

The Perioperative Anesthetic Management of Conjoined Twins: A Case Report

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ABSTRACT

The perioperative management of conjoined twins poses unique anatomic, physiologic, logistic, and even ethical challenges, necessitating a multidisciplinary team approach. We present a case of omphalopagus conjoined twins separated at the second month of life (39 2/7 weeks post-conceptual age). This paper highlights significant components in the anesthetic management of conjoined twins for separation surgery, in light of modern medicine and technological advancements.

Keywords: conjoined twins, omphalopagus, anesthesia

INTRODUCTION

Conjoined twinning is a rare phenomenon wherein there is an incomplete splitting of monozygotic (identical) twins after day 12 of embryogenesis. It is estimated to occur in 1 of 50,000 to 200,000 births. Of every four cases, three are females, and there is a higher prevalence in Southwest Asia and Africa.¹ No two sets of twins are the same—the extent of shared viscera and cross-circulation vary from one pair to another, altering anatomy and physiology.

The complexity of the management of conjoined twins necessitates a multidisciplinary approach involving various specialists and disciplines. The pediatric anesthesiologist is a vital part of the team, involved not only during the separation surgery, but also the preliminary investigations and the post-operative critical care. Advances in anesthesiology and technology have contributed significantly to the global successes of conjoined twin surgery.²

CASE PRESENTATION

Preterm twins joined at the abdomen were admitted on the 4th day of life (DOL) at the University of the Philippines – Philippine General Hospital for further management. The patients were born preterm at 31-32 weeks age of gestation (AOG) to a 22-year-old mother via cesarean section due to malpresentation at a local hospital. Upon birth, Twin A had an Apgar score of 7, 8, 9, while Twin B had an Apgar score of 7, 9. Their combined weight was 3.1 kg, and the length of Twin A was 43 cm while Twin B was 44 cm.

On physical exam, both babies presented with grunting and subcostal retractions upon birth, managed as mild respiratory distress syndrome with neonatal pneumonia. They both had soft, non-distended abdomens, conjoined at the abdominal area (Figure 1) with a diameter of 3.5 cm

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Figure 1. Photo of the twins on admission showing conjoining of the abdominal area.

and circumference of 11 cm. The rest of the physical exam was unremarkable, with no other noted gross congenital anomalies. Echocardiography eventually done also had normal results.

An initial multidisciplinary conference (MDC) was held on the 3rd hospital day. Participants included specialists from Neonatology, Pediatric Surgery, Pediatric Anesthesia, and Plastic Surgery services. Consensus during the MDC was to optimize the patients first prior to surgery and wait until they are term. In the interim, the patients were weaned from

oxygen support, given antibiotics, and underwent feeding progression.

On the 38th hospital day, the patients were already on the 42nd day of life (38 weeks post-conceptual age). By this time, they already had a combined weight of 3.5 kg and were off oxygen support and antibiotics. The patients underwent abdominal computed tomography (CT) scan with contrast to assess the extent of organ sharing, as part of the planning for surgery. Two teams were organized for the procedure, one to take care of each twin, with all the equipment needed in two sets. The CT scan was done under monitored anesthesia care (MAC), a specific anesthesia service in which an anesthesiologist has been requested to participate in the care of a patient undergoing a diagnostic or therapeutic procedure and includes all the usual aspects of anesthetic care. During MAC, the continuous attention of the anesthesiologist is directed at optimizing patient comfort and safety.³

During the CT scan, the patients were snugly wrapped with a thin blanket and fell asleep while listening to white noise. Plastic wraps and a forced-air warming blanket were placed on the patients to ensure thermoregulation. A modified Bier block using Lidocaine 1 mg/kg IV was done prior to administration of contrast. For this modified Bier block, a single tourniquet was used and was released after 60 seconds. The procedure was successfully done without the use of any sedative hypnotics, the distribution of which may be unpredictable due to the uncertain extent of shared circulation of the twins. The set-up during the CT scan procedure is shown in Figure 2.

The CT scan revealed transperitoneal herniation of the right liver lobe of Twin B into Twin A with no definite liver conjoinment. There was also minimal renal excretion of contrast by Twin A on administration of contrast into Twin B, indicative of minimal cross-circulation.



Figure 2. Conjoined twins during CT scan showing use of linen to position patients while protecting pressure points, and of plastic wraps and forced-air warming blankets for swaddling and thermoregulation. Note the use of rain sounds to keep the patients asleep.

Now with the advantage of having high-resolution imaging results, another MDC was subsequently conducted. For this second MDC, Radiology service joined as well to provide insight on the CT scan findings. Intricacies of the surgery was planned, and a simulation was then done a few days after to enact the critical steps of patients handling and anticipate possible difficulties, including positioning during intubation. There were two teams consisting of surgeons, anesthesiologists, neonatologists, and nurses arranged to handle each twin, and two sets of anesthesia workstations, airway management equipment, and medications were prepared. Color-coding using stickers were used to designate equipment for each twin to avoid confusion.

The surgery was held on the 47th hospital day, and the patients underwent internal jugular cutdown right (Twin A), internal jugular cutdown left (Twin B), separation of conjoined twins under general endotracheal anesthesia. Total surgery time was 2 hours and 28 minutes, and the patients were extubated to oxygen support via nasal cannula. The patients underwent close monitoring at the post-anesthesia care unit for the first 48 hours post-operatively before they were transferred to regular ward. They were then sent home well on the 7th post-operative day.

DISCUSSION

Omphalopagus twins are conjoined twins bound at the abdomen, and the presentation may vary from a very simple conjunction to one that is exceedingly complex.² Shared organs usually involve the liver, digestive system, diaphragm, among other organs.¹ Once there is fusion of the liver, surgery may be particularly challenging with major blood loss expected. The extent of organ sharing and cross-circulation between twins determine the possibility and prognosis of a separation procedure.⁴

The case highlights the need for a collaborative approach in every step of decision-making in the management of conjoined twins. Initial challenges include answering the questions of whether or not separation is feasible, and if yes, when to perform the actual separation. The perspective of each discipline is equally important to ultimately provide the best possible care for both patients. In the case presented, given the prematurity and respiratory problems of the patients, after conducting an initial MDC, it was deemed necessary to upbuild and optimize them first prior to any intervention to increase their chances of survival.

The possibility of separation can only be ascertained after thorough investigations to determine the extent of organ-sharing and cross-circulation^{4,5}, and both the separation and necessary investigations have anesthetic implications. Through advances in technology in the field of radiology, high resolution CT scans are now able to provide a clearer image of anatomy, which aids surgeons in surgical planning.^{6,7} Extent of shared circulation can now also be determined through the use of contrast, whereas in the past, only the

presence of cross-circulation can be determined, rather than the degree.^{8,9} This was done by administering a medication such as atropine to the first twin, and then observing for the same drug effects in the second twin.^{10,11} The administration of drugs to ascertain cross-circulation in conjoined twins may pose risks, especially in certain patient profiles.

Radiologic investigations require absolute stillness of the patients, hence the need for the expertise of pediatric anesthesiologists. The use of white noise is an innovative way of inducing sleep in such settings. It is suggested that white noise may help soothe a baby by recreating the sounds in the womb, and may also block out sudden noises that may cause awakening. A study by Pratiwi et al. also concluded that using white noise is effective in the hypno sleeping process in infants aged 0-6 months.¹² White noise has also been found to control pain, reduce crying time, and positively effect vital signs in neonates¹³, possibly promoting sleep.

Currently, there is no literature available in the utilization of white noise in monitored anesthesia care for imaging procedures in the pediatric population. However, it proved to be a useful adjunct to swaddling in this case, especially in the setting of unknown presence and extent of cross-circulation. It eliminated the use of any sedative hypnotic in the face of uncertain pharmacodynamics and pharmacokinetics, and hastened recovery time.

Additionally, assigning roles during surgery and rehearsal in the operating room prior to the actual procedure continue to be an invaluable component of perioperative care of conjoined twins. It promotes organization and accountability during the surgery, and anticipates possible intraoperative challenges so that the members of the team can further prepare.

The vital role of a multidisciplinary approach in minimizing morbidity and optimizing care continues through the recovery period and post-operative care. While surgeons manage surgical sequela, anesthesiologists and neonatologists work hand-in-hand to maintain physiology. Anesthesiologists also manage pain in an effort to enhance and hasten recovery. Best outcomes are achieved through interdisciplinary communication and cooperation.

CONCLUSION

A multidisciplinary approach remains to be a key component in the successful management of conjoined twins. Teamwork, communication, and cooperation is a recurrent theme throughout the pre-, intra- and post-operative course of conjoined twin separation. Prior to separation surgery, now-available enhanced technology allows for more detailed imaging for surgical planning and modalities to ascertain the degree of cross-circulation in conjoined twins. Monitored anesthesia care with the help of white noise and a modified Bier block allows the conduct of diagnostic imaging procedures without the use of sedatives, which is beneficial in conjoined twins in whom cross-circulation is yet to be determined.

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