GASTROSCHISIS REPAIR

Surgical Specialty:	General Surgery
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Background:

- General Considerations
  - This abdominal wall defect results from occlusion of the omphalomesenteric artery at 7- 10 weeks of gestation, causing ischemia and atrophy of part of the abdominal wall with subsequent herniation of abdominal contents.
  - Incidence is 1/2000 births and seems to be increasing worldwide. It is more common in pregnancies in women less than 20 years old.
  - The degree of herniation can vary from minor to major amount of bowel and viscera.
  - Unlike omphalocele, the herniated contents of gastroschisis are not covered by membranes. The defect is usually at the right side of a normal umbilical cord.
  - Uncovered abdominal contents are exposed to amniotic fluid in utero, resulting in inflammation, edema, dilation, and functionally abnormal bowel.[1]
  - Exposed bowel and viscera post-birth increases susceptibility to extracellular fluid loss, hypothermia, and infection.
  - Herniated bowel is also at an increased risk for ischemia, intestinal malrotation, and obstruction.
  - Due to the considerations above, surgical repair is usually initiated as soon as possible within 24 hours of birth.[2]
- Patient Considerations
  - Premature delivery is more common due to the uncovered gut causing uterine irritation.[2]
  - Intrauterine growth restriction is common.
  - Some teams induce delivery as soon as signs of bowel compromise (thickening of the intraamniotic bowel) are present on weekly ultrasound surveillance: vaginal delivery is usually preferred to cesarian delivery.
  - Respiratory insufficiency may result from prematurity.
  - The patient may be hypovolemic due to increased insensible losses from exposed abdominal contents.
  - Additionally, the exposed viscera is a source of heat loss, leading to hypothermia.
  - Patients may experience gastrointestinal obstruction associated with herniation.

- Associated Comorbidities/Syndromes
    - Unlike omphalocele, gastroschisis is not usually associated with genetic, cardiac, or inherent metabolic abnormalities.
        - However, another malformation is present in up to 32% of cases: bowel atresia (often called "complex gastroschisis").
    - Patients may have a patent ductus ateriosus and a patent foramen ovale due to neonatal status and frequent premature delivery.

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Additional labs/tests indicated during work-up
        - CBC, especially hematocrit, electrolytes, BUN and creatinine, and glucose levels
        - Although associated congenital anomalies are not as common with gastroschisis as with omphalocele, it may be prudent to evaluate the following based on the clinical picture:
            - Echocardiogram: investigating for presence and direction of persistent fetal shunts, evaluating pulmonary pressures, and excluding any anomaly
            - Chest X-ray and abdominal X-ray
            - Renal ultrasound
            - Sacral/vertebral ultrasound
    - Obtain adequate intravenous access: peripheral IV for volume resuscitation, and possibly a Peripherally Inserted Central Catheter (PICC) for postoperative Total Parenteral Nutrition (TPN) administration.
    - Maintain adequate perfusion to herniated viscera.
        - Administration of full-strength, balanced salt solution.[2]
        - Prior to surgery in the NICU, patients are often administered a dextrose, amino acid, and saline solution at a greater than maintenance rate to counter third-space losses and support increased metabolic demands.[3] It is prudent to continue these fluids intraoperatively at a rate decreased by 30-50% and discontinue any associated intralipid infusions.[4]
        - Albumin 4-5% may be added in case of hypovolemia or hypotension
    - Reduction of fluid loss from exposed viscera
        - Mucosal surfaces are covered with warm saline-soaked gauze.
        - Placing the lower part of the body in a sterile plastic wrap immediately after birth to prevent evaporative loss, hypothermia, and bacterial contamination.
    - Discussions to have with surgeon/family
        - Code Status
        - Need for blood transfusion
        - Possibility for primary closure vs. staged reduction and closure.
- Specific or Unique Room Set-up Requirements
    - Airway
        - Patients may already be intubated in the NICU if being managed for respiratory insufficiency.
        - Otherwise, an orogastric tube should be placed to drain gastric contents prior to induction.
        - An appropriately sized cuffed or cuffless endotracheal tube (ETT). Consider that an ETT with a low-pressure leak may not provide an adequate seal if

higher inspiratory pressures are required for ventilation after the reduction of abdominal contents (decreased thoracic compliance).
        - A low compression volume anesthesia circuit should be used.
        - If the patient is intubated with controlled ventilation, and transfer to the OR is required, it may be advantageous to use an identical ventilator with similar settings to those in the NICU.
    - Drugs/Infusions
        - Several options are available for induction and maintenance, but institutional practices may vary.
            - Volatile maintenance, if surgery is performed in the OR.
                - Volatile anesthetic agents cause a dose-dependent decrease in cardiac output to a greater degree than in adults. Premature infants require even lower concentrations than otherwise term neonates.[4]
                - Avoid nitrous oxide to prevent bowel distension.
            - Narcotics: Fentanyl or any other synthetic opioid, morphine titrated to effect.
            - Neuromuscular blocking agents:
                - Rocuronium is used most commonly
    - Monitors
        - Standard ASA monitors
        - Pulse oximetry: preductal and postductal. A postductal SpO2 lower than the preductal SpO2 is indicative of a right to left shunt (pulmonary pressure higher than systemic pressure) at the level of the ductus arteriosus. Postductal oxygen saturations in the lower extremity may serve to monitor circulation to the lower extremities during abdominal wall closure.
        - Arterial line if present, will help guide fluid resuscitation and adequacy of perfusion, and will allow for intermittent monitoring of arterial blood gases.
        - Cerebral NIRS
        - Urine output aids as an indicator of adequate fluid resuscitation, keeping in mind that it is a late indicator of hypovolemia. For example, 1 mL/kg/h in a 3 kg neonate means 3 mL/h, which is barely measurable.
        - Intragastric pressure has been used to determine the feasibility of primary closure and avoid abdominal compartment syndrome. More to follow below.
    - Blood Availability (if indicated)
        - Usually not required.
    - NICU
        - Will require NICU for close monitoring and management.
        - Surgical correction is frequently performed at the bedside in the NICU.

Intraoperative Considerations:

- General
    - The patient will experience ongoing fluid and heat loss in the operating environment.
        - Maintain adequate fluid resuscitation while avoiding fluid overload.
        - Use measures to prevent heat loss.
    - Close monitoring for adequate perfusion using blood pressure, pulse oximetry, and urine output.
    - Maintain adequate muscle relaxation to facilitate the reduction of abdominal contents.

  - Intraabdominal pressures increase during abdominal wall closure: there is a risk of postoperative abdominal compartment syndrome.
- Induction
  - Rapid sequence induction vs. modified rapid sequence induction.
  - Ketamine or etomidate are better choices than propofol (risk of hypotension)
  - Avoid bag-mask ventilation with peak inspiratory pressure > 10-12 cm H2O to prevent bowel distension.
- Positioning
  - Supine
  - If the procedure is performed at the bedside in the NICU, access to the patient may be limited.
- Maintenance
  - Drugs as above
  - Cardiac output is heart rate dependent, so high concentrations of inhalational agents may not be tolerated. Opiate-based techniques may be beneficial.
  - Muscle relaxation is necessary to facilitate the reduction of visceral contents and abdominal closure.
  - Avoid nitrous oxide due to potential intestinal distension.
  - Retinopathy of prematurity: inspired oxygen guided by pulse oximetry to maintain at a preductal SpO2 95%[1-2,4].
- Hemodynamic/Physiologic goals
  - Avoid hypothermia
  - Continued fluid resuscitation without fluid overload
  - Maintain adequate perfusion to herniated abdominal contents.
  - Maintain adequate muscle relaxation throughout the reduction of abdominal contents and abdominal wall closure.
  - Maintain consistent peak inspiratory pressures and tidal volumes throughout the surgery.
  - Abrupt increases in peak pressures or decreases in volumes may indicate increased abdominal pressures.
- Surgical Considerations (such as neuromonitoring, muscle relaxation, anticipated blood loss)
  - Muscle relaxation is necessary to allow for visceral reduction and abdominal wall closure.
  - Intragastric pressure less than or equal to 20 mm Hg is conducive to primary closure of abdominal wall defect. For pressure above 20 mm Hg, placement of a silo to cover abdominal contents with delayed closure is warranted.
- Emergence/Disposition
  - Patients almost always require postoperative ventilatory support.
  - Prior to emergence, peak inspiratory pressures should be compared to baseline.
- Post-op Care
  - Patients should be monitored aggressively for infection/sepsis, abdominal compartment syndrome, and peripheral circulation status (pulse oximetry, blood pressure, urine output)

Case-Specific Complications/Pitfalls

- Infection/Sepsis
    - The defect is covered in sterile wet dressings to prevent infection until the time of operation.
    - Care must be taken to not cause twisting of the intestines thus compromising blood flow while transporting the patient to the operating room.
- Hypothermia
    - Exposure of large surface areas of the intestines can be a source of heat loss.
    - Heating the room to 80-85 degrees F (26.7-29.4 degrees C).
    - Careful and vigilant use of radiant warming units, often available in the NICU, or part of the NICU crib.
    - Use of forced-air warming devices.
    - Adding humidity to inspired gases.
- Ongoing fluid loss
    - Large exposed surfaces of the intestines lead to increased insensible losses.
    - Aggressive fluid replacement is essential.
- Increased intra-abdominal pressure
    - After the reduction of the intestines and abdominal closure, there is an increase in intra-abdominal pressure.
    - If intra-abdominal pressures are too high, this can lead to respiratory compromise, decreased perfusion to the abdominal organs and lower extremities, and decreased venous return and cardiac output.
    - Close monitoring of the peak inspiratory pressures and tidal volumes (depending on the ventilatory mode used) throughout the surgery, especially during abdominal wall closure, and close communication with the surgeon is essential to prevent this complication.
    - The use of an intragastric pressure monitor can aid guide whether primary abdominal wall closure or a staged repair is necessary.
- The postoperative course is often complicated by a slow return to normal bowel mobility, abdominal infections, or short-bowel syndrome requiring total or partial parenteral nutrition.

References

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