Prenatal ultrasonic diagnosis of conjoint twins

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ABSTRACT

Background: A primipara with multiple pregnancy who booked for antenatal care at 21 weeks presented with vaginal bleeding and pedal oedema at the 27th week. She had had five previous ultrasound scans that gave conflicting results. **Materials and Methods:** The patient was therefore sent for a confirmatory scan, which was performed using a linear 3.5 MHz transducer of a dynamic imaging dedicated, concept MC ultrasound scan machine. **Results:** A diagnosis of conjoint twins was made. The twin was joined from the mandible to the abdomen. Although they cried at birth, they died a few minutes after. The pregnancy was terminated by caesarian section at the patient's request. The patient did well postpartum and was discharged on the 5th postoperative day. Records confirm that this is the first case seen in this hospital, which has been in existence for 18 years. **Conclusion:** To diagnose conjoint twins by ultrasound, one needs the expertise and careful scanning techniques, as the diagnosis may be easily missed, especially if the union is extensive.

Key words: Conjoint twins, CT, MRI, prenatal, ultrasound

DOI: 10.4103/0189-6725.62856

INTRODUCTION

The psychological trauma of carrying a congenitally abnormal baby, especially for a primipara, cannot be overemphasized. Early diagnosis helps to reduce the burden of perinatal morbidity and mortality by providing to the doctor and patient an option of termination or prenatal intervention.[1]

The conjoint twin is a rare and interesting congenital malformation. It is shrouded in mystery and considered a taboo in this part of the world. They are usually born dead.[2]

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We present a case of cephalothoraco-omphalopagus conjoint twins, discussing the challenges of prenatal diagnosis, a brief literature review and a mention of other relevant imaging modalities.

CASE REPORT

A healthy, 29-year-old booked primipara, who attended antenatal clinics in a teaching hospital.

She registered for antenatal care at a gestational age of 21 weeks + 1 day, with a history of vaginal bleeding in early pregnancy and pedal oedema. She had a positive family history of twinning. Past medical history was insignificant.

Routine antenatal laboratory investigations were all normal. However, she had had several conflicting ultrasound scan findings (five preceding ultrasound scan examinations at 12 weeks 4 days, 14 weeks 6 days, 23 weeks 0 days, 26 weeks 5 days and 27 weeks 5 days). The first two scan examinations diagnosed normal twin pregnancy. A diagnosis of hydrops fetalis in a singleton pregnancy was made at the third examination. The fourth examination diagnosed a twin pregnancy with two hearts while the fifth said it was a single foetus with two heads and one heart with pleural effusion, ascites and an enlarged heart. Polyhydramnious was also noted. At the 27th-week gestational age, she experienced a recurrence of vaginal bleeding. She was admitted and referred to the radiology department for a conclusive ultrasound scan because of the discrepancies in the previous scan reports.

Using a 3.5 MHz transducer, the patient was found to be carrying a conjoint twin, presenting breech, with two fully developed heads facing each other [Figure 1], joined at the neck and thorax and sharing a common abdomen [Figure 2].

A large liver, two kidneys and bowel loops were shared by the twin. Two upper and lower limbs were seen for each foetus. Both spines were well developed and normal. Two viable foetal hearts, pleural and pericardial effusion and ascites were part of the findings [Figure 3].



Figure 1: Ultrasound showing two faces, facing each other and joined around the mouth

No separating membrane was seen and there was associated polyhydramnios. Their biparietal diameters and head circumferences were 67 mm ≈27 weeks, 261 mm ≈28 weeks 3 days for the first twin and 68 mm ≈27 weeks 2 days, 254 mm ≈27 weeks 4 days for the second twin, respectively. One of the femur lengths was 50 mm ≈27 weeks 2 days.

The couple was then informed of the ultrasound findings and counseled on the various management options. They insisted on the termination of pregnancy.

The babies were delivered by caesarian section and the operative findings include a fairly well-formed lower segment, polyhydramnious, live female conjoint twins (cephalo thoraco-omphalopagus), each with an Apgar score of 3. They both presented breech, two upper and two lower limbs each, with two heads and a single large umbilical cord and placenta. They both cried immediately after delivery but died a few minutes after. The maternal ovaries and fallopian tubes were normal.



Figure 3: Ultrasound showing two spines, pericardial effusion and ascites



Figure 2: Picture of the delivered conjoint twin

A plain X-ray of the foetuses showed a conjoint twin joining at the mandible, neck and thorax, ventrally [Figure 4].

The head and spines of both foetuses were normal. One abdomen and a single large umbilical cord were seen.

Postoperatively, the patient's condition was satisfactory and she was discharged on the 5th day.

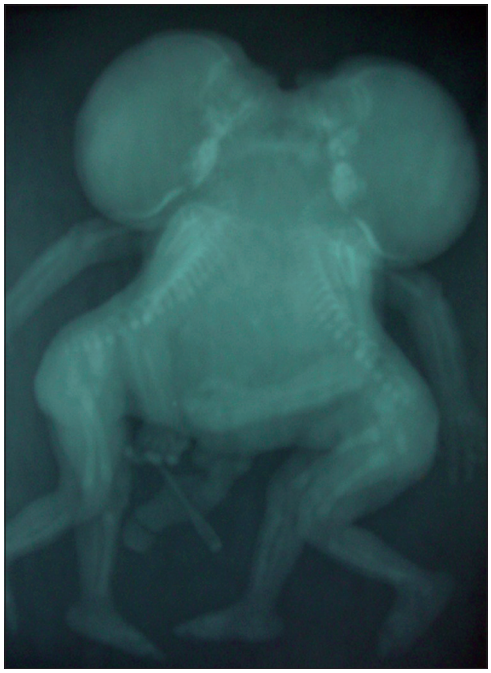


Figure 4: X-ray showing two foetal heads, extremely flexed backwards and normal spines

DISCUSSION

A conjoint twin is a very rare condition, with a reported frequency of 0.1–0.35:10,000 births (1 in 50,000, 1 in 100,000), and hardly any risk of recurrence.[2-5] Its incidence is higher in Asia and Africa, accounting for 1–2% of monozygotic twins.[2,3] It should be suspected in all monochorionic, monoamniotic, monozygotic twin pregnancies.[2,6] It is more common in females, with a ratio of 3:1.[3] Most of them are born prematurely, 40% are stillborn and 35% die within 24 h, as occurred in this case.[3,4,7] The overall survival rate is 25%.[3]

It has been attributed to incomplete division of the embryonic disc after the 13th day of conception, leading to failure of complete separation of both foetuses.[2,4,8] The precise cause of conjoint twinning is not known, but two contradictory theories are used to explain its origin; "fission", where the fertilised egg splits partially and "fusion", in which a fertilised egg completely separates but the stem cells that search for similar cells find like stem cells on the other twin and fuse the twins together.[2,3,8]

Common associated anomalies include cardiac malformations, congenital diaphragmatic hernia, abdominal wall defects and imperforate anus. Conjoint twins should be suspected in a gravid mother with multiple gestations and an abnormal foetal attitude.[9]

Preoperative imaging is helpful in determining the feasibility of separation.[10] Early prenatal diagnosis and precise characterisation of conjoint twins are therefore essential for optimal obstetric intervention and postnatal management.[2] Advances in prenatal diagnosis of conjoint twins broaden the scope of available management options.

The classification of conjoint twin depends on the site of union and the suffix "pagus" is used, meaning fastened.[6] They could be joined at the thorax (thoracopagus), abdomen (omphalopagus), chest and abdomen (thoraco-omphalopagus), which is one of the most common types, and the iliac bone (ileopagus).[2,6,8]

When the twins are extensively connected, as in our patient, they are then named by their duplicated part, e.g. two heads with one body (dicephalus), one body with four arms (dicephalus tetrabrachius),[3] xipho-omphalopagus,[5] cephalo-thoracopagus (joined at the head and chest), dicephalus parapagus (single trunk and two heads),[3,4] craniopagus (joined at the head), rachipagus (dorsal union of the head and trunk)[2] and pyopagus (joined at the back)[3] and parapagus (share a conjoint pelvis, one symphysis pubis and one or two sacrum).[4] When the union is limited to the abdomen and pelvis and does not involve the thorax, it is called dithoracic parapagus.[4] A single head with two faces and a trunk is called prosopic parapagus.[4]

With extreme fusion, a conjoint twin can be mistaken for a singleton pregnancy,[4] as occurred in earlier scans in the patient presented, although this might also be due to the sonographer's expertise.

Organ union may also be seen. Cardiac defects are the most common association (20–30% cases).[2] The heart is often conjoint, unlike this case where two foetal hearts were seen. Prognosis, obstetric management and treatment planning are determined by the degree of fusion and the extent of foetal organ union.[2]

A conjoint twin is often a mirror image of its partner, especially in the dicephalus twins.[2]

Polyhydramnious was seen in 50–75% of the cases,[4] as was confirmed in this case.

Antenatal diagnosis by ultrasound is by identification of any of the following classical signs that may suggest diagnosis: both foetal heads in the same plane, unusual backward flexion of the cervical spine, no change in the relative position after maternal movement and manual manipulation, inability to separate foetal bodies even with careful scanning techniques and observation, lack of separating membrane, presence of foetal anomalies and identification of more than three vessels in a single umbilical cord.[2,8] Almost all these findings were seen in this patient. All sonographers must be familiar with signs associated with conjoint twins and careful ultrasound assessment should be made to identify shared organs.

Current imaging technology provides a basis for early diagnosis and, therefore, a better prognosis. In a retrospective study of 14 sets of conjoint twins by Mackenzie *et al.*, the earliest age of diagnosis was 9 weeks and prenatal imaging with ultrasound, echocardiography and ultrafast foetal magnetic resonance imaging (MRI) accurately defined the shared organs in all cases.[9]

Diagnosis is possible using ultrasound scan in the first trimester, but details are better seen in the second trimester.[7] As was found by Quiroz *et al.*, the

ultrasound findings in this patient included the inability to separate the foetal bodies ventrally and the face to face position.[6] Ultrasound, which plays a crucial role not only in diagnosis but also in establishing the degree of conjoining, is noninvasive and nonionising.

MRI is complementary to ultrasound and provides additional anatomically precise clinical data. **[10]** It is noninvasive and has a large diagnostic window, allowing total foetal imaging with excellent resolution of tissue composition.[10] Unlike a computed tomography scan, it does not use ionising radiation, although it is very expensive and lacks real-time imaging capabilities.[10]

Separation of the conjoint twin is a complicated procedure requiring a multidisciplinary approach and the prognosis is usually predetermined by the underlying anatomy.[2]

Overall prognosis however depends on the degree of organ sharing.[8] Because of the increased morbidity and mortality of vaginal delivery in monoamniotic twins, even if not conjoint, elective caesarian section at term will ensure the best chance for survival.[6]

Ultrasound diagnosis of conjoint twins requires the expertise and careful scanning techniques available at a second-level abnormality screen, which should always be employed for multiple gestations.

REFERENCES

1. Akinola RA, Akinola OI, Disu E, Jinadu FO, Balogun BO, Akintomide TE, *et al*. Audit of congenital fetal anomalies as seen on ultrasound in a teaching hospital in Ikeja, Lagos, Nigeria. Nig Jr of Hel and Bio Sci 2008;7:61-5.
2. Sethi SK, Solanki RS, Hemal U. Conjoined twins: A case report. Ind J Radiol Imag 2004;14:67-9.
3. Conjoined twins From Wikipedia, the free encyclopedia. Retrieved from "http/en.wikipedia.org/wiki/Conjoined twins" 2007.
4. Kharat A, Karamchandani A, Singh A, Shetty SS. Antenatal ultrasound appearance of dithoracic parapagus conjoint twins. Ind J Rad Img 2009;14:159-60.
5. Shi CR, Cai W, Jin HM, Chen F, Zhou Y, Zhou DX. Surgical management to conjoined twins in Shanghai area. Pediatr Surg Int 2006;22:791-5.
6. Quiroz VH, Sepúlveda WH, Mercado M, Bermúdez R, Fernández R, Varela J. Prenatal ultrasonographic diagnosis of thoracopagus conjoined twins. J Perinat Med 1989;147:297-303.
7. Dr Joe Anthony. Ultrasound images of conjoint twins. I discuss sonographic images of conjoined or Siamese twin. Available from http://drjoea.googlepages.com/obstetric-2.
8. Grutter F, Marguerat P, Maillard-Brignon C, De Grandi P, Pescia G. Thoracopagus fetus. Ultrasonic diagnosis at 16 weeks. J Gynecol Obstet Biol Reprod (Paris) 1989;18:355-9.
9. Mackenzie TC, Crombleholme TM, Johnson MP, Schnaufer L, Flake AW, Hedrick HL, *et al*. The natural history of prenatally diagnosed conjoined twins. J Pediatr Surg 2002;37:303-9.
10. Turner RJ, Hankins GD, Weinreb JC, Ziaya PR, Davis TN, Lowe TW, *et al*. Magnetic resonance imaging and ultrasonography in the antenatal evaluation of conjoined twins. Am J Obstet Gynecol 1986;155:645-9.

Source of Support: Nil, **Conflict of Interest:** None.