Volume 15 (2), 2013

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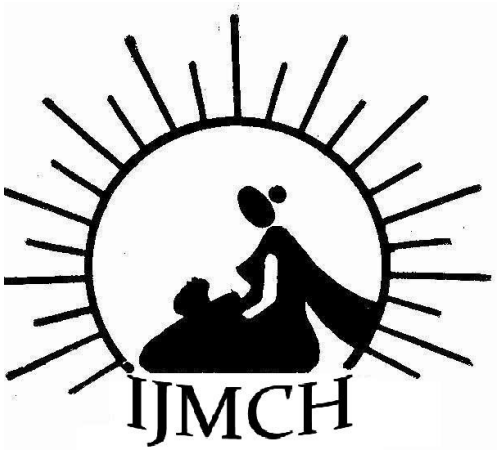

INDIAN JOURNAL OF MATERNAL AND CHILD HEALTH

Conjoined twin is a rarely seen congenital anomaly together with severe mortality and morbidity. We are reporting one case of an eight hour old male conjoined thoraco-omphalopagus twin, with 3.1 kg weight, admitted in a neonatal intensive care unit of a tertiary care centre.

Late Detection of Conjoined Twins: The Lessons to Learn

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ABSTRACT:

Conjoined twin is a rarely seen congenital anomaly together with severe mortality and morbidity. We are reporting one case of an eight hour old male conjoined thoraco-omphalopagus twin, with 3.1 kg weight, admitted in a neonatal intensive care unit of a tertiary care centre. It was diagnosed by ultrasonography in a 26 year old second gravida, who was referred for routine obstetrics sonographic screening in the late second trimester. Making an early diagnosis with ultrasonographic examination gives the parents a chance to elect pregnancy termination with minimal maternal morbidity. It is also important to motivate ASHA workers and all public health staff to educate the women and their spouses on the need for proper antenatal checkups including routine obstetric ultrasonographic examinations.

Key Words: Conjoined twins, Thoraco-omphalopagus, Sonography, Homozygous twins.

INTRODUCTION:

The conjoined twins represent one of the rarest forms of twin gestation, occurring roughly 1 in every 200 identical twin pregnancies [(1)] with incidence ranging from 1 in 50,000 to 1 in 100,000 live births [(1, 2)]. Various degrees of fetal fusion result from incomplete division of the inner cell mass 13-15 days after fertilization. Although the precise causes are unknown, many authors believe that factors responsible for monozygosity may play a role in conjoined twins [(3)].

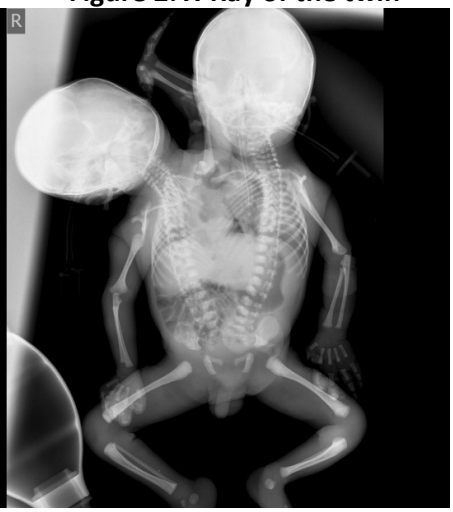
Conjoined twins can cause dystocia with the risk of rupture of the uterus, and quite often require cesarean section which may have negative consequences for the obstetrical future of the mother. The risk that the condition recurs in a subsequent pregnancy may be considered negligible [(3)]. Two contradicting theories (theory of fission and theory of fusion of fertilized egg) exist to explain the origins of conjoined twins [(2)]. Although, prognosis for all types of conjoined twins is extremely poor, a careful anatomical and vascular mapping to determine the extent of organ sharing is of paramount importance to determine the prognosis. Diagnosis prior to 12th week of gestation has been reported by several authors [(5)].

CASE REPORT:

An eight hour old male conjoined twin with 3.1 kg, was admitted in a neonatal intensive care unit of a tertiary care centre in North Karnataka. As shown in figure 1, the newborn conjoined twin had two heads separated upto neck. The thorax and abdomen were fused. It had four well formed limbs- two upper and two lower with a rudimentary upper limb between two heads. As seen in X-ray of the conjoined twins (Fig. 2), the separate skulls with a small rudimentary limb near the skull was noted. Two separate spines were noted with no fusion in entire extent. There was a single pelvic cavity. The upper and lower limbs were well formed showing the bones. The epiphysis of the distal end of femur was not noted, suggesting the foetal age to be less than 36 weeks of gestation. The rib cage was fused in its entire extent with a single cardia (?). The abdominal organs were fused. However, lateral X-ray would have given the additional information about the visceral organs. In spite of resuscitation in neonatal intensive care unit, baby could not be survived and succumbed after 12 hours of life.

Figure 1: Photograph of the twin

Figure 2: X-Ray of the twin



Retrospectively, parents were interrogated for the detailed obstetric and family history. A 26 year old, second gravida was referred for the first time for routine obstetric sonography to a tertiary care centre in her late second trimester. Her exact date of last menstrual period was not known. She had no personal and family history of twins. She had one 3 year old healthy female child and her medical history was not remarkable. She received 2 doses of tetanus toxoid and haematinics by peripheral health workers. Ultrasonography revealed the presence of conjoined twins (thoraco-omphalopagus) of 32 to 34 weeks gestation. The parents were informed about the malformation and the twins' poor chance of survival. The parents decided to terminate the pregnancy. After taking informed written consent from the family and the ethical clearance from institutes Ethics Committee, pregnancy was terminated by caesarean section. However, the twins succumbed at 12 hours of life. The parents did not allow for postmortem examination of the twin and took it away against the institutes advice.

DISCUSSION:

Conjoined twins have been a source of fascination for both general public and medical profession since time immemorial. Their birth was initially viewed as an ominous sign of impending disaster. This was followed, in western by prolonged periods through the Middle Ages and into the nineteenth century when they were regarded as freaks or monstrosities and were exhibited with substantial financial reward at circuses and sideshows [(1)]. The conjoined twins represent one of the rarest forms of twin gestation, occurring roughly 1 in every 200 identical twin pregnancies [(1)] with incidence ranging from 1 in 50,000 to 1 in 100,000 live births [(1, 2)] and the ratio of females to males is 3:1 [(2)]. The prevalence of symmetrical conjoined twins is higher than those asymmetrical twins or incomplete conjoined twins. In a study conducted in Hungary 1970-1986, for example, symmetrical conjoined twins had predominance of 92.3% over the asymmetrical [(6)]. Various degrees of fetal fusion result from incomplete division of the inner cell mass 13-15 days after fertilization. Although the precise causes are unknown, many authors believe that the factors responsible for monozygosity may play a role in conjoined twins [(3)].

The pattern of fusion has been studied by Barth and coworkers. The nomenclature is based upon the joined regions. The anatomic site of fusion is named followed by the suffix “pagus” from the Greek fastened. Typical fusions are thoraco (chest), omphalo (umbilical/abdominal), pygo (sacral), ischio (pelvis), and cranio (head). Very extensive zones of fusion may be named by the prefix “di” (meaning two) followed by the portion of the twins that is unfused like di- cephalus (two heads in one baby), dipygus (single head and torso with separate pelvis-four legs). Asymmetric forms are extremely rare [(7)]. The most common forms of conjoined twins are those found at the trunk level, such as thoracopagus, omphalopagus, and xiphopagus [(7)].

Conjoined twins can cause dystocia with the risk of rupture of the uterus, and quite often require cesarean section which may have negative consequences for the obstetrical future of the mother. However, an early ultrasound diagnostic can modify prognosis and allow medical termination of pregnancy in case of seriously malformed conjoined twins. The risk that the condition recurs in a subsequent pregnancy may be considered negligible [(3)]. Two contradicting theories exist to explain the origins of conjoined twins. The traditional theory is fission, in which the fertilized egg splits partially and conjoined twins represent delayed separation of the embryonic mass between 13^{th} and 15^{th} days of fertilization. The second theory is fusion, in which a fertilized egg completely separates, but stem cells (which search for similar cells) find like-stem cells on the other twin and fuse the twins together [(2)]. Conjoined twins share a single common chorion, placenta, and amniotic sac, although these characteristics are not exclusive to conjoined twins as there are some monozygotic but nonconjoined twins that also share these structures in utero [(2, 4)].

Although, prognosis for all types of conjoined twins is extremely poor, a careful anatomical and vascular mapping to determine the extent of organ sharing is of paramount importance to determine the prognosis. The higher prevalence of embryonic and fetal anomalies in cases of multiple gestation, especially in those that are monochorionic monoamniotic

argues for diagnostic ultrasonographic imaging early in pregnancy. Diagnosis prior to 12th week of gestation has been reported by several authors [5]. First trimester sonographic scanning to rule out conjoined twins is indicated in all pregnant women who are at high risk for multiple gestation by careful observation of yolk sacs, the chorion and the amnion [5]. Once conjoined twins have been diagnosed, characterization of the type and severity of the abnormality can be performed with ultrasound, three-dimensional ultrasound, computed tomography, or magnetic resonance imaging [9].

PUBLIC HEALTH IMPLICATIONS:

This case emphasizes the need for regular antenatal checkups with routine prenatal ultrasound screening for foetal growth and development. Mid trimester routine ultrasound examination can achieve a detection rate of congenital anomalies comparable with tertiary centres. One stage ultrasound examination at mid trimester gives acceptable results concerning congenital anomalies with a few false positive results [10]. As serious malformations including conjoined twins are associated with a high perinatal mortality, its early diagnosis with sonographic examination gives the parents a chance to elect pregnancy termination. Therefore, there is a need to improve our health care delivery system to make such services available and accessible to all our pregnant women in the periphery. Similarly, it is also important to motivate ASHA workers and all public health staff including medical officers of peripheral health institutions to educate the women and their spouses on the need for proper antenatal checkups including routine obstetric ultrasonographic examinations.

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