Congenital abdominal wall defects

Objectives: to study the neonatal abdominal wall defect, embryology, etiology, diagnosis, how to differentiate between omphalocele and gastroschisis and management.

Abdominal wall defects are divided into omphalocele and gastroschisis. The incidence in live birth is approximately 1 in 4000.

**Embryology:**

The abdominal wall forms during the 4th week of gestation from differential growth of the embryo causing infolding in the craniocaudally and mediolateral direction. The lateral abdominal folds of the embryo meet in the anterior midline and surround the yolk sac eventually constricting the yolk sac into yolk stalk that becomes the umbilical cord.

During the 6th week of gestation rapid growth of the intestine causes herniation of the midgut into the umbilical cord. Elongation and rotation of midgut occurs. By week 10, the midgut has returned to the abdominal cavity.

An omphalocele occurs if the intestine fails to return to the peritoneal cavity.

The etiology of gastroschisis less clear, many theories

1. Failure of mesoderm to form in the anterior abdominal wall.
2. Failure of lateral folds to fuse in the midline leaves a defect to the right side of the umbilicus.
3. Thrombosis of the right omphalomesenteric vein (umbilical vein) causing necrosis to the surrounding abdominal wall leading to right side defect.
4. Rapture of an omphalocele in utero
Diagnosis

- U/S these two conditions are often diagnosed on prenatal ultrasonography and are easily differentiated by the location of the defect and by the presence or absence of a sac surrounding the eviscerated bowel.
- Elevation of maternal α-feto protein

Gastroschisis:

Perinatal care:

Due to prolong exposure of the bowel of a neonate with gastroschisis to the damage effect of amniotic fluid, bowel edema, poor motility and malabsorption is noticed significantly.

The delivery for fetuses with gastroschisis is better to be carried in tertiary perinatal center so as to provide immediate neonatal and pediatric surgical experience.
Neonatal resuscitation and management:

- Intravenous fluid resuscitation, the neonates with gastroschisis have significant evaporative water loss from the open abdominal cavity and exposed bowel.
- Nasogastric decompression to prevent gastric and intestinal distension.
- The herniated bowel should be wrapped in warm saline soaked gauze.
- The infant placed with the bowel and legs in a plastic bag to reduce evaporative losses.
- Although gastroschisis most often is an isolated anomaly, thorough examination of the neonate should be done to exclude the coexistence of other congenital anomalies.

Surgical management:

The primary goal is to return the viscera to the abdominal cavity while minimizing the risk of damage to the viscera due to direct trauma or to increased intra abdominal pressure.

**Options**

Primary closure

- Is practiced for neonate when reduction of the herniated viscera is thought to be possible
- Or using the prosthetic options when primary fascial closure cannot be achieved, non-absorbable mesh or bioprosthetic material such as dura or porcine small intestinal sub mucosa can be used.

Staged closure

Consist of placing the bowel into a prefabricated silo with a circular spring that is positioned under fascial opening. Can be inserted at delivery room or bedside without general anesthesia, the bowel is
reduced once or twice daily into the peritoneal cavity, when the reduction is completed the definitive closure can be performed. This process takes between 1-14 days. This procedure is used to avoid ischemic injury to the viscera due to the high intra-abdominal pressure.

**Postoperative course**

- Enteral feeding is delayed for few weeks while awaiting return of bowel function.
- Nasogastric decompression.
- Parenteral nutrition.
- Prokinetics for treatment of GIT dysmotility e.g erythromycin, metoclopramide, domperidone, cisapride.

The long term outcomes for patients born with gastroschisis are generally excellent. The presence of bowel atresia is the most important prognostic determinant for poor outcome.

**Omphalocele**

**Perinatal care**

- Mode of delivery should be decided by the obstetrician. But in giant omphalocele cesarean section is preferable because of the fear of liver injury.
- Delivery at a tertiary center is preferable for immediate access to neonatal and pediatric surgical expertise.

**Neonatal resuscitation and management**

- A thorough search for associated anomalies should be done,(cardiac evaluation ,renal, neonatal hypoglycemia for possibility of Bechwith-
Weidemann syndrome, and blood sample for genetic evaluation if indicated.

- Intravenous access and fluid resuscitation, infants with omphalocele do not have as significant fluid and temperature losses as those with gastroschisis but the loss are higher than those with intact abdominal wall.
- The omphalocele itself can be dressed with saline soaked gauze to minimize those losses.
- Nasogastric tube.

**Surgical management**

Treatment options depend on the size of the defect, gestational age and the presence of associated anomalies.

**Immediate primary closure:** is used in infants with small defects or larger but still easy to close.

**Staged neonatal closure:** this method utilize the existing amnion sac with serial inversion, or the sac is excised and replaced with mesh and then closed over time.

**Delayed staged closure:**

Used for large omphalocele, the sac is excised and a silastic silo is used with serial reduction similar to that of gastroschisis.

**Scarification treatment**

Non operative technique is used for neonates who can't tolerate operation or the defect is too large that initial repair will result in potential life-threatening abdominal compartment syndrome. So an agent is used that allows an eschar to develop over the intact amnion sac. This eschar will epithelialize over time leaving ventral hernia that require repair later in life. Iodine, silver sulfadiazine, and neomycin ointment is used.
Postoperative course

- Most of patients require mechanical ventilation after primary closure for few days.
- Nasogastric tube for gastric decompression.
- Feeding can begin when nasogastric tube output is no longer bilious and bowel activity has occurred.
- Antibiotics for 48 hours (if there is no infection).
- If a hernia develops, closure usually can be done after age of one year.

Post-operative complications

1. Increase intra abdominal pressure
2. Acute hepatic congestion
3. Renal failure
4. Bowel infarction