Uncorrected tetralogy of Fallot and pregnancy: a case report

C.P. Katsetos[1], M.B. Kontoyannis[1], A. Koumousidis[1], Ol. Petropoulou[2], C. Delos[3], M. Katsoulis[1]

[1]Department of Obstetrics and Gynaecology, [2]Department of Anaesthesiology, [3]Department of Cardiology
"Tzaneio" General Hospital of Piraeus, Piraeus (Greece)

Summary

We report a case of pregnancy in a 34-year-old woman with uncorrected tetralogy of Fallot (TOF). There are more risks in patients without surgical correction. In our case, haemoglobin and haematocrit were higher, oxygen saturation was lower, and right ventricular enlargement was observed. Pregnancy was resolved successfully by caesarean section. Improvement of fetomaternal outcome may be related to corrective procedures before conception to achieve better functional heart capacity. Delicate multidisciplinary medical management is essential for these limited cases to achieve optimal prognosis.

Key words: Tetralogy of Fallot and pregnancy; Heart disease in pregnancy; Heart disease and mode of delivery; TOF and mode of delivery.

Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease (15%), characterized by ventricular large septum defect, pulmonary stenosis, aortic riding of septum communication, and right ventricular hypertrophy. TOF is treated surgically. For cases of TOF surgery is the only possible therapy - sometimes a temporary operation is performed when the patient is a small child. Later in life complete surgical repair must be carried out [1].

Case Report

A 34-year-old woman, gravida 2, para 0, was admitted to hospital at 38 weeks of gestation for elective caesarean section. There was a medical history of tetralogy of Fallot (TOF) with a palliative Blalock-Taussig shunt, osculation between the subclavian artery and pulmonary arteries, in addition to mild renal failure and mild cyanosis. The patient had explicitly refused for years to submit to complete surgical treatment of her cyanotic heart disease. There was no other significant family history.

The drugs used during pregnancy were allopurinol (100 mg) and lasix tablets (1 x 2 bid). Cardiologic examination revealed a heart rate of 80-100 bpm, blood pressure (BP) of 120/87 mmHg with a normal sinus rhythm, right ventricular hypertrophy, left atria enlargement, a rightward axis and nonspecific T-wave abnormality. There was also a huge interventricular septal defect, situs solitus, overriding of the ectatic aorta over the interventricular septum, severe constriction of the exitus of the right ventricle and pulmonary artery. The patient's pregnancy had been uneventful and continued normally without complications.

A week before the scheduled caesarean section, a full clinical examination revealed dyspnoea during stress, mild cyanosis, pulse of 98 bpm, BP of 135/90 mmHg, haematocrit of 52%, haemoglobin of 16.5 g/dl, systolic murmur of the pulmonary artery and continuous puffs in the precordium.

On admission to the hospital at 38 weeks of gestation, the presentation of the foetus was cephalic and the cardiotocography (CTG) trace was normal. There was no uterine activity noted. At the vaginal examination there was no effacement of the uterine cervix. Cervical dilation was 4 cm and the membranes were intact. The patient's pulse was 84 bpm and her blood pressure 95/70 mmHg. Endocarditis prophylaxis was given before the surgical procedure.

On admission to the theatre epidural anaesthesia was performed and right after that a low transverse elective caesarean section was completed. A female neonate was delivered weighing 2,140 g, with an Apgar score of 8-10, at 1 and 5 min, respectively. The baby was in good condition. The patient remained stable during the operation as well as after the delivery without any haemodynamic or ECG changes. The mother was eventually transferred to the Intensive Cure Unit (ICU) on antibiotic treatment with ampicillin IV. Due to long term use of cardiological medication she was advised against lactation. During her stay in the ICU she had three episodes of ventricular tachycardia and swelling of the lower extremities. No proteinuria was noted. She was treated with hydrochloride amiodarone IV (150 mg daily).

There was rapid clinical progress over the next 24 hours so the patient was transferred to the ward. Blood tests revealed the following results: Hct 40.6%, Hb 13.5 g/dl, PLT 166,000 k/ul (with no signs of renal failure), urea 49 mg/dl, and creatinine 1.0 mg/dl. LDH 296 U/l, fibrinogen 4.32 g/l and INR 1.02 were all within normal values. Mother and baby were both discharged seven days after the caesarean section with appropriate diuretic therapy through salt restriction and a special low calorie diet. She was also advised bed rest.

At the follow-up appointment almost a month after the delivery, there were no signs of cardiac disturbances. Five months after delivery she was admitted to the hospital in the cardiology clinic with ventricular tachycardia and episodes of unconsciousness. She was discharged 24 hours later on amiodarone (200 mg tablets bid) and lasix tablets (1 x 2 bid). Unfortunately she did not attend the follow-up appointments the first or second year.

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Discussion

TOF is the most common cyanotic congenital heart disease (15%), characterized by ventricular large septum defect, pulmonary stenosis, aortic riding of septum communication, and right ventricular hypertrophy. TOF is treated surgically. In our case a temporary operation was performed a Taussig-Blalock shunt at the age of seven years.

Uncorrected TOF during pregnancy belongs to Group 2 - moderate risk (5-15%) of maternal mortality. A fall in peripheral resistance during pregnancy and hypotension during labour may increase the right to left shunt and aggravate preexisting cyanosis. A rise in blood volume and venous return to the right atrium along with a fall in systemic vascular resistance increase the right to left shunt and cyanosis. Close monitoring of systemic blood pressure and blood gases during labour is essential. Any further systemic (drug induced) vasolidation should be avoided [1, 2]. Maternal and foetal complications are tied to the degree of maternal cyanosis. The risk is high when oxygen saturation is ≤ 85%. There is a risk of 5% probability of transmission to the offspring and in 15% studies have shown deletion of the short arm of chromosome 22 (genetic cause). Recurrent risk when the father is affected is 1.5%; with an affected mother it is 2-3%, so in one sibling the risk is 2.5 times and in two siblings 8 times [1].

Maternal death rates have been quoted as high as 50% with preterm delivery rate at 55%, IUGR rate at 30% and perinatal mortality rate at 28% [3].

During pregnancy the risk of right ventricular insufficiency and hypoxic attack is increased [4]. There may also be an increased shunt and worse acidosis. Uncorrected TOF in pregnancy leads to deterioration, when the arterial saturation of oxygen is < 85% and the Hct > 60%. In those patients the most significant predictors of foetal hypoxia are the mothers' persistent cyanosis and congestive heart failure. The prognosis is less favourable if there is already myocardial compromise before pregnancy. In addition, the frequency of abortion, premature birth, foetal distress and congenital malformation of the child is 57% [5]. The indication of assisted delivery in those patients depends on the severity of the disease. In our case caesarean section was indicated and the technique of anaesthetics (epidural anaesthesia) was carefully chosen [6].

On the contrary, the risk of pregnancy in repaired patients depends on their haemodynamic condition. Generally, the risk is low in patients with efficient repairs. In patients with significant residual right ventricular out flow obstruction, severe pulmonary regurgitation with or without tricuspid regurgitation and/or RV dysfunction, the increased volume load of pregnancy may lead to RV failure and arrhythmias [2, 7].

In a study of the American College of Cardiology [8] on 72 women, 43 women with TOF had 112 pregnancies. Eighty-two of these were successful. Eight women had uncorrected TOF at the time of their 20 successful pregnancies. One percent of these women had preterm labour, 8.5% of foetuses were small for their gestational age, six out of seven were low birth weight infants and 6% of infants had major congenital abnormalities (e.g., congenital heart disease/stomach outlet obstruction, etc.). Finally, 24% of the above pregnancies had adverse obstetrical outcomes [8].

To our knowledge, there is one report as well as four case reports of delivery in uncorrected TOF, but only one without aortic regurgitation [8-10]. One case had a successful delivery by caesarean section - a patient with TOF following a Taussig-Blalock shunt [11, 12]. In that case, there was intrauterine growth restriction and pregnancy-induced hypertension. Our case did not demonstrate such features.

All patients with TOF should have genetic counselling preconception with assessment in case of 22q11 deletion syndrome using fluorescent in situ hybridisation (FISH). In its absence the risk of defects in the foetus is low (4%) [13].

It is essential for high-risk patients with congestive heart disease like TOF either uncorrected or with a Tausig-Blalock procedure when becoming pregnant to be supervised by a multidisciplinary team including foetal medicine specialists, cardiologists and anaesthetists.

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Address reprint requests to:
A. KOUMOUSIDIS, M.D.
Grigoriou Lambraki 112-114
Pireas, pc 18532 (Greece)
e-mail: kumusidi@yahoo.gr