The 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) dilation 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 mmHg, RV 72/15, ratio 0.8, RVOT gradient 23 mmHg.

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 prestented by using a 36-mm-long Max LD stent mounted on the aforementioned 18-mm Bالي Crystal balloon. A Medtronic Melody Valve was prepared and successfully deployed to 20 mm inside the prestented 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 mmHg. 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 dia-
stolic 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 improve-
ment 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, collima-
tion to a minimum field, screening as close to posteroanterior
as possible, and a fluoroscopy frame rate at 2 frames per sec-
ond as previously described.3,4 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 gesta-
tion when the woman presented in premature labor with the
predicted (from uterine artery Doppler measurements) bio-
chemical 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 car-
diac 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 conse-
quence 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 malign-
ancy by <1:1.7 million. The background incidence of child-
hood 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.8

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.9,10 It may be argued that pulmonary
valve replacement should be performed before planned con-
ception 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 percuta-
neous 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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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.