

Dicephalus dipus tetrabrachius conjoined twins of Zaria: Case report and literature review

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Abstract

One of the most interesting congenital malformations to manage is a conjoined twin. Conjoined twins are rare occurrences in obstetric/pediatric practice. More commonly known as Siamese twins, this phenomenon is shrouded in mystery and considered a curiosity by general public. Current technology is lending a helping hand in the early diagnosis of these conditions. Frequently, the twins are born dead, but there are few cases in which the twins survive. We presented a case of dicephalus dipud conjoined twins; a rare type of conjoined twins.

Key words: Conjoined twins, dicephalus, dipus, tetrabrachius

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Introduction

Conjoined twins are very rare, with a variable reported world incidence of about 1 in 50,000 to 1 in 100,000 births^[1,2] and the dicephalus dipud type is among the rarer forms of the congenital anomaly.^[3] The incidence of conjoined twins in Nigeria is not known. To our knowledge, 15 cases have been reported from Nigeria so far. This probably does not reflect the true incidence in Nigeria as most of the cases are not reported on account of the associated stigma.^[4] Cases of conjoined twins seen in hospital are rare and pose a management challenge when they present because of paucity of experience in managing them.^[4] The present case is of interest because of its extreme rarity, and the fact that it is the first such dicephalus dipud type to have been managed in our centre.

Case Report

A 29 year old woman gravida3, para2, with uneventful gestation delivered conjoined twins at home. It was a full term, abnormal delivery with breech presentation. There was single placenta and umbilical cord. The babies were

noted to be joined but were alive. She presented to our hospital two days after delivery at home. She did not receive antenatal care and she had no antenatal ultrasound scan report. Her maternal grandmother had a history of twins.

There was no history of local herbs or drug use or alcohol consumption in early pregnancy. Her maternal grandmother had a history of twins. On clinical examination it was noted that the twins were joined from level of xiphisternum onwards. The twins had two heads, two pairs of upper limbs, a shared pelvis and a single pair of lower limbs (dicephalus, tetrabrachius, and dipus twins). There was a single penis and anus. Both twins weighed 5.3 Kg, but the right twin appeared bigger than left. Heartbeat was auscultated for the twin on the right with a heart rate of 146 beats per minute, no murmur was evident clinically. No heartbeat was detected in the thoracic cavity of the twin on the left; however, the right twin had respiratory rates of about 45 cycles per minute and none was observed on the left twin. However, there was movement in the upper limbs of the left twin.

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An infantogram [Figure 1] done to assess the twins further confirmed the clinical findings. The babies had two heads, two thoraces, two pairs of upper limbs and a shared abdomen, pelvis and a single pair of lower limbs. Echocardiography revealed that the heart in the right twin was morphologically normal, with no evidence of any gross cardiac anomaly, but the heart in the left twin was rudimentary. Computerized tomographic scan [Figures 2a and 2b] revealed that the twins had two separate thoraces, paired thoracic diaphragm, hearts and vertebral columns, a shared liver and spleen and a single pair of hydronephrotic kidneys in the right twin. The right twin had normally formed intestine, but the intestines in the left were atretic.

An emergency surgical operation was carried out to separate the twins in view of progressive sepsis in the twin on the left on the third day of admission. The operative findings were similar to that of the imaging findings, the anus and external genitalia were given to the twin on the right. Nasogastric tube was inserted in both twins as part of resuscitation measures and there was no oesophageal atresia on both sides. The twin on the left stopped breathing shortly following the separation. The twin on the right survived for two weeks but later succumbed to severe sepsis and disseminated intravascular coagulopathy (DIC) and died on the 21st post operative day.

Discussion

There are various explanations for the development of conjoined twins. Four days after fertilization the trophoblast (chorion) differentiates, if the split occurs before this time the monozygotic twins will implant as separate blastocysts each with their own chorion and amnion. Eight days after fertilization the amnion differentiates, if the split occurs between the 4th and 8th day, then the twins will share the same chorion but have separate amnions. If a split occurs



Figure 1: An infantogram revealed conjoint twins with two heads, two thoraces, two pair of upper limbs, a shared abdomen, a single pelvis and a pair of lower limbs

after 8th day and before the 13th day, then the twins will be sharing of their chorion and amnion. This is a very rare condition and accounts for 1-2% of monozygotic twins. The embryonic disk starts to differentiate on the 13th day. If the split occurs after day 13, then the twins will share body parts in addition to sharing of their chorion and amnion.^[5]

The classification of conjoined twins is based on the site of union. The suffix-pagus is used meaning fastened. Thoracopagus means shared thorax, with 90% having a shared heart. Omphalopagus means shared abdomen. Thoraco-omphalopagus is one of the most common types where both the thorax and abdomen are shared. Ileopagus twins are connected at the iliac bone. When the twins are extensively connected then the duplicate part is named, for example, dicephalus refers to two heads with one body.^[5] Regardless of the site of union, variations occur with regard to the internal organs.^[3] Certain organs may be common to both twins or these may be separate. In the thoracopagus, the heart is often conjoined with associated cardiac anomalies.^[5] In this case report, there were two separate thoraces and two hearts, although a rudimentary heart was found in the left twin. In omphalopagus the liver is often conjoined as seen in this case report. Prognosis, obstetric management and treatment planning are determined by degree of fusion and extent of joining of fetal organs.^[6] Cesarean section is recommended in most of the cases of third trimester deliveries because of the high incidence of dystocia with resultant fetal damage.^[6] In this case report, the patient did not attend antenatal clinic throughout her pregnancy and also she delivered per vaginam at home and therefore did not benefit from any prenatal care. However, the conjoined twins did not suffer any dystocia with resultant fetal damage.

Antenatal diagnosis by ultrasound is possible in modern day obstetrics. Ultrasonographic identification of any of the following classical signs may suggest the diagnosis: Both fetus heads in the same plane, unusual backward flexion of the cervical spine, no change in the relative position after maternal movement and manual manipulations and inability to separate fetal bodies after careful observation.^[7]

Dicephalus dipud conjoined twins form a rare variant of conjoined twins. Such variants are usually still born or die

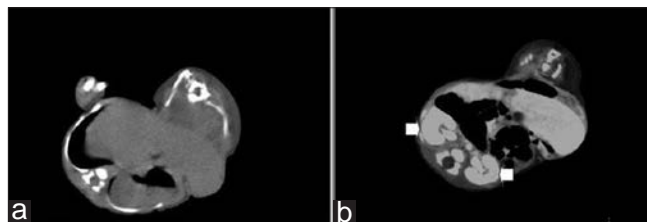


Figure 2: (a and b) Computerized tomographic scan revealed conjoint twins with two separate thoraces, hearts and vertebral columns, a shared liver and spleen and a single pair of hydronephrotic kidneys in the right twin. It also revealed right twin with normal formed intestine, but the left is atretic

immediately after birth, but some have lived for a number of years.^[3] In this case report, the twins were brought to the hospital alive after delivery at home. As most of the studies concentrate on the obstetric problems, the anatomical description of the dicephalus twins is often incomplete.^[3] A conjoined twin is frequently the mirror image of its partner and this is particularly true in dicephalus twins.^[3] For example the right set of lungs may be the mirror image of the left.^[3] But in this case report, the right twin had normal lungs while its left counterpart had hypo plastic lungs. The liver is always single^[3] as seen in this case report. There is usually only one spleen that is in the twin on left.^[3] In our case the only one spleen present was found in the right twin. The stomach in the right twin appeared normal i.e., J shaped while it was absent on the left. This is contrary to earlier findings by Sethi *et al.*^[3] that the stomach is always normal in the left twin and may be atrophic but usually present in the right twin. The intestines almost always join either just distal to the duodenum or at the level of Meckel's diverticulum.^[3] In this case the intestines joined just distal to the duodenum. Sheti *et al.*^[3] also reported that diaphragmatic hernias are also frequent in dicephalus, dipud conjoined twins but this was not found in this case report. Sheti *et al.*^[3] stated that the right sided twin of the dicephalus dipud twins will usually have complex cardiovascular anomalies not amenable to surgical correction, but we found a normal heart in the right sided twin and a rudimentary heart in the left. The possibility of cardiovascular anomalies should be taken into account if the surgical separation of this variant of conjoined twins is planned.^[8]

Separation of conjoined twins is a complicated procedure. The importance of a multidisciplinary team with rehearsal of all aspects (surgical, anesthetic and nursing) of the operative procedure cannot be overemphasized.^[8] Although the outcome is influenced by careful planning and organization from all participants, the prognosis is often predetermined by the underlying anatomy which may preclude successful separation.^[9-11] Besides there may not be sufficient time for adequate planning if an emergency separation becomes expedient, as was in the present case. Although conjoined twins are rare, occurring in approximately 1 per 50,000 to 1 per 100,000; it should be suspected in all monochorionic, monoamniotic twin pregnancies, and careful Sonographic assessment should be performed to identify the presence of shared fatal organs.^[12]

Conclusion

Siamese twins or double monsters have always been a subject of curiosity and mystery for the general public. This

case emphasizes the need for antenatal care with prenatal ultrasound monitoring of high-risk pregnancies in order to determine the nature of the perinatal management required. When serious malformations that are incompatible with postnatal life are diagnosed early enough, the family has the option of terminating the pregnancy. Therefore, there is a need to improve our health care delivery system to make such services available and accessible to all our pregnant women. Similarly, it is important to educate the women and their spouses on the need for proper ANC.

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