CASE REPORT

Complex congenital heart disease in a complicated and precious pregnancy

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SUMMARY

A single ventricle is a rare congenital heart disease that accounts for less than 1% of all congenital heart diseases. A woman was assessed in our obstetric clinic for the first time at the gestational age of 28 weeks and found to have placental bleeding. She also had complex congenital heart disease and atrial fibrillation requiring anticoagulation. Echocardiography revealed double-inlet single ventricle with right and left atrioventricular valves entering into this chamber and levo-transposition of the great arteries. After an extensive discussion with the patient regarding the risks and benefits of anticoagulation including risk of stroke, the agreed plan was to start her on intravenous heparin with close observation and to continue pregnancy for at least 32 weeks in order to reduce the postpartum risk for the fetus. The pregnancy progressed without any further complications and the patient had elective caesarean section at 33 weeks of gestation and delivered a healthy baby boy.

BACKGROUND

To the best of our knowledge, this is the first case of pregnancy in an individual with a single ventricle, which poses a high risk of thromboembolism with double-inlet right ventricle (RV) and intact inferior vena cava flow, and atrial fibrillation requiring anticoagulation, complicated by placental bleeding.

CASE PRESENTATION

A 22-year-old woman presented to our obstetric clinic for the first time on 2/9/2014, with a history of having her last menstrual period occurring 7 months earlier. She was married 1 year prior and this was her first pregnancy. Obstetric ultrasound showed a viable male fetus with a gestational age of approximately 28 weeks, with no obvious congenital malformation. There was a 13×6.5 cm heterogeneous, hypoechoic area with some debris seen in the outer part of the placenta. The radiologist's opinion was that the hypoechoic area likely represented placental bleeding leading to a haematoma formation (figure 1). On being asked for her detailed history, the patient said that she was having regular follow-up for her cardiac problem in another hospital. Her medical report from that hospital revealed that she had complex congenital heart disease and atrial fibrillation.

According to the report, the patient had a univentricular heart with both atria connecting to the morphological RV as main chamber and levo-transposition of the great arteries (L-TGA). She had cardiac surgery twice during early childhood.

During her first surgery at the age of 3 months, in 1992, she had an atrial septostomy and Blalock-Taussig shunt. She had her second surgery at the age of 7 years, where a Glenn procedure was carried out. Owing to her atrial fibrillation with complex cardiac disease, she was being treated with warfarin as her regular anticoagulation. Her warfarin was switched to half therapeutic dose of enoxaparin when she became pregnant.

INVESTIGATIONS

An urgent transthoracic echocardiography was performed, which revealed a univentricular heart with morphological RV as main chamber and an L-TGA. The left ventricle was rudimentary and was located posterior to the RV. RV ejection fraction was about 35–40%. The aorta arose from the main chamber (RV). There were no signs of aortic stenosis. Trace aortic regurgitation was seen. The pulmonary artery was posterior to the aorta and arose from the side of the rudimentary left ventricle (figure 2).

There was severe valvular and subvalvular pulmonary stenosis with some forward flow. The RV, which was acting as the main chamber, had a double-inlet with the right and left atrioventricular (AV) valves entering into it with no obstruction (figure 3). Both valves had trace to mild regurgitation. The aortic arch was right sided. There was a continuous flow through the previous Blalock shunt (systemic pulmonary shunt). The previously performed Glenn shunt was patent with good flow. The next day, an obstetric ultrasound was repeated, which revealed no change in the size or nature of the hypoechoic, most likely representing the bleed.

DIFFERENTIAL DIAGNOSIS

The single ventricle differential includes mainly hypoplastic left ventricle, tricuspid atresia and, to a lesser extent, as in our case, double-inlet RV.

Figure 1 Ultrasound showing placental haematoma.

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Figure 2 Right ventricle as the main chamber for inlet and outflow to aorta.

TREATMENT

Owing to stasis, endothelial dysfunction and other factors, single ventricle patients are prone to thromboembolism. In our case, the patient was already at high risk due to atrial fibrillation, a structurally abnormal heart and the free mixing of the left and right circulation with its risk of systemic embolisation. She was on long-term warfarin, followed by enoxaparin; subsequently, due to the placental haematoma, this was replaced by intravenous heparin infusion, until the carrying out of the caesarean section (C-section), after which, when it was safe, warfarin was resumed. Her treatment also included bed rest until her elective C-section at 33 weeks of gestation.

OUTCOME AND FOLLOW-UP

Pregnancy progressed without any further complications and the patient had elective C-section at 33 weeks of gestation. Inspection and histopathology of the placenta confirmed bleeding into the placenta.

The patient was discharged from hospital after 3 days. The baby boy was placed in the neonatal intensive care unit for 3 days, where he remained stable, and was then transferred to the nursery ward, from where he was discharged home after a week.

We scheduled a 2-week follow-up with the mother after discharge. She was doing fine and her anticoagulation was within therapeutic range.

Figure 3 Flow from left atrium to morphological right ventricle.

DISCUSSION

A single ventricle is a rare congenital heart disease that accounts for less than 1% of all congenital heart diseases. Optimal physiology of the univentricular heart requires good ventricular function without AV valve regurgitation, an unrestrictive atrial septal defect–ventricular septal defect and well-balanced systemic and pulmonary blood flow.^{1 2}

This is currently accomplished by an aortopulmonary shunt, such as a modified Blalock-Taussig shunt, or bidirectional cavopulmonary anastomosis (Glenn shunt). A Fontan procedure, which separates the pulmonary circulation from the systemic circulation, is completed sometime between 18 months and 4 years of age.³ These patients have multiple problems, and pregnancy is challenging, with increasing orthopnoea and a higher thromboembolic risk.⁴

There are case reports that show successful pregnancy in patients with single ventricles and their risks are controlled with collaborative efforts from all teams involved.^{5–7}

Our patient, in addition to having a single ventricle, had atrial fibrillation and a placental haematoma, which was managed with bed rest, intravenous heparin and C-section. She had an uneventful in-hospital course and she and her baby were discharged in good health.

This case highlights the challenges faced by other disciplines of medicine due to the ever increasing population of patients with grown-up congenital heart (GUCH) disease. An important measure is counselling these patients on coping with adult life, explaining what to expect during pregnancy, when participating in sports, while gaining employment and when facing insurance issues. The management of GUCH defects is evolving as a new specialty.⁸ Our case highlights the challenges faced by the cardiology and obstetrics teams to optimise the management of such a high-risk patient and to achieve an ideal outcome in a precious pregnancy such as this.

Learning points

- Counselling of patients is the key to success and should be carried out by a multidisciplinary team.
- Pregnancy in some cases of grown-up congenital heart (GUCH) defects is possible, but requires prior arrangements made to ensure the patient is under the care of a GUCH team.
- Pregnancy in a female with single ventricle double-inlet right ventricle, placenta bleeding and atrial fibrillation was safe and achieved mainly through bed rest, careful intravenous heparins and a caesarean section.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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