CASE REPORT

Conjoined Twins: First Trimester Diagnosis at 9 Weeks

Amandeep Singh[1], Bikramjit Singh Sidhu[2]

Abstract

Introduction: Conjoined twins are an extremely rare occurrence registered in 1:50,000 to 1:100,000 live births. Due to the increased risk of perinatal morbidity and mortality it is crucial to do the prenatal evaluation and early diagnosis of conjoined twins by ultrasound scan in the first trimester. Early diagnosis in the first trimester gives the parents an option of safe termination of pregnancy.

Case report: A 27-year-old G2P1L1 female was referred to our department for routine first trimester antenatal ultrasound at 9 weeks 4 days according to last menstrual period (LMP). Ultrasound examination detected a monochorionic-monoamniotic twin pregnancy. The two fetuses were seen in a face-to-face fixed position with two separate fetal heads and chests seen. Two separate hearts were seen. The twins were found to be joined at the lower abdomen and pelvis. Based on ultrasound findings ischiopagus conjoined twins diagnosis was made. Parents were counseled and they opted for termination of pregnancy.

Conclusion: Conjoined twins are a very rare complication of monoamniotic-monochorionic pregnancies. Diagnosis on antenatal ultrasound can be made, if in a twin pregnancy, both fetuses persistently maintain a fixed position relative to each other. Early antenatal diagnosis by ultrasound gives the parents a chance to choose an option of safe termination.

Keywords: Conjoined twins, Early antenatal diagnosis, Ischiopagus, Monoamniotic-monochorionic twins.

Donald School Journal of Ultrasound in Obstetrics and Gynecology (2022): 10.5005/jp-journals-10009-1931

Introduction

Conjoined twins are an extremely rare occurrence happening in 1:50,000 to 1:100,000 live births.[1] Conjoined twins occur in about 1% of monochorionic pregnancies. The pathogenesis of this malformation is delayed aberrant division of a single zygote after 13–15 days of the embryonic period.[2] Monochorionic-diamniotic conjoined twins are even rarer and occur due to secondary fusion of two originally separate embryonic disks.[3] The common types of conjoined twins are thoracopagus (joined at the thorax), omphalopagus (joined at the abdomen), and thoraco-omphalopagus (joined at thorax and abdomen).[4] Ischiopagus variety is rare representing only about 12% of cases.[5] Conjoined twins prognosis depends on which body parts are fused. Approximately 60% of cases die *in utero* while about 35% are stillborn or die within the first 24 hours after birth.[6] Due to the high risk of perinatal morbidity and mortality, it is of vital importance to do antenatal evaluation and diagnosis of conjoined twins using ultrasound in the first trimester. Ultrasonography is a safe, reliable, and quick method for evaluation of conjoined twins. Early antenatal diagnosis of conjoined twins is crucial for accurate counseling of parents so that they can make an informed decision to choose early termination or, if pregnancy continues, to make plan for prenatal, perinatal, and postnatal care. Here the authors will present a rare case of conjoined twins (ischiopagus) diagnosed by ultrasound at 9 weeks 4 days of gestation.

[1,2]Department of Radiodiagnosis and Imaging, Sri Guru Ram Das Institute of Medical Sciences and Research, Amritsar, Punjab, India

Corresponding Author: Amandeep Singh, Department of Radiodiagnosis and Imaging, Sri Guru Ram Das Institute of Medical Sciences and Research, Amritsar, Punjab, India, Phone: +91 9872454954, e-mail: dr.amancs@gmail.com

How to cite this article: Singh A, Sidhu BS. Conjoined Twins: First Trimester Diagnosis at 9 Weeks. Donald School J Ultrasound Obstet Gynecol 2022;16(2):163–165.

Source of support: Nil

Conflict of interest: None

Case Description

A 27-year-old gravida 2, para 1, living 1 female was referred to our department for routine first trimester antenatal ultrasound at 9 weeks 4 days according to LMP. Basic investigations including complete blood count, bleeding time, clotting time, prothrombin time test, HIV, hepatitis B surface antigen, hepatitis C virus, and antenatal ultrasound were advised to her. The patient was in good general condition. There was no history of twin pregnancy in her family. The present pregnancy was a spontaneous conception. Present ultrasound examination revealed a monochorionic-monoamniotic twin pregnancy with two fetuses seen in a persistently fixed face-to-face position. A single yolk sac was seen. Two separate fetal heads and

chests were seen. Two separate hearts with separate cardiac pulsations were visualized. The twins were found to be joined at lower abdomen and pelvis (Fig. 1). One of the twins had mild ascites. Despite the repeated attempts there was no observed change in the relative positions of the fetuses. Subsequently, a three-dimensional (3D) volume dataset of the twins was acquired for further evaluation. Multiplanar views and surface-rendered images of the embryos were acquired which accurately delineated the level of fusion (Fig. 2). Based on ultrasound findings a diagnosis of ischiopagus conjoined twins has been made. Ultrasound findings were presented to the parents. All the possible management options were explained to them and they chose to terminate the pregnancy, which was done the following day.

Discussion

Conjoined twins are classified depending upon the site of fusion. The various types of conjoined twins include thoracopagus (fused at the thorax), omphalopagus (fused at the abdomen), craniopagus (fused at the level of the cranium), ischiopagus (fused at lower abdomen and pelvis), thoraco-omphalopagus (fused at thorax and abdomen), pyopagus (fused at sacrum and perineum), rachipagus (fused at vertebral column), and heteropagus (parasitic twins).[7] Ischiopagus variety is rare representing only about 12% of cases.[5] Most of the conjoined twins are found to be females. M:F ratio is 1:3. Chances of survival depend upon the type of fusion however overall survival rates are between 20 and 25%.[8] Conjoined twins in a triplet pregnancy are very rare, their incidence being less than one in a million deliveries, however, we should be aware of the possibility of conjoined twins in triplet pregnancies as well as the rate of complication in these pregnancies is even higher.[9]

The exact pathogenesis of this malformation is not known however two theories have been proposed. According to the fission theory, it is thought that monozygotic twin pregnancies occur as a result of delayed aberrant division of a single zygote after 13–15 days of the embryonic period whereas according to fusion theory monozygotic twin pregnancies occur by the secondary fusion of two separate mono-ovulatory embryonic disks.[10] Various drugs including clomiphene citrate, valproate, and griseofulvin used in periconceptual period have been identified as risk factors. No mutations have yet been established as causative factors in conjoined twinning. If a pregnancy is complicated by conjoined twins, there is no evidence of increased recurrence risk in subsequent pregnancies.[11]

Conjoined twins prognosis depends on the fused body parts. Apart from the conjointment, various other congenital anomalies such as congenital heart diseases, gastrointestinal anomalies like imperforate anus, genitourinary, and central nervous system anomalies are known to occur in these cases. Due to such complications, approximately 60% of cases die *in utero* and about 35% are stillborn or die within the first 24 hours after birth.[6] Surgical approach to separate the conjoined twins is a complex and multidisciplinary process and it is often difficult to surgically treat this condition. In most cases, surgery for separation is extremely risky and life-threatening and therefore antenatal imaging is essential to diagnose the condition and to provide required anatomical information if, at all, a surgical division of conjoined twins is planned.[12]

Due to the high risk of perinatal mortality and morbidity, it is of paramount importance to have antenatal detection of conjoined twins in the first trimester. Ultrasonography is a safe, reliable, and quick method for evaluation of conjoined twins. Using ultrasound, conjoined twins can be diagnosed even in early pregnancy (as early as 8–10 weeks).[13] Suspicious ultrasound findings which may point towards the diagnosis include: both fetal heads visualized in same plane, no change in the relative position despite maternal movement and manual manipulation, fetal scoliosis, contiguous skin, unusual limb positions, and more than three vessels in the umbilical

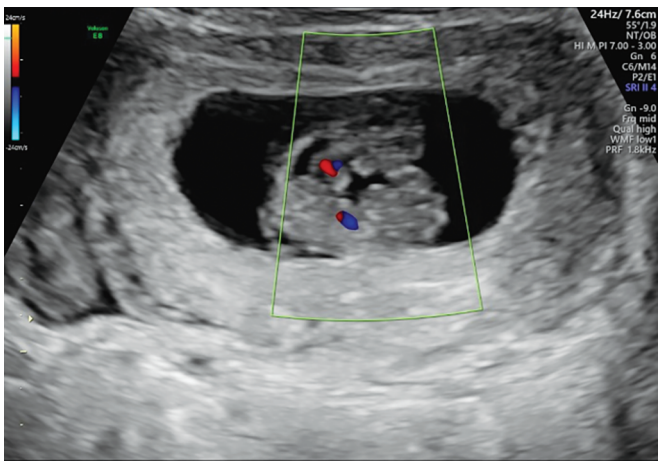

Fig. 1: Two-dimensional gray scale ultrasound image showing a single gestational sac with two fetuses seen lying in a face-to-face position. Two separate fetal heads, chests, and hearts with separate cardiac pulsations are seen. The twins are fused at lower abdomen and pelvis

Fig. 2: Three-dimensional ultrasound image accurately delineates the level of fusion to be at lower abdomen and pelvis

cord.[14] However, there are several pitfalls which result in false-positive sonographic diagnosis of conjoined twins. These include thin dividing membrane which may not be easily visible, close proximity of the fetuses which may make it appear that they are conjoined, and polyhydramnios may be a late finding, and therefore may be absent at the time of initial assessment. Inseparable skin contours are the most important finding which, if present, avoid a false-positive diagnosis. Also, discordant presentation does not completely exclude the diagnosis, particularly in omphalopagus twins.[15]

Other imaging techniques including 3D examination by ultrasound, echocardiography as well as magnetic resonance images, may be necessary to determine associated anomalies, delineate the anatomy, and determine the extent of fusion deformity.[16] Detailed imaging evaluation is necessary for accurate counseling of the parents about prognosis and to prepare for possible postnatal separation surgery.

Early diagnosis of conjoined twins has the following advantages. First, it helps parents to decide whether to choose early termination, which reduces the risk of trauma during vaginal delivery and minimizes maternal mortality and morbidity. Secondly, if the parents decide to continue the pregnancy, it is necessary to do early evaluation of the site and degree of fusion which will help in deciding optimal time and strategy of the treatment.[17] Evaluation of conjoined twin pregnancies should be multidisciplinary. Obstetricians, radiologists, pediatricians, and pediatric surgeons need to be involved to decide what is the best time for the pregnancy to be interrupted and to define what are the chances of postnatal separation. Ethical dilemmas arising in such pregnancies need to be analyzed and discussed with the healthcare team and the family.[18] In our case, antenatal diagnosis of conjoined twins was made with the use of two-dimensional ultrasound. However, using 3D ultrasound imaging with surface rendering significantly improved image clarity of the characteristic features of ischiopagus twins and increased our diagnostic confidence. It also helped the parents to better understand the complex anomalies present in their fetuses and they chose to terminate the pregnancy.

Conclusion

Conjoined twins are an extremely rare and complex occurrence. Since the associated complications lead to high perinatal mortality, making early diagnosis by ultrasound is an essential requirement. It gives an option of safe and early termination of pregnancy to the parents thereby preventing maternal mortality and morbidity.

References

1. Nahar N, Khan N, Islam SM, et al. Conjoined twins with single heart and liver—a case report. Mymensingh Med J 2015;24(1):172–176. PMID: 25725685.
2. Osmanağaoğlu MA, Aran T, Güven S, et al. Thoracopagus conjoined twins: a case report. ISRN Obstet Gynecol 2011:238360. DOI: 10.5402/2011/238360
3. Ernst A, Schlattmann P, Waldfahrer F, et al. Die Behandlung des M. Menière mit Betahistin: Kritische Anmerkungen zur BEMED-Studie. Laryngorhinootologie 2017;96(8):519–521. DOI: 10.1136/fetalneonatal-2012-301809.62
4. Lumme RH, Saarikoski SV. Monoamniotic twin pregnancy. Acta Genet Med Gemellol (Roma) 35(1-2):99–105. DOI: 10.1017/s0001566000006309
5. Zikulnig L, Hecher K, Bregenzer T, et al. Prognostic factors in severe twin-twin transfusion syndrome treated by endoscopic laser surgery. Ultrasound Obstet Gynecol 1999;14(6):380– 387. DOI: 10.1046/j.1469-0705.1999.14060380.x
6. Kavecan I, Obrenovic M, Privrodski JJ, et al. Parasitic Twin Presenting Rudimentary Upper Limbs Causes a Unique Spectrum of Anomalies of Autosite. Balkan Med J 2018;35(6):445–446. DOI: 10.4274/balkanmedj.2018.0781
7. Seo JW, Lee YS, Chi JG. Cross-sectional illustration on major types of conjoined twins. J Korean Med Sci 1988;3(1):19–25. DOI: 10.3346/jkms.1988.3.1.19
8. Chiu CT, Hou SH, Lai HS, et al. Separation of thoracopagus conjoined twins. A case report. J Cardiovasc Surg (Torino) 1994;35(5):459–462. PMID: 7995841.
9. Liu H, Deng C, Hu Q, et al. Conjoined twins in dichorionic diamniotic triplet pregnancy: a report of three cases and literature review. BMC Pregnancy Childbirth 2021;21(1):687. DOI: 10.1186/s12884-021-04165-x
10. Spencer R. Theoretical and analytical embryology of conjoined twins. Part I: embryogenesis. Clin Anat 2000;13(1):36–53. DOI: 10.1002/(SICI)1098-2353(2000)13:1<36::AID-CA5>3.0.CO;2-3
11. Czyz W, Morahan JM, Ebers GC, et al. Genetic, environmental and stochastic factors in monozygotic twin discordance with a focus on epigenetic differences. BMC Med 2012;10:93. DOI: 10.1186/1741-7015-10-93
12. Tannuri AC, Batatinha JA, Velhote MC, et al. Conjoined twins: twenty years' experience at a reference center in Brazil. Clinics (Sao Paulo) 2013;68(3):371–377. DOI: 10.6061/clinics/2013(03)oa14
13. Cuillier F, Dillon KC, Grochal F, et al. Conjoined twins: what ultrasound may add to management. J Prenat Med 2012;6(1):4–6. PMID: 22905303.
14. Alkhateeb M, Mashaqbeh M, Magableh S, et al. Early prenatal diagnosis of thoracopagus twins by ultrasound. Acta Inform Med 2015;23(1):60–2. DOI: 10.5455/aim.2015.23.60-62
15. Pajkrt E, Jauniaux E. First-trimester diagnosis of conjoined twins. Prenat Diagn 2005;25(9):820–826. DOI: 10.1002/pd.1267
16. Brizot ML, Liao AW, Lopes LM, et al. Conjoined twins pregnancies: experience with 36 cases from a single center. Prenat Diagn 2011;31(12):1120–1125. DOI: 10.1002/pd.2843
17. Saw PE, Jiang S. The significance of interdisciplinary integration in academic research and application. BIO Integr 2020;1(1):2–5. DOI: 10.15212/bioi-2020-0005
18. Kirtan K, Doopadapalli D, Shailaja N, et al. An interesting case of undiagnosed conjoined twins. Donald School J Ultrasound Obstet Gynecol 2015;9(3):327–330. DOI: 10.5005/jp-journals-10009-1419