

## Case Report

# Parapagus dicephalus conjoined twin: a case report

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**Received:** 08 September 2019

**Revised:** 16 October 2019

**Accepted:** 02 November 2019

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## ABSTRACT

A conjoined twin is a rare event. As per recent literature, 60% of conjoined twins are aborted spontaneously and hence true incidence of conjoined twin is approximately 1 in 200,000 live births. Classification is based upon site of attachment out of which parapagus with dicephalic variety is extremely rare (0.5% amongst reported cases). Author report a case of dicephalic twin born at 36 weeks of gestation to a multigravida mother in central India.

**Key words:** Conjoined, Dicephalic, Monoamniotic, Monochorionic, Monozygotic, Parapagus, Twin

## INTRODUCTION

Conjoined twin is born as a random event due to abrupt embryogenesis. True incidence of conjoined twin is approximately 1 in 200,000 live births.<sup>1</sup> There are two well-known theories of development of conjoined twin namely fission theory (incomplete fission of a single embryonic disc 13 to 15 days after the ovum is fertilized) and fusion theory (secondary fusion of two separate monovular embryonic discs). Fusion theory is no longer believed to be the basis of conjoined twinning.<sup>1-4</sup>

The incomplete separation of monozygotic twins occurring after first two weeks of conception results in conjoined fetus that share one placenta, one amniotic sac and one chorionic sac. The extent of separation and the stage at which it occurs determine the type of conjoined twins.<sup>1,3</sup>

Anatomical classification of conjoined twin describes them in terms of their shared anatomy. Incidence of Ventral type is more common (87%) than Dorsal type (13%).<sup>4</sup> In parapagus, the lower part of the body, usually from the lumbo-sacral region caudally, is single, while the region rostral to this is duplicated. The shared anatomy of

internal organs may vary from case to case. It has implications on survival and quality of life for parapagus twin. Cases that have more complicated sharing of internal organs do not even survive until the age where surgical correction is possible. Author report similar case diagnosed in 36 weeks of gestation and succumbed on day 3 of life due to complex anatomy of internal organs.<sup>2,4</sup>

## CASE REPORT

A conjoined twin of male sex was born at 36 weeks of gestation to a multigravida mother in central India. The twin cried immediately at birth. The twin had a single trunk, two arms, two legs and two separate heads joined at the trunk (Figure 1). Infantogram done were suggestive of two separate vertebral columns from cervical to lumbosacral region (Figure 2). Ultrasonography findings revealed two separate thoracic cavities fused in the midline with one pair of lungs and one large sized heart, a Multicystic dysplastic kidney on the left and a normal kidney on the right, a single partially distended gall bladder with normal bowel loops. Neurosonogram of both the heads was suggestive no ultrasonographically identifiable structural abnormality.

The twin had respiratory distress on admission, hence was given on O<sub>2</sub> by prongs. The respiratory distress gradually progressed requiring intubation and mechanical ventilation. The twin subsequently succumbed on day 3 of life. Postmortem examination could not be done due to unwillingness of parents.



**Figure 1: Parapagus and dicephalic twins.**



**Figure 2: Infantogram showing two Separate vertebral columns.**

## DISCUSSION

Due to the appearance and uniqueness of shared anatomy of internal organs, conjoined twin is always a subject matter of attraction to the researchers. A very well-documented early case of parapagus conjoined twins is the Chulchurst Sisters of Biddenden (Bondeson, 1992; Kokcu et al, 2007). They were born in 1100 A.D. and died at age 34 unseparated. The most famous and well-known case of conjoined twins is Chang and Eng Bunker of Siam (Mitchell, 2003). The term “Siamese twins” was used for them and they were one of the longest living conjoined twins documented (63 yrs).<sup>1</sup>

The classification of conjoined twin is based upon the site of attachment either ventral or rostral. The most common varieties encountered were thoraco-omphalopagus (28%), thoracopagus (18.5%), omphalopagus (10%), parasitic twins (10%) and craniopagus (6%) (15). Second classification depends upon the structures, which are unfused. Examples include dicephalus (two heads on one body) and dipygus (single head and torso with separated pelvis and four legs).<sup>2</sup>

As per the literature roughly 40% of conjoined twins are still born and 35% die in the first 24 hours of life. Moreover, only 60% of surgically treated conjoined twins survive. Still birth and mortality rate are extremely high in dicephalus twins. In those Dicephalic twin who are born the duration of survival in cases reported is very short varying from 15 min to 11 days.<sup>2</sup> The reason could be the complexity of sharing of anatomy of internal organs which may be malformed. Ultrasonographic findings revealed one pair of lungs in this case. Most reports have shown two sets of lungs, which may be underdeveloped or anomalous.<sup>2</sup> Kidney on one side was Multicystic dysplastic. Presence of two heads brings in lot of ethical issues in the surgical separation dicephalus twin.<sup>5</sup> Antenatal ultrasound examination is useful for early detection of conjoined twin especially in monochorionic, monoamniotic twin pregnancy. Criteria for ultrasound examination for conjoined twin include the absence of separating amniotic membrane, inseparable fetal bodies, lack of relative change of bodies, and fetal head on repeated examinations.<sup>6</sup>

## CONCLUSION

Conjoined twin is a rare event out of which Parapagus with dicephalus variety is extremely rare (0.5% amongst reported cases). Spontaneous abortion rate in conjoined twins is high (roughly 60 %). Amongst those who are born, survival largely depends upon the shared anatomy of internal structures. Though there are cases with medium to long survival among conjoined twins the quality of life is greatly impacted amongst those who survive. Surgical correction can be offered in very rare cases and almost impossible in dicephalus parapagus twin due to presence of two heads. In the present scenario, early antenatal detection and choice of medical termination of pregnancy can only be offered to the parents. High index of suspicion should be kept from early antenatal period and possibility of conjoined twin should be ruled out when one is dealing with monochorionic, monoamniotic twin pregnancy.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

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**Cite this article as:** Mahajan S, Chauhan U, Gholap S, Yelam B. Parapagus dicephalus conjoined twin: a case report. *Int J Contemp Pediatr* 2020;7:217-9.