

Cephalothoracopagus Conjoined Twin - Ultrasound Diagnosis at 26 Weeks. A Rare Case Report

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Abstract

A case of cephalothoracopagus conjoined twin was diagnosed at 26 weeks of gestation in spite of the mother having had five ultrasounds done before. The fetus had single large head, fused thoraces, fused abdomen, common umbilicus but had two pelvises and two sets of genitalia. The fetus had four legs however three arms. Antenatal ultrasound images are complemented by post-natal photographs. The limitation of ultrasound is that a relatively small zone of fusion may not be detected and with an extreme degree of fusion, the twins may be mistaken for a singleton. This case report is an exceptional in daily obstetric ultrasound practice. That's why we reported this case.

Keywords: *Cephalothoracopagus; Conjoined Twin; Ultrasound*

Introduction

Conjoined twins are identical twins joined in uterus [1]. Conjoined twinning is usually thought to occur with late division of the embryonic disc and "conjoined" might actually be a contradiction since it is a failure of fission. Although first antenatal diagnosis of conjoined twins reported in year 1970 by using B-mode ultrasound it is currently possible without difficulty by using 2D and 3D ultrasound facilities [2]. 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) [4] here 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 [5]. Thoracopagus (18.5%) [4] having two bodies fused from the upper thorax to lower belly. The heart is always involved in these cases [5]. 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%) [4], here 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 [5].

Parasitic twins (10%) [4] where the twins that are asymmetrically conjoined, resulting in one twin that is small, less formed, and dependent on the larger twin for survival. Craniopagus (6%) [4] where the 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 [5].

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 twins generally cannot survive due to severe malformations of the brain. Also known as janiceps or syncephalus [5]. In Syncephalus, one head with a single face but four ears, and two bodies [5]. 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 [5,6]. 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 [5]. A famous example is Chang and Eng Bunker. In ischiopagus twin fused lower half of the two bodies, with spines conjoined end-to-end at a 180° angle. These twins have four arms; one, two, three or four legs; and typically one external set of genitalia and anus [5].

Omphalo-Ischiopagus: Fused in a similar fashion to ischiopagus twins but facing each other with a joined abdomen akin to omphalopagus. These twins have four arms, and two, three, or four legs [5]. 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 [5]. Craniopagus parasiticus: Like craniopagus, but with a second bodiless head attached to the dominant head. Pygopagus or Iliopagus: Two bodies joined at the pelvis [5]. Rachipagus: Twins joined along the back of their bodies, with fusion of the vertebral arches and the soft tissue from the head to the buttocks [7].

We reported a case of Cephalothoracopagus conjoined twins which was diagnosed antenatally at 26 weeks and delivered by Cesarean section. This case was initially diagnosed in different centers as a single pregnancy with large head by ultrasound. However final diagnosis was done in Bangladesh Assisted Contraceptive Center Women and Children hospital, Dhaka where dedicated obstetrical ultrasound center are existing. The limitation of conjoined ultrasound is that a relatively small zone of fusion may not be detected and with an extreme degree of fusion, the twins may be mistaken for a singleton. This case report is an exceptional in daily obstetric ultrasound practice. That's why we reported this case.

Antenatal detection of conjoined twins is of supreme importance as it offers wide range of management options. The early diagnosis of conjoined twins allows reasonable obstetric maneuvers and informing parent about options for monitoring, abortion on therapeutic grounds.

Case Report

A 27-year-old healthy woman, gravida 1 was referred from a tertiary level hospital for level II anomaly scan at 26 weeks of her gestational age. 2D and 3D ultrasound was done with a digital scanner (Voluson S 6 Pro V with curvilinear probe). The most significant ultrasound findings included the double heart, having regular cardiac activities (heart rate 144 b/min and 139 b/min) (Figure 1) the two fused heads (Biparietal diameter was 6.2 cm and 4.9 cm respectively and fused head circumference corresponds with 10.3 cm) (Figure 2), single body, spine fused near neck however separated from the lumbar regions, 4 lower limbs and 4 upper arms. Single fetal stomach, two kidneys and single fetal urinary bladder was seen and single placenta present at the anterior and fundal region. Two pelvises were separated with two male external genitalia and four legs; and the cord had three vessels. The amniotic fluid volume was almost nil due to premature rupture of membrane. The patient was informed about the rare form of conjoined twin and was terminated after proper parental counseling. The fetuses were both having male external genitalia and they joined head, neck, chest and abdomen with separate pelvis, upper and lower limbs (cephalothoracopagus) consistent with ultrasound findings (Figure 3 and 4).

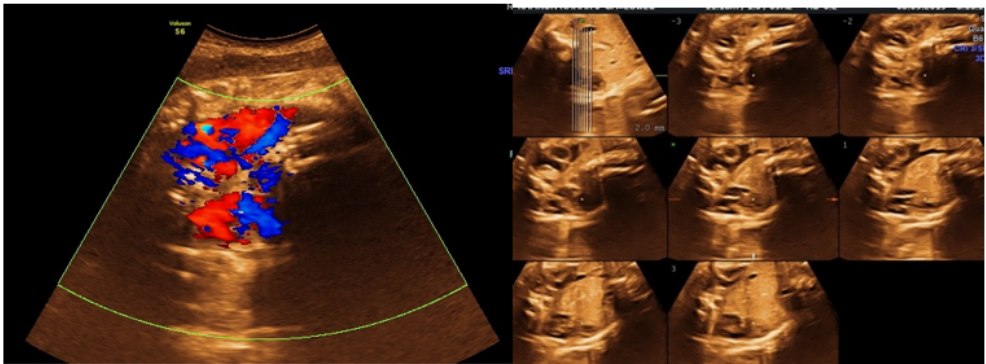


Figure 1: Two heart beats regularly in one thorax.

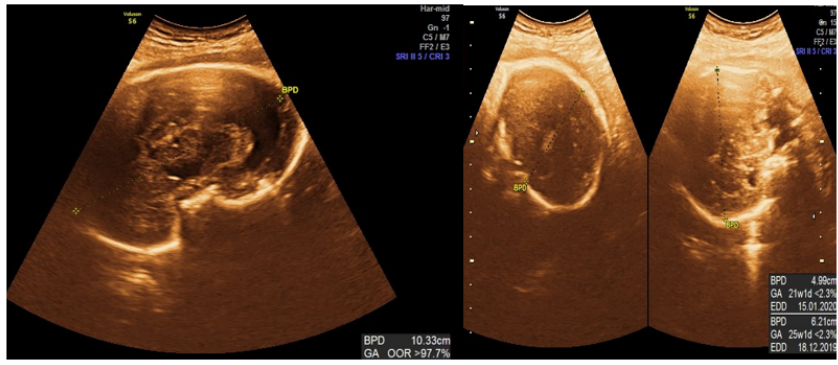


Figure 2: Two head fused, large BPD and after delivery, single face, short neck.



Figure 3: Showing single body with 2 sets of lower limbs and upper limbs after delivery.



Figure 4: One chest, 4 hands, 2 pelvis and four lower limbs after delivery.

Discussion and Conclusion

Conjoined twins are monozygotic twins that result from an incomplete fission of the inner cell mass, probably during the third week after fertilization [8]. The nature of embryo splitting determines the type of conjunction [9]. The earliest prenatal diagnosis reported in the literature was made by vaginal ultrasound examination at 9 weeks 'gestation in two cases of dicephalus with a shared heart [9,10].

Conjoined twins are generally incompatible with life i.e. 65% of cases are stillborn while of those that are born alive, 35% die within the first 24 hours [11]. Sabih, *et al.* [12] reported a case at 29 weeks of pregnancy having similar features of our case, though the patient had 2 - 3 time ultrasound before and not diagnosed as conjoined twin.

The low incidence and poor outcomes of this condition necessitates acceptable observance on part of the radiologist and the obstetrician so as to avoid problems during the later parts of the pregnancy. Surgical separation of nearly complete conjoined twins may be fruitful when organs essential for life are not shared. The prognosis of cephalothoracopagus is extremely poor because single brain and heart are present with fused gastrointestinal tracts [5]. Our case was diagnosed at 26 weeks of gestational age after the mother had undergone five ultrasounds at 22 to 24 weeks and no abnormality was reported except macrocephaly. Then she was referred to us for evaluation of level II anomaly scan. Surgical separation was not possible in our case as the twins shared the brain thorax and also abdomen. This case highlights the features of an extremely rare fetal malformation and concludes with the suggestion that everyone's mind should be kept open to all possibilities when making a diagnosis, as rare causes are rare but do occur in everyday practice of obstetrical ultrasound [12].

The limitation of ultrasound is that a relatively small zone of fusion may not be detected and with an extreme degree of fusion, the twins may be mistaken for a singleton [5].

Early diagnosis and detailed depiction is vital for proper obstetric and postnatal planning. This case highlights the essential role of ultrasound in terms of early detection, appropriate interference as well as recommending of the patients to decrease the psychological disturbance.

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