

Cephalothoracoomphalopagus: A Rare Type of Conjoined Twin

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ABSTRACT

We present a case of female cephalothoracoomphalopagus conjoined twin, which is extremely rare type of conjoined twins. We also review the contemporary knowledge regarding incidence, etiopathogenesis, antenatal diagnosis and outcome or the prognosis of conjoined twins. The case belong to hindu female, no history of consanguineous marriage, ingestion of drugs or exposure to any radiation. History of one abortion and one twin delivery present. Conjoined twin was cephalothoracoomphalopagus type, delivered vaginally at 30 wks of gestation to a 25 yrs old multigravida. Management of conjoined twin still remain challenges because of multiple congenital anomalies and poor outcome is seen.

Key words:

Cephalothoracoomphalopagus, conjoined twin, outcome

INTRODUCTION

Twin who share their vital organ and body parts, are referred as conjoined twin. Types - cephalopagus-fusion from top of the head to umbilicus, thoracopagus - united from upper thorax down to the umbilicus. Omphalopagus-fetuses are joined primarily in the area of umbilicus. Craniopagus-united on any portion of skull except face. Rare types - cephalo thoracopagus, cephalo thoraco omphalopagus,^[1,2] etiopathogenesis.

The precise etiology of conjoined twinning is unknown. The most common explanation is fission of single zygote, or alternatively fusion of two dizygotic or monozygotic embryos in their very early embryonic development between 13 and 15 days after conception. Because conjoined twins develop after differentiation of the chorion and amnion all conjoined twins are monochorionic-monoamniotic.^[3,4]

CASE REPORT

This was a case report of a 25-year-old, multigravida women (gravida 3, para 2, abortion 1) who was admitted in Department of Obstetrics and Gynecology with labor pain at 30 weeks of gestation. History of previous preterm twin delivery present, twins died after delivery. There was no history of exposure to teratogenic agent in pregnancy, she has not gone for any antenatal checkup anywhere, or ultrasonography evaluation before admission to our hospital. Vaginal delivery was performed. Conjoined twins were born with apgar score 1, weighing 1.08 kg. Baby was immediately shifted to Sick Newborn Care Unit of Department of Pediatrics.

On examination, it was seen that the twins were joined from the head down to thorax with one head, one face, one neck,

a single thorax and abdomen, and a single umbilical cord. On the face there were two eyes, one nose and two ears. On the thorax, at the front and back, there existing total four nipples, being two on either side and there were four upper limbs and single spine [Figures 1 and 2]. They were separate from pelvis with four lower limbs and two female genitals. On cardiac examination apex was localized to single left site, on per abdomen examination single liver was palpable they were rare type of conjoined twins-cephalothoraco omphalopagus.

There was no improvement after all intensive care management and the conjoined twin died after 20 min following delivery.

DISCUSSION

The exact frequency of conjoined twins is not established and estimated incidence varies in the literatures. Spontaneous twinning occurs in 1.6% of all

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Figure 1: (Anterior view) Single head, face, neck, thorax and abdomen with single umbilicus and separate upper, lower limbs with separate pelvis and genitals

human pregnancies, of which 1.2% are dizygotic and 4% are monozygotic. In India and Africa 1:14,000 birth and in Europe 1:250,000 live birth has been estimated, suggesting an increase in incidence in black population in addition to this conjoined twins are 3 times more common in female fetuses than male, but its incidence does not vary with maternal age, parity. If we see the incidence of various types-thoraco omphalopagus (28%), thoracopagus (18.5%), omphalopagus (10%), craniopagus (6%). Cephalopagus (5%) while cephalo thoracopagus and cephalo thoraco omphalopagus are extremely rare.^[4,5,6]

Prognosis is very poor among conjoined twins. In a study of 14 cases of prenatally diagnosed conjoined twins, 28% of cases died in utero, 54% died immediately after birth, and only 18% survived out of which 50% died postoperatively.^[4,6,7] In another study the prognosis was unfavorable, with approximately 40% of cases stillborn. The prognosis of cephalo thoracopagus is extremely poor because single brain and heart are present with fused gastrointestinal tracts.^[4,7]



Figure 2: (Posterior view) Two vertebral columns with separate sets of upper and lower limbs

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