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Case Report

Dicephalus dipus: a rarer siamese

Pradipprava Paria*, Debasree Guha, Sibnath Gayen, Probodh Chandra Mondal, Sabyasachi Som

Department of Pediatrics, R G Kar Medical College, West Bengal University of Medical Sciences, Kolkata, India

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***Correspondence:**
Dr. Pradipprava Paria
E-mail: drpradip83@gmail.com

ABSTRACT

Dicephalus twins are very rare form of conjoined parapagus twins where there is a common body with two heads. They are subdivided into many groups depending on the number of upper limbs, lower limbs and number of torso. A dicephalus dibrachus dipus conjoined twin is reported here. Most of the cases are still born or died immediately after birth. Early diagnosis is important for management of conjoined twins.

Keywords: Conjoined twins, Parapagus, Dicephalus

INTRODUCTION

Conjoined twins is a rare phenomenon with a prevalence of 1.47 per 100,000 births.[1] Amongst them, parapagus is the rarest one (<0.5% of all the reported cases of conjoined twins).[2] Parapagus means there is side-to-side fusion of body.[3] Dicephalus is a subset of parapagus, in which the twins share a common body from upper chest downwards.[4] It may be subdivided into many groups depending upon the number of upper limbs or lower limbs present. There is a high incidence of mortality in parapagus twins due to cardiorespiratory malformation. Here, we report a dicephalus conjoined twin with two upper and lower limbs.

CASE REPORT

A term baby of 3.6 kg with congenital anomaly and severe perinatal asphyxia (Apgar score 1 at 1 min) was sent to us from labour room. Baby was delivered by emergency cesarean section for obstructed labour. The 27 years old second gravid mother did not undergo any antenatal ultrasonography study. There was no history of consanguinity, teratogenic drug intake, history of in vitro fertilization or family history of twining. On examination, we found that, the baby had two completely separated head with fully formed facial features, two neck (left head had smaller neck than the right one) but one fused torso and one pair of upper and lower limbs (Figure 1). Head circumference of right one was 31.5 cm and of left one was 32 cm, chest circumference was 35 cm and abdominal girth at the level of umbilicus was 34 cm. Baby was in gasping condition and heart rate was 40/min. Cardiac apex was on the left side of the hemi thorax. Liver was palpable 2 cm below right costal margin. No other palpable mass was there on per abdominal examination. One umbilical cord was there having one umbilical vein and single umbilical artery. Upper limbs did not reveal any abnormality except left shoulder dislocation. But, both the lower limb were arthrogrypotic with pterigyum formation and feet had equinovarus deformity. Sex could not be determined due to ambiguous genitalia. No anal opening was there in perineum. On dorsal examination, we found two spinal column lying side by side and seemed to be fused in lumbar region. Baby died 2-3 min after birth. Imaging studies and postmortem examination could not be done due to religious constrain.

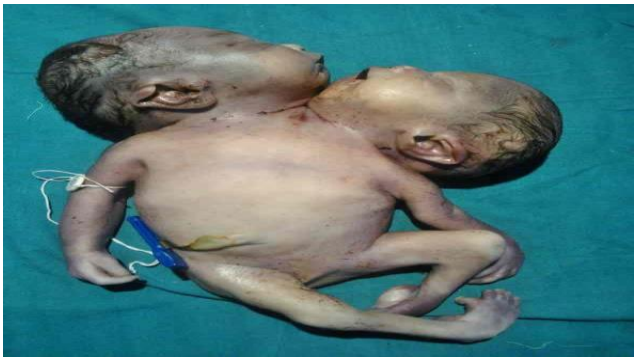

Figure 1: Dicephalus dibracheus dipus: having two completely separated head, one torso, two upper limb and two lower limb. Lower limbs are arthrogrypotic and feet having equinovarous deformity.

DISCUSSION

Conjoined twining is very rare form of congenital malformations where there is fusion of different body parts of genetically identical, monozygotic twins. The condition is more frequently found among females, with a ratio of 3:1.[5] No significant genetic, environmental or demographic factors have been identified and there is no increased chance of recurrences in subsequent pregnancies.[6] Conjoined twins are classified according to the body part at which fusion occurs.[7] Thoracopagus is the commonest variety (40%), followed by omphalopagus (32%), pyopagus (19%), ischiopagus (6%), and craniopagus (2%). Parapagus is the term used where there is extensive side-to-side fusion of body parts. Dicephalus is considered an unusual variant of parapagus where craniofacial duplication occurs. The phenotype ranges from partial duplication (diprosopus) of a few facial structures to complete dicephalus.[8] Regardless of the site of union, variations occur with regard to the internal organs. Some organs may be common to both twins or these may be separate.

Two opposing theories are proposed as embryological basis of conjoint twining- fission and fusion theories. According to fission theory, the inner cell mass divides around day 13 to 15 of fertilization which leads to conjoined twins. Dicephalus twins are as a result of fission at the cranial end alone. Fusion theory tells that, two separate embryos fuse with each other at areas where surface ectoderm is absent. This happens early in the embryonic life within hours to days.[9,10] Conjoined twins share a single common chorion, placenta, and amniotic sac.

Antenatal diagnosis is possible by ultrasound. Signs that may suggest the diagnosis are: bifid appearance of the fetal pole in first trimester, absence of separating membrane, more than three vessels in the umbilical cord, fetal heads are in the same plane, no change in the relative position after maternal movement and inability to separate fetal bodies.[11] Dicephalus variant of conjoined twins are usually stillborn or die shortly after birth. Those who survive will usually have complex cardiovascular anomalies and are not amenable to surgical correction.[12]

CONCLUSION

Early and accurate diagnosis by 3-D ultrasound is thus important so that parents can be counselled for either medical termination or further obstretic management and possible post natal outcome.

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