Dicephalus Dibrachius Dipus Conjoined Twins in a Triplet Pregnancy

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Abstract
Conjoined twins occurring in a triplet pregnancy is a rare occurrence. We present a case of undiagnosed diencephalic conjoined twins occurring in a multigravida with triplet pregnancy delivered by caesarian section. The anatomical and pathologic findings in these twins after their demise are described with a brief review of the literature.


Introduction
Conjoined twinning is a rare phenomenon, occurring in 1 in 30000-100000 live births. A rarer form of conjoined twining is the diencephalic dipus dibrachius twins in which the baby has two heads on one body with 2 upper limbs and two lower limbs. The incidence of conjoined twins in our environment is unknown but there have been previous reports from Nigeria.

We recently encountered a case of undiagnosed diencephalic conjoined twin in an unbooked patient with triplet pregnancy that died within 36 hours of birth from cardiopulmonary failure. The anatomical, pathological and prognostic feature of the twins is presented with a review of the literature.

Case Report
An unbooked 35 years old Gravida5Para4+0 all alive was admitted in early labor with multiple pregnancies at gestational age of 37 completed weeks. The fundal height was 47 cm. Two distinct heads were palpated in the lower uterine segment while another was felt in the fundus. The fetal heart sounds picked with the aid of sonicaid were heard in two distinct places and were normal. The uterine contractions were weak and she was in respiratory distress. All other clinical signs were within normal limit. A diagnosis of triplet pregnancy with respiratory embarrassment was made and a decision was made to deliver the babies through caesarian section.

At surgery a set of conjoined twin was delivered in the first gestational sac. Both had two heads and one body and two upper limbs and lower limbs (diencephalic dipus dibrachius twins) and weighed 3.0kg with poor Apgar score of 3, 5, and 8 at 1, 5 and 10 minutes respectively. The movements of the limbs on the two sides appeared to be independent of each other. From the second sac was delivered a healthy male baby weighing 2.65kg.

The parents of the babies raised objection to either the treatment or the survival of the conjoined infants but the babies were transferred to the neonatal intensive care unit for full evaluation by the pediatricians, pediatric surgeons and the neurosurgical team. The babies however died 36 hours post delivery. Post mortem plain X rays and computerized tomogram was done which showed the fusion of the vertebral column and the spinal cord from the level of Thoracic vertebra (Figures 1 and 2). A limited autopsy was also performed the findings of which is summarized below: Each foetus had a separate head and neck, which joined, a single chest.

Thoracic Organs: A single trachea in each fetus entering in the thoracic cavity from each twin with normal bifurcation of the trachea into right and left main bronchus. The right and left lungs made up of 4 lobes each and tightly packed into the single thoracic cavity. The thymus glands were duplicated. The heart is single and is univentricular.

Abdominal Organs: There were 2 separate oesophagus entering through separate hiatus in the diaphragm into2 separate stomach. There was shared intestine from the second part of the duodenum to the anus. The liver and the pancreas were shared while the single biliary ductal system joining the fused second part of the duodenum. There was only one spleen located in the left hypochondrium.

Genito-Urinary System: There were two kidneys drained by two ureters into a single urinary bladder. There was a single urethra, a single vagina, uterus and right and left ovaries.

Musculoskeletal system: Y-shaped vertebra column with fusion at the level of the thoracic vertebra (T9).

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Discussion
Conjoined twins complicating triplet pregnancy have been reported but are very rare. It is also rare for diencephalic conjoined twin to occur as part of triplet pregnancy and to our knowledge there has been only one previous report of occurrence of Diencephalus dibrachius dipus twins in triplet pregnancy. Two main models for the embryogenic origin of conjoined twins have been proposed. They are either due to incomplete division of the monozygotic embryo in the early period of embryogenesis (fission theory) or due to secondary union of two originally separate monovular embryonic disc (fusion theory). Microsurgical dissection of a specimen of diencephalic dibrachiatus twin by Crauiciuc et al and their observation makes them to support the fusion theory in these fetuses.

Figure 1: Diencephalus Twins Immediately After Birth

Figure 2: Plain X-Ray Showing Fusion of the Spine of the Diencephalus Twins

Most cases of Diencephalus dibrachius dipus conjoined twins are either still born or died shortly after birth because of severe cardiopulmonary malformations as in our patients and therefore termination of pregnancy may be an option. However cases of diencephalic twins living to maturity have been described in the literature. Groner et al did a multiple imaging studies of a Diencephalus twin and concluded that separation should not be attempted in this instance because of the complexity of the shared organs in these twins. We agreed with this because the morbid study of our twins showed that separation is impossible. However if the babies were to have survived there would have been dilemma regarding the care of the infants as the parents of the twins were against the treatment or survival of the infants because of cultural taboo. We would therefore advise that termination of pregnancy should be done in our environment if the diencephalic twin is detected early in utero.

With the advent of high-resolution ultrasonography conjoined twins can be picked up as early as the 8th week of gestation and with fetal echocardiography as well as ultra fast magnetic resonance imaging (MRI), evaluated for possibility of postnatal survival. Where the fetus is unlikely to be viable because of severe associated anomalies selective termination of the abnormal twins can be offered when there is a triplet pregnancy. However all these facilities are not available in our institutions and moreover many of the patients do not register for antenatal care as the mother of these twins, therefore prenatal diagnosis is unlikely in our environment. Undiagnosed conjoined twins may cause dystocia in labor leading to emergency operative delivery thereby jeopardizing the survival of the twins and the normal singleton. There is therefore need to improve the health care delivery system to make it available and accessible to all our pregnant women. Also it is important to educate the women on the need for antenatal monitoring for safe delivery and early detection of abnormalities in the fetus so that proper measures could be taken to ensure satisfactory outcome of the pregnancy.

References