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Conjoined Thoracopagus Siames Twins : A Case Report from the Casablanca University Hospital

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Abstract

Conjoined twins are a particularly rare condition, requiring perfect understanding of their anatomy and multidisciplinary intervention to free them from their attachments. Thoracopagus twins are connected by the chest and umbilicus and are the most common of all variants, but have a low survival rate. We report the case of a 18-year-old female patient, pregnant at 12 weeks amenorrhoea, who consulted in the maternity emergency department with a clinical picture of a miscarriage in the process of expulsion of a conjoined thoracopagus siames twins.

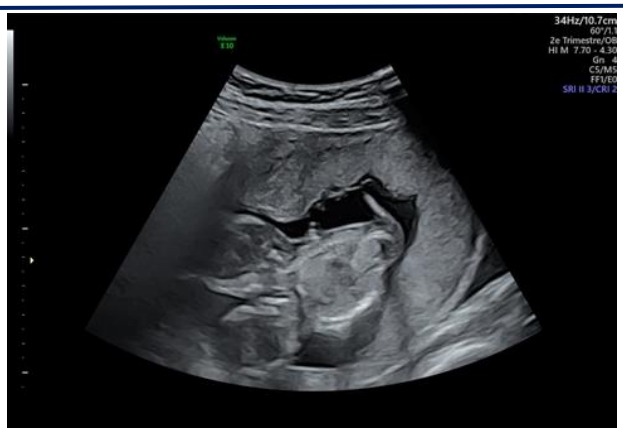
Keywords: Conjoined Twins, Thoracopagus, Siames Twins, Case Report.

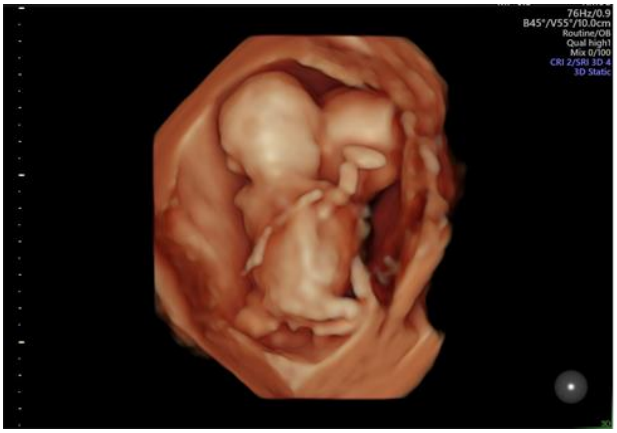
Introduction

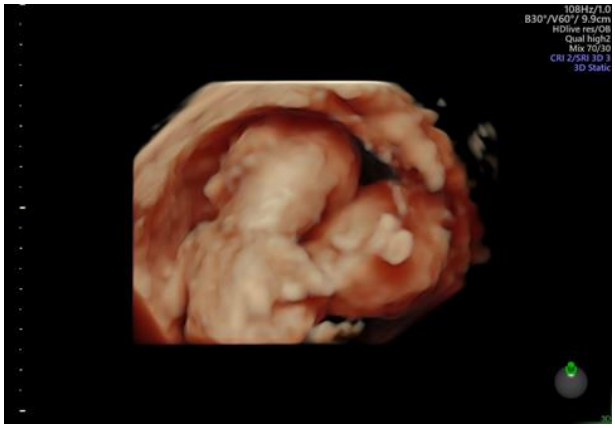
Siamese twins are one of the rarest forms of twin pregnancy, and are homozygotic twins **[1]**. They occur in 1/50000 to 1/100000 births, with a female predominance of approximately 70% **[2]**. Early detection relies on obstetrical ultrasound, so therapeutic termination of pregnancy may be indicated. The prognosis depends essentially on the location, type and extension of common organs, as well as the association with other malformations **[2]**. Based on this observation and a brief discussion, we will review this rare pathology and the importance of early ultrasound diagnosis for a better therapeutic and decision-making approach.

Case report:

The patient was Mrs A.B, 18 years old, primiparous, a chronic smoker and Cannabis user, with no hereditary diseases. The father was 24 years old, with no consanguinity, and the pregnancy was followed in a health center by a general practitioner. She attended a prenatal consultation where an ultrasound scan was performed, showing a pregnancy with thoracophagous conjoined twins. A first-trimester morphological ultrasound was indicated, concluding in a monochorionic, monoamniotic twin pregnancy of 12 - 13 SA with thoracophagous conjoined fetuses, two distinct cephalic poles joined by the thorax, the thorax is common with a single cardiac structure, the heart has four balanced chambers **(Figures 1,2,3)**. The patient consulted the maternity emergency department with a clinical picture of miscarriage in the process of expulsion, and a fetopathological study was carried out.







Figures (1,2,3): Morphological ultrasound of a pregnancy with thoracophagic conjoined twins, with a single cardiac structure.

Discussion:

Conjoined twins are popularly known as Siamese twins, named after the birthplace of the original Siamese twins born in 1811 in Thailand **[3]**. The first case reported in the literature was that of sisters Mary and Eliza Chulkhurst , They were born in 1100, in Kent, England, and were joined at the hips and shoulders **[1,3]**. Nevertheless, the most famous Siamese twins are Chang and Eng Bunker, born in Siam (now Thailand) in 1811 and died in 1874 at the age of 63. They were xiphopaguses united at the lower part of the thorax. The first ultrasound diagnosis of conjoined twins was made in a 35-week pregnancy in 1976 **[4]**. Nowadays, diagnoses are made earlier and earlier, even at seven weeks **[5]**. The first successful operation to separate a pair of Siamese twins was performed in Switzerland in the 17th century, when the twins were joined at the umbilicus **[1]**. The last separation operation took place in Lyon, France, on November 13, 2019, and lasted seven hours. The Siamese twins were one year old **[6]**.

Monozygotic twin pregnancies result from early division of the same egg. Bichorionic and biamniotic twins are described (20-30%) when division occurs within three days of fertilization. Monochorionic and biamniotic twins (70-80%), when division occurs between days 4 and 8. When embryonic

cleavage occurs between days 9 and 13: the cells of the lineage destined to form the chorion and amnion are differentiated, and any cleavage of the egg from this point onwards will result in a monoamniotic, monochorionic twin pregnancy. These are the pregnancies with the greatest risk of complications. From the fourteenth day after fertilization, late division will result in conjoined twins. This phenomenon is extremely rare. However, the etiopathogenesis of conjoined twins is poorly understood. There is no associated chromosomal abnormality. Race, heredity, parity and consanguinity are not involved in the process. Two opposing theories have been proposed to explain this phenomenon. On the one hand, the theory of incomplete and late fusion of a single embryo (split theory) is the most widely accepted **[3]**.

Classifications have been described according to site of fusion, common organs. According to Spencer's classification, we find dorsal junctions (13%) from the neural tube or ventral (87%) from the anterior line. This results in eight types of complete fusion: cephalopagus, thoracopagus, omphalopagus, ischiopagus, parapagus, craniopagus, pygopagus and rachipagus twins **[1,3,4]**. To this classification we can add duplications and rare forms: diprosopes, dicephales, dipygus, parasitic twins and fetus in fetus **[5]**.

Early diagnosis of the conjoined twin is based on ultrasound. This is possible in the first trimester, between 10 and 13 weeks' amenorrhea (SA) **[2,4]**. Some incomplete forms may be difficult to diagnose, and an ultrasound scan performed at 22 weeks' amenorrhea helps to elucidate these forms, locate the attachment zone more precisely, and look for associated malformations, particularly cardiac anomalies **[4]**. Magnetic resonance imaging of the uterine contents, 3D imaging, is useful for diagnosis and provides good mapping of lesions for a better prognostic approach **[3]**.

In most cases, the ultrasound diagnosis of Siamese twins indicates therapeutic termination of pregnancy **[7]**. For undetected conjoined twins, three attitudes are possible, depending on the prognosis: immediate surgical separation, delayed surgical separation or no separation at all. When separation is indicated after multidisciplinary discussion, surgical intervention should be performed between 6 and 12 months of age, to ensure better adaptation of the conjoined twins **[7]**.

A similar complex surgical separation of a pair of Siamese twins connected from chest to pelvis was successfully performed by a group of surgeons at Texas Children's Hospital in Houston in 2015 using a 3D-printed model. The prognosis for conjoined twins remains very guarded in developing countries. Their survival depends on shared organs and other associated anomalies **[4]**. Surgical intervention to separate Siamese twins can be very complex as a result. Neonatal mortality of conjoined twins is generally high **[8]**.

Conclusion:

Conjoined twins are an extremely rare congenital anomaly. Ante-natal diagnosis relies on ultrasound, which must be carried out by experienced obstetricians, to determine the anatomical structures of the conjoined twins, carry out a malformative assessment, deliver the twins in a third-level maternity hospital and pre-establish appropriate neonatal care. Successful separation of Siamese twins requires the combined efforts of a multidisciplinary team for the best possible care.

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