

## Patient Care > Omphalocele

### Introduction

**An omphalocele is a congenital defect in which the abdominal (belly) wall at the umbilicus (belly button) fails to close** (see [Embryology/Pathology](#), below). The defect allows the abdominal organs (intestine, liver, bladder), usually contained within a thin membranous sac, to protrude out of the abdomen. Omphalocele occurs once in every 5000 live births.

### Antenatal Diagnosis:

**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 pediatric surgeon. A prenatal cardiac ultrasound may be obtained by a pediatric cardiologist.

### Embryology/Pathology:

**During normal embryologic development the intestine (bowel) protrudes into a sac at the level of the umbilicus.** The intestine undergoes a set rotation and returns into the abdominal or body cavity where it continues its normal rotation. For reasons that are unclear, the intestine occasionally remains within the sac outside of the abdominal cavity, resulting in omphalocele. Depending on whether the defect is high or low on the abdominal wall, the intestine may be accompanied by the liver (commonly) or bladder (rarely).

### Associated Anomalies:

**Omphalocele is associated with other anomalies approximately 30% of the time.** These defects are primarily in the cardiac and urinary systems. Nearly all newborns with omphalocele will have malrotation of the intestine as the intestine has not returned into the abdominal cavity and undergone its normal rotation. Many of the babies also have altered pulmonary mechanics, and may require prolonged mechanical support (ventilator). There may be other defects which will be discussed with you by the neonatologist, pediatric surgeon, or other newborn specialists.

### Treatment:

**Scheduled delivery for the baby facilitates 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 care givers, to understand the baby's problems and potential treatments, and to answer any specific concerns. In this way, we hope to allay as much anxiety as possible.

**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 is passed into the stomach to prevent secretions and air from entering the intestine, and to prevent vomiting and aspiration of stomach contents into the lungs. An intravenous line is placed and antibiotics started.

**In the neonatal intensive care unit**, 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 anomaly. An ultrasound of the abdomen may be performed if a urinary anomaly is suspected. 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.

**Once the newborn is cleared for surgery**, the baby is moved to the operating room and one of the following is performed:

**for small omphaloceles**, the sac is removed, the intestine returned into the abdominal cavity (an appendectomy may or may not be performed), the abdominal wall reconstructed, and the anesthesiologist is consulted regarding the effect of increased intra-abdominal pressure on ventilation.

**for large omphaloceles** which may contain a significant amount of liver and intestine, whose return will cause respiratory compromise, a sac of synthetic material (often called a silo) is sewn to the abdominal wall surrounding the intestine and liver. Antibiotic ointments and solutions will cover the synthetic sac to minimize the risk of infection. Then every day or every other day the sac is made smaller allowing the intestine and liver to return gradually into the abdomen and to allow the abdominal cavity to enlarge. When the abdominal organs have been returned into the abdomen, the baby is taken back to the operating room and the abdominal wall closed.

**For giant omphaloceles** whose protruded organs cannot be returned into the abdomen, synthetic material may be used to stabilize the abdominal wall enclosing the organs. The skin is mobilized and used to cover the synthetic material. Depending on the size of the synthetic material used, it may be removed later or left in place.

**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 the abdominal wall is not stabilized, there is no stimulus for it to grow together. In fact, it tends to retract as the abdominal organs protrude into the skin sac which stretches over time. This leads to many chronic care problems.

### **Post-operative Complications and Long Term Problems:**

**Any baby undergoing general anesthesia and surgery has the risk of reaction to anesthetic agents, bleeding, infection, and bowel obstruction from scarring.** If the omphalocele requires multiple stages to close, there may be a prolonged ileus (bowel does not move) requiring intravenous nutritional

support. Such support, while necessary, may cause liver problems. This will be discussed with you by the neonatologists. The central catheter to administer the intravenous solutions may become infected. The use of any synthetic material incurs the risk of infection. Because the abdomen is closed under a variable amount of tension, there is approximately 10% incidence of incisional hernias and separations of the abdominal wall that will need to be surgically repaired. When the liver protrudes into the defect, the orientation of the diaphragm (muscle separating the chest and abdomen) is changed and gastroesophageal reflux is not uncommon. While most reflux can be managed medically, occasionally surgery is needed for correction. As mentioned earlier, because the intestine protrudes into the sac it does not rotate and fixate properly. As a consequence, the appendix may not end up in the right lower quadrant. If the appendix is not removed at surgery, the parents should always bear in mind as the child grows that abdominal pain anywhere in the abdomen may be the first sign of appendicitis and the child's care giver should be so informed.

**Very large omphaloceles with a significant amount of liver involved with tax both the parents and surgeon.** Some may require many operations over months or even years.

**Survival of neonates with omphaloceles is well over 90%.** The majority of babies who do not do well have significant associated 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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