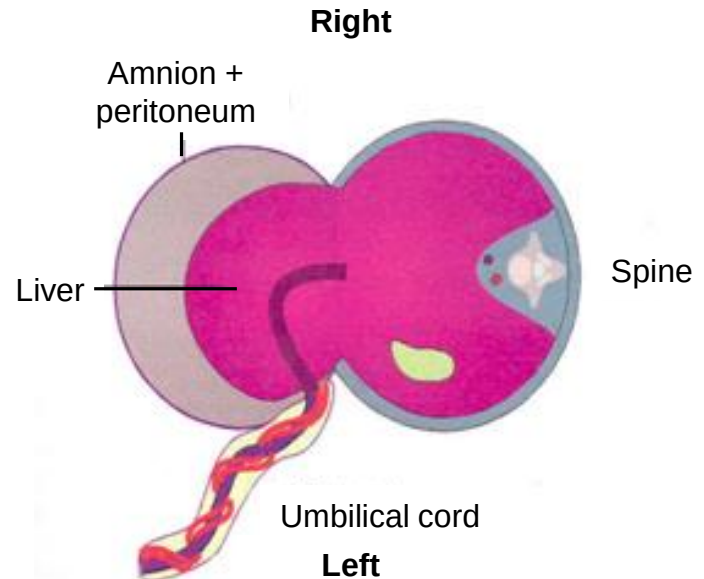
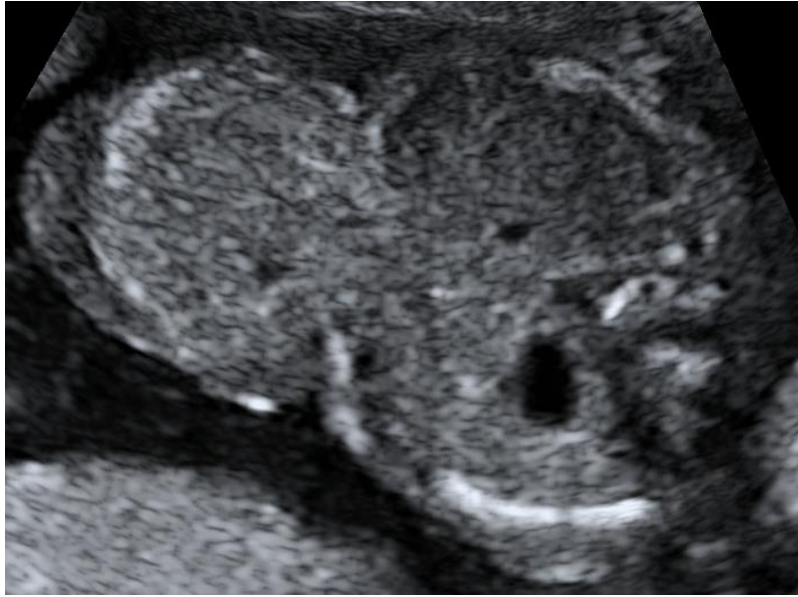


Omphalocele

Definition and incidence



An omphalocele is a congenital abdominal wall defect characterized by herniation of the abdominal viscera through the abdominal wall at the umbilicus. The protruding abdominal organs are covered by a two-layered membrane composed of amnion and peritoneum into which the umbilical cord inserts.

An omphalocele occurs in 3.4 of 10.000 pregnancies and 1-2 per 10.000 live births.

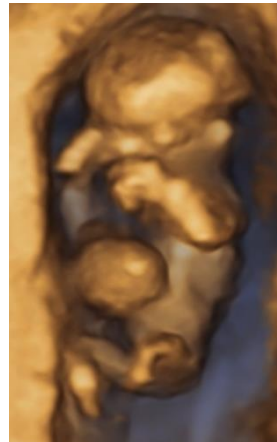
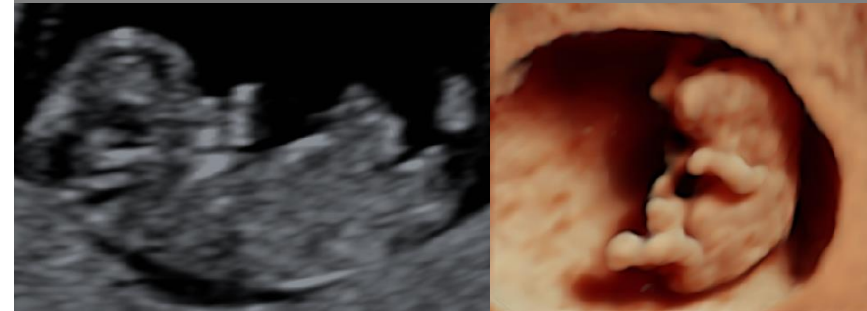
Omphalocele

Pathogenesis

At eight weeks gestational age the abdominal cavity is too small for its contents and the bowel protrudes the abdomen at the level of the umbilical cord insertion. This herniation is a temporary physiological midgut herniation which is visible on ultrasound from 9-12 weeks gestational age.



9 weeks' gestation



If this herniation is present after 12 weeks gestational age it is no longer physiological and defined as an omphalocele. If the liver protrudes into the base of the umbilical cord at any gestational age, the herniation is considered pathological.

Omphalocele Pathology

Omphaloceles can be divided into small and giant omphaloceles. A small (or minor) omphalocele can often be closed primarily, typically within 48 hours after birth.

13 weeks' gestation



20 weeks' gestation



30 weeks' gestation



If the postnatal defect size equals or is larger than 5 cm, with liver (partly) protruding, closure is usually delayed due to viscerocranial disproportion. Infants with these 'giant' omphaloceles are at risk for pulmonary hypoplasia, feeding problems and prolonged hospital stay.

12 weeks' gestation



20 weeks' gestation

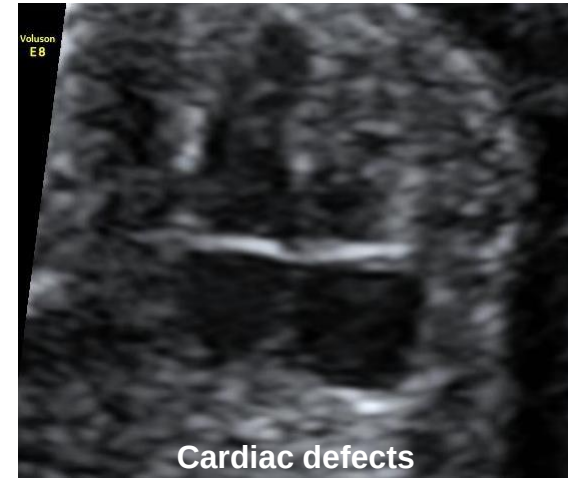


30 weeks' gestation



Omphalocele

Associated anomalies



In up to 80% of fetuses with omphalocele associated anomalies are present, including structural anomalies and syndromes like Beckwith-Wiedemann syndrome. When additional abnormalities are present, chromosome abnormalities, including trisomy 18 are common.

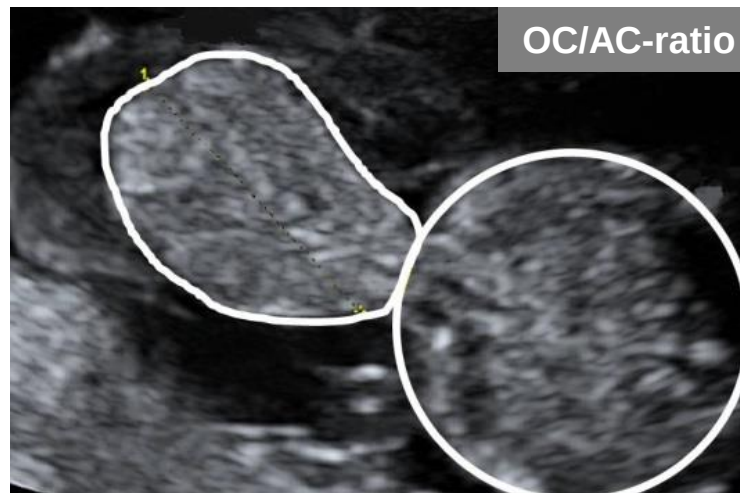
The most common associated abnormalities are cardiovascular abnormalities, followed by gastrointestinal and urogenital anomalies.

Omphalocele

Diagnosis

When an omphalocele is suspected, a thorough search for associated anomalies is recommended. Also follow-up scans to check for fetal growth, amount of amniotic fluid and calculation of the OC/AC-ratio are recommended.

Evaluation of organomegaly of the liver, kidney and tongue to screen for Beckwith-Wiedemann syndrome is suggested. The OC/AC-ratio is measured by dividing the omphalocele circumference by the abdominal circumference at the level of the defect with exclusion of edema. An OC/AC-ratio in second trimester of 0.66 or higher may predict delayed closure of the defect after birth (unpublished data).



Omphalocele Management

Type of postnatal surgical closure consists of a primary or a delayed closure.

Primary closure is mostly done within 48 hours after birth for neonates with a small omphalocele (left)

Delayed closure (below) includes initial epithelization and later surgical closure. In infants with a 'giant' omphalocele (postnatal defects size $>5\text{cm}$ and liver herniation) a delayed closure may be performed



Omphalocele

Prognosis

Neonates with isolated omphaloceles have a good prognosis, and up to 96% survive. Depending on the type and number of associated anomalies, survival rates decrease to 14%.

Infants with a giant omphalocele are at risk for chronic lung disease, feeding problems, prolonged hospital stay, a lower chance of survival, and difficulties with surgical closure of the defect.

Mental function is typically normal. Motor delays at 2 years of age are more common in children with giant omphalocele compared to those with a small omphalocele.