

Thoraco-Omphalopagus: A Case of Conjoined Twin with a Single Heart

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Received: January 14, 2016; **Published:** January 29, 2016

Abstract

A conjoined twin is a rare disorder affecting 1/ 30,000 to 100,000 births. A 33 years old multigravida of G3P2, with previous II CS at 24 weeks of gestation was referred for an anomaly scan for suspecting fetal anomalies. Ultra sound revealed monochorionic, mono-amniotic living conjoined twins of 24 weeks with Thoraco-Omphalopagus type with a single heart. The poor prognosis in terms of perinatal morbidity and mortality was explained to the parents in such twins because of a single heart and poorly equipped neonatal units that can handle this case in our country. so, repeat elective Hysterotomy was done and the babies were conjoined at delivery. They died few hours after birth.

Keywords: Conjoined twins; Thoracopagus

Introduction

Conjoined twins is a rare disorder affecting 1 / 200 monozygotic twin pregnancies, 1 / 900 twin pregnancies, and 1 / 25,000 to 1,00,000 births [1,2]. Successful surgical separation of thoracopagus twins with separate hearts has been reported, but the success rate for those with a single heart is very low. So, their antenatal diagnosis should be made early in pregnancy. Color Doppler imaging is useful for imaging the cardiovascular anatomy in suspected conjoined twins having a fused hearts [3].

Conjoined twins who survive after separation are mostly females, at a ratio of 3:1 over males, the reason is not known. Conjoined twins usually result from the incomplete division of the early embryo with two fetuses. Incomplete division of the embryo is associated with incomplete formation of the organs and thus conjoined twins with fused organs usually have incompletely developed hearts and liver [4].

The thoracopagus is the most common type, accounting for 40% of all conjoined twins cases . Thoracopagus conjoined twins have the highest incidence of cardiovascular abnormalities; 90% share a common pericardial sac. The type of cardiac fusion is a critical factor for mortality [5,6].

Case report

A 33- years- old multi gravida G3P2 with history of previous II CS at 24 weeks of gestation, was referred to our hospital for anomaly scan for suspected fetal anomalies with twins. Her children are free and with no anomalies. No family history of twins. No history of assisted reproduction or relevant drug history. The vital signs and general examination findings were normal. Height of uterus was 28 weeks size, multiple fetal parts were palpable. On auscultation single fetal heart sound was heard.

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Blood investigations were within normal limits. Ultrasound showed monochorionic, mono amniotic living conjoined twins of 24 weeks thoraco-omphalopagus twin with single heart multi chambered with single umbilical cord with polyhydraminous. There was cleft upper lip in one of the babies (Figure 1,2,3).

The patients requested termination by caesarean section rather than vaginal delivery after counseling about the incompatibility of life of such twins in view of single heart and poor prognosis of these babies in Egypt. The patient delivered live female conjoined twins with poor APGAR score. Birth weight of both babies together was nearly one kilogram. Both babies have separate heads with two upper and two lower limbs with single umbilical cord and fusion at thorax and abdominal levels (Figure 4). They died few hours after birth. Post-operative period of mother was uneventful.



Figure 1: 3D ultrasound showing fused twins at thoracoabdominal levels with one cord and cleft upper lip of one baby.



Figure 2: 2D ultrasound showing single heart in chest of both twins.



Figure 3: 3D showing tightly fused both twins with cleft lip of upper lip.



Figure 4: Postoperative photo of conjoined twins with single umbilical cord.

Discussion

A conjoined twin occurs predominantly in females (3:1 female to male ratio). It results from incomplete division of the embryonic inner cell mass that occurs probably before the second week after fertilization. In the United States conjoined twins are commonly called as Siamese twins after Chang and Eng Bunker of Siam (Born in Thailand in 1811) who was displayed worldwide by P.T. Burnam. They were 63 years old when they died [2].

Conjoined twins are classified according to the site of union that could be ventral in 87% of the cases and dorsal in 13% of the cases. Ventral unions include the following (Spencer 1996):- Thoracopagus (19%), Omphalopagus (18%) and Ischiopagus (11%) (11%) The distribution of dorsal unions is Craniopagus (5%), Pygopagus (6%), and Rachiopagus (2%) [1]. In our case, ventral union of thoracopagus with a single heart was detected.

Early diagnosis of conjoined twins is possible at first trimester by ultrasound examination. A compressive ultrasound examination between 18 and 22 weeks may be useful to determine the anatomy of the shared organs and to detect associated malformations which allow the parents to decide whether continue or terminate the pregnancy. The following ultrasound findings increase the probability: twins face each other, heads are at the same level and plane, the thoracic cages are in unusual proximity, fetal heads are hyper extended

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and no change in the relative position of the fetuses with movement or in repeat examination [1]. Repeated 2D and 3D ultrasound examination, MRI scanning are useful for cardiac connections between the twins.

The outcome of conjoined twins is poor. Approximately 40% of them will be still born, 35% will die within one day after delivery, and the overall survival rate of conjoined twins is between 5-25%. Consultation with pediatric surgeon is needed for decision making. Termination of pregnancy is an option when the heart or brain is shared because in these cases attempt to separate usually fail. Plans should be made for caesarean delivery unless there is a special circumstance indicating safe vaginal delivery [7]. The only hope of independent life is through surgical separation. The first successful surgical separation of conjoined twins was achieved in 1953. Separation is feasible in Ischiopagus, Pygopagus, Omphalopagus and Parastic twins. Ideal time for separation at 9-12 months of age.

Thoracopagus conjoined twins with a fused heart always have complex abnormal cardiac anatomy [8-12]. The degree of cardiac fusion is most often the important factor in the prognosis. Leachman, *et al.* [8] classified twinning on the basis of cardiac fusion as follows: separate normal hearts (type A), fused atria with separate ventricles (type B), and fused atria and ventricles (type C). Andrews, *et al.* [9] reported echocardiography results in the evaluation of 23 sets of conjoined twins with thoracic level fusion, and classified them into four groups as: group A: separate hearts, separate pericardium; group B: separate hearts, common pericardium; group C: fused atria, separate ventricles; and group D: atrial and ventricular fusion. Thoracopagus and thoraco-omphalopagus twins have complex cardiac abnormal anatomy. The present twins were thoraco-omphalopagus with a four-chambered heart.

McMahon, *et al.* [12] reported cardiac malformations in a review of 1262 cases of all types of conjoined twins. Of all the cases studied, 314 twins were thoracopagus and typically shared a single multi-chambered heart; there were from one to four atria and from one to four ventricles, with several variations of the great vessels, and triple-outlet connection was observed in only three cases of all 1262 twins. Our twins died on the first day due to heart failure and progressive metabolic acidosis.

Conclusion

A conjoined twin is a well-known rare entity. It is very important to make a diagnosis in time to prevent complications. An ultrasound and color Doppler are of great help in making the early diagnosis. Detailed anomaly scan at 18-20 weeks helps to determine the shared organs and to detect malformation. Consultation with pediatric surgeon often facilitates parental decision making. Although conjoined twins, especially thoracopagus type, are known to present several cardiac anomalies, a shared single four-chambered heart a rare condition.

Conflicts of Interest

The authors declare that they have no conflict of interest.

Consent

The authors declare that:

1. They have obtained informed consent for the publication of the details relating to the patient.
2. They safeguard the identity of the patient.
3. This submission is compliant with the requirements of local research ethics committees.

Bibliography

1. Fernando Arias, *et al.* "Practical guide to high-risk regency & delivery". *Elsevier* (2008): 306-307.
2. Cunningham, *et al.* "Williams obstetrics". 20th Edition 38 (1996): 875.
3. N Uma D. "A Rare Case of Thoracopagus-Conjoined Twins with Single Heart". *Journal of Evolution of Medical and Dental Sciences* 4.28 (2015): 4912-4915.
4. Torako Omfalopaguslu yapışikizler. "Thoraco-Omphalopagus conjoined. *OlguSunumları Case Reports*". 13 (2013): 278-85.

5. Kalibushi BJ, *et al.* "Thoracopagus conjoined twin with one heart - Uncommon case". *International Journal of Current Microbiology and Applied Sciences* 4.4 (2015): 426-428.
6. Pandey S., *et al.* "Conjoined Twins with a Single Heart: A Rare Case Report". *Australasian Medical Journal* 4.4 (2011): 145-147.
7. Kapoor Mridula, *et al.* "Conjoined twins Case Report". *The journal of obstetrics and gynaecology of India* 63.1 (2013): 70-71.
8. Leachman RD, *et al.* "Cardiovascular evaluation of conjoined twins". *Birth Defects* 3 (1967): 52-65.
9. Andrews RE., *et al.* "Echocardiographic assessment of conjoined twins". *Heart* 92.3 (2006): 382-387.
10. McMahon CJ, *et al.* "Cardiac catheterization in diagnosis and management of congenital heartdisease in thoracopagus conjoined twins". *Catheterization and Cardiovascular Interventions* 51.2 (2000): 159-167.
11. Liu HC., *et al.* "Various modalities for evaluation of a fused heart in conjoined twins". *Pediatric Cardiology* 33.1 (2012): 192-200.
12. McMahon CJ and Spencer R. "Congenital heart defects in conjoined twins: out come after surgical separation of thoracopagus". *Pediatric Cardiology* 27.1 (2006): 1-12.

Volume 2 Issue 3 January 2016

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