

## Diprosopic Parapagus : A Rare Type of Conjoint Twin

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Conjoint twins are rare to begin with and diprosopic parapagus are even rarer. We had an opportunity to deliver one such in our hospital recently. She was a post caesarean pregnancy and presented with a scan report suggesting a dicephalus variety of conjoint twin. After extensive counselling about the poor prognosis she opted for termination of pregnancy by surgical method.

**Keywords:** Diprosopic, Parapagus, Dicephalus, Conjoint Twin

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## Introduction

Conjoined twins are identical twins [1] joined in utero. A rare phenomenon, the occurrence of which is estimated to range from 1 in 50,000 births to 1 in 200,000 births, with a higher incidence in Southwest Asia and Africa. [2] Approximately half is stillborn, and others born alive have abnormalities incompatible with life. The overall survival rate is approximately 25%. [3]

The condition is more common in females, with a ratio of 3:1. [2] Two theories exist to explain the origins of conjoined twins. The old theory is fission, in which the fertilized egg splits partials.

The second and more generally accepted theory is fusion, in which a fertilized egg completely separates, but stem cells (which search for similar cells) finds like-stem cells on the other twin and fuse the twins together. Conjoined twins share a single common chorion, placenta, and amnioticsac, (these characteristics are not exclusive to conjoined twins).[4]

While monozygotic twinning may be induced experimentally.the mechanism of induction of spontaneous twinning in the humans remains unknown.

## Case Study

A 30 yrs. Old p1+0 (post c/s) patient visited the ANOPD of RGKMCH in 27th week of her pregnancy with a USG report showing SLF of 33wk with polyhydramnios? Hydrocephalus.

NO 1st trimester USG was done. She was advised an urgent USG WITH DOPPLER VELOCIMETRY. Her USG DOPPLER VELOCIMETRY report showed live conjoint twin (DUPLICATA INCOMPLETA DICEPHALUS) of approx. 27 wk. 1 day gestational age. The patient was counseled regarding the prognosis and she opted for termination of pregnancy by hysterotomy although VBAC was offered to the patient.

A hysterotomy was performed on the patient and 1.140 kg male DICEPHALIC PARAPAGUS conjoint twins were delivered. The babies were alive for 3hr15mins after birth. The post op period of the mother was uneventful. She was discharged after a period of 7 day.



## Discussion

Conjoint twins are a rare occurrence especially the DIPROSOPIC PARAPAGUS type.

It was first of its type at our hospital.there has been only one such case report where an unnamed boy Dicephalic parapagus twin born in Jaipur , Rajasthan in India in july 2013.the boys head and backbone were separate joined at the pelvis, they shared the ribcage and the shoulder girdle.

They died after 6 days, which is the most common outcome in such cases.(2)

### Types of Conjoint twins

Conjoined twins are typically classified by the point at which their bodies are joined.

The most common types of conjoined twins are:

- **Thoraco-omphalopagus**(28% of cases):(7) Two bodies fused from the upper chest to the lower chest.(8)
- **Thoracopagus**(18.5%):(7) Two bodies fused from the upper thorax to lower belly. The heart is always involved.(8)
- **Omphalopagus**(10%):(7)Two bodies fused at the lower chest ,heart is never involved in these cases;(8)
- **Parasitic twins** (10%):(7)Twins that are asymmetrically conjoined, resulting in one twin that is small, less formed, and dependent on the larger twin for survival.
- **Craniopagus**(6%):(7) Fused skulls, but separate bodies.

Other less-common types of conjoined twins include:

- **Cephalopagus:** Two faces on opposite sides of a single, conjoined head; the upper portion of the body is fused while the bottom portions are separate. These conditions are fatal due to severe brain malformations.. [8]
- **Syncephalus:** One head with a single face but four ears, and two bodies.(8)
- **Cephalothoracopagus:** Bodies fused in the head and thorax. In this type of twins, there are two faces facing in opposite directions, or sometimes a single face and an enlarged skull. (8)(9)
- **Xiphopagus:** Two bodies fused in the xiphoid cartilage, which is approximately from the navel to the lower breastbone. These twins almost never share any vital organs, with the exception of the liver.(8)
- **Ischiopagus:** Fused lower half of the two bodies, with spines conjoined end-to-end at a 180° angle. These twins have four arms; two, three or four of genitalia and anus..(8)
- **Omphalo-Ischiopagus:** Fused in a similar fashion as ischiopagus twins, but facing each other with a joined abdomen akin to omphalopagus. These twins have four arms, and two, three, or four legs.(8)
- **Parapagus:** Fused side-by-side with a shared pelvis. Twins that are **dithoracic parapagus** are fused at the abdomen and pelvis, but not the thorax. Twins that are diprosopic parapagus have one trunk and two faces (this is the case that we are reporting) twins that are Dicephalic Parapagus are Dicephalic, and have two (Dibrachius), three (Tribrachius), or four (Tetrabrachius) arms.(8)
- **Craniopagus parasiticus:** Like craniopagus, but with a second bodiless head attached to the dominant head.
- **Pygopagus (Iliopagus):** Two bodies joined at the pelvis.(8)
- **Rachipagus:** Twins joined along the dorsal aspect (back) of their bodies.(10)

**Diagnosis:** Early prenatal diagnosis of conjoint twins allows better counseling of the parents regarding the management options, including

Continuation of pregnancy with post-natal surgery. With the introduction of high-resolution and transvaginal ultrasound imaging, accurate prenatal diagnosis is possible early in pregnancy. Although first-trimester diagnosis of conjoined twins is feasible, false-positive cases are common before 10 weeks because, earlier in gestation, fetal movements are limited and monoamniotic twins may appear conjoined. Even when the parents opt for conservative management, half of the fetuses die in utero and another 44% will die during the neonatal period. .11 <the 3-d usg offers no special benefits over the 11th to 14th wk usg>

### Criteria for Ultrasonographic Diagnosis of Conjoint Twins Include [15]

Absence of separating amniotic membrane,. Inseparable fetal bodies,. Lack of change in relative positions of bodies and fetal heads on repeated examinations, hyperextension of spine, unusual proximity of limbs ,bifid appearances of first trimester fetal pole, complex anomalies ,more than 3 umbilical vessels, polyhydramnios occurs in 50 to 76 percent of cases.

3D USG, MRI , ECHO before birth and ANGIO ,CARDIAC CATHETERISATION ,RADIONUCLEIDE SCANNING ,CYSTOGRAPHY,URETHROGRAPHY and GI CONTRAST STUDIES after birth may clarify degree of conjoining ,potential of separation and ideal obstetric and perinatal management .[16]

**Deliveri:** There are many reported vaginal deliveries of conjoint twins. Although compressible fetal tissue may facilitate vaginal birth, dystocia uterine rupture or maternal soft tissue injury can occur .When conjoint twins are identified before birth, cesarean section is preferred to avoid maternal trauma and to facilitate treatment of viable fetuses. However VD may be attempted with small premature nonviable fetuses. Craniotomy, evisceration, decapitation or amputation may be needed as a last resort during VD. But since these practices are obsolete in modern day obstetrics LSCS is the preferred method in an antenatally diagnosed conjoint twin.[15]

## Conclusion

Conjoint twins are rare and PARAPAGUS DICEPHALIC twins represent only a small proportion .Complex anomalies of heart and

Abdominal organs occur in most of them .Early diagnosis facilitate termination or optimized obstetrics or perinatal preparation.

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