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Diecephalic thoraco-omphalopagus conjoined twins: case report

Mbuga MN, Odawa FXO, Maranga ISO

Department of Obstetrics and Gynaecology, University of Nairobi

Correspondence to: Dr. M.N. Mbuga. Email: nyakimbuga@gmail.com

Abstract

We present a case report of conjoined twins discovered at 17 weeks gestation on routine trans-abdominal ultrasound in a 27 year old woman. The twins were joined at the chest and upper abdomen. They shared a heart, liver, sternum and diaphragm. The patient opted for termination of the pregnancy which was performed by cervical ripening after which she had a successful vaginal delivery. The importance of early ultrasound scans by an expert is emphasized.

Key words: Conjoined, Thoraco-omphalopagus, Kenya, Monozygotic, Gestation

Introduction

Conjoined twins are popularly known as Siamese twins (1). The incidence ranges from 1:50,000 to 100,000 live births (2). They are more common in females with a female to male ratio of 3:1. It is thought that the XX genotype tends to survive better than the XY (3). Conjoined twins are invariably monozygotic although usually they will be different individuals with different personalities sharing similar body parts. Due to its rarity it still poses a challenge in management due to poor prognosis and fatal outcomes as well as limited human resource expertise especially in the developing countries. It's however unfortunate that most of the cases will be missed prenatally due to limited access to obstetric ultrasound in the country. Several such unpleasant surprises have been referred to our facility either in labour or in early neonatal life without prior planning. This in turn poses difficult management and ethical dilemmas in decision making. We present a case of conjoined twins identified antenatal to emphasise the importance of routine anomaly scan as part of comprehensive prenatal care.

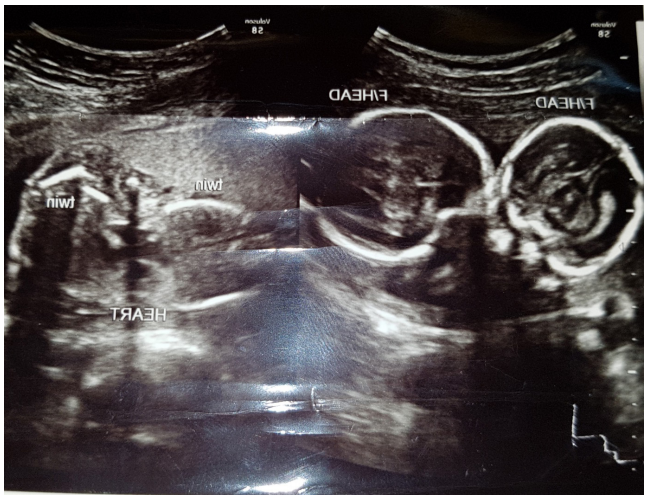
Clinical presentation

A 27 year old in her second pregnancy, was referred to us from a local District hospital with a diagnosis of conjoined twins discovered on routine ultrasound at 17 weeks.

The woman was otherwise well. She was normotensive. Her antenatal profile was unremarkable. The index pregnancy through natural conception with no prior use of any fertility treatment. Her previous pregnancy was uneventful and she had had a spontaneous vaginal delivery to a live male infant whose weight was 3600gms. She was in good general condition with normal vital signs. No pallor, no icterus. The symphysio-fundal height was of 28cm and multiple foetal parts that could not be well defined were felt.

We reviewed the earlier obstetric ultrasound done. It showed twins conjoined at the thorax and with a shared heart. The twins had ill-defined abdominal and thoracic structures. Different foetal spines and heads were discerned with each twin having their own pair of limbs that were observed to be normal. They shared a single umbilical cord. A diagnosis of a diecephalic thoraco- omphalopagus conjoined twins was made (Figure 1).

Figure 1: 2D ultrasound image showing conjoined twins sharing a heart



The prognosis of the twins was discussed with the patient and she opted for immediate termination of the pregnancy. This was done using combined 100 mcg of misoprostol given vaginally and extra-amniotic Foley's catheter with saline as per the unit termination protocol.

She delivered two female infants that died within minutes. They had a combined weight of 590 grams. They were joined at the chest up to the level of the umbilicus. An autopsy was performed. It further revealed that the babies shared sternum,

heart, diaphragm, liver and umbilical cord which had one vein and 4 arteries. All other abdominal and reproductive organs appeared grossly normal and had normal development. The placenta weighed 281 grams (Figures 2-4).

Figure 2: Diecephalic thoraco- omphalopagus conjoined female twins

Figure 3: Shared sternum, liver, heart

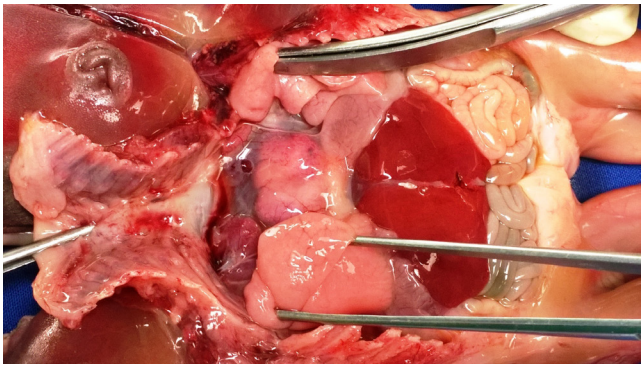
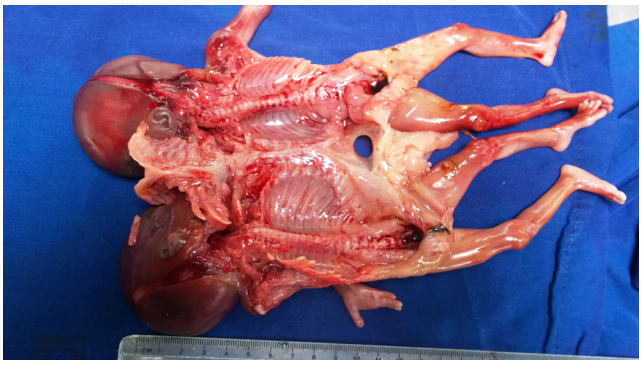


Figure 4: Post organ removal image showing the bony structure



Discussion

Conjoined twins are classified based on their site of union namely; ventral which is the most common at 87% and dorsal types at 13% (1–3). The suffix "pagus" is used meaning fixed to demonstrate the area of fusion. The ventral types are; thoracopagus (joined at the thorax), omphalopagus (joined at the anterior abdominal wall), ischiopagus (joined at the buttocks), parapagus (joined at the pelvis and variable trunk) and cephalopagus (joined at the head and neck). The commonest ventral types are thoracopagus and omphalopagus. These commonly occur together as thoraco – omphalopagus which constitutes 70% of all conjoined twins (2).

The dorsal types are; craniopagus (joined at the cranium), rachiopagus (joined at the vertebral column) and pyopagus (joined at the sacrum) (1–3).

The aetiology is unknown. However, there are theories that have been addressed and these include; **fission theory**; entails late separation of the developing embryo beyond day 13 after fertilization and the **fusion theory;** a fertilized egg separates completely but during development, alike stem cells find each other, fuse and develop as one unit (1–3).

Diagnosis can be made as early as 12 weeks by ultrasound (1). The features detected are anatomical structures that are not separate, lack of a separating membrane and foetal heads that are fixed in position. Severity and type of abnormality can further be investigated by 3D ultrasound, CT-Scan or MRI (1,4).

Management is either termination of the pregnancy or expectant. Generally, most conjoined twins are incompatible with life (2). Termination of pregnancy can be recommended if there is fusion of sensitive organs such as the brain-cerebral region and the heart as they are associated with a poor prognosis and separation is rarely successful (2,3). If severe deformities are anticipated post an attempt to separate the twin's then termination of the pregnancy is recommended (1). Vaginal delivery after induction of labour and cervical ripening can be attempted before 24 weeks of gestation. Reason being the largest diameter of the twins has not been shown to be an impediment to birth. Beyond 25 weeks of gestation vaginal delivery has been associated with a risk of dystocia and uterine rupture hence caesarean deliveries are preferred (5).

Expectant management can be done with the mode of delivery being an elective caesarean delivery after 32 weeks following antenatal corticosteroids (6). Surgical separation post-delivery will depend on the severity of the abnormality and if the life of the twins is at risk (5).

Conclusion

Conjoined twins occur rarely and there are no known preventive measures. They pose a challenge in developing countries due to late commencement of antenatal visits and few early ultrasound scans. Professional expertise to deal with live born conjoined twins is also inadequate. Emphasis is therefore made on the need for early diagnosis by use of ultrasound and MRI to better characterize the deformity. Termination of pregnancy through the vaginal route can be done at gestations less than 24 weeks. Caesarean deliveries can be done if separation of the foetuses is possible and viability considered possible.

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Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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