# Cephalothoracopagus Conjoint Twin – A Case Report

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**ABSTRACT**

Conjoined twins are a rare anomaly, with a prevalence between 1:50 000 and 1:200 000 births. The mechanism for the development of conjoined twins is not known, it is postulated as an alteration in the normal developmental process of monozygotic twins. Two theories were hypotheses related to the subject. A 23 years old primigravida woman was referred for a level I (NT/NB) scan at 13 weeks of gestation. She had no personal or family history of twins. Sonography was performed using Mindray DC N3 USG machine at a govt hospital Jhansi. The ultrasonography revealed two fetuses with 2 arms and 2 legs corresponding to the gestational age.

**KEYWORDS**
Cephalothoracopagus, Oropharyngeal, Monozygotic twinning, Conjoint twins

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## INTRODUCTION

Conjoined twins are a rare anomaly, with a prevalence between 1:50000 and 1:200000 births. The mechanism for the development of conjoined twins is not known, it is postulated as an alteration in the normal developmental process of monozygotic twins. Two theories were hypotheses related to the subject. The fissure theory postulates that there is a failure of complete separation of the embryonic disc in the 15$^{th}$ to 17$^{th}$ days of gestation. The fusion theory postulate that a second fusion occurs between the two originally separated embryonic discs. As conjoint twins carry high risk, early diagnosis and management of delivery are extremely important. Broadly conjoint twins are classified into eight sub -categories depending on their site of fusion. Cephalopagus conjoined twins represent about 11 % of all conjoined twins. These are ventrally fused twins with fused heads, thorax, and abdomen, each of them having two arms and two legs. Due to the fusion at the level of the thorax, they are also called Cephalothoracopagus. They are fused by the oropharyngeal membrane according to the fusion theory. These twins are considered non - viable due to complex anatomical fusion of CNS and often die in utero.[1-4]

## CASE PRESENTATION

A 23 years old primigravida woman was referred for a level I (NT/NB) scan at 13 weeks of gestation. She had no personal or family history of twins. Sonography was performed using Mindray DC N3 USG machine at a govt hospital Jhansi. The ultrasonography revealed two fetuses with 2 arms and 2 legs corresponding to the gestational age. The twins were fused ventrally at their head, thorax, and abdomen. A single head was noted without any identifiable facial structure. Two separate heart was visualized, and the visualized abdomen show two sets of liver and kidneys. A single umbilical cord is noted. On basis of ultrasound, a diagnosis of conjoint twin cephalothoracopagus subtype was made. Parents were counseled regarding the diagnosis and possible adverse outcomes of pregnancy. The parents decided to terminate the pregnancy. Written consent was obtained from the parents and induction for termination was done. There was complete delivery of conjoined twins without complication. The abortus external feature showed a single face and the other fetus's head was fused posteriorly. Both the fetus was fused at their head, thorax, and upper abdomen level. No dissection of the abortus was done to examine the internal organs (Figures 1 and 2).[5-8]

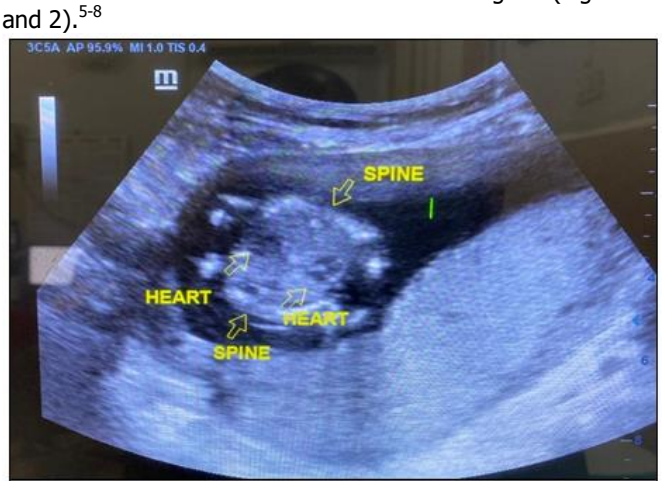


Figure 1. Thoracic Region Fusion with Two Cardiac.

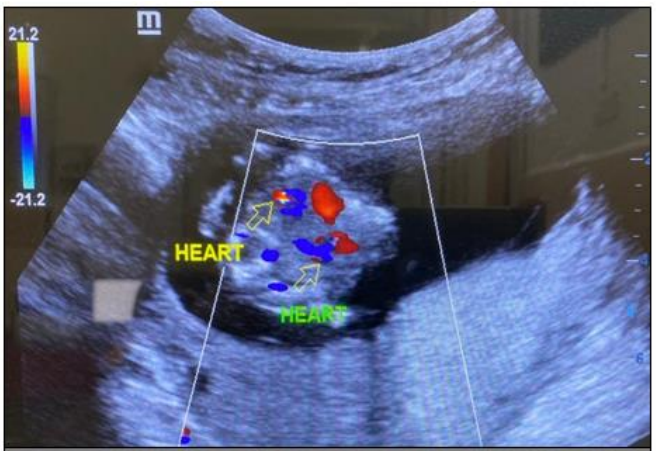


Figure 2. Doppler Scan Showing Two Separates Cardiac with Fusion of Ventral Soft Tissue.

## DISCUSSION

Conjoined twins are a rare variation of monozygotic twinning. Conjoined twins develop from a single fertilized ovum. There are two hypotheses on the development of conjoined twins, the fissure theory and the fusion theory. The fissure theory postulate that there is incomplete separation of the embryonic discs in the 15$^{th}$ to 17$^{th}$ days of gestation. According to the fusion theory, advocated by Spencer and other authors, the development of various types of conjoined twins can be explained through the fusion between two distinct embryonic discs occurring during the early embryonic stage. The accurate mechanism giving rise to conjoined twins is not known. Conjoined twins were classified by various classifications but the most favored ones are the classifications according to the fusion sites. Some classifications include an extension of the junction, like cephalothoracopagus, thoraco - omphalopagus, and others. In this case, we had classified according to the classification proposed by Spencer in 1996, divided into eight major types, grouped according to their ventral or dorsal attachments. In our case, the specimen was cephalopagus or cephalothoracopagus variety according to the classification proposed by Spencer. This variety is extremely rare among conjoined twins with the incidence of one in 58 cases of conjoined twins (Figure 3).

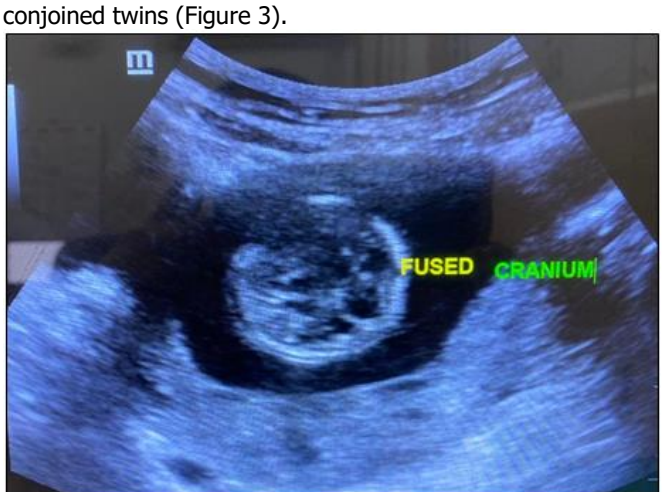


Figure 3. Single Fused Cranium.

As the general incidence of conjoined twins is rare, we can conclude that this particular variety is indeed extremely rare.

This variety has a fused cranium and is often also fused at the level of the thorax and upper abdomen. These twins generally have normally developed four arms, four legs, and a separate lower abdomen and pelvis. A variation of this group is described as Janiceps named after the two-faced roman god Janus because of conjoined twins with two faces present one on each side of the head. In our case, only one face is noted with the fusion of the other fetus's cranium without any discernable second face. The conjoined twins had a set of normal upper and lower limbs. The thoracic viscera appearance of cephalothoracopagus twins was described by Badawy and Shehata. They have described two hearts, one of them well developed and the other one much smaller and rudimentary in form. In our case, two hearts were seen along with a set of lungs for both fetuses. These varieties of conjoined twins are usually considered non - viable due to numerous malformations especially involving the central nervous system. It is to be expected to have multiple developmental anomalies in the brain and spinal cord in these conjoined so postnatal surgical separation will be difficult. In this particular variety of conjoined twins, most do not recommend surgical intervention. In the majority of cases, this type of twin does not survive until birth or dies shortly afterward. Conjoined twins cause mental agony to the parents and there is a need to counsel these parents that these are random events and future pregnancy outcomes are not affected (Figure 4).

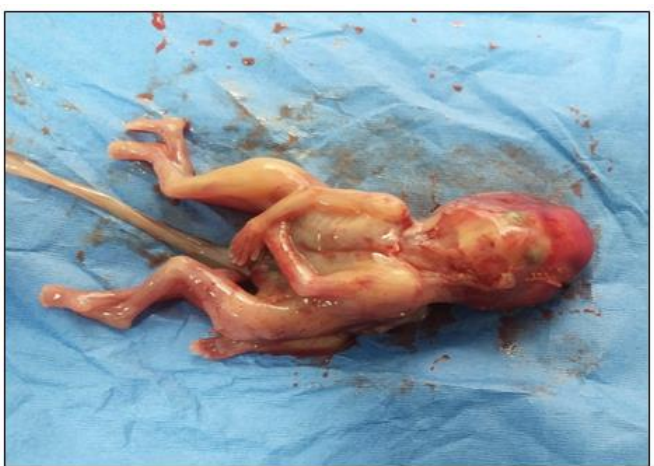

**Figure 4. Post Abortus Showing Cephalothoracophagus Twin with Single Face.**

## CONCLUSION

Cephalopagus or cephalothoracopagus is an extremely rare variety of conjoined twins. They are fused in their ventral aspect at their head, thorax, and upper abdominal cavities. Each twin generally has normal appearing two upper and lower limbs, the heart and lungs are present for each of the twins; usually, one of the hearts is less developed and rudimentary. These twins generally have abnormal development of the superior central nervous system, so surgical intervention is not advised. The prognosis being very poor termination of pregnancy is advised in most of the cases.

## REFERENCES


1. Osmanagaoglu MA, Aran T, Guven S, et al. Thoracopagus Conjoined Twins: A Case Report. ISRN Obstet Gynecol. ISRN Obstet Gynecol 2011;28:1–3.
2. Saravagi G, Chawla S, Bhagat HK, et al. Cephalothoracopagus: A rare conjoined twins, pre and postnatal findings. Med J Armed Forces India 2017;78(1):258-261.
3. Kulhari S, Rastogi A, Kanaujia A, et al. Anesthetic management of premature conjoined twins posted for nonseparation emergency surgery. Paediatr Anaesth 2016;26(12):1204–1205.
4. Boer LL, Franke ANS, Oostra RJ. Two is a Crowd. Clin Anat 2019; 32(5):722–741.
5. Spencer R. Theoretical and analytical embryology of conjoined twins: Part I: Embryogenesis. Clin Anat. 2000;13(1):36–53.
6. Spencer R. Anatomic description of conjoined twins: A plea for standardized terminology. J Pediatr Surg 1996; 31(7):941–944.
7. Singh M, Singh KP, Shaligram P. Conjoined twins' cephalopagus Janiceps monosymmetros: A case report. Birth Defects Res Part A Clin Mol Teratol 2003; 67(4):268–272.
8. Badawy Ah, Shehata R. Cephalothoracopagus: clinical and anatomic study of a case. Obstet Gyneco 1961; 18:106-112.