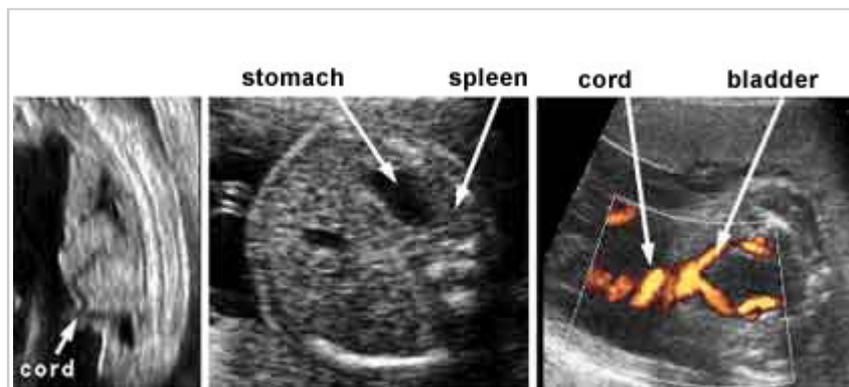


Chapter 6

Anterior abdominal wall

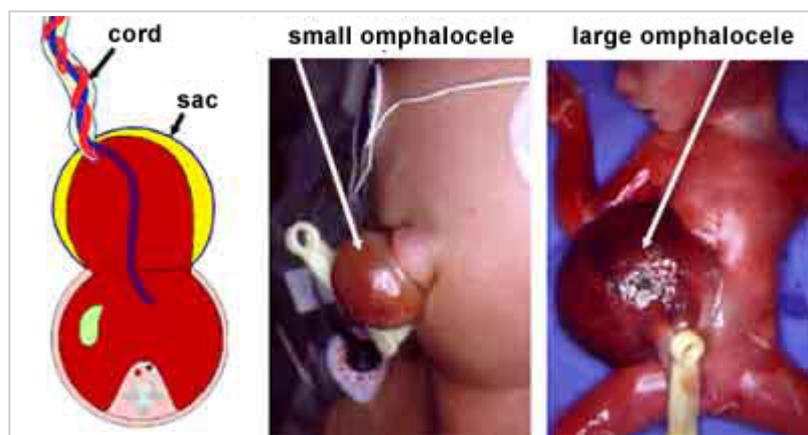
NORMAL SONOGRAPHIC ANATOMY

Normal development of the anterior abdominal wall depends on the fusion of four ectomesodermic folds (cephalic, caudal and two lateral). At 8–10 weeks of gestation, all fetuses demonstrate herniation of the mid-gut that is visualized as a hyperechogenic mass in the base of the umbilical cord; retraction into the abdominal cavity occurs at 10–12 weeks and is completed by 11 weeks and 5 days. The integrity of the abdominal wall should always be demonstrated; this can be achieved by transverse scans demonstrating the insertion of the umbilical cord. It is also important to visualize the urinary bladder within the fetal pelvis, because this rules out exstrophy of the bladder and of the cloaca.



EXOMPHALOS

Exomphalos results from failure of normal embryonic regression of the mid-gut from the umbilical stalk into the abdominal celom. The abdominal contents, including intestines and liver or spleen covered by a sac of parietal peritoneum and amnion, are herniated into the base of the umbilical cord.



Less often there is an associated failure in the cephalic embryonic fold, resulting in the pentalogy of Cantrell (upper mid-line omphalocele, anterior diaphragmatic hernia, sternal cleft, ectopia cordis and intracardiac defects) or failure of the caudal fold, in which case the omphalocele may be associated with exstrophy of the bladder or cloaca, imperforate anus, colonic atresia and sacral vertebral defects. The Beckwith–Wiedemann syndrome (usually sporadic and occasionally familial syndrome with a birth prevalence of about 1 in 14 000) is the association of omphalocele, macrosomia, organomegaly and macroglossia; in some cases there is mental handicap, which is thought to be

secondary to inadequately treated hypoglycemia. About 5% of affected individuals develop tumors during childhood, most commonly nephroblastoma and hepatoblastoma.

Prevalence

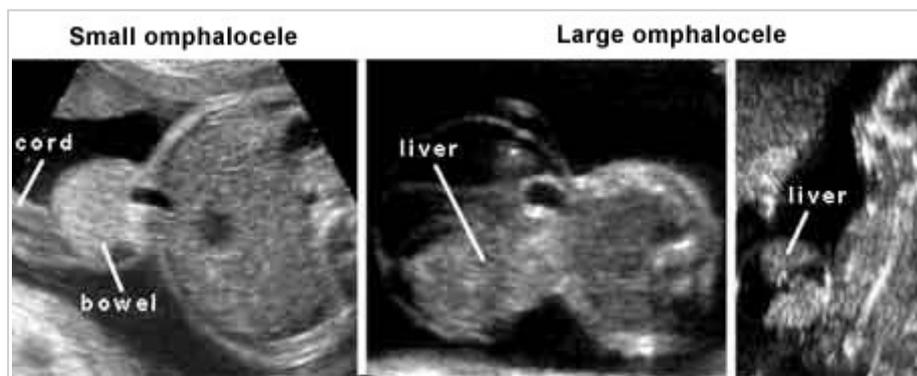
Exomphalos is found in about 1 per 4000 births.

Etiology

The majority of cases are sporadic and the recurrence risk is usually less than 1%. However, in some cases, there may be an associated genetic syndrome. Chromosomal abnormalities (mainly trisomy 18 or 13) are found in about 50% of cases at 12 weeks, 30% of cases at mid-gestation and in 15% of neonates. Similarly, in Beckwith-Wiedemann syndrome, most cases are sporadic, although autosomal dominant, recessive, X-linked and polygenic patterns of inheritance have been described.

Diagnosis

The diagnosis of exomphalos is based on the demonstration of the mid-line anterior abdominal wall defect, the herniated sac with its visceral contents and the umbilical cord insertion at the apex of the sac. Ultrasonographic examination should be directed towards defining the extent of the lesion and exclusion of other malformations.

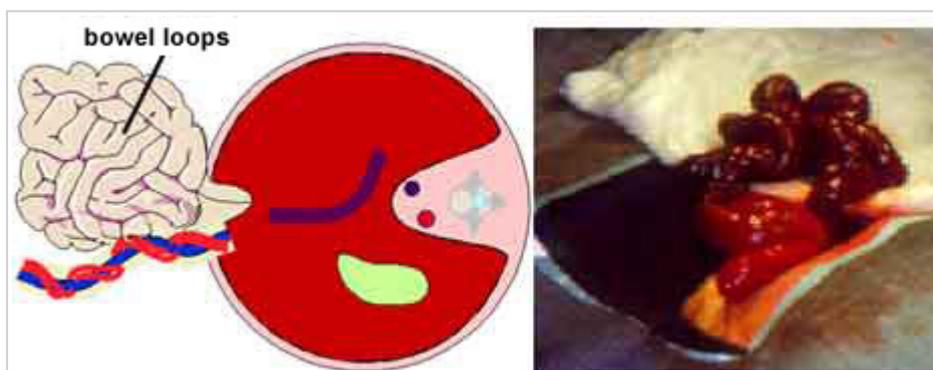


Prognosis

Exomphalos is a correctable malformation in which survival depends primarily on whether or not other malformations or chromosomal defects are present. For isolated lesions, the survival rate after surgery is about 90%. The mortality is much higher with cephalic fold defects than with lateral and caudal defects.

GASTROSCHISIS

In gastroschisis, the primary body folds and the umbilical ring develop normally and evisceration of the intestine occurs through a small abdominal wall defect located just lateral and usually to the right of an intact umbilical cord. The loops of intestine lie uncovered in the amniotic fluid and become thickened, edematous and matted.



Prevalence

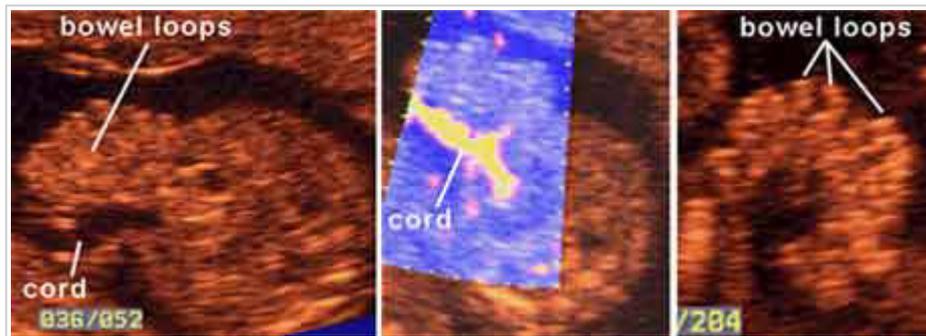
Gastroschisis is found in about 1 per 4000 births.

Etiology

This is a sporadic abnormality. Associated chromosomal abnormalities are rare, and, although other malformations are found in 10–30% of the cases, these are mainly gut atresias, probably due to gut strangulation and infarction in utero.

Diagnosis

Prenatal diagnosis is based on the demonstration of the normally situated umbilicus and the herniated loops of intestine, which are free-floating and widely separated, and usually on the right of the cord insertion. In the midtrimester, the bowel loops are uniformly echogenic.



In the third trimester, chemical peritonitis causes distension and thickening of the walls of the intestine.



About 30% of fetuses are growth-restricted but the diagnosis can be difficult because gastroschisis as such is associated with a small abdominal circumference.

Prognosis

Postoperative survival is about 90%; mortality is usually the consequence of short gut syndrome. In this condition, the infants require total parenteral nutrition and they usually die within the first 4 years of life from liver disease.

BODY STALK ANOMALY

This abnormality is characterized by the presence of a major abdominal wall defect, severe kyphoscoliosis and a rudimentary umbilical cord.

Prevalence

Body stalk anomaly is found in about 1 per 10 000 pregnancies.

Etiology

This is a sporadic abnormality. The pathogenesis is uncertain but possible causes include abnormal folding of the trilaminar embryo during the first 4 weeks of development, early amnion rupture with amniotic band syndrome, and early generalized compromise of embryonic blood flow.

Diagnosis

The ultrasonographic features are a major abdominal wall defect, severe kyphoscoliosis and a short or absent umbilical cord. Typically, the liver is directly attached to the placenta without an interposed umbilical cord and there is major distortion of the spine.



In the first trimester, it is possible to demonstrate that part of the fetal body is in the amniotic cavity and the other part is in the celomic cavity. The findings suggest that early amnion rupture before obliteration of the celomic cavity is a possible cause of the syndrome.

Prognosis

This is a lethal abnormality.

BLADDER EXSTROPHY AND CLOACAL EXSTROPHY

Bladder exstrophy is a defect of the caudal fold of the anterior abdominal wall; a small defect may cause epispadias alone, whilst a large defect leads to exposure of the posterior bladder wall. In cloacal exstrophy, both the urinary and gastrointestinal tracts are involved. Cloacal exstrophy (also referred to as OEIS complex) is the association of an omphalocele, exstrophy of the bladder, imperforated anus, and spinal defects such as meningomyelocele. The hemibladders are on either side of the intestines.

Prevalence

Bladder exstrophy is found in 1 per 30 000 births and cloacal exstrophy is found in about 1 in per 200 000 births.

Etiology

Both bladder exstrophy and cloacal exstrophy are sporadic abnormalities.

Diagnosis

Bladder exstrophy should be suspected when, in the presence of normal amniotic fluid, the fetal bladder is not visualized (the filling cycle of the bladder is normally in the range of 15 min); an echogenic mass is seen protruding from the lower abdominal wall, in close association with the umbilical arteries.

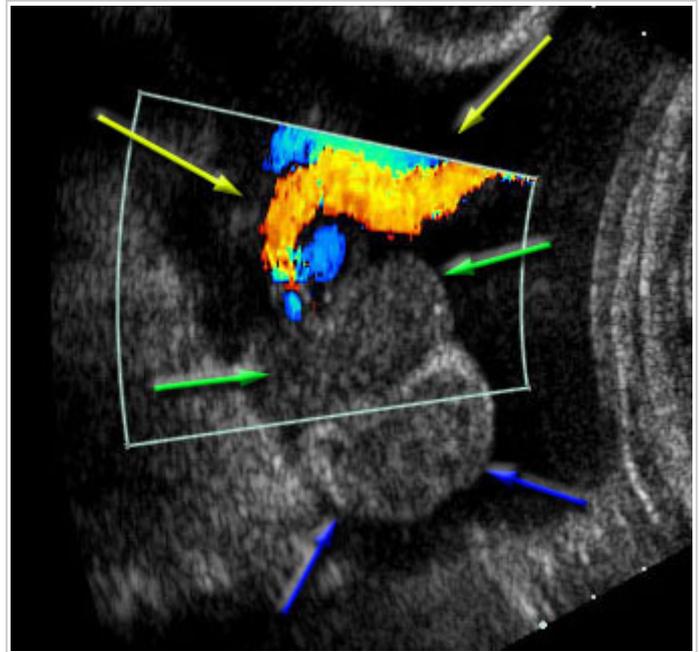
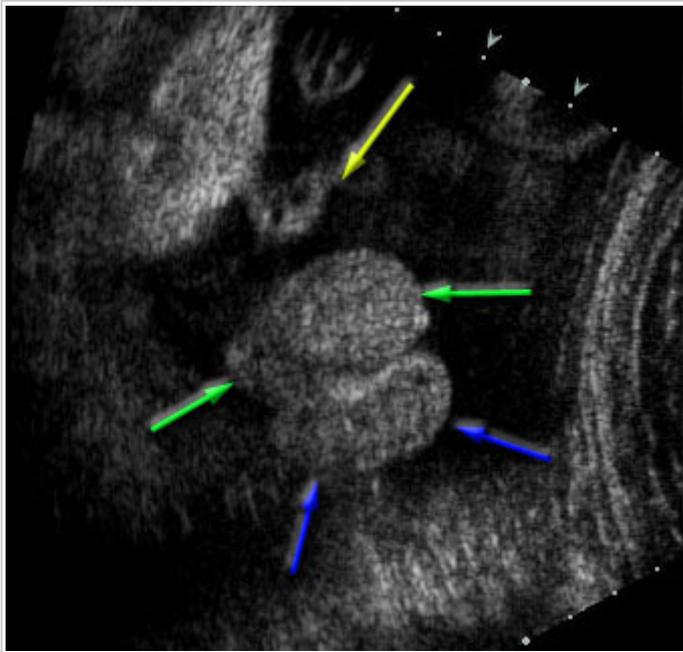
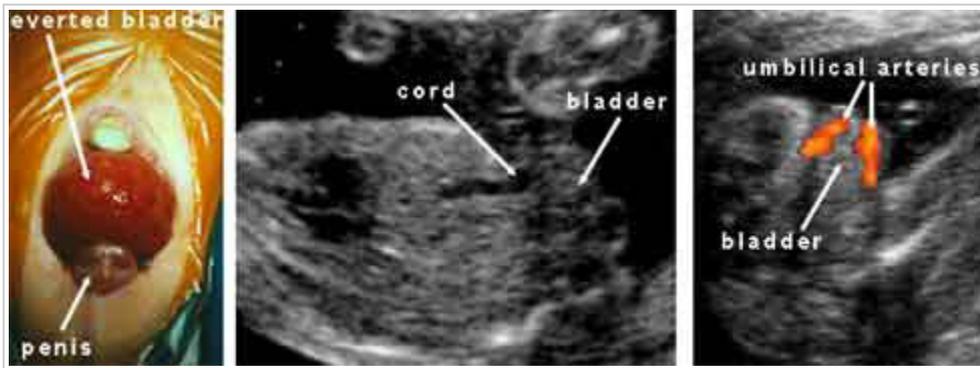


Figure 01
Bladder Exstrophy and absence of the penis - an echogenic mass is seen protruding from the lower abdominal wall, in close association with the umbilical arteries (Color Doppler).
green arrows: everted bladder; blue arrows the scrotum; yellow arrows: umbilical cord

Figure 02 with color doppler



Bladder Exstrophy and absence of the penis
green arrows: everted bladder; blue arrows the scrotum; yellow arrows: umbilical cord

In cloacal exstrophy, the findings are similar to bladder exstrophy (large infraumbilical defect that extends to the pelvis), but a posterior anomalous component (represented by herniated bowel and/or a meningocele) is present. Other findings include single umbilical artery, ascites, vertebral anomalies, club foot and ambiguous genitalia (in boys, the penis is divided and duplicated).

Prognosis

With aggressive reconstructive bladder, bowel and genital surgery, survival is more than 80%. Although it has been suggested that gender re-assignment to females should occur, psychological follow-ups of such patients suggest that both male and females with this condition are capable of a normal lifestyle with normal intelligence, although some form of urinary tract diversion is required for all. Furthermore, both sexes have been reported to be fertile after surgery.