Double-outlet right ventricle is a rare congenital cardiac anomaly resulting in intracardiac mixing of oxygenated and deoxygenated blood. Surgical palliation involves staged conversion to Fontan circulation, with an intermediate stage using a Glenn shunt. We report the case of a patient at 36 weeks of gestation, with a partially palliated double-outlet right ventricle and a Glenn shunt, who presented with severe dyspnea and worsening cyanosis. After preoperative optimization, a combined spinal-epidural technique was successfully used for cesarean delivery. The anesthetic concerns and perioperative management of patients with complex cardiac physiology are discussed in this report. Carefully titrated combined spinal-epidural technique can be safe and effective for such cases.

**Keywords:** Obstetric anesthesia, congenital heart defect, epidural anesthesia, Glenn shunt.

Patients with double-outlet right ventricle (DORV) demonstrate a single-ventricle physiology, with pulmonary and systemic blood flow in parallel to one another, instead of in a series circuit. A patient with DORV with obstruction to pulmonary blood flow (pulmonary stenosis or atresia) usually presents with reduced blood flow to the lungs, leading to severe cyanosis. In contrast, patients with DORV without obstruction to pulmonary blood flow have pulmonary hyperemia and may present with congestive features or may eventually develop pulmonary hypertension and heart failure. Double-outlet right ventricle is a complex cardiac lesion without effective means of surgical correction. The aim of palliative surgery is to improve pulmonary blood flow and to convert the parallel circulation into a series circulation. Staged surgical palliation is usually undertaken. The first stage is a Glenn shunt or a hemi-Fontan procedure, in which the superior vena cava is anastomosed to the pulmonary artery, bypassing the heart. This procedure is usually undertaken by 6 months of age. The second stage is completion of the Fontan procedure, usually between 18 months and 4 years of age, whereby both the superior and inferior vena cava are diverted to drain directly into the pulmonary artery. The stages of palliation are shown in the Figure.

With the advent of sophisticated antenatal screening of fetal cardiac anomalies and greater access to cardiac surgical intervention, the number of patients with uncorrected or partially palliated complex heart defects is drastically reducing. Added to this, better awareness of the dangers of pregnancy in such patients and increased willingness to adopt family planning measures mean that such a patient presenting for obstetric management is exceedingly rare. We therefore feel it is worthwhile to report the successful management of such a case, occurring toward the end of pregnancy with severe cardiac decompensation.

**Case Summary**

A 26-year-old primigravida woman, with a singleton pregnancy, at 36 weeks of gestation, was referred to our hospital with the complaint of gradually increasing shortness of breath for the past 4 weeks. She had a history of cardiac surgery at 6 years of age, after which she had been poorly compliant to follow-up. No past medical records were available from the patient. On admission, she was dyspneic at rest (New York Heart Association functional class 4); cyanotic, with an oxygen saturation measured by pulse oximetry ($\text{SpO}_2$) of 78% on room air, blood pressure of 110/84 mm Hg, and heart rate of 110/min. Basal crepitations were present. Ultrasound evaluation revealed a reassuring biophysical profile of the fetus, with a Manning score of 8 of 10.

The patient was initially managed with humidified oxygen (60%) with continuous positive airway pressure (CPAP) using a cushion-sealed face mask, in a 45-degree propped-up position. Intravenous furosemide (40 mg immediately, followed by 20 mg every 12 hours) was ad-
ministered, and fluid intake was restricted to 1 L over 24 hours. Continuous electronic fetal monitoring was initiated. Tachypnea settled within 2 hours of CPAP therapy, and after 6 hours, she was able to comfortably maintain \( \text{SpO}_2 \) of 90% to 92% on oxygen by face mask at 5 L/min. Her condition continued to improve over the next 48 hours.

An echocardiogram revealed a DORV with a large subpulmonic ventricular septal defect and moderate pulmonary stenosis, with a patent bidirectional Glenn shunt. Ventricular function was normal (ejection fraction of 55%, no diastolic dysfunction). An electrocardiogram showed normal sinus rhythm with peaked p waves. Her hemoglobin concentration was 12.6 g/dL.

Once the patient had stabilized, a decision was made by the combined team of treating obstetricians and anesthesiologists to deliver the baby. Because the hemodynamic stress caused by the severe sympathetic stimulation during labor may have been detrimental to this patient, it was decided to proceed with an elective lower-segment cesarean delivery as opposed to induction of labor. After careful deliberation over the suitability of general anesthesia or regional anesthesia for this case, a combined spinal-epidural (CSE) approach, with low-dose intrathecal local anesthetic and opioid, along with a lumbar epidural catheter, was chosen. The patient was fasted for 8 hours before surgery. Sips of water were allowed for up to 2 hours before she was taken to the operating room. Ranitidine (150 mg) was administered orally the previous night and at 90 minutes before surgery. Intravenous amoxicillin-clavulanate (1.2 g) was administered 30 minutes before surgery for infective endocarditis prophylaxis. Blood products (4 units each of packed cells, platelets, and fresh-frozen plasma) were cross-matched and kept on standby.

The patient was kept in the left lateral position, with supplemental oxygen administered nasally, during transfer to the operating room and during the neuraxial block. Two wide-bore 16-gauge intravenous lines were secured and radial artery cannulation was performed under local anesthetic infiltration. Appropriate dilutions of atropine, esmolol, diltiazem, epinephrine, phenylephrine, norepinephrine, milrinone, and nitroglycerin infusions were prepared and primed. Etomidate and suxamethonium were kept on hand for emergent general anesthesia.

A CSE injection was performed at the L3-L4 interspace. Fluid coloading was performed with 300 mL of Ringer’s lactate to prevent a sudden fall in preload. The subarachnoid injection consisted of 7 mg of hyperbaric bupivacaine (0.5%) with 20 \( \mu \)g of fentanyl. The patient was then turned supine, maintaining a 15-degree leftward tilt. At 5 minutes the sensory block reached T8, after which a minimal (10-degree) Trendelenburg tilt was applied. Surgeons were allowed to start once the sensory block reached the T6 level (after another 2 minutes). Mean arterial pressure was maintained throughout the procedure between 70 and 100 mm Hg. There were 4 episodes of hypotension (defined as mean arterial pressure < 70 mm Hg), which were successfully treated with intermittent boluses of intravenous phenylephrine (50 \( \mu \)g). Intraoperative \( \text{SpO}_2 \) ranged between 85% and 90%. Episodes of a fall in \( \text{SpO}_2 \) corresponded with episodes of hypotension and responded promptly to phenylephrine.

A healthy female baby (birth weight of 2.5 kg; Apgar scores of 8 at 1 minute and 10 at 5 minutes) was delivered with gentle fundal pressure. An oxytocin infusion at 20 IU/h was started immediately after delivery and gradually reduced once uterine tone recovered. Blood loss was approximately 800 mL. One unit of packed red blood
cells was transfused. Bilateral tubal ligation was done with the consent of the patient. Epidural supplementation with 3-mL boluses of 0.5% bupivacaine was started once the block height began receding. At the end of the procedure, 3 mg of morphine was given via the epidural catheter, after which the patient was transferred to the intensive care unit.

Humidified oxygen via face mask and oxytocin infusion were continued for the first 24 hours. Postoperative analgesia was maintained with intravenous paracetamol and epidural morphine (3-mg boluses every 12 hours). Epidural local anesthetic was omitted to avoid any sudden, unexpected hemodynamic alterations. The postoperative course was uneventful. The epidural catheter was removed on the third postoperative day. Written consent was obtained from the patient for the purpose of this report.

Discussion

Pregnancy complicates cyanotic heart disease as the decrease in systemic vascular resistance (SVR) worsens the right-to-left shunt. The effect is further compounded by neuraxial anesthesia for cesarean delivery. Pregnancy in patients with cyanotic heart disease is associated with a more than 30% incidence of maternal cardiovascular complications. Maternal oxygen saturation less than 85% may be predictive of increased risk.

The principal anesthetic concern in the patient described in this case report involved the effect of anesthesia on the intracardiac resistance-dependent bidirectional shunt. The status of this patient was unique because the hemi-Fontan stage is a combination of both Fontan physiology (univentricular system in which forward flow is aided by maintaining low total vascular resistance) and Fallot physiology (intracardiac shunt with shunt fraction dependent on the relative ratio of pulmonary and SVR). A sudden increase in SVR could lead to acute failure of the single ventricle and a decrease in cardiac output. On the other hand, a sudden fall in SVR or rise in pulmonary vascular resistance (PVR) could lead to a catastrophic increase in right to left shunting and hence cyanosis. Arrhythmias are poorly tolerated because adequate filling of the functionally single ventricle is largely dependent on atrial function. Therefore, hemodynamic goals in such a patient are to maintain adequate intravascular volume and sinus rhythm to ensure good ventricular filling, maintain myocardial contractility by avoiding use of agents with the potential for myocardial depression, and avoid any sudden fall in SVR or rise in PVR.

Neuraxial anesthesia was preferred in this case because it avoids the risks involved with general anesthesia for cesarean delivery. This includes the possibility of a catastrophic increase in PVR due to intubation response or positive pressure ventilation. The CSE technique is a suitable choice for such patients because the lower dose of subarachnoid local anesthetic required in CSE, compared with single-shot spinal epidural, ensures a lesser degree of sympathetic block, thus avoiding sudden hemodynamic fluctuations. The epidural catheter allows titration of additional local anesthetic, if required, and provides a route for excellent postoperative analgesia. Careful fluid coloading, prevention of aortocaval compression, and timely replacement of losses with fluids and blood products is essential.

Earlier case reports have described the successful management of parturients with DORV undergoing cesarean delivery using CSE. These reports, however, deal with patients whose cardiac morphologies follow Fallot physiology. Our case, being partially palliated with a Glenn shunt, is a unique combination of Fallot and Fontan physiology, making its anesthetic management more challenging. Wendling et al reported a case of tricuspid-atresia with a Glenn shunt for elective cesarean delivery. They preferred pure epidural anesthesia in their case. Catarci et al described the management of a partially palliated case of transposition of great vessels with a Glenn shunt, for urgent cesarean delivery, using a CSE technique. Although a carefully titrated epidural potentially offers the greatest hemodynamic stability, the CSE technique provides faster and more reliable block onset and is therefore invaluable in cases where time is critical.

Patients with cyanotic heart disease need to maintain a higher hemoglobin concentration to compensate for the large fraction of desaturated hemoglobin. Hence, it is prudent to set a lower threshold for blood transfusion. In our case, for example, a preoperative hemoglobin level of 12.6 g/dL and saturation of 84% effectively meant only 10.6 g/dL of saturated hemoglobin. We transfused 1 unit of packed red blood cells to compensate for the loss of 800 mL of blood, with the aim of maintaining an effective saturated hemoglobin concentration above 10 g/dL. Postpartum hemorrhage can be catastrophic in such patients. Prophylaxis with oxytocin is essential. Methylergometrine and carboprost must be avoided because they may cause severe pulmonary vasconstriction, worsening cyanosis.

Other anesthetic concerns in patients with complex cyanotic heart disease include a high risk of infective endocarditis, hepatic dysfunction due to chronic congestion, and deranged kidney function due to glomerulosclerosis from chronic hypoxia. Patients with partial or complete cavopulmonary bypass may have hypoalbuminemia due to protein-losing enteropathy caused by the chronic high venous pressure. A high risk of paradoxical embolism mandates meticulous de-airing of intravenous lines. These patients are prone to hypercoagulability due to venous stasis, polycythemia, and hyperviscosity. It is preferable to avoid central venous cannulation in such patients because it predisposes to the risks of catastrophic shunt thrombosis, paradoxical embolism, and infective endocarditis.
In conclusion, this case report describes the successful use of the CSE technique for cesarean delivery in a rare, partially palliated case of DORV with pulmonary stenosis in the hemi-Fontan stage. Each patient with a complex cardiac defect is unique and requires an anesthetic plan that is carefully tailored to his or her cardiac morphology and physiology. The goal is to maintain perioperative hemodynamics as close to the patient’s own stable preoperative status as possible. This involves meticulous attention to individual physiologic parameters as well as to the global condition—sensorium, urine output, and patient comfort.

REFERENCES

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The authors have declared no financial relationships with any commercial entity related to the content of this article. The authors did not discuss off-label use within the article.