



Case Report

Parapagus dicephalus conjoined twins with tribrachius and a single heart – A scarce variant of conjoined twins: A case report

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ARTICLE INFO

Keywords:

Parapagus dicephalus
Conjoined twins
Tribrachius
A single heart

ABSTRACT

Conjoined twins occurred in one of every 58,000 births. Parapagus dicephalus tribrachius with a single heart is a scarce variant of conjoined twins. A male parapagus conjoined twins with tribrachius was born by cesarean section with severe asphyxia, weighing 3170 g from 25 years old mother with G1P0A0, 35–36 weeks gestation. There is one umbilical cord, and the placenta consists of 1 amnion, one chorion. The baby has a single heart, a pair of the lung, a single stomach, duodenum, liver, double columna vertebralis with the single pelvis, one penis, undescended right, and left testes, one anus, and a pair of inferior extremities. After 28 h of treatment, the patient's condition became unstable, deteriorated, and died. This case reminds us of the importance of antenatal care and ultrasound examination since early pregnancy.

1. Introduction

Conjoined twins (CT) are very rare. Tan et al. reported the incidence of conjoined twins 1 in 58,000 births and 1 in 546 multiple births. [1] Generally, CT stillbirth or are only a few hours or days old. [1] Dicephalus twin is 11% of all CT. [2] Mutchinick reported that of 26,138,837 births recorded from 21 Clearinghouse Surveillance Programs, the prevalence of CT was 1.47 per 100,000 births with a female-male ratio of 2:1. [3] The female-male ratio in the type of thoracopagus 3.27: 1, parapagus dicephalus 2.23: 1, omphalopagus 4: 1. [3] This paper will discuss a rare case of parapagus dicephalus CT.

Case presentation:

A mother of 25 years old, G1P0A0, 35–36 weeks gestation, twins came to the hospital for vaginal water discharge. In the family, there is no history of twins. The mother only makes one consultation with the obstetrician at seven months' gestation (because the family unafforded financially for an ultrasound examination), and the doctor declared twins. The doctor said twins with rapture membranes, so an emergency caesar section should be done.

A male CT was born at RSD Idaman Banjarbaru on October 6, 2020. Twins are born attached to the chest and abdomen with two heads, two legs, three arms (two normal arms and one arm at the top of the body, abnormal but five fingers). APGAR score right baby is two at 1 min and three at 5 min, and left baby is one at 1 min and one at 5 min. Birth weight 3170 g, birth length of right baby 42 cm, birth length of left baby

33.5 cm. Right baby head circumference 30 cm, left baby 32 cm. There is one umbilical cord (consists of one large vein and two small arteries), and the placenta consists of 1 amnion, one chorion. The baby has one penis and undescended left and right testicles. The anus is patent, and only one; 2 ½ hours later, there is meconium.

On physical examination, Lungs: bronchovesicular breath sounds. Heart sound was heard on the left and the right, with no murmur. The twin was stable. An umbilical cord infusion was placed, and an oral gastric tube for each baby. The picture of the CT can be seen in Fig. 1.

In Fig. 2, the X-rays babygram showed the heart is only one, and the size appears enlarged. The lung is only one (right and left) with a normal appearance. One pair of superior extremities showed the humeral, radial, and ulnar. Single stomach and duodenum, single liver. The contour of the right and the left kidney is not visible. Double columna vertebralis (cervical to lumbar) with fusion on the ribs 3,4,5, 6,8, 9, 10, 11,12, VL 1 medial part. Single pelvis and a pair of inferior extremities. Thus, the baby was fused from the thorax to the pelvis. The diagnosis is parapagus dicephalus tribrachius with a single heart.

Considering the condition is not viable to separate and not transportable to be referred to a tertiary hospital, it was decided to be treated at RSD Idaman Banjarbaru. After 28 h of treatment, the patient died.

2. Discussion

The etiology of CT is indecisive. The most accepted theory is an

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Fig. 1. Gross photograph of parapagus dicephalus tribrachius.



Fig. 2. Conventional babygram of the conjoined twins.

incomplete division of the monozygous embryo that occurs 13–15 days after fertilization. CT can be equal or unequal depending on the duplication structure. [4] The development of fetal embryology until parapagus CT is very complicated. Parallel duplication of two notochords that are very close together will cause twins to conjoined laterally. The parapagus is always attached anterolateral, united in the caudal part, and separated in the cranial part. In the parapagus, there are generally parts of the body missing because of the two notochord-distance that is so close. The origin of the two embryos' fusion determines the extent of the fusion resulting in many differences in these twins. [4,5] Generally, parapagus are conjoined to the upper chest. Parapagus, united laterally, always share a conjoined pelvis with one symphysis pubis and one or two sacrum. Dithoracic parapagus when the two chests are separated, and the fusion is confined to the pelvis and abdomen. If there is the union of the entire trunk but not the heads, they are dicephalic parapagus. The heart, liver, and diaphragm are fused, but there is a duplication of the respiratory tract and upper digestive tract; the viscera organs are fused. There are two arms, two legs, and two complete spinal cord and vertebral column. The number of limbs varies from 4 to 7, rarely with four legs. Generally, each lung is in a separate lung cavity. The fusion of lungs will be rare. The configuration of the conjoined pelvis is a diagnostic-one complete pelvic ring, with a single anterior symphysis pubis, and with two laterally fused sacrum, there is usually only one rectum. [5]

Ischiopagus are united ventrally from the umbilicus down to a sizeable conjoined pelvis with two sacrum and two symphyses pubis. Craniopagus are united on any portion of the skull except the face and the foramen magnum. Pygopagus are joined dorsally; they share the sacrococcygeal and perineal regions, sometimes even the spinal cord. Rachipagus are fused dorsally above the sacrum. The union may involve the occiput. The cephalopagus are fuses from the umbilicus to the top of the head. Usually, the pelvis and lower abdomen are not fused. Thoracopagus are united face-to-face from the upper thorax down to the umbilicus. Omphalopagus are joined face-to-face, primarily in the umbilicus area. The pelvis is not united. [5]

The pure parapagus is two heads, two hands, two legs, two hearts, two pairs of lungs. [6] In this case, the parapagus, not pure parapagus, showed rare variants, namely two heads, three arms, two legs, one pair of lungs, a single heart, liver, pelvis, undescended right, and left testes. This case suited to literature that said the dicephalus with 2–3 arms is usually accompanied by one heart or severe heart defects or is accompanied by an intricate fusion of the intraabdominal organs. It is generally stillborn or survives only a few hours to a few days. [7,8] This case cannot be separated.

The obstetrician did not expect twins born to be CT. The baby was born with severe asphyxia. The patient's condition was worsened by infection may be due to premature membrane rupture. Usually, patients with a shared heart and lung are only viable in a matter of minutes or hours. In this case, the baby lived 28 h. Willobee et al. reported that out of 240 CT infants, mortality was highest in hospitals not designated as children's hospitals (72%) compared with children's hospitals (44%) ($p = 0.007$). [9]

Supposed the patient had checked with ultrasound examination from the beginning, then the possibility of CT diagnosis would have been established earlier. Termination of pregnancy can be considered. Early diagnosis can reduce the mother's mental stress during pregnancy and face the fact that her child cannot be saved by separation. [10]

3. Conclusion

This case reminds us of the importance of antenatal care and ultrasound examination since early pregnancy.

4. Ethics statement

This case report has been approved by The Ethical Committee of

Medical Research, Medical Faculty, University of Lambung Mangkurat, Banjarmasin – Indonesia, No. 519/KEPK-FK ULM/EC/I/2021.

Funding

No funds, no grants, or other support was received.

Patient Consent Statement

All patients gave written informed consent.

Declaration of Competing Interest

The author declares that the author has no known competing financial interests or personal relationships that could have influenced the work reported in this paper.

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