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THORACOPAGUS CONJOINED TWIN WITH PATHOLOGICAL AUTOPSY FINDINGS -A RARE CASE REPORT

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ABSTRACT
Conjoined twins is an unique anomaly of multiple gestation and has been a fascinated entity both for general population and medical fraternity. It is seen in 10.25 per million births. 75% of them occur in females. These are associated with severe mortality and morbidity. Thoracopagus is a common type where fusion starts at chest and also involves the heart. Autopsy in case of conjoined twins plays a vital role in deciding the type of fusion of the body and also the status of heart, the great vessels and liver etc. which they share. It thus helps couples to plan future pregnancies and avoids the fear associated with this rare anomaly as there is no chance of recurrence in subsequent pregnancies. We discuss a case of conjoined twin (thoracopagus) because of its rarity, with documentation of incidence and type with review of literature.

KEYWORDS: Autopsy, Conjoint twin, Thoracopagus.

INTRODUCTION
Conjoined twins represent one of the rarest forms of twin gestation accounting 1 in 100 sets of monozygotic twins, 1 in 50,000 gestations or 1 in 250,000 live births.[1] Because this situation carries high risk, early diagnosis and management of pregnancies is extremely important. Conjoined twin, a rare anomaly refers to an incomplete splitting of monozygotic twins after 12 days of embryogenesis. 75% of them occur in females. They are classified according to the point of union; the label used is the Greek word pagus, which means "that which is fixed." Depending upon the site of fusion or non-fusion types of twins may differ, either grossly separable (duplica completa), or grossly inseparable (duplica incompleta). The most common varieties encountered were thoracoomphalopagus (28%), thoracopagus (18.5%), omphalopagus (10%), parasitic twins (10%) and craniopagus (6%).[2] We present the autopsy findings in a case of thoracopagus who were females.

CASE REPORT
A 26 year old female, G2P1L1 with 28 weeks of pregnancy presented in labour with complaints of decreased fetal movements since two days to Obstetrics & Gynecology outdoor of our hospital. She was an unbooked case and had no previous antenatal check ups or any reports of ultrasonography done in the past. Per vaginum examination revealed cervix was effaced and fully dilated and head being palpated. All routine haematological investigations were within normal limits. Ultrasonography confirmed it to be a twin pregnancy. She delivered two still born, joined female fetuses vaginally. After taking due consent from parents the fetuses were sent to our department for detailed pathological autopsy. External examination revealed that the twin was fused from thorax till lumbar and sacrococcygeal region together weighing 2.1 kg (Fig 1). Hair, ear cartilage and nails were present in both the babies. However anus and nipples were absent. Placenta was single weighing 350 gms with single umbilical cord having two arteries and one vein. Anthropometry measurements revealed head circumference-24cms (first fetus tied with gauze piece), 24.5cm of (second fetus), chest circumference was 22.5cms. Crown heel length and crown rump length couldn't be taken. Foot length of each foot of both the fetuses was 4.5 cms. Internal examination revealed single heart, single enlarged liver measuring 44.6gms and two lungs each measuring 4.7gm (Fig 2). Single heart with bifid apex measuring 10.9gms (Fig 3). There was normal gastrointestinal tract but imperforate anus.(Fig 4) Histopathological examination didn't reveal any abnormality. So a final autopsy diagnosis of thoracopagus with single heart and single liver was given.

Fig 1: Showing fetuses of conjoint twin joined at thorax.

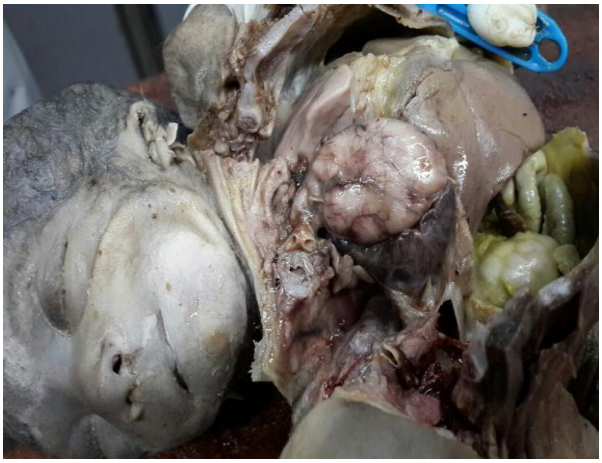

Fig 2: Gross photograph showing abdominal cavity with single heart, enlarged liver & intestine.

Fig 3 (a): Showing heart with bifid apex, (b) weight of heart measured as 10.9 gms.

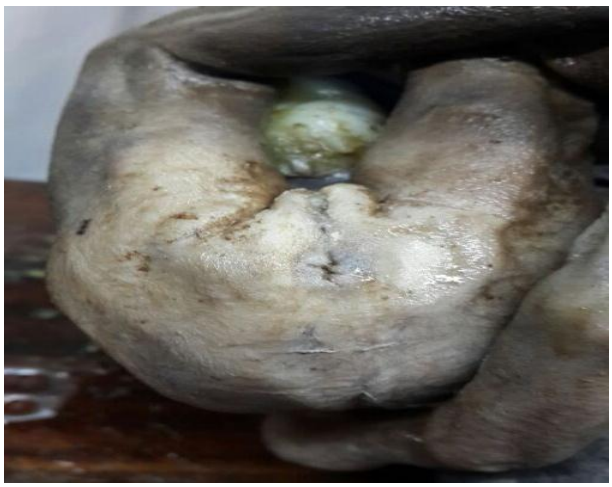

Fig 4: Photograph of conjoint twin showing absence of anus.

DISCUSSION

Conjoined twins also referred as Siamese twins were first reported after the birth of famous conjoined twins Chang and Eng Bunker in 1811 in Siam (now Thailand). They were xiphopagus joined at lower chest by a narrow flesh band that connected their lives and remain unseparated for 63 years due to lack of medical facilities. These twins are monozygous (have same sex) and majority of cases have a monochorionic and monoamniotic placenta. Various theories have been proposed to explain the etiology of conjoined twins. Collision theory is that, two previously duplicate embryonic axes fuse before tissue differentiation and fission theory is the one in which the embryonic tissue divides incompletely remaining fused at some points (or) point.[3] The latter theory is more acceptable. At about 2nd week of normal twining process, i.e the blastula stage, the inner cell mass splits into two separate and equal halves each usually producing a single individual. Conjoined twins result from an arrest in division of the inner cell mass and the non separated parts of otherwise normal twins remain fused throughout the development.[4]

Conjoined twins are classified depending on the site of conjunction. Thoracopagous accounting for 40% of cases is the most common showing fusion at the level of the chest with common sternum, thoracic cage, diaphragm and abdominal walls down to umbilicus. Others are Omphalopagous or Xiphipagous (34%), Pyopagous

(18%), Ischiopagous (6%), Craniopagous (1-2%) and Heteropagous (rare).[5]

Apart from junction at one anatomical site, conjoined twins can result from joining of multiple sites like in craniothoracopagus, xiphoomphaloischiopagus. Prenatal diagnosis of conjoined twins is essential for appropriate obstetric management . Diagnosis of conjoined twins can be made based on different clinical, ultrasound, and MRI criteria. Once diagnosed it becomes essential to check the chorionicity and amnionicity. For management of conjoint twins, cesarean section is recommended in most of the third trimester deliveries however vaginal delivery is reserved for still births and for forms of conjoined twins that are incompatible with life. Emergency separation is required in certain cases of conjoined twins with the death of one twin as was pbserved by Woldeyes WS for the survival of one fetus.[6]

CONCLUSION

Conjoined twins are generally incompatible with life i.e 39% of conjoined twins are still born, and 34% die within first day of life. Thus the overall survival rate for conjoined twins is approximately 25% who are considered for surgical separation. Surgery to separate conjoined twins is extremely risky and life-threatening and pose a surgical challenge for the surgeons as it depends on the point of attachment and the internal parts that are being shared by the twin. Prognosis is usually better if separation is delayed till the infants are 6–12 months of age. Timely diagnosis is essential as one can get sufficient time to counsel the parents whether to continue with the pregnancy and proper intervention can be planned. On the other hand, undiagnosed cases may be first recognized in labour room or after they have caused some labor-related complications. So regular antenatal check ups and prenatal assessment of the fetus by USG as and whenever required during pregnancy can diagnose such anomalies and prevent complications associated with this entity. Perinatal autopsy in such cases can help in assessing the site of junction and type of the conjoint twin which can help in documentation, guide management in subsequent cases and counsel parents about future family planning.

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