Our experience with anesthetic management of conjoined twins' separation surgery

ABSTRACT

Conjoined twins are one of the most fascinating human malformations. Here, we report the anesthetic management and challenges faced in performing the successful separation surgery of 4-day-old thoraco-omphalopagus conjoined twins, born at term to a multigravida by elective caesarean section weighing 3.5 kg with APGAR score of more than 7. Computerized tomography scan revealed fused anterior surface of the left lobe of liver with common left portal vein. Confirmation of cross-circulation between the twins was done by giving intravenous midazolam to one of the conjoined twins, but no effect seen in the other one. We highlighted the responsibility of anesthesia team in anesthetizing sequentially the two patients who are joined together, technical difficulty of intubating the twins facing each other, need of careful monitoring, anticipation of complications such as massive blood loss, hemodynamic instability, desaturation, and hypothermia, and preparedness for their management and vigilant postoperative care.

Key words: Anesthetic management; conjoined twins; separation surgery; thoraco-omphalopagus

Introduction

Conjoined twins are rare unnatural phenomenon ranging from 1 in 50,000 births to 1 in 200,000 births, with a somewhat higher incidence in Southwest Asia and Africa. Of these, about 40% were stillborn and 60% live born, although only about 25% of those that survived to birth lived long enough to be candidates for surgery. Till now, about 250 separation surgeries have taken place around the world and with very rare incidence of successful separation surgery in Indian subcontinent as per the current literature available. It was a daunting challenge for us, the anesthesiologists, as we need to care for two patients at the same time instead of just one.

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Case History

Four-day-term female conjoined twins born to a multigravida by elective caesarean section weighing 3.5 kg with APGAR score of more than 7 were posted for separation surgery. They were thoraco-omphalopagus twins, the most common (42%), joined ventrally at epigastrium from xiphisternum to umbilicus, facing each other [Figure 1]. There was nothing significant in history and examination.

Evaluation of the extent of shared organ systems done with computerized tomography scan and magnetic resonance imaging revealed around 3-cm-wide isthmus-like part in the

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Figure 1: Twins before separation surgery

left lobe of the liver along with left portal vein to be common between them with separate biliary system [Figure 2].

The twins were named and labelled B1 and B2, and all the monitoring cables, intravenous (i.v.) lines, and tubings were color-coded (red for B1and blue for B2) to avoid confusion. Separate 24G i.v. lines were secured in both individuals. One anesthesia team each was designated for each of the twins, and dedicated anesthesia work stations and monitoring equipments were assembled. All standard monitoring was attached as per American Society of Anaesthesia.^[1] Prior assessment of the extent of cross-circulation between the twins was done by giving i.v. midazolam 0.75 mg/kg to baby B1. No appreciable change was noticed in B2 suggesting no cross-circulation.

Induction

After preoxygenation with Jackson Rees modification of the Ayre's T-piece, sequential induction was done. All drugs and i.v. fluids were calculated based on the total weight, and half was given to each of the twins. Babies were induced separately with fentanyl 1 μ g/kg and sevoflurane at 5–6 volume% with preservation of spontaneous ventilation due to anticipation of difficult bag and mask ventilation and intubation as both babies were facing each other. But fortunately no difficulty was encountered in oral intubation done in the lateral position with uncuffed endotracheal tube size 3 mm and then inj. cisatracurium 0.4 mg i.v. was given to each of the babies. Maintenance was carried out with oxygen (50%) and air (50%) and 1 volume% of isoflurane. Caudal block was given to both the twins with 2 mL of bupivacaine 0.25% with fentanyl 1 μ g for perioperative analgesia.

Intraoperative course

Both babies were ventilated on pressure control mode of ventilation. Incision was made on the skin bridge connecting

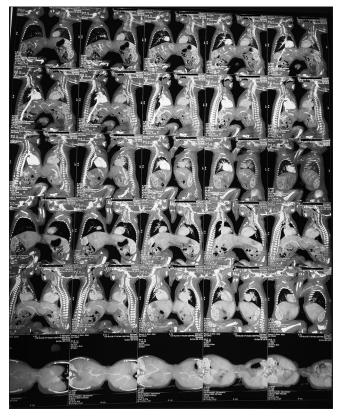


Figure 2: Magnetic resonance imaging of the twin babies revealing extent of organ sharing

both the twins. Excision of common isthmus was done and livers were separated with ligation of common left portal vein. Both the twins were separated successfully and twin baby B2 was shifted to another table. Repair of abdominal wall defect was done by the surrounding abdominal wall muscle flap.

Intravenous maintenance fluid given was 60 mL of Ringer's lactate with 1% dextrose to each baby. Serial blood gases and blood sugar done at 2-h interval revealed no significant derrangements. The total estimated blood loss was approximately 40–50 mL, which was replaced with 20 mL of fresh warm blood to each twin based on clinical assessment and blood gas analysis. Both the babies remained hemodynamically stable throughout the surgery period of 4 h with no eventful episodes [Figure 3].

Recovery and postoperative course

Both the babies were extubated on return of spontaneous ventilation after giving reversing agent (inj. neostigmine 0.1 mg and inj. glycopyrollate 0.02 mg) to each baby and shifted to the neonatology intensive care unit. Monitoring for hemodynamic instability and hypothermia was done. Blood sugar was checked periodically. Hypoxemia, electrolyte imbalance, and acidosis were evaluated through serial blood gases.



Figure 3: Twins after successful separation surgery

Outcome and follow-up

The postoperative course was uneventful. Nasogastric tube feeding was started on postoperative day 2. Twin neonates finally got discharged on the sixth postoperative day as two single separate individuals.

Discussion

Conjoined twins are identical twins (monozygotic and monochorionic) whose bodies are joined *in utero*. Approximately 75% of conjoined twin pairs are females. Conjoined twins are classified by the point of union like craniopagus type (joined at cranium), thoracopagus type (anterior union of upper half of trunk), omphalopagus (joined at chest or abdomen), pyopagus type (joined at sacrum), and ischiopagus (joined at pelvis).

Our case was conjoined baby of thoraco-omphalopagus type in which highest rate of separation survival is reported.

Kobylarz described the anesthetic management and associated problems for surgical operations in 10 sets of conjoined which include maintenance of patent airway, lung ventilation, optimal positioning of patients on the operating table, extremely high blood losses, long duration of surgery, and involvement of many members of medical personnel.^[2]

This case was more challenging and risky as twins were decided to be operated at an early age of 4 days on parents' insistence as they were having social stigma pertaining to the condition of the babies. Hence, they wanted to get them separated before taking them back to their native place. Operative survival is reported to be 50% in those operated in the neonatal period, but 90% in those over 4 months of age.

Estimation of circulatory mixing is a must in this case prior to induction to help calculate drug dosage and fluid replacement during surgery.^[3]

Many authors stress the fact of difficult face mask ventilation and intubation in thoraco-omphalopagus conjoined twins as their heads face each other. Hence, in our case also intubation was done with preservation of spontaneous ventilation.^[4]

Theodore *et al.* emphasized the importance of teamwork, communication, and advanced planning required in separation of 11-month-old craniopagus twins.^[5]

Hence, the goals concerning such kind of separation surgery involve the following:

- 1. Prior information about cross-circulation and organ sharing with their anesthetic implications
- 2. Massive fluid shifts and blood loss owing to long surgery
- 3. Meticulous planning and team effort in handling two patients simultaneously
- 4. Psychosocial counselling of parents.

Preservation of normothermia during separation surgery is an important factor affecting the surgical outcome owing to extensive heat loss due to long duration of surgery.^[6]

Parents should make the final and informed decision on separation only after considering the physical, ethical, and psychological aspects of possible separation.^[7,8]

Therefore, conjoined twins remain a topic of scientific speculation and public interest.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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