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PERIPARTUM CARDIOVASCULAR DISEASE MINI-FOCUS ISSUE

BEGINNER

CASE REPORT: CLINICAL CASE

Pregnancy in a Patient With Tetralogy of Fallot and Borderline Pulmonary Arterial Hypertension

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ABSTRACT

Management of pregnancy in patients with complex congenital heart disease and pulmonary arterial hypertension has always been a challenge. We are presenting a patient with complex congenital heart disease and borderline pulmonary arterial hypertension who complicated with pulmonary embolism during pregnancy. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:91–5) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 29-year-old woman with tetralogy of Fallot (ToF), pulmonary atresia, and borderline pulmonary arterial hypertension (PAH) expressed the wish to become pregnant. New York Heart Association (NYHA) functional class was II. At clinical examination there were midline sternotomy and left thoracotomy scars. There was a mild diastolic 2/6 murmur heard at the right upper sternal edge. Sinus rhythm, saturation was 98%. There were no signs of peripheral edema. Her blood pressure was 105/58 mm Hg.

LEARNING OBJECTIVES

- To underline the fact that patients with tetralogy of Fallot and systemic-to-pulmonary shunts may develop pulmonary arterial hypertension.
- To discuss level of surveillance, management, and complications of pregnancy in patients with congenital heart disease and pulmonary arterial hypertension.
- To underline the need of expert pre-pregnancy counseling and assessment and follow-up during pregnancy.

PAST MEDICAL HISTORY

Her history included multiple surgical and transcatheter interventions. She underwent Waterston shunt at 3 weeks of age. She had a left Blalock-Taussig (BT) shunt at 16 months. At 4 years, both shunts were taken down and establishment of continuity between the 2 pulmonary arteries (PAs) and connection to the right ventricle (RV) was constructed. Soon, the left BT shunt was re-established due to failure to RV-to-PA connection, and at 7 years of age she underwent surgical patch angioplasty of left PA (LPA) stenosis. One year later, she underwent complete repair of pulmonary atresia with transatrial patch closure of ventricular septal defect, suture closure of patent foramen ovale, and establishment of continuity between RV and PA confluence with 19 mm aortic valve homograft, takedown of the modified left BT shunt,

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Informed consent was obtained for this case.

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ABBREVIATIONS AND ACRONYMS

BT = Blalock-Taussig

LPA = left pulmonary artery

MAPCA = major aortopulmonary collateral artery

NYHA = New York Heart Association

PA = pulmonary artery

PAH = pulmonary arterial hypertension

PAP = pulmonary artery pressure

PCWP = pulmonary capillary wedge pressure

RPA = right pulmonary artery

RV = right ventricle

ToF = tetralogy of Fallot

and dilation of the LPA and right PA (RPA). At 9 years of age, she had RPA balloon dilation, and 1 year later, she had a RPA balloon angioplasty and RPA stenting, as well as an LPA balloon angioplasty. The catheter after the last intervention revealed an elevated mean PA pressure (PAP) of 32 mm Hg at 10 years of age. There was a 6 mm Hg gradient among the RV, PA, and LPA and gradient of 9 mm Hg between the RV and RPA. No major aortopulmonary collateral arteries (MAPCAs) were mentioned in her past medical or surgical notes. She has remained clinically stable over the years.

DIFFERENTIAL DIAGNOSIS

As there were no MAPCAs, PAH was attributed to long-standing (7 years) systemic-to-pulmonary shunts (1).

INVESTIGATIONS BEFORE PREGNANCY

In order to evaluate the patient before pregnancy, complete functional and imaging assessment was performed. Oxygen consumption was 24 ml/kg/min (60% predicted). Holter monitoring did not reveal significant arrhythmia. Echocardiography revealed normal size and function of the left ventricle and aorta dilation 42 mm at the sinuses of Valsalva. The right ventricle (RV) was mildly dilated with borderline normal ventricular function. There was mild-to-moderate pulmonary regurgitation, and there was no significant gradient at the level of pulmonary valve, moderate aortic regurgitation, and mild tricuspid regurgitation. RV systolic pressure was estimated 50 to 55 mm Hg (**Figure 1**).

Cardiac magnetic resonance showed normal left and right systolic function, moderate aortic regurgitation, patent RV-to-PA aortic homograft with moderate regurgitation, aorta dilation up to 42 mm, and patent RPA stent with good peripheral flow (**Figure 2**, Video 1).

Right heart catheterization showed RV pressure of 54/10 mm Hg, PAP of 50/6, mean PAP of 22 mm Hg, and mean pulmonary capillary wedge pressure (PCWP) of 12 mm Hg. There was gradient between the PA and LPA, and a 3 mm Hg gradient between the main PA and RPA. No MAPCAs were revealed by aortogram.

She had complex congenital heart disease, and the aorta was dilated in the setting of ToF up to 42 mm, which was not considered as high risk for rupture during pregnancy. The main concern was borderline PAH, keeping in mind that mean PAP was 32 mm Hg at 10 years of age. It was decided that she could go through pregnancy. It was explained to the patient and family that there was a high risk of complications (heart failure and deterioration of PAH during pregnancy), while quoted risk of death was <1%.

MANAGEMENT

During pregnancy, she was under close follow-up, with monthly echocardiography and Holter monitoring. Owing to multiple ventricular extrasystoles she was put on metoprolol. Up to the 33rd week, the pregnancy was uneventful but then she presented with deterioration of NYHA functional class III peripheral edema and PA systolic pressure up to 80 mm Hg. There was no deep venous thrombosis detected and the patient declined computed tomography pulmonary angiography that was offered.

She was treated with therapeutic dose of low-molecular-weight heparin, as differential diagnosis of RV systolic pressure elevation should include pulmonary embolism and pregnancy-induced volume overload. Her oxygen saturation was not impaired, and there was no hemodynamic compromise. Before delivery, the left ventricle had good systolic function, the RV was dilated with mildly impaired systolic function, and there was moderate aortic regurgitation, while RSVP reduced to 65 to 70 mm Hg.

She delivered a healthy boy weighing 2,500 g by elective caesarean section under general anesthesia at the 36th week of pregnancy. The decision for caesarean section was made by a multidisciplinary team, as she was in NYHA functional class III.

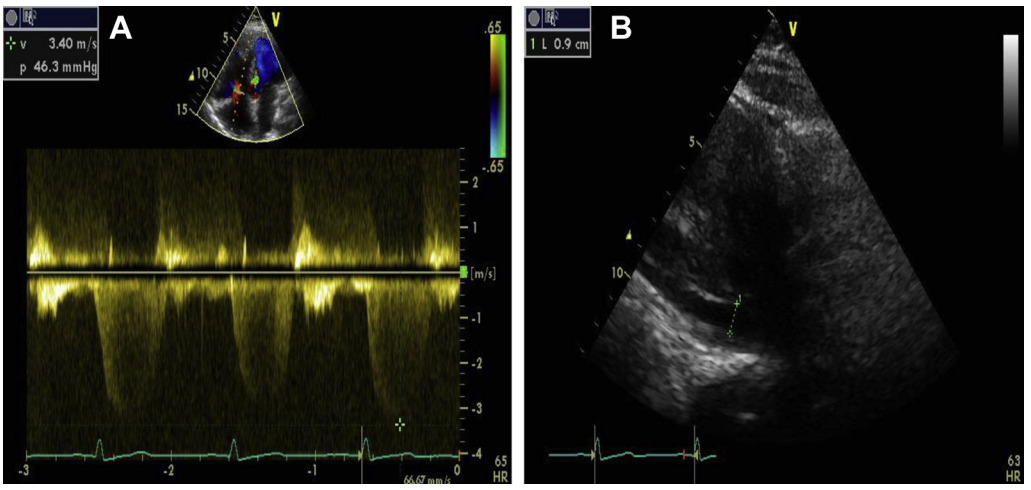
Post-delivery, the patient underwent elective computed tomography pulmonary angiography that revealed a pulmonary embolism (**Figure 3**), and she was discharged on oral anticoagulation. On oral anticoagulation and diuretics 6 weeks post-delivery, her NYHA functional class was II.

DISCUSSION

Pregnancy in patients with repaired ToF without PAH is usually well tolerated (2). Long-standing systemic-to-pulmonary shunts may be the cause of PAH in the setting of ToF.

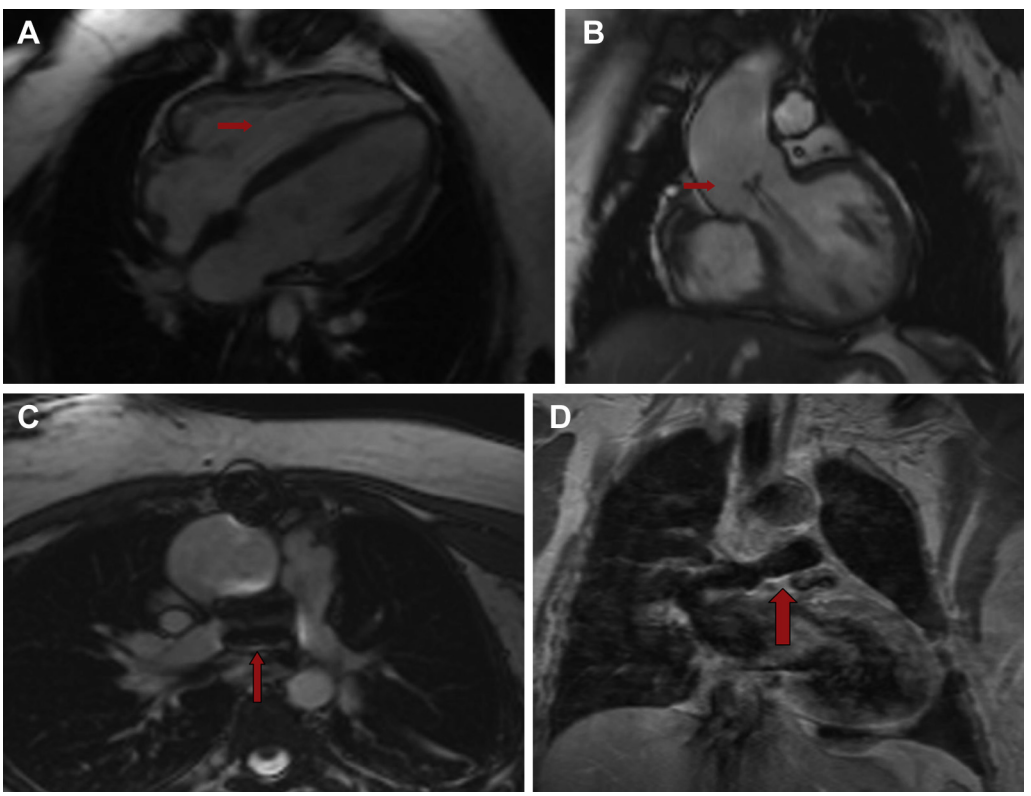
As our patient's mean PAP was 22 mm Hg, <25 mm Hg (3), she was classified as World Health Organization risk category III based only on complex congenital heart disease and aorta dilation (4). Clinical significance of a mean PAP between 21 and 24 mm Hg was previously characterized as unclear (3). According to new classification, mean PAP

FIGURE 1 **Pre-Pregnancy Echocardiography**



Right ventricular systolic pressure was estimated at 50 to 55 mm Hg: **(A)** tricuspid regurgitation trace and **(B)** inferior vena cava.

FIGURE 2 **Cardiac Magnetic Resonance Imaging Pre-Pregnancy**



(A) Axial 4-chamber cine view: mildly dilated right ventricle **(arrow)**. **(B)** Oblique coronal left ventricular outflow tract cine view. Moderate aortic regurgitation with dilated aortic root **(arrow)** and ascending thoracic aorta. **(C)** Patent right pulmonary artery stent **(arrow)** axial cine view. **(D)** Patent right pulmonary artery stent **(arrow)** well seen in coronal dark blood spin echo image.

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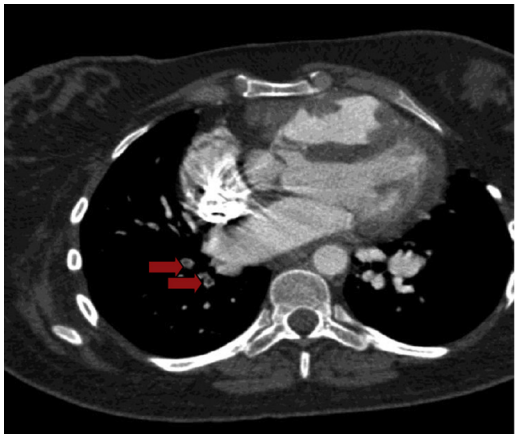
FIGURE 3 Computed Tomography Pulmonary Angiogram Post-Delivery



Pulmonary artery branches to right lower lobe filling deficits **(arrows)**.

>20 mm Hg with a mean PCWP <15 mm Hg is considered as PAH (5). Our patient's postpartum mean PAP was 24 mm Hg and mean PCWP was 10 mm Hg.

Several predictors of maternal cardiovascular events for congenital heart disease patients have been proposed (6,7). The presence of pulmonary hypertension further increases the risk of ToF pregnant women (8). In the recent ROPAC (Registry Of Pregnancy And Cardiac disease) registry, patients with PAH demonstrated the highest mortality (9%) (9). The greatest period of risk is the puerperium and early postpartum. Pulmonary hypertensive crisis, pulmonary thrombosis, and right heart failure are the most common causes of death (2,10). Thromboembolism is a major risk for pregnant patients with PAH and anticoagulation should be considered (2). Our patient was not on anticoagulation as she had no risk factors for thromboembolism. Unexpected pulmonary embolism complicated the pregnancy and may have contributed in the clinical deterioration at the end of pregnancy.

For patients with heart disease detailed consultation should precede pregnancy. Severe systemic ventricular dysfunction, severely dilated aorta, PAH, severe symptomatic aortic or mitral stenosis, NYHA functional class >II and complex congenital heart disease, carry significant risks for maternal morbidity and mortality (3).

These women should be evaluated and managed by a multidisciplinary team, in an expert center for pregnancy and cardiac disease.

FOLLOW-UP

Catheterization 1 year post-delivery: RV pressure 65/4 mm Hg, PAP 40/10 mean 24 mm Hg, mean PCWP 10 mm Hg. Treatment with macitentan was initiated for PAH. Her medication includes oral anticoagulation, metoprolol, and low-dose furosemide. Three years post-delivery she is in NYHA functional class II, taking care of her boy.

CONCLUSIONS

Patients with repaired ToF are in general at low risk, while pulmonary hypertension is a major risk factor for adverse maternal and fetal outcomes. Complication with thromboembolism further increases the risk. Even in patients with meticulous pre-pregnancy counseling and adequate management, severe complications may occur. Complex patients require expert care, given the increased risk for pregnancy complications.

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KEY WORDS pregnancy, pulmonary atresia, pulmonary hypertension, tetralogy of Fallot

APPENDIX For a supplemental video, please see the online version of this paper.