Double-outlet right ventricle (DORV) is a rare cardiac condition in which both the aorta and pulmonary artery arise from the right ventricle, resulting in parallel systemic and pulmonary circulations. Usually, DORV is present with ventricular septal defect; however, the location of the ventricular septal defect and presence of pulmonary stenosis (PS) result in various physiological features and subtypes of DORV. Because DORV without PS causes congestive cardiac failure and DORV with PS results in cyanotic heart disease, anesthesia management varies widely according to the resultant physiological characteristics. Reports of anesthesia management in a parturient with DORV undergoing cesarean delivery is scarce because of the low incidence of DORV and the discouragement of these patients to conceive. Only 8 known previous such cases are reported, and almost all these patients were administered regional anesthesia. Here we describe a parturient with DORV, to whom general anesthesia was administered because of incidental antiphospholipid syndrome with low platelets. To the best of our knowledge, this scenario has not been described previously.

Keywords: Anesthesiology, cardiac, double-outlet right ventricle, pulmonary stenosis.

In a normal heart, pulmonary artery arises from the right ventricle, which contains deoxygenated blood, and the aorta arises from the left ventricle, which contains oxygenated blood. Oxygenated blood from the left ventricle is carried via the aorta to the whole body. Double-outlet right ventricle (DORV) is a rare cardiac condition with a probable incidence of less than 1 in 3,000. In DORV, both the aorta and the pulmonary artery arise from the right ventricle. In this congenital heart condition, as the aorta arises from the right ventricle (which contains deoxygenated blood), a ventricular septal defect (VSD) is also present to allow oxygenated blood of the left ventricle to pass to the right ventricle, from where the aorta carries this mixed blood to the whole body. Although not optimally oxidized, the mixed blood helps in survival of these patients. Pulmonary stenosis (PS) is another important factor of cardiac physiology in DORV. If PS is not present in DORV, too much blood flows to the heart through the pulmonary artery and results in congestive cardiac failure. On the other hand, with presence of PS, passage of blood through the pulmonary artery is restricted, resulting in cyanosis.

To improve survival, oxygenated blood to the right ventricle is maximized by a series of palliative surgical procedures undertaken from an early age with the aim to establish Fontan circulation. In Fontan circulation, the systemic venous return is connected to the pulmonary arteries without the interposition of an adequate ventricle. Its advantages are near-normalization of the arterial saturation and abolishment of the chronic volume overload so that cardiac output is no longer determined by the heart but by transpulmonary flow, which in turn depends on pulmonary vascular resistance (PVR). For establishing Fontan circulation, the first surgical procedure is usually a Glenn shunt, in which the superior vena cava is anastomosed to the pulmonary artery (Figure 1). With advancement in palliative surgery, the average survival has improved to a normal life expectancy. However, because of the low incidence of DORV and the discouragement of these patients from getting pregnant, anesthesia management in a parturient with DORV undergoing cesarean delivery is scarce.
The anesthetic goal in a parturient with DORV is to maintain Fontan circulation, primarily established by increased cardiac preload, the driving force of forward blood flow. Neuraxial techniques because of associated sympathectomy decrease the preload and thus may not be ideal for cesarean delivery in these patients. On the other hand, general anesthesia may not decrease preload but can be detrimental to Fontan physiology because of associated sympathetic stimulus at endotracheal intubation, decreased venous return, preload, and oxygenation with positive pressure ventilation and anticipated difficult airway of a parturient. Thus, almost all published reports of patients with DORV undergoing cesarean delivery demonstrate they were given regional anesthesia and had a favorable outcome because of the gradual establishment of neuraxial blockade and perioperative maintenance of higher preload.6-12

Antiphospholipid syndrome (APS) is an autoimmune, hypercoagulable state caused by antiphospholipid antibodies and is often present with low platelet counts.13 To the best of our knowledge, APS has previously not been reported in a patient with DORV. We describe such a patient who received general anesthesia for cesarean delivery because of PS with DORV and subnormal platelet count.

Case Summary
A 23-year-old woman, 50 kg in weight, with a complicated obstetric history of gravida 5, para 1, aborta 4, was scheduled for an emergency lower-section cesarean delivery at 34 weeks because of absent end-diastolic flow in the fetus. She received a diagnosis of DORV in childhood and was operated on 11 years earlier with a Glenn shunt. In the present pregnancy, APS was diagnosed as the cause of her previous multiple miscarriages, and for this condition, subcutaneous enoxaparin sodium, 40 units once a day, was prescribed from the second trimester. She was also receiving oral supplements of iron, calcium, L-arginine, and aspirin, 75 mg, once a day. She reported for regular antenatal checkups, was asymptomatic until the 29th week of gestation, and was managed conservatively with oral tablets of furosemide, 20 mg.

The patient was scheduled for elective cesarean delivery at 34 weeks of gestation. On preanesthesia history, the patient complained of marked limitation in daily activities and was comfortable only at rest (New York Heart Association grade 3). She had cyanosis, pedal edema, and clubbing of the toes (Figure 2) and of the fingers (Figure 3). On auscultation, a systolic murmur was heard over the pulmonic valve area. Her present peripheral oxygen saturation ($SpO_2$) in room air was 76% to 82%. Electrocardiograms showed a normal sinus rhythm. Echocardiograms revealed a DORV with a large subaortic VSD, a large atrial septal defect (20 mm) with a bidirectional shunt, severe valvular PS, mild tricuspid regurgitation, trivial aortic regurgitation, a Glenn shunt with normal laminar flow (peak $\Delta P$, 65 mm Hg), and normal biventricular function. Preoperative arterial blood gas analysis revealed the following values: pH, 7.40; $PCO_2$, 24.9 mm Hg; $PO_2$, 50.2 mm Hg; $SpO_2$, 85%; hematocrit, 52%; lactate, 1.5 mmol/L; and bicarbonate, 15.7 mmol/L. Biochemistry laboratory reports revealed a hemoglobin level of 16 g/dL and platelet count of 88,000/mm$^3 (x10^3/\mu L)$. Liver function and renal function test results were within normal limits.

Before transport to the operating room (OR), the patient was given intravenous ampicillin, 2 g; gentamicin, 80 mg; ranitidine, 50 mg; and metoclopramide, 10 mg, through a 20-gauge intravenous cannula, which was already inserted on the ward. In the OR, monitors of electrocardiography, noninvasive blood pressure, and
SpO2 were attached, which showed a heart rate of 62/min, blood pressure of 140/90 mm Hg, and SpO2 of 84%. In the OR with the patient under local anesthesia, an 18-gauge intravenous cannula was secured in the dorsum of the left hand, an arterial line was inserted in left radial artery, and a 7F central line was inserted in the right internal jugular vein. The following drugs were kept ready: etomidate, ketamine, succinylcholine, fentanyl, vecuronium, oxytocin, lignocaine, diltiazem, and phenylephrine. The base of the tongue and the posterior pharyngeal wall were sprayed with 10% lignocaine spray. The patient was optimally positioned with a pillow under her shoulder and a left tilt of the OR table. Preoxygenation was done with 100% oxygen for 3 minutes. Simultaneously, obstetricians disinfected the surgical site with povidone-iodine (Betadine) and alcohol-based disinfectant (Sterillium) and draped the patient with a sterile drape.

After 3 minutes, anesthesia was induced with gradual administration of intravenous etomidate, 0.2 mg/kg (12 mg), and ketamine, 1 mg/kg (50 mg), over 1 minute. Cricoid pressure was applied (the patient was educated about pressure on the neck before induction). One minute after intravenous succinylcholine, 1 mg/kg (50 mg), was injected, a senior anesthetist intubated the trachea with a cuffed endotracheal tube size 7 mm within 10 seconds. Heart rate and blood pressure remained within 20% of baseline at anesthesia induction and endotracheal intubation. The operation was then commenced. Anesthesia was maintained with oxygen in air (50% + 50%) and 1.5% isoflurane. Muscle relaxation was achieved with intermittent boluses of intravenous vecuronium, 0.02 mg/kg. A tidal volume of 7 to 8 mL/kg and respiratory rate of 10/min, with no positive end-expiratory pressure (PEEP), was set on the ventilator, maintaining end-tidal carbon dioxide (ETCO2) between 34 and 38 mm Hg. Peak airway pressure was 14 cm H2O. Estimated blood loss was less than 150 mL. Intraoperatively the patient's heart rate and mean arterial pressure remained stable in the range of 61/min to 82/min and 68 to 84 mm Hg, respectively. The SpO2 values remained in the range of 91% to 94%, and central venous pressure remained between 18 and 21 cm H2O. The surgical procedure lasted 1 hour. At the end of the operation, intravenous fentanyl, 2 μg/kg, and midazolam, 0.02 mg/kg, were injected, and a bilateral transversus abdominis plane (TAP) block was given with 20 mL of 0.25% ropivacaine and 8 mg of dexamethasone on each side under ultrasound guidance. Neuromuscular blockade was reversed with neostigmine (0.05 mg/kg) and glycopyrrolate (0.01 mg/kg), and the trachea was extubated. The patient was pain free at the end of the surgical procedure, and her vital signs remained within normal limits.

Postoperative analgesia was maintained with intravenous acetaminophen (paracetamol), 20 mg/kg 4 times a day. The patient's stay in the hospital was uneventful. The patient and her baby were discharged on the seventh postoperative day with advice to follow up in the cardiology department. Patient permission was obtained for reporting, including publishing of photographs.

**Discussion**

In a normal heart, the pulmonary artery carries deoxygenated blood from the right ventricle to the lungs, and the aorta carries oxygenated blood from the left ventricle to the rest of the body. Double-outlet right ventricle is a rare form of congenital heart disease with a low incidence of 1 in 3,000.4 In DORV, both great vessels arise from the right ventricle. For survival, the aorta has to carry oxygenated blood; thus, a VSD is co-present in these patients, which enables oxygenated blood of the left ventricle to mix with the deoxygenated blood in the right
ventricle. Apart from the VSD, other heart conditions that might be present in DORV are PS, transposition of the great arteries, pulmonary atresia, coarctation of the aorta, and mitral valve abnormalities. Pulmonary stenosis is an important factor that determines blood flow to the lungs.\(^1\,^2\) A patient with DORV with obstruction to pulmonary blood flow (PS or atresia) usually presents with reduced blood flow to the lungs, leading to cyanosis. In contrast, patients with DORV without obstruction to pulmonary blood flow have pulmonary hyperemia and may present with congestive features, eventually developing into pulmonary hypertension and heart failure.\(^1\,^2\)

Double-outlet right ventricle is a constellation of abnormalities, with various classifications of DORV described in the literature.\(^10\) Traditional classification is according to the location of VSD: subaortic, subpulmonic, doubly committed, or noncommitted.\(^3\) Classification by the Society of Thoracic Surgeons divides DORV according to the resultant physiological characteristics.\(^3\) To facilitate administration of anesthesia, this classification may be simpler to comprehend the specific physiological consequences of each subtype of DORV.

- VSD type: In this, DORV is present with subaortic or doubly committed VSD and no PS. Anesthetic management is similar to the child with a large VSD.
- Tetralogy of Fallot type: This DORV type is characterized by either a subaortic or doubly committed VSD and PS. The clinical presentation and anesthesia management is similar to that for tetralogy of Fallot.
- Transposition of great arteries type: This type of DORV is characterized by a subpulmonary VSD with or without PS. These patients usually present with cyanosis and physiological features similar to the patient with transposition of the great arteries/VSD.
- Remote VSD type: This type of DORV is characterized by a remote VSD with or without PS. The VSD is usually
located in the muscular septum and is distant from both the pulmonary artery and aorta. Anesthetic management is similar to that of a patient with a large VSD.

- DORV and atrioventricular septal defect: This type of DORV is characterized by an atrioventricular septal defect and may be accompanied by heterotaxy, right ventricular outflow tract obstruction, and total anomalous pulmonary venous return. In these patients, the presence of PS causes cyanosis and without PS, cardiac failure is present. Thus anesthesia management is accordingly.

In pregnancy, physiological changes such as increased blood volume, decreased hematocrit, decreased systemic vascular resistance, decreased PVR, increased heart rate, and increased cardiac output occur. To accommodate these changes in a parturient with normal cardiovascular anatomy and function, the anesthesia provider easily can increase the cardiac output by increasing stroke volume and heart rate. But in a parturient with DORV, acclimating to the aforementioned changes is frequently not possible because stroke volume is dependent on the weaker right ventricle and the pulmonary venous circulation is not able to accommodate the increased preload of pregnancy. This increases the pulmonary venous pressure and may result in heart failure, even though a decrease in PVR with pregnancy is a favorable physiological factor in a DORV-affected parturient.

Preload, and its effect on PVR, is the single most important determinant in maintaining cardiac output in these patients. For anesthesia in a parturient with DORV, the goal is to maintain Fontan circulation by keeping an increased preload, which is the driving force of forward blood flow. Neuraxial techniques cause sympathectomy, which decreases the preload and thus may not be ideal for cesarean delivery in these patients. However, of the 8 published reports on anesthetic management of parturients with DORV scheduled for cesarean delivery, almost all patients were administered regional anesthesia5-12 (Table). Of these, 3 patients were given graded epidural anesthesia; 2, combined spinal epidural anesthesia; and 1, a subarachnoid block.6-12 All had a favorable outcome, attributed to gradual establishment of neuraxial blockade and increased preload. General anesthesia for cesarean delivery has previously been administered in 1 asymptomatic parturient with DORV who underwent a Fontan procedure, but unlike the present patient, that woman had normal cardiac physiological findings. The report gives no details of anesthetic drugs.5

General anesthesia is challenging in parturients with DORV and can be detrimental to Fontan physiology because of (1) associated sympathetic stimulus at endotracheal intubation; (2) decrease in venous return, preload, and oxygenation with positive pressure ventilation, which causes an increase in intrathoracic pressure, hampering flow from the superior vena cava to the pulmonary arteries; (3) hypotension due to negative inotropic effects of general anesthetics; and (4) potential difficult airway of a parturient.

Despite the challenges of general anesthesia in patients with DORV, it was favored in the present patient for various reasons. First, the patient had cyanosis, for which the anesthetic goals are prevention of a fall in SVR and prevention of an increase in the right to left shunt, both of which are a challenge with neuraxial blocks. Gradual establishment of the neuraxial block with a combined spinal epidural technique or a pure epidural anesthesia was considered; however, the platelet count was below normal, and the report available was a day old. Regional anesthesia has been safely undertaken in patients even with a platelet count of 50,000/mm3. However, a spinal hematoma with regional anesthesia was previously reported in a parturient with DORV who had a normal platelet level of 1.73,000/mm3 and strict adherence to guidelines. The cause of low platelet count in the present patient was the incidental presence of APS.

General anesthesia was undertaken in the present patient with several precautions. At induction of anesthesia, hypotension and a fall in PVR were avoided by using a combination of ketamine and etomidate. A phenylephrine infusion was kept ready in the OR to treat hypotension. Sympathetic stimulation at endotracheal intubation was prevented by spraying the oral cavity with lignocaine spray, and a senior anesthetist performed the endotracheal intubation in less than 10 seconds to prevent an increase in PVR. All factors known to increase PVR were avoided, such as aortocaval compression, hypoxia, hypercapnia, unusually high or low lung volumes during positive pressure ventilation, high levels of PEEP, and high mean airway pressures to prevent a decrease in pulmonary blood flow, which may decrease cardiac output. Intraoperative hypoxia was prevented by administration of a high fraction of inspired oxygen of 0.7 with SpO2 between 91% and 94%. Hypercarbia was prevented by maintenance of ventilatory parameters to maintain ETCO2 around 35 mm Hg. Acidosis was prevented by avoidance of hypercarbia through ventilation and checking of pH in arterial blood gases, especially monitoring serum lactate levels (for peripheral hypoperfusion). PEEP was not administered to prevent deterioration of PS and nitrous oxide was avoided to prevent an increase in PVR. Adequate intravascular volume was maintained to ensure good ventricular filling. Blood and blood products were kept ready for the patient. Oxytocin was administered in a low dose, followed by infusion to prevent hypotension. Methylergometrine and carboprost were avoided because they cause severe pulmonary vasoconstriction and worsen cyanosis. The patient’s core oral temperature was maintained at more than 36°C to avoid vasoconstriction secondary to hypothermia. We ensured that no air bubbles were present in the tubing of the intravenous line.
In conclusion, general anesthesia may safely be administered in a parturient with DORV and PS scheduled for cesarean delivery, by careful selection of anesthetic drugs to maintain increased cardiac preload and prevent an increase in PVR. An understanding of the physiologic characteristics of DORV is key for successful anesthesia because each patient presents with unique physiological features and challenges.

REFERENCES


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