T

he number of women of childbearing age who have congenital heart disease is increasing. A similar proportion of
these women can be expected to attempt pregnancy as women in general. Although most of these women can be expected to
tolerate pregnancy well, particularly if the pregnancy is carefully planned following appropriate clinical evaluation, not all
pregnancies are planned, and some will occur in women with unsuitable hemodynamics.

We report the case of a 21-year-old woman born with pulmonary atresia and a ventricular septal defect (VSD), ultimately palliated by a fenestrated closure of VSD and a valved homograft conduit placement between the right ventricle and the main pulmonary artery at the age of 3 years, who presented at 16 weeks gestation with a severely degenerate conduit.

Case Report

A 21-year-old woman had been born with pulmonary atresia and a VSD. She was initially palliated with bilateral Blalock-Taussig shunts (classical left and modified right) in the neonatal period because her pulmonary artery confluence was hypoplastic. At 3 years of age, she underwent reparative surgery consisting of closure of the VSD with a fenestrated patch and the placement of a 19-mm homograft valved conduit between the right ventricular outflow tract (RVOT) and the pulmonary artery bifurcation, thereby stimulating growth of the hypoplastic pulmonary artery confluence. The bilateral Blalock-Taussig shunts were ligated. At the age of 18 years, both of her pulmonary arteries were stented because of distorted and stenotic branch pulmonary arteries. The fenestration of the VSD patch was closed interventionally by the use of a muscular VSD closure device. At 20 years of age, there was clinical and echocardiographic evidence of degeneration of the homograft conduit, and she was advised against pregnancy pending further evaluation.

Approximately 6 months later, she presented at 16 weeks gestation of an unplanned pregnancy. At this stage, she was asymptomatic and clinically well. At 20 weeks, she remained asymptomatic but was showing clinical evidence of hemodynamic decompensation with raised jugular venous pressure and echocardiographic evidence of worsening right ventricular (RV) dilatation with high RV pressure. There was severe pulmonary regurgitation and significant RVOT obstruction with Doppler evidence of RV hypertension. Obtaining a good-quality Doppler signal across the RVOT was difficult but was at least 3 m/s. There was evidence of fetal growth retardation from an obstetric/fetal well-being assessment. At 23 weeks, the otherwise healthy female fetus had an estimated weight of 480 g (expected mean weight at 24 weeks is 600 g).

The clinical view was that the patient was at risk of sudden hemodynamic decompensation or life-threatening arrhythmia. There was concern that the pregnancy might not be sustained long enough for the fetus to reach a viable maturity.

The management options available were to let the pregnancy continue, deliver the baby at the earliest possible gestation, surgically replace the conduit, or implant a new pulmonary valve percutaneously during the pregnancy. The patient was evaluated with cardiac MRI. This demonstrated stenosis of the homograft with anatomy indicating that she would be suitable for placement of a Medtronic Melody stent-mounted valve prosthesis (Figure 1).

At 23 weeks and 4 days, the patient underwent cardiac catheterization. Pulmonary angiography demonstrated good angiographic results from the branch pulmonary artery stenting, but with severe pulmonary regurgitation (Figures 2 and 3). The aortic pressure was 90/46 mean 61 mm Hg, RV 72/15, ratio 0.8, RVOT gradient 23 mm Hg.

A RV angiogram confirmed the conduit stenosis in the outflow tract.

An 18-mm Crystal balloon was inflated to predilate the conduit, while, simultaneously, the left coronary artery was opacified to demonstrate that it would not be compressed by stent implantation. The conduit was prestenated by using a 36-mm-long Max LD stent mounted on the aforementioned 18-mm Balt Crystal balloon. A Medtronic Melody Valve was prepared and successfully deployed to 20 mm inside the prestenated area with the use of an Ensemble delivery system (Figure 4).

Repeat hemodynamics demonstrated that the aortic pressure had risen to 111/54 mean 73, RV 76/12, ratio 0.68, RVOT gradient 27 mm Hg. Angiography in the main pulmonary artery showed negligible pulmonary regurgitation. Although the RVOT gradient increased, the concomitant increase in the aortic pressure suggested that the cardiac output had increased.
and the fall in systemic to RV pressure ratio and RV end diastolic pressure suggested that RV function had benefited immediately from the intervention.

The patient and fetus made an uneventful recovery from the anesthetic. The patient's central venous pressure normalized. The following day, her tricuspid regurgitation velocity was measured at 3.4 m/s. Echocardiography at 30 weeks described a substantial reduction in RV size (Figure 5) with improvement in both RV contractility and septal movement consistent with a reduction in RV pressure resulting in a reduction in left ventricular compression with improved filling (Movies I and II in the online-only Data Supplement).

Radiation to the patient and fetus was minimized by using single-plane fluoroscopy only above the diaphragm, collimation to a minimum field, screening as close to posteroanterior as possible, and a fluoroscopy frame rate at 2 frames per second as previously described. The dose area production to the patient was recorded as 437 μGy/m².

The fetus remained small, but, rather than becoming increasingly growth retarded, she grew on approximately the 5th centile. The pregnancy continued to 32 weeks gestation when the woman presented in premature labor with the predicted (from uterine artery Doppler measurements) biochemical evidence of preeclampsia. A 1.5-kg female infant was delivered by cesarean delivery. The mother and the child made an uneventful recovery from delivery, and the infant has subsequently thrived, having almost doubled her birth weight by her expected date of delivery.

Discussion
The prevalence of women of childbearing age with significant cardiac pathology is increasing. This is mainly due to the increase in the number of women with congenital heart disease surviving until childbearing age.1 Also, there is a small number of women with acquired cardiac pathology as a direct consequence of pregnancy: peripartum cardiomyopathy and pregnancy-related arrhythmia with rate-related cardiomyopathy that warrant expert care.

Ideally, women with congenital heart disease considering pregnancy should have their cardiac status optimized before embarking on pregnancy.5 There are reports of pulmonary valve replacement6 or percutaneous valve implantation within a right-sided conduit to facilitate pregnancy in such patients.7 Not all pregnancies are planned, and it is likely that there will be an increasing number of unplanned pregnancies in women with RV to pulmonary artery conduits whose cardiac status may not be well suited to pregnancy.

Radiation to a pregnant woman causes concern, but the fetus of a woman with serious cardiac pathology is at very high risk. In this patient, the calculated uterine dose was 0.01 mGy. During the second trimester of pregnancy, the consequence of radiation exposure to the fetus is an increased risk of childhood malignancy. The uterine dose exposed to this fetus was calculated to increase the risk of childhood malignancy by <1:1.7 million. The background incidence of childhood malignancy in the United Kingdom is 1:650.

A severely hemodynamically compromised mother is a liability to the fetus’ well-being, the intrauterine growth retardation demonstrated by this case is a testament to that; and, with the further risk of premature onset of labor, it is important that the mother is treated. Irradiation of a fetus in the second or third trimester does not risk teratogenicity but rather may increase the risk of malignancy in childhood. With suitable care and technique, radiological investigations and procedures can be safely performed during pregnancy.9 Percutaneous pulmonary valve replacement is a well-established strategy for patients with severe pulmonary valve disease of degenerate homograft conduits between the RV and pulmonary artery in patients with pulmonary atresia and Tetralogy of Fallot.8,9 It may be argued that pulmonary valve replacement should be performed before planned conception in such patients. The hemodynamic changes in the cardiac pathology associated with this unplanned pregnancy yielding a growth-retarded fetus and impending cardiac decompensation in the mother led us to undertake percutaneous pulmonary valve implantation. We feel that we have demonstrated that, in exceptional circumstances, the complex ratios of risk/benefit to both the mother and the fetus can be served by a carefully performed percutaneous pulmonary valve implantation.

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Disclosures
None.

References
Figure 1. Cardiac magnetic resonance image of long axis of right ventricular outflow tract showing severe degeneration of homograft conduit and severely dilated right ventricle. RV indicates right ventricle.

Figure 2. Preprocedure pulmonary angiogram systolic frame demonstrating a good long-term result from pulmonary artery stenting.

Figure 3. Preprocedure pulmonary angiogram diastolic frame demonstrating severe pulmonary reflux. PR indicates pulmonary reflux.

Figure 4. Postprocedure pulmonary angiogram showing fully competent Melody Valve.

Figure 5. Left and right ventricular cavity dimensions over time. LVIDd indicates left ventricular internal dimension-diastole; and RVDd, right ventricular dimension-diastole.
Percutaneous Pulmonary Valve Replacement During Pregnancy
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Movie Legend

Movie 1. Parasternal long axis imaging immediately before and 7 weeks post intervention. Best viewed with Windows Media Player.

Movie 2. Parasternal short axis imaging immediately before and 7 weeks post intervention. Best viewed with Windows Media Player.