Case report

Dicephalus parapagus tribrachius - A rare variety of conjoined twins: A Case Report

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Introduction

Due to abrupt embryogenesis, conjoined twins are born as an unexpected occurrence. One in 200,000 live births actually result in conjoined twins.1 There are two widely accepted theories for how conjoined twins develop: the fission theory, which describes the incomplete fission of a single embryonic disc on 13 to 15 days after ovulation, and the fusion theory, which describes the secondary fusion of two distinct monovular embryonic discs. Conjoined twinning is no longer thought to be based on fusion theory.1-4

Conjoined twins, commonly referred to as Siamese twins after Chang and Eng Bunker of Siam (Thailand), are a rare congenital condition that affects 1/50,000 to 1/100,000 pregnancies.5,6 Due to significant morbidity and mortality that are associated with such conjoined twins as well as the associated ethical, moral, and legal challenges, caring for and treating twins can be extremely difficult for both parents and medical professionals.7 Parapagus (anterolaterally fused) or dicephalus (two-headed) type conjoined twins typically have a poor prognosis because of their shared thoraco-abdominal and spinous anatomy and extremely rare occurrence.8-10 These dicephalic twins share a set of reproductive organs as well as a body that extends from the neck or upper thorax below. We outline the findings of the tribrachial dicephalus parapagus twins' autopsy.

Case Report

In Southern India, an adolescent primigravida, age 19 years, with no congenital defects or twins in the family, and no significant past medical history, delivered at 36 weeks of gestation, a male conjoined twin. There was no history of teratogenic agent exposure during pregnancy, and before to admission, she had not undergone any antenatal checkup or had an ultrasonography evaluation. Her husband aged 59 years and the age gap between the couple being 40 years has to be noted. Live conjoined twins with two heads, two necks, three arms (two normal arms and one arm at the top of the torso fused with ten fingers), and two legs coming from one trunk were the result of an emergency caesarean delivered via caesarean section at SNR district hospital Kolar. The various anomalies of the thoracic, abdominal cavity and central nervous system are illustrated.

Autopsy Findings

External Examination:

Radiographs taken prior to autopsy revealed dicephalic parapagus twins fused anterolaterally, with two heads, fused upper limbs up to the wrist but separated hands, and fused upper limbs revealing two separate humerus, ulna, and radius. The radiographs also showed two separate spines which are not fused.
The conjoined twin was centrally and peripherally cyanosed. Conjoined twin had 3 upper limbs, one on either side, one fused upper limb with two palms, ten fingers and 2 lower limbs with normal fingers and toes. They share a single external male genitalia and a single anus which was patent. En mass evisceration of organs was done by a “y” incision, exposing the unfused vertebral bodies. Additionally, ribs were seen extending between the thoracic vertebrae.

**Hepatobiliary System:** There were two unfused gallbladders and two livers which were fused in midline.

**Lymphoreticular System:** One normally appearing spleen was observed. There were grossly normal lymph nodes in the hilar, retroperitoneal, and intra-abdominal regions.

**Genitourinary System:** There were two kidneys, each of which had a single ureter that empties into a single urinary bladder. Right testis was undescended and was found in the right superficial inguinal ring, left testis was descended till scrotum.

**Discussion**

One set of conjoined twins are born alive every 200,000 live birth. This is a random occurrence that has nothing to do with maternal age, parity, or genetics. Although modern research implies the fusion of two monovular embryonic discs based on examination of numerous conjoined twins, the classic theory for conjoined twinning is incomplete separation of a single embryonic disc at day 13–15 post fertilization phase.

Conjoined twins are categorized according to site they attach, either ventrally or rostrally. The most common types discovered were thoraco omphalopagus (28%), thoracopagus (18.5%), omphalopagus (10%), craniopagus (6%) and cephalopagus (5%), while cephalo thoracopagus and cephalo thoraco omphalopagus are incredibly rare.11 The unfused structures provide the basis of the second classification. Examples include dipygus, which has a single head and torso with a divided pelvis and four legs, and dicephalus, which has two heads on one body.2

Based on fusion (Spencer et al.), we further categorise them into three groups: twins with dorsal fusion, twins with lateral fusion, and twins with ventral fusion. The ventral union group is composed of the cephalopagus (fused with the head), thoracopagus (linked with the chest), omphalopagus (joined with the umbilicus), and ischiopagus (fused with the hips).12

Twins with a dorsal union fall in to one of three categories: Union of cranium (Craniopagus), sacrum (Pyopagus), or spine (Rachipagus). The only twin type with lateral fusion indicated in the Rachipagus type is Parapagus (fused with the side).13

According to the literature, 35% of conjoined twins die during the first 24 hours of life, and 40% of them are stillborn. Furthermore, only 60% of conjoined twins who undergo surgery survive.
Dicephalus twins have an exceptionally high rate of stillbirth and mortality. The documented survival times for those Dicephalic twins are quite brief, ranging from 15 minutes to 11 days. The sharing of the internal organs with complicated anatomy, some of which may be malformed, could be the cause. Conjoined twins from the Parapagus have a wide range of autopsy findings. In this study, the majority of the midgut and all of the structures in the hindgut were solitary, but the foregut and its derivatives were all double. Literature demonstrates that parapagus conjoined twins typically have two pairs of lungs, one of which is hypoplastic (typically the right pair). Our study had three lungs, none of the three lungs showed any evident signs of underdevelopment and appeared normal. Conclusion

Conjoined twin are a rare event, and the Parapagus dicephalus type is extremely rare (only 0.5% of all reported cases). Conjoined twins have a high rate of spontaneous abortion (about 60%). The management of conjoined twins is difficult, and their death and morbidity rates are high. The question of parapagus twin survival and quality of life is debatable because of the broad extent of shared organs. The majority of cases die early before they can undergo surgical separation.

The age gap between the couple can be one of the associated factor which could be explored in future studies. In all monochorionic, monoamniotic twin pregnancies, conjoined twins should be suspected. Careful sonographic assessment should be performed to rule out any classic conjoined twin signs and to determine the severity of the shared foetal organs for perinatal care and management.

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References