

Ultrasound-based differential diagnosis of fetal abdominal wall defects in early pregnancy

Mircea-Octavian
Poenaru^{1,2},
Delia-Maria
Bogheanu²,
Romina-Marina
Sima^{1,2},
Liana Pleș^{1,2}

1. "Carol Davila" University
of Medicine and Pharmacy,
Bucharest, Romania

2. "Sf. Ioan" Emergency
Hospital,
"Bucur" Maternity,
Bucharest, Romania

Corresponding author:
Romina-Marina Sima
E-mail: romina.sima@umfcd.ro

Abstract

Fetal abdominal wall defects (AWDs) refer to a variety of congenital anomalies characterized by the incomplete closure or disruption of the abdominal wall in the early stages of embryonic development. Based on the 2011 European Surveillance of Congenital Anomalies (EUROCAT) report, the general prevalence of fetal abdominal wall defects is documented to be six cases per 10,000 births. AWDs pose significant challenges for prenatal diagnosis and management. Ultrasound has emerged as an indispensable tool in diagnosing and characterizing various abdominal wall defects early in pregnancy, enabling precise prenatal counseling and appropriate antenatal management strategies. This article provides a general review of the specialized literature regarding various types of abdominal wall defects, such as omphalocele, gastroschisis, pentalogy of Cantrell, bladder exstrophy, and limb body wall complex, highlighting the significant sonographic features observed during ultrasound examination. Additionally, differential diagnoses are explored, emphasizing the importance of accurately distinguishing between these conditions to ensure optimal management strategies. This article aims to enhance the understanding and familiarity of ultrasound practitioners and obstetricians with the spectrum of fetal abdominal wall defects and their prenatal sonographic appearance, thus facilitating improved prenatal care and patients' counseling.

Keywords: ultrasound, prenatal diagnosis, abdominal wall defects, fetal anomalies, antenatal management

Rezumat

Defectele fetale ale peretelui abdominal se referă la o varietate de anomalii congenitale caracterizate de închiderea incompletă a peretelui abdominal în stadiile timpurii ale dezvoltării embrionare. Conform raportului European Surveillance of Congenital Anomalies (EUROCAT) din 2011, prevalența globală a defectelor fetale ale peretelui abdominal este de șase cazuri la 10000 de nașteri. Defectele fetale ale peretelui abdominal reprezintă o provocare semnificativă în ceea ce privește diagnosticul prenatal și managementul acestora. Ecografia este un mijloc indispensabil în diagnosticarea și caracterizarea diferitelor defecte ale peretelui abdominal în stadiile timpurii ale sarcinii, permițând consilierea prenatală precisă și strategiile adecvate de management antenatal. Acest articol prezintă o analiză a literaturii de specialitate referitoare la diferitele tipuri de defecte ale peretelui abdominal, cum sunt omfalocelul, gastroschizisul, pentalogia lui Cantrell, extrofia vezicii urinare și complexul de membru-corp-perete, evidențiind caracteristicile semnificative sonografice observate în timpul examinării cu ultrasunete. În plus, sunt explorate diagnosticile diferențiale, subliniind importanța de a distinge cu precizie între aceste condiții pentru a asigura strategii optime de management. Acest articol își propune să îmbunătățească înțelegerea și familiarizarea cu spectrul defectelor fetale ale peretelui abdominal fetal și cu aspectul lor sonografic prenatal, facilitând astfel îngrijirea prenatală și consilierea pacienților.

Cuvinte-cheie: ecografie, diagnostic prenatal, defecte ale peretelui abdominal, anomalii fetale, management prenatal

Submission date:
28.02.2024
Acceptance date:
6.03.2024

Diagnosticul diferențial ecografic al defectelor de perete abdominal fetal în primul trimestru de sarcină

Suggested citation for this article: Poenaru MO, Bogheanu DM, Sima RM, Pleș L. Ultrasound-based differential diagnosis of fetal abdominal wall defects in early pregnancy. *Ginecologia.ro*. 2024;43(1):30-35.

Introduction

Fetal abdominal wall defects (AWDs) encompass a wide range of congenital anomalies that involve the incomplete closure or disruption of the abdominal wall during the embryonic development.

According to the 2011 European Surveillance of Congenital Anomalies (EUROCAT) report, which analyzed congenital anomalies in Europe from 1980 to 2008, the overall prevalence of fetal abdominal wall defects is reported to be six cases per 10,000 births⁽¹⁾. The most frequently encountered fetal abdominal wall defects are gastroschisis and omphalocele⁽²⁾. Other AWDs include

pentalogy of Cantrell, cloacal exstrophy, bladder exstrophy, and limb body wall complex. The association of other congenital abnormalities with AWDs is commonly observed, and these additional abnormalities carry crucial prognostic and management implications⁽³⁾. These defects can have severe health implications and potential risks, which highlight the importance of precise prenatal diagnosis and effective management strategies.

Ultrasound (US) has emerged as a fundamental tool in screening and diagnosing these conditions, providing a noninvasive and safe approach to visualize and assess fetal abdominal wall defects. The American Institute

of Ultrasound in Medicine guidelines mandate the visualization of umbilical cord insertion on the fetal abdomen during second- or third-trimester ultrasound examinations in order to exclude a potential AWD⁽⁴⁾. In cases where an abdominal wall defect is detected, a specific diagnosis is determined by assessing the relationship between the defect and the site of umbilical cord insertion.

This paper provides a general review of the specialized literature regarding various types of abdominal wall defects, namely omphalocele, gastroschisis, pentalogy of Cantrell, bladder exstrophy, and limb body wall complex, highlighting the significant sonographic features observed during ultrasound examination. This includes aspects such as the precise location, contents and any associated anomalies. Additionally, differential diagnoses are explored, emphasizing the importance of accurately distinguishing between these conditions to ensure optimal management strategies. To gather pertinent articles on prenatal diagnosis of abdominal wall defects, an extensive search was conducted using databases such as PubMed, Elsevier and Google Scholar. The search utilized the keywords “abdominal wall defect”, “ultrasound”, “omphalocele”, “gastroschisis”, and “pentalogy of Cantrell”.

Omphalocele

Omphalocele refers to a condition where there is a defect in the closure of the abdominal wall, specifically affecting the insertion of the umbilical cord. It arises when loops of the intestines do not return to the abdominal cavity as expected after their natural herniation into the umbilical cord from the sixth to tenth week of development⁽⁵⁾. It is a membrane-covered herniation, where the organs that have protruded are enclosed within a protective sac made up of two layers: the peritoneum and the amnion⁽³⁾. During the prenatal period, omphalocele stands out as one of the most frequently encountered abnormalities affecting the abdominal wall⁽⁶⁾, having a global prevalence of 2.6 per 10,000 births⁽⁷⁾.

There are two distinct variants of omphalocele, categorized based on whether the liver is present within the sac or not. These two variants exhibit different patterns of embryogenesis and can have different prognoses, primarily due to the varying risks of accompanying chromosomal abnormalities⁽³⁾. The presence of associated abnormalities is prevalent among affected fetuses, with approximately 67-88% exhibiting additional anomalies⁽⁶⁾. In cases of omphalocele, cardiac anomalies are frequently observed, occurring in approximately 50% of affected individuals⁽⁶⁾. Among these cardiac abnormalities, ventricular septal defect, tetralogy of Fallot, and *ectopia cordis* are the most commonly seen conditions⁽⁷⁾. Following cardiac anomalies, gastrointestinal abnormalities are encountered in approximately 40% of fetuses affected by omphalocele⁽⁶⁾. Diaphragmatic hernia, intestinal atresia and enteric duplications are the most frequently observed gastrointestinal abnormalities in this context⁽⁷⁾. Other organ system anomalies include

musculoskeletal anomalies, genitourinary anomalies and central nervous system anomalies⁽⁷⁾. Chromosomal abnormalities most commonly associated with omphalocele are trisomy 18 and trisomy 13⁽⁸⁾.

Ultrasound is the imaging method of choice for prenatal assessment of the fetus, enabling early detection of omphalocele. The earliest possible identification is around 12 weeks of menstrual age, as earlier it can be mistaken for physiological herniation⁽¹¹⁾.

On ultrasound, an omphalocele can be visualized as a protruding structure that exhibits the following features: it originates from the anterior abdominal wall, and it contains abdominal viscera such as the liver and/or bowel, as well as the convex side of the bulge displays the insertion of the umbilical cord⁽³⁾. The differential diagnosis should be made, as mentioned before, with the physiological herniation, gastroschisis, limb body wall complex (LBWC) and cloacal exstrophy. In gastroschisis, there is no membrane to cover the herniated organs and the umbilical cord insertion is normal, while in an omphalocele it inserts on the top of the sac⁽³⁾. The limb body wall complex is distinguished by distortion of the body's anatomy, resulting in severe scoliosis, as well as abnormalities in the limbs and skull⁽³⁾. While omphalocele commonly exhibits midline bowel herniation, gastroschisis is characterized by a defect that is typically right-sided, and in limb body wall complex, the defect is typically left-sided⁽¹²⁾. In cases of cloacal exstrophy, the omphalocele extends downwards, involving the region below the umbilicus⁽¹²⁾. This condition is accompanied by bladder exstrophy and abnormalities in the external genitalia⁽⁹⁾. Other nonchromosomal syndromes associated with omphalocele, beside the cloacal exstrophy, are Beckwith-Wiedemann syndrome (omphalocele + macroglossia + somatic hemihypertrophy + polycystic kidney) and pentalogy of Cantrell (midline anterior ventral wall defect + diaphragmatic defect + cleft distal sternum + defect of the apical pericardium with communication into the peritoneum + congenital heart defects)⁽³⁾.

The rates of neonatal morbidity and mortality exhibit a direct correlation with both the presence and severity of associated anatomic and chromosomal anomalies⁽¹³⁾.

Gastroschisis

Gastroschisis is characterized by a paramedian defect in the abdominal wall, frequently right-sided, through which loops of the bowel herniate and freely float within the amniotic fluid, not being covered by any membranes^(3,9). It is an AWD that does not involve the umbilical cord. The incidence of gastroschisis is 3 per 10,000 pregnancies⁽¹⁴⁾. There may be accompanying fetal anomalies primarily affecting the bowel. These anomalies involve motility dysfunction, intestinal atresia or stenosis, bowel obstruction, perforation, peritonitis, short gut syndrome, and necrotizing enterocolitis⁽¹⁵⁾. Other anomalies are quite rare⁽⁸⁾, but it is important to be looked for, as among fetuses with gastroschisis, approximately 14% were found to have additional anomalies,



Figure 1. Free floating bowels at 12 weeks of gestation

including central nervous system and cardiac malformations⁽¹⁶⁾. The utilization of ultrasound enables the identification of this birth defect *in utero* with a high level of both specificity and sensitivity. The diagnosis of this condition depends on identifying the presence of bowel loops freely floating outside the fetal abdomen (Figure 1 – image from personal collection) as early as 12 weeks of gestation with normal umbilical cord insertion (Figure 2 – image from personal collection)⁽¹¹⁾.

It is believed that prolonged exposure of the exteriorized bowel to the amniotic fluid leads to the leakage of important proteins, which in turn results in poor fetal growth⁽¹⁷⁾. Wall thickening in the herniated bowel



Figure 2. Sagittal view of the fetus at 12 weeks of gestation with normal cord insertion and paraumbilical wall defect through which the bowel protrudes

may occur due to inflammation and direct trauma, causing a series of bowel complications like atresia, perforation and infarction⁽⁹⁾. It is advisable to perform serial examinations to monitor fetal weight and fetal bowel diameters to help predict bowel complications. It is important to take into consideration that fetal weight might be underestimated in cases of gastroschisis due to the smaller abdominal circumference resulting from the extrusion of intraabdominal contents⁽¹⁸⁾. In highly exceptional scenarios, certain authors have documented instances where, following a significant dilation of an ileal loop caused by obstruction, the dilated loop could vanish entirely⁽³⁾. This disappearance is attributed to the complete necrosis of the affected loop, a phenomenon commonly referred to as “vanishing gut”. The differential diagnosis should be made with omphalocele, physiological hernia and LBWC. The ultrasonographic differences between these entities have been discussed earlier. It is important to diagnose the AWDs not earlier than 12 weeks of gestation, as physiological hernia can be mistaken for gastroschisis. As an example, in Figure 3 we present the case of a physiological hernia, analyzed in our clinic, mistaken for gastroschisis at 10 week and 4 days, that completely disappeared at almost 12 weeks of gestation. Typically, the prognosis for gastroschisis is favorable, unless complications such as perforation, infarction or infection of the herniated loops arise⁽³⁾.

Limb body wall complex

LBWC is a fetal condition characterized by the presence of at least two out of three following specific features: exencephaly, and either facial clefts, thoraco- and/or abdominoschisis, along with limb defects⁽²⁴⁾. The terminology of LBWC has been variably interchanged with body stalk anomaly (BSA) in the literature^(9,11). The prevalence of this rare disorder is approximately one in 10,000- 40,000 live births⁽²⁵⁾. The condition is characterized by the visceral organs being attached to the placenta and a shortened or absent umbilical cord⁽¹¹⁾. Russo et al. describe two phenotypes for LBWC: one consisting of an encephalocele with facial cleft and craniofacial placental attachment, and the other comprising an abdominal defect, abdominal placental attachment, a short or absent umbilical cord, and visceral abnormalities⁽²⁵⁾.

On ultrasound, the characteristic features of LBWC include the fetus typically being in a fixed position and frequently exhibiting complete evisceration of abdominal contents⁽¹¹⁾. These abdominal contents are often found adherent to the surface of the placenta. The identification of the cord insertion site is not possible, and there are no observable free-floating loops of the umbilical cord. In the placentocranial variant, notable craniofacial abnormalities are observed alongside severe defects in the limbs, spine and internal organs⁽⁹⁾. These abnormalities may be present along with or without abnormal craniofacial placental attachment⁽⁹⁾. Early diagnosis of LBWC is critical for providing parental counseling, since this condition is incompatible with life.

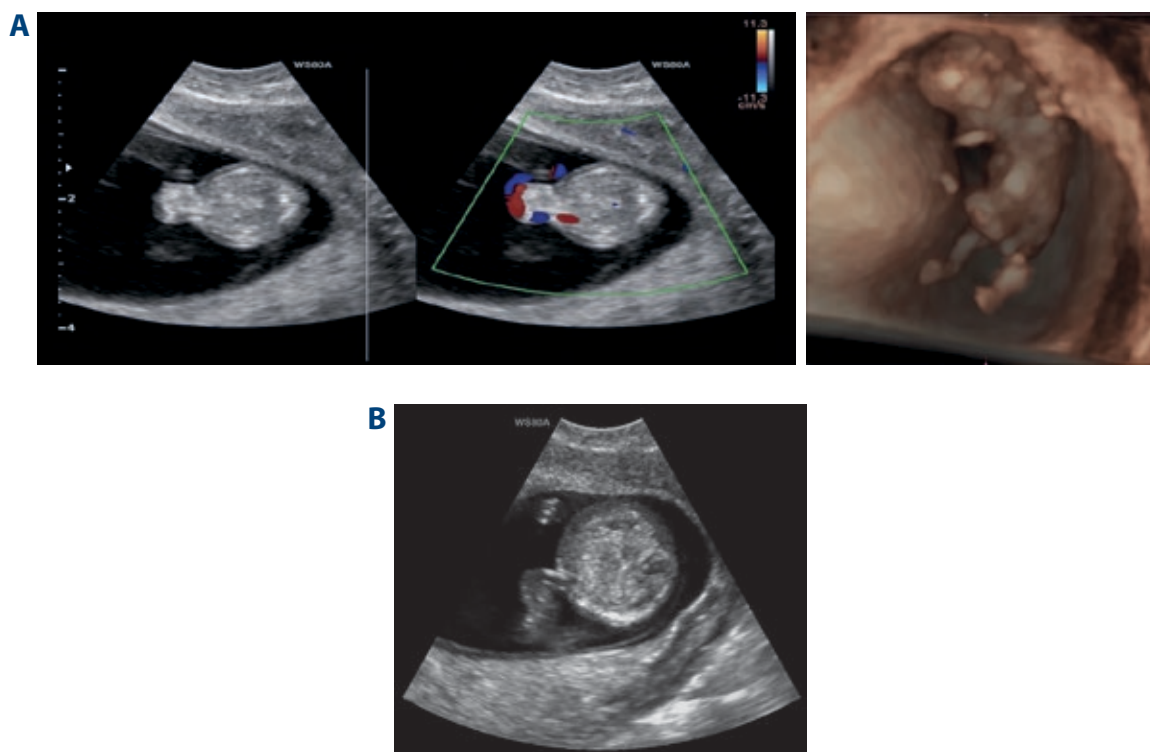


Figure 3. Ultrasound imaging at 10 weeks and 4 days. **A)** The scan revealed an echogenic structure with irregular borders emerging from the right side of the umbilical cord insertion, suggesting an abdominal wall defect with features consistent with gastroschisis. The main features of the image were the irregular border, the lack of a membrane to cover it, and also the lateral position relative to the umbilical vessels. **B)** Ultrasonography at 11 weeks and 4 days showing the complete resolution of the discussed anomalies, with complete reintegration of the bowel into the abdomen

Bladder and cloacal exstrophy

Bladder exstrophy is a condition that manifests in approximately one out of every 30,000 births, with a higher incidence among males⁽¹⁾, whereas cloacal exstrophy, a rare and serious congenital abnormality, has an incidence of approximately one case in 200,000-400,000 births, with higher rates observed among women⁽¹⁹⁾.

In normal embryological development, the caudal growth of the urorectal septum leads to its fusion with the cloacal membrane, effectively and completely partitioning the cloaca into two distinct parts, the bladder anteriorly and the rectum posteriorly⁽²⁰⁾. Around the sixth week of development, a normal perforation occurs in the cloacal membrane, leading to the formation of separate openings for urogenital and anal pathways⁽²⁰⁾. The urogenital sinus, an embryonic structure, serves as the precursor for several important anatomical features. It gives rise to the bladder and urethra in both male and female fetuses⁽⁵⁾. Additionally, in female fetuses, a section of the vagina also originates from the urogenital sinus⁽⁵⁾. Marshall and Muecke stated that the migratory failure of the lateral mesodermal folds to the cloacal membrane is responsible for these two AWDs⁽²¹⁾. The absence of mesodermal reinforcement in this context

leads to the premature rupture of the cloacal membrane. The timing of this rupture determines the severity and extent of the abdominal wall defect, as well as the degree of involvement of the urogenital tract⁽²⁰⁾. Cloacal exstrophy occurs when the rupture of the cloacal membrane happens prematurely, before its fusion with the urorectal septum, whereas bladder exstrophy arises from the rupture of the cloacal membrane right after its fusion with the urorectal septum⁽⁹⁾.

Ultrasound of bladder exstrophy reveals an AWD inferior to the cord insertion on the abdominal wall with the hallmark of “absent” urinary bladder because of the bladder being open to the abdominal wall, with urine eliminated immediately into the amniotic cavity⁽¹¹⁾. By visualizing the perivesical iliac arteries, it becomes possible to locate the bladder within the protruding mass, thus providing confirmation of the exstrophy⁽⁹⁾. The inflammation of the exposed mucosa can lead to an abnormal thickening of the posterior bladder wall which can be observed during ultrasound examination as an irregular contour of the lower anterior abdominal wall⁽¹¹⁾.

Cloacal exstrophy on ultrasound, like bladder exstrophy, is characterized by a low anterior abdominal wall defect and the absence of a visible bladder.

Furthermore, it involves the exstrophy not only of the bladder, but of all the other structures that contribute to the formation of the cloaca, including the rectum and lower genitourinary tract⁽⁹⁾. Genital abnormalities occur in the bladder exstrophy as well, and the most frequent for males are epispadias, a short or split penis and maldescended testes, whereas for females, there are commonly observed the presence of a bifid clitoris, as well as anomalies affecting the uterus and vagina⁽¹¹⁾. Cloacal exstrophy is generally accompanied by an omphalocele of varying size⁽²⁰⁾. On ultrasound, in the case of cloacal exstrophy, the omphalocele constitutes the upper part of a larger and more intricate abdominal wall defect that extends downwards to the pubis⁽¹¹⁾. The classical ultrasound image is the “elephant trunk” formed by the bowel herniation between the bladder halves. It is worth noting that other abnormalities affecting the urogenital, gastrointestinal, skeletal, and neurospinal axis are commonly seen alongside cloacal exstrophy⁽⁹⁾, like the OEIS complex which stands for omphalocele, exstrophy, imperforate anus, and spinal defects. The diagnosis of OEIS complex is established through ultrasound examination by focusing on the identification of the aforementioned defects. The visualization of the anal orifice and the internal genitalia poses the highest level of difficulty during sonography scans, fetal MRI being a valuable tool in this situation. OEIS complex has an unfavorable outcome attributed to the notable morbidity and mortality observed among infants at birth, primarily due to the necessary postnatal procedures⁽⁹⁾.

Pentalogy of Cantrell

Pentalogy of Cantrell (POC) is a rare thoraco-abdominal anomaly characterized by defects in the supraumbilical anterior abdominal wall, inferior sternum, anterior diaphragm, and pericardium⁽²²⁾. It is often accompanied by various cardiac abnormalities like pulmonary stenosis, septal defects, tetralogy of Fallot and ventricular diverticulum⁽⁹⁾. Its incidence is 1 in 65,000-200,000 births⁽²³⁾. The expression of the syndrome is variable, and it may manifest with all five components, enabling a definitive diagnosis⁽⁹⁾. Alternatively, it can present with three to four components, making the diagnosis probable⁽⁹⁾. Pentalogy of Cantrell can be accompanied by craniofacial and limb anomalies⁽⁹⁾. Additionally, it is frequently linked to trisomies 18 and 13 and Turner syndrome, so that amniocentesis should be performed⁽¹¹⁾.

Ultrasound is sufficient to make the diagnosis, allowing the visualization of the characteristic anomalies⁽³⁾. Omphalocele is better seen on sagittal and transverse views, whereas the cardiac ectopia are better revealed in the sagittal view⁽⁹⁾. Color Doppler ultrasound can be used as a helpful tool in detecting cardiac ectopia during medical examinations. During the early stages of the first trimester, it is possible for the inferior sternal and anterior diaphragmatic defect to be difficult to detect. In such instances, the presence of a pericardial effusion in conjunction with an omphalocele can serve as

an indication of the condition⁽³⁾. When considering a diagnosis of pentalogy of Cantrell, it is important to differentiate it from other conditions such as isolated omphalocele, LBWC and *ectopia cordis*. The first two have already been discussed. Unlike *ectopia cordis*, where the heart is positioned outside of the chest in an abnormal manner, POC is distinguished by the heart being positioned inside the chest cavity, typically with a noticeable shift in the cardiac axis that results in the apex of the heart being vertically oriented at the upper edge of the defect in the abdominal wall⁽³⁾. Color Doppler ultrasound can be a valuable tool for detecting cardiac ectopia. The prognosis of POC depends on the form of manifestation. Patients who have milder forms of the disease generally have a more favorable prognosis. The treatment approach to these individuals may involve either a single surgical repair or a staged surgical repair, depending on the specific combination of findings observed⁽³⁾.

Postnatal management and prognosis

More data regarding management are available for less complex abdominal wall defects, as some of the more intricate anomalies in the abdominal region are inherently lethal. It is crucial to accurately diagnose each condition in order to provide appropriate prenatal counseling and ensure accurate postnatal management.

Generally, gastroschisis is not lethal. Typically, fetuses affected by these congenital anomalies are delivered through a scheduled caesarean section, carefully managed and performed around the 36th week of gestation⁽⁹⁾. The purpose of this approach is to minimize the risks of neonatal and maternal morbidity that may arise from a spontaneous delivery. A study conducted in USA on 4803 newborns showed that, with appropriate obstetric and neonatal care, the survival rates are more than 90% for infants born with gastroschisis⁽²⁷⁾. The same study concluded that infants with an increased risk of mortality are born weighing less than 2500 grams or prior to 34 weeks of gestation⁽²⁷⁾. In a 2019 review of the literature performed for 207 pregnancies with fetal gastroschisis, spontaneous labor was associated with adverse neonatal outcomes, particularly in those born preterm⁽²⁸⁾.

LBWC – which is characterized as a lethal condition – is often accompanied by a high rate of spontaneous abortion⁽⁹⁾. Only a limited number of cases have been reported where infants with this condition have undergone extensive postnatal surgery and survived to early childhood.

Omphalocele is surgically corrected after birth in a single procedure or, depending on the size of the omphalocele, it may necessitate a two-step approach. The prognosis is highly favorable when the omphalocele is isolated and the liver is not located within the sac, whereas when the liver is in the sac, the prognosis is relatively poor, and the surgical correction do not allow the complete closing of the abdominal wall, and sometimes the complete closure require waiting until the growth of the abdominal wall allows accommodation of the liver without an abnormal increase in intraabdominal pressure⁽⁹⁾.

Following birth, bladder exstrophy is typically treated through either primary bladder closure or staged closure. The surgical repair is commonly conducted during the early stages of the neonate's life. However, some medical centers opt to delay the closure until the infant reaches 2-4 months of age. This delay allows for parent-child bonding and allows organs to grow and mature before undertaking the complex surgical procedure⁽⁹⁾.

The typical approach for the surgical management of cloacal exstrophy involves a collaborative effort between pediatric surgery and urology, and it is usually performed within the first 48-72 hours of the newborn period, being a very complex procedure that requires multiple surgical interventions⁽²³⁾. The primary objectives of the reconstruction procedure are to achieve a stable closure of the abdominal wall and bladder, to protect renal function, prevent short gut syndrome, and create genitalia that are both functional and esthetically satisfactory⁽³⁾. Joint planning by a multidisciplinary team, consisting of a radiologist, maternal-fetal medicine specialist, neonatologist, pediatric surgeon, and other surgical subspecialists, is essential in order

to establish a coordinated strategy for patient's counseling and postnatal care⁽¹¹⁾. Parental counseling should encompass a comprehensive discussion of topics such as bowel and urinary control, as well as sexual function, emphasizing the chronic nature of this condition and its impact on the patient's quality of life.

Conclusions

Fetal abdominal wall defects encompass a diverse group of conditions characterized by a wide range of associated multisystem anomalies and manifestations. Accurate prenatal characterization and classification of these defects are crucial in offering effective parental counseling throughout pregnancy and facilitating timely referral to a multidisciplinary management team. Ultrasound is the primary imaging modality used to accurately establish the prenatal diagnosis of AWD, and plays a crucial role in prenatal counseling, management, and surgical planning. A comprehensive understanding of the sonographic features, associated findings, and limitations of ultrasound is essential for effectively managing these complex congenital anomalies. ■

References

- Boyd PA, Haeusler M, Barisic I. EUROCAT Report 9: Surveillance of congenital anomalies in Europe 1980-2008. *Birth Defects Res A Clin Mol Teratol*. 2011;91 Suppl 1:S1.
- Bence C, Wagner A. Abdominal wall defects. *Transl Pediatr*. 2021;10(5):1461-9.
- Paladini D, Volpe P. Ultrasound of congenital fetal anomalies: differential diagnosis and prognostic indicators, 2nd Ed., CRC Press, London, 2014:267-307.
- American Institute of Ultrasound in Medicine. AIUM practice guideline for the performance of obstetric ultrasound examinations. *J Ultrasound Med*. 2013;32(6):1083-101.
- Sadler TW. *Langman's Medical Embryology*. Lippincott Williams&Wilkins, 2010:155-64.
- Mann S, Blinman TA, Wilson DR. Prenatal and postnatal management of omphalocele. *Prenat Diagn*. 2008;28(7):626-32.
- Nembhard WN, Bergman JEH, Politis MD, et al. A multi-country study of prevalence and early childhood mortality among children with omphalocele. *Birth Defects Res*. 2020;112(20):1787-801.
- Wilson RD, Johnson MP. Congenital abdominal wall defects: an update. *Fetal Diagn Ther*. 2004;19(5):385-98.
- Revels JW, Wang SS, Nasrullah A, et al. An algorithmic approach to complex fetal abdominal wall defects. *AJR Am J Roentgenol*. 2020;214(1):218-31.
- Chih-Ping C. Chromosomal abnormalities associated with omphalocele. *Taiwanese J Obstet Gynecol*. 2007;46(1):1-8.
- Pakdaman R, Woodward P, Kennedy A. Complex abdominal wall defects: appearances at prenatal imaging. *Radiographics*. 2015;35(2):636-49.
- Emanuel PG, Garcia GI, Angtuaco TL. Prenatal detection of anterior abdominal wall defects with US. *Radiographics*. 1995;15(3):517-30.
- Brantberg A, Blaas HG, Haugen SE, et al. Characteristics and outcome of 90 cases of fetal omphalocele. *Ultrasound Obstet Gynecol*. 2005;26(5):527-37.
- Friedman AM, Ananth CV, Siddiq Z, D'Alton ME, Wright JD. Gastroschisis: epidemiology and mode of delivery, 2005-2013. *Am J Obstet Gynecol*. 2016;215(3):348.e1-9.
- Salihi HM, Boos R, Schmidt W. Omphalocele and gastroschisis. *J Obstet Gynaecol*. 2002;22(5):489-92.
- Barisic I, Clementi M, Häusler M, et al. Evaluation of prenatal ultrasound diagnosis of fetal abdominal wall defects by 19 European registries. *Ultrasound Obstet Gynecol*. 2001;18(4):309-16.
- Payne NR, Simonton SC, Olsen S, Arnesen MA, Pfliegerhaer KM. Growth restriction in gastroschisis: quantification of its severity and exploration of a placental cause. *BMC Pediatr*. 2011;11:90.
- Centofanti SF, Brizot Mde L, Liao AW, Francisco RP, Zugaib M. Fetal growth pattern and prediction of low birth weight in gastroschisis. *Fetal Diagn Ther*. 2015;38(2):113-8.
- Luehr B, Lipsett J, Quinlivan A. Limb-body wall complex: a case series. *J Matern Fetal Neonatal Med*. 2002;12(2):132-7.
- Sahinoglu Z, Uludogan M, Arik H, et al. Prenatal ultrasonographic features of limb body wall complex: a review of etiopathogenesis and a new classification. *Fetal Pediatr Pathol*. 2007;26(3):135-51.
- Russo R, D'Armiento M, Angrisani P, Vecchione R. Limb body wall complex: a critical review and a nosological proposal. *Am J Med Genet*. 1993;47(6):893-900.
- Feldkamp ML, Botto LD, Amar E, et al. Cloacal exstrophy: an epidemiologic study from the International Clearinghouse for Birth Defects Surveillance and Research. *Am J Med Genet C Semin Med Genet*. 2011;157C(4):333-43.
- Woo LL, Thomas JC, Brock JW. Cloacal exstrophy: A comprehensive review of an uncommon problem. *J Pediatr Urol*. 2010;6(2):102-11.
- Marshall VF, Muecke EC. Variations in exstrophy of the bladder. *J Urol*. 1962;88(6):766-96.
- Williams AP, Marayati R, Beierle EA. Pentalogy of Cantrell. *Semin Pediatr Surg*. 2019;28(2):106-10.
- Desselle C, Herve P, Toutain A, Lardy H, Sembely C, Perrotin F. Pentalogy of Cantrell: sonographic assessment. *J Clin Ultrasound*. 2007;35(4):216-20.
- Brebner A, Czuzoj-Shulman N, Abenhaim HA. Prevalence and predictors of mortality in gastroschisis: a population-based study of 4803 cases in the USA. *J Matern Fetal Neonatal Med*. 2020;33(10):1725-31.
- Osmundo Junior GS, Mohamed SHM, Nishie EN, et al. Association of spontaneous labor onset with neonatal outcomes in pregnancies with fetal gastroschisis: A retrospective cohort analysis. *Acta Obstet Gynecol Scand*. 2019;98(2):154-61.

CONFLICT OF INTERESTS: none declared.

FINANCIAL SUPPORT: none declared.



This work is permanently accessible online free of charge and published under the CC-BY.