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The Twin Trouble and A Shared Conundrum- A Case Report on Urinary Malformation in Siamese Twins

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Abstract

Aim: To demonstrate how timely antenatal registration and regular follow up helps in better maternal and fetal care and outcomes.

Background: Conjoined twin gestation, an extremely rare entity, carries a high obstetric risk. Early detection and timely management are imminent to reduce maternal and fetal morbidity and mortality.

Case description: Here, we discuss a 30 year old, Gravida 2 Parity 1 Living 1, at 21 weeks of gestation, referred to us with an abnormal malformation scan, only to reveal a conjoined twin gestation post medical termination of pregnancy.

Conclusion: Early detection of non viable anomalies and its timely management lead to better maternal outcomes and prevents poor progeny.

Clinical significance: Emphasis on early antenatal registration and regular follow up even in seemingly low risk pregnancies.

Keywords: Conjoined Twins, Malformation Scan, Urogenital Malformation, Medical Abortion.

Case Description

A 30 year old, gravida 2 parity 1 living 1, at 21 weeks of gestation was referred to our tertiary health care centre with an abnormal malformation scan. She presented with no active complaints and past history revealed no medical or surgical high risk factors. The marriage was non consanguineous, pregnancy was conceived naturally with no family history of twinning. On examination, she was vitally stable with abdomen corresponding to the gestational age and regular fetal heart sounds. Routine blood investigations were found to be within normal limits. Targeted imaging for fetal anomalies revealed

features suggestive of body stalk anomaly with infraumbilical abdominal wall defect with hypotelorism, corpus callosal agenesis, closed neural tube defect, kyphoscoliosis and possibility of bladder exstrophy. The couple opted for medical termination of pregnancy in view of substantial risk that if the child was born, it would suffer from such physical or mental abnormalities so as to be seriously handicapped. The process was initiated using tablet Mifepristone 200mg orally which was followed 48 hours later by 3 doses of tablet Misoprostol 400mcg 4 hours apart. Products aborted and on examination revealed conjoined twins with craniophagus and thoracophagus with bladder exstrophy (Fig 1). Post abortal scan of the products confirmed the clinical findings with sharing of the brain and heart between the two twins.

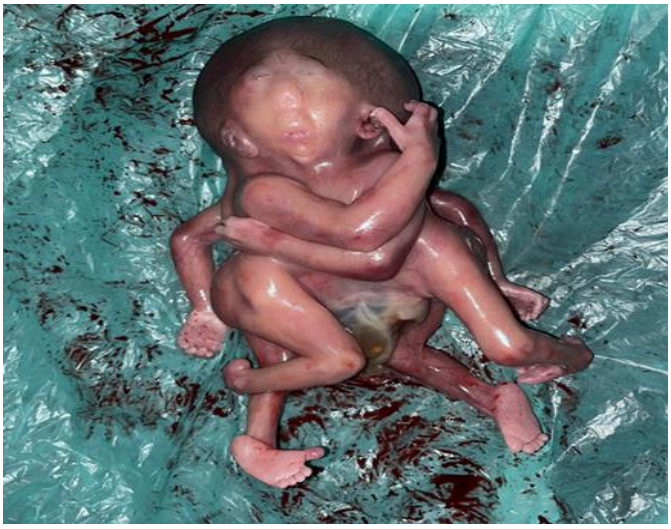

Figure 1: Conjoined twins with craniophagus and thoracophagus with bladder exstrophy

Discussion

Conjoined twins occur in approximately one of every 50,000 to 200,000 births, predominantly females, with about 60% of conjoined twins being stillborn. The occurrence of conjoined twins has shown association neither with increasing maternal age, nor with any genetic, demographic, or environmental factors. The genesis of conjoined twins still remains an unsolved puzzle. While most of the available literature cite it as partial fission or secondary fusion, definite answers on its plausibility remain largely absent. Classically, they are believed to arise when the monozygote splits after 2 weeks of conception, hence having the same sex, a single placenta, a single amnion and a single chorion. The extent of splitting determines the type of the conjoined twins (Table 1), the most common being thoracophagus. The occurance of an abnormally large cloacal membrane leads to improper migration of mesenchymal tissue medially causing ventral wall defects, like omphalocele (Fig 2) and some rarer anomalies like bladder exstrophy, as seen in this case. While most conjoined twins who are born alive do not make it beyond a week or two, some very rare ones like the famous Siamese twins live to lead a prosperous life. This is mainly determined by the organs and the kind of vasculature the twins share. If and when these twins live, their treatment and surgical correction requires an extremely skilful multidisciplinary team, most often involving the ethics committee. When medically managed, shared circulation between the twins affecting the pharmacokinetics of the drugs must be taken into consideration. With the ever-advancing medical care, incorporating computer aided designs and modelling with three dimensional printers, doctors were successful in separating craniophagus twins at 10 months of age. Numerous other successful separation stories including but not limited to omphalopagus and pyopagus have been reported from all over the world, however the general prognosis of conjoined twins remains poor with scope for improvement in imaging, surgical techniques, use of tissue expanders and potential cadaveric transplantation. In this particular case, the twins structurally shared a brain, a heart and had 3 lungs apart from an omphalocele, bladder exstrophy, Corpus

callosum agenesis among other findings which made them incompatible with life. Timely intervention resulted in an uneventful vaginal abortion hence alleviating the need for hysterotomy.

Table 1: Types of conjoined twins

Type	Description
Thoracopagus	Fusion from the upper chest to the lower chest, often share a heart and may share a part of the digestive system
Omphalopagus	Fusion at the lower abdomen. Do not share a heart, but often share a liver, digestive system, diaphragm and other organs
Pygopagus	Fusion back-to-back at the base of the spine and buttocks. Some share the lower gastrointestinal tract, genital and urinary organs
Ischiopagus	Fusion at the pelvis, either face-to-face or end-to-end. Often share the lower digestive tract, liver, and genital and urinary organs. Each twin may have two legs or they may share two or three legs
Parapagus	Fusion side-by-side at the pelvis and part or all of the abdomen and chest, but have separate heads Can have multiple limbs
Craniopagus	Heads are joined at the back, front or side, but not the face or base of the skull
Rachipagus	Fusion at the dorsal aspect with face away from each other, may result in sharing of vertebral column and spinal cord

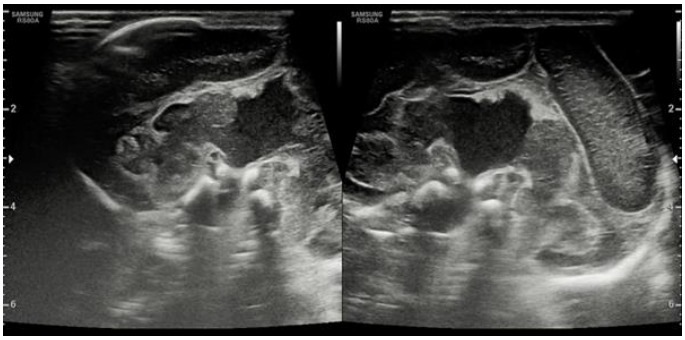

Figure 2: Omphalocele

Conclusion

Early detection of non-viable anomalies and its timely management lead to better maternal outcomes and prevents poor progeny. It is important to emphasise on early antenatal registration and regular follow up even in seemingly low risk pregnancies.

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