CASE REPORT

Early diagnosis and management of inseparable conjoint twins. A low-middle-income country experience

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ABSTRACT

Objectives: To discuss the crucial early diagnosis of conjoined twins to determine the type and prognosis.

Case Report: A 27-year-old woman was referred to the type A referral hospital with suspicion of congenital abnormalities at 17 weeks of pregnancy. Ultrasound results showed intrauterine monochorionic monoamniotic twins with babies fused on their heads to the urogenital part. The MRI showed a craniopagus, suspected meningomyelocele, and severe bilateral hydronephrosis in the second baby. Due to non-separable cases and a bad prognosis for the fetus, the pregnancy was terminated using misoprostol induction and inserting a balloon catheter. The fetus was born weighing 400 g and 20 cm in length. Conjoined twins obtained the rostral type: a fused head with two faces, a fused thorax until the lower abdomen with one umbilicus, and two pairs of hands and feet. The diagnosis of conjoint twins becomes a problem in early pregnancy, mostly in developing countries. Early diagnosis of conjoined twins during prenatal examination is critical for ascertaining the prognosis of the fetus, guiding parental counseling over appropriate courses of action, and potentially enabling the termination of the pregnancy to prevent maternal stress and complications.

Conclusion: Conjoined twins should be identified as soon as feasible to establish the best course of management for both mother and fetus. Ultrasonography and MRI are modalities for determining the diagnosis and prognosis of conjoined twins.

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Highlights:

1. Conjoined twins have captivated mankind for centuries due to the rarity of this birth type. Nonetheless, physicians have constantly encountered difficulties in dealing with conjoined twins.
2. Early diagnosis of conjoined twins during prenatal examination is critical for ascertaining the prognosis of the fetus, guiding parental counseling over appropriate courses of action, and potentially enabling the termination of the pregnancy.
3. First-trimester ultrasonography and MRI are complementary modalities in determining the diagnosis and prognosis of conjoined twins in early pregnancy.

INTRODUCTION

Conjoined twins occur due to abnormalities in the process of embryogenesis in monoamniotic monochorionic pregnancies, which are proposed to have resulted from either fission or fusion. Due to the imperfect division of one fertilized ovum, conjoined twins are identical monozygotic twins that do not entirely separate from one another but are still partially linked to one another.[1] The incidence of conjoined twins is 1 in 50,000 to 100,000 births. However, because 60% of them die in the womb or shortly after birth, the actual incidence can reach 1 in 250,000 live births. The ratio of women to men babies is 3:1.[2]

Conjoined twins are classified based on the area of the body and internal organs that are fused: 11% cephalopagus (joined at the head), 19% thoracopagus (joined at the chest with one heart), 18% omphalopagus (lower abdomen), 11% ischiopagus (lower abdomen and thoracic system genitourinary), 28% parapagus (joined on the sides of the body and pelvis), 5% craniopagus (cranium), 2% rachipagus (vertebral column), and 6% pyopagus (sacrum).[3] Based on the prognosis, conjoined twins are classified into 2, survived and non-survived. Survived conjoined twins consist of separable and non-separable twins. Cephalophagus is frequently missed as a singleton pregnancy because of the extreme degree of fusion. The type of conjoined twins, cephalothoraco-abdominopagus itself, is a scarce type of conjoined twins with a poor prognosis. Prenatal diagnosis of conjoined twins is crucial to determine the prognosis so that it can properly manage both the mother and the fetus. The tools that can be used for early detection are ultrasonography and MRI.[4]

Conjoined twins can be challenging to diagnose accurately, especially in the early stages of pregnancy. Sometimes, a diagnosis might be ambiguous, necessitating additional imaging or specialist testing. Referral to a specialized prenatal facility or consultation with a fetal medicine expert may be advised under challenging circumstances. This case report will describe a rare conjoint twin case that could be diagnosed early in pregnancy, and can be appropriately managed in early pregnancy.

CASE REPORT

A 27-year-old woman, gravida 4, para 1, abortion 2, child alive 1, was referred to the type A referral hospital with suspicion of multiple congenital abnormalities with two hearts. The patient received antenatal checks twice at the public health care and once times at an obstetrician during pregnancy. Anamnesis obtained a history of twin pregnancies from the husband's grandmother. General examination was recorded within normal limits. An obstetric examination found fundal height three fingers below the umbilical, a positive fetal heart rate was observed, and an ultrasound examination revealed multiple congenital anomalies. The patient had been taking pregnancy vitamins and folic acid since five weeks of gestation.

Ultrasound results at 17/18 weeks of gestation at Dr. Soetomo Hospital showed conjoined twins fused from head to urogenital part (one thalamus, two cerebellum, two hearts, two lungs, fetal hydrops, and spina bifida abnormalities were seen in one of the fetuses). The MRI confirmation showed a craniopagus, suspected meningomyelocele, and severe bilateral hydronephrosis in the second baby. Based on the ultrasound and MRI results, it showed a bad prognosis because it was categorized as non-separable and non-survived conjoint twins. The multidisciplinary conjoint twin team discussion decided to terminate the pregnancy.

In non-separable cases, if it is discovered early in pregnancy, there is a possibility of terminating the pregnancy or maintaining it until term or the patient goes into labor spontaneously. However, if the decision waits until term, there is a possible risk of having a cesarean section at the time of delivery. If the diagnosis is early, it allows for early termination so that no surgery is required and prevents stress on the mother. The patient was informed of her pregnancy's prognosis and management options. The patient and her family decided to terminate it. The fetus was terminated at 20 weeks of pregnancy by inducing a combination of misoprostol 200 mcg vaginally every 6 hours and the insertion of a balloon catheter. The fetus was born weighing 400 g and has a length of 20 cm. There were rostral conjoined twins, two heads with two faces in opposite positions, each with two eyes, one nose showing nasal proboscis, two ears, and one mouth. The head is fused to the thoracic and lower abdomen, and an omphalocele was obtained in one fetus with one umbilicus and one placenta. Siamese twins have two pairs of arms and legs. However, the patient refused to undergo an autopsy.

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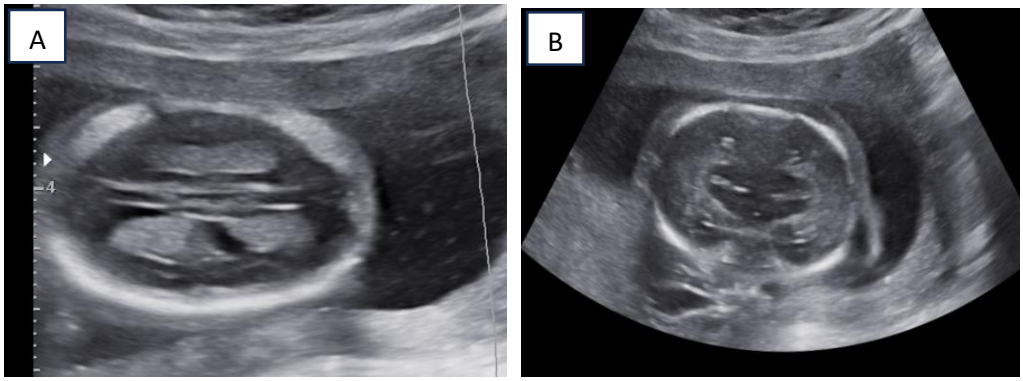

Figure 1. 2D ultrasonography results in a) Arrows indicate two choroid plexuses, b) Two fetal heads fused at the thalamus.

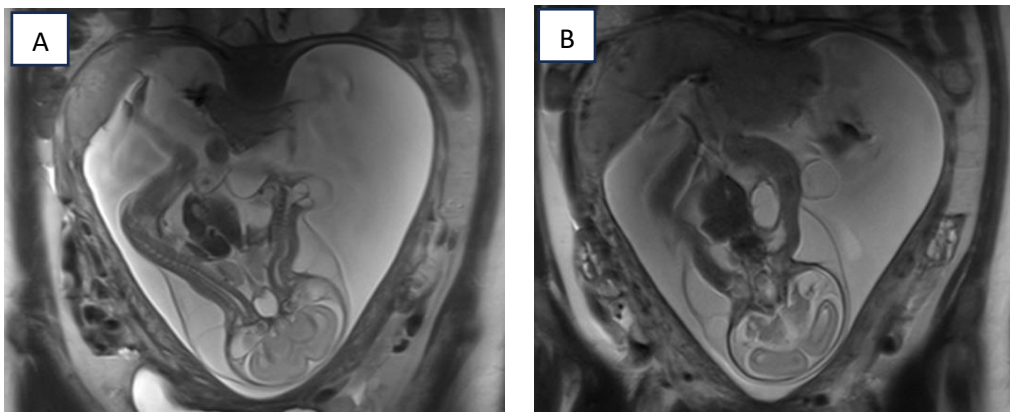

Figure 2. MRI results show two cerebellum lobes, two spines, two hearts, two lungs, and two livers. This organ fused on head to abdomen. Hydronephrosis and meningomyelocele are also seen in this small fetus.

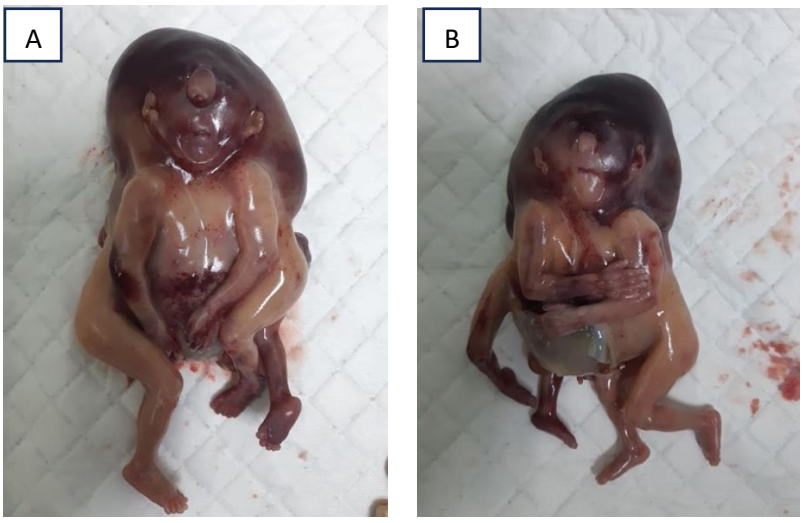

Figure 3. a) anterior part, b) posterior part

DISCUSSION

Conjoined twins have captivated mankind for centuries due to the rarity of this birth type. Nonetheless, physicians have constantly encountered difficulties in dealing with conjoined twins.[5] The embryological process of conjoined twins has yet to be discovered clearly. There are various theories on the formation of conjoined twins: a) Fission theory, which states that conjoined twin results from division in the early stages of embryo formation that comes from the fertilization of one ovum; b) Fusion theory, which suggests that conjoined twins result from two, initially separate monozygotic embryos, which coalesce and become secondarily and homologously fused (cephalo-pagus, thoracopagus, omphalopagus, ischiopagus, and parapagus).[3] Research on conjoined twins finds that union is homologous: head to head, buttocks to buttocks, chest to chest, back to back, sides to sides, but never head to buttocks or chest to back.[6]

Conjoined twins are classified based on the area of the body and the fused internal organs. The most common conjoined twins found are the thoracopagus, omphalopagus, and thoracic-omphalopagus types, with an incidence of around 56% of the total number of conjoined twins.[7] Cephalopagus conjoined twins fused from head to umbilicus are the rarest type. This type has one cranium and two opposite faces, with one face usually rudimentary. Another finding in cases of cephalopagus can be found in the lower abdomen and pelvis, which are separated by two pairs of hands and feet.[8]

Seventy percent of conjoined twins die within twenty-four to forty-eight hours after delivery or have a lethal congenital disease caused by the untimely diagnosis, which delays the implementation of optimal surgical treatment. Thus, early diagnosis and treatment are preferred.[9,10] Prior studies utilized ultrasonography to diagnose conjoined twins between 11 and 13 weeks of gestation.[11] Recent studies have documented the diagnosis of fetal abnormalities in twin pregnancies as early as 8 weeks gestation. Nevertheless, precise assessment of shared anatomical components remains unattainable.[5] Pregnancy termination remains the most effective course of action, regardless of gestational age, but especially during the early stages.

The diagnosis of conjoined twins in early pregnancy typically relies on prenatal ultrasound imaging. Some steps involved in diagnosing conjoined twins during the early stages of pregnancy are: early ultrasound examination, identification of gestational sac and embryos, assessment of fetal anatomy, evaluation of the placenta and umbilical cord, and proper follow-up examination. The ultrasound examination enables the identification of conjoined twins during prenatal diagnosis as early as 12 weeks of gestation. However, examination at 18-20 weeks of gestation is recommended for a more comprehensive assessment. Another modality that can be used is magnetic resonance imaging (MRI), which offers enhanced precision in providing anatomically detailed radiological images compared to ultrasound.[12–14]

In the present case, a prenatal ultrasound assessment is performed during the 17th to 18th weeks of pregnancy. The result showed that the cranium and the thalamus are joining, affecting the brain's structure. Furthermore, one of the fetuses exhibited the presence of an omphalocele and spina bifida. A magnetic resonance imaging (MRI) test was conducted to clarify the diagnosis further. The magnetic resonance imaging (MRI) scan reveals an image depicting two vertebrae, each fetus exhibiting distinct anatomical structures such as individual hearts, livers, and kidneys. Additionally, both fetuses exhibit the presence of a single pair of hands and feet. The tests showed that the conjoined twin fetuses, which were diagnosed as cephalo-thoraco-abdominopagus, are babies that cannot be separated and will not live.

In general, conjoined twins have a poor prognosis. The survival rate is indeed 7.5%. Survival rates for surgically separated cases are as low as 60%.[15] An improved prognosis could result from antenatal imaging, postnatal surgery, tissue expansion during surgery, and cadaveric transplantation for important organs shared by the twins, if applicable.[16] Legal abortion should be contemplated in Indonesia when a fetus has a life-threatening congenital abnormality with a poor prognosis, especially cephalon-thoraco-abdominopagus twins, whose survival rate is low and unlikely to be successfully separated.[17]

Early diagnosis of conjoined twins during prenatal examination is critical for ascertaining the prognosis of the fetus, guiding parental counseling over appropriate courses of action, and potentially enabling the termination of the pregnancy. First-trimester ultrasonography continues to be the most effective diagnostic modality in early pregnancy. Additionally, prenatal magnetic resonance imaging can assist in tissue characterization, conjunction type identification, and the detection of embryological malformations.[18] Once applicable, contemporary techniques such as 3D printing may facilitate surgical pre-planning and subsequent separation.[13] A viable pregnancy is easier to terminate vaginally if the diagnosis is made before that time, which may lessen the risk of trauma.[19] As the diagnosis progresses, the probability of achieving termination via cesarean section augments. Early

pregnancy termination is considered a safer option due to its potential to mitigate the emotional impact on the couples, which could be exacerbated by the numerous interdisciplinary follow-ups that are required throughout the prenatal and postnatal phases.[20] The effective management of conjoined twins necessitates the close collaboration of a multidisciplinary team.[21]

CONCLUSION

Conjoined twins of the cephalon-thoraco-abdomino-pagus type are a very rare type of conjoined twin with a poor prognosis. An early prenatal diagnosis provides a good outcome for the mother. Ultrasonography and MRI are complementary modalities in determining the diagnosis and prognosis of conjoined twins.

DISCLOSURES

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Conflict of interest

The author reports there are no competing interests to declare.

Patient consent for publication

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