Emergency separation of a xipho-omphalopagus twin in a developing country

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ABSTRACT

Female conjoined twins (thoraco-omphalopagus) were delivered by emergency caesarean section in a private nursing home. On examination, one of the twins was dead and was threatening the survival of the surviving twin (twin A). An emergency separation was performed to salvage the surviving twin.

Key words: Conjoined, emergency separation, female, newborn, surgery, twins

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INTRODUCTION

Conjoined twins are a clinical rarity having an incidence that varies from 1 in 50,000[1] to 1 in 200,000[2] newborns. More than 600 cases have been reported in the world literature until 1986, most of which are autopsy reports.[3] Although xipho-omphalopagus is the commonest variety with less anatomical complexity, the complex physiology and gross congenital malformation of one can be detrimental to the other. We report a rare case of emergency separation of xipho-omphalopagus conjoined twins in neonatal period. The indication of neonatal separation, complex anatomy and physiology are discussed.

CASE REPORT

A set of full-term female conjoint twins was delivered by primigravida through an emergency caesarean section because of prolonged, and failure of progression of, labor in a private nursing home. The pregnancy was supervised antenatally and antenatal ultrasonography done at 5th, 7th and 9th months of gestation showed presence of live and moving twins. There was no family history of twins. Because of prolonged labour and failure of progression of presenting part, an emergency caesarean section was performed in the nursing home. Two hours (the time taken to transfer the twins) after delivery, the twins were brought to our department for expert management, and on examination, it was found that one of the twins unfortunately was already dead. Both were females, the combined weight was 2.6 kg and they were joined by a bridge extending from xiphisternum to the umbilicus [Figure 1]. There was a single umbilical cord attached on the inferior aspect of the bridge. A soft tissue mass, probably liver tissue, could be palpated beneath the skin of the bridge. There were no palpable bowel loops in the isthmus. The surviving twin (twin A) was active with normal tone, pink in color and temperature was normal. Her pulse rate was 140/min and had good air-entry on both sides; however, she had a systolic murmur.

The other twin (twin B) had multiple congenital malformations including cyanotic heart disease and by the time this twin (twin B) came to us he was already clinically dead. The twins were referred from the nursing home with a note from the attending neonatologist. Twin "B" (who subsequently died when he reached our centre) was examined by the neonatologist in the nursing home and was suspected to have cyanotic heart disease. The clinical examination provided ample evidence that twin "B" was dependent on twin "A" and was threatening the survival of twin "A". In order to save time, only a few investigations were performed and decision of emergency separation was taken.

Twin "A" was started on intravenous fluid and she received vitamin K at the outset. Plain X-ray revealed well-aerated lungs in twin "A" and the bowel contained gas. The ultrasonography of the connecting bridge revealed liver tissues with wide channels in the substance. 2 Dimensional Echocardiography was not done to save time.

Emergency separation of the twins was performed 1 hour after admission, under a general anaesthesia. An incision on the anterior surface of the bridge connecting both lobes of liver exposed the liver tissue (diameter:

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Figure 1: Twin A (healthy twin) on left and twin B (dead) on right



Figure 2: Healthy twin at the time of discharge

5 cm wide); both peritoneal cavities were separate extending up to the centre of the isthmus. There were two falciform ligaments; the right contained an enlarged umbilical vein. The bowel of twin "A" contained gas (an intraoperative finding) and the rest of the viscera were positioned normally. The extrahepatic biliary apparatus in both of them was separate and patent. Once the peritoneal cavities of both the twins were opened, the isthmus was divided using bipolar cautery. This was an autopsy exploration of the dead baby. This completed the separation. Multiple interrupted figure of eight sutures approximated the cut margins of the shared liver. The ultrasonography report revealed the liver of body "A" was connected with that of "B" by a bridge of tissue. That bridge was divided and repaired. The posterior skin bridge was cut in line with the anterior incision. Inferiorly, the cord was left attached to twin "A". The abdomen of twin "A" could be closed easily. The procedure lasted 1 hour and 30 ml of blood was transfused.

Twin "A" was nursed in the neonatal Intensive Care Unit (ICU) under strict monitoring. Post-mortem examination of twin "B" showed hypoplastic, nonaerated lungs on both sides with atretic larynx and malformed heart.

The surviving twin (twin "A") did well in the postoperative period. Her echocardiography showed a large perimembranous Ventricular Septal Defect (VSD) which already had been enlisted for repair by cardiothoracic surgeon. The patient was discharged on the 10th postoperative day [Figure 2]. On follow up, the patient is doing well 2 months after surgery.

DISCUSSION

Thoraco-omphalopagus forms the commonest variety of conjoined twins.[3] The first reported surgical separation of conjoined twins is by Farius in 1689.[4] In a case report by B Birmole *et al.,* both the separated conjoined twins died after emergency separation.[5] In 1660, Bohm[6] separated a 2-day-old xiphopagus twins with no common organs. After 1660, no other neonatal separation was reported until 1952, when Reitman *et al.*[7] described the separation of 1-day-old xiphopagi with a bridge of skin, subcutaneous tissues and cartilage but without organ or cavity sharing. Only nine cases of surgical separation in newborn period were found in English Literature till 1968 by Gans *et al.*[8]

Most surgeons prefer not to operate on conjoined twins in the newborn period. However, emergency surgery is indicated in cases of (1) death or impending death of one twin that threatens the survival of the other; (2) existence of severe malformation that threatens the survival of twins and requires immediate correction and (3) significant damage to connection bridge.[8,9] In a series by Spitz *et al*., where seven pairs of conjoined twins underwent emergency separation, only four of the twins survived.[10] In another series by Watanatittan, only two patients of the three pairs survived.[11]

We performed an emergency separation because the dead twin ("B") threatened the survival of the partner (twin "A"). The results of clinical evaluation confirmed by autopsy showed that twin "B" was indeed a parasite of twin "A". A team approach and meticulous operative and postoperative management contributed to the success achieved in this case, even with the limited facilities in our setup.

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