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Novel Insights from Clinical Experience

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Severe Pregnancy-Induced Deterioration of Truncal Valve Regurgitation in an Adolescent Patient with Repaired Truncus Arteriosus

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Established Facts

- Due to the progress in the field of thoracic surgery and diagnostics, a growing number of patients with complex congenital heart disease (will) reach childbearing age.
- It has become clear that patients with different types of repaired complex congenital heart disease (e.g., tetralogy of Fallot, pulmonary atresia, transpositions of the great arteries) can successfully carry pregnancies to term.
- Little information is available on the outcome of pregnancy after truncus arteriosus repair.

Novel Insights

- Successful pregnancy after repair for type 1 truncus arteriosus is possible.
- However, as illustrated by this case, worsening of regurgitation of the truncal valve may develop during pregnancy, but can recede spontaneously after delivery.

Key Words
Pregnancy · Truncus arteriosus

Abstract
Truncus arteriosus, a rare and complex congenital heart disease, is hallmarked by a single great vessel (truncus) that arises over a large ventricular septal defect and provides both the pulmonary and systemic circulation. Pregnancy reports after repair for truncus arteriosus are scarce. Therefore, the maternal and offspring outcomes are unknown. We report the outcome of a pregnancy in an 18-year-old woman with repaired truncus arteriosus. Despite severe and symptomatic deterioration of truncal valve regurgitation, she successfully delivered a healthy child, and the valve function recovered within 2 weeks postpartum.

Case Report

An 18-year-old young woman with repaired type 1 truncus arteriosus visited our congenital heart disease outpatient clinic and appeared to be 25 weeks pregnant. Her medical history consisted of a successful surgical repair of the truncus arteriosus at the age of 4 months (weight 4 kg). During this intervention, the ventricular septal defect was closed using a Dacron patch, thereby

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assigning the native truncal valve to the systemic (left-sided) ventricle. The main pulmonary arteries were isolated from the truncus and reimplanted on a Hancock valved conduit connected to the pulmonary (right) ventricle. Reoperation was necessary 5 years later; the obstructed Hancock conduit was replaced by a pulmonary homograft. Throughout childhood and early adolescence, she remained in good clinical health (NYHA class I). No important residual lesions were recorded.

At presentation, she was still asymptomatic, and physical examination showed a normal blood pressure of 120/60 mm Hg and an average heart rate of 65 bpm. On cardiac auscultation, a grade 3/6 systolic murmur without radiation and a grade 2/6 diastolic murmur in the third left intercostal space were heard. No signs of heart failure were recorded. The electrocardiogram showed sinus rhythm, a normal axis and normal repolarization. A transthoracic echocardiogram revealed normal left and right ventricular dimensions and function, a mild to moderate truncal valve regurgitation, an aortic trunk diameter of 40 mm, and a mild stenosis of the pulmonary homograft was documented. These findings were consistent with pre-conceptional examination.

During follow-up, at 30 and 34 weeks, the truncal valve regurgitation deteriorated significantly. At 34 weeks, the echocardiogram documented severe truncal valve regurgitation, but without left ventricular or aortic trunk dilatation. For the first time during her pregnancy, the patient now complained of dyspnoea on exercise (NYHA II). Physical examination revealed a blood pressure of 140/30 mm Hg and a positive Duroziez sign as parameters of a significant truncal valve regurgitation. Both the systolic and diastolic murmur had increased to grade 4/6. There were no signs of heart failure. The following week, the patient's exercise tolerance decreased further (NYHA III) and we then decided to induce labor at 36 weeks of gestation to guarantee maternal and fetal well-being. After multidisciplinary evaluation, induction of vaginal delivery by syntocinon infusion and under epidural anaesthesia was performed. No obstetric complications arose. The patient gave birth to a healthy child weighing 2,940 g. A prenatal echocardiogram had already excluded congenital heart disease of the child. Postpartum, the signs and symptoms resided and the truncal valve regurgitation returned to the pre-conceptional 'mild to moderate degree' on transthoracic echocardiogram performed 2 weeks after delivery. No dilatation of the aortic trunk occurred.

Discussion

Truncus arteriosus, a rare congenital heart disease, is believed to occur less than once in every 10,000 live births. It is frequently (35% of cases) associated with chromosome 22q11 deletion [1]. Without surgical intervention, the first-year mortality is approximately 80%. Nowadays, most patients are repaired in the neonatal period or early infancy. Survival after repair depends on the type of surgical approach, the presence of an interrupted aortic arch in association with truncus arteriosus and the need for reoperation [2, 3]. Pulmonary conduit failure and severe regurgitation of the truncal valve are the main reasons for reintervention [4].

Pregnancy in complex congenital heart disease, even after successful repair, is associated with increased maternal and fetal risks [5–8]. Maternal risks are for the most part related to substantial haemodynamic changes during pregnancy and labour. Circulatory changes inherent to pregnancy include shifts in cardiac output (up to 50% above pre-pregnancy levels), volume loading and alterations in systemic vascular resistance. During labour, the catecholamine secretion (subsequent to anxiety, exertion and/or pain), the auto-transfusion of 500 ml of blood sequestered in the uteroplacental vascular bed and vena cava inferior decompression cause superfluous haemodynamic shifts [5]. Maternal risks are deterioration in NYHA class and valve regurgitation, clinically significant heart failure, especially if fixed obstructive cardiac lesions are present, arrhythmias and thromboembolic events. The frequency of these complications depends on the underlying congenital heart disease, the presence and type of surgical correction, the residual sequelae and the functional capacity of the heart.

Higher rates of miscarriage, premature delivery and offspring mortality are the main fetal risks. In addition, more children are small for gestational age, and recurrence of congenital heart disease also needs to be taken into account.

The outcomes of pregnancy in women with repaired truncus arteriosus are virtually unknown. In the only available report, the outcome of pregnancy in a 31-year-old woman who had undergone repair for truncus arteriosus is described [9]. In this patient, pregnancy was well tolerated. Importantly, due to the late repair (at 13 years of age), this patient also suffered from pulmonary hypertension. Despite this risk factor, pregnancy and delivery could be managed without complications. Our patient was corrected in early infancy and had not developed pulmonary hypertension. Our main concerns were the possible increase in the pre-existing truncal valve regurgitation and the potential dilatation of the aortic (former common) trunk due to the physiological increase in cardiac output and intravascular volume load. Aortic (left-sided) valve regurgitation may increase during pregnancy; however, it is generally well tolerated due to the reduced systemic vascular resistance [10]. Indeed, in the third trimester, the truncal valve regurgitation deteriorated and resulted in a diminished exercise tolerance. Nevertheless, the situation could be handled conservatively. This would support the idea that also in truncal valve regurgitation, conservative management in first instance is warranted.

We advocate multidisciplinary assessment prior to delivery; in the present case report, the decision to induce vaginal delivery was discussed elaborately. A vaginal delivery was chosen based on the fact that the haemodynamic shifts during Caesarean section are generally more pronounced. Moreover, to reduce maternal stress and alleviate the heart, epidural anaesthesia was administered. It needs to be taken into account that epidural anaesthetic regimens may cause hypotension, mainly through their effects on the systemic vascular resistance. On the other hand, the expected mild decrease in systemic vascular resistance would improve the truncal valve regurgitation. In the patient described by Perry, a Caesarean section was performed based on obstetric grounds.

The fact that this young woman became pregnant unexpectedly at the age of 17 illustrates the importance of informing patients early and repeatedly about possible risks associated with pregnancy. The increasing number of women with congenital heart disease reaching child bearing age makes it crucial for the physician and the patient to be informed about potential risks of pregnancy, also in uncommon cardiac malformations.

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