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Conjoined twins - a review of literature with two case studies

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they were found to be of the commonest variety i.e. thoraco-omphalopagus type. The fusion extended from the xiphisternum down to the umbilicus (Fig. 1) Both were found to be suffering from postural cyanosis and dyspnea which was aggravated by placing them on one side. Besides the routine examination done the ultrasound and echocardiogram showed two hearts contained within a single pericardial cavity. The atria of the two hearts were found to be fused (Fig. 2) In addition, fusion of the liver was also noted with two separate gall bladders. While the twins were being further investigated and being fed with nasogastric tubes their condition deteriorated and they died after six days of admission of cardiopulmonary failure. A postmortem was performed which confirmed the atrial fusion and the fusion of the liver mass. In addition, although they had independent GIT, one of the twins had a persistent Meckel's diverticulum and the same twin also had ureteropelvic junction obstruction in one of these kidneys. The rest of the abdominal viscera were independent in each. Besides the face perhaps these twins were non-separable because of the cardiac fusion, the fact is highlighted that the proper facilities for cardiopulmonary bypass that might have been required were not available to us.

Fig. 1.



Fig. 1: Thoraco-omphalopagus fusion extending from the sternum down to the umbilicus.

Fig. 2.



Fig. 2: Postmortem of twin showing single pericardial cavity containing two hearts which are fused at the atria. Note also below the diaphragm a fused liver.

Case II

An eight year old boy presented to us with an omphalopagus fusion as a case of heteropagus twinning. One twin was complete while the other had an appearance only of abdominal, gluteal and limb element (fig. 3). On ultrasound the liver was noted to be shared by the twins and on x-ray showed the incomplete twin to have the complete vertebral axis, pelvis and complete lower limb skeleton. The fact that this boy had grown that long without having any treatment was unfortunately not due to illiteracy but due to the ulterior motives of the parents who had set out to earn money by displaying the child in different exhibition through out the country and by charging the public money to exploit their superstitious and unfortunate beliefs. Surgery was offered with the interest of the boy in mind but was vehemently refused.

Fig. 3.



Fig. 3: Eight year old twin, used by the parents and owner for display in exhibition.

Summary

Since time immemorial conjoined twins have fascinated both people and physician alike. Little is known about their etiology. They have been classified on the basis of anatomical union but with the help of the latest diagnostic tools both prenatal detection and subsequent management can be chalked out. But this gives them only a fighting chance even in the best of centres. Unfortunately our centers lack most of these facilities accounting for the high mortality.

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livery. In the most common type of conjoined twin, Thoracopagus, 73% have varying degrees of cardiac fusion and upto 90% have pericardial fusion.

In omphalopagus twins the fusion of liver, gastrointestinal and urinary tracts should be anticipated along with peritoneal complications. An omphalocele and imperforate anus may complicate the repair⁽¹⁾. In pygopagus twins usually the rectum and urinary tract are complete in each twin and spinal cord are also separate but the vertebrae are usually fused⁽¹⁾.

Ischiopagus twins present with fusion extending from the umbilical area downward including the lower trunk and pelvis. They may present with three limbs (tripus) or four limbs (tetrapus). They often present with fusion of gastrointestinal tract from the level of Meckel's diverticulum distally. Various degrees of fusion of the bladder, vagina, and colon should be anticipated⁽¹⁾.

Craniopagus are classified into partial and total subtypes. In the partial subtype the two brains are separated by bone and dura and each brain should have separate leptomeninges. The other variety has an extensive connection of brain tissue or separation only by the arachnoid mater.

The diagnosis of conjoined twins is important to evaluate whether the continuation of pregnancy is feasible. The most common and useful tool for antenatal diagnosis is ultrasonography. The antepartum ultrasound diagnosis of this abnormality was first described in 1977 by Wilson et al. (2 & 8).

Sonographic findings of relevance include:

- Fetal heads and body parts at the same level.
- Constant relative fetal position.
- En face fetal position.
- Bi-breech or less commonly bi-vertex presentation.
- Fetal extremities in unusual proximity.
- Hyperextension of one or both cervical spines.
- Solitary umbilical cord with more than three

vessels.

- Non-separable skin contour (1,2,3).

Amniography and amniocentesis provide additional information for diagnosis and prognosis⁽³⁾.

The success of possible postnatal separation depends on good preoperative assessment. In this regard investigation depending on the organs involved should be carried out. CT scan, ultrasound, echocardiogram, gastrointestinal contrast study, oral and intravenous cholecystogram, angiography and radionuclide studies may be carried out as the case may warrant⁽³⁾. In recent years MRI has been a useful adjunct to other investigations. One group of surgeons have used MRI to detect an avascular plane in the tissue bridge for the line of surgical separation of conjoined twins postnatally. MRI has been proved to be the most useful diagnostic tool for both prenatal and postnatal diagnosis and anatomical evaluation of conjoined twins and also proved to be the most useful in deciding the necessary surgical procedure⁽⁴⁾.

The choice and timing of operative procedure depends entirely on the complexity of attachments and associated congenital abnormalities. Careful planning of surgery for individual cases is of prime importance. The absolute indications for surgery at the time of birth include;

- Death / Critical condition of one twin threatening the life of other.
- Severe injury to the connecting bridge.
- Associated severe abnormalities not compatible with life but surgically correctable.

Generally speaking, the operation is recommended after three months of age. Successful surgical separation depends on cohesive team work. Proper hospital and operative facilities, proper preoperative evaluation, and intensive care management are essential.

Case I

A pair of conjoined male twins, 48 hours old, after having been spontaneously delivered at home in a village were admitted to the NICU of The Children Hospital, Islamabad. On examination

CONJOINED TWINS — A REVIEW OF LITERATURE WITH TWO CASE STUDIES

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Abstract

Two cases of conjoined twins have been reported. A review of literature is given.

Key Words: Twins, Conjoined Twins.

Introduction

The most famous conjoined twins in history and from where the term Siamese originates were Chang and Eng Bunker, born in 1811 near Bangkok in Siam. They were joined together at the umbilicus by a bridge of tissue 17cm in circumference. They remained united for the rest of their lives until they both died in 1874, one of pneumonia and second of hypovolemic shock just 30 minutes later. In between they led "normal lives" marrying twin sisters and producing 22 children between them. Since then the term Siamese twins is synonymous with conjoined twins^(1,2)

The first documented attempt at surgical separation of these twins was done by Farius in 1689 the operation was not successful. The first successful separation was reported by Konig in Germany only a year later (1690). Konig's twins were united only by an abdominal skin bridge⁽³⁾.

The incidence of conjoined twins is between 1:50,000 to 1:100,000. There is a geographical variation and there is also a female to male ratio of between 2 to 3:1⁽⁶⁾. There is also a high incidence of congenital malformations in these twins. There is a higher incidence in non-caucasians⁽¹⁾.

Conjoined twins are the result of an incom-

plete twinning process. They are always monozygotic, monochorionic and mono amniotic. The normal twinning process in a monozygote occurs between the 5th and 12th day after fertilization. In conjoined twins the process occurs between the 13th and 15th day, hence the incomplete separation. (6&7) Zimmerman believes that this is caused by interference with the inducers or organizers that determine the formation of body regions and organs⁽⁵⁾.

No definite causative factor has as yet been identified although in animal research some agents like butyric acid, manganese deficiency, radiation and acetone have been shown to induce the defect. Maternal age appears to have no role and there is also no explanation for female preponderance⁽³⁾.

Conjoined twins are classified by combining the anatomical area of union with Greek word pagos meaning "that which is fixed". There are five types namely; thoracopagus (joined at chest 40%); xiphi and omphalopagus (joined at ant. abdominal wall - 34%); pygopagus (joined at the buttocks - 18%); ischiopagus (joined in a linear fashion at the pelvis-6%) craniopagus (joined at heads - 2%)⁽³⁾. There could be a combination of the above types for e.g. thoracoomphalopagus which is the most common type.

The presentation of conjoined twins is much varied depending on the organs shared and associated abnormalities. 40% of conjoined twins are still born and 35% survive for only a day. As home delivery is very common in our country and the referral system poor, mortality rate is much higher in Pakistan. The causes of death in these conjoined twins are the associated severe congenital abnormalities and birth trauma during de-

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