

Thoraco-Omphalopagus Conjoined Twins: Imaging and Antenatal Diagnosis

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A B S T R A C T

Introduction: Conjoined twins are a rare occurrence with a reported incidence of 1 per 33,000-165,000 births in developed countries. Since still birth rate amongst conjoined twins is 40- 60% the incidence becomes approximately 1 per 200,000 live births. Early diagnosis by ultrasound scan is very important as it provides the parents with an option of safe termination. An alternative would be MRI which will give better tissue contrast.

Case Report: A 28-year-old G2P1 female had an antenatal ultrasound done for the first time at 16 weeks of gestation for assessment of fetal being and to know the gestational age. An ultrasound examination done by us showed two fetal heads at the same level and in the same plane. Both heads had a fixed relation with each other. The fetal chests and abdomen were found to be fused. Based upon ultrasound findings a diagnosis of thoraco-omphalopagus conjoint twins was made. Parents opted for medical termination of pregnancy hence MTP was done. An MRI was done before termination for confirmation of diagnosis, ruling out associated congenital anomalies and for academic purpose. MRI confirmed the ultrasound findings and ruled out presence of associated congenital anomalies.

Conclusion: Conjoined twins are a very rare malformation seen exclusively in monoamniotic monochorionic twins. It can be suspected on antenatal ultrasound if in a twin pregnancy both fetal heads persistently maintain same level. Early antenatal diagnosis is essential to provide the parents with an option of safe termination.

Keywords: Monoamniotic-Monochorionic Twins, Thoraco-Omphalopagus, Conjoined Twins, Antenatal Diagnosis

INTRODUCTION

Conjoined twins are amongst the rare and fascinating congenital human malformations. The incidence is very rare and reported to be between 1:33000 to 1:165000 pregnancies.¹ Approximately 75% of the affected fetuses happen to be female. The common types of conjoint twins include thoracopagus and omphalopagus. The other less common forms of conjoint twins include thoraco-omphalopagus (joined at chest and abdomen), pyopagus (joined at buttocks), ischiopagus (joined at the ischium) and craniopagus (joined at the head).² These twins are further divided respectively in symmetrical or asymmetrical type depending upon whether there are 2 well developed babies or there is a small part of the body that is duplicated. The widely accepted mechanism for development of conjoint twins is incomplete splitting of monozygotic twins after 12 days of embryogenesis.³ Due to high morbidity and mortality associated with conjoint twins antenatal diagnosis is imperative. All the monozygotic twins should be screened for a possibility of conjointment. If conjointment is present then associated

congenital anomalies like complex congenital heart diseases, Lower GI anomalies like imperforate anus, genitourinary and CNS anomalies should be ruled out.⁴ The antenatal diagnosis is usually done by ultrasonography which is safe, accurate, reliable and quick. An alternative would be obstetric MRI which will give a better tissue contrast and further give better details of associated congenital anomalies.⁵ Here we are presenting a rare case of conjoined twin (Thoraco-omphalopagus) diagnosed by ultrasound and MRI at 16 weeks of gestation.

CASE REPORT

A 28 years old Gravida 2, Para 1 living 1 lady at 16 weeks period of gestation, had come for antenatal ultrasound at radiology department of our institute. She was advised basic investigations including CBC, HIV, HBsAg and antenatal ultrasound. Ultrasonography performed at our department revealed monoamniotic-monochromic twins with four extremities, 2 spines, single liver and single heart and fused chest walls. Both fetal heads were found to be at the same level and in the same plane and had fixed relation with each other. The fetal chests and abdomen

were found to be fused. Two stomach single bubbles were seen. Ultrasound diagnosis of conjoined twin (Thoraco-omphalopagus) was made on the basis of these findings [Fig. 1].

An Obstetric MRI was advised to confirm ultrasound findings and for teaching and academic purpose with the informed consent of the patient. MRI showed complete fusion of the chest wall and the thoracic cavities. Two separate spinal cords, single heart and a single liver was demonstrated on MRI. Additionally short spines with scoliosis were noted on MRI. No Other congenital anomaly was found. Thus these findings confirmed the MRI diagnosis of Thoraco-omphalopagus conjoined twins [Fig. 2].

After proper counseling patient opted for termination of pregnancy. A medical termination of pregnancy was carried at 16 weeks of gestational age. Gross Examination Of the abortus confirmed the diagnosis of thoraco-omphalopagus conjoint twins [Fig. 3].

DISCUSSION

The classification of conjoint twins depends upon the site of union and they are classified into Thoraco-omphalopagus (chest, abdomen, or both), thoracopagus (chest), omphalopagus (abdomen), Pygopagus (buttocks), Ischiopagus (ischium) and Craniopagus (head) depending upon the site where they are united. The analysis of survived and aborted fetuses showed that most of the conjoint twins are females with a M: F ratio of 1:3. The survival depends upon the type of union but overall survival rates reported range between 20-25%.⁶

The exact cause of this is not known but it is widely accepted that conjoint twins occur due to incomplete splitting of monozygotic twins after 12 days of embryogenesis. The risk factors may include intake of clomiphene citrate, valproate and griseofulvin during periconceptual period. No mutations are yet identified to be associated with conjoint twins. Some researchers proposed the theory of abnormal X-chromosome inactivation while some other researchers have refuted this theory. There is no evidence of increased recurrence risk in case of pregnancy complicated by conjoint twins.⁷

The presentation and prognosis depends upon the type of conjoint twins. In case of craniothoracopagus where there is union at the level of cranium and thorax prognosis is poor as there is only one brain and surgical separation is not an available option but fortunately this type of conjoint twins are rare. Craniopagus twins may be partial or complete depending upon whether they significantly share dural venous sinus system. In the bicephalus conjoint twins there is presence of 2 heads on 1 trunk. In cases of omphalopagus twins the fusion occurs at abdominal wall. Omphalopagus may be isolated or associated with union at the level of thorax in which case they are called thoraco-omphalopagus. These types of conjoint twins may

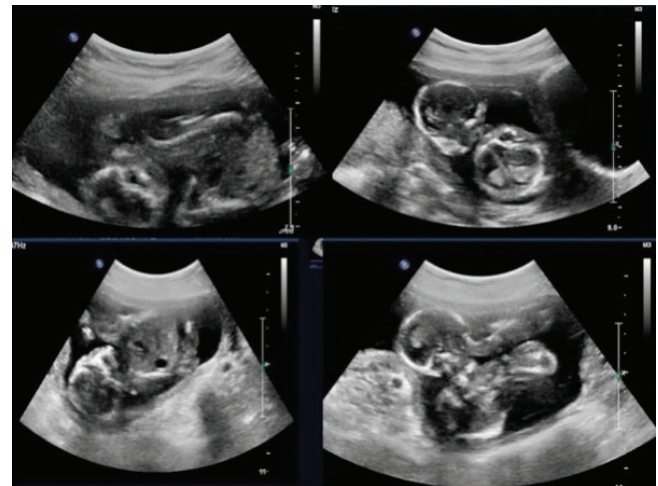


Figure-1: Antenatal ultrasound showing thoraco-omphalopagus conjoint twins .Note fused upper chest to the lower chest and abdomen ,2 separate heads and spines and single heart and liver.

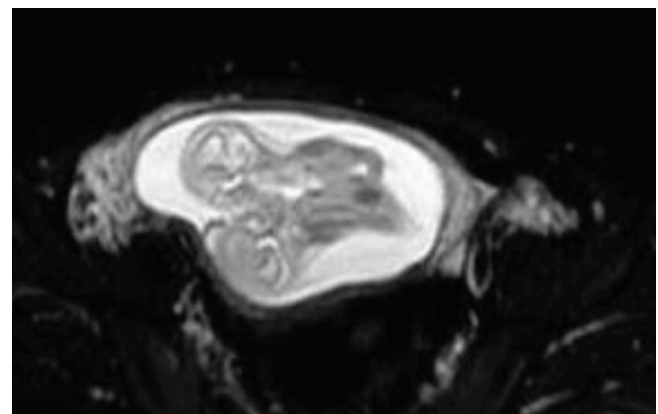


Figure-2: Antenatal MRI showed fused chest wall, scoliosis, single Liver and Heart confirming the diagnosis.



Figure-3: Gross specimen of aborted conjoint twins confirmed the ultrasound /MRI findings

undergo complex surgery for separation if they are found not to be sharing vital organs. Other less common forms of conjoint twins include parapagus (union at the trunk), pyopagus (union at sacrum) and rachipagus (fusion at vertebral columns).⁸

Early detection of this condition is of paramount importance so as to provide the parents with an option of safe termination of pregnancy. Ultrasound is a safe

and quick method by which conjoint twins can be diagnosed even in early pregnancy (as early as 8-10 weeks). The twin fetuses maintaining a fixed alignment is a sign of conjoint twins. This may be associated with polyhydramnios.⁹ Transvaginal scan may provide a better view and inability to separately see different parts of the fetuses may help in early diagnosis. Once the diagnosis of conjoint twins is confirmed by ultrasound it is of critical importance to find out the exact site of union, number of vital organs including brain, heart and liver and presence of any associated congenital anomaly like congenital heart disease, urogenital anomalies or neural tube defects. Magnetic resonance imaging has the advantage of not being operator dependant and having better tissue contrast. Moreover MRI can pick up presence of associated congenital anomalies at an early stage. The option of medical termination of pregnancy can be offered to the family if diagnosed before 20 weeks of gestational age.¹⁰

Since in this case the diagnosis has been made at 16 weeks and because the parents chose termination of pregnancy medical termination was performed. An MRI was done before termination of pregnancy for finding out presence of any associated congenital anomaly and for teaching purpose. MRI confirmed the ultrasound findings and there was no evidence of any additional congenital anomalies. After termination examination of gross specimen of conjoint twins confirmed the presence of thoraco-omphalopagus conjoint twins.

CONCLUSION

Conjoined twins are a rare and complex occurrence. Associated high perinatal mortality makes early diagnosis by ultrasound an essential requirement so as to provide parents with an option of safe medical termination of pregnancy.

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