

# An Interesting Case of Undiagnosed Conjoined Twins

<sup>1</sup>Krishna Kirtan, <sup>2</sup>Divyasree Doopadapalli, <sup>3</sup>Nageshu Shailaja, <sup>4</sup>Lingegowda Krishna, <sup>5</sup>BS Bhat

## ABSTRACT

A 20 years old primigravida who presented to us at 36 weeks 3 days gestation with labor pains. She was perceiving fetal movements well, booked and immunized at a private hospital in Hosur. On examination, she was 7 cm dilated, well effaced, breech at 0 station. She was taken up for emergency lower segment cesarean section (LSCS) in view of primi with breech and mother not willing for vaginal delivery. An alive term 3.5 kg female paraphagus dicephalus twins was extracted. Extraction was difficult. Apgar was 3 at 1 minute and 5 at 5 minutes. The babies died after 45 minute. External examination showed that the twins were conjoined from the level of thorax downward. The twins had two heads, two upper limbs and a shared thorax, abdomen, pelvis and a single female external genitalia and single pair of lower limbs with CTEV (dicephalus, dibrachius, dispinous, thoraco-omphalopagus twins).

**Keywords:** Conjoined twins, Dibrachius twins, Dicephalus twins, Dispinous twins, Paraphagus, Thoraco-omphalopagus twins.

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## INTRODUCTION

The description of conjoined twins is legendary. Its earliest record occurred in 945 B.C., in Constantinople. In this case, the twins were joined at the abdomen and attempted separation occurred after the death of one of them, at age 30. However, the other twin died 3 days later.<sup>1</sup>

Since then, several reports of new conjoined twins are recorded, but the first published report of successful separation was described by Konig in 1689. The surgeon, Johannes Fatio, operated the twins joined at the ischium.<sup>2</sup>

Conjoined twins are rare and complex complication of monozygotic twinning (MC). The incidence of conjoined

twins is reported as approximately 1/30,000 to 100,000 and 1/600 of twin births.<sup>3</sup>

As 60% are stillborn or die shortly after birth, the true incidence is around 1 in 200,000 live births.<sup>5</sup> Girls predominate in the ratio of 3:1.<sup>4,5</sup>

There are two theories explaining the etiology of conjoined twins: According to the 'Fission theory', 13 to 15 days after fertilization, the embryonic disk undergoes an incomplete separation, whereas in the 'Fusion theory', two separate mono-ovulatory embryonic disks undergo a secondary association. Recently, the 'Fusion theory' has been accepted, because it can explain all the conjoined twin phenomena.<sup>6</sup>

Conjoined twins with the classification of two-dimensional (2D) ultrasound are too difficult because of their complex three-dimensional structures. Three-dimensional (3D) ultrasound, reveal its complex anatomical spatial relationships, may be useful in determining the complex fetal anatomy of conjoined twins.<sup>7</sup>

Early prenatal diagnosis of conjoined twins allows better counseling of the parents regarding the management options, including continuation of pregnancy with post-natal surgery, termination of pregnancy or selective fetocide.<sup>8</sup>

## CASE REPORT

We report a 20 years old primigravida who presented to us at 36 weeks 3 days gestation with labor pains. She was perceiving fetal movements well, booked and immunized at a private hospital in Hosur. She was married for 10 months and no history of twins in the family. A scan was done 3 days prior to admission which was normal. No anomalies were noted. On examination she was 7 cm dilated, well effaced, breech at 0 station. She was taken up for emergency lower segment cesarean section (LSCS) in view of primi with breech and mother not willing for vaginal delivery. An alive term 3.5 kg female paraphagus dicephalus twins was extracted. Extraction was difficult. An inverted T incision was made on the uterus to facilitate delivery of the baby.

Apgar was 3 at 1 minute and 5 at 5 minutes. The babies died after 45 minutes. Resuscitation was stopped as the attenders were not willing for resuscitation and NICU care after the prognosis was explained. Autopsy was not performed as the parents refused to give consent.

<sup>1,2</sup>Assistant Professor, <sup>3</sup>Associate Professor  
<sup>4</sup>Professor and Unit Head, <sup>5</sup>Professor and Head

<sup>1-5</sup>Department of Obstetrics and Gynecology, PES Institute of Medical Sciences and Research, Kuppam, Andhra Pradesh, India

**Corresponding Authors:** Krishna Kirtan, Assistant Professor Department of Obstetrics and Gynecology, PES Institute of Medical Sciences and Research, Kuppam, Andhra Pradesh India, Phone: +9989448753, e-mail: kirtank1@gmail.com

External examination showed that the twins were conjoined from the level of thorax downward. The twins had two heads, two upper limbs and a shared thorax, abdomen, pelvis and a single female external genitalia and single pair of lower limbs with CTEV (dicephalus, dibrachius, dispinous, thoraco-omphalopagus twins) (Figs 1 to 3).

### Investigations

#### Antenatal

Ultrasonography (USG) on 10/5/13 revealed SLIUG of 36 weeks 1 day with breech presentation with no obvious fetal abnormality, posterior placenta and adequate liquor. Neither nuchal translucency nor anomaly scan was performed.

#### Postnatal

Ultrasonography of the neonatal abdomen revealed a shared liver, a single spleen on the left side, single pair

of kidneys, two vertebral columns, bilateral inferior vena cavae and a single left abdominal aorta and a single uterus.

There were two hearts which were not fused. Both the hearts had single atrial chamber communicating with both the ventricles through two atrioventricular valves (Fig. 4). The atrium received right-sided IVC without any drainage of pulmonary veins. A single great vessel, aorta, originated from it, forming a right aortic arch. Pulmonary artery was not seen in right-sided heart.

A single infantile uterus was noted (Fig. 5).

- Infantogram revealed that the twins had two heads, two vertebral columns, two upper limbs and a shared abdomen, pelvis and a single pair of lower limbs (Fig. 6).

### Diagnosis

Primigravida with 36 weeks 3 days gestation with breech presentation in active phase of labor with moderate anemia.



Fig. 1: A dicephalus, dibrachius, dispinous, thoraco-omphalopagus twins



Fig. 2: A dicephalus, dibrachius, dispinous, thoraco-omphalopagus twins with congenital talipes equinovarus



Fig. 3: Dorsal surface of dicephalus, dibrachius, dispinous, thoraco-omphalopagus twins with congenital talipes equinovarus and occult spina bifida

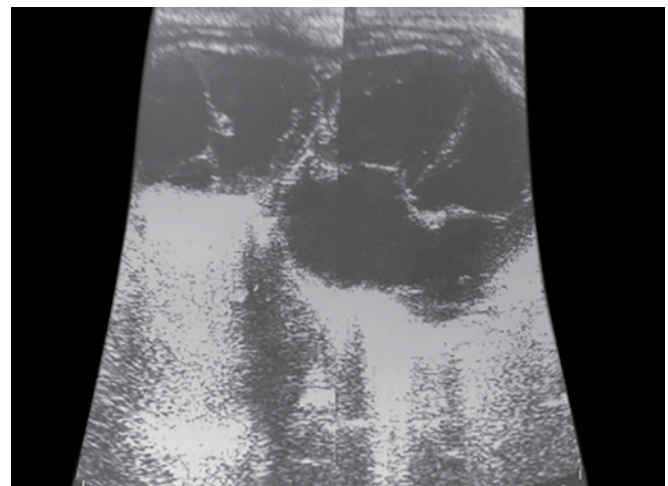
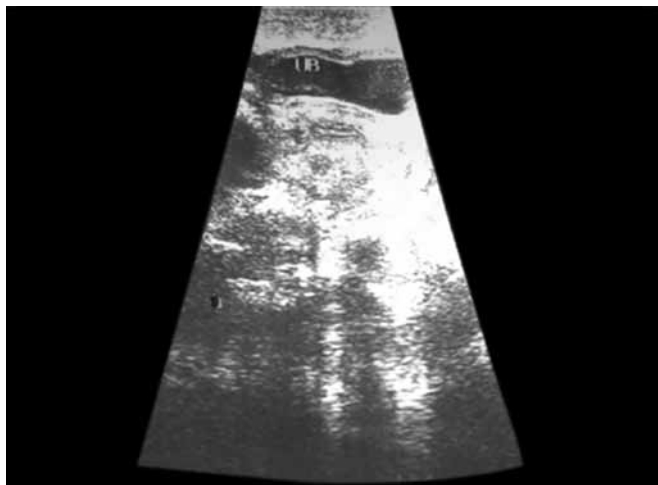
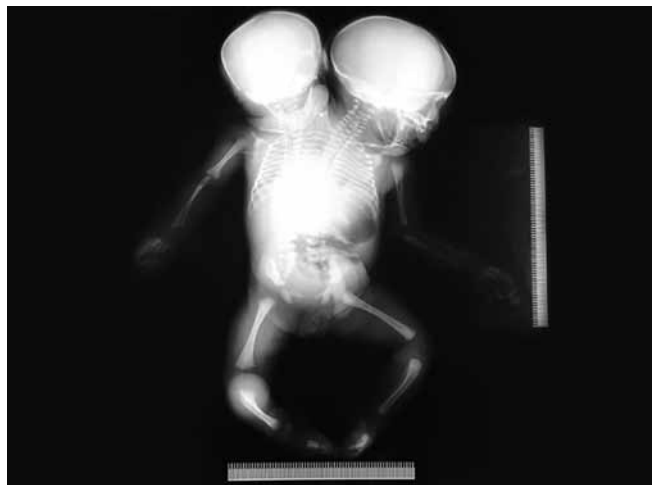


Fig. 4: Postnatal ultrasound of the two fetal hearts which were not fused. Both the hearts had single atrial chamber communicating with both the ventricles through two atrioventricular valves



**Fig. 5:** Postnatal ultrasound of the pelvis showing a partially filled bladder and an infantile uterus



**Fig. 6:** Infantogram which revealed that the twins had two heads, two vertebral columns, two upper limbs and a shared abdomen, pelvis and a single pair of lower limbs

## DISCUSSION

Prenatal diagnosis of twins is the most important aspect in the management of conjoined twins. The parents should be counseled regarding the various treatment options and prognosis following delivery.

Conjoined twins should be suspected in case of a monochorionic monoamniotic twins. On ultrasound presence of two distinct placentae rules out conjoined twins. Presence of polyhydramnios and bi breech or face to face presentation are indicative of conjoined twinning. There are reports of increased levels of alpha fetoprotein are indicative,<sup>9</sup> however there is no data regarding the sensitivity and specificity of this marker for detection of conjoined twins is not available. In order to prevent missing the diagnosis the scan should be repeated at least once.

When ultrasound is done by an experienced radiologist conjoined twins can be detected early in the first trimester. There have been several such case reports. There is not much improvement in the diagnosis of twins with 3D ultrasound.

The main stay in diagnosis of conjoint twins is use of 2D ultrasound with Doppler at 11 to 14 weeks. Specific features suggestive of conjoined twins are:

- Bifid appearance of the fetal pole in the first trimester
- Absence of separating membrane between fetuses
- Nonseparation of fetal bodies which persist and the fetal parts are seen at the same level
- More than three vessels in the umbilical cord
- Unusual extension of the fetal spines
- Unusual proximity of the fetal limbs
- The fetuses do not change position to one another after movement or manipulation.<sup>10</sup>

Conjoined twins may be misdiagnosed as singleton pregnancy in cases with extensive fetal conjunction.

Hence, the duplication of all anatomical parts should be identified. In the third trimester identification of conjoint twins becomes difficult because one of the fetal head may be engaged while the other head is unengaged and may be mistaken as separate. Newer modalities like 3D, 4D, power Doppler, fetal rapid MRI can be used as an adjunct in diagnosis.

The aim of diagnosis is to identify the type of fetal conjunction is to predict the immediate postnatal viability of the fetuses and the effect of surgical separation on both the babies. The overall mortality rate of conjoined twins is as high as 28% prenatally, 54% immediate postnatally, while long time survival rates are only about 18%.<sup>11</sup> It is important to diagnose in 2nd trimester before 20 weeks as termination of pregnancy by the vaginal route is possible.

## CONCLUSION

Conjoined twins should be suspected in all monochorionic, monoamniotic twin pregnancies and careful prenatal diagnosis should be undertaken to exclude any of classical signs which are suggestive of conjoined twins and to identify the severity of the shared fetal organs for perinatal management. This proves the importance of anomaly scan being done by a trained sonologist. If they are diagnosed before viability, the parents should be offered termination of the pregnancy. Hence, the evaluation of fetuses with imperfect twinning should be multidisciplinary, involving mainly obstetricians, pediatricians, and pediatric surgeons, to decide the best time to interrupt pregnancy and define the chances of postnatal separation. However, such dilemmas may constitute true ethical dilemmas, in which different aspects should be discussed and analyzed, along with the healthcare team and the family.

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