

Case Report

Conjoined twins – Thoraco-omphalopagus: A case report



Y.J. Bhosale*, L. Rajgopal, M.L. Kothari

Department of Anatomy, Seth G S Medical College and KEM Hospital, Mumbai 400012, India

ARTICLE INFO

Article history:

Received 27 April 2016

Accepted 25 July 2016

Available online 4 August 2016

Keywords:

Conjoined twins

Thoraco-omphalopagus

Spencer's spherical theory

ABSTRACT

Conjoined twins are a rare congenital anomaly of twinning which borders on monstrosity. Of the various types of conjoined twins, thoraco-omphalopagus is a commonly occurring type. A recent theory postulates fusion of two embryonic discs as more of a cause for conjoined twinning than fission of a single embryonic plate. In this paper, the findings of a pair thoraco-omphalopagus twins are outlined with a review of the embryological theories that surround their formation.

© 2016 Anatomical Society of India. Published by Elsevier, a division of RELX India, Pvt. Ltd. All rights reserved.

1. Introduction

A statue excavated from a neolithic shrine near Anatolia shows the earliest depiction of conjoined twins – a pair of female ischiopagus twins. The story of conjoined Molionides brothers is the earliest written document about conjoined twins that appears in Greek mythology in 700 BC.¹ 16th century work of Ambrose Pare, “Of Monsters and Prodigies” mentions conjoined twins as one of the 11 generally accepted causes of monstrosities. Conjoined twins were inappropriately called as “Siamese twins” by P.T. Barnum who used this term to denote Chang and Eng Bunker whose mother was half Siamese and father Chinese.² Here we present a case of thoraco-omphalopagus twins and discuss the various theories pertaining to the embryological basis of conjoined twins.

2. Case history

A female patient of non-consanguineous marriage with obstetric history of G1, P0, came for routine USG check up in her 16th week of pregnancy. On USG the presence of monochorionic, monoamniotic conjoined twins was diagnosed. There was no history of drug intake or exposure to radiation. There was no family history of twinning. The parents opted for medical termination of pregnancy.

2.1. External features

The twins were joined in the region of thorax and abdomen (Fig. 1). The palpebral fissures were closed. Both showed

imperforate anus and female external genitalia. Meconium was seen coming out of the external genitalia of one of the twins (Fig. 2). The faces and the limbs were well-developed.

2.2. Internal features

There was a single heart shared by both the twins (Fig. 1). The liver, biliary system and the small intestines of the twins were fused (Fig. 3). The large intestine was ending abruptly at the level of rectum showing rectal atresia. There was presence of recto-vaginal fistula in one of the twins which explains the presence of meconium on the external genitalia. Urachus and umbilical vessels were seen in

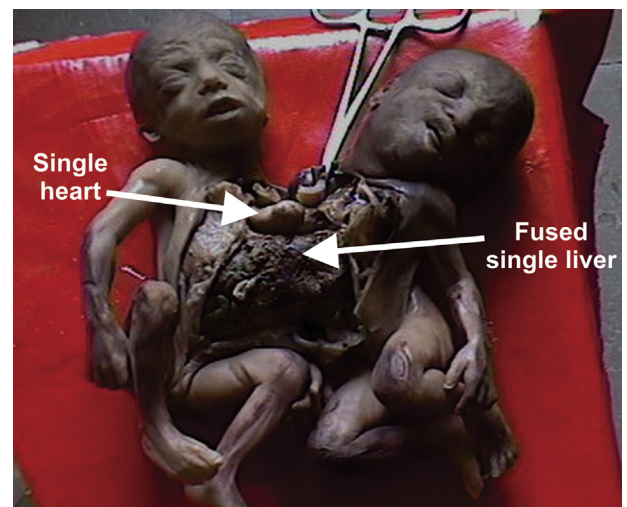


Fig. 1. Conjoined twins showing thoraco-omphalopagus.

* Corresponding author at: Department of Anatomy, Seth G S Medical College and KEM Hospital, Parel, Mumbai 400012, India.

E-mail address: dryuvaraj@gmail.com (Y.J. Bhosale).

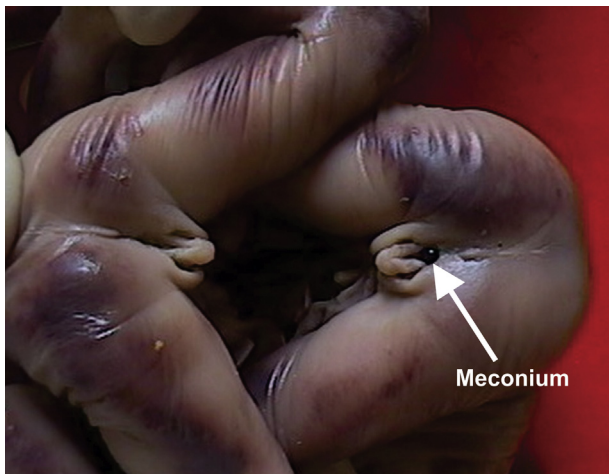


Fig. 2. Female external genitalia of the twins.

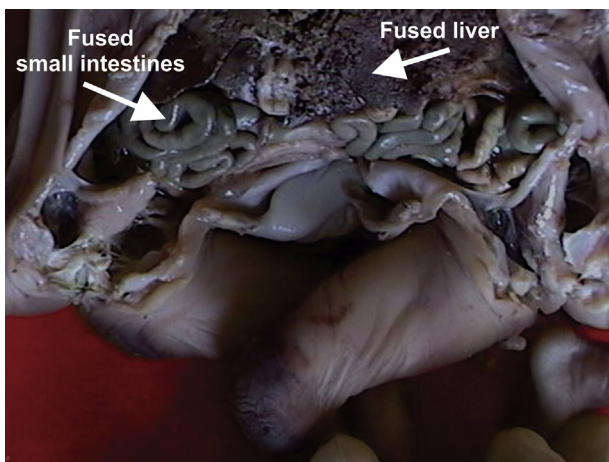


Fig. 3. Fused abdomen of the conjoined twins.

both. The kidneys were lobulated. Suprenals were well-developed. The internal genitalia were well-developed. There was presence of well-developed uterus and fallopian tubes (Fig. 4) in both.

3. Discussion

The incidence of conjoined twins is supposed to range from 1 in 50,000 to 1 in 89,000 in some communities. 40–60% are still born

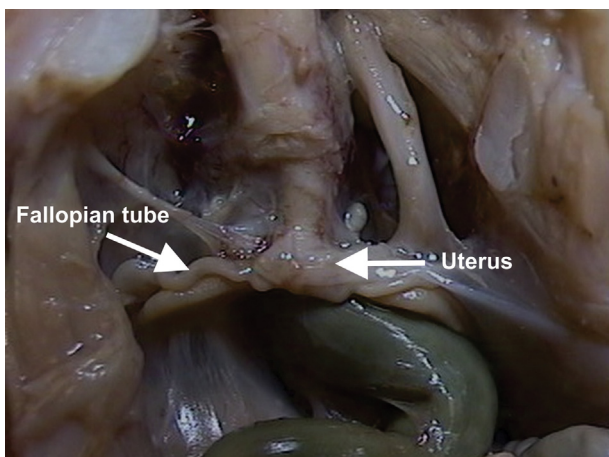


Fig. 4. Internal genitalia of the twins.

Table 1
Terminology of conjoined twins.

Term used	Fused parts
Craniopagus	Cranial vault (Helmet) region
Cephalopagus	Heads
Thoracopagus	Chest
Omphalopagus or Xiphopagus	Ventral part of abdomen
Thoraco-omphalopagus	Chest and ventral part of abdomen
Pygopagus	Sacral and coccygeal region
Ischiopagus	Hip region
Rachipagus	Back (spine)
Parapagus	Side to side fusion

and 75% of those born alive are females. Non-Caucasians have a higher incidence than Caucasians. Increased maternal age is not linked to its incidence.²

Two main categories of conjoined twins are described depending on the symmetry: Symmetrical (Equal) and Asymmetrical. In the former two well-developed babies are fused at some point of the body whereas in the latter an incompletely developed twin or a duplicated body part like limbs or lower half of the body or only the head and neck, called parasite, is attached to a fully developed baby.³

4. Terminology

Symmetrical conjoined twins are named according to the sites of body that are fused: e.g. thoracopagus – (thoraco-chest region, pagus-fixed or attached).

At least nine different types are described as given in Table 1. Of these thoraco-omphalopagus type is the commonest one (74%).

5. Embryological basis

There are two theories which explain the phenomenon of conjoined twinning. In both the theories, the timing of the anomaly is said to be after 13th–14th day of fertilization. In the “Fission Theory”, it is said that the bilaminar embryonic disk after the 14th day splits incompletely and then each portion of the disk develops into an individual. The site where the split has not occurred remains fused.⁴ In the “Fusion theory”, it is said two monovular embryonic discs develop and fuse at similar sites giving rise to conjoined twins. In a refinement of “Fusion Theory”, Spencer⁵ postulated that embryonic discs float either in the outer surface of one sphere, i.e. the yolk sac or on the inner surface of another sphere, i.e. the amniotic cavity. The embryonic discs which float on the surface of the yolk sac, if join rostrally will result in cephalopagus or thoracopagus or omphalopagus; if they join caudally, will result in ischiopagus; if they fuse laterally, it will result in parapagus. The embryonic discs which float on the inner aspect of the amniotic cavity if join rostrally, will result in craniopagus; if fuse back to back, will result in rachipagus; if join at the lower back, will result in pygopagus.⁶

In the present case, there was fusion of the ventral aspect of the chest and abdominal walls and hence the term thoraco-omphalopagus. Since the twins were sharing a common heart, separation could not be possibly attempted. Hence the parents opted for medical termination of pregnancy.

6. Conclusion

Conjoined twinning is a fascinating anomaly whose embryological basis is quite intriguing. Whether it is formed by fission or

by fusion, surgical separation to lead to two fully functional individuals will remain a challenge.

Conflicts of interest

The authors have none to declare.

Acknowledgement

The authors would like to acknowledge the help and support rendered by Dr. P.S. Bhuiyan, Professor & Head, Department of Anatomy and Dr. A.N. Supe, Dean, Seth GS Medical College & KEM Hospital, Mumbai.

References

1. Tithonus JP. Conjoined twins. Available from: <http://thehumanmarvels.com/?p=10> Accessed 13.04.16.
2. Votteler TP, Conjoined twins. 4th ed. Welch K, Randolph JG, Ravitch MM, O'Neil Jr JA, Rane RI, eds. *Pediatric Surgery*. vol. 2. Chicago: Yearbook Manual Publishers Inc.; 1986: 771–779.
3. Kamal K. Conjoined twins. Available from: <http://emedicine.medscape.com/article/934680-overview> Accessed 13.04.16.
4. Moore KL, Persaud TVN. Placenta and foetal membranes. In: Moore KL, Persaud TVN, eds. In: *The Developing Human – Clinically Oriented Embryology* 7th ed. New Delhi: Saunders (Elsevier); 2003:148–149.
5. Spencer R. Theoretical and analytical embryology of twins: Part I – embryogenesis. *Clin Anat*. 2000;13:36–53.
6. Waldhausen JHT, Conjoined twins. Oldham KT, Colombani PM, Foglia RP, Skinner MA, eds. *Principles and Practice of Pediatric Surgery*. vol. 2. Philadelphia: Lippincott, Williams and Wilkins – Wolters Kluwer; 2006: 1795–1803.