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Repeated pregnancies in uncorrected double outlet right ventricle: a rare occurrence

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Brief Report

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Abstract

Pregnancy in a woman with double outlet right ventricle, ventricular septal defect, and pulmonary stenosis is presented. Her previous two pregnancies ended in fetal losses. In the current pregnancy, she had thrombocytopenia, placenta previa, and severe fetal growth restriction. She underwent an emergency caesarean section followed by post-partum haemorrhage and COVID-19 infection. Literature review has been performed.

Double outlet right ventricle, ventricular septal defect, and pulmonary stenosis is a congenital cyanotic heart disease with a pathophysiology similar to tetralogy of Fallot. These patients have a right-to-left shunt with low pulmonary blood flow. Unoperated patients have dismal survival with only 25% surviving till 1 month of age.[1] However, a small percentage of patients do survive and reach adulthood. Pregnancy may, therefore, rarely be encountered in women with double outlet right ventricle. Double outlet right ventricle belongs to modified WHO class 3 risk during pregnancy indicating significantly increased risk of maternal mortality or severe morbidity.[2] Only a few reports describing uncorrected double outlet right ventricle in pregnancy are described in the literature.[3,4] We report three pregnancies in a woman who also developed a COVID-19 infection after delivery.

Case report

A 30-year-old G_3P_{200} at 30+4 weeks, known case of double outlet right ventricle with a ventricular septal defect and pulmonary stenosis since childhood was presented to our emergency labour room complaining of painless spotting per vaginum. She was unable to get the surgical repair of the cardiac defect due to financial constraints and had not been under regular supervision of a cardiologist and obstetrician. There was a gradual increase in breathlessness as pregnancy advanced. She complained of dyspneoa on walking for 100 m and was NYHA functional class III at presentation. Cyanosis was present since infancy.

She had been married for 9 years and had two pregnancies, 7 years and 3 years back. These were not supervised under a multidisciplinary team and she delivered in a small hospital. During these pregnancies, her symptoms remained largely unchanged. However, severe intrauterine growth restriction was present in both pregnancies. Preterm deliveries occurred at 30 and 32 weeks, respectively. In the first pregnancy, there was intrauterine fetal demise while in the second, a live-born boy of 1100 g was delivered. The neonate expired on day 8 of life due to respiratory distress.

At initial evaluation, she had central cyanosis and acropachy. Oxygen saturation was 82% on room air. Apex was present in the left 5th intercoastal space in the mid-clavicular line. Systolic thrill was present in the left parasternal region. The second heart sound was single with a harsh long ejection systolic murmur best heard in the left parasternal edge. She had normal vesicular breath sounds with normal liver span. Uterus was 26 weeks size, relaxed with regular fetal heart sounds.

Thrombocytopenia was detected since 28 weeks of gestation with platelet counts ranging between 40,000 and 60,000 per µl, haemoglobin was 16 g/dl. Thyroid function was normal. Level II ultrasound at 17+6 weeks was suggestive of a low-lying placenta with no gross fetal malformation. Fetal echocardiogram was normal. Biochemical and coagulation profiles were normal. Electrocardiogram showed sinus tachycardia, right axis deviation, P pulmonale, and right ventricular hypertrophy (Fig 1). Echocardiography showed a large mal-aligned peri-membranous ventricular septal defect with inlet extension, aortic override of more than 70%, and severe pulmonary stenosis with a side-by-side relationship of great vessels (Fig 2).

Ultrasound revealed placenta previa covering internal os with fetal parameters of 24–26 weeks with an estimated fetal weight of 639 ± 93 g (<3rd centile) suggesting severe growth restriction. Doppler of the umbilical vessels showed reversal of blood flow at places. The patient was given dexamethasone for fetal lung maturity.

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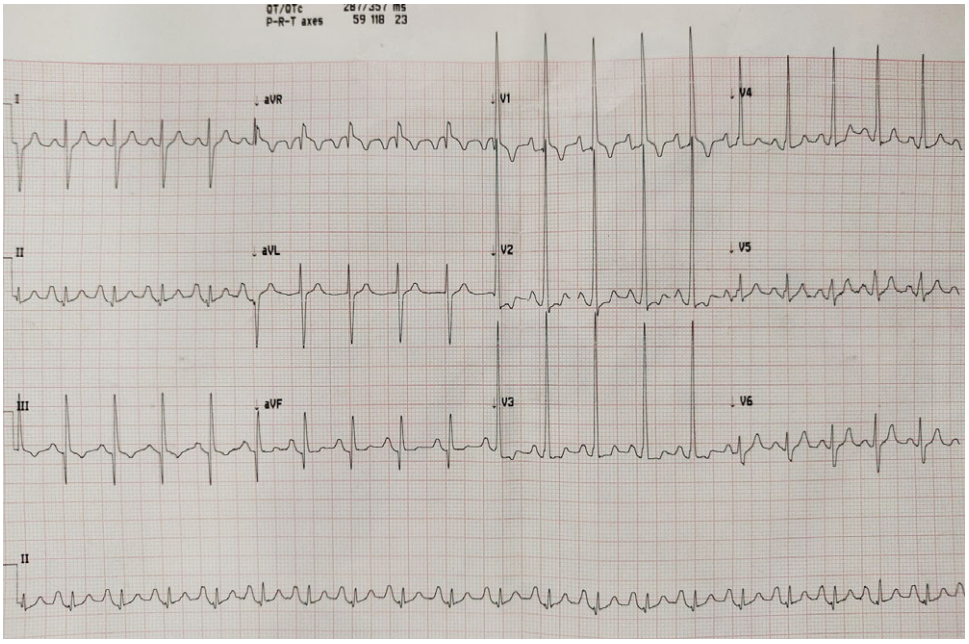


Figure 1. ECG showing right axis deviation, clockwise loop, right atrial enlargement, right ventricular hypertrophy, and prominent Q waves in inferior leads.

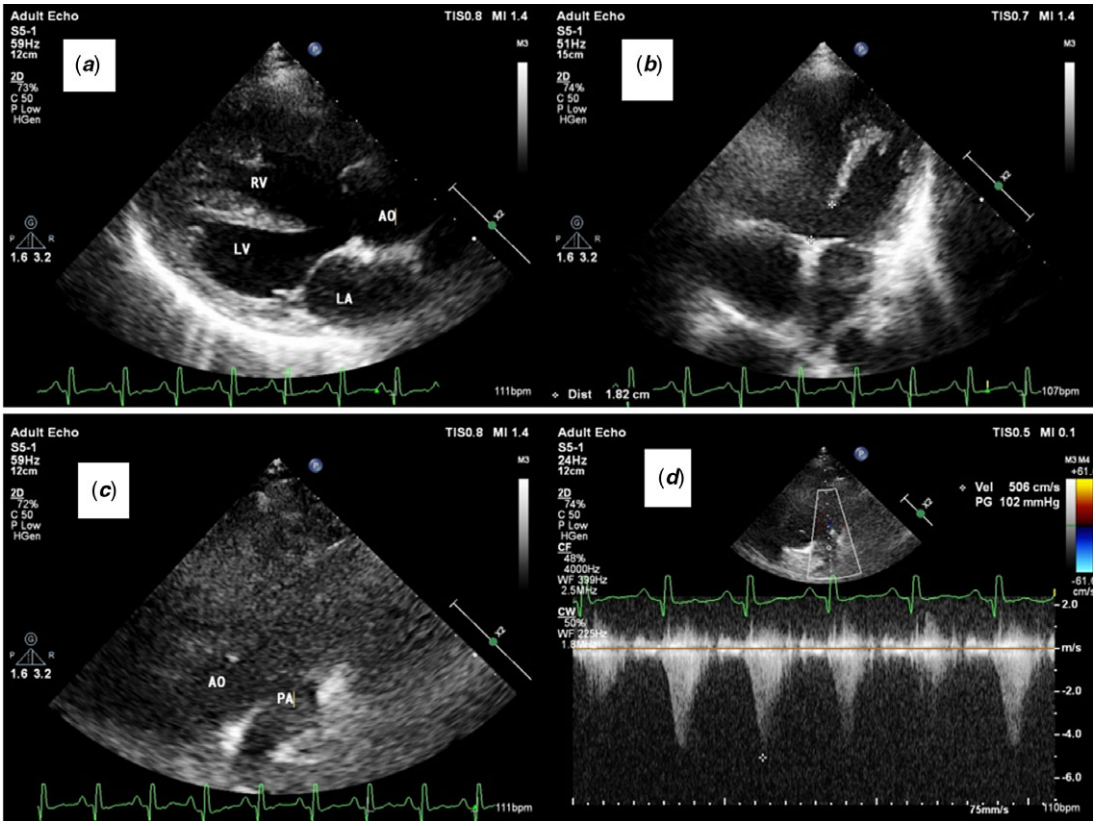


Figure 2. (***a*** and ***b***) Echocardiogram showing large mal-aligned peri-membranous VSD with inlet extension, with an aortic override of more than 50% (***a***) with a side-by-side relationship of great vessels (***c***) and severe pulmonary stenosis (***d***).

At 31+1 weeks, she had a sudden episode of heavy bleeding and was taken up for an emergency caesarean section. Infective endocarditis prophylaxis was given. General anaesthesia was administered with injection ketamine 1 mg/kg and injection propofol 1 mg/kg. Succinylcholine 1.5 mg/kg was given to facilitate endotracheal intubation through rapid sequence induction. Anticipating a drop in blood pressure, phenylephrine infusion was started at 50 mcg/min. Intraoperatively, the placenta was anterior low lying and delivered a live-born girl weighing 545 g with APGAR of 7, 8 and no gross malformation. The patient had an atonic post-partum haemorrhage with blood loss of around 1L. She was managed medically with oxytocin infusion and intramyometrial PGF2 alpha. There was a sudden drop in mean arterial blood pressure to 50 mmHg. Oxygen saturation fell to less than 80%. This sudden

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hypotension could be due to the vasodilatory effects of oxytocin. Vasodilatation also increases right-to-left shunt and aggravates systemic desaturation. 100 μg of phenylephrine bolus was given with immediate improvement in vitals. She received two units of packed cells and platelet transfusions. She was extubated on the table and shifted to a high-dependency unit. On the 4th post-operative day, she developed fever and myalgia. RT-PCR of nasopharyngeal swab was positive for COVID-19 infection. Fever subsided in 4 days. She had an uneventful course and was discharged on the 8th post-LSCS day. The baby is doing well with a weight of 1365 g at 2 months post-caesarean.

Discussion

Double outlet right ventricle with a ventricular septal defect and pulmonary stenosis is a rare complex congenital cyanotic heart disease with a right-to-left shunt and reduced pulmonary blood flow. Without surgical repair, only a few individuals survive into adulthood.[1] Thus, this condition is rarely reported in pregnancy.[5,6]

Women with underlying complex congenital cyanotic heart disease do not adapt well to the physiological changes of pregnancy and are at significant risk of maternal and fetal adverse events. The major risks during pregnancy include heart failure, thrombotic complications, infective endocarditis, and arrhythmias.[2] Reduced peripheral vascular resistance that occurs in pregnancy may increase the right-to-left shunt and worsen the cyanosis. Risk of recurrent miscarriages, intrauterine growth restriction, fetal deaths, and operative delivery is also increased. This is especially true in women with oxygen saturation of <85% as in our patient. In these women, live birth rate of only 12% is reported.[7] Thus, pregnancy should be avoided in women with cyanotic heart disease and saturations of <85%. However, if they conceive, pregnancy should be supervised by the pregnancy heart team, under a multidisciplinary care.

There are only a few reports of pregnancies in this disorder with mixed outcomes. Tilak et al reported a third gravida with double outlet right ventricle with a ventricular septal defect and pulmonary stenosis. She had previous two second-trimester abortions. She was presented at 22 weeks of gestation with NYHA class III–IV dyspnoea and haemoptysis. Oxygen saturation was only 78%. Intrauterine fetal demise was diagnosed and pregnancy terminated by hysterotomy.[4] A successful pregnancy in a woman with double outlet right ventricle with ventricular septal defect, pulmonary stenosis, and associated atrial septal defect has also been reported. She had an uncomplicated normal vaginal delivery.[3] She had the previous history of a first-trimester abortion.

Our patient tolerated the pregnancy well in the early months, however, there was a gradual increase in dyspnoea. This is because haemodynamic changes including plasma volume peak at around 28–30 weeks. Thrombocytopenia has been reported in 25% of pregnancies in women with cyanotic CHD.[8] Mechanisms potentially responsible for this are decreased megakaryocyte production, decreased platelet production from megakaryocytes, increased platelet destruction, and increased platelet activation. Post-partum haemorrhage occurs in 6–21%, and is an important complication in women with CHD though the mechanisms are not well understood.[9,10] Post-partum haemorrhage is the second most common cause of maternal mortality in these women after pre-eclampsia with an incidence of 6.1%.[10] In our patient, post-partum haemorrhage was due to placenta previa and was managed with packed cell and platelet transfusions.

Like other cyanotic CHD, the preferred mode of termination is vaginal delivery using epidural anaesthesia.[2,11] In our patient, however, caesarean section was done for symptomatic placenta previa. General anaesthesia was chosen as the patient was actively bleeding and had significant thrombocytopenia. Hypotension is poorly tolerated and can increase cyanosis due to an increase in right-to-left shunt. Systemic vasoconstrictors like phenylephrine reduce the shunt, and thus improve the systemic saturation.[11] Oxytocin is the first-line uterotonic in women with cyanotic heart disease.[12] Our patient may have developed hypotension and worsening saturation due to the vasodilatory effects of oxytocin. The infusion should thus be slow, and the blood pressure was closely monitored. Haemodynamics showed immediate improvement with intravenous phenylephrine. Misoprostol and carboprost are the second-line uterotonics.[12] Ergometrine can cause hypertension, precipitate heart failure, and should be avoided.

An additional maternal complication developed in the post-operative period in our patient was a COVID-19 infection. Despite an underlying congenital cyanotic heart disease, she had an uncomplicated course.

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Conflicts of interest. None.

Ethical standards. Patient data has been anonymised.

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