

Prenatal Ultrasound Diagnosis Of Cephalothoracomphalopagus Janiceps Monosymmetros.

SANDHYA SUDHIR KAMBLE¹, FAIZAL BIN SHARIF², INDULEKHA ANAYATH³

¹Maternal and Child Health Clinic, Seria Health Centre, Belait,

² Department of Radiology, Suri Seri Begawan Hospital,

³Department of Obstetrics and Gynaecology, Suri Seri Begawan Hospital.

ABSTRACT

The role of antenatal ultrasound is established in detecting fetal well-being and abnormalities. Cephalothoracomphalopagus conjoined twin is a rare occurrence and typical ultrasound features have been described in the literature. We report a case of cephalothoracomphalopagus conjoined twin diagnosed by ultrasound performed in a peripheral Maternal and Child Health Centre and later confirmed at a district hospital. The objective of this case report is to emphasize the possibility of fetal abnormalities even in low risk antenatal case at early gestational age and highlight the ultrasound features of this rare condition.

Keywords: Conjoined twin, conjoint twins, antenatal ultrasound, cephalothoracomphalopagus, prenatal diagnosis

INTRODUCTION

Cephalo-thoraco-omphalopagus is an extremely rare variety of conjoined twin where the twins are joined with a single head, neck, thorax and abdomen but have 4 limbs.¹ Majority of conjoint twins are not viable and are still birth and an additional third will not survive more than a day.¹ With routine use of antenatal ultrasound, an early diagnosis can be made. We report a case of Cephalo-thoraco-omphalopagus conjoined twin in a young primigravida who did not have any recognized risk factors for congenital anomalies and discussed about the incidence and causes of such disorder.

CASE REPORT

A 29-year-old healthy, clinically uncomplicated primigravida, was first seen at the Maternal and Child Health clinic at Seria Health Centre, Brunei Darussalam for routine second trimester ultrasound as internationally recommended.² This was a spontaneous pregnancy with a married life of less than one year. The patient had no major medical illness, consanguinity or previous history of twin pregnancy. There was no significant family history of multiple gestations or any congenital anomaly. There was no history of smoking, drugs consumption, exposure to teratogenic medications or irradiations, assisted reproduction or infectious diseases during the pregnancy.

The initial ultrasound scan revealed an abnormal alive conjoined twin. The fetus had single large head with fused thalamus

Correspondence author: Sandhya Sudhir KAMBLE, P.O. Box 585, Seria Post Office, KB1190, Brunei Darussalam.
Email: sdhrkamble@yahoo.co.in
Contact number: +6738930721

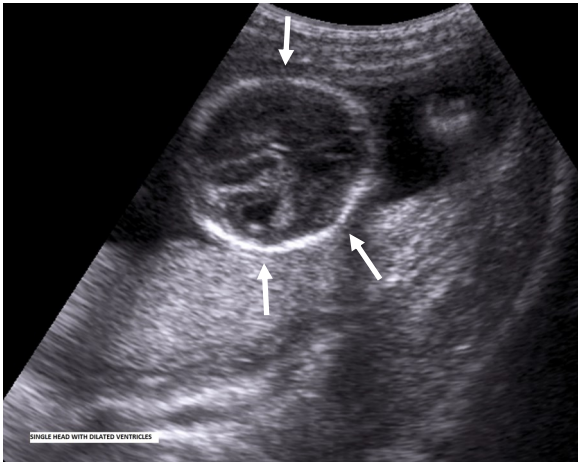


Figure 1: Ultrasound scan image showing a single large head with dilated ventricles (White arrows).



Figure 2: Ultrasound scan image showing conjoint twin with two separate spines (White arrows).

and dilated ventricles (Figure 1). There was a single thorax with two lower abdomens and pelvic girdles. There was complete independent cervico-thoraco-lumbosacral vertebral columns diverged inferiorly (Figure 2). The twin had a single neck (Figure 3). There were two pairs of arms and legs, with single placenta and polyhydramnios.

She was then referred to the district hospital for detailed ultrasound scan to be performed by the radiologist. In addition to the above findings that were confirmed, the conjoined twin revealed shared lungs, heart and stomach. The liver was wide and shared into two halves.

The ultrasound findings were explained to the couple and they were counseled extensively. The couple opted for termination of pregnancy. The patient was admitted under the care of the Obstetrician. The pregnancy was terminated at 21 weeks of gestation. The patient delivered vaginally, a female conjoined twins with birth weight of 750 grams. On examination of twins, the findings of ultrasound examination were confirmed including a single face with large head. Both babies had normal shoulders, arms, hands, legs and feet. Thorax and abdomen were fused. There was a single placenta with a single umbilical cord. Both eyes were normal, the tongue was en-

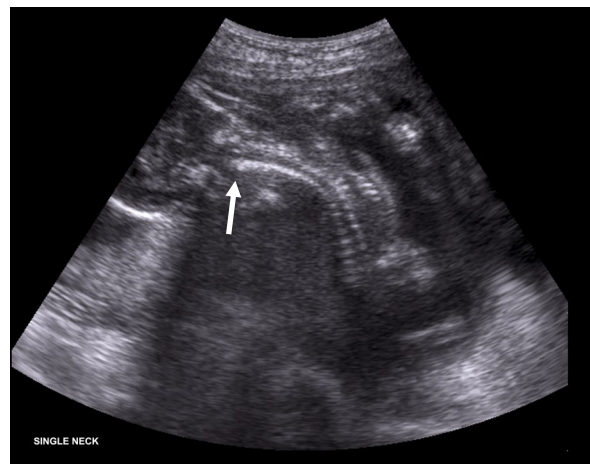


Figure 3: Ultrasound scan image showing conjoint

larged with a small cyst of 0.5 cm hanging outside the mouth. Considering the sensitivity of the matter and parents request, no photographs were obtained or any postmortem examination conducted. amination conducted.

DISCUSSION

Twins are categorized into dizygotic twin and monozygotic twin. Dizygotic twin is a phenomenon resulting from fertilization of 2 ova separately by 2 sperms. The monozygotic twin occurs due to fertilization of a single ovule to form one zygote, which then divides into 2 separate embryos.³ The dizygotic twin is constrained by conditions such as heredity, race, maternal age, parity and history of as-

assisted reproduction. The monozygotic twin in turn does not depend on any condition, occurring independently and randomly.³⁻⁵

Conjoined twin is a variety of monozygotic twins which results from an incomplete division of embryonic disk after 13th day of conception. There is incomplete separation between both the embryos, which by design will occur in single amniotic cavity and same placenta.^{3,6,8} This is associated with high risk of perinatal morbidity and mortality.^{3,8}

There are several types of conjoined twins, which are named for the site of union followed by suffix *pagus* meaning 'fixed'.⁴⁻⁶ There can also be overlap with more than one type involved.⁵ Conjoint twins are classified by Spenser according to the site of union: Craniopagus-joined at cranium, Pyopagus-joined at sacrum and coccyx, Thoracopagus-joined at thorax, Thoraco-omphalopagus-joined at thorax, Thoraco-omphalopagus-

joined from thorax to abdomen; Ischiopagus-joined at pelvic area, Cephalo-thoraco-omphalopagus- joined from cranium to thorax and abdomen (Figure 4).^{4,6-8}

Although historically there have been many reported cases of conjoined twins through-out the centuries, the most famous case was of Chang and Eng Bunker born in Siam in 1811.^{4,5} The incidence of conjoined twins is in the range of 1:50000 to 1:100000.^{3,6,9} However the incidence rate of the rarer variety of cephalothoracomphalopagus conjoint twin is 1:3000000 pregnancies or 1:58 conjoined twins.^{9,10} The ratio of females to males is 3:1.^{4-6,8} Thus cephalothoracomphalopagus is a rarest type of conjoined twin pregnancy described as imperfect division of head, chest and abdomen at the area of umbilicus, but separated vertebral columns, limbs and pelvis.

The term janiceps is derived from the 2-faced Roman god named Janus. Janiceps twins are categorized into 2 subgroups: disymmetros and monosymmetros. In janiceps disymmetros the 2 faces are symmetrical and identical on a single head, in which the orientations of 2 notochord axes are perfectly ventroventral. In janiceps monosymmetros variety there is asymmetric union of 2 faces due to 1 poorly developed notochord.^{3,9,10}

Our case is classified as cephalothoracomphalopagus janiceps monosymmetros which is the rarest type of conjoined twin with features described earlier. There have been few cases of conjoined twin reported in the late second and third trimester of pregnancy in the last ten years with the outcome of termination of pregnancy.^{4,6,9} Another case of conjoined twin was diagnosed early in first trimester.⁵ Our case was diagnosed at 19 weeks of gestation and the pregnancy was terminated at 21 weeks. This substantially reduced the psychological stress and anxiety

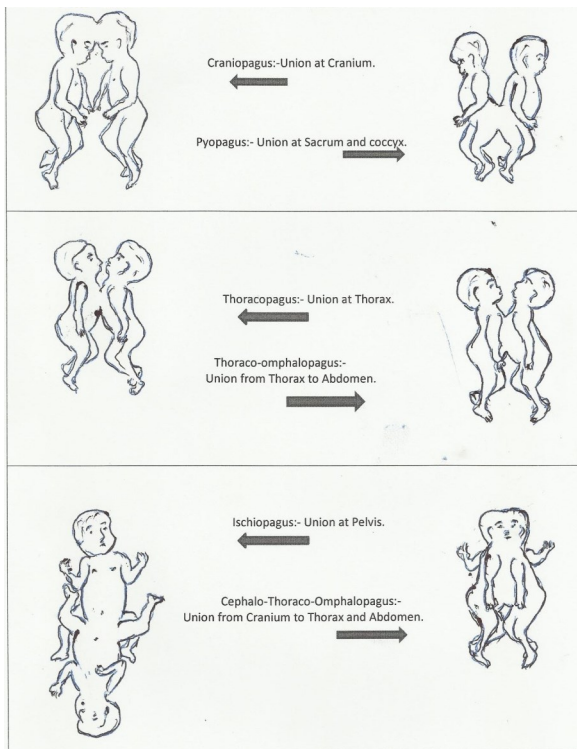


Figure 4: Spenser classification of Conjoint twins.

of the couple.

The antenatal diagnosis of conjoined twins is very important at early gestation of pregnancy in order to counsel the parents about interruption of pregnancy. The neonatal prognosis is extremely poor and surgical separation of the complex defects is usually not offered.³ Conjoined twins can cause dystocia with the risk of rupture of uterus and quite often require cesarean section which may have negative consequences for the obstetrical future of the mother.⁶ Before 24 weeks of gestation, the termination of pregnancy by vaginal route is opted, whereas after 24 weeks of gestation termination by hysterotomy is seen prudent.^{3,10}

The present case study highlights two important points related to clinical practice. It is possible to detect serious congenital abnormalities on antenatal scan performed at peripheral health center and it can occur even in low risk cases.

CONCLUSION

In conclusion, Cephalothoracomphalopagus janiceps momosymmetros is extremely rare variety of conjoined twin and the prenatal diagnosis with typical ultrasound features at earlier gestation is very important in order to counsel the parents.

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