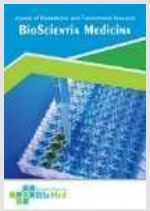
eISSN (Online): 2598-0580



Bioscientia Medicina: Journal of Biomedicine & Translational Research

Journal Homepage: www.bioscmed.com

Anesthetic Management of a Single Ventricle in Pediatric Patient Undergoing Open Duodeno–Duodenostomy Surgery: A Case Report

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ARTICLE INFO

Keywords:
Baby
Duodenal atresia
Functional single ventricle
Open duodeno-duodenostomy

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All authors have reviewed and approved the final version of the manuscript.

https://doi.org/10.37275/bsm.v8i8.1045

A B S T R A C T

Background: Duodenal atresia is a rare congenital disorder characterized by blockage of the small intestine. Open duodeno-duodenostomy is the surgery of choice to correct this condition. **Case presentation:** We report the case of a three-day-old baby girl with duodenal atresia and a functional single ventricle who underwent open duodeno-duodenostomy. The patient was born via caesarean section due to indications for a previous caesarean section in the mother and congenital abnormalities in the fetus. At birth, the patient showed signs of intestinal obstruction and cyanosis. Preoperative physical and laboratory examinations confirmed duodenal atresia and cardiac abnormalities. The patient underwent an open duodeno-duodenostomy without complications. The patent ductus arteriosus must be kept open to maintain systemic perfusion by maintaining PaO_2 at 40 to 45 mmHg and SaO_2 at 70% to 80%, along with the administration of prostaglandin agents. Good perioperative management and improvements in surgical procedures will increase the life expectancy of patients with single ventricle problems, especially hypoplastic left heart syndrome. The use of low-dose fentanyl induction agent, 1 MAC sevoflurane, and atracurium has been proven to produce favorable outcomes in these patients. **Conclusion:** Open duodeno-duodenostomy is a safe and effective operation for duodenal atresia. The patient in this case recovered well after surgery. Open duodeno-duodenostomy should be considered as the primary treatment option for duodenal atresia.

1. Introduction

Hypoplastic left heart syndrome (HLHS) is the most common single ventricle defect (accounting for 25% of all single ventricle lesions).[1] Knowledge of single ventricle physiology (SVP) becomes very important for anesthesiologists in making a clinical decision. The management strategy and the actual delivery of care from the time of birth (or at the time of diagnosis) through the postoperative period are crucial to optimize the short-term and long-term outcomes.[2] Single ventricle defects affect approximately 31 per 100,000 live births. They are characterized by the absence of two well-developed ventricles, one of which is typically rudimentary or hypoplastic.[1]

SVP is a very specific and precise term used to describe a circulation wherein the complete mixing of pulmonary venous and systemic venous blood occurs at the atrial or ventricular level and where the ventricles then distribute output to both the systemic and pulmonary vascular beds.[3] Depending on the patient's condition, less intervention may be most beneficial during initial management prior to surgery.

In the otherwise stable and compensated patient, interventions such as providing supplemental oxygen therapy and/or mechanical ventilation may actually upset the balance of the circulation, resulting in hemodynamic instability and metabolic disturbance, resulting in end-organ dysfunction.[2] Different studies identify a variety of factors associated with a high risk of perioperative complications in patients with congenital heart defects for noncardiac surgery, of which are how complicated are the disease, the general and physiological condition, the type of operation, and the smaller the child.[4]

2. Case Presentation

A three-day-old baby girl, who is being treated in the NICU with ventilator support, is planning to undergo an open duodeno-duodenostomy procedure. The patient has a history of being born by cesarean section due to maternal with a history of first cesarean section indication and fetal congenital abnormalities including polihydramnions, duodenal atresia, and functionally single ventricle with left ventricular hypoplasia. At birth, the patient was reported to cry immediately, appeared cyanotic, had weak muscle tone, and no chest wall retractions were observed with a weight of 2460 grams and length of 46 cm. The amniotic fluid was clear, with an Apgar score of 8-9.

The patient was consulted with a pediatric cardiologist and received therapy including oral misoprostol at a dose of 10 mcg/kg/dose (25 mcg) every 6 hours, and continuous infusion of heparin at a dose of 500 IU/kg (1260 IU in 50 ml NaCl 0.9% at a rate of 10 IU/kg/hour). On preoperative physical examination, found lethargic, with a respiratory rate of 55 breaths per minute and peripheral oxygen saturation of 88% using PCAC ventilator mode (VG VT 10.8 PEEP Pmax 25 RR 50 FiO_2 21%), heart rate 147 beats per minute, axillary temperature 36.7°C. Cardiovascular examination revealed single and regular S1 and S2 heart sounds with no murmurs or gallops. The abnormal result from laboratory examination showed HGB 9.20 g/dL, HCT 27.20%, prolongation of faal hemostasis with PPT 18.9 seconds, APTT 50.3 seconds, INR 1.37, hyperkalemia (K 5.94 mmol/L), hyperbilirubinemia with total bilirubin 4.90 mg/dL, direct bilirubin 0.26 mg/dL, indirect bilirubin 4.64 mg/dL. A babygram X-ray showed a suspected duodenal atresia with a double bubble sign appearance and suspected pneumonia in the lungs, with a normal heart size. Echocardiography revealed functionally single ventricle, LV hypoplasia, patent VD stenting of small tortuous vertical duct type C L to R shunt using Cre8 DES 4.0x12 mm (4/11/23), Large ASD II bidirectional shunt, mitral atresia, pulmonary atresia without MPA. malposition aorta.

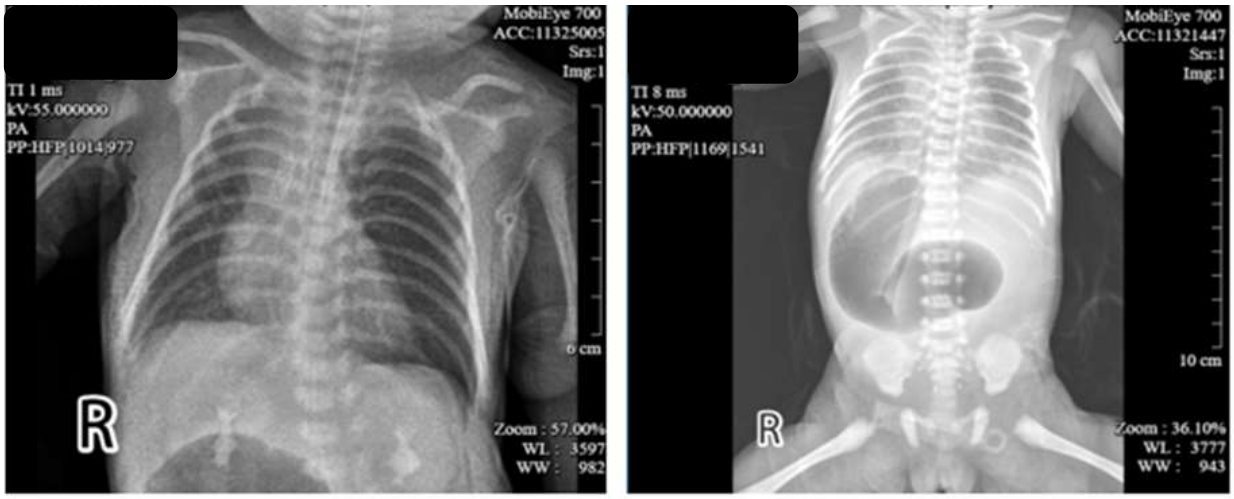

Figure 1. Preoperative babygram.

Preoperative management included administering 21% oxygen fraction with mechanical ventilation PCV mode and misoprostol as a prostaglandin agent to maintain PDA patency. On the day of surgery, she was anesthetized using general anesthesia. No premedication was administered, and a warming mattress and plastic wrap were prepared to maintain the patient's warmth during the procedure. Intraoperative management involved administering fentanyl 1.5 mcg/kg IV, sevoflurane 1 MAC, and atracurium 0.5 mg/kg IV to facilitate the surgery and anesthetic maintenance along with intermittent atracurium. The oxygen fraction was maintained at 21% without hyperventilation manipulation, with a peripheral oxygen saturation target at 80 - 85%. The surgery lasted for two hours, and the patient's vital signs remained stable. She received 68 ml of crystalloid fluid with an estimated bleeding of 7 ml and a urine output of 5 ml during surgery. In the surgery, we found the liver, appendix, and gallbladder on the left side. The stomach and Treitz's ligament were found on the right side. Upon evaluation, a duodenal atresia was identified. After surgery, the patient was promptly transferred back to the NICU for care, where she remained on a ventilator with post-operative analgesia using fentanyl 120 mcg/day in NaCl 0.9% 10 ml, administered at a rate of 0.4 ml/hour (4.8 mcg/ hour).

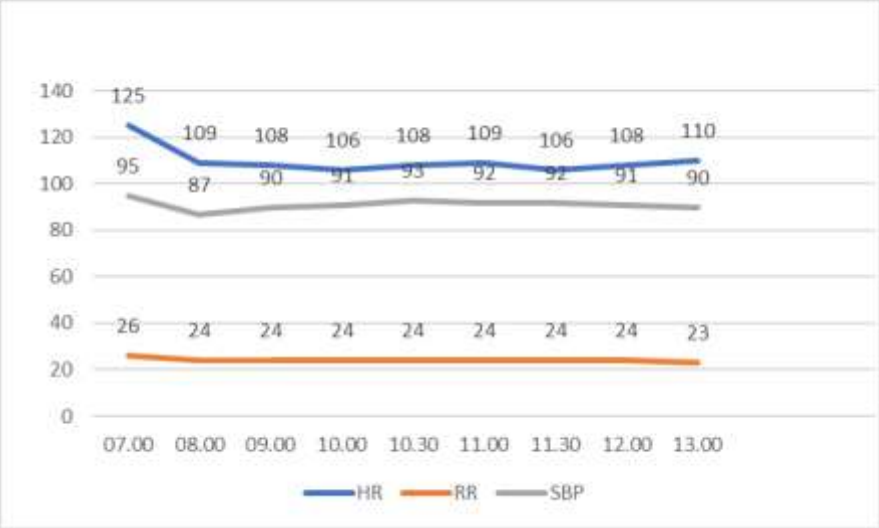

Figure 2. Vital signs during surgery.

3. Discussion

Infants with single ventricle disease are typically dependent on a patent ductus arteriosus (DA) to achieve adequate SBF and DO_2 or to maintain PBF adequate for gas exchange.[2] In this case, preoperative management included mechanical ventilation with PCV mode with a tidal volume of 10.8, PEEP 5, Pmax 25, and RR 50, with an oxygen fraction of 21%, and misoprostol as a prostaglandin agent. This corresponds to the literature where hypercarbia combined with a 21% FiO_2 is employed to elevate PVR while decreasing pulmonary blood flow. Avoid a high FiO_2 unless the PaO_2 is less than 35-40 mmHg. A target PaO_2 of 40 to 45 mmHg and SaO_2 of 70% to 80% are associated with adequate systemic O_2 delivery because exceeding this value can lead to an increase in pulmonary blood flow (lung overflow/over-shunted).[5,6] Continuous infusion of PGE_1 is used to maintain ductal patency in patients with ductal-dependent blood flow.[5] We used oral prostaglandin agents due to the unavailability of intravenous prostaglandin agents. Common adverse effects of prostaglandins include respiratory depression and apnoe, CNS disturbance, electrolyte imbalances, pyrexia, and systemic hypoperfusion. In children with ductus arteriosus–dependent congenital heart

disease, closure of the duct will lead to impaired systemic perfusion and cardiorespiratory collapse if the duct is needed for blood flow to the systemic circulation (eg HLHS).[1] Single ventricle physiology (SVP) is a very specific and precise term used to describe a circulation wherein the complete mixing of pulmonary venous and systemic venous blood occurs at the atrial or ventricular level and where the ventricle(s) then distributes output to both the systemic and pulmonary vascular beds.[3]

Anesthesia induction is performed using low-dose inhalation medication. The most crucial aspect is to maintain optimal SVR to prevent additional shunting from right to left. Sevoflurane is the appropriate choice because its effect on SVR is minimal.[6] Induction for this patient involves the use of fentanyl 1.5 mcg/kg IV, sevoflurane 1 MAC, and atracurium 0.5 mg/kg IV to facilitate incision with maintenance sevoflurane and intermittent atracurium without hyperventilation manipulation to optimize systemic oxygen delivery and perfusion pressure. The primary goal in the management of patients with SVP is the prevention of end-organ injury. Systemic oxygen delivery and perfusion pressures are optimized by balancing the systemic and pulmonary circulations (QP:QS ~1:1). QP:QS > 1:1 is associated with a decrease in systemic oxygen delivery due to a decrease in systemic blood flow. QP:QS < 1:1 is associated with a decrease in systemic oxygen delivery due to a decrease in systemic oxygen content.[3] We opted for a low dose of fentanyl, specifically 1.5 mcg/kg in this case because higher doses can lead to cardiovascular instability and muscle rigidity. The typical suggested intravenous dosage of fentanyl ranges from 1 to 2 mcg/kg.[7,8] As for atracurium, we administered it at 0.5 mg/kg for induction, aligning with literature suggesting that this standard dose results in a more rapid 95% depression of twitch in neonates. The adverse effects associated with atracurium relate mainly to histamine release. The cardiovascular changes are dose-related and usually occur at doses greater than twice the ED_{95}.[9-11]

Non-cardiac surgical procedures in single ventricle (SV) patients present a unique set of challenges for anesthesiologists. The physiological complexity of SV, combined with the need to maintain systemic perfusion, demands a careful multidisciplinary approach and individualization of anesthetic strategy. This manuscript aims to discuss in depth the important aspects of anesthesia in SV patients, from pathophysiology to anesthetic techniques and perioperative management. SV is a congenital heart defect in which there is only one functional ventricle, either the right ventricle (RV) or the left ventricle (LV). This causes a significant imbalance in the pulmonary and systemic circulation, resulting in hypoxemia and potential heart failure. SV patients often have coexisting cardiac abnormalities, such as pulmonary valve atresia or pulmonary stenosis, further compounding the physiologic challenges. The main goal of anesthesia in SV patients is to maintain perfusion of vital organs, especially the brain and heart. Maintaining patency of the ductus arteriosus (DA) is a vital blood vessel that connects the aorta and pulmonary artery, allowing oxygenated blood flow from the right ventricle to the systemic system. Keeping the DA open during surgery is critical to maintaining systemic perfusion. Achieving and maintaining adequate arterial oxygen saturation (SaO_2) of at least 70-80% is necessary to prevent hypoxemia and organ dysfunction. Arterial blood pressure (TA) should be maintained within a narrow range to avoid hypotension or hypertension, which can worsen organ perfusion. SV patients have limited cardiac reserve, so myocardial stress during surgery should be minimized. SV patients are at high risk of bleeding complications, so careful coagulation management is essential.[12-14]

The choice of anesthetic agent and anesthetic technique in SV patients should be based on the individual risk profile and type of surgery planned. In general, low-dose induction agents such as fentanyl and propofol are preferred to minimize negative inotropic effects. Sevoflurane, a volatile inhalation agent, is often used for the maintenance of anesthesia because of its mild vasodilator effects. Atracurium, a non-depolarizing neuromuscular paralytic agent, can

be used for muscle relaxation, but the dose should be titrated carefully to avoid negative inotropic effects. Anesthesia in patients with a single ventricle (SV) is like walking on a thin rope. On the one hand, we must ensure that adequate analgesia and muscle relaxation are achieved for the operation. On the other hand, we must be careful not to aggravate the already fragile physiology of the heart. Selection of the appropriate anesthetic agent and anesthetic technique is key to success in this challenging scenario. Initiating anesthesia in SV patients requires a gentle touch. Low-dose induction agents such as fentanyl and propofol are often preferred because of their minimal negative inotropic effects. Fentanyl, a potent opioid analgesic, provides potent initial analgesia and helps reduce anxiety. Propol, a fast-acting intravenous anesthetic, induces hypnosis smoothly and minimizes adverse cardiovascular effects. Maintaining anesthesia in SV patients requires a careful balance between analgesia, muscle relaxation, and hemodynamic stability. Sevoflurane, a volatile inhalation agent, is often the primary choice for the maintenance of anesthesia. Its mild vasodilator properties help maintain blood pressure and organ blood flow. Sevoflurane also provides good analgesia and minimizes negative inotropic effects. Adequate muscle relaxation is necessary for successful surgery. However, in SV patients, the selection of neuromuscular paralytic agents must be done with caution. Atracurium, a non-depolarizing agent, is frequently used because of its minimal negative inotropic effects. However, the dose should be titrated carefully to avoid excessive muscle weakness that could worsen ventricular function. Several other factors need to be considered when selecting an anesthetic agent for a SV patient: History of allergies: The patient may have allergies to certain anesthetic agents, which need to be taken into consideration; Liver and kidney function: Impaired liver or kidney function may affect the metabolism and excretion of anesthetic agents, requiring dosage adjustments; Medications currently being consumed: Some medications the patient is taking may interact with anesthetic agents, requiring dose adjustments or selection of alternative agents. There is no one-size-fits-all approach when it comes to anesthesia for SV patients. The choice of anesthetic agent and anesthetic technique should be based on the patient's individual risk profile, type of surgery planned, and comorbid medical conditions. Careful consultation with an anesthesiologist and pediatric cardiologist is essential to develop an individualized and comprehensive anesthesia plan that maximizes the chances of success and minimizes risks in these complex patients.[15,16]

Before stepping into the operating room, thorough preparation is the key to success in patients with single ventricle (SV). A comprehensive preoperative evaluation is like a road map that guides the medical team in identifying individual risks and optimizing the patient's physiological status. Consultation with a pediatric cardiologist is like the main gateway to understanding the heart anatomy and unique physiology of SV patients. A thorough understanding of the underlying cardiac abnormality, such as pulmonary valve atresia or pulmonary stenosis, is essential to estimate perioperative risks and develop appropriate management strategies. A patient's medical history is like a book that holds valuable information about comorbid medical conditions, drug allergies, and previous surgical history. These details assist the medical team in selecting the appropriate anesthetic agent and anticipating potential complications. The function of the patient's vital organs, like the pillars that support health, must be studied carefully. Laboratory examinations, including blood tests and liver and kidney function tests, provide insight into the patient's physiological status and help in identifying anesthesia-related risks. A comprehensive heart and lung assessment is like a compass that guides the medical team in understanding the patient's heart function and blood flow. Echocardiography, chest x-ray, and pulmonary function tests provide important information about heart status, blood pressure, and oxygenation. Armed with the information gathered during the preoperative

evaluation, the medical team can create an individualized anesthesia plan tailored to the SV patient's unique needs and risks. This plan should include the choice of anesthetic agent, anesthetic technique, and perioperative monitoring strategy. Close collaboration between the anesthesiologist, pediatric cardiologist, and surgical team is essential to ensure optimal SV patient care. Open communication and constant exchange of information help in identifying and addressing potential complications and improves surgical outcomes. Comprehensive preoperative evaluation is like a solid foundation that supports successful surgery in SV patients. With a deep understanding of the patient's cardiac anatomy, physiology, and overall health status, the medical team can develop an individualized anesthesia strategy that maximizes the chances of success and minimizes risks. Collaboration between specialists and open communication are the main keys to ensuring optimal SV patient care.[17,18]

Upon entering the operating room, the medical team is faced with a challenging but crucial task: maintaining the physiological stability of a patient with a single ventricle (SV) during surgery. Careful hemodynamic monitoring, appropriate fluid management, and judicious use of inotropic and vasopressor agents are key to achieving this goal. A patient's vital signs are like a window that provides a glimpse into his or her physiological status. Blood pressure (TA), heart rate, cardiac output, and arterial oxygen saturation (SaO_2) should be closely monitored during surgery. This information allows the medical team to detect physiological changes early and take appropriate corrective action. Intravenous fluid administration during surgery should be titrated carefully to avoid volume overload, which can cause pulmonary edema and worsen cardiac function. The medical team must consider the patient's hemodynamic status, urine output, and fluid requirements to maintain optimal fluid balance. Inotropic agents and vasopressors may be necessary to support blood pressure and cardiac output in SV patients who experience hypotension or reduced cardiac output. The choice of agent and dosage should be tailored to the patient's individual needs and response to therapy. The patient's lung function should also be carefully monitored during surgery. Supplemental oxygen may be required to maintain adequate SaO_2. The medical team should be alert for signs of hypoxemia, such as shortness of breath and cyanosis. Hypothermia can worsen cardiac function and increase the risk of complications in SV patients. The medical team must maintain the patient's body temperature by using warm blankets and warm fluids. Open communication and close collaboration between the anesthesiologist, surgeon, and the rest of the medical team are essential to ensure optimal SV patient care during surgery. Constant exchange of information and shared decision-making helps in identifying and addressing potential complications and improves surgical outcomes. Careful hemodynamic monitoring, appropriate fluid management, and judicious use of inotropic and vasopressor agents are key to maintaining physiological stability in SV patients during surgery. Open communication and close collaboration between the medical team are also important to achieve optimal results. With this comprehensive and individualized approach, the medical team can maximize the chances of successful surgery in these complex SV patients.[18,19]

The journey of a single ventricle (SV) patient does not end in the operating room. Intensive post-operative care is like a bridge that leads them back to health. Careful monitoring of physiological status and appropriate supportive interventions are key to ensuring optimal recovery. Vital signs are like a window that opens insight into the patient's physiological status. Blood pressure, heart rate, cardiac output, arterial oxygen saturation (SaO_2), and diuresis should be closely monitored to detect potential complications such as hypotension, arrhythmia, heart failure, hypoxemia, and renal failure. SV patients may require postoperative mechanical ventilation to assist their breathing. Ventilators help ensure adequate alveolar ventilation

and tissue oxygenation. Ventilator parameters, such as tidal volume, respiratory rate, and FiO_2 settings, must be adjusted to the patient's individual needs. Inotropic agents, such as dobutamine, may be necessary to support cardiac function in SV patients who experience postoperative hypotension or ventricular dysfunction. The dose and choice of inotropic agent should be tailored to the patient's individual needs and carefully monitored to avoid adverse side effects. Intravenous fluid therapy should be continued postoperatively to maintain hydration and organ perfusion. However, fluid balance must be carefully maintained to avoid volume overload and pulmonary edema. The patient's diuresis and hydration status should be closely monitored to guide adjustments in fluid intake. Postoperative pain can cause significant discomfort and interfere with recovery. Intravenous analgesics, such as morphine, can be used to control pain effectively. Multimodal analgesic techniques, such as regional nerve blocks and oral analgesics, may also be considered to provide optimal pain control. Venous thromboembolism and infection are common postoperative complications in SV patients. Venous thromboembolism prophylaxis with heparin or oral anticoagulant drugs and infection prophylaxis with prophylactic antibiotics may help reduce the risk of these complications. Early mobilization, such as getting out of bed and sitting up in bed as soon as possible after surgery, can help improve blood flow, prevent pulmonary complications, and speed recovery. Physical therapists can assist patients in developing a safe and effective mobilization plan. Surgery and recovery can be a stressful experience for SV patients and their families. Psychological support, such as counseling and group therapy, can help patients overcome stress, anxiety, and depression, and improve their quality of life. Intensive postoperative care is essential for optimal recovery of SV patients. Close monitoring of vital signs, mechanical ventilation when necessary, inotropic support, appropriate fluid therapy, effective pain management, prevention of complications, early mobilization, and psychological support are key elements in a successful recovery journey. Open communication between the medical team and patients and their families is critical to ensuring patient-centered care and optimal outcomes.[19,20]

4. Conclusion

Non-cardiac surgical procedures in patients with single ventricles are a unique challenge for anesthesiologists because they require a multidisciplinary approach from various fields of expertise, where we must maintain the physiological condition of the single ventricle physiology (SVP). The patent ductus arteriosus must be kept open to maintain systemic perfusion by maintaining PaO_2 at 40 to 45 mmHg and SaO_2 at 70% to 80%, along with the administration of prostaglandin agents. Good perioperative management and improvements in surgical procedures will increase the life expectancy of patients with single ventricle problems, especially hypoplastic left heart syndrome. The use of low-dose fentanyl induction agents, 1 MAC sevoflurane, and atracurium has been proven to yield favorable outcomes in this patient. In the future, cases involving patients with a single ventricle still require studies with larger samples to determine the appropriate combination of anesthetic agents for the patient's condition and surgery.

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