Case Study Article

CONJOINT TWIN

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INTRODUCTION

Conjoined twins also known as **Siamese twins** refers to twins that are physically fused in utero and consequently at birth. The first mention of conjoined twins historically comes from the neolithic period. The condition is proposed to result from either fission or fusion. It happens when a monozygotic twin pregnancy cleaves more than 13 days after fertilization. Conjoined twins are monochorionic (one placenta) and monoamniotic (one amniotic sac). Conjoined twins exist in a female to male ratio of 3 to 1. This type of pregnancy is a complicated phenomenon that requires an interprofessional approach to manage it effectively.

ETIOLOGY

The most commonly believed etiology of conjoined twinning is either partial fission (partial splitting of zygote) or secondary fusion (similar stem cells on the other twin and fuse the twins together). Both of these processes can be used to explain the embryological and anatomical findings observed in conjoined twins. However, the fission and fusion theories do not unequivocally account for every possible conjunction and cannot be applied to the full spectrum of findings in conjoined twins. Conjoined twins can be broadly categorized into non-dorsally conjoined twins (which includes ventral, lateral, and caudal conjunction) and dorsally conjoined twins.

Non-dorsal conjunction may have a varied presentation but typically demonstrates a single shared umbilical cord along with the sharing of organs. The dorsal conjunction presents with two umbilical cords and mostly separate internal organs. It is proposed that non-dorsal conjunction may be initiated by

duplication of the axial primordia, which leads to a change in the axial orientation and/or interaction aplasia. Dorsal conjunction may occur because of secondary fusion of initially separate monozygotic twins

EPIDEMIOLOGY

A very rare phenomenon, the occurrence is estimated to range from 1 in 49,000 births to 1 in 189,000 births, with a somewhat higher incidence in Southwest Asia and Africa. Approximately half are stillborn, and an additional one-third die within 24 hours. Most live births are female, with a ratio of 3:1.

PATHOPHYSIOLOGY

Conjoined twinning is the result of cleavage or axis duplication that occurs after day 13 of fertilization. They are described according to the site of fusion.

The following are the types of conjoined twins along with their frequency:

- Thoraco-omphalopagus (joined at thorax and abdomen) 28%
- Thoracopagus (joined at the thorax) 18.5%
- Omphalopagus (joined at the abdomen) 10%
- Heteropagus (parasitic twins) 10%
- Craniopagus (joined at the level of the cranium) 6%

Less commonly observed conjoined twins include:[7]

- Pyopagus (joined at sacrum and perineum)
- Rachipagus (joined at vertebral column)
- Ischiopagus (joined at lower abdomen and pelvis)
- Cephalopagus (joined from head to umbilicus)

The site of fusion and the organs involved are a primary consideration for separation surgery. Typically, 25% of live births live long enough to be candidates for surgery.

EVALUATION

First-trimester ultrasonography remains the best modality of diagnosis early in the pregnancy. Prenatal magnetic resonance imaging can also help with identifying the type of conjunction, embryological malformations, and tissue characterization. Prenatal imaging should be used to inform the postnatal magnetic resonance imaging.[9] Modern methods like 3D printing may aid with surgical pre-planning and subsequent separation if applicable.

DIFFERENTIAL DIAGNOSIS

In utero differential include cystic hygroma, teratoma, and lymphangioma.

CASE STUDY

Conjoined twin sisters Mercy and Goodness Ede were born joined at the chest, abdomen, and liver. The operation to separate them was carried out at the National Hospital in Abuja, Nigeria using equipment supplied by the Kids Operating Room. The girls shared a diaphragm and liver, but because all their other organs were separate, doctors determined they could live independently. Plastic surgeons on the team had to create artificial skin to cover a large area of the girl's chests which was open and at risk of infection once they were separated. The surgery took 13 hours and was carried out by a team of 78 staff in total.

TREATMENT / MANAGEMENT

The conjoined twins that survive until birth can be broadly categorized into two categories, those who can be surgically separated and those who cannot. The determination of which set of conjoined twins can be separated is an interprofessional effort employing imaging modalities and multiple reviews

Surgical management requires identification of the anatomic relationship of the conjunction between the twins. Three-dimensional modelling and simulations help the team in making surgical decisions. The use of tissue expansion preoperatively is very helpful due to issues with the closure of the abdominal wall. At least 1 simulation of the separation event is recommended preoperatively as that helps with the outcome of conjoined twins.

COMPLICATIONS

The intrauterine demise of the twins is likely given the complicated nature of the pregnancy. Postnatally, complications related to the complex anatomy of the twins may lead to death. If surgery is an option, complications related to separation include organ failure, skin defects, surgical infection, bleeding, injury to internal organs and/or vasculature, and failure to complete the procedure.

PROGNOSIS

Conjoined twins generally have a poor prognosis. The total survival rate is 7.5%. Only 60% of the surgically separated cases survive.[14] Antenatal imaging, postnatal surgery if applicable, use of tissue expansion in surgery, and cadaveric transplant for vital organs that are shared between the twins, might lead to a better prognosis.

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