

A Case of Ischiopagus Dicephalus Conjoined Twins with Tetrabrachius Bipus from Dessie, Ethiopia

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Abstract: The development of conjoined twins always catches the eyes of researchers and clinicians. Beyond the rareness of the cases, how they develop is a debatable issue. This report presented a case of ischiopagus conjoined twins who had two heads (dicephalus), four upper extremities (tetrabrachius), and were joined below the chest with two lower extremities (bipus). The twin's mother was referred from a primary hospital to Dessie Comprehensive Specialized Hospital for proper management of twin pregnancy, where a cesarean section was performed. The mother and her husband have no family history of birth defects or exposure to known teratogens. On imaging, the twins had separate hearts, lungs, and kidneys but a single liver, spleen, stomach, and intestine. They also shared genitourinary structures: a single penis with sub-coronal hypospadias and one imperforate anus. In addition, their placenta was single with one umbilical vein and two umbilical arteries. The conjoined twins had multiple accompanying cardiovascular anomalies but no external craniofacial, extremity, or brain anomalies. They passed away after 36 hours of follow-up in the neonatal intensive care unit at Dessie Comprehensive Specialized Hospital.

Keywords: conjoined twins, bipus, dicephalus, ischiopagus, tetrabrachius, Dessie Hospital

Introduction

Conjoined twins are developed from fertilization of a single ovum by a single sperm cell and separation of the embryo at an early embryonic period, beginning from the third week of intrauterine development.¹ Conjoined twins can also be formed through the fusion of previously formed monozygotic twins.² Conjoined twins are classified based on the site of fusion. Thoracopagus (fusion at the chest), omphalopagus (fusion in the abdomen), thoraco-omphalopagus, ischiopagus, and parapagus are the most commonly reported varieties of conjoined twins.³ In addition to the union sites, the conjoined twins may have variable limbs. Dicephalus (two heads), tetrabrachius (four upper limbs) and bipus (two lower limbs) is one of the rarest forms of conjoined twins.⁴

Generally, multiple factors, such as alcohol consumption, drug use, radiation or chemical exposure, and lack of folic acid supplementation during pregnancy, as well as family history, are reported risk factors for the development of congenital anomalies.^{5,6} Specific to conjoined twins, the exact etiology has not been discovered yet. However, in many literatures,^{2,7,8} the theory of fusion (the two identical twins united together at some specific site) and the theory of fission (no complete division of the embryo) are mentioned to explain the embryologic basis of conjoined twinning.

The fission theory states that the twins arise from the splitting of a zygote at specific stage of embryonic development.⁹ The time of splitting matters the extent of sharing embryonic structures and fetal membranes (having a common or separate amnion, chorion, or placenta). If the separation occurs at the bilaminar embryonic disc stage, the twins can have one placenta and common chorionic and amniotic membranes.⁹ When splitting occurs after germ layer formation (third to fourth week of development), the twins can share specific organs.¹

Currently, some researchers advocate the theory of secondary fusion of monozygotic twins as a cause of conjoined twins.^{2,8,10,11} They stated that conjoined twins are formed by the union of initially distinct embryos. The union may not

occur when the skin or ectoderm is intact, but when ectodermal agenesis or break exists.¹¹ According to the assumption of Spencer (2003),¹² the union always occurs in the midline. Twins which are dorsally united can be formed by secondary fusion of two initially separate monozygotic twins.¹³

In addition to the theory of fission and fusion, a third theory, the crowding theory, is advocated to explain the causes of conjoined twins. In this theory, the presence of “initial duplication of axially located morphogenetic potent primordia in one inner cell mass” is the initiating factor in the development of ventrally, caudally, and laterally united twins.¹³

Many congenitally anomalous pregnancies are terminated before birth.¹⁴ This situation reduces the true incidence of reported congenital anomalies. The incidence of conjoined twins has been reported as 1:50,000 to 1:100,000.¹⁵ However, the actual incidence can reach 1:250,000.³ The final outcome and survival of conjoined twins depend on the site of fusion or the structures they share. Almost half of the conjoined twins are stillborn.³ Craniopagus/cephalopagus (fusion at the head/cranium) and thoracopagus (union at the chest) types of conjoined twins are usually difficult to separate and have a poor survival rate.^{14,16}

In Ethiopia, the true incidence is not reported, but there are limited reports disclosing the cases of conjoined twins. Some of these are omphalopagus conjoined twins,¹⁷ parasitic twins,¹⁸ dicephalus tetrabrachius,¹⁹ and thoraco-omphalopagus conjoined twins.²⁰ Our article reported a case of liveborn ischiopagus dicephalus tetrabrachius bipus conjoined twins.

Case Report

The neonates (ischiopagus dicephalus tetrabrachius dipus conjoined twins) were referred to the neonatal intensive care unit of Dessie Comprehensive Specialized Hospital (DCSH) following a cesarean section delivery. The twins were born to a 29-year-old para two mother who did not remember her last normal menstrual period but claimed to be amenorrheic for the last 9 months. The mother had antenatal care follow-up at a nearby primary hospital but was not supplemented with iron and folic acid during her pregnancy. The area where she lived was a war zone when the Tigray People's Liberation Front (TPLF) forces fought with the Ethiopian army. She had obstetric ultrasound imaging at fifth and seventh months and was told she had a normal twin pregnancy. She had a family history (her sister) of normal twin delivery. She has no other self or family history of significant risk factors for congenital anomalies and was not exposed to known teratogens during her pregnancy. Her husband is 30 years old and has no history of smoking, drug intake, and chronic diseases, as well as self or family history of congenital anomalies.

Her labor started spontaneously at home, and then she went to a nearby primary hospital where she was immediately referred to DCSH for further management of the twin pregnancy. At DCSH, an ischiopagus dicephalus tetrabrachius dipus conjoined twin was detected while undergoing ultrasound imaging and an immediate caesarean section was performed to deliver the twin. At birth, the newborns were of male sex, alive, active, weighed 4.7 kg together, and had a single placenta and umbilical cord with two umbilical arteries and one umbilical vein. The APGAR score was 8 and 9 (right) as well as 7 and 9 (left) in the first and fifth minutes, respectively. The neonates were immediately referred to the neonatal intensive care unit for further evaluation and care by pediatricians.

On physical examination, the twins were conjoined below the chest (ischiopagus) with two heads (dicephalus), four upper limbs (tetrabrachius) and two lower limbs (dipus), [Figure 1](#). The right-side twin was acutely sick-looking and in respiratory distress with central and peripheral cyanosis. Moreover, the head circumference and height of the right neonate were, respectively, 37 cm and 50 cm. In addition, he has a clear chest with good air entry, but subcostal and intercostal retraction as well as use of accessory muscles for respiration were observed. On cardiovascular examination, there was a grade III holosystolic murmur at the left lower sternal border and also a high-pitched, mid-systolic ejection murmur best heard at the right upper sternal border radiating to the neck, possibly due to aortic stenosis. On echocardiography, there was severe left ventricular hypertrophy secondary to aortic stenosis and a large ventricular septal defect. The laboratory investigation results for the right-side neonate were as follows: blood group; B+, white blood cell count (WBC); 17,220/ μ L, haematocrit; 42%, and platelet count; 221,000/ μ L.

On the other hand, the left neonate (height 49 cm and head circumference 36 cm) had no significant physical findings on the respiratory and cardiovascular systems, but dextrocardia was observed on echocardiography. The complete blood



Figure 1 Photograph of ischiopagus dicephalus conjoined twins with tetrabrachius bipus live born in Dessie Comprehensive Specialized Hospital.

count (CBC) for the left neonate was blood group; B+, white blood cell count (WBC); 15,030/ μ L, haematocrit; 39.6%, and platelet count; 68,000/ μ L. The abdominal ultrasound revealed that they have a common liver, stomach, spleen, and intestine. However, the kidneys were four, two for each. The genito-urinary system (cloacal region) is shared. As a result, they have common and imperforated anus, as well as a male type external genitalia with single penis, testis in the scrotum and subcoronal hypospadias. The twins had no external craniofacial anomalies, limb deformities, or brain anomalies. Finally, both new-borns died after 36 hours of follow-up in the neonatal intensive care unit.

Discussion

Conjoined twinning is a rare congenital anomaly and the exact mechanism by which it develops is still a debating agenda. In Ethiopia, like many Africans, giving birth to an anomalous baby is stressful to the parents; they feel anxious and guilty. It is considered a punishment for sin or an expression of God's anger.²¹ The current case report presented one of the uncommon types of conjoined twins that are joined below the chest but have two heads and four upper extremities.

Etiopathogenesis of conjoined twins is still a controversial issue. The older concept, predominantly found in text books, is the fission theory, which states that splitting of a zygote at some embryonic period results in conjoined twins,⁹ whereas, currently, fusion of an already formed monozygotic twins^{2,11} and the presence of initial duplication of morphogenetic potent primordia are considered as a cause of conjoined twins. In this case, it seems that the caudal part of the embryo failed to completely separate and gave rise to a fused lower abdomen, pelvis, and lower extremities. This suggests that the cause of the current conjoined twin, as explained by Kaufman (2004),¹ may be an incomplete separation of the embryo.

Recently, the presence of imaging techniques has facilitated the early diagnosis of conjoined twins.²² In our case, even though the mother had two episodes of obstetric ultrasound imaging in the fifth and seventh months of her pregnancy, she was not aware of conceiving conjoined twins. Thus, she has lost the chance to terminate her pregnancy. This may be due to the poor health care delivery system or lack of experienced obstetricians in the primary hospital. The diagnosis was known at DCSH immediately before the cesarean section was performed. When the anomaly is

incompatible with postnatal life, termination of the pregnancy has been recommended and this option should be made available to the families.⁴

In the conjoined twins, the structure they share or own independently is one point of discussion. In our case, the twins have separate lungs, which are paired for each. In some other case reports, the bilateral lungs are reported to be mirror images of each other²³ or one lung is well developed, whereas the counter part is hypoplastic.⁴ Sethi et al²³ reported the presence of different cardiac anomalies, especially on the right side of dicephalus twin. Similar to this, in our case, severe left ventricular hypertrophy, aortic stenosis, and ventricular septal defect in the right-side twin and dextrocardia in the left side twin were observed. On the contrary, Ibinaiye et al⁴ reported no anomalies in the right-side heart but a rudimentary heart on the left-side twin. This indicates that the degrees of heart-related anomalies are variable and greater attention should be paid to this region, especially while trying to separate the twin. In the current case, there was a single liver, for both twins. This is consistent with other similar case reports.^{4,23} In addition to this, we found a single spleen, stomach, intestine, and four kidneys (two for each).

The prognosis of conjoined twins depends on the structure they share and the site of union. In the current case, both twins died after 36 hours of care and follow-up in the intensive care unit without getting the option of surgical separation. This may be due to the presence of accompanying cardiac anomalies in both twins. Different degrees of survival, after surgical separation or joined together in situ, have been reported by different investigators.^{24,25} However, most conjoined twins are still born or die in the early neonatal period.²⁵

In conclusion, our case report presented one of the rare types of conjoined twins, probably the second case from Ethiopia, twins with two heads, four upper extremities, and two legs. In the antenatal care service, early diagnosis of conjoined twins should be maintained and the option of early termination of anomalous pregnancy should be provided to the families. In addition, a complete antenatal care service, including folic acid supplementation, should be provided for every pregnant mother.

Ethical Approval and Consent Form

Written informed consent was obtained from the twin's mother and father for taking the picture and publishing the case. We have also obtained ethical approval from the hospital.

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Disclosure

All authors declare no conflicts of interest in this work.

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