

# A Rare Case of Cephalothoracophagus - Conjoined Twins Fused at Head and Thorax Terminated at 13 Weeks of Gestation

Akshay Kumar K K

## 1. Introduction

Conjoined twins represent one of the rarest forms of twin gestation. They occur in roughly 1 in every 200 identical twin pregnancies and are always identical. The incidence ranges from 1 in 50 000 to 1 in 100 000 live births [1]. Because this situation carries high risk, early diagnosis and management of delivery is extremely important.

## 2. Case report

25yr old primi married since 1 year, presented to our Family planning OPD at 13w 6d for MTP. Her antenatal history was

uneventful. Her marriage was non consanguineous and her's was a spontaneous conception. And she had no significant past medical, surgical or family history. Her NT scan at 13 w showed conjoined twins with fused head and thorax, single heart but separate pelvis and limbs - cephalothoracophagus and was referred to our institute. A repeat 3D scan was also done.

She was induced with Mifegest 200 mg Q8H 3 doses and PGE1 400 ug pv and delivered a conjoined twin of 30g after 4 days of starting induction.



Figure 1, 2, 3 Shows the scans.



Figure 4: Shows the Twins after expulsion

## 3. Discussion

Conjoined twinning is one of the most fascinating human malformations and has also been reported in other animals—mammals, fishes, birds, reptiles, and amphibians. [11, 12, 13, 14] The term Siamese twins, once commonly used for conjoined twins, has fallen out of favor because it seems to convey that these individuals are circus freaks or monsters.

Approximately 75% of conjoined twins are female, and 70% are fused at the thorax (thoracopagus) or abdomen (omphalopagus). The union can be in the frontal, transverse, or sagittal plane. (2) The two main categories of conjoined twinning are as follows:

Symmetrical or equal conjoined twins (ie, two well - developed babies)

Asymmetrical or unequal conjoined twins (ie, a small part of the body is duplicated, or an incomplete twin is attached to a fully developed twin)

Volume 13 Issue 3, March 2024

Fully Refereed | Open Access | Double Blind Peer Reviewed Journal

[www.ijsr.net](http://www.ijsr.net)

In broad terms, conjoined twins may be regarded as a doubling anomaly. The later the incomplete embryologic separation occurs, the higher the likelihood of a complicated fusion.

Conjoined twins are classified according to the most prominent site of conjunction: thorax (thoracopagus), abdomen (omphalopagus), sacrum (pygopagus), pelvis (ischiopagus), skull (cephalopagus), and back (rachipagus). Depending on the aspect of the embryonic disc, the most common types are thoracopagus (19%) [3] (. Its etiology is unknown, but an incomplete division of the zygote between 13th and 15th days after fertilization probably occurs [4]. The overall survival rate for conjoined twins is approximately 25% [5]. The condition is more frequently found among females, with a ratio of 3 : 1 [4]. Two theories have been proposed to explain this observation: the process of X - inactivation overlaps with the timing of monozygotic twinning and thus may directly contribute to development of monozygotic twins, and the XX karyotype may confer a survival benefit [2].

Two contradicting theories exist to explain the origins of conjoined twins. The traditional theory is fission, in which the fertilized egg splits partially and conjoined twins represent delayed separation of the embryonic mass after day 12 of fertilization. The second theory 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 [4, 6, 7]. 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 nonconjoined twins that also share these structures in utero [4, 6].

Early diagnosis of conjoined twins was previously reported, but not before the 10th week of gestation [8]. Our case was diagnosed at 13 weeks. On careful transvaginal sonography and serial scanning, there appears to be an inability to separate between the anatomical parts of the fetuses. Once conjoined twins have been diagnosed, characterization of the type and severity of the abnormality can be performed with ultrasound, three - dimensional ultrasound, computed tomography, or magnetic resonance imaging [9, 10]. Termination of pregnancy can be offered to the family. In the present study, the diagnosis has been performed in the first trimester, and the family has chosen termination of this pregnancy. Surgery to separate conjoined twins may range from relatively simple to extremely complex, depending on the point of attachment and the internal parts that are shared. Most cases of separation are extremely risky and life - threatening.

Treating conjoined twins can be a daunting challenge for the surgeon.

Successful separation of a set of omphalopagus twins was first reported in 1689 in Basel, Switzerland. Whereas series of experience with conjoined twins have been reported in the literature successful separation of large numbers of conjoined twins has been limited to a few centers in the world, including the following.

The prognosis for Cephalothoracopagus is extremely poor because surgical separation is not an option, in that only a

single brain and a single heart are present and the gastrointestinal (GI) tracts are fused.

#### 4. Conclusion

Conjoined twins are associated with a high perinatal mortality; therefore, making an early diagnosis with ultrasonographic examination of conjoined twins gives the parents a chance to elect pregnancy termination.

#### References

- [1] Rees AEJ, Vujanac GM, Williams WM. Epidemic of conjoined twins in Cardiff. *British Journal of Obstetrics and Gynaecology*.1993; 100 (4): 388–391. [PubMed] [Google Scholar]
- [2] Chitnis S, Derom C, Vlietinck R, Derom R, Monteiro J, Gregersen PK. X chromosome - inactivation patterns confirm the late timing of monoamniotic - MZ twinning. *American Journal of Human Genetics*.1999; 65 (2): 570–571. [PMC free article] [PubMed] [Google Scholar]
- [3] Schnauffer L. Conjoined twins. In: Raffensperger JG, editor. *Swenson's Pediatric Surgery*.4th edition. New York, NY, USA: Appleton Century - Crofts; 1980. pp.910–920. [Google Scholar]
- [4] Abossolo T, Dancoisne P, Tuailon J, Orvain E, Sommer JC, Rivière JP. Early prenatal diagnosis of asymmetric cephalothoracopagus twins. *Journal de gynécologie, obstétrique et biologie de la reproduction*.1994; 23 (1): 79–84. [PubMed] [Google Scholar]
- [5] Stone JL, Goodrich JT. The craniopagus malformation: classification and implications for surgical separation. *Brain*.2006; 129 (5): 1084–1095. [PubMed] [Google Scholar]
- [6] Spencer R. Theoretical and analytical embryology of conjoined twins: part I: embryogenesis. *Clinical Anatomy*.2000; 13 (1): 36–53. [PubMed] [Google Scholar]
- [7] Spencer R. Theoretical and analytical embryology of conjoined twins: part II: adjustments to union. *Clinical Anatomy*.2000; 13 (2): 97–120. [PubMed] [Google Scholar]
- [8] Hubinont C, Kollmann P, Malvaux V, Donnez J, Bernard P. First - trimester diagnosis of conjoined twins. *Fetal Diagnosis and Therapy*.1997; 12 (3): 185–187. [PubMed] [Google Scholar]
- [9] Kuroda K, Kamei Y, Kozuma S, et al. Prenatal evaluation of cephalopagus conjoined twins by means of three - dimensional ultrasound at 13 weeks of pregnancy. *Ultrasound in Obstetrics and Gynecology*.2000; 16 (3): 264–266. [PubMed] [Google Scholar]
- [10] Kingston CA, McHugh K, Kumaradevan J, Kiely EM, Spitz L. Imaging in the preoperative assessment of conjoined twins. *Radiographics*.2001; 21 (5): 1187–1208. [PubMed] [Google Scholar]
- [11] Afzal AR, Montero FJ. *Conjoined Twins*. Treasure Island, FL: StatPearls; 2022. [Full Text].
- [12] Mazzullo G, Macrì F, Rapisarda G, Marino F. Deradelphous cephalothoracopagus in kittens. *Anat*

Histol Embryol.2009 Oct.38 (5): 327 - 9. [QxMD MEDLINE Link].

- [13] Hartwell S. Feline medical curiosities: conjoined kittens. Messybeast. Available at [http://www.messybeast.com/freak - conjoined. htm](http://www.messybeast.com/freak-conjoined.htm).2016; Accessed: August 15, 2022.
- [14] Kompanje EJ, Hermans JJ. Cephalopagus conjoined twins in a leopard cat (*Prionailurus bengalensis*). J Wildl Dis.2008 Jan.44 (1): 177 - 80. [QxMD MEDLINE Link].