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Role of high-resolution ultrasound in early diagnosis of conjoined twins

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Abstract

Conjoined twins are rare, with the estimated prevalence is 1:50000 to 1:200000 [1]. Fusion theory and fission theory are two theories behind the origin of conjoined twins [2]. The antenatal diagnosis of conjoined twins can be made with high-resolution ultrasound as early as 12 weeks gestation and is important for optimal obstetric management because early termination of pregnancy has less risk of complications. Fetal echocardiography should be done optimally at 18 weeks because the degree of cardiac fusion will determine the likelihood of successful separation and postnatal viability of the twins. During ultrasound, conjoined hearts are easier to examine in utero because the amniotic fluid acts as a buffer.

Keywords: Twins, Conjoined, Monochorionic, Dichorionic, Ultrasound

Introduction

Conjoined twins are a rare form of monozygotic twins fused in utero. The incidence of conjoint twins falls in the range of 1:50,000 to 1:100,000 live births [1]. It depends on duration of separation of embryonic plates [2] in a monozygotic twin pregnancy as described in the following table 1.

Table 1: Shows type of twins depending on days of separation

Type of Twins	Day Of Separation
Dichorionic Diamniotic	1-3 Day
Monochorionic Diamniotic	4-7 Day
Monochorionic Monoamniotic	8-13 Day
Conjoined Twin	13 Days

The prognosis of conjoined twins is poor and depends upon which vital organ system is shared [3]. The data suggests that 40-60% cases die in utero and almost 35% cases die within a day after birth [4].

Conjoined twins are classified according to their prominent site of connection as described in the following table 2.

Table 2: Classification of conjoined twins according to their prominent site of connection

Parapagus	Twins are fused anterolaterally, depending on the site of fusion they are called bicephalic (two separate heads with fused thorax) or bithoracic (two separate heads and thorax). They usually have a common pelvis and have various GIT and genitourinary abnormalities. They may be bipus, tripus or tetrapus depending on the number of legs attached to the pelvis.
Thoracophagus	Two separate heads, facing towards each other with fused thorax and shared heart. The abdomen and pelvis are separated with all 4 limbs in each of the twins
Omphalophagus	Two separate heads, facing towards each other with fusion from the lower thorax to the umbilical region. The pelvis is separated with all 4 limbs in each of the twins
Cephalophagus	Fused heads with two separate faces in which one face is well developed and the other is rudimentary. Other body parts of the twins may or may not be fused
Craniophagus	Heads are fused with 2 separate well-developed faces. Thorax, abdomen, and pelvis are also separated. Only cranium and meninges are shared with each other, even the brain parenchyma is separated so it has a much better prognosis than cephalopagus twins
Rachiphagus	Twins are fused by their spines thus they are facing opposite to each other. Thoraco-lumbar vertebrae are the most common part to be involved. Twins have separate spinal cords.
Pyophagus	Twins have separate faces, thorax and abdomen facing opposite to each other fused dorsally via sacral vertebrae. They have a common anus although each twin has a separate GIT including the rectum. Twins have separate spinal cords.
Ischiophagus	Twins are fused in their pelvic region with separate heads, thorax, and abdominal regions. The shared pelvis has complicated anatomy with abnormal connections of GIT and the reproductive system of each of the twins.

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In this article, we demonstrate the appearances of conjoined twins with different sites of connection using ultrasound. Material and methods.

An observational institutional study was done over a period of May 2019 to December 2019. All 4 cases were diagnosed antenatally by using an ultrasound machine – ALOKA with a curved array transducer (1–5 MHz). Detailed histories of parents were taken regarding exposure to medications, use of assisted reproductive techniques, and family history of twins.

Examination of each placenta was done for any abnormality and number. Each fetus of the 4 twin cases was individually evaluated from the head to the foot. The site of fusion was noted. The numbers of vital organs were evaluated such as heart and color Doppler was applied for identifying vascular connections between the twins. Liver and GIT were evaluated for complications of the hepatobiliary system. The pelvis was evaluated for the bladder, genitals, and for fusion abnormalities. The number of limbs was also noted.

Table 3: Comparison of various types of conjoined twins in our study




Type	Post Natal Image	Head	Heart	Liver and Git	Pelvic Organs	Limbs	Vertebral Column
Thoracophagus		Separate	Common	Separate	Separate	Separate	Separate
Thoraco-Omphalopagus		Separate	Common	Common	Separate	Separate	Separate
Thoraco – Omphalopagus		Separate	Common	Common	Separate	Separate	Separate
Thoracopagus	N/A	Separate	Common	Separate	Separate	Separate	Separate

Table 4: Congenital anomaly in various organ system

Organ System	Associated Congenital Anomaly
Msk	Ctev, Kyphoscoliosis, DDH
Thorax	Congenital Diaphragmatic Hernia, Tapvc, Dextrocardia
Git	Anomalous Hepatic Venous System, Bowel Atresia, Colovesical Fistula, Meckel’s Diverticulum
Genitourinary System	Renal Dysplasia, Duplax Pelvic System, Puj Obstruction, Vuj Obstruction

The first case was a pair of thoracophagus twins (fig 1) with 14 weeks of gestational age diagnosed in a 23-year primigravida. USG with curved array probe revealed single placenta, single umbilical cord with conjoined twins from thorax region till the upper part of the abdomen facing towards each other with two

separate heads, fused thorax with shared heart (fig 2A and 2B). They had two unfused spines and two separate pelvis. Each of the twins had two pairs of limbs. One of the twins had Occipital encephalocele (fig 2C) and club foot deformity in both lower limbs (fig 2D)



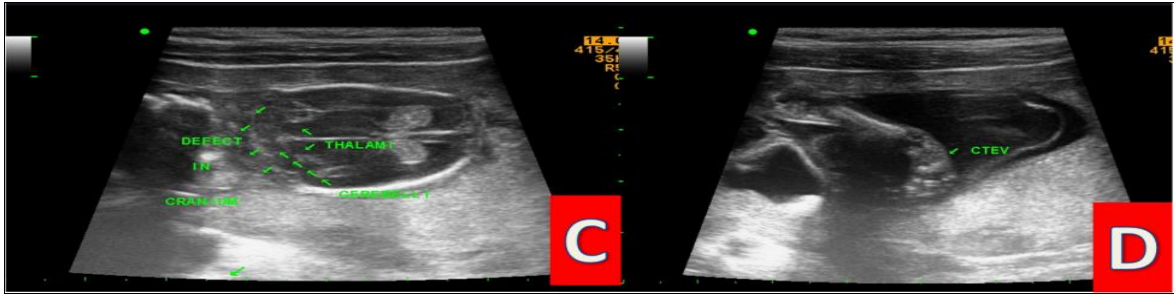


Fig 1: Thoracophagus twin (USG images)

The second case also was a pair of thoraco-omphalopagus twins (fig 3) with 15 weeks of gestational age diagnosed in a 24 yrs old primigravida. USG with curved array probe revealed a single placenta with a single umbilical cord and conjoined twins fused from the thorax till the umbilical region (fig 4A and 4B). Twins

had two separate heads with the shared sternum, diaphragm, and abdominal wall till the umbilicus (fig 4C). They were sharing a common heart (fig 4D) and liver. Pelvis and urinary tract were separated. Each of the twins had a two pair of limbs.



Fig 2: Thoraco-omphalopagus twin

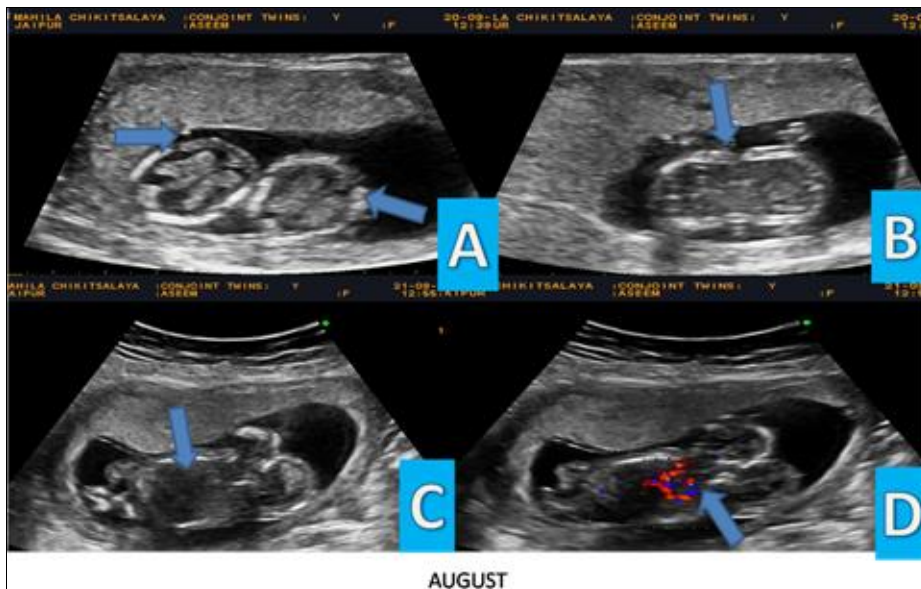


Fig 3: USG images of thoraco-omphalopagus twin

The third case was a pair of thoraco-Omphalophagus twins (fig 5) with 20 weeks of gestational age diagnosed in a 27-year-old second gravida with past obstetric history of normal vaginal delivery of term child. USG with curved array probe revealed single placenta with a single umbilical cord, conjoined twins had

2 separate heads (fig 6A) and they were fused from the thorax till the umbilical region with sharing of a common heart and liver(fig 6B and 6C). Pelvis and urinary tract were separated. Each of the twins had two pairs of limbs.

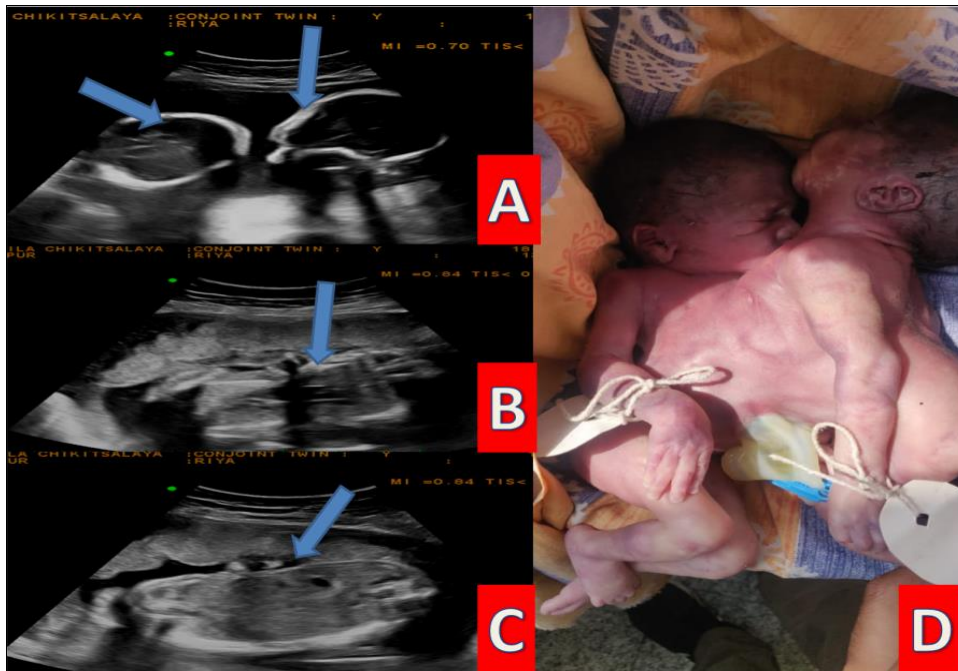


Fig 4: Thoraco omphalophagus twin

The fourth case was a pair of thoracophagus twins of 13 weeks of gestational age diagnosed in a 24-year-old primigravida. USG with curved array probe revealed single placenta with single umbilical cord conjoined twin from thorax region till the upper part of the abdomen facing towards each other with two separate

faces, fused thorax with shared heart, two unfused spines and two separate pelvis (fig7A). Each of the twins had two pairs of limbs. 3D ultrasound was done for better visualization of the thoracophagus twins (fig 7B).

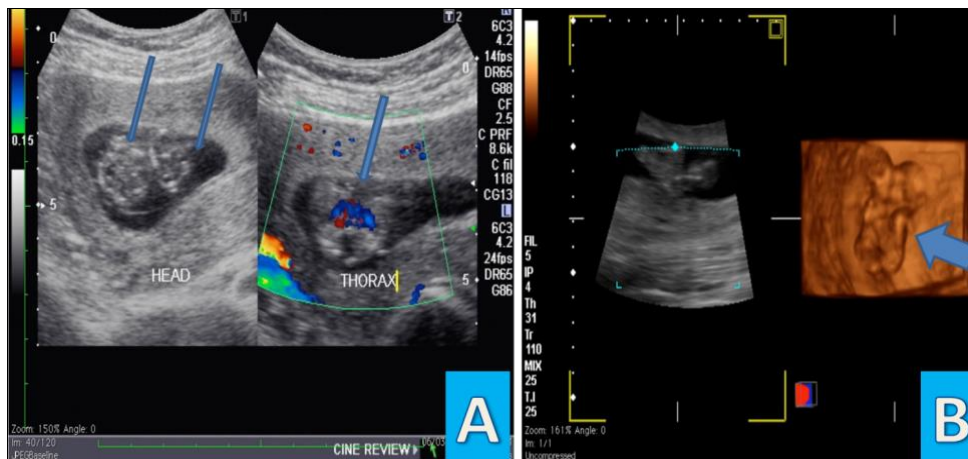


Fig 5: ultrasound (a) 2d (b) 3d images of a case of thoracophagus twin of 13 weeks gestational age.

Results

After detailed counseling and obtaining written consent from all the parents, both the thoraco-omphalopagus and thoracopagus twin pregnancies were terminated by medication. No complications were observed in the postoperative period in all four cases.

Discussion

Conjoined twins are rare, with the estimated prevalence is 1:50000 to 1:200000 [1]. Fusion theory and fission theory are two

theories behind the origin of conjoined twins [2]. According to the fusion theory, two separate mono-ovulatory embryonic discs undergo a secondary association after which conjoined twins develop. According to the fission theory, about 13–15 days post-fertilization, the embryonic disc undergoes an incomplete separation leading to some parts of the twin remaining fused.

Antenatal Assessment

The antenatal diagnosis of conjoined twins can be made with high-resolution ultrasound as early as 12 weeks gestation and is

important for optimal obstetric management because early termination of pregnancy has less risk of complications. Fetal echocardiography should be done optimally at 18 weeks because the degree of cardiac fusion will determine the likelihood of successful separation and postnatal viability of the twins. During an ultrasound, conjoined hearts are easier to examine in utero because the amniotic fluid acts as a buffer. The other US features that suggest the possible diagnosis of conjoined twins include constant relative positions of the fetuses with heads and other body parts persistently at the same level on repeat scans, Fetus facing towards each other, fused body parts with shared organs, fewer limbs than expected and a single umbilical cord with more than three vessels.

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