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Pregnancy with Didelphys Uterus and Omphalocele Infant: A Case Report

Leony Lim[1], I Wayan Artana Putra[2], I Nyoman Bayu Mahendra[2], Endang Sri Widiyanti[2]

[1]Resident of Obstetric and Gynecology Department, Faculty of Medicine Udayana University, Sanglah Hospital, Bali-Indonesia
[2]Obstetric and Gynecology Department, Faculty of Medicine Udayana University, Sanglah Hospital, Bali-Indonesia

Abstract: ***Didelphys uterus is a congenital anomaly characterized by the failure of Mullerian duct fusion and is a rare condition. Pregnancy with didelphys uterus may cause various complications to the fetus. Abnormalities of the fetus in the form of omphalocele also become a complication in the pregnancy with didelphys uterus. This article reviews the case report of a 30-year-old female patient, G3P1102, gestational age 20 – 21 weeks with a history of two times of caesarean section, presented with a complaint of acute abdominal pain. Initially, the patient was suspected of having an ectopic pregnancy. However, physical and ultrasonography examination revealed an abnormality of didelphys uterus with pregnancy in the right uterus. A fetal congenital anomaly of omphalocele was also found. Termination of pregnancy was not immediately carried out since there were no signs of emergency in both the mother and fetus. The patient had caesarean section delivery at 36 weeks of gestation and the infant underwent conservative treatment in an incubator according to omphalocele management. Pregnancy with didelphys uterus and omphalocele infant may increase the risk of various complications to both the mother and fetus. Therefore, antenatal screening should be performed as early as possible, especially in the first or second trimester, and is recommended to detect a congenital anomaly that may complicate pregnancy earlier.***

Keywords: Didelphys uterus, mullerian duct anomaly, omphalocele

1. Introduction

Didelphys uterus is a congenital anomaly that occurs due to the failure of Mullerian duct fusion during the early female internal genitalia system development.[1] This anomaly is characterized by the presence of two uterine corpora and two cervices that are separated by a vaginal longitudinal septum.[2] The incidence of Mullerian duct anomalies is approximately 0.5% - 5% of the normal population, with the didelphys uterus prevalence of 8.3%.[1] The incidence of didelphys uterus is also found on 0.1% - 0.5% of the fertile population and based on research, about 45% of which may achieve term pregnancy.[1,3] Pregnancy with didelphys uterus may develop various maternal and perinatal outcome, ranging from miscarriage (21%), prematurity (24%), ectopic pregnancy (2%), intrauterine growth restriction (11%), to an increased incidence of caesarean section (CS) (84%).[4]

Omphalocele is a congenital abnormality that occurs due to the failure of intra-abdominal (midgut) organs to return to the abdominal cavity after going through the physiological umbilical herniation process. This condition results in a defect of anterior abdominal wall closure, accompanied by abdominal viscera herniation that is covered by the membrane connected to the umbilical cord.[5] The incidence of omphalocele is estimated to occur in 1:1100 pregnancies. However, because of the high rate of miscarriage in these pregnancies, it is estimated that the incidence range between 1:4000 to 1:7000 live births.[6,7]

Considering the complications and difficulties that may happen to both the mother and the fetus in the pregnancies with didelphys uterus and omphalocele, a multidisciplinary approach involving obstetricians and gynecologists, pediatricians, and anesthesiologists throughout the pregnancy and delivery process is needed.

2. Case Report

A-30-year-old female patient, G3P1102, 20 – 21 weeks of gestational age, with a history of previous two sectio caesarea, was referred to Sanglah General Hospital Denpasar with the diagnosis of ectopic pregnancy. The patient complained of abdominal pain 1 day before hospital admission without any vaginal discharge. The patient denied any history of dyspareunia (pain during intercourse), dysmenorrhea (painful menstruation), cyclical abdominal pain, or recurrent miscarriage, however, the patient said that there was a history of preterm labor in two previous pregnancies and acute abdominal pain in the current pregnancy. The fetal movement was still felt and active. Based on the physical examination, it was found that the patient had a good general condition with a bodyweight of 57 kg and a height of 152 cm. The patient's vital signs were within normal limits. From the abdominal examination, a solid, immobile, 20 x 20 cm, well-demarcated mass with a flat surface was found. Obstetric examination revealed pain, no contraction, and fetal heart rate (FHR) of 158 beats per minute. A vaginal touche examination was not performed.

Complete blood count revealed leukocytosis with a leukocyte count of 17.7 x 10^3/µL. Subsequently, the patient underwent an ultrasonography (USG) examination. The USG showed a single live fetus with a gestational age of 20 weeks 1 day, weighing 342 gram, anterior placenta, and sufficient amniotic fluid volume. USG examination also revealed a ventral abdominal defect with a diameter of 2,94 cm with the impression of an omphalocele. The patient was then diagnosed with G3P0202 19 weeks 6 days, single live fetus, history of 2 CS, suspected abdominal pregnancy, and fetal congenital anomaly of omphalocele. The patient did not undergo exploratory laparotomy because there were no signs of emergencies.

The next day, the patient underwent another USG examination at our hospital for re0evaluation. The result of the re-USG examination showed a single, live, intrauterine fetus with an abnormality of omphalocele. In addition, from USG examination also showed didelphys uterus with septum (Figure 1 and 2), thus the patient's diagnosis was changed to G3P0202 19 weeks 6 days single live fetus, history of 2 CS, didelphys uterus, and fetal congenital anomaly (omphalocele). The patient was then assigned to undergo amniocentesis, karyotyping, and fetal echocardiography at 22-24 weeks of gestation. In this patient, a routine USG examination was carried out every 2 weeks to monitor the fetal condition at the Obstetrics and Gynecology Polyclinic of Sanglah General Hospital Denpasar.

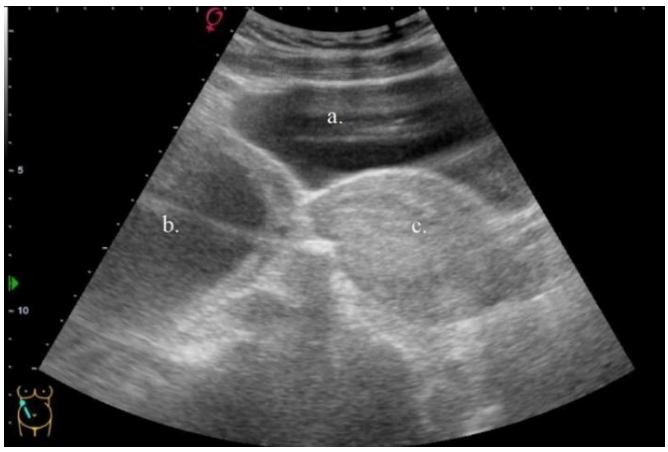

Figure 1: Ultrasonography (USG) revealed (a) and Didelphys uterus (b and c). Pregnancy in the right uterus (b).

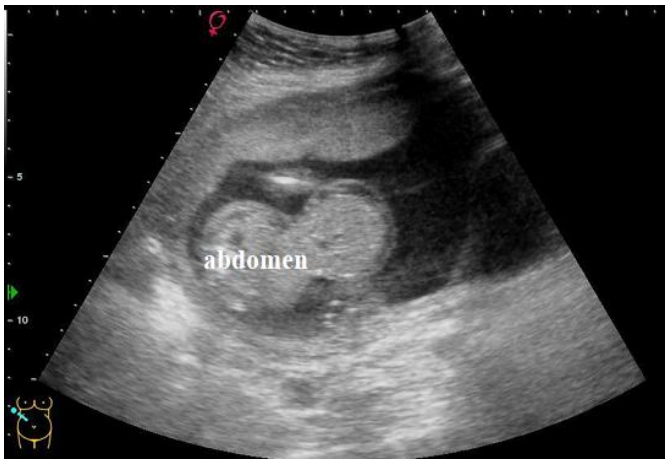

Figure 2: Transversal section of USG showed a fetus with omphalocele (white arrow)

Two weeks later the patient returned to the polyclinic for amniocentesis and routine USG examination at 22-24 weeks of gestation. From the amniocentesis, clear amniotic fluid with a volume of ± 30 ml was obtained. Karyotyping examination revealed a fetal chromosome of 46 XY. Furthermore, the patient also underwent fetal scanning at 30 weeks of gestation. Fetal scanning showed a hyperechoic lesion in the ventral abdomen with the impression of omphalocele and no other major abnormalities were found. The patient was planned for an elective CS at 38 weeks of gestation.

At 36 weeks of gestation came back for control. The patient complained of waxing and waning abdominal pain 3 hours ago and vaginal discharge 2 hours ago. Vital signs examination and general conditions were within normal limits. Obstetrics examination showed contractions 2 – 3 times in 10 minutes with a duration of 20 – 25 seconds and FHR of 155 beats per minute. Subsequent vaginal speculum examination showed a longitudinal vaginal septum (Figure 3) without blood the presence of blood and mucus. Based on the vaginal toucher examination, a vaginal opening of 2 cm, 50% effacement, no amniotic membrane, two palpable cervices separated with a longitudinal vaginal septum, palpable fetal head with transverse sagittal suture, a Hodge I decrease, and no small parts/fetal umbilical cord were found. Cardiotocography examination was in accordance with category I that indicates no fetal distress. The patient was then diagnosed with G3P0202 36 weeks 0 days single live fetus, history of 2 CS, didelphys uterus, fetal congenital anomaly (omphalocele), and 1st stage of labor. The patient was then decided to undergo termination of pregnancy with CS, followed by bilateral Pomeroy tubectomy.

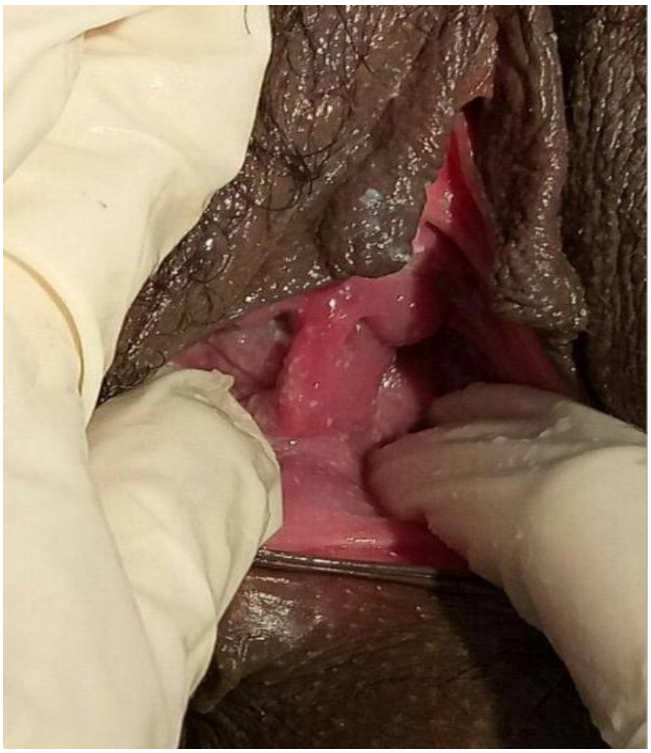

Figure 3: Longitudinal vaginal septum (white arrow)

During the surgery, there were two uteri that supported the diagnosis of didelphys uterus with the pregnancy occurring in the right uterus (Figure 4). Post-surgery, a male infant was delivered with a birth weight of 2300 grams, body length of 43 cm, APGAR score 5-6, New Balllard Score of 30 – 36 weeks, and a congenital abnormality in the form of an omphalocele measuring 5 x 5 cm (Figure 5). The patient then underwent hospitalization for 2 days after the surgery. The patient was allowed to go home with outpatient medicine of Cefixime 100 mg every 12 hours orally, paracetamol 500 mg every 8 hours orally, ferrous sulphate 300 mg every 12 hours orally, and methylergometrine 0,125 mg every 8 hours orally. The patient was also advised to return to the polyclinic 2 days later.

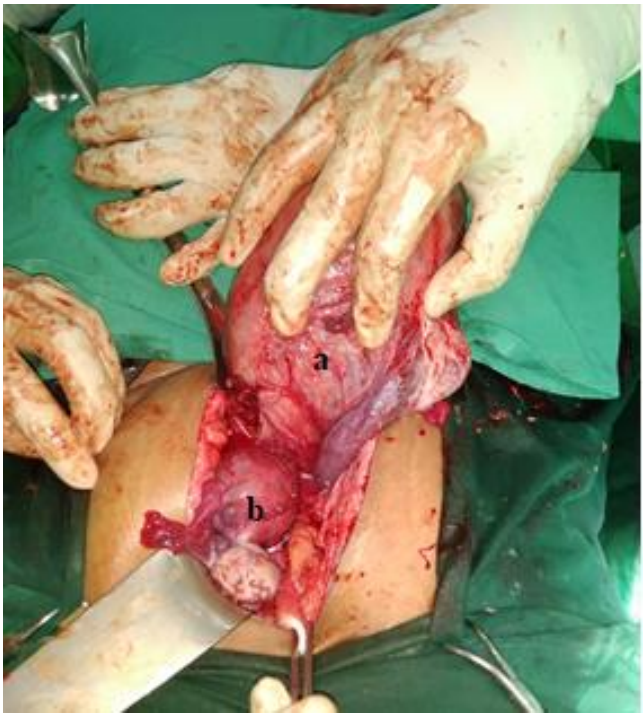

Figure 4: Didelphys uterus; (a) right uterus and (b) left uterus

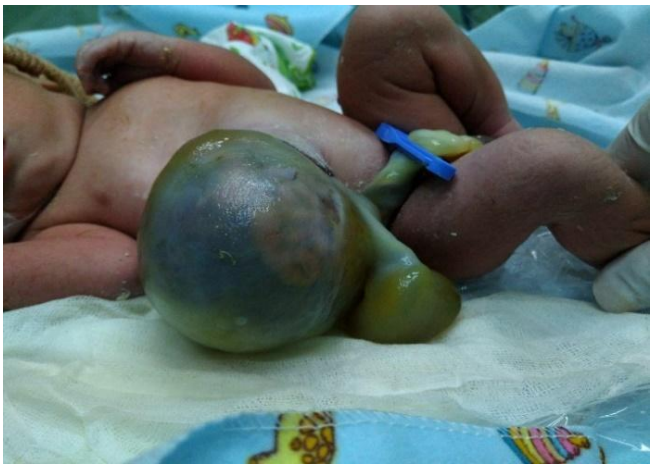

Figure 5: Delivered infant with omphalocele

The delivered infant was treated with omphalocele conservatively in an incubator using nebacetine powder and covered with wet gauze to stimulate neo-epithelization, however the infant passed away at the age of 1 month due to sepsis.

3. Discussion

This case report describes a pregnancy with didelphys uterus and omphalocele infant. Initially, our patient was suspected of having an ectopic pregnancy, however from the anamnesis, physical, and supporting examination, it was found that the patient was pregnant with didelphys uterus and omphalocele infant. Until now, the diagnosis of didelphys uterus still faces many obstacles, since many of these patients do not have significant symptoms and the appearance of the uterus is also difficult to be differentiated from other Mullerian duct anomalies, such as the bicornuate uterus and septate uterus.

During the initial presentation, the patient complained of acute abdominal pain and was initially suspected of ectopic pregnancy. The symptoms experienced by the patient were not specific however, based on a subsequent physical and supporting examination, two cervices and masses apart from the pregnancy itself were discovered. Based on these results, we have to determine whether the mass is a uterus (which indicates an anatomical anomaly and should be differentiated between didelphys uterus, bicornuate uterus, or septate uterus, because they all give the same impressions, namely two uterine cavities), or pregnancy with subserous myoma, or ectopic pregnancy (abdominal pregnancy).

Most didelphys uterus patients were asymptomatic, but patients with Mullerian duct anomalies sometimes had a history of dyspareunia or dysmenorrhea. Pain may occur because of the presence of a longitudinal septum of the vagina that sometimes obstructs one of the uterine corpora. This obstruction then causes hematometra (blood retention in the uterine corpus) and cyclical abdominal pain. In addition, due to the presence of structural uterus anomaly, complaints related to the reproductive function can also be asked such as a history of recurrent miscarriage, preterm labor, a history of cyclical abdominal pain, without any abdominal pain during pregnancy.[3,8] Suspicion of pregnancy with subserous myoma or abdominal pregnancy is higher in patients with complaints of acute abdominal pain. This occurs due to the effect of the mass' pressure on the surrounding organs, causing various disorders, ranging from urinary or defecation disorder to acute abdominal pain in the event of torsion or necrosis of the myoma.[9]

Our patient denied any complaint of dyspareunia, dysmenorrhea, recurrent miscarriage, or cyclical abdominal pain, however, our patient reported a history of preterm labor in two previous pregnancies and acute abdominal pain in the current pregnancy. Based on the anamnesis, we cannot immediately determine the patient's abnormality. Based on the literature, it is said that both didelphys uterus and bicornuate uterus are often associated with the increased incidence of preterm labor, in contrast to septate uterus. In the case of septate uterus, an increased incidence of miscarriage was found. This happens due to the disruption of fetal implantation process, that occurs in the avascular septum (less blood flows to the septum compared to the normal myometrium tissue). However, sudden abdominal pain that our patient experienced may suggest a pregnancy with subserous myoma or abdominal pregnancy.[9,10,11]

On physical examination, an abdominal mass with a measurement of 20x20 cm was found, which supported the diagnosis of pregnancy with subserous myoma in our patient. However, a further examination found that there were two cervices separated by the longitudinal vaginal septum which supports the diagnosis of a didelphys uterus, in which the diagnostic criteria for the didelphys uterus are the findings of two uterine corpora and two cervices which are separated by the longitudinal vaginal septum of varying consistency, ranging from a thin and elastic septum to the thick and inelastic one..[4,10] The discrepancy between complaints of acute abdominal pain and the finding of two cervices on physical examination complicate the diagnosis in this patient.

Therefore, investigations are carried out to help distinguish the existing abnormalities.

Further examinations that can be done to help support the diagnosis are USG or magnetic resonance imaging (MRI). Each procedure has its advantages and disadvantages. However, MRI examination is the gold standard with a higher degree of accuracy for diagnosing existing abnormalities when compared with ultrasound. Ultrasound examination is generally performed because it is relatively easy to use, especially to assess the contours of the external fundus. Several subtypes of Mullerian duct anomalies such as didelphys, arcuate, bicornuate, and septate uterus can also be assessed using ultrasound. An example of the USG image below explains the difference between Mullerian duct anomaly with fusion disorders (bicornuate) and resorption disorders (septate). The main point of diagnosis is based on the presence of a gap in the uterine fundus, with a coronal cut oriented at the uterine fundus. If the apex of the fundus is more than 5 mm above the line drawn between the two tubal ostia, the abnormality is a septate uterus (Figure 6a). However, if the apex of the fundus is below or less than 5 mm of the line drawn between the two tubal ostia, the abnormality is a bicornuate uterus (Figures 6b and 6c).[12]

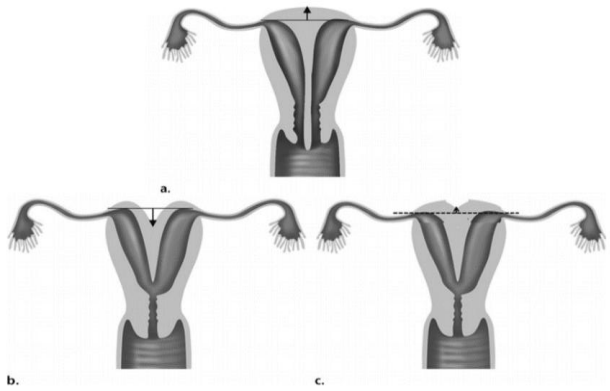

Figure 6: Various Mullerian duct anomalies (a) Didelphys uterus, (b) Septate Uterus, and (c) Bicornuate uterus.[12]

In this case, the only examination carried out was USG (without MRI), which revealed a pregnancy with a left-sided mass. This examination, in turn, raises a question, whether this pregnancy is a pregnancy with two uteri (didelphys uterus, bicornuate uterus, or septate uterus), a pregnancy with subserous myoma, or abdominal pregnancy. However, USG examination result showed a picture of intrauterine pregnancy (the fetus is bounded by the uterine wall), with the uterine wall separating the bladder and fetus, accompanied with intrauterine placenta, without any fetal body parts adjacent to the intra-abdominal organs of the mother, indicating that this pregnancy is an intrauterine pregnancy with two uterus corpora, thus the diagnosis was made to intrauterine pregnancy with didelphys uterus. Several differences between pregnancies with didelphys uterus, subserous myoma, and abdominal pregnancy are described in Table 1.

Management of pregnancy with didelphys uterus includes two things, namely the management of the pregnancy itself and the management of the didelphys uterus. The management of our patient should consider several factors since the patient had a history of 2 CS, a fetus with omphalocele, and the presence of didelphys uterus. Didelphys uterus alone is not an absolute indication for CS. Several thin and elastic septum allow the patient to undergo vaginal delivery. However, dystocia may occur if the septum has a thick and stiff consistency. Meanwhile, the mode of delivery of a fetus with omphalocele itself still becomes a matter of debate. Existing retrospective studies suggest that CS has not been shown to improve the survival rate of infants with omphalocele. However, current recommendations suggest vaginal delivery in infants with small defects (<5 cm) and CS for infants with large defects (≥ 5 cm).[1,7]

Table 1: Differences of Didelphys Uterus, Subserous Myoma, and Abdominal Pregnancy

	Didelphys Uterus	Subserous Myoma	Abdominal Pregnancy
Anamnesis			
- Dyspareunia	+/-	-	-
- Dysmenorrhea/cyclical abdominal pain	+/-	-	-
- Recurrent miscarriage	-	-	-
- Premature delivery	+	+/-	-
- History of malpresentation	+/-	+/-	-
- Acute abdominal pain	-	+/-	+/-
- Urinary or defecation problem	-	+/-	+/-
Physical Examination			
- Abdominal mass	-	+	+
- Two cervix and two vagina separated by longitudinal vaginal septum	+	-	-
Supporting examination (USG/MRI)			
- Mass with endometrial layer	+	-	+
- Whorled appearance	-	+	-
- Intra-mass vascularization	-	+/-	-
- Absence of uterus wall between the fetus and bladder	-	-	+
- Placenta outside the uterus			
- Fetus part adjacent to the mother's intraabdominal organs	-	-	+
- Oligohydramnion	-	-	+
	-	-	+

In our patient, the chosen mode of delivery was CS, considering that the history of two previous CS may increase the incidence of uterine rupture during vaginal delivery and the size of omphalocele was ≥ 5 cm. At the time of surgery, two separate uteri was found, supporting the diagnosis of didelphys uterus. Operative management of the Mullerian duct anomaly is generally performed in patient with habitual miscarriage, without any other cause. The surgical technique that can be used is metroplasty, which joins the two endometrial cavities into one.[8,13] However, in this patient, the treatment is only done for the obstetric emergency through CS to deliver the infant, without the reconstruction of the uterine cavity.

Omphalocele is a congenital disorder that occurs due to failure of the intraabdominal organs (midgut) to return to the abdominal cavity after going through the physiological umbilical herniation process. This condition causes abdominal viscera herniation covered by a membrane connected to the umbilical cord.[14] Diagnosis of omphalocele can be done as early as possible, starting from 12 weeks of gestation through transvaginal USG, if a mass that resembles fetal abdominal diameter next to the ventral abdomen was discovered. In this case, the transabdominal USG in the 19 – 20 weeks of gestation revealed a fetal abdomen with a defect in the ventral side covered with a membrane, suggesting an omphalocele.

The management of omphalocele includes operative and non-operative methods. Management of newborn infants includes airway stabilization and defect closure with sterile moist gauze to prevent hypothermia and excessive fluid loss. Topical drug application (silver sulfadiazine) can be given to stimulate neo-epithelization. Non-operative methods involve the administration of antibiotics, intravenous hydration, and visceral organ decompression to prevent the incidence of distention. The main objective of non-operative management is to return the visceral organ into the abdominal cavity, followed by the closure of skin and fascia. If the omphalocele is large, the repair is carried out gradually using a silo sutured to the fascia layer until the extra-abdominal organs slowly return to the abdominal cavity in 3 – 10 days. After the mass is reduced, the silo can be removed, and then a repair was performed to close the existing defect. In this patient, a conservative method with defect closure using sterile-wet gauze and administration of topical drugs was performed however, the infant passed away at the age of 1 month due to neonatal sepsis.[15]

4. Conclusion

Diagnosis of didelphys uterus in pregnancy is easier to be made if the patient has checked herself during early pregnancy, and becomes more difficult if the patient had her antenatal examination in the mid/late pregnancy. Differential diagnosis of didelphys uterus in pregnancy includes other uterine abnormalities (bicornuate or septate uterus), pregnancy with myoma, or abdominal pregnancy.

Omphalocele is one of the most common congenital abnormalities. The prognosis was determined by the presence or absence of any accompanying abnormalities (chromosomal abnormalities) and the size of the omphalocele. Diagnosis and management can be done by performing screening since early pregnancy, chromosome analysis, and delivery planning that involves anesthesiologists, pediatricians, and pediatric surgeons. Therefore, antenatal screening in the 1st or 2nd trimester was recommended to detect congenital abnormalities in the fetus.

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