Anesthetic Considerations for a Preterm Two-Day Old Omphalopagus Conjoined Twins with Imperforate Anus For an Emergency Procedure: A Case Report*

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ABSTRACT

Conjoined twins are one of the most intriguing malformations in human beings. This report describes the anesthetic management of a two-day-old pretermomphalopagus conjoined twins posted for an emergency diverting colostomy for imperforate anus. These conjoined twins were born to a primigravid via cesarean section for fetal indication. They were born preterm at 36 weeks age of gestation with a collective birth weight of4.1 kg and an APGAR score 8,9. Computerized tomography scan of the chest and abdomen revealed Omphalopagus twins with left lower lobe atelectasis for Twin 2, fused liver, separate stomach and small intestines with distal fusion, imperforate anus, horseshoe kidney, bladder fistula, and the shared lower extremity appears to derive its blood supply from Twin 1. Evaluation of cross-circulation between the twins was done pre-induction by giving atropine IV to Twin 1. Atropine flush and increase in heart rate were noted in Twin 1, however, were not appreciated in Twin 2. Anesthesia for conjoined twins can be the most daunting and intimidating procedure that an anesthesiologist can handle in his lifetime. We highlight the challenges encountered with anesthesia preparation, preoperative planning, positioning, airway management, preservation of hemodynamic stability, as well as monitoring in an emergency setting.

Keywords: conjoined twins, atropine flush, omphalopagus, cross circulation

INTRODUCTION

Conjoined twins are an uncommon phenomenon worldwide ranging from 1 in 50,000 to 1 in 200,000 live births, with a somewhat higher incidence in Southwest Asia and Africa[1]. This condition is more frequently found among females with a ratio of 3:1[2]. About 40% are still born and 60% liveborn, although only about 25% of those that survived to birth lived long enough to be candidates for separation surgery[1]. One third of live births who die within the first few days are usually associated with complex anatomical conjunction, multiple congenital anomalies, and different degrees of cross circulation[3]. In some local context, giving birth to conjoined twins puts a social stigma giving the mother the perception that she delivered monster kids[4]. The occurrence of conjoined twins was initially explained by the fission theory arising from a partial / incomplete separation of the zygote around the 20th gestational day. More recently, the fusion theory has been proposed and is now more generally accepted. During fusion, stemcells find a like stem cells in the other twin and cause them to fuse together. The union of the embryos occurs around the 4th gestational week[5,2]. Conjoined twins are classified according to the anatomical site of conjunction: 40% for thorax (thoracopagus), 30% for abdomen (xiphopagus and omphalopagus), 18% for sacrum (pygopagus), 6% for pelvis (ischiopagus), and 1-2% for the head (craniopagus)[5]. Thoracopagus, thoraco-

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omphalopagus, and omphalopagus comprise approximately 70% of all conjoined twins[2]. Detailed evaluation of shared organs and associated anomalies is significant for anesthetic management. Here, we describe our experience regarding the successful anesthetic management of omphalopagus conjoined twins posted for emergency colostomy.

Case Description

Two-day-old preterm 36 weeks AOG conjoined twins, Twin FFA boy and Twin FFC boy, born to a 32-year old primgravid via primary cesarean section secondary to fetal indication, with a collective birthweight of 4.1 kg and Apgar Score of 8,9 was posted for an emergency diverting colostomy for an imperforate anus.

Antenatal history showed complete prenatal checkups from the first month of pregnancy up to delivery. 4 months age of, On prenatal check up, the parents were informed off et al conjunction through an ultrasound, Upon admission for birth, the Main Service informed the Section of Pediatric Surgery for co-management of conjoined twins based on prenatal ultrasound. Immediately after birth, the diagnosis of an imperforate anus was made. The conjoined twins were then formally referred to Pedia surgery service. Seen by Pedia surgery service on the 3rd hour of life, Assessment made was complete colonic obstruction secondary to absent fecal outlet. Hence, an emergency diverting colostomy was called. Further diagnostics were ordered which included cranialutz, baby gram and ct scan of the chest and abdomen. The conjoined twins were also referred to the Anesthesia service for evaluation and anesthetic management.

Physical examination by the Anesthesia service was done on the 17th hour of life. The conjoined twins were noted to be active, with good cry and appeared to be well-hydrated. Vital signs range forTwin 1 – 70/50 mmhg, 140 bpm, 40 cpm, 98% O2 sat; Twin 2 – 70/45 mmhg, 140 sbpm, 40cpm, 100% O2 sat. An OGT French 8 was noted to be in place for each twin, no fluid was seen draining from it. The chests were not shared and were noted to have adequate expansion. The lungs had good air entry and with clear breath sounds. Cardiac sounds were distinct, no murmurs were appreciated. The abdomen was noted to be slightly distended, the conjunction was observed to be just above the umbilicus up to the pelvis. There was a single genitalia that was ambiguous, and a shared imperforate anus. Twins have complete upper limbs with no distal limb deformities of the fingers and toes however, one twin has a pair of lower limbs and the other has a fused lower limb located posteriorly.

Assigning which of the twins was Twin 1 and which one was Twin 2, was based on taking the reference of the anesthetist's position on the head part of the operating table. Twin FFA (Twin1) was identified as the one on the left lying laterally on his right side, while Twin FFC (Twin2) was the one on the right lying supine. There was enough space between their faces to allow for intubation.

Umbilical catheter was in place and hooked to D5IMB with calcium gluconate. Separate gauge 26 intravenous line was also cited on the upper extremity of each twin and attached to heplock. No urinary catheter was attached because of ambiguous genitalia. Urine output was assessed by weighing the diaper and approximated at 1cc/kg/hr. There were no episodes of hypoglycemia and cyanosis at the NICU. Due to the urgency of the procedure, both twins were only partially evaluated for the extent of organ sharing as well as the presence of other associated anomalies. Based on perinatal workup, the pedia service assumed a shared circulation between the twins hence only one set of labs was taken. Laboratory results showed acceptable limits for preterm neonates except for leukopenia. Cranial ultrasound was unremarkable. Baby gram result showed a singular

abdominal cavity, while CT scan of the chest and abdomen revealed Omphalopagus twin with left lowerlobe atelectasis for twin 2, fused liver, separate stomach and small intestines with distal fusion, imperforate anus, horse shoe kidney, bladder fistula, and the shared lower extremity appears to derive its blood supply from Twin 1. No angiogram was done.

The twins were then prepared for surgery. Parents' consent was obtained for the surgical and anesthetic plan. The twins were maintained on NPO. One aliquot of 100mL cross matched maternal blood group-packed red blood cells was requested. They were given Ampicillin at 100 mg/kg, Gentamicin at 4 mg/kg and Metronidazole at 0mg/kg. Given the limited time for preparation, the two Anesthesia teams were immediately organized and planned. A dedicated anesthesia team was assigned for each of the twins. Each team consisted of 1 senior resident and 1 junior resident. The two teams were oriented and primed as to the plan and what to anticipate. In addition to the 2 teams, there was a single Team Captain (a pediatric anesthesiologist) who served as the central analyst and integrator of all of the data of the two patients. The monitors and anesthesia workstations were also designated for each twin. Infusion pumps, syringe pumps, as well as suction machines were positioned separately; anesthetic drugs were drawn and aspirated accurately, while emergency drugs were kept readily available. The operating room, the bed, as well as the fluids were warmed. The Pediatric Surgery team was also consulted for their site of incision in order to guide the Anesthetists' positioning of their workstations, for a more fluid flow during the time of operation.

The twins were received at the prewarmed operating room on the 28th hour of life and hooked to standard monitors as per American Society of Anesthesiologists' Basic Anesthetic Monitoring[6]. Even though the monitors and tubings were not color-coded, care was made sure to avoid entanglement. Vital signs were within normal limits. Peripheral line was patent on the upper extremity of each twin, as well as a central umbilical catheter hooked to a dextrose-containing intravenous fluid with calcium gluconate incorporation. Patency of all indwelling catheters and lines were checked prior to induction. The anesthetic plan was general endotracheal anesthesia. The twins were preoxygenated using Jackson Rees modification of the Ayre's T-piece. Since both twins were similar in size, for estimation of fluid and blood replacement as well as calculation of drug dosage to be given, the weight used for Twin 1 was 2kg, similar to Twin 2. Evaluation of the extent of cross-circulation between the twins was assessed by giving atropine 0.1mg IV to Twin 1 via the umbilical catheter. Atropine flush and increase in heart rate were noted in Twin 1, however, were not appreciated in Twin 2,partially ruling out cross-circulation but still on continued vigilance for unexpected depression of Twin 2. Hence, a sequential general endotracheal anesthesia induction was planned. Umbilical catheter was removed, main lines shifted to peripheral line for each twin and hooked to D5IMB with calcium gluconate incorporation. Twin 1 was then positioned for induction by placing a shoulder roll and the body shifted slightly to the left for better ventilation, while Twin 2 was supported with linen paddings. After positioning, Twin 1 was given fentanyl at 2mcg/kg IV, then sevoflurane was started at 1-2 MAC. Preparation for muscle paralysis of Twin 1 was facilitated upon an N-PASS (Neonatal Pain, Agitation, Sedation Scale) of -2 and after confirmation of ease of gentle manual ventilation, atracurium 0.5mg/kg IV was given thereafter. During intubation, Twin 2 was lifted a little higher over Twin 1 for better airway access. Intubation of Twin 1 was uneventful. Sequential induction of Twin 2 proceeded at the same doses of drugs as Twin 1. Intubation of Twin 2 did not need Twin 1 to be lifted because the former was already positioned supine. However, care was taken not to accidentally extubate Twin 1 while attempting to intubate Twin 2. Two attempts were made on

intubation of Twin 2 because of a change to a smaller uncuffed endotracheal tube size from 3.5 to 3. No desaturation, cyanosis, hypotension, nor bradycardia was observed during induction of both twins. Thereafter, Ultrasound-guided central venous access was performed on both twins - Left internal jugular vein for Twin 1, and the Right internal jugular vein for Twin 2. This was hooked to Sterofundin for fluid shift replacement.

Maintenance of anesthesia was carried out using 50% oxygen, 50% medical air, and1-2 MAC sevoflurane. Intraoperative Vital signs range of both twins were as follows: Twin 1 70/40 mmhg, 145 bpm, 35 cpm and 100% O2 sat; Twin 2 65/40 mmhg, 149bpm, 35 cpm, 95% O2 sat. The twins were kept warm all throughout the procedure. Active forms of heat transfer were applied. Blood glucose was monitored hourly intraoperatively and showed normal results. The monitoring of urine output was not really accurate because no urinary catheter was inserted due to the ambiguous genitalia. However, urine output was estimated based on the spillage on the under pad.

Surgery was uneventful and was concluded after two hours. On discontinuation of sevoflurane, the twins were later noted to have active movement of extremities, with good respiratory effort and volume. There was good air entry and breath sounds were clear. They were eventually safely extubated. Total duration of surgery was 2 hours, while total anesthesia time lasted 4 hours. Blood loss was estimated to be less than 5mL. Total amount of fluids given for Twin 1 was 100cc (12.5cc/kg/hr) and for Twin 2 was 80 cc (10cc/kg/hr). Total urine output was estimated at 1.3cc/kg/hr. They were transported back to NICU after the procedure, both were stable and with maintenance of thermo regulation on transport. They were given paracetamol IV at 7.5 mg/kg/ dose each and were continued every six hours for two days. Rescue dose of nalbuphine at 0.2mg/kg IV as needed every 6 hours was ordered for each twin for restlessness and incessant crying. The final procedure done was Exploratory laparotomy, ileostomy with distalmucus fistula, ultrasound-guided central venous access left internal jugularvein, Twin 1, and right internal jugularvein, Twin 2.

Monitoring for desaturation, hemodynamic instability and hypothermia were continued in NICU. Blood sugar was checked periodically. Hypoxemia, electrolyte imbalance, and acidosis were evaluated through serial blood gas. Describe the stay in NICU – Was nalbuphine rescue given, how many doses? How long was NICU stay, were there any problems encountered in NICU? What were the labs requested in NICU? How many days did the twins stay in the ward? On what postoperative day were the twins started on feeding? On what postop day were they finally sent home?)

DISCUSSION

Anesthetizing conjoined twins present several challenges to the anesthesiologists. While there is a number of data regarding the management of conjoined twins for elective surgery, we found paucity of literature of conjoined twins for an emergency procedure. Our case is one of emergent non-separation surgery.

The preparation of conjoined twins for an emergency procedure is nothing different from that for an elective. Because of the rarity and complexity of the case, an elaborate understanding of its pathophysiology should be obtained in order to anticipate potential problems, and prepare for its management. Additional pressure is put on the needs of not only one, but two neonates who are especially preterms, just like in our case. They are often delivered preterms because obstetric intervention is planned around the 36-38 weeks of gestation, once the pulmonary maturity has already been achieved, avoiding a complicated vaginal delivery or an emergency cesarean section. Emergency cesarean

procedures have a mortality rate of 70% versus 20% of the electives[5,7].

Careful and meticulous planning is vital because of the complexity associated with organ sharing, cross-circulation, as well as the presence of other congenital anomalies. In our case, time constraint was a limiting factor - The work-up was limited, the planning was short, and no practice rehearsal was done before the procedure. However, it did not affect the outcome of the procedure. As per The American Society of Anesthesiologists, the Risk Classification for these preterm conjoined twins were 4E, which means an absolute mortality rate of 7.8-25.9% plus 0.001-0.026% risk for postoperative complications for an emergency surgery[8.]

Cardiorespiratory insufficiency is the main cause of immediate death[5]. Thus, Anesthesia preparation is indispensable. A duplicated supply of equipment such as anesthesia machines, medical air, suctioning ands cavenging systems, and monitors should be available. The anesthesiologist should be prepared for multiple changes in positions, ET tube must be secured as well as arterial and venous lines for large fluid shift procedures. Care should be taken to avoid disconnection of monitoring lines as well as entanglement of IV lines leading to cross-dosing of drugs-which is why color-coding of tubings and monitors is ideal. We were not able to do this, however, we made sure that all the lines were fixed and organized in reference to the patient's body. Analgesics, anesthetic agents, muscle relaxants as well as emergency drugs should be prepared for each twin. Operating room should be arranged based on the type of conjoined twins to be operated on. In our case, two anesthesia machines were arranged on the head part of the twins, one on each side of the operating room table. Lastly, controlling the environment of the operating room is essential, noise and congestion should be limited both for the patients and the medical team's interest. Room temperature should also be regulated between 24-26C[5]. These challenges should be discussed with pediatric surgeons and with the neonatologist[5,6].

Difficult airway management is one of the anticipated potential problems encountered during the perioperative period. In non separation procedure, airway difficulties are more common in twins who are conjoined high on the chest, when their faces are facing each other or when there is hyper extension of the head and neck with exaggerated lordosis. In ample time such as in elective procedure, a mock drill and simulation is usually done a day ahead of surgery which involves stitching two dolls at the same site of conjunction seen in the conjoined twins. This is in order to increase the confidence of the anesthetist, and to avoid confusion and mismanagement on the day of surgery[3]. We were not able to do this because of the emergency nature of surgery. However, we did not encounter extreme difficulty during ventilation and intubation because their chests were not fused, so there was enough space in between their faces to maneuver. Access to their airway was also improved when proper positioning was done. Caution for auto transfusion was performed during lifting of Twin 2 over Twin1[5]. Signs of hypo volemia in Twin 2 such as increase in heart rate and poor pulses were, however, not observed.

As to blood loss, the type of fusion that is commonly associated with extensive blood loss is the thoraco pagusor ischiopagus twins. They require blood transfusion between 10% and 450% of estimated blood volume especially in separation surgery involving hepatic fusion[5].There is more extensive blood loss when there is greater fusion of organs. Cross matching of blood products is vital especially for separation surgery. Maintaining hemodynamic stability is essential during the surgical procedure. Blood loss assessment can be done by aspirating the volume of the operative field, checking for hematocrit variation, and

weighing of surgical gauzes. When there is circulatory communication there is no need to separately estimate the blood loss of each twin, as the volume and hematologic variation are shared[5]. Cross circulation is seen more often in craniopagus and thoracopagus twins[9,10]. Drug pharmacokinetics and pharmacodynamics are dependent on cross circulation[10]. This can be seen when an anesthetic agent administered in Twin 1 may have altered and unpredictable response in Twin 2[3,11]. Cross circulation also poses a risk of major bleeding during separation surgery[10]. In our case, there was no large fluid shift because our omphalopagus twins underwent a non-separation surgery and blood loss was only less than 5 mL.

Normothermia is another challenge in large neonatal surgeries. Drop in central temperature is expected because of the thermal self-regulation breakdown caused by the anesthesia, especially in the first hours due to heat redistribution. The unique physiology of these neonates pose additional clinical implications and challenges to the anesthesiologist: they have thin skin with large surface area which predispose them to rapid heat loss and hypothermia. In addition, a cephalic and more anterior glottis, poorly compliant lung, easy fatigability of the respiratory muscles, heart rate-dependent cardiac output, as well as hepatic and renal immaturity. The organ systems are not mature, the heart is not prepared for hypervolemia or hypovolemia. The kidney has difficulty retaining sodium under stress and there is tubuloglomerular imbalance, with urine concentration difficulty. The liver has not yet developed mechanisms for drug metabolism. The concentration of albumin and alpha1-acid glycol protein that bind to drugs is small which predisposes to a higher percentage of free drugs[5]. All these complicate the management of neonates, more so preterm conjoined twins.

In our case, we attribute the success to multiple factors: 1. The good medical status of the twins apart from the fact that they were conjoined, however, they had no congenital cardiac lesions which could pose additional problems hemodynamically; 2. The surgery was straightforward, it did not take long, and did not require changes in positions, which could otherwise have clinical implications in terms of monitoring, airway maintenance, and blood loss; 3. The knowledge and skill of the team captain pediatric anesthesiologist having had clinical experience with two separation surgeries in the past and one of which involved hepatic fusion with a good outcome; 4. The skill, vigilance, and coordination within the team and between the teams of the two anesthesia teams in handling neonates.

All these cumulative factors have played significantly toward the success of the procedure. Therefore, it cannot be over emphasized that the key factor in the success of surgery for conjoined twins lies in the multidisciplinary team approach with meticulous preoperative planning, perioperative vigilance, and interdisciplinary communication.

CONCLUSION

Anesthetizing conjoined twins is demanding and complex Hence, It cannot be overemphasized, that the key factors in success lies in the multidiciplinary team approach with meticulous preoperative planning, perioperative vigilance and interdisciplinary communication.

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Table and Figures

Figure 1: Conjoined twins at the Operating Room

Figure 2: Double setup prepared for the operation of the conjoined twins

Figure 3: Preoperative laboratory results

CBC	
HEMOGLOBIN	153g/l
HEMATOCRIT	0.46
WBC	5.61x 10/ul
PLATELET	140
PT	22.5
INR	1.7
%ACT	47%
APTT	75.5
CHEMISTRY	
NA	142g/l
K	3.6 mmol/l
CA	1.8 mmol/l
MG	0.56 mmol/l
CREATININE	44 umol/l

REFERENCES

1. Rawat, A., Rai,R., Paswan, A.K., & Pandey,V.(2020). Our experience with anesthetic management of conjoined twins' separation surgery. Saudi Journal of Anaesthesia, 14 (1), 100. https://doi.org/10.4103/sja.sja_142_19 Smith Anesthesia for Infant and Children 8th edition davidson, clavis and motoyama Grant M. Stuart MBChB, FRCA The anaesthetic managemen of conjoined twins https://www.sciencedirect.com/science/article/abs/pii/S1055858615000694?fbclid=IwAR0kep6P1kYkYT9QHvnw70WCicpq8PNCujYtpYe9qo0scLc7BQV-e9tIOCg
2. Deepak, C., Vandana, S., Pradeep, B. D., & Nikhil, K. (2019). Anesthetic management of separation of conjoint twins: Challenges and limitations. Saudi Journal of Anaesthesia, 13 (2),140–143. https://doi.org/10.4103/sja.sja_589_18
3. Anesthesia for successful surgical separation of conjoined twins in ... (n.d.). https://juniperpublishers.com/jaicm/pdf/JAICM.MS.ID.555765.pdf
4. De Freitas, M. H. B., Lima, L. M., De Menezes Couceiro, T. C., Da Costa, M.C., & De Freitas, M. R. (2019). Anesthesia and perioperative challenges for surgical separation of thoraco-omphalopagus twins: case report. Anesthesia and Perioperative Challenges for Surgical Separation of Thoraco-omphalopagus Twins, 69 (2), 214–217.https://doi.org/10.1016/j.bjane.2018.10.004
5. Módolo, N.S.P., Amorim, R.B., Castiglia,Y.M.M., Dias,S.R., & Guasti,V. M. (2002). Anestesia para separação de gêmeos isquiópagos no período neonatal:relatodecaso.RevistaBrasileiraDeAnestesiologia,52(4).https://doi.org/10.1590/s0034-70942002000400008
6. Daabiss, M. (2011). American Society of Anaesthesiologists physical status classification. Indian Journal of Anaesthesia, 55 (2),111.https://doi.org/10.4103/0019-5049.79879
7. Chalam, K. S. (2009, June 1). Anaesthetic Management of Conjoined Twins' Separation Surgery PubMed Central (PMC). https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2900119
8. Dogar, S.A., Hamid, M.R., Khan, A.U., Shamim, F., & Khan, F.A.(2017). Anaesthetic management of thoraco-omphalopagus twins with congenital heart disease for separation surgery. Anaesthesia Cases. https://doi.org/10.21466/ac.amottwc.2017
9. Shank, E. S., Manohar, N. S., & Schmidt, U. (2005). Anesthetic Management for Thoracopagus Twins with Complex Cyanotic Heart Disease in the Magnetic Resonance Imaging Suite. Anesthesia & Analgesia. https://doi.org/10.1213/01.ane.0000140952.35504.4d
10. Smith Anesthesia for Infant and Children 8th edition davidson, clavis and motoyama