Conjoined Twin: Review with a Case Report

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Abstract

Conjoined twins are suggested to result from aberrant embryogenesis. The two main theories proposed to explain the phenomena are fission and fusion. The incidence rate is 1 in 50,000 births; however, since about 60% of the cases are stillborn, the true incidence is approximated at 1 in 200,000. There is a higher predisposition towards female than male gender with a ratio of 3:1. Conjoined twins are classified based upon the site of attachment. The extent of organ sharing, especially the heart, determines the possibility and prognosis of a separation procedure. Meticulous preoperative evaluation, planning, and preparedness of the team are crucial for a successful separation. Separation of conjoined twins poses several technical, legal, and ethical issues. With the aid of diagnostic imaging techniques, such as ultrasonography (US) and three-dimensional magnetic resonance imaging (3D-MRI), physicians are able to make prenatal diagnoses**.**

Keywords: *Conjoined twins, cephalopagus, Monozygotic, Prenatal diagnosis, Surgical separation*

Introduction

This is a 22 years PRIMIGRAVIDA with four months of amenorrhea attended Gynaecology OPD of city hospital, Cuttack for her routine pregnancy check up. She was married for last 6 months and was only taking folic acid after her urine examination revealed beta grav index positive. No history of taking any other medicines. Family history was not suggestive of any multiple pregnancy. On General examination all parameters were normal. All systemic examination was also normal. Obstetric examination revealed 16 weeks size Uterus with external ballotment positive .She was advised to do all blood investigations and USG to confirm the period of gestation as well as to rule out any congenital anomaly[1,2].

USG Report Revealed: Conjoined twin with one head (cephalopagus), two bodies joined anteriorly with two pairs of upper limbs and two pairs of lower limbs and single placenta.

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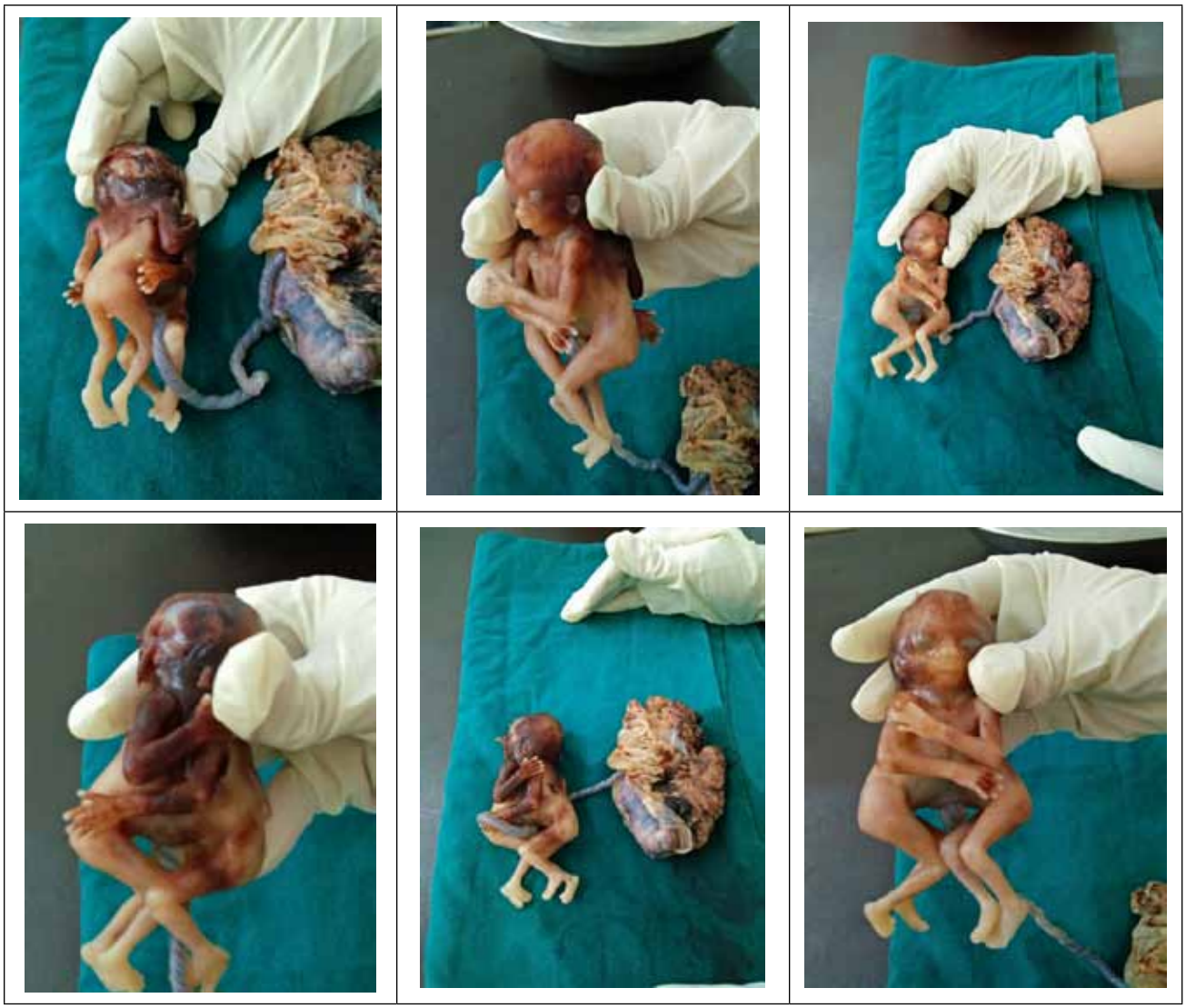
Patient was explained fully about the prognosis. She wanted termination of pregnancy, She was admitted to the City Hospital O&G ward and termination was done with Misoprostol (200mg), 2 tabs 6 hourly[3].

Discussion

Conjoined twins are identical twins joined in utero. An extremely rare phenomenon, the occurrence is estimated to range from 1 in 49,000 births to 1 in 189,000 births, with a somewhat higher incidence in Southeast Asia, Africa and Brazil. Approximately half are stillborn, and an additional one-third die within 24 hours. Most live births are female, with a ratio of 3:1. Most stillborns are male[4,5].

Two contradicting theories exist to explain the origins of conjoined twins. The more generally accepted theory is FISSION, in which the fertilized egg splits partially. The other theory, no longer believed to be the basis of conjoined twinning, is FUSION, in which a fertilized egg completely separates, but stem cells (which search for similar cells) find like-stem cells on the other twin and fuse the twins together. Conjoined twins share a single common chorion, placenta, and amniotic sac, although these characteristics are not exclusive to conjoined twins as there are some monozygotic but non-conjoined twins that also share these structures in utero[6,7].



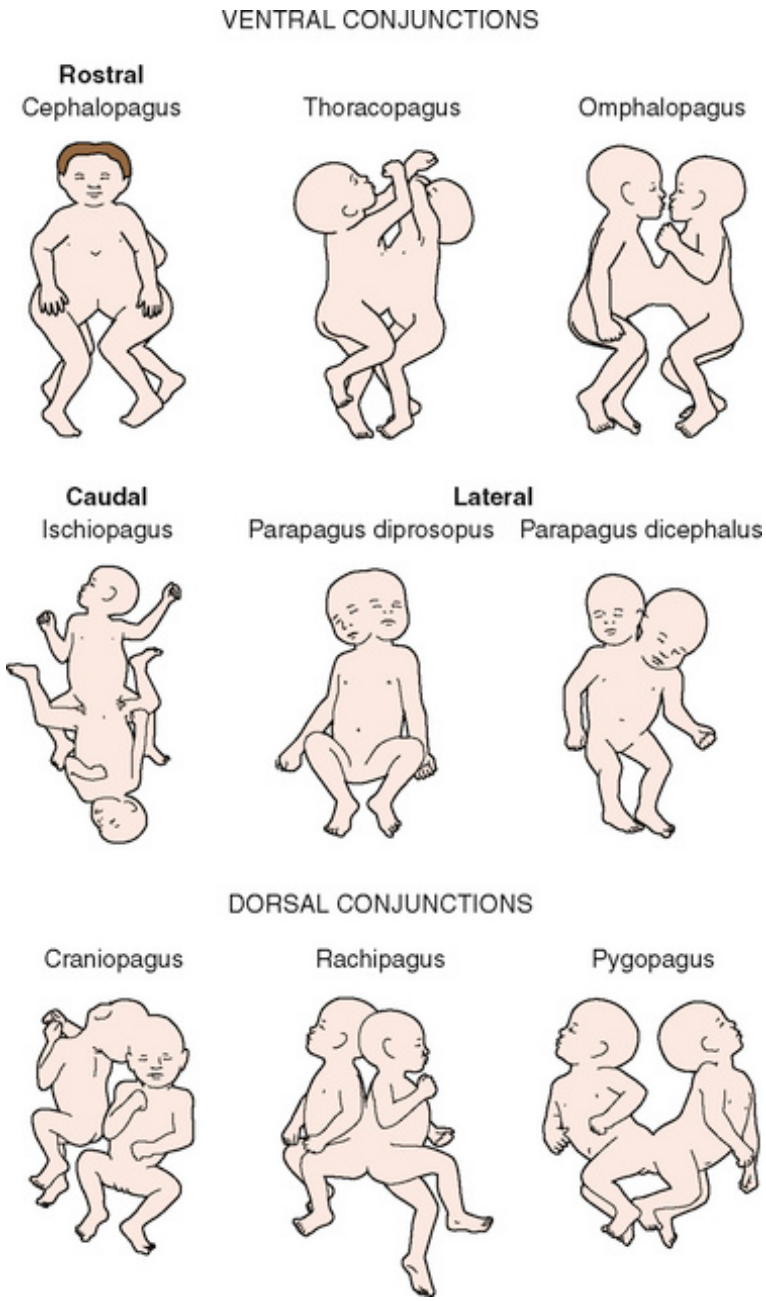
Picture 1: Foetus and the placenta.

Types of conjoined twins[8]: 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): Two bodies fused from the upper chest to the lower chest. These twins usually share a heart, and may also share the liver or part of the digestive system.
- **Thoracopagus** (18.5%): Two bodies fused from the upper thorax to lower belly. The heart is always involved in these cases. As of 2015, separation of a genuinely shared heart has not offered survival to two twins; a designated twin may survive if allotted the heart, sacrificing the other twin.
- **Omphalopagus** (10%): Two bodies fused at the lower abdomen. Unlike thoracopagus, the heart is never involved in these cases; however, the twins often share a liver,digestive system, diaphragm and other organs.
- **Parasitic twins** (10%): Twins that are asymmetrically conjoined, resulting in one twin that is small, less formed, and dependent on the larger twin for survival.
- **Craniopagus** (6%): Fused skulls, but separate bodies. These twins can be conjoined at the back of the head, the front of the head, or the side of the head, but not on the face or the base of the skull.

Other less-common types of conjoined twins include[9]:

- **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 twins generally cannot survive due to severe malformations of the brain. Also known as janiceps (after the two-faced god Janus) or syncephalus.
- **Syncephalus**: One head with a single face but four ears, and two bodies.
- **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
- **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. A famous example is Chang and Eng Bunker.
- **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 legs; and typically one external set of genitalia and anus.



Picture 2: Types of Conjuctions

Picture 3: Siamese Twins

- **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.
- **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. Twins that are **dicephalic parapagus** have one trunk and two heads, and have two (dibrachius), three (tribrachius), or four (tetrabrachius) arms.
- **Craniopagus parasiticus**: Like craniopagus, but with a second bodiless head attached to the dominant head.
- **Pygopagus (Iliopagus)**: Two bodies joined at the pelvis.
- **Rachipagus**: Twins joined along the dorsal aspect (back) of their bodies, with fusion of the vertebral arches and the soft tissue from the head to the buttocks

The most famous pair of conjoined twins was Chang and Eng Bunker (1811–1874), Thai brothers born in Siam, now Thailand. They travelled with P.T. Barnum's circus for many years and were labeled as the **Siamese Twins**. Chang and Eng were joined by a band of flesh, cartilage, and their fused livers at the torso. In modern times, they could have been easily separated. Due to the brothers' fame and the rarity of the condition, the term "Siamese twins" came to be used as a synonym for conjoined twins[10].

Conclusion

Conjoined twins arise from abnormal embryogenesis. There have been many well-observed cases of conjoined twins throughout history, as well as attempts of separation. While separation is normally opted for, there have also been many examples of unseparated conjoined twins leading fulfilling lives. Conjoined twins are classified based on the attachment site. This, as well as the extent of attachment, determines their prognosis. Due to the near infinite variations in conjoined twins, the outcome is highly case-specific. Prenatal diagnosis is possible using ultrasound, MRI, and CT, and can be performed as early as 12 weeks of gestation. Due to the presence of several congenital anomalies associated with conjoined twins, preoperative, intraoperative, and postoperative management is essential to improve prognosis. Cardiovascular and respiratory failures are the two major risk factors for death of the twins postseparation. There are many ethical challenges associated with treating conjoined twins, such as difficult decisions regarding early termination and determining the individuality of each twin.

Ethical Permission: Obtained

Conflict of Interests: None

Funding: None

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