

Case Report

Successful Vaginal Delivery in a Woman with Tetralogy of Fallot and Pulmonary Atresia After Four Cardiac Operations

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Key words:

Congenital heart diseases, tetralogy of Fallot, pregnancy, delivery, reoperation.

We describe a woman with tetralogy of Fallot and pulmonary atresia, with a history of four previous cardiac operations, who decided to bear her own children. Under interdisciplinary counselling and the appropriate medical care, she underwent a successful vaginal delivery and had a healthy baby.

Evolution in paediatric cardiology and cardiac surgery has given the opportunity to an increasing number of women with congenital heart disease to bear their own children. During pregnancy, a number of haemodynamic changes occur, such as a decrease in systemic vascular resistance, an increase in circulating blood volume and cardiac output, that can cause functional cardiovascular deterioration. The challenge of the expert cardiologist, obstetrician and anaesthesiologist working as a team is to estimate the real risk for mother and foetus, give the appropriate counsel and then to achieve the best possible outcome of the patient's decision. The majority of women with congenital heart diseases do not face special problems during pregnancy and continuation of the pregnancy should be suggested by the specialists.

We describe the case of a 32-year-old woman with tetralogy of Fallot and pulmonary atresia, with a history of 4 previous cardiac operations, who underwent a successful delivery at a tertiary hospital.

Case presentation

A 32-year-old woman with operated te-

tralogy of Fallot and pulmonary atresia decided to bear her own children. She underwent her first operation with full correction, using a conduit, 25 years ago. After 3 years, she underwent a Björk-Shiley valve insertion in the pulmonary position. Sixteen years ago, she had an aortic homograft conduit placed in the pulmonary position. Five years ago, she underwent her fourth operation, due to conduit stenosis and regurgitation, and a composite conduit with a porcine valve was inserted as the previous calcified homograft was removed.

Since then, she had been under scheduled follow up with clinical and echocardiographic examination, and she was in NYHA functional class I to II with good cardiac function. On clinical examination the patient was pink (saturation 96%, on room air) and well with no peripheral oedema. She had a normal first heart sound, single second, and a very soft ejection systolic heart murmur at the upper left sternal edge 1/6 with no diastolic murmur. Her chest was clear and the rest of the examination was normal. Chest X-ray showed situs solitus, levocardia, right aortic arch, mild cardiomegaly with promi-

Manuscript received:
December 14, 2008;
Accepted:
October 11, 2009.

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nant central pulmonary arteries, and evidence of previous median sternotomy. ECG examination revealed sinus rhythm with a superior axis and a right bundle branch block pattern with a QRS duration of 150 ms. Echocardiographic study showed mild aortic root dilation (3.8 cm) with no aortic regurgitation. Her pulmonary valve prosthesis was functioning well, with no pulmonary stenosis or regurgitation. Left ventricular function was normal, with a fractional shortening of 38%. Pulmonary artery systolic pressure was slightly increased (25 mmHg). The FISH test was negative regarding 22q deletion (Di George syndrome).

The patient decided to bear her own child and the appropriate counselling was given by a cardiologist experienced in adult congenital heart diseases, who explained the potential risks to her.

During the pregnancy she was in NYHA class II and she was under scheduled follow up with clinical and echo examinations. During gestation the patient developed mild dilatation of the right heart chambers (Figure 1). Decision about the time and mode of delivery was made after an interdisciplinary discussion (cardiologist specialising in adult congenital heart diseases, obstetrician and neonatologist). The patient was admitted to a tertiary hospital a week before the delivery for monitoring of the foetus. A 35-week healthy boy (weight 3.5 kg, APGAR 9) was born by a vaginal delivery without the use of oxytocin. Eighteen months later the mother is currently in NYHA class I to II with good cardiac function and the boy is clinically healthy. Right ventricular size returned to normal dimensions.

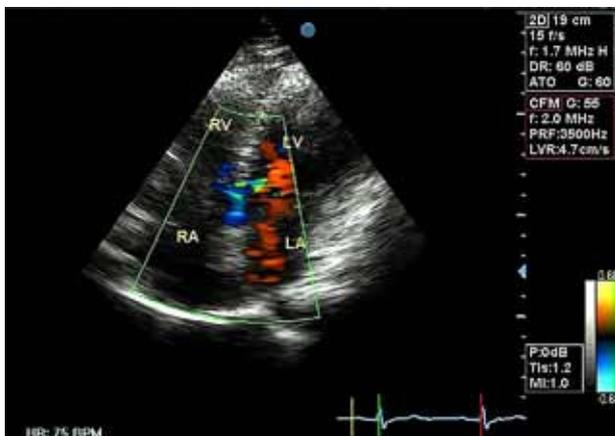


Figure 1. Cardiac echo (4-chamber view) showing mild dilatation of right ventricle and right atrium, at 32 weeks' gestation. RV – right ventricle; RA – right atrium; LV – left ventricle; LA – left atrium.

Discussion

It is mandatory for cardiologists to be aware of the risks that women with congenital heart disease could face during pregnancy, so they can give the right preconception counselling and the optimal care during pregnancy, delivery and postpartum period.¹ Preconception counselling has to address how pregnancy may affect not just the mother but the foetus too, and the final decision belongs to the family. The counselling should be provided by a specialised obstetrician and cardiologist with special training in adult congenital heart disease.² Before pregnancy, a complete clinical assessment should be made, including medical history, clinical examination, oxygen saturation, ECG, chest radiography and echocardiography. Treadmill exercise testing has a significant utility in order to define the functional class of the patient, as women in NYHA classes I and II have better outcomes. Invasive methods may be required in some groups of patients.³ Further predictors of a poor pregnancy outcome are also the history of a previous cardiac event, the presence of left heart obstruction, an ejection fraction below 40% and when the saturation on room air is under 90%.³

Candidate mothers should also be aware of the risk of recurrence of congenital heart disease and the potential need for their hospitalisation during pregnancy. The reported risk of their foetus having structural cardiac defects ranges from 3% to 12%, in comparison with a background risk of 0.8% for the general population.⁴ Patients with Eisenmenger or other causes of pulmonary arterial hypertension, Marfan syndrome with aortic root diameter >4 cm, or severe left side obstructive lesions are considered as a group with high maternal morbidity and mortality and if the patient chooses to proceed, the need for care in a tertiary hospital is mandatory.²

Classically, corrected tetralogy of Fallot is considered a low-risk lesion in terms of both maternal and foetal outcomes.⁵ Seven percent of the patients with repaired tetralogy of Fallot developed adverse cardiovascular events, including supraventricular tachycardia, heart failure, pulmonary embolism and progression of right ventricular dysfunction during pregnancy.^{5,6} In our case, the patient developed mild right ventricular dilatation, which was reversed after delivery. Children of mothers with tetralogy of Fallot are more likely to have congenital heart disease, with a reported incidence of 3%. In addition, it is believed that 15% of patients with tetralogy of Fallot have Di

George syndrome, which is inherited with autosomal dominant pattern.^{5,6}

Pulmonary valve regurgitation is a risk factor in patients with surgically corrected tetralogy of Fallot. Exercise limitation, secondary development of tricuspid regurgitation, presence of supraventricular or ventricular arrhythmias, and the risk of sudden death are the most important complications of pulmonary valve regurgitation.^{7,8} The choice of the appropriate timing for pulmonary valve replacement (need for reoperation) is very important and should be considered before the development of irreversible right ventricular dysfunction.^{7,8} Furthermore, mean QRS duration tends to be longer (>180 ms) in populations of patients with tetralogy of Fallot who suffer sudden death, rather than in healthy counterparts.⁸

Patients with operated tetralogy of Fallot need periodic follow up in order to achieve timely pulmonary valve replacement before the development of complications due to volume overload of the right ventricle.

Regarding pulmonary atresia, a retrospective study showed that this group of patients developed significant complications during pregnancy, even the patients with surgical repair. Important risk factors are elevated right ventricular pressures and the existence of residual systemic to pulmonary collaterals.^{5,9}

Decisions about timing and mode of delivery should be reached after an interdisciplinary discussion that also involves the patient. Vaginal delivery has a lower risk of complications for both the mother and foetus.² Cardiac indications for caesarean section are Marfan syndrome with aortic root >4 cm, aortic dissection and warfarin treatment within the preceding two weeks, due to the risk of cerebral haemorrhage for the foetus.³ Low dose epidural anaesthesia does not cause excessive vasodilation and is the analgesia of choice.³ The American Heart Association guidelines do not recommend antibiotic prophylaxis in vaginal or caesarean delivery,¹⁰ but in practice,

most centres give prophylaxis, as we did in our patient because of the existence of the porcine valve conduit. Oxytocin and ergometrine have unpredictable effects on the haemodynamic status and should be avoided.¹¹ Close observation and continuous haemodynamic monitoring are mandatory during the delivery and up to one week postpartum. No anticoagulation was administered to our patient during the pregnancy or puerperium.

Our case suggests that pregnancy in a woman with congenital heart disease and a history of multiple cardiac operations, although carrying substantial risks for foetus and mother, is feasible under interdisciplinary counselling and the appropriate medical care.

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