Dicephalic Parapagus Tribrachius Conjoined Twins in a Triplet Pregnancy: A Case Report

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Abstract

Dicephalic parapagus tribrachius conjoined twin is a very rare condition.

We present a case of 5-days-old male dicephalic parapagus conjoined twins. The conjoined twins were diagnosed in 4th month of pregnancy by ultrasonography. The pregnancy terminates at 36 weeks by cesarean section and triplets were born. The babies were male conjoined twins and another healthy male baby. Many congenital defects of interest can now be detected before birth. Sever form of this malformation precludes postpartum life. It is supposed that with advances in screening methods for prenatal diagnosis these cases are terminated in first or second trimester of pregnancy.

Keywords: Dicephalic Parapagus Tribrachius, Conjoined Twins, Triplet Pregnancy

Introduction

Conjoined twin is a rare phenomenon, occurring 1 in 50,000 to 100,000 (1). However since 60% are stillborn or die shortly after, the true incidence is around 1in 200,000 live births. A rarer form of conjoined twinning is the dicephalic parapagus twins which fused side-by-side with a shared pelvis (2). They are dicephalic and have two (dibrachius) three (tribrachius) or four (tetrabrachius) arms.

Case Report

A 6-hour -old male conjoined twins with three arms and two heads were brought in our hospital. They were pregnancy products of a 27 years old mother in her second delivery.

Her pregnancy was triplets. Ultrasonography in 15^{th} weeks of pregnancy showed a male conjoined

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Dr. Mahbod Kaveh, Department of Neonatalogy, Tehran University of Medical Science, Bahrami Hospital, Tehran, Iran Email: mahbodkaveh@yahoo.com twins and a healthy male. The pregnancy terminates at 36 weeks by caesarian section. Mother did not have any problem in her first pregnancy and her first child is a 4 years old healthy girl.

There is no history of abortion, infertility, medical problem or drug history in mother. All of pregnancy products were alive at the time of birth but twins died after 5 days .The twins were male and white. The pattern of our conjoined twins was dicephalic parapagus tribrachius and is referred as a side-by-side fusion with a shared pelvis and three arms. There was a history of conjoined twins in their grandmother.

The conjoined twins' birth weight was 3080 gr. In physical exam they had two heads which left's head circumference was 32 cm and right's head was 32/5cm. There was one trunk and two normal upper extremities and a common and abnormal arm between their heads.

Heart & lungs looked normal in physical exam. There was no abnormality in chest wall except wide chest wall dimension. They had a single abdomen with no obvious organomegaly or abnormality of abdominal wall. The conjoined twins had two normal lower extremities.

Their genitalia were normal and male. In the neurologic examination the pupils were symmetric and reactive to light. The left head had responded to the left side stimulation and the right head had responded to the right side stimuli.

In echocardiography only one normal heart is seen in the left side. Right heart seems to be only an Atrium draining into the left heart. No pathology was seen in left heart. The twins had one Aortic Arch.

Brain sonography of both heads was normal and did no sign of intraventricular hemorrhage, germinal matrix hemorrhage, periventricular leukomalacia or gross anomaly of brain was seen. In abdominal sonography of conjoined twins they had single liver in midline with two gallbladders, one common bile duct and portal vein. Also they had only one spleen in the left side. The babies had three kidneys which two of them were in the left side and one of them was in the right side. They had single urinary bladder in midline. Also they had only one inferior vena cava and Abdominal aorta in midline. Twins had two stomachs and in chest X-ray was seen two vertebral column .with a shared pelvis. Also left lung of the right one (neonate) and right lung of the left one (neonate) were hypoplastic and common at the midline (Figures 1, 2).



Figure 1: Ultrasonographic appearance of triplet pregnancy with conjoined twins

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Dicephalic Parapagus Tribrachius Conjoined Twins



Figure 2: Conjoined twins

Discussion

Conjoined twins, a rare entity in obstetrical practice, is a fascinating congenital abnormality with devastating consequences both for the twins and the family. Various congenital anomaly was reported in conjoined twins such as an encephaly, heart anomaly, Gastrointestinal anomaly, urogenital anomaly cleft lip, cleft palate and lumbosacral meningomyelocele (3).

There are two theories of how conjoined twins are formed. The more widely accepted one is the "fission theory" which states that conjoined twins occur when a fertilized ovum begins to split into identical twins, but is somehow interrupted during the process and develops into two partially formed individuals who are stuck together (4). It is argued that conjoined twinning cannot possibly result from a "fission event", and can result from the fusion of mono amniotic twins (5). He proposed that two monoovular embryonic discs may lie adjacent to one another at various angles, and may be become secondarily united dorsally, caudally, laterally or dorsally and symmetrically or asymmetrically but always homologous (6). It might seem logical to assume that dicephalus twins arise from two separate, nearly Parallel notochords on one embryonic disc, very close together caudally, but with varying degrees of separation rostrally (7).

Ultrasound is helpful in diagnosing conjoined twins. In case of monochorionic and monoamiotic twins, careful assessment should be done to rule out shared parts between the fetuses.

Conclusion

Conjoined twins should be suspected in all monochorionic, monoamniotic twin pregnancies and careful sonographical assessment should be undertaken to exclude any of classical signs which are suggestive of conjoined twins and to identify the severity of the shared fetal organs for perinatal management. When serious malformations that are incompatible with post natal life are diagnosed in early gestational age, termination of pregnancy should be advised.

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