Congenital Diaphragmatic Hernia Anomaly in Multigravida at 36 Weeks Gestation with One Previous Cesarean Section, Single Live Fetus, Cephalic Presentation: Case Report

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Abstract
Background: Case of a multigravida at 36 weeks of gestation with one previous cesarean section carrying a single fetus was diagnosed with diaphragmatic hernia. This case aims to address the challenges posed by this complex scenario of diaphragmatic hernia and the importance of specialized care to ensure optimal maternal and fetal outcomes.
Case Report: Referred from Muhammadiyah Hospital Palembang, the patient at 36 weeks of gestation with G3P2A0 status present a single live fetus and was diagnosed with diaphragmatic hernia. Following prior midwife care where fetal heartbeats were not detected, the patient was referred to Dr. Mohammad Hoesin Central General Hospital Palembang. The management plan includes a one-week follow-up and folic acid, calcium carbonate, and iron supplementation.
Discussion: Congenital diaphragmatic hernia (CDH) is a developmental defect causing diaphragmatic discontinuity, diagnosed prenatally with 40% to 90% accuracy via ultrasound. The treatment aims to minimize lung hypoplasia and reduce mortality, typically performed at 26-28 weeks for severe cases and 30-32 weeks for moderate ones. The optimal delivery timing for CDH remains controversial, with lung-to-head ratio as a widely used prognostic indicator.
Conclusion: Congenital diaphragmatic hernia (CDH) exhibits lower survival rates on the right side (50% vs. 75%), with lung area to head circumference ratio (LHR) as a common prognostic parameter. Recent minimally invasive techniques like FETO aim to improve prognosis by reducing pulmonary hypoplasia and mortality.

Key words: Congenital Diaphragmatic Hernia, Prior Cesarean Section, Multigravida

Laporan Kasus: Multigravida Hamil 36 Minggu Belum Inpartu Bekas Seksio Sesarea Satu Kali Janin Tunggal Hidup Presentasi Kepala Dengan Anomali Kongenital Hernia Diafragmatik

Abstrak
Latar Belakang: Kasus multigravida hamil 36 minggu dengan riwayat operasi caesar janin tunggal hidup yang didiagnosis hernia diafragma. Tujuan laporan kasus ini untuk mengatasi tantangan yang ditimbulkan oleh skenario kompleks hernia diafragma dan menunjukkan pentingnya perawatan khusus untuk memastikan hasil akhir ibu dan janin yang optimal.
Laporan Kasus: Pasien usia kehamilan 36 minggu dengan status G3P2A0 janin hidup tunggal dengan diagnosis hernia diafragma dirujuk dari RS Muhammadiyah Palembang setelah sebelumnya diperiksa oleh bidan dan tidak terdeteksi detak jantung janinnya sehingga memerlukan rujukan ke RSUP Dr. Mohammad Hoesin Palembang. Rencana penatalaksanaannya mencakup tindak lanjut selama satu minggu, bersamaan dengan suplementasi asam folat, kalsium karbonat, dan zat besi.
Diskusi: Hernia diafragma kongenital (CDH) merupakan kelainan perkembangan yang menyebabkan diskontinuitas diafragma dan didiagnosis sebelum lahir dengan akurasi 40% hingga 90% melalui ultrasonografi. Tatalaksana bertujuan untuk meminimalkan hipoplasia paru-paru dan mengurangi angka kematian, biasanya dilakukan pada minggu ke 26 sampai 28 untuk kasus yang parah dan 30 - 32 minggu untuk kasus yang sedang. Waktu persalinan yang optimal untuk CDH masih kontroversial, dengan rasio paru-paru sebagai indikator prognosis yang banyak digunakan.
Kesimpulan: Hernia diafragmatika kongenital (CDH) menunjukkan tingkat kelangsungan hidup yang lebih rendah pada sisi kanan (50% vs. 75%), dengan rasio area paru terhadap lingkar kepala (LHR) sebagai parameter prognosis yang umum; teknik invasif minimal terkini bertujuan untuk meningkatkan prognosis dengan mengurangi hipoplasia paru dan kematian.

Kata kunci: Hernia Diafragma Kongenital, Riwayat Operasi Caesar Sebelumnya, Multigravida

Background

Congenital diaphragmatic hernia (CDH) is a diagnosis of developmental closure defect resulting in discontinuity of the diaphragm. This allows abdominal contents to enter the chest cavity. CDH occurs in approximately 1 in 3000 live births and results in high neonatal morbidity and mortality. It is also associated with severe pulmonary hypoplasia and pulmonary hypertension. The left posterolateral side of the diaphragm is the most common localization (75-90% of cases), but the defect, can also be right-sided (10-15% of cases) or even bilateral (1-2% of cases). Some studies report a slightly higher incidence of CDH in male fetuses. The prevalence of CDH does not seem to be associated with maternal age.[1,2]

In most cases, CDH occurs sporadically. However, a few cases with autosomal recessive, autosomal dominant, and X-linked inheritance patterns have been reported. Sporadic cases occur possibly due to de novo mutational events in genes for normal diaphragm development or reflect polygenic or multifactorial inheritance. The etiology of this defect remains unknown in more than 70% of individuals with CDH. The estimated risk of recurrence of congenital diaphragmatic hernia in future siblings in the absence of a family history is 1-2% after one affected child.[1,3,4]with extremely high neonatal mortality. This paper presents a review of the available literature on prenatal diagnosis, management and treatment options for CDH. In selected cases, a prenatal procedure to improve neonatal survival is possible. The authors of this manuscript believe their work might contribute to a better understanding of congenital diaphragmatic hernia and patient selection for the FETO (fetal endoscopic tracheal occlusion

The diagnosis of CDH can be made using prenatal ultrasound examination, which is accurate in 40% to 90% of cases. Prenatal diagnosis by ultrasound can detect more than 50% of CDH cases at an average 24 weeks of gestational age. Three-dimensional ultrasound imaging, fetal echocardiography, and fetal magnetic resonance imaging (MRI) are other prenatal diagnostic modalities used in assessing the severity and prognostics of CDH.[5,6]

Antenatal management can be given using antenatal corticosteroids to the mother to improve lung maturation in neonates with CDH. Surgery Maternal condition determines the condition of the fetus. Optimal surgical intervention in pregnant women is as important as prompt and appropriate diagnosis for the good of the mother and fetus. Fetal endoluminal tracheal occlusion (FETO) procedure can improve neonatal survival at 30 days and 6 months in patients with severe CDH. However, this procedure may increase the risk of premature rupture of membranes. The recent discovery of minimally invasive surgery, FETO, provides a better prognosis. The main goal of FETO is to minimize pulmonary hypoplasia and reduce mortality.[1,7,8]with extremely high neonatal mortality. This paper presents a review of the available literature on prenatal diagnosis, management and treatment options for CDH. In selected cases, a prenatal procedure to improve neonatal survival is possible. The authors of this manuscript believe their work might contribute to a better understanding of congenital diaphragmatic hernia and patient selection for the FETO (fetal endoscopic tracheal occlusion

The survival prognosis in CDH depends on several factors. If CDH is associated with a chromosomal abnormality, the long-term prognosis depends on the type of genetic abnormality and the co-existing abnormalities with CDH (particularly central nervous system and cardiac abnormalities). Several studies have shown that infants with CDH on the right side have a lower survival rate compared to CDH with left-sided lesions

(50 vs. 75%). For isolated cases of congenital diaphragmatic hernia, the most common prognostic parameter is the assessment of the amount of lung tissue in the fetal chest.[1] Hence, it is crucial to discuss the diagnosis and determine an appropriate intervention strategy for this uncommon scenario involving a multigravida with a previous cesarean section, carrying a live single fetus diagnosed with congenital diaphragmatic hernia.

Case Report

Mrs. KO was under-month pregnant with a congenital anomaly. The patient was referred from RS Muhammadiyah Palembang and diagnosed with G3P2A0 13 weeks of pregnant, live single fetus, with suspected diaphragmatic hernia. The patient previously had a history of antenatal care with a midwife and was told that the fetal heartbeat was not found, so the patient was referred to Dr. Mohammad Hoesin Hospital Palembang for further examination. There are no symptoms of heartburn, nausea and vomiting, abdominal pain, water discharge, or blood and mucus discharge. There was no history of trauma. There was no history of taking medicines and herbs. There was no history of sequestration. History of previous pregnancy with congenital anomaly was denied. Family history of pregnancy with congenital anomaly was denied. The patient admitted to be under-month pregnant with fetal movement still felt. The patient had Menarche at the age of 15 years, had regular menstruation with 5-7 days length of menstruation, and fist day of last menstruation was in March 2022. Moreover, the patient is married once for a duration of 7 years. The patient had a history of giving birth two times: the first child was spontaneous IUFD, the second child was SC on induction failure indication, and this pregnancy.

We found physical examination and vital signs within normal range, with general physical examination within normal limits. Obstetric examination TFU 3 fingers was below the umbilicus (30 cm), elongated, left dorsal, under the head, U 5/5 DJJ: 145x/m, TBJ: 2790g. Laboratory examination obtained Hb 10.1 g/dL, Leukocytes 11.86 x 106/mm3, Hematocrit 33%, RDW-CV 16.20%, Alpha Feto Protein 176.74 ng/ml. Ultrasound imaging is shown in Figures 1-5.

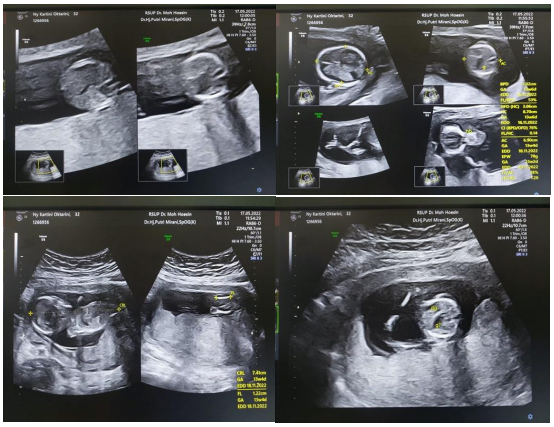

Figure 1 Ultrasound (17/05/22). 13 weeks pregnancy with a 1-time SC with a single live intrauterine fetus diagnosed with suspected diaphragmatic hernia.

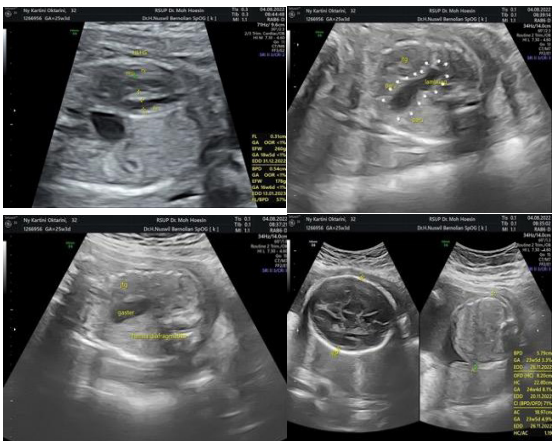

Figure 2. Ultrasound (22/09/22). Thirty-one weeks pregnancy with a single live fetus head presentation with suspected diaphragmatic hernia.

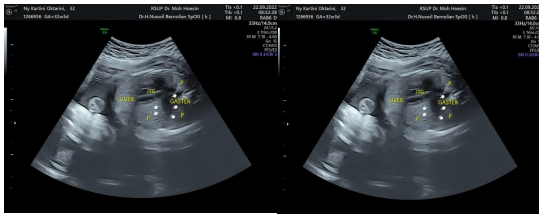

Figure 3 Ultrasound (06/10/22). A Fetal stomach was seen filling the thorax cavity. 34 weeks pregnancy with single live fetus head presentation with diaphragmatic hernia.

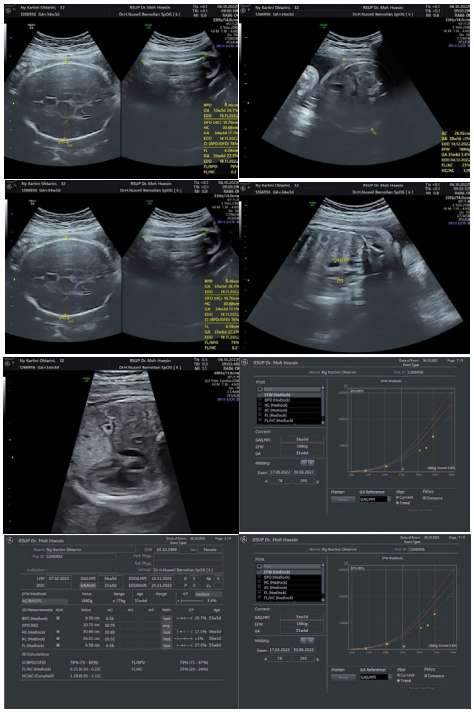

Figure 4. Ultrasound (20/10/22). 36 weeks pregnancy with a former 1-time SC live single fetus head presentation with diaphragmatic hernia

Based on anamnesis, physical examination, and comprehensive ultrasound examination, we diagnosed the patient with G3P2A0 36 weeks pregnancy, not in labor time, one previous cesarean section, single live fetus, head presentation with congenital anomaly (diaphragmatic hernia). The Mother had relatively good prognosis, while the baby was in dubia prognosis. We administered 400 mcg of folic acid once daily, 500 mg CaCo3 three times a day, and FeSo4. The patient then asked to check her condition again in the next seven days.

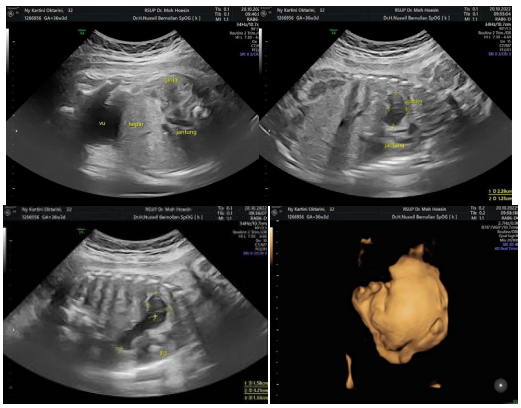

Figure 5 Lung-to-Head Ratio Measurement (25/10/22). Outcome 0.52 survival rate 61%.

Discussion

Congenital diaphragmatic hernia (CDH), is a developmental closure defect resulting in discontinuity of the diaphragm. This allows abdominal contents to enter the chest cavity. The prevalence of CDH does not seem to be related to maternal age.[1,2]

The diaphragm originates from the septum transversum, pleuroperitoneal folds, derivatives from the body wall, dorsal mesentery, and a pair of pre-muscular masses located opposite the fourth cervical segment. Under normal conditions, the pleuroperitoneal folds fuse with the septum transversum, esophageal mesentery, and muscular growths from the body wall fuse with the folds, forming the muscular portion of the diaphragm. Incomplete fusion of any of these components may result in a Morgagni hernia, pleuroperitoneal defect, hiatal hernia, or diaphragmatic eventration.[1,2,9–11]

Lung development usually occurs at 14-16 weeks of intrauterine life. Disruption of normal lung development leads to decreased bronchiolar branching as well as pulmonary

hypoplasia, excessive muscularization of pulmonary artery branches, and pulmonary hypertension. As well as, Abnormal lung development results in surfactant system dysfunction in late pregnancy and after birth. Pulmonary hypoplasia and abnormal pulmonary vascular development and function occur on both sides. In addition, Vasoconstriction with also abnormal pulmonary vasoreactivity contributes to poor pulmonary blood flow.[1,12]with extremely high neonatal mortality. This paper presents a review of the available literature on prenatal diagnosis, management and treatment options for CDH. In selected cases, a prenatal procedure to improve neonatal survival is possible. The authors of this manuscript believe their work might contribute to a better understanding of congenital diaphragmatic hernia and patient selection for the FETO (fetal endoscopic tracheal occlusion

The etiology of CDH remains largely unknown and is currently considered to have a multifactorial etiology. Most of the cases have diaphragmatic hernia with pulmonary hypoplasia and pulmonary hypertension of newborns (PPHN). CDH may be associated with cardiac, gastrointestinal, with genitourinary anomalies or chromosomal aneuploidies such as trisomy. Several genetic factors, along with environmental exposures and nutritional deficiencies, are possible etiologies for CDH.[6,7,13]

Posterolateral hernia, also known as Bochdalek's hernia, is the most common type (70-75%), with the majority occurring on the left side (85%) and less commonly on the right side (13%) or bilateral (2%). Anterior defect or Morgagni hernia (23-28%) and central hernia (2-7%) are other types of CDH. The diaphragm starts developing at around weeks of gestation and is fully formed by 12 weeks of gestation. Defects can range from defects in the posterior muscle rim to the complete absence of the diaphragm.[6]

The embryological basis of CDH remains controversial. It was originally thought that the defect occurred secondary to the failure of parts of the diaphragm to fuse to produce a patent pleuroperitoneal duct. In experimental studies, mouse models showed defects in the primordial diaphragm called pleuroperitoneal folds. This allows the intestines to enter the thoracic cavity when returning from the extraembryonic umbilicus.[6]

Alpha-fetoprotein (AFP) is a glycoprotein produced in the gut and liver during fetal life and serve a precursor to fetal albumin. AFP is a plasma protein produced by the embryonic yolk sac and fetal liver. AFP levels in serum, amniotic fluid, and urine serve as a screening test for congenital defects. Elevated levels of alpha fetoprotein levels in maternal serum and amniotic fluid are reliable indicators of fetal abnormalities. Although used to screen for neural tube defects, AFP is also elevated in defects such as omphalocele, gastroschisis, and sacrococcygeal teratoma, where fetal serum transudation increases.[14,15]

CDH can be diagnosis of prenatal ultrasound examination and is accurate in 40% to 90% of cases. Prenatal diagnosis by ultrasound can detect more than 50% of CDH cases at an average gestational age of 24 weeks. Three-dimensional ultrasound imaging, fetal echocardiography, and fetal magnetic resonance imaging (MRI) are other prenatal diagnostic modalities used in assessing the severity and prognostics of CDH.[5,6]

Ultrasound examination can be performed during routine pregnancy check-ups and when there is a suspicion of polyhydramnios. Polyhydramnios has been reported in approximately 80% of fetuses in pregnancies with CDH. The mechanism of polyhydramnios is thought to be due to the bending of the gastroesophageal junction by translocation of the stomach into the thorax with resultant obstruction of the anterior bowel. Diagnosis of CDH using ultrasound is advised by observing whether there is

abdominal content present in the fetal chest. Fetuses with CDH usually have a small abdomen due to impaired swallowing. If the diaphragmatic defect is on the right side, the liver may cause hernia site tamponade and obscure the diagnosis. CDH on the presence of left side may be characterized by a heterogeneous mass in which the abdomen may be filled with fluid or intestine.[5]

In addition to diagnosis, prenatal ultrasound may also be useful in prognostic prediction by using quantitative techniques to estimate the severity of fetal pulmonary hypoplasia in the lungs of fetuses with CDH. Three-dimensional estimation of fetal lung volume, calculation of the ratio of right lung area to thoracic area, and calculation of the ratio of lung to thoracic circumference are three different measurements that may correlate with neonatal outcomes. But, the lung-to-head ratio has been the most widely used prognostic indicator.[5]

After birth, the spectrum of respiratory symptoms in infants with CDH is determined by the degree of lung hypoplasia and reactive pulmonary hypertension. The majority of infants develop respiratory symptoms within the first 24 hours of life. Infants with CDH usually have a scaphoid abdomen and an asymmetrically distended thorax. The thorax may become more distended as ingested air passes into the stomach and intestines. Further gastrointestinal distension may compress the lung parenchyma and affect its ventilatory characteristics. This may lead to mediastinal compression with contralateral lung damage. Physical examination may reveal an abnormal thoracic size, with no breath sounds on the side with CDH. The diagnosis of CDH can be confirmed with a thoracic plain photograph showing bowel loops in the thorax. The location of the gastric bubble should also be noted, and its position can be confirmed by the placement of an orogastric tube.[5]

Antenatal management in the form of antenatal corticosteroids is given to the mother to improve lung maturation in neonates with CDH.[7] The condition of the mother largely determines the condition of the fetus. Optimal surgical intervention in pregnant women is as important as prompt and appropriate diagnosis for the good of both mother and fetus.[7] Regardless of the surgical approach, there are four important steps to correct CDH: reduce the size of the hernia; lyse the adhesions; reconstruct the hernia after complete excision of the hernia sac from the posterior pleural cavity; and repair the hernia defect.[16]the Bochdalek hernia (BH

Primary closure repair for hernia defects is the best treatment. Fetal endoluminal tracheal occlusion (FETO) procedure may improve neonatal survival at 30 days and 6 months in patients with severe CDH. However, this procedure may increase the risk of premature rupture of membranes. The recent discovery of minimally invasive surgery, FETO, provides a better prognosis. The main goal of FETO is to minimize lung hypoplasia and reduce mortality. Since its clinical introduction, FETO has been performed for severe cases of CDH at 26-28 weeks of gestation and for moderate cases at 30-32 weeks of gestation.[1,7,8]with extremely high neonatal mortality. This paper presents a review of the available literature on prenatal diagnosis, management and treatment options for CDH. In selected cases, a prenatal procedure to improve neonatal survival is possible. The authors of this manuscript believe their work might contribute to a better understanding of congenital diaphragmatic hernia and patient selection for the FETO (fetal endoscopic tracheal occlusion

The optimal timing for delivering a baby with CDH is controversial. Infants delivered by elective cesarean section at 37-38 weeks of gestation are associated with less extracorporeal membrane oxygenation (ECMO) use (22 vs. 35.5%) compared to those at 39-41 weeks of gestation. However, a more recent study found reduced mortality

at later gestational ages.[7]

The delivery of an infant with CDH should take place in a medical center with capabilities in the management of infants with CDH and associated complications. Insertion of central or peripheral venous access is required to administration of fluids and medications. Umbilical arterial line is required to determine the value of the umbilical arterial line, which reflects post-ductal arterial oxygen pressure (PaO2) and leads to an increased fraction of inspired oxygen (FIO2). The optimal ventilation mode for infants with CDH and hypoplastic lungs is unknown. Most medical centers use conventional mechanical ventilation (CMV) for respiratory support and optimize ventilation by adjusting PIP and respiratory rate. Invasive blood pressure monitoring is preferred over noninvasive monitoring. Dopamine is the most common cardiovascular drug in the NICU and is given to maintain systemic BP appropriate for gestational age. Dobutamine is preferred in infants with poor myocardial contractility. Norepinephrine and epinephrine may be used as first-line agents in some institutions secondary to their potent vasoconstrictor effects.[8,17–20]thereby allowing abdominal viscera to herniate into the thoracic cavity and subsequently interfering with normal lung development. At birth, pulmonary hypoplasia leads to respiratory insufficiency and persistent pulmonary hypertension (PHT

The survival prognosis in CDH depends on several factors. If CDH is associated with a chromosomal abnormality, the long-term prognosis depends on the type of genetic abnormality and the co-existing abnormalities with CDH (particularly central nervous system and cardiac abnormalities). Several studies have shown that infants with CDH on the right side have a lower survival rate compared to CDH with left-sided lesions (50 vs. 75%). For isolated cases of congenital diaphragmatic hernia, the most common prognostic parameter is the assessment of the amount of lung tissue in the fetal chest. This can be seen in the lung area to head circumference ratio (LHR).[1] However, lung volume growth in the developing fetus is different compared to head growth. To correct for gestational age, LR can be expressed as a percentage of normal LHR. Several predictors assess the fetal pulmonary vasculature. The McGoon index (MGI) on ultrasound and modified McGoon index on MRI are calculated as the sum of the right and left pulmonary artery diameters measured at the bifurcation and divided by the aortic diameter. According to some authors, suggest that this index may be useful for predicting neonatal survival and the severity of postnatal pulmonary hypertension.[1,13,21]with extremely high neonatal mortality. This paper presents a review of the available literature on prenatal diagnosis, management and treatment options for CDH. In selected cases, a prenatal procedure to improve neonatal survival is possible. The authors of this manuscript believe their work might contribute to a better understanding of congenital diaphragmatic hernia and patient selection for the FETO (fetal endoscopic tracheal occlusion

The intra-abdominal position of the fetal abdomen is associated with a better prognosis. Whereas, the presence of the abdomen within the thorax during the fetal period or neonate has been shown to correlate with a less favorable prognosis. Fetal abdominal postion is predictive of postnatal survival and the need for patch repair.[1,13,21]with extremely high neonatal mortality. This paper presents a review of the available literature on prenatal diagnosis, management and treatment options for CDH. In selected cases, a prenatal procedure to improve neonatal survival is possible. The authors of this manuscript believe their work might contribute to a better understanding of congenital diaphragmatic hernia and patient selection for the FETO (fetal endoscopic

tracheal occlusion

Conclusion

A arotid cavernous fistula (CCF) is an abnormal shunt from the carotid artery into the cavernous sinus, with symptoms depending on neural and vascular involvement. The decision to terminate pregnancy is influenced by increasing gestational age and should be promptly executed, keeping the patient under observation. Ethical, legal, medical, and religious considerations should guide the evaluation of terminating pregnancies in mothers with a history of CCF. Postnatally, infants born to mothers with CCF history can be managed conservatively.

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