

Review Article

The problem of separation and management of conjoined twins

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Sri Lanka Journal of Child Health, 2001; **30**: 72-5

(Key words: conjoined twins, separation, management)

Introduction

Identical twins have intrigued people all over the world. Their similarity in appearance and their complex mental connection with each other in some cases have been a source of debate over the ages.

Conjoined twins are by definition two near normal individuals united by a bridge of tissue that may range from skin alone or involve vital organs and bone. Early in this century, a boy of 9 had a fetal "twin" consisting of fetal parts enclosed in a sac, connected by a short umbilical cord, within his abdomen. This tumour grew in size ultimately killing its host. This would not qualify as a conjoined twin existing within the body. In the same context, should a twin born without a functional heart be labeled a twin or a parasite?

Identical twins are the product of a single fertilised ovum. One third of all twin births are monozygotic. They share the same chorionic membrane; 75% share the same placenta and are of the same blood group. They occur randomly in all races and do not follow the hereditary pattern of dizygotic twins. Monozygotic twins usually are split images of each other. Although sharing the same hereditary information, modifications may occur during embryonic development. Shunting of blood in conjoined twins may lead to disparity in development in one twin.

Identical twins may share the same emotions, thoughts and physical disabilities even though they may have lived apart from each other since early childhood. The Smithsonian 1980 reported the instance of Jim Lewis and Jim Springer, identical twins separated and adopted by two families. They were reunited after 39 years. Both had married twice, women with same first name. Their dogs had the same name, both chain smoked, both drank beer, both holidayed at the same location and had the same jobs.

Both drove the same cars. Medically, both had high blood pressure, migraine and severe heart attacks. Both had vasectomies. Their brain wave patterns were identical.

The diversity of the site and degree of union is considerable. Conjunction occurs at corresponding sites - the head, chest, abdomen, pelvis. Rotation in the vertical or horizontal axis of the fetal anlage before conjunction may occur. Unusual unions produce monsters. Organs may be shared. Mortality is directly proportional to extent of union, extent of visceral communication and presence of cardiovascular anomalies. Cardiovascular anomalies in thoracophagus twins are frequent. A common pericardium occurs in 90% and a shared heart in 75% of thoracophagus twins¹. The commonest site of union is the upper abdomen and lower chest. Thoracophagus occurs in 75% of conjoined twins. Two previous sets of thoracophagus twins recorded in Kenya were inoperable due to complex cardiac abnormalities. One set had a two chambered heart and a normal heart; the second set had normal hearts; the third set had fused hearts and poor pulmonary and systemic flow in one twin².

Incidence

The incidence varies with race, 1 in 250,000 births Bender³, 1 in 66,000 Milham⁴, 1 in 250,000 births Towey² in Kenya.

Historical aspects⁵

The earliest recorded case in the 12th century is the twin sisters of Biddenden in Kent, joined at the hip and shoulders. They had a single pair of upper limbs. There is a statue in the village of Biddenden in Kent in memory of the conjoined sisters. The next recorded case was the brothers from Scotland during the reign of James III. They were joined from the waist downwards. Probably the best known are the Siamese twins Chang and Eng Bunker born 1811, who earned their living in the United States as circus attractions in the Barnum and Bally circus and later

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as farmers. They lived long and fruitful lives, both married and fathered 21 children between them. They died at the age of 63. Two of their grand daughters produced 2 sets of normal fraternal twins.

The first successful separation was as far back as 1902 by Doyen who separated two sisters one of whom died. This was followed in 1912 by an unnamed R.A.M.C. officer with one survivor. Mc Laren⁶ in 1936 separated twins and both survived. Aird⁵ in 1954 separated thoracophagus twins from Kano and one survived: Peterson⁷ in 1960 separated thoracophagus twins and both survived. Mulcare⁹ in 1970 at Korat separated twins and one survived. Spitz in 1997 at Great Ormond Street Hospital separated conjoined twins and both survived.

Mortality is high and many succumb at or shortly after birth. 75% die of complex cardiovascular¹⁰ or cerebral anomalies. Others die of pulmonary complications. Survival is also dependent on the availability of assisted labour and centres capable of assessing and undertaking the separation of the twins. It is not surprising that successes are few and far between.

Patient

A 28 year old Kikiyu mother was delivered of conjoined twins in 1978 (Figure 1). A prenatal

diagnosis of conjoined twins was made as the twins were lying in the "kissing" double breech position. Labour was prolonged and final delivery was by caesarean section. The twins were united by a 13 cm bridge from the umbilicus to the nipple. They weighed 5.17 kg. There was a common umbilicus. Both twins were well formed. Respiratory and cardiac rates were asynchronous.



Figure 1. Conjoined twins at birth

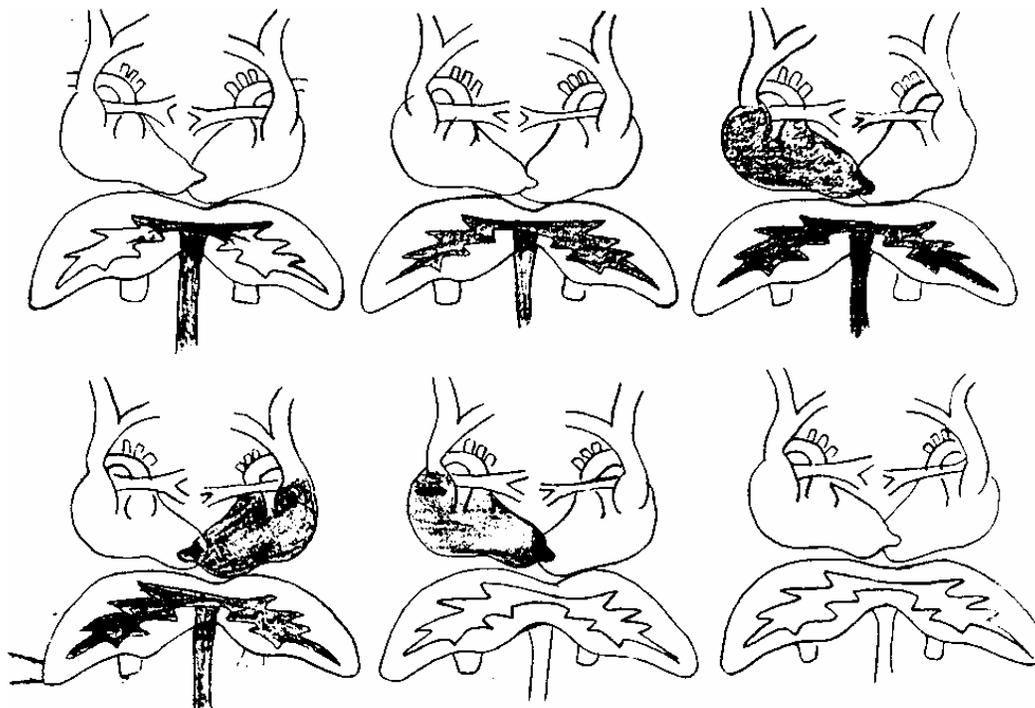


Figure 2. Angiogram (graphic) Conjoined twins

Investigations

Cardiac catheterisation through the umbilical vein revealed two hearts within a common pericardium. The right ventricle of each heart had a nipple like prolongation which was evidence of an earlier fistula between the two hearts. The hearts contracted asynchronously on the angiogram (Figure 2). There were separate hepatic and inferior caval systems. The livers united by a bridge of liver. There were twin gall bladders. The lower sternum was bifid with common costal cartilages. The gastrointestinal and renal systems were normal. There was a single umbilicus.

Clinical management

Although surgery was planned for the sixth month, the smaller twin showed signs of respiratory distress, tachypnoea and sweating 28 days after birth. The date for surgery was advanced to the first month after birth. Each twin was allotted a separate anaesthetic, surgical and nursing team. They were given a detailed program of the anaesthetic and surgical procedures. Barrier nursing and dummy runs were practised prior to the operation. One hour before surgery hydrocortisone 10 mg, atropine 15 mg and vitamin K 1 mg were given. During surgery each twin received an equal share of medication as cross circulation is unreliable. A heated mattress was used. The twins were intubated awake, relaxed with tubocurarine and manually ventilated with 50% nitrous oxide in oxygen. Sodium bicarbonate was given for correction of acidosis. The central venous pressure, ECG, EEG, rectal temperature, blood gas and urinary output were monitored. Blood pressure was monitored by neonatal cuff rather than by intra arterial monitoring which is difficult in neonates. Blood loss was measured by careful swab weight.

Surgery

The common abdominal cavity revealed a fold of peritoneum attached to a liver bridge connecting the livers. There were 2 separate gallbladders. The line of union between the livers was oblique. The liver bridge was divided between clamps. Bleeding was controlled with mattress sutures. The abdominal incision was extended up into the lower chest. The common pericardial sac was opened and the separation of the twins completed. The sternal defect was closed with a Teflon patch. Relief skin incisions were made in the loins for skin shortage. The skin defect in the loins was later skin grafted.

Postoperative management

Blood loss for Twin A was 50 ml and for Twin B 45 ml. In addition to blood replacement each twin received 40 ml of 10% dextrose and 15meq sodium bicarbonate to correct acidosis. Total operative time was 5 hours.

Twin A needed respiratory support for 21 days. This is necessary because paradoxical respiration persists till the defect in the lower sternum is stabilised. She gained weight and was discharged from hospital.

The smaller Twin B had persistent tachycardia and metabolic acidosis and died 10 hours after operation. Post mortem of Twin B showed an insignificant post ductal coarctation and left ventricular hypertrophy.

Follow up of Twin A 5 and 10 years later revealed that she was mentally and physically normal.

Discussion

The feasibility of separation should be evaluated against twins with anatomical or functional disability after separation. Separation of conjoined twins is ideal at the age of 6 months. If there are signs of deterioration in one twin surgical intervention should be advanced to save the life of the other. Endocrine dependence of one twin's adrenocortical tissue may determine the final outcome¹⁰. Instability of the mediastinum and the upper abdominal wall deficit consequent to the separation of the twins requires prolonged respiratory support. Absence of the cough reflex and respiratory distress is one of the factors that contributes to the high mortality after separation of the twins⁷. Accurate estimation of blood loss is critical. Hypovolaemia is a major factor in postoperative mortality¹².

Recent advances

Ultrasonographic diagnosis permits intra uterine evaluation of the degree of conjunction and the presence of vascular anomalies¹¹. Bleeding from the liver is reduced if the liver is drained of blood and the blood vessels sealed with glue while dividing the liver bridge. There is always shortage of skin for closure after separation of the twins. Gap closure with synthetic material is prone to infection. Current technique is to insert temporary teflon packs under the abdominal skin of the twins to act as skin expanders two weeks prior to the main operation. Operating time is reduced by team work and briefing of all the personnel involved.

This case was operated on at the Kenyatta National Hospital, Nairobi in 1978.

Acknowledgments

My thanks to Mr Roy Miller, Senior Surgeon, Kenyatta National Hospital, leader of the team and Prof. Aiyim for the anaesthetic notes.

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