Perioperative Management of a Parturient After Atrial Switch Surgery for Dextro-Transposition of the Great Vessels

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DEXTRO-TRANSPOSITION OF THE great arteries (d-TGA) is a congenital cyanotic heart disease with abnormal connections of the great vessels (Fig 1A).[1] With d-TGA, there is atrial situs solitus, D-looped ventricle, dextroposition of the aorta (ie, the aorta is right and anterior to the pulmonary artery), atrioventricular concordance, and ventriculoarterial discordance.[2] Atrioventricular concordance indicates the morphologic right atrium (RA) and left atrium (LA) are connected to the morphologic right ventricle (RV) and morphologic left ventricle (LV), respectively. Ventriculoarterial discordance means the aorta arises from the RV, and the pulmonary artery arises from the LV. The atrioventricular valves, the tricuspid and mitral valves, follow the morphologic RV and LV, respectively. The semilunar valves, the aortic and pulmonary valves, follow the aorta and pulmonary artery, respectively.[2] Before corrective surgery, oxygen delivery depends on the presence of intracardiac shunt via atrial septal defect, ventricular septal defect, or patent ductus arteriosus.

Senning[3] described the first successful surgical palliation for d-TGA. He created a baffle for diverting blood flow from the vena cava into the morphologic LA using autologous tissue and implanted the pulmonary veins into the RA. This correction allowed the morphologic RV to receive oxygenated blood and supply the systemic circulation via the aorta. Mustard et al[4] later described an alternative by performing atrial septectomy and using pericardium to create a baffle to divert the blood flow (Fig 1B). Both surgeries resulted in >80% survival over 20 years,[5–8] allowing girls born with d-TGA to reach childbearing age.

The authors describe successful perioperative management of such a parturient status post-Mustard procedure, undergoing vaginal delivery followed by tubal ligation.

CASE PRESENTATION

A 29-year-old woman (gravida 2, para 1), born with d-TGA, status post-Mustard procedure, presented for elective induction of labor at 39 weeks' gestation. She had undergone Mustard surgery as definitive palliation at the age of 9 months without further surgeries. Her medical history was significant for sinus node dysfunction not requiring a pacemaker (baseline normal sinus rhythm) and nonsustained atrial tachycardia. She had New York Heart Association (NYHA) class 1 functional status. She had not required any chronic medical therapy so far in life for her cardiovascular condition.

A transthoracic echocardiogram (TTE) performed 1 month prior to scheduled induction demonstrated the superior vena cava and inferior vena cava surgically baffled to the LA and pulmonary veins baffled to the RA without obstruction. The morphologic LV was qualitatively normal in appearance and supplied the pulmonary circulation. The morphologic RV, supplying the systemic circulation, was dilated and hypertrophic with mild systolic dysfunction, based on qualitative and quantitative analysis. Echocardiographic images are shown in Figure 2 and Video Clip 1.

The patient's delivery plan, as agreed upon by obstetrics, anesthesiology, and the adult congenital heart disease (ACHD) team, was for vaginal delivery using patient-controlled epidural analgesia followed by tubal ligation. The authors' institution is a university medical center with all of the aforementioned teams available around the clock. This delivery plan was formulated based on mild systolic RV dysfunction with adequate Mustard repair and NYHA class 1 functional status. The patient's adequate repair, RV function, and functional status allowed the authors to manage her as a high-risk delivery, but without the need for invasive hemodynamic monitoring. The hemodynamic goals were to avoid an increase in systemic vascular resistance with adequate pain control and to maintain euvolemia to avoid right ventricular overload. Continuous telemetry was used to monitor for arrhythmia.

Epidural placement was uneventful, and 3 mL of 1.5% lidocaine with epinephrine 1:200,000 were used as the test dose. No fluid bolus was administered prior to epidural placement to avoid fluid overload. Fluid status was monitored using clinial symptoms, noninvasive blood pressure, and urine output. Labor progressed uneventfully, and the infant was delivered without complication. The authors performed the tubal ligation 2 hours after the delivery to facilitate use of the epidural catheter for surgical anesthesia. In the operating room, the authors performed a controlled bolus of the epidural catheter with lidocaine 2% with epinephrine 1:200,000 in small-volume (2-3 mL) incremental doses to minimize hemodynamic changes. A total of 15 mL of solution were administered over a period of 30 minutes to achieve a dense surgical block to a dermatomal level of T7. Noninvasive monitoring was performed, and the patient was positioned her with arms out at 90° for additional arterial and venous access if necessary. A cardiac anesthesiologist supervised the case with immediate availability of echocardiography, in the event that the clinical situation warranted it. The patient underwent uneventful tubal ligation and was discharged home 2 days later. Follow-up visits revealed no cardiac complications.

DISCUSSION

Cardiovascular Changes During Pregnancy and Complications Associated With d-TGA

Cardiovascular changes during pregnancy include 45% to 50% increase in blood volume and 40% to 50% increase in cardiac output. During labor, cardiac output may increase further by 25% to 50%, and an additional 15% to 30% during contractions.[9] The morphologic RV supplies systemic circulation. Therefore, increased blood volume and cardiac output

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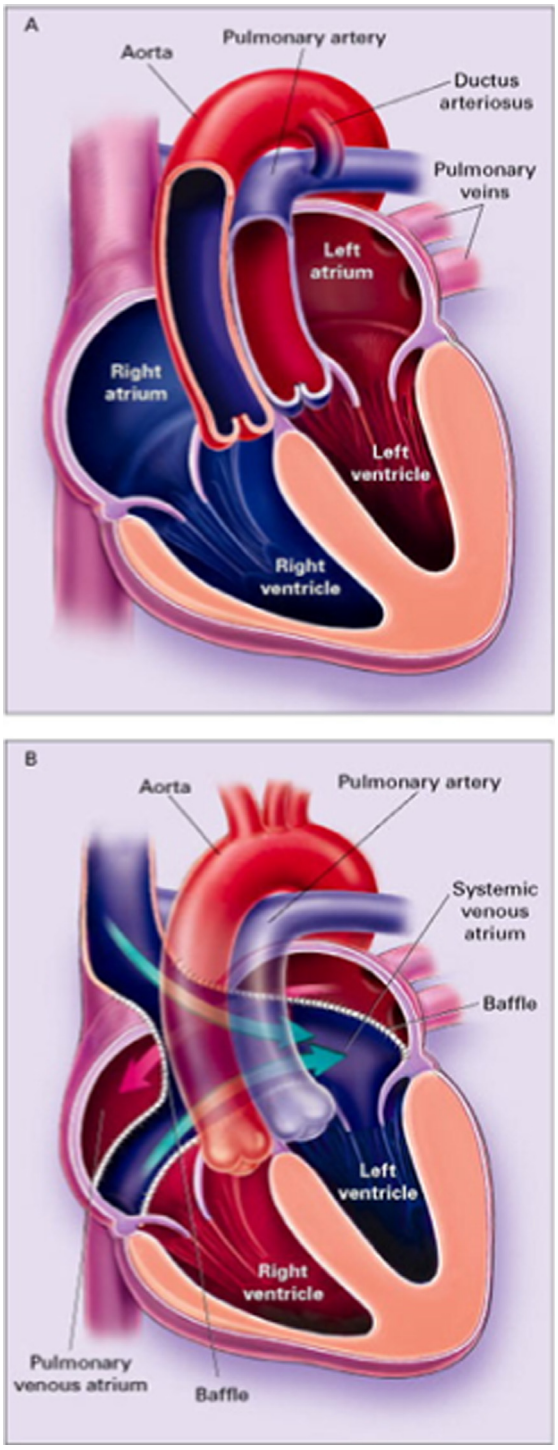

Fig 1. Transposition and atrial switch of the great arteries. (A) The anatomic relationship in d-TGA prior to surgical correction. (B) The results of atrial switch via the creation of a baffle. d-TGA, dextro-transposition of the great arteries. Reproduced with permission from Brickner et al.[1] Copyright Massachusetts Medical Society.

place considerable risk for RV failure,[10] as demonstrated in the literature review summarized in Table 1. Pregnancy after atrial switch usually is well tolerated, but it is associated with a risk of deterioration in RV function and a small but permanent reduction in functional class.[24]

Arrhythmias, including supraventricular tachycardia and sinus node dysfunction, also are common following atrial switch surgery.[25,26] Beta-blockers appear to be protective against life-threatening arrhythmias and should be considered in surgically corrected d-TGA patients with known risk factors for sudden cardiac death (eg, history of arrhythmias, RV dysfunction, and QRS prolongation).[25] Electrical cardioversion is safe and can be used for hemodynamic instability.[9] Baffle obstruction, leak, and sudden cardiac death are other known complications of atrial switch surgery.[5,7,8,27]

Because of the aforementioned complications, delivery of such patients should be considered high risk and carried out in a specialized medical center with expertise in managing ACHD patients. At the authors' medical center, management of such patients is a collaborative effort of anesthesiologists, ACHD specialists, and obstetricians, with services available around the clock.

The goals of anesthetic management are determined based on oxygen saturation demonstrating adequacy of surgical repair, the presence of RV function/dilation, functional status of the individual, presence of arrhythmias, and any baffle-associated complication. The goals are geared toward maintaining adequate oxygen delivery, avoiding increase in afterload against which systemic RV is working by providing adequate pain control, maintaining euvolemia, and monitoring and treating arrhythmia. Clinical cardiovascular decompensation, such as dyspnea, arrhythmias, or pulmonary edema, necessitate admission for bed rest. It is recommended to proceed with delivery under these circumstances if gestational age allows.[9] Cesarean section under general anesthesia with invasive hemodynamic monitoring should be considered in face of clinical decompensation.

Cardiovascular Monitoring

All women with d-TGA should undergo cardiac assessment, including TTE, early in pregnancy to determine baseline status.[25] Serial follow-up is essential and should include follow-up during the peak hemodynamic period (28-32 weeks' gestation) as well as postpartum. Clinical evaluation includes current functional status (NYHA classification), signs of venous congestion, and ejection and regurgitant systolic murmur, indicating subpulmonic outflow obstruction and tricuspid regurgitation, respectively. Electrocardiogram may show RV hypertrophy, escape rhythm, atrial flutter, or other arrhythmias. Automatic implantable cardioverter-defibrillator (AICD) or antiarrhythmia drugs should be used as needed. The evaluation of systemic RV is achieved best using cardiac magnetic resonance imaging (CMR).[28,29] CMR has been used during pregnancy without gadolinium contrast.[30]

However, TTE remains the first-line diagnostic modality to assess ventricular function and the degree of tricuspid regurgitation.[31] Contrast echocardiography is useful in demonstrating baffle obstruction or leak.[28] RV fractional shortening, RV ejection fraction (RVEF), fractional area of change (FAC), tricuspid annular plane systolic excursion (TAPSE), rate of systolic RV pressure increase (dP/dt), peak systolic tissue