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Case report

Successful pregnancy by in vitro fertilization after Mustard operation for transposition of the great arteries

Hamza Sunman^a, Uğur Canpolat^{a,*}, Hikmet Yorgun^a, Timur Gürkan^b,
Lale Tokgözoglu^a

^a Department of Cardiology, Hacettepe University, Ankara, Turkey

^b Department of Obstetrics and Gynecology, Hacettepe University, Ankara, Turkey

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Summary We report on a 32-year-old woman who had Mustard operation for transposition of great arteries and who underwent successful pregnancy by in vitro fertilization and without peripartum complications.

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Introduction

Transposition of the great arteries (TGA) is a complex cardiac malformation with worse prognosis without surgical correction. Before initiation of surgical treatment, most children with TGA died in early infancy. Since the initiation of surgical procedures such as the Mustard operation, an increasing number of patients may reach adulthood and experience pregnancy [1]. Long-term complications after Mustard operation are composed of heart failure, arrhythmias, pulmonary edema, venous return stenosis, and hemodynamic changes during pregnancy may deteriorate to produce life-threatening complications in these patients [2]. To the best of our knowledge, this case is the first report

of successful pregnancy by in vitro fertilization (IVF) and delivery after Mustard operation for TGA.

Case report

A 32-year-old woman consulted our department for cardiac evaluation by a perinatologist before IVF procedure. IVF was planned because of male factors due to poor sperm morphology. We learned that she underwent Mustard operation for TGA when she was 2 years old. Past medical history was unremarkable except corrected congenital heart disease. She was asymptomatic and had no complications due to her congenital heart disease until childbearing age. She was able to climb more than three flights of stairs. Cardiac examination revealed 3/6 pansystolic murmur at mitral and tricuspid foci. Cardiac catheterization that was performed when she was 18 years old, disclosed that there was no obstruction of venous return and there was normal functioning of atrial baffles with a reasonable size. Before her preg-

* Corresponding author at: Hacettepe University, Faculty of Medicine, Department of Cardiology, Sıhhiye-06100, Ankara, Turkey. Tel.: +90 312 305 1780; fax: +90 312 305 4137.

E-mail address: drucanpolat@yahoo.com (U. Canpolat).

nancy, transthoracic echocardiography (TTE) showed normal ventricular systolic function, right ventricular end-diastolic diameter (RVEDD) of 2.5 cm, moderate valvular regurgitation at anatomic areas (moderate tricuspid regurgitation, mild mitral and aortic regurgitation), normal functioning conduits on both sides, and no interventricular defect. Posteroanterior chest-X-ray showed cardiothoracic ratio of 0.52 with no other changes. Due to her clinical and echocardiographic findings, she was allowed to undergo IVF procedure. Long protocol was used for IVF procedure. Pituitary down-regulation with leuprolide was followed by superovulation with recombinant follicle stimulating hormone (r-FSH). Ten eggs were collected, and six of them fertilized normally. Three embryos were transferred, and the patient had a positive pregnancy test 14 days after the embryo transfer procedure. There was no cardiac and gynecologic complication during the IVF program and pregnancy follow-up. She was closely followed up clinically and by control TTE at all visits during pregnancy. There was only minimal increment in RVEDD (which was 2.7 cm at 2nd trimester and 2.55 cm 6 months after pregnancy) with no changes in valvular regurgitations during the pregnancy period. We have learned that she had successful delivery at term in another center with a healthy newborn. She was asymptomatic and had normal systemic ventricular function at 6th month, first, and second year visits after pregnancy.

Discussion

This case report illustrates IVF pregnancy and successful delivery in a patient who underwent Mustard operation for TGA. Due to improvement in survival, most of these patients have reached their childbearing age and consulted cardiologists and obstetricians because they plan a pregnancy or are already pregnant.

Pregnancy causes hemodynamic and physiologic changes [3], some of them may adversely affect hemodynamic status of patients with congenital heart disease. During pregnancy, there is a 40–50% increase in blood volume and heart rate increases by about 10–20 beats/min. There is also a decrease in systemic and pulmonary vascular resistance. All these changes result in a 30–50% increase in cardiac output, with a maximum rise around 32 weeks of gestation [4,5]. Because of these hemodynamic changes during pregnancy, the risk of complications (systemic RV failure, tricuspid regurgitation, arrhythmias, and pulmonary venous or systemic venous baffle obstruction) may be increased [2,6]. Our patient was asymptomatic and had normal ventricular systolic functions before pregnancy. Due to these findings, we reminded her about risks and allowed her to become pregnant.

Although many patients with surgically corrected congenital heart disease are reaching childbearing age, there is limited experience about normal pregnancy [2,6–9]. Also there was no consensus about optimal maternal age in this type of congenital heart disease. But complication risk increases with older age. Guedes et al. [1] reported the impact of pregnancy on the systemic RV after Mustard operation for TGA in 16 women (mean age at pregnancy was 27 ± 5 years) who completed 28 pregnancies with New York Heart Association heart failure class I-II. There was progression

in RV dimensions (31%), RV systolic dysfunction (25%), and tricuspid regurgitation (50%). Clarkson et al. [9] reported the clinical outcome of 15 pregnancies in 9 asymptomatic women who remained free of cardiac symptoms during or after pregnancy and no significant changes in ejection fraction or degree of tricuspid regurgitation (TR) was seen. Although pregnancy after Mustard operation is well tolerated clinically, it carries the risk of RV dysfunction, which can be irreversible. In our patient, there was no deterioration in functional status, RV systolic function, and tricuspid regurgitation. These preserved functions continued to be normal after the postpartum period.

There are no data for IVF pregnancy outcomes in patients with Mustard operation for TGA. An IVF program carries a risk due to some complications. During an IVF program, the most serious complication which is related to the cardiovascular system is ovarian hyperstimulation syndrome (OHSS). OHSS is an acute and self-limiting but potentially life-threatening condition appearing within the first week after ovulation is induced. It is secondary to circulatory dysfunction caused by simultaneous occurrence of increased vascular permeability and arteriolar vasodilation that leads to ascites formation, arterial hypotension, tachycardia, increased cardiac output, reduced peripheral vascular resistance, stimulation of renin-angiotensin system, sympathetic nervous system, hemoconcentration, oliguria, and in extreme cases, renal failure and thrombotic events [10–13]. Also, problems associated with twin and higher order pregnancies have assumed major importance, with international debate about multiple pregnancies, the biggest risk with IVF. The latest data from the USA [14] concerning multiple pregnancies showed that 32% of deliveries after IVF were twins and 7% were triplets or more. Multiple pregnancy causes maternal and fetal complication risk. So, IVF programs before successful pregnancy carry additional risk for these patients. Because there is no evidence related to increased cardiac complications of IVF pregnancies in asymptomatic patients with Mustard procedure for TGA, pregnancy by IVF was allowed in our case. There was no complication in our patient related to the IVF program. She had an uneventful pregnancy with frequent follow-up visits. She did not experience any serious complication such as arrhythmia, heart failure, or pulmonary edema during the pregnancy.

This to our knowledge is the first case of a patient who underwent IVF pregnancy and successful delivery after Mustard operation for TGA. It should be highlighted that collaboration between perinatologists and cardiologists experienced in the care of patients with congenital heart disease is important during follow-up of these patients.

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