# Transitional Care for Young Adults With Congenital Heart Disease: A Case Study

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Congenital heart disease, transitional care, young adults

## INTRODUCTION

Congenital heart defects, any of myriad innate problems with the structure and function of the heart, affect nearly 1% of births in the United States annually (Centers for Disease Control and Prevention, 2016). Historically, a congenital heart defect was regarded as an inevitable fatality. However, immense strides in diagnosis, operative techniques, and critical care over the past 60 years have increased an infant's chance of survival into adulthood to 85% to 90% (Gurvitz et al., 2016). In fact, adults account for two-thirds of the overall congenital heart disease (CHD) population (Jacobs et al., 2015). This transition from mortality to vitality is an epic achievement, but it is not lacking in controversy. On the contrary, adult CHD poses very unique challenges to patients and health care providers alike. Most prominent is the ambiguity surrounding which providers should follow adult patients with pediatric heart conditions. The following case elucidates the nature of this novel dilemma, leading to a discussion of the critical role of the pediatric nurse practitioner (PNP) in the transition of care as children with CHD become adults.

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## CASE PRESENTATION

### Chief Complaint and History of Present Illness

> Adult CHD poses very unique challenges to patients and health care providers alike.

A 34-year-old female presented to a pediatric cardiology clinic for a surgical consultation. Her cardiac history results were positive for pulmonary valve stenosis, which was present (and diagnosed) at birth. At the time of the visit, her chief concern was her desire to become pregnant. She recognized that she was approaching advanced maternal age and that certain risks are more common in the pregnancies of older women. As a result, she hoped to conceive soon. Unfortunately, the combination of the stress of pregnancy together with a diagnosis of severe pulmonary valve stenosis increased her risk for complications such as arrhythmias, heart failure, miscarriage, and preterm delivery (Warnes, 2015). Consequently, this woman's consultation focused on the risks and benefits of an elective pulmonary valve replacement procedure.

### Past Medical History

This woman was diagnosed at birth with Noonan syndrome, an autosomal dominant disorder of variable expression. Noonan syndrome occurs in approximately

1 in 1,000 to 1 in 2,500 live births and is characterized by distinctive facial features, developmental delay, and cardiac defects (Prendiville et al., 2014). The woman in this case had neither distinctive facial features nor developmental delay. However, she was also diagnosed with pulmonary valve stenosis, pulmonary insufficiency, an atrial septal defect, and right ventricular dilation at birth. She underwent a successful open pulmonary valvectomy, transannular patch placement, and primary atrial septal defect closure at 6 months of age. Fortunately, with regular, biannual follow up from her cardiology team, this woman neither suffered complications nor required hospitalization during childhood. Her past medical and surgical history results were otherwise negative. Her birth was unremarkable, her immunizations were up to date, and she did not suffer from any adult-onset conditions.

### Family History

The woman in this case reported having no significant family history. Of note, her family history was negative for Noonan syndrome, CHD, arrhythmia, or sudden death.

### Personal/Social/Developmental History

This 34-year-old woman lived with her husband of 2 years with whom she planned to have children. She had positive relationships with her mother, father, and younger sister. She ate a varied diet and exercised approximately three times per week. She denied tobacco, drug, or excessive alcohol use. She graduated from college and reported working fulltime.

### Review of Systems

At the time of this surgical consultation, the woman was completely asymptomatic. The review of systems results were negative for shortness of breath, chest pain, loss of consciousness, cyanosis, and fatigue.

### Pertinent Physical Examination Findings

The woman's physical examination findings were within normal limits in all body systems with the exception of cardiovascular. Upon auscultation of her precordium, a Grade 3/6 murmur and intermittent right ventricular gallop could be appreciated best in the pulmonic position. The systolic crescendo–decrescendo ejection murmur increased with inspiration. There were no palpable heaves or thrills.

### Diagnostic Studies

Pulmonary valve stenosis can lead to changes in the right ventricle and pulmonary artery. The right ventricle hypertrophies in proportion to the degree of valvular stenosis (Elzouki, 2012). It is critical, therefore, to regularly perform diagnostic studies to assess for these changes. Before her surgical consultation, the woman in this case underwent stress testing with bicycle ergometer, an echocardiograhy, and cardiac magnetic resonance imaging (MRI).

The stress test involved a study of the woman's whole body oxygen consumption, carbon dioxide production, 12-lead electrocardiogram, work level, and blood pressure. She completed the test successfully and did not suffer from chest pain, lightheadedness, palpitations, or syncope.

The echocardiogram and cardiac MRI showed a severely dilated right ventricle and severe pulmonary insufficiency but good myocardial function. Marked dilation of the right ventricle is indicative of severe obstruction (Elzouki, 2012).

### Plan

Female CHD patients must receive counseling and intervention before pregnancy (Canobbio et al., 2017). The hemodynamic changes that occur during pregnancy—an increase in heart rate, central venous pressure, and cardiac output—are associated with risks to both the mother and fetus (Canobbio et al., 2017). Mothers with CHD have an increased risk of heart failure, arrhythmia, thromboembolism, aortic dissection, and pregnancy-induced hypertension, and the fetus faces increased risks of preterm birth, intrauterine growth retardation, spontaneous abortion, and fetal/neonatal death (Lu et al., 2015). As illustrated in this case, it has been recommended that when planning for a pregnancy, a patient receive a full assessment of any pulmonary regurgitation because of associated increased risks of pulmonary regurgitation with right ventricular dilatation. It is therefore critical that women with CHD be counseled on the additional risks to mother and fetus resulting from the underlying CHD (Roos-Hesselink & Johnson, 2017).

To provide the woman in this case with the best possible treatment plan, there were three major options posed to her: surgical tissue pulmonary valve replacement, transcatheter pulmonary valve replacement (tPVR), or nonintervention. Of course, each choice was replete with its own unique advantages and disadvantages.

Surgical tissue pulmonary valve replacement involves implanting a bioprosthetic valve in place of the affected pulmonic valve. Evidence indicates that this may be the preferable option before considering pregnancy, if the woman meets specific surgical criteria (Roos-Hesselink & Johnson, 2017). This has typically been regarded as the criterion standard for young adults with pulmonary regurgitation, and it is the most commonly performed procedure in adults with CHD (Jacobs et al., 2015; Suleiman et al., 2015). After infantile valvectomy, the likelihood of right ventricular dilation and dysfunction in adulthood increases—as was the case for this woman (Jacobs et al., 2015). It is therefore indicated that a competent pulmonary valve be used as a means of preventing further valve