With omphalocele presentation,

careful attention should be made around the cardiopulmonary status as there maybe unsuspected pulmonary hypoplasia. ¹

- Following stabilization, an oro-gastric tube should be inserted to decompress the stomach and avoid intestinal distension
- The infant should be dried and a warm environment maintained whilst protecting the exposed viscera to prevent heat loss. Due to potential evaporative water losses from the bowel the abdominal wall defect should be kept moist by gently covering with plastic wrap. This is best achieved by initially wrapping the plastic film around the defect and then over the top. This preserves body heat and minimizes insensible water loss.
 - **NOTE:** This is a two-person procedure. Care should be taken not to twist or kink the bowel by ensuring the plastic wrap is not applied too tightly.
- Ensure the cord is cut as long as possible – this is very important as the cord may contain bowel contents
- Cannulation of the umbilical vessels is difficult for an omphalocele due to the abnormal insertion and position of the vessels ¹. Insert a peripheral intravenous access to provide fluids.
- It is important to position the baby and support the exteriorized bowel in such a way as to maintain best possible circulation to the bowel by taking tension off the mesenteric vessels. This is achieved by having the baby in the right lateral posture with support under the bowel (very important for the large/giant omphalocele)¹.
- For an infant with an omphalocele if the covering membrane is intact a non adherent dressing may be applied.
- If the sac is ruptured in an omphalocele, treat the same as for a gastroschisis. Refer to CPG “Gastroschisis management in the neonate” JHCH_NICU_16.03\



Infant with a ruptured omphalocele

<http://emedicine.medscape.com/article/975583-overview> ¹³

Review of an infant with Omphalocele following delivery in NICU

- Once stable in NICU a thorough examination of the neonate should be undertaken by the Neonatologist/Fellow/Nurse Practitioner.
- Newborns with omphalocele should be examined and investigated to exclude cardiac and renal anomalies.
- Surgeons should discuss treatment options and obtain consent from parents
- The aim of the surgical team is to reduce the external bowel and organs into the abdomen using a method that minimises risks to the baby.

Pre-operative care of defect

- Sterile latex free gloves should be used when handling bowel to minimise infection
- Ensure defect remains covered with plastic wrap
- Wet gauze/cotton must NOT be used (threads stick to bowel and lead to more inflammation; and wet gauze also causes hypothermia)
- Prepare infant for a ward reduction or theatre
- Refer to CPG: [‘Surgery-Preparation and care of infant in NICU’ 5.8.2](#)

Fluid management

- Infant is placed Nil By Mouth
- A peripheral IV should be inserted preferably in upper limbs and not lower limbs (as there may be poor venous return due to compression of the vena cava)¹⁴
- Maintenance IV Therapy of 10% glucose to be commenced at 80-100ml/kg/day
- For an intact omphalocele additional fluid management should be tailored with review of BP, perfusion and clinical assessment of fluid status by the neonatologist.
- For a ruptured omphalocele treat as for a gastroschisis-CPG JHCH_NICU_16.03 ‘Gastroschisis in the neonate’
- Accurate urine output should be measured and recorded (an indwelling urinary catheter may be required in some cases)
- Regular BP monitoring should occur (with either an arterial line or using NIBP monitoring with a cuff)
- FG 8-10 oro-gastric tube should be inserted and placed on free drainage. It then should be aspirated at least 2nd hourly
- Bloods to be taken for FBC, group and x-match
- Blood gas and BSL to be taken 4th hourly, EUC 8th hourly initially, then as per neonatologist

Drug and pain management

- Antibiotics should be administered to the baby as indicated
- Inotropic support if required
- Pain assessment score should be undertaken at least once a shift

- Pain relief will often be required and can be given in discussion with surgeon and neonatologist and review of pain score. Consideration should be given to allowing sufficient time for the desired effect of pain relief medications prior to surgical management. Refer to CPG [JHCH NICU 03.04: 'Assessment and Management of pain in the Newborn'](#)

Surgical management

Defects that are <2cm are usually managed surgically by a primary closure attended to in theatre soon after delivery. This minimises the risks associated with bacterial contamination and sepsis, acidosis and hypothermia. This may be achieved by excision or inversion of the sac with closure of the fascia and skin. The surgeon may also use a flap that mobilises the muscle, fascia and skin of the abdominal wall to allow midline fascial closure. In straight forward cases repairs may be done as a ward procedure within 4 hours.

Medium to large defects require a staged procedure. This may involve a silastic silo to cover the bowel, similar to treatment for a gastroschisis. The silo is then attached to an external frame to allow elevation of the silo and gravity to assist the internal organs to settle into the abdominal cavity over time. The silo can be reduced gradually by reapplying clips (generally over 3-7 days) until the size is small enough for final closure of the abdominal wall. Another option if the sac is thick is to ligate the sac gradually to enable reduction of the viscera.

Another form of staged closure is 'escharotic therapy' which results in gradual epithelialization of the omphalocoele sac, commonly using silver sulfadiazine. Tissue expanders have also been used to increase the size of the abdominal cavity.

Time for complex cases will be considered in light of other issues such as prematurity and other anomalies ⁸

Postoperative Care

- Vital signs monitored for tachycardia, thread pulses, hypotension, poor perfusion
- Prevent hypothermia
- Nurse the baby supine – especially for the first 24-48 hours post repair
- Care must be given to avoid accidental dislodgement of silo if insitu
- Fluid balance to be carefully monitored with electrolytes being done 6th hourly;
- Large bore gastric tube to remain on free drainage and is aspirated 2-4th hourly. Aspirate losses should be replaced intravenously as prescribed
- Accurately measure and record urine output – 6th hourly urinalysis (signs of hypovolemia may be ↓ urine output and ↑ specific gravity)
- Observe for metabolic acidosis – ABG, serum lactate and BSL 4-6th hourly until stable
- Observe for progressive discolouration of the bowel, whether silo insitu or not, and report if noted

- Observe for respiratory compromise due to increased bowel returned into peritoneal cavity causing increased pressure on the diaphragm
- Observe for inferior vena cava compression – mottling, cyanosis, and oedema of lower extremities, systemic hypotension¹⁴
- Observe wound/suture line for colour, discharge, and signs of infection or breakdown
- Monitor pain score and provide appropriate analgesia;
- PICC line should be considered and PN commenced for nutrition in consultation with the neonatologist.
- Introduce oral feeding in consultation with surgical team;
- Provide support for parents and follow the Partnership in Care policy

Other abnormalities associated with Omphalocoele

- There is an increased risk of associated cardiac defects so close examination of cardiac status is essential.
- Renal ultrasound should be undertaken to exclude renal defects
- Close examination of glucose status should be observed as there is also an increased risk of Beckwith-Wiedemann syndrome and therefore hypoglycaemia may become a problem

General care and management

- Generally the care is the same as a Gastroschisis but if possible ensures that the sac is protected and remains intact. If the sac perforates then treat it as a gastroschisis by covering it with plastic wrap to reduce water loss and temperature loss
- Surgical treatment depends on the size of the defect, gestational age, and the presence of associated anomalies.

Prognosis

The prognosis for a patient with an omphalocoele often depends more on the associated anomalies than the presence of the omphalocoele itself.

Newer techniques

In the case of a giant omphalocele a Vacuum Assisted Closure (VAC) dressing maybe used ¹² however there is no evidence suggesting that this provides a better outcome. This device uses negative pressure to assist in healing wounds and drawing wound edges together. For care of a negative pressure wound dressing see Appendix 3.

Fig 2 Vac device
Photograph and information
provided by Kerry Sullivan
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Health Policies

NSW Health Policy Directive PD 2010_062 Antenatal Maternal Referral/Transfer: Known Congenital Structural Malformations - Early Surgery (downloaded 9th October 2013).

http://www0.health.nsw.gov.au/policies/pd/2010/PD2010_062.html

APPENDICES

1. Flow Guide for omphalocele
2. Equipment for Ward reduction
3. Steps to applying a Vac dressing

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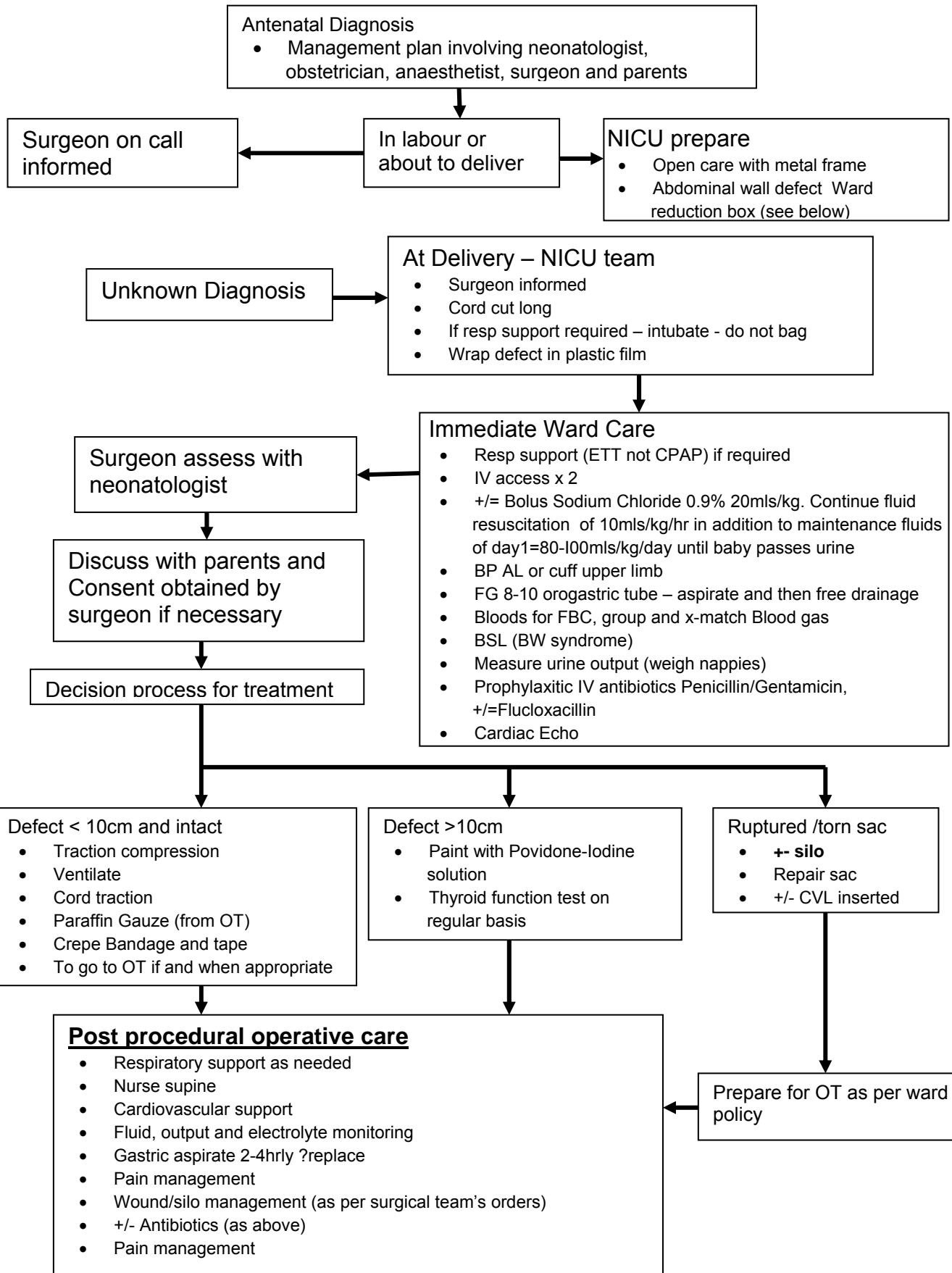
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APPROVED NICU Executive: 10/09/2014

Appendix 1

Flow guide for care of a baby with Omphalocoele Major in JHCH NICU



Appendix 2

Equipment required for Ward reduction

Boxes (kept in back storeroom), frame (on top of medication cupboard) and gladwrap roll (in 1st row of NICU storeroom)

1 Rectal washout – performed by surgeon

- 200mls of warm sodium chloride 0.9%
- 1 x 50ml Toomey syringe
- 1 x 20 ml syringe
- 1 x FG 10 Foley catheter
- 1 x Nelaton Catheter 10FG
- “Bluey” pad
- 1 x small bowl

2) Abdominal wall ward reduction box - Equipment for reduction

- 1 x each eye & ear drape
- 1 x sterile drape F6
- 2 x Steristrips 12mm
- 2 Large and small op sit IV 3000
- Skin closure sutures 3/0 / RB1 vicryl
- Benzoin compound (Friar’s Balsam)
- Hypafix - unsterile (10cm wide)
- Silk 0 suture (cord tie)

3) Extras

- Face masks with eye protection
- Sterile Gloves for 3 people
- Plastic Drapes, One large one small
- Gowns for 3 people
- Glad wrap roll

4) Surgeon brings

- Silo
- Mini instrument set (OT brings it with them)

5) Items for Omphalocoele Major reduction

- Paraffin Gauze
- Crepe Bandage and tape
- Large Cobain

Post reduction

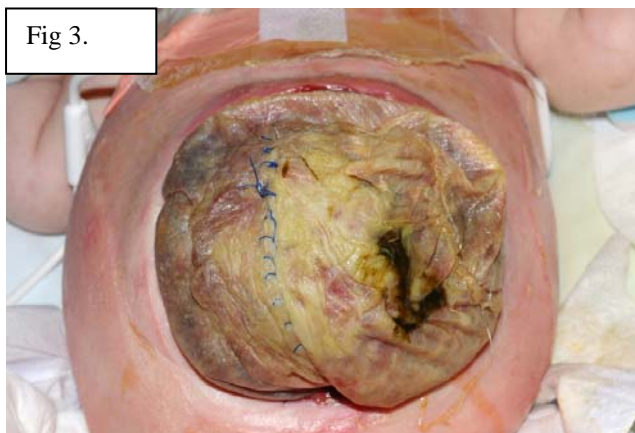
Please identify what you used on stock replacement sheet (in box), sign form and then place box in the TA room

Appendix 3

This document details basic equipment and process. Each surgeon may request different products depending on the defect.

Steps to Applying VAC Dressing

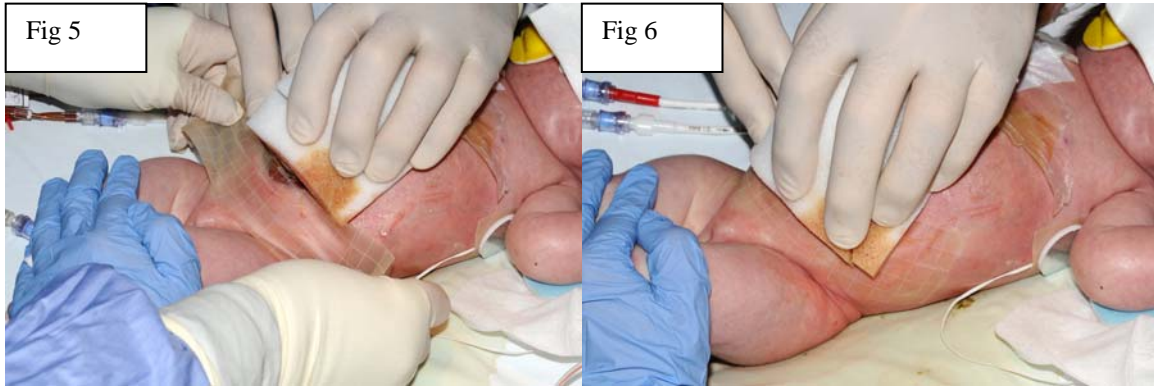
- 1 Assemble all equipment as follows:
 - a. VAC canister
 - b. VAC white foam
 - c. VAC suction tubing and drape
 - d. Comfeel
 - e. Basic dressing pack
 - f. Gauze X 7
 - g. Sterile scissors X 5
 - h. Plastic drape (for trolley)
 - i. Large sterile drape
 - j. Sterile gloves various sizes
 - k. Benzoin compound
- 2 Wash hands and apply gloves
- 3 Apply Benzoin compound (Friars Balsalm) around wound margins (Fig 3).



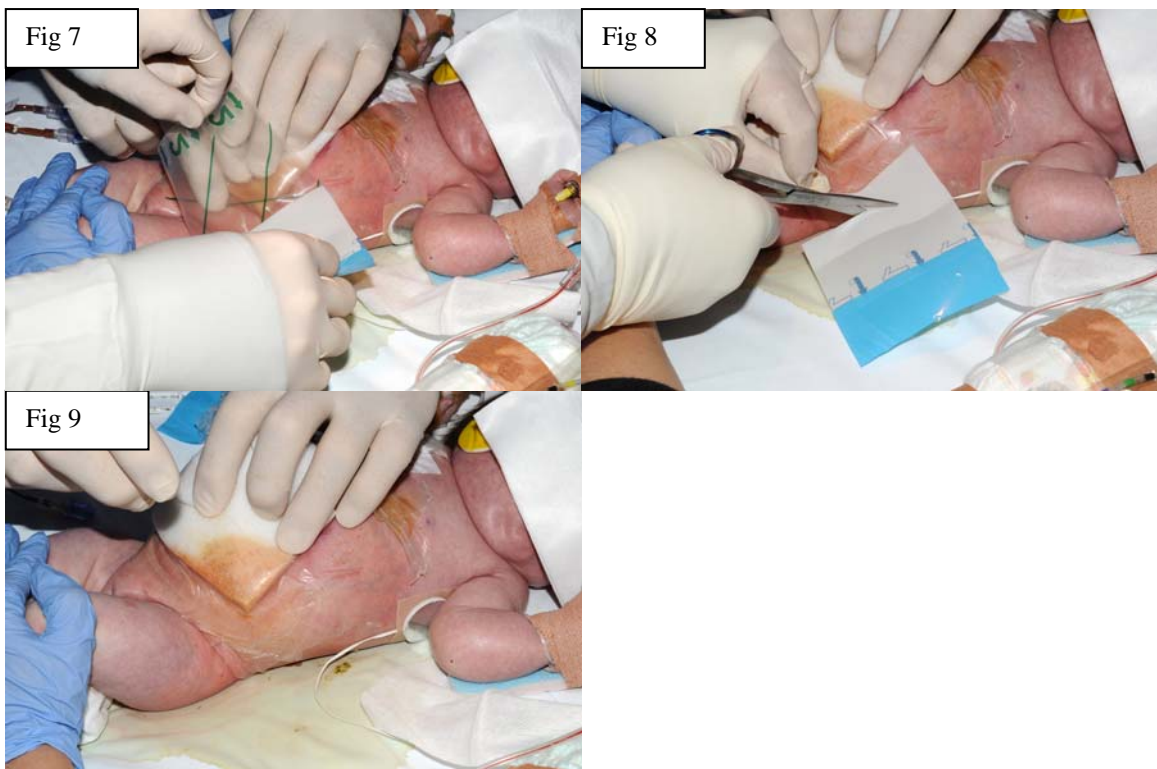
- 4 Apply VAC foam over exposed bowel. Other dressings may also be used under the foam dressing such as Acticoat, Atruman depending upon the surgeons' preference and the appearance of the bowel (Fig 4).



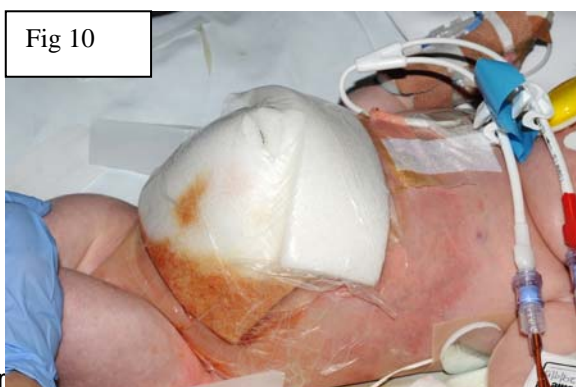
- 5 Apply Comfeel to wound margins to protect healthy skin (Fig 5,6)



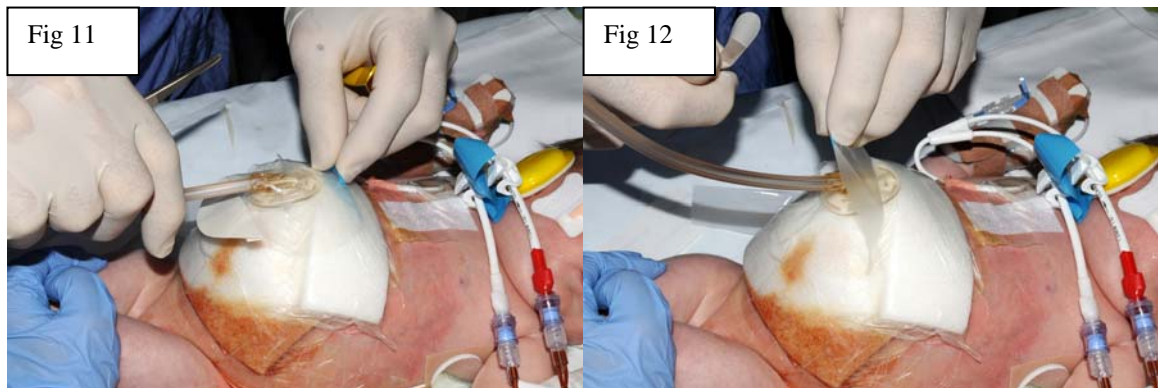
- 6 Ensure area around the wound is dry to help occlusive dressing stick
- 7 Attach occlusive VAC drape over the foam. It is easier to use strips of drape rather than one large piece to avoid wrinkling and unwanted sticking. (Fig 7, 8, 9)



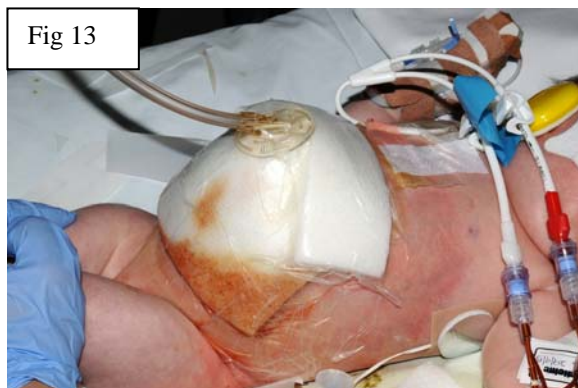
- 8 When area is completely covered with occlusive dressing, pierce a small hole in the occlusive dressing with sterile scissors. (Fig 10)



- 9 Attach the VAC suction piece over the small hole. (Fig 11, 12)



- 10 Connect suction and test to see if the dressing is sealing. If there is a leak, it needs to be found and repaired. (Fig 13)



- 11 Cover dressing with a crepe bandage (Fig 14)



Photographs and information provided by Kerry Sullivan Surgical CNC JHCH.