Successful separation of Xipho-Omphalopagus twins

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ABSTRACT

Xipho-omphalopagus twins are conjoined twins sharing some part of gastrointestinal system and lower sternum. These types of twins have best chances of survival if successfully separated. We report a case of successfully separated Xipho-omphalopagus twins, highlighting the importance of proper planning and team effort in such separations.

KEY WORDS: Conjoined twins, omphalopagus, twin separation, Xipho-omphalopagus

INTRODUCTION

The separation of conjoined twins poses a unique challenge to pediatric surgeons due to its rarity, complex anatomy and physiology involved. A case of female xipho-omphalopagus twins sharing a common liver was delivered by elective caesarian section. The conjoined nature of the twins was not diagnosed antenatally. One of the babies (Baby A) had a major cardiac anomaly, while the other (Baby B) had a pelvic ectopic kidney. Babies were nursed in ICU and underwent successful separation at 6 months of age. Meticulous planning and complete clinical and radiological workup for the delineation of anatomy ensured a successful separation. The procedure was performed by a team comprising of pediatric surgeons, hepatobiliary surgeon, plastic surgeon, neonatologist and anesthetists. At 10 months of follow up, both the babies are doing well.

CASE HISTORY

A 30-year-old multipara underwent elective caesarian section and delivered female Xipho-omphalopagus twins and were then referred to our hospital. The combined birth weight of babies was 4.8 kg. The babies were designated as “Baby A” and “Baby B” and shall hereafter be referred to with these names. The babies were fused from xiphisternum to umbilicus and had a single umbilical cord with 3 arteries and 2 veins. The bridge of tissue was firm 5 x 6 cm with palpable bowel underneath [Figure 1]. The babies were investigated at birth and a decision was taken to defer the separation till the age of 6 to 12 months for adequate weight gain and to provide optimum survival advantage.

Investigations were repeated prior to surgery at the age of 6 months.

Investigations

Abdominal sonography revealed the fusion of the livers with other organs being separate. The left kidney of baby B was not visualized. Doppler examination confirmed that the IVC and portal venous systems were separate. Various investigations, including CT scan, MRI scan and Doppler studies, could not give exact anatomical picture regarding the individual extra hepatic biliary tree.

Contrast CT scan and MRI scan of the thorax and abdomen showed the fusion of liver. The differential injection of an intravenous contrast agent into the twins on CT scan delineated the area of overlap of the blood supply and the

Figure 1: Twins at birth showing fusion between xiphisternum and umbilicus
presence of three cross-communicating vessels traversing the bridge of liver [Figure 2]. The left kidney in Baby B was ectopically situated in the pelvis. Oral contrast given to one baby (A) confirmed that the bowels were separate. The rest of the organs were found to be normal.

Echocardiography of Baby A revealed a large VSD with bidirectional flow and a small PDA with moderate to severe pulmonary hypertension.

The babies were nursed in the surgical neonatal intensive care for 6 months during which the periodic assessment of babies was conducted, and meetings were held involving pediatric surgeons, hepatobiliary surgeon, plastic surgeons, anesthesiologists, neonatologist, cardiologists and nursing staff in presence of the parents of the twins for the planning of the surgery. At the end of 6 months, the babies had grown to 9.5 kg and the bridge of tissue had increased in size (10 cm). In the viewpoint of the adequate weight gain and the possibility of the increasing pulmonary hypertension of Baby A affecting the other twin, surgery was planned at 6 months of age. Written informed consent was taken with the provision of saving the baby with the better chance of survival in the event of inability to save both of them. A mock drill was conducted a day prior to surgery by the surgical and anestheisa teams.

The babies were taken for surgery on single operation table with adjacent operation table kept ready for closing the abdomen after separation. Two anaesthesia teams and two surgical teams were allocated to manage each baby. Baby A was first induced and intubated, following which Baby B was intubated.

After preparation and draping, incision was taken anteriorly with a rectangular flap of skin and subcutaneous tissue of Baby B being reflected on to Baby A. The falciparum ligament was divided, peritoneum was opened and liver bridge was defined [Figure 3]. The bridge of liver was divided with Harmonic scalpel and the bridging vessels were coagulated. The capsule of liver was approximated with horizontal mattress sutures of 2-0 vicryl, tied over haemostatic agent (surgicel fibrillar®). Posteriorly, a rectangular flap of skin and subcutaneous tissue from Baby A was reflected on to Baby B. The fused xiphisternum was divided with scissors. After separation, Baby B was transferred to the adjoining operating room and the abdominal closure was done. Both babies tolerated the procedure well and were extubated on the table and epidural analgesia was used in postoperative period. They were nursed in ICU for 48 h. Baby A was digitalized in view of the cardiac problem. Both the babies were discharged after 1 week. At 6 months of follow-up, both the babies were thriving well with adequate weight gain [Figure 4]. Baby A weighed 8 kg and Baby B weighed 9.5 kg.

DISCUSSION

Conjoined twins are rare, the estimated prevalence in the literature varies widely from 1:50,000 to 1:200,000, and from the reported cases, around 250 surgical separations have been reported thus far. The largest
The separation of conjoined twins presents a unique challenge to pediatric surgeons because of its rarity. Although omphalopagus twins have the best chances of survival, adequate team management and preoperative planning is required in relation to the radiological investigations for the evaluation of the shared organs, presence of anomalies, presence and extent of cross circulation and most importantly anesthesia management. The timing of surgery is controversial; however, a delay of few months gives better chances of survival. Early separation is indicated when one twin threatens the life of the other.

Conjoined biliary tract is reported in 25% of cases of omphalopagus twins. The routine evaluation of cross circulation is performed using many methods like Tc-99m microcolloidal human serum albumin (HSA), Tc-99m HIDA, injection of indigo carmine and the examination of its excretion in urine of the other twin. One method of evaluating cross circulation is contrast CT scan, which in our case showed a watershed zone in the centre of liver bridge. Some have advocated glucose tolerance test for the evaluation of parasitism. Intravenous fluorescein can be used to demarcate the large liver juncture.

Surgical separation should take the following into account: shared organs, soft tissue and bony defects that ensue after separation, age of the child and associated anomalies. In the absence of associated anomalies, xipho-omphalopagus twins have a better chance of survival.

The outcome of surgery and the long-term survival of the separated twins depend on numerous factors. The previous reports of the infant's death during or shortly after attempting the surgical separation have emphasized the dangers of insidious surgical shock due to adrenal insufficiency and recommended routine use of steroids in postoperative periods. Although major cardiac anomalies are contraindications of separations, variations in the cardiac functions have been observed with otherwise normal hearts. Accurate preoperative investigations, a team approach, previous experience and meticulous operative and postoperative management contribute to the success rate. The outcome is better in twins who do not share vital organs such as the heart or brain and the best results are reported for omphalopagus twins and pygopagus twins without neural involvement.

After separation, there are often large areas devoid of skin; despite the use of preoperative tissue expansion, primary wound closure is not always possible, increasing the risk of postoperative sepsis in these individuals. Tissue expanders and prosthetic patches, i.e., prolene or vicryl mesh, can be used.

The index case thus adds to the overall statistics of successful separation of conjoined twins.

REFERENCES


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