

Omphalocele

What is an Omphalocele?

An Omphalocele is a congenital defect in which the abdominal (belly) wall at the umbilicus (belly button) fails to close. The defect allows the abdominal organs including the intestine, liver and bladder, to protrude out of the abdomen. The organs are usually contained within a thin membranous sac. Omphalocele occurs once in every 5000 live births.

When is it Diagnosed?

An Omphalocele is usually detectable by prenatal ultrasonography. Early detection allows ample time for the parents to have detailed discussions with the obstetrician or perinatologist and a pediatric surgeon. A prenatal cardiac ultrasound may be obtained by a pediatric cardiologist.

What Causes an Omphalocele?

During normal embryologic development the intestine protrudes into a sac at the level of the umbilicus (belly button). The intestine rotates and returns into the abdominal cavity. For reasons that are unclear, the intestine occasionally remains within the sac outside of the abdominal cavity, resulting in Omphalocele. Depending on the size of the defect, the intestine may be accompanied by a part of the liver (commonly) or bladder (rarely).

Are There Associated Anomalies?

Omphalocele is associated with other anomalies approximately 50% of the time. These defects are primarily in the cardiac and urinary systems. Nearly all newborns with Omphalocele will have malrotation, abnormal position of the intestines, which is rarely a problem in this scenario. Many of the babies also have respiratory problems and may require a ventilator. There may be other defects including neurologic and chromosomal anomalies, which will be discussed with you as needed.

What is the Treatment for an Omphalocele?

- **Scheduled delivery:** this helps facilitate care, minimizes the risk of infection, and maintains integrity of the sac which surrounds the abdominal organs. Long before delivery, conferences with the pediatric surgeon, perinatologist, and neonatologist allow parents to become acquainted with their baby's future caregivers, understand the baby's problems and potential treatments, and to answer any specific concerns.
- **Immediately after birth:** saline-soaked sponges are applied to the defect and covered with plastic wrap, minimizing contamination and preventing heat loss. A naso-gastric tube, a tube from the nose to the



stomach, is passed to prevent secretions and air from entering the intestine, and to prevent vomiting. An intravenous line is placed and antibiotics started.

- **Neonatal intensive care unit:** after birth, the baby will be fully evaluated by the neonatologist, pediatric surgeon, and other consultants. A pediatric cardiologist generally performs an ultrasound of the heart to determine whether there is any associated heart anomalies. An ultrasound of the abdomen is performed to look for a possible urinary anomaly. It is not critical that the baby undergo immediate surgery. Rather, it is more important to find any additional anomalies and problems that may affect the risks of anesthesia and surgery.

How is an Omphalocele Surgically Corrected?

- **Small Omphalocele:** the sac is removed, the intestine returned into the abdominal cavity and the abdominal wall is reconstructed.
- **Large and Giant Omphalocele:** can contain a significant amount of liver and Intestine, where returning the organs into the abdomen will cause respiratory compromise. If this occurs the sac can be covered with a solution to make it thick and act as a cover for the organs. This technique is known as “paint and wait”. This allows for the organs to be placed back into the abdomen at a later time. Once the abdominal organs have been returned into the abdomen, the baby is taken to the operating room for final closure of the abdominal wall. In some cases, a silo may be placed to gradually reduce the contents back into the abdomen until enough space is made for closure of the abdominal wall
- **In the face of infection,** the skin may be mobilized and used as a covering for the intra-abdominal organs. Once the infection is controlled (several weeks), the defect needs to be repaired by one of the above means. If it were to be left alone, the muscle would slowly retract as the abdominal organs protrude into the skin sac which stretches over time. This could lead to many chronic problems.

Post-operative Complications and Long Term Problems:

- **Ileus:** If the omphalocele requires multiple stages to close, there may be a prolonged ileus (intestine does not function properly) requiring intravenous nutritional support. Such support, while necessary, may cause liver problems if needed for a prolonged period of time. This will be discussed with you by the neonatologists.
- **Central Line Infection:** The central catheter to administer the intravenous solutions may become infected, requiring treatment with antibiotics and sometimes removal of the catheter. The use of any synthetic material incurs the risk of infection.
- **Incisional Hernias:** There is an approximately 10% incidence of separation of the abdominal wall or incisional hernias that will need to be surgically repaired in the future because the abdomen is often closed under tension.
- **Gastroesophageal Reflux:** When the liver protrudes into the defect, gastroesophageal reflux is not uncommon. While most reflux can be managed medically, occasionally surgery is needed for correction.
- **Malrotation:** As mentioned earlier, because the intestine protrudes into the sac, it does not rotate properly and is not fixed in a normal position. As a consequence, the appendix may not end up in the right lower quadrant. Parents should always bear in mind, as the child grows, that appendicitis may not present with the typical location of abdominal pain. Parents should always mention the history of omphalocele to the child’s caregivers.
- Very large omphaloceles with a significant amount of liver involved may require many operations over months or even years.

Survival of neonates with omphalocele is 90% when there are no other associated anomalies. The survival rate with other anomalies varies significantly depending on the other anomalies.

Disclaimer: Your child's condition is unique. The information contained on this web site is not intended to substitute for advice from a doctor or nurse. If you are unsure about any aspect of your patient's care, please contact us at 303-839-6001, or talk to your pediatrician.

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