

Parapagus Dicephalus Tribrachus Tripus Conjoined Twin: A Case Report

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Abstract

Dicephalus twins are very rare form of conjoined twins and are a subtype of parapagus conjoined twin. Dicephalus twins share 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 stillborn dicephalus tribrachus tripus conjoined twin is reported here. There are presence of six fingers and three toes in extra limbs and external genitalia are ambiguous. Presence of third lower limb makes the case even more unusual among the rare type. There is a high incidence of mortality in conjoined twins. Early and accurate diagnosis is important in conjoined twins.

Key words: Conjoined twins, Dicephalus, Parapagus, Tripus

INTRODUCTION

Conjoined twins have fascinated people for centuries.¹ Conjoined twinning is a rare phenomenon and in a recent epidemiological study, it was found that the total prevalence of conjoined twin was 1.47 per 100,000 births.²

Parapagus is the term used where there is extensive side-to-side fusion, and this is a rare form of conjoined twins.³ Dicephalus is a subset of parapagus, in which the twins share a common body from the neck or upper chest downwards.⁴ This anomaly represents <0.5% of all the reported cases of conjoined twins.⁵

They are subdivided into many groups depending upon the number of upper limbs, lower limbs and number of torso.⁶ Accordingly they are dicephalus tetrabrachius dipus, dicephalus tribrachius dipus and dicephalus dibrachius dipus.

There is a high incidence of mortality in conjoined twins. In a study conducted by Mackenzie *et al.*, 28% of conjoined twins died during the intrauterine period, 54% during immediate postnatal period and 18% survived.⁷

Here, a parapagus dicephalus conjoined twin is reported with three upper and lower limbs. Thus, the present case is a parapagus dicephalus tribrachus tripus conjoined twin. In most of the literature bipus dicephalus conjoined twins were reported, but tripus dicephalus conjoined twins are unusual.

CASE REPORT

A stillborn full-term two headed conjoined twin was brought to the Department of Anatomy from the department of obstetrics and gynecology with birth weight of 3.5 kg, and was delivered by caesarean section to a 30-year-old female who did not undergo any antenatal ultrasonography. There was no family history of twinning, but consanguinity was present. History of radiation exposure or intake of drugs was absent. Informed consent was taken from the guardians for examination.

On external examination, twin had two heads and necks with the right head had smaller neck than the left one (Figure 1). The two heads had fused trunk below the neck. Externally, there were normal trunk, abdomen and a single

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Month of Submission : 12-2014
 Month of Peer Review : 01-2015
 Month of Acceptance : 01-2015
 Month of Publishing : 02-2015

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umbilical cord. A single set of ambiguous genitalia and an imperforate anus were seen (Figure 2).

On dorsal examination of the twin, an upper limb was found to attach on the cervical region of the back of the twin (Figure 3). The hand of the limb was abnormally developed with six fingers of abnormal sizes (Figure 4).

The most unusual part of the present case report which makes the case a rare one is that there was a third lower limb attached on the lower lumbar region of the back of the twin (Figure 3). The foot of the limb was elongated with three toes (Figure 5).

DISCUSSION

Conjoined twinning is one of the rare, most interesting and most challenging congenital malformations.⁸ Conjoined

twinning arises when the twinning event occurs at about the primitive streak stage of development, i.e., at about 13-14 days after fertilization and it is always associated with the monoamniotic, monochorionic type of placentation.⁹

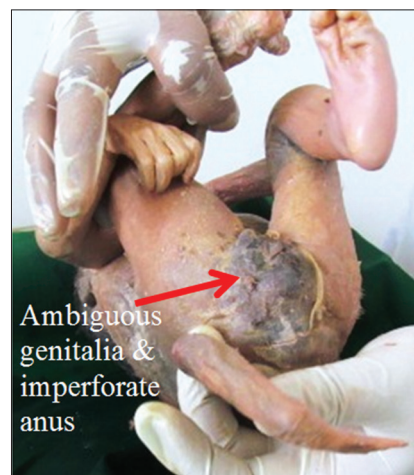


Figure 3: Ambiguous genitalia and imperforate anus in the conjoined twin



Figure 1: Two headed conjoined twin with right head having smaller neck than that of left one



Figure 4: Hand of third extra upper limb of the conjoined twin with abnormally developed six fingers

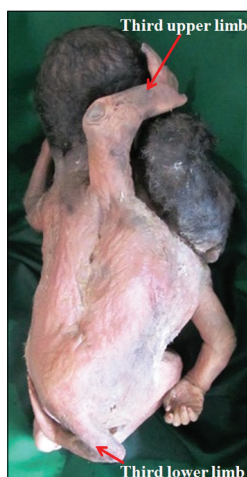


Figure 2: An extra upper limb and a lower limb are attached on cervical and lumbar region of back of the conjoined twin respectively



Figure 5: Elongated foot of the third extra lower limb with three toes

Dicephalus is considered an unusual variant of craniofacial duplication in conjoined twinning. The phenotype comprises a wide spectrum and ranges from partial duplication (diprosopus) of a few facial structures to complete dicephalus.¹⁰

A significant male predominance in parapagus type of conjoined twin has been detected.² In the present case, the external genitalia is ambiguous.

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.¹¹ It is argued that conjoined twinning cannot possibly result from a “fission event,” and can result from the fusion of mono amniotic twins.¹² He proposed that two mono-ovular embryonic discs may lie adjacently to one another at various angles and may become secondarily united dorsally, caudally, laterally or dorsally and symmetrically or asymmetrically, but always homologous.¹³ 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.¹⁴

CONCLUSION

Early and accurate diagnosis is important in conjoined twins so that parents can be counseled for options of

termination of pregnancy. Ultrasound imaging is useful diagnostic tools in early pregnancy. Three-dimensional scan is superior to two-dimensional scan as it detects anomalies more precisely.

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How to cite this article: Sahu SK, Choudhury PR, Saikia M, Das TK, Talukdar KL, Bayan H. Parapagus Dicephalus Tribrachus Tripus Conjoined Twin: A Case Report. *Int J Sci Stud* 2015;2(11):211-213.

Source of Support: Nil, **Conflict of Interest:** None declared.