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Case report : Dicephalic parapagus conjoined twins

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***Abstract*---**Dicephalic twins account for around 11% of all conjoined twins. Dicephalic parapagus is an uncommon form of lateral partial twinning characterized by two heads on one trunk. A 25-year-old primigravida was referred to our clinic with a conjoined twin diagnosis, which was made for the first time at 29 weeks of gestation. The fetus had two heads, two separate necks, two vertebrae, a fused heart with two apexes, four ventricles, three atriums, two gastrics, a fused liver, four kidneys (one of which had hydronephrosis and the other concurrent hydroureteric), two bladders, one male gender, two arms, and two legs, according to an ultrasound. A second case is a pair of conjoined twins that were discovered at 20 weeks of gestation and are 29 years old. The results of an ultrasonographic test showed that the fetus has two vertebrae, one heart, one hepar, one umbilical insertion, and one foot with a congenitally equinovarous talus and a rocker bottom. Both pregnancies ended due to family requests for termination and after a conjoined twin committee meeting in our hospital was approved and the unseparable conjoined twin fetus diagnosis was made. The size of two heads necessitated a Caesarean operation. The degree of organ fusion and the severity of the associated abnormalities are both related to the prognosis of conjoined twins. If requested by the family and accepted by the hospital committee, early antenatal identification may have an impact on the treatment of pregnancy termination.

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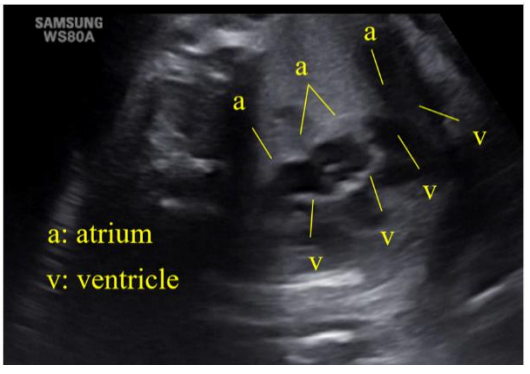
Introduction

Monozygotic monoamniotic twins with conjoined twins are an uncommon abnormality that affects 1 in 50,000 to 1 in 100,000 newborns (Guessan et al., 2020). Conjoined twins, according to the fission theory, are a result of an incomplete division of a fertilized ovum. This syndrome is caused by two monovular embryos that are originally different, in accordance with the secondary fusion idea (Watanabe et al., 2016).

The location of the body fusion is typically used to categorize conjoined twins. The seven varieties of conjoined twins that are most frequently found are thoracocephalopagus, thoracopagus, omphalopagus, craniopagus, cephalopagus, and xiphopagus. Conjoined twins that have dicelike heads are a rare type. Before implantation, the zygote fails to divide at neck level, resulting in the twins fusing below the neck, leading to this uncommon dicephalic head abnormality (Singh et al,. 2017). When the line of fusion extends caudally from the lower abdomen to the pelvis on the abdominal side, it is called a parapagus. Parapagus dicephalus are parapagus twins with two distinct heads (Karn & Mahato, 2021).

Case Reports

Our first case was a 25-year-old primigravida at 29/30 weeks gestation who was referred due to suspicions of having conjoined twins. The patient was suspected of having conjoined twins at 28/29 weeks gestation based on the history of antenatal exams. After that, the patient was sent to a tertiary hospital for a diagnosis confirmation. The patient had requested a second opinion from a different physician. Conjoined twin parapagus dicephalic was identified from the ultrasound testing results. In order to be sent to a tertiary hospital, the patient consented. In an ultrasound examination, it was discovered that the fetus had two heads, two separate necks that were fused at the clavicle, two vertebrae, a heart with two fused apexes, four ventricles, three atria, two stomachs, a fused liver, four kidneys (one of which had hydronephrosis and the other concurrently had hydroureteric kidney), two bladders, one male gender, two arms, and two legs A parapagus dicephalic conjoined twin was found to be non-survival and non-separable.



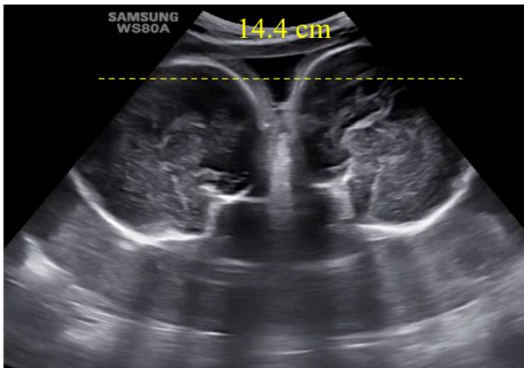


Figure 1. The fused heart

Figure 2. Diameter of both heads

Figure 2. Parapagus dicephalic

Figure 3. Infantogram

Following discussions with the family and the RSUD dr. Soetomo Surabaya Conjoined Twin committee over the status of the fetus, the family decided that the pregnancy should come to a stop. After signing the informed consent and informed to consent forms, it was decided to end the pregnancy by cesarean section, taking into account that the two fetal heads measured 14.4 cm in diameter. The newborn's APGAR score was 3-5-7, and their weight was 2500 grams. The babies were given ventilator care, but it only kept them alive for 27 hours until they passed away from heart failure caused by disorders connected to cardiac fusion.

In the second case, a 29-year-old lady was referred to the clinic with a diagnosis of second gravida, 20/21 weeks gestation, conjoined parapagus dicephalic twins with one heart, and cardiomegaly. The fetus I (left) had Congenital Talipes Equina Varus, a rocker bottom, and an absent right lower extremity when an ultrasound was done. It also had thickening in the occipital area of 1.5 cm, lymphangioma colli dextra of 2 x 1.4 cm, scalp edema, and bladder filling (single foot). Fetus II (right) BPD21/22 weeks, determined to have Congenital Talipes Equina Varus, a full bladder, fused liver, heart, and liver, as well as cardiomegaly and a single umbilicus insertion. concluded that the conjoined twins were parapagus dicephalic and unable to be separated.

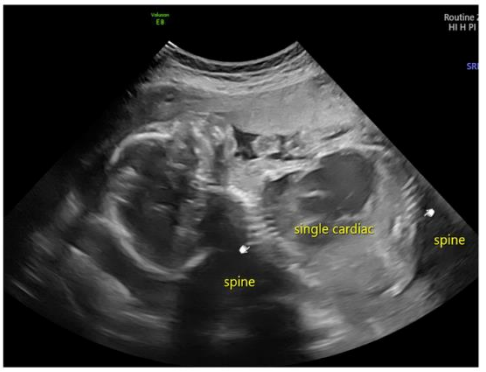


Figure 5. Single cardiac Figure

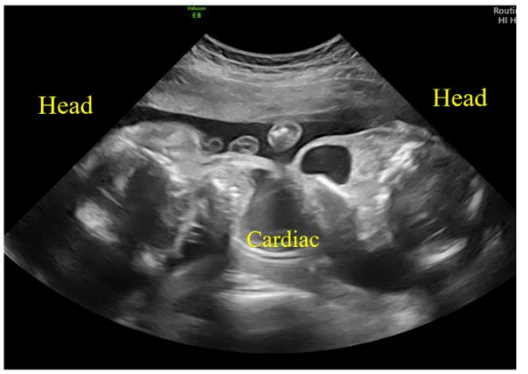


Figure 6. Dicephalic

Figure 7. Parapagus dicephalic conjoined twin

We spoke about the state of the fetus with the family and the RSUD dr. Soetomo Surabaya Conjoined Twin committee. In the end, the family wanted the pregnancy to be aborted. The decision was made to end the pregnancy by cesarean section after signing the informed consent and informed to consent forms, taking into account that the two fetal heads measured 11 cm in diameter. The newborn's APGAR score was 1-1-0 and their weight was 1000 grams. The infants passed away after 20 minutes due to heart failure brought on by a single cardiac ailment.

Discussion

Two competing ideas about embryology have been put forth to explain the clinical polymorphism of this abnormality. According to the fission theory, it is the second fusing of two previously split embryonic discs after the embryonic disc's final

incomplete separation (fusion theory) (Singh et al., 2017). The area of fusion between the two fetuses determines how conjoined twins are categorized. The ventral and dorsal sides of this union predominate (Guessan et al., 2020).

The joining of the twins' sides is referred to as the "parapagus" phenomenon. Parapagus conjoined twins either have one head with two different faces (dicephalic parapagus) or the same body but two distinct heads (diprosopus parapagus). They may have two to four legs and two to four arms (Guessan et al., 2020). The first case had two limbs and two legs, whereas the second case had four arms and three legs, one of which was underdeveloped.

Starting in the tenth week of pregnancy, two-dimensional ultrasonography allows for the early prenatal identification of this condition (Guessan et al., 2020). Fetal head in the same plane, no change in relative position following maternal movement, lack of a dividing membrane, more than three vessels in the cord central, and failure to separate the fetal body are all symptoms that can support the diagnosis (parai at al., 2016). Ideally, a diagnosis should be made in the first trimester so that the delivery can be scheduled to minimize maternal morbidity (Singh et al. 2017). In this report, the conjoined twin was discovered for the first time at 29 weeks and 20 weeks of gestation, respectively, making vaginal delivery impossible due to the combined size of the fetal head diameter. We decided to have a cesarean section for that reason which raises maternal morbidity.

Approximately 40% of conjoined twins are stillborn, and 35% pass away during the first 24 hours of life, according to the literature. Additionally, only 60% of postoperatively treated conjoined twins made it through the procedure. Dicephalus twins have extraordinarily high rates of stillbirth and mortality. The documented survival times for Dicephalic twins range from 15 minutes to 11 days, which is a fairly short time. This is because important organs including the heart, lungs, and liver have fused, resulting in anatomical abnormalities (Mahajan et al., 2020).

Size of the fusion of the essential organs determines the prognosis for conjoined twins, and in cases where there are no additional organ anomalies and surgical separation operation is possible, the decision to continue the pregnancy should be made (Taflc et al., 2010). The level of cardiac fusion is a prognostic factor for follow-up care. Twins with joined hearts are rarely successfully separated through surgery; instead, survival is typically influenced by how severe the cardiac abnormality is (Ince et al., 2012). In the first case, the infants made it through 27 hours before eventually passing away from cardiac fusion-related heart failure. While in the second case, after 20 minutes, infants passed away from heart failure brought on by a single cardiac arrest.

Conclusion

Parapagus with dicephalus variation is highly uncommon in conjoined twins, which is a rare occurrence (0.5 percent of reported cases). The shared anatomy of internal systems among individuals born is crucial to survival (6). The majority of dicephalic twins experience heart and abdominal organ defects, which in our cases resulted in death.

Especially in monochorionic and monoamniotic twin pregnancies, ultrasound examination during the first trimester is helpful for early detection of conjoined

twins. This allows for vaginal termination of conjoined twins if they are determined to be inseparable and unable to survive, reducing maternal morbidity.

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