

Pregnancy with Uncorrected Tetralogy Of Fallot :Anaesthetic Management Of A Case For LSCS

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Abstract

Among the congenital heart diseases causing intra cardiac R-L shunts Tetralogy of Fallot is the most common syndrome, characterized by presence of VSD, aortic overriding, pulmonary artery outflow obstruction and right ventricular hypertrophy. If remains uncorrected it can cause significant morbidity and mortality to the patient. Pregnancy in such a patient presents furthermore challenges and worsening of symptoms. Anaesthetic management of patients with TOF requires thorough understanding of anatomical defects and its physiological adaptations, and also the events and drugs that can alter the magnitude of R-L shunt. Problems in such patients are of chronic hypoxia, polycythemia and coagulopathy, CHF, embolism, episodic and reactive pulmonary vasoconstriction and altered acid base status. We present a case of uncorrected tetralogy of fallot (TOF) who underwent emergency caesarean section.

INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect and accounts for 5% to 6% of congenital heart malformations¹. Its hallmark anterior and superior infundibular septal displacement gives rise to the tetrad of ventricular septal defect, aortic override, infundibular obstruction, and right ventricular (RV) hypertrophy². Without surgical repair 25~35% die in the first year of life, 40~50% die by the age of 4, 70% by 10 years, 95% by 40 years³ and With complete repair 85% survive to adulthood.

Women with uncorrected TOF do poorly during pregnancy and maternal mortality approaches 10%⁴. Any disease complicated by severe maternal hypoxemia is likely to lead to miscarriage, poor fetal growth, preterm delivery or fetal death. There is a relationship between chronic hypoxemia and the polycythemia it causes with the outcome of pregnancy. When hematocrit rises above 65%, pregnancy wastage is virtually 100%⁴.

Stillbirth rates of 14% and fetal growth retardation of 36% of pregnancies in women with cyanotic heart disease has been reported⁵. With satisfactory surgical correction prior to pregnancy, maternal risks are decreased dramatically, and fetal environment is improved. Intracardiac repair has permitted survival into the childbearing years and excellent quality of life⁶. Long-term complications usually relate to

functional competence of the RV outflow tract and its secondary effects on ventricular and atrial myocardial function. Pulmonary regurgitation or stenosis may result in RV dysfunction and failure, progressive tricuspid valve regurgitation, atrial and ventricular arrhythmias, and sudden cardiac death⁷.

CASE REPORT

A 28 year female primigravida was posted for emergency cesarean section, having complaints of dyspnoea, early fatigability and history of cyanotic spells during heavy exertion before her pregnancy. All her symptoms exaggerated during pregnancy and she was diagnosed a case of "tetralogy of fallot" with right to left shunt. She had history of taking beta blocker (propranolol) and digitalis off on. Her cesarean section was decided because of failed progression of labor because of cephalo-pelvic disproportion. She was found cyanotic and her SpO₂ was 85-86%. Clubbing and murmur was present. Her ABG showed PaO₂-61mmHg PaCO₂-31mmHg pH 7.46. Haemoglobin in ABG 16 gm% . As patient came in emergency her other investigations were not available at time of surgery. General anaesthesia was planned for her surgery. Ranitidine and Metaclopromide was given half an hour before surgery. Amoxicillin was also given for prophylaxis. ECG, NIBP and SpO₂ monitors were attached. 500 ml Ringer lactate given and patient was induced with

ketamine 2mg/kg and Thiopentone 1mg/kg. Scholine 1.5mg/kg was given and her trachea was intubated with rapid sequence. Vecuronium 0.08mg/kg was given when effect of scholine was washed out. Anaesthesia was maintained with 50% O₂ and 50% N₂O and sevoflurane. Phenylephrine 50 µgm bolus repeated on 20 minute to increase SVR and maintain saturation above 95%. After delivery of baby there was sudden fall of saturation, ringer's lactate 200ml and phenylephrine 50 µgm given but saturation was not improved and it was diagnosed as Tet spell and Propranolol 1mg was given after which she gained SpO₂ 90%. Fentanyl 2µgm/kg was also given. Oxytocin was avoided. Uterus contracted slowly spontaneously. On completion of surgery patient was reversed with neostigmine 0.05mg/kg and glycopyrolate 0.01 mg/kg. and she gained consciousness. In postoperative period she had SpO₂ of 85-86%. She was further referred to cardiothoracic department.

DISCUSSION

Maternal heart disease complicates 0.2 to 3 % of pregnancies, and congenital heart lesions now constitute at least half of all these cases⁸. The classical and most commonly encountered (10% of all CHD) congenital cardiac lesion in pregnancy is the tetralogy of Fallot (TOF). A maternal mortality report from UK had 35 death associated with cardiac disease, 29% were due to congenital heart disease, 15 % due to IHD and the remainder due to other acquired cardiac conditions⁹.

The complex of anatomic malformation results from an anterior displacement of the conoseptum toward the right ventricle creating a malalignment VSD and a narrowing of the outflow tract of the right ventricle (RV)². The aorta is displaced anteriorly, straddling the muscular septum and arising from both ventricles. The obstruction to outflow of the RV usually involves the infundibulum of the RV but can arise from the pulmonary valve, its annulus, the main pulmonary artery or even in the peripheral pulmonary arteries.

Elevated pressures in the RV from outflow obstruction and exposure to systemic pressure from overriding aorta lead to compensatory RV hypertrophy. The main characteristic of TOF is cyanosis. Cyanosis can result from three separate Mechanisms. Inadequate pulmonary blood flow, right to left shunting or intrinsic pulmonary disease. In TOF, cyanosis results from a right-to-left shunt at the level of ventricles and inadequate pulmonary blood flow. Because of the outflow

obstruction, blood ejected from RV crosses the VSD and enters the overriding aorta. This reduces the amount of pulmonary blood flow available for oxygenation and adds desaturated blood to the systemic circulation. Pressures in the right ventricle are near to the systemic pressure. The likelihood of a favorable outcome for the mother with TOF depends upon the functional cardiac capacity of the patient before pregnancy, other complications that further increase cardiac load, and quality of medical care provided throughout pregnancy and surgical correction of the anomaly before conception. Pregnant mothers with TOF are affected differently depending upon if they remain uncorrected, have palliative or definitive procedure or they have residual defects after these procedures.

The principle danger for a pregnant woman with TOF is cardiac decompensation because of inability to meet the additional demands imposed by the physiological changes of pregnancy and parturition. If present, infection, hemorrhage and thrombo-embolism compound the risk. The cardiovascular changes of pregnancy may unmask residual or recurrent TOF in patients with corrective procedures, who have been asymptomatic throughout their life after TOF repair⁶.

Before successful intracardiac repair of TOF was introduced in the 1950s, few patients reached childbearing age, and successful pregnancy was uncommon. Pregnancies were characterized by spontaneous abortions, stillbirths, and premature deliveries. Presbitero et al. demonstrated that the most important risk factor for adverse fetal outcome in cyanotic patients was the degree of cyanosis. These authors suggested that an arterial oxygen saturation >85% and a hemoglobin concentration <18 g/dl were more likely to result in live birth, whereas hemoglobin concentrations >20 g/dl were associated with adverse fetal outcome¹⁰.

Chronic hypoxemia in such patients leads to adaptations to provide adequate tissue oxygenation ie. polycythemia, increased blood viscosity, vasodilatation, hyperventilation and chronic respiratory alkalosis. Such adaptive mechanisms may limit cardiac reserve and O₂ delivery during stress¹¹. As no specific technique is suggested for such patients and anaesthesia carries considerable risk, the management should be based on avoiding changes that would increase the magnitude of R – L shunt, dehydration should be avoided to improve the circulating volume by decreasing the blood viscosity¹².

Both general and regional techniques have been employed

successfully in parturient with TOF¹³. Regarding cesarean section, general anesthesia (GA) is probably the technique of choice¹⁴. GA with endotracheal intubation provides airway protection, eliminates work of breathing and may reduce oxygen consumption. The complications of controlled mechanical ventilation include decreased venous return as well as ventricular dysfunction, compression of pulmonary vessels, hypoxemia, hypo or hypercarbia and acidemia. The choice of anesthetic drugs may not be of prime importance. In patients with dynamic right ventricular outflow obstruction, increases in heart rate and contractility should be avoided as they will worsen the obstruction and cyanosis. Anesthetic drugs and adjuncts having vagolytic or sympathomimetic effects should preferably be avoided. Regional anesthesia allows spontaneous respiration with little disruption of V / Q relationships, which may be critical in parturient with less severe TOF. Epidural catheter techniques offer continuous, titrated anesthesia or analgesia.

In this case we used general anaesthesia as patient came as emergency with known TOF but without taking any medicine regularly and with cardiac decompensation. She had not gone any palliative or corrective procedure. Our goal was to maintain SVR and decrease PVR¹⁵. To achieve this ketamine was used for induction and phenylephrine to increase SVR. She was preloaded with 500ml RL before induction. Sevoflurane was used for maintenance in low concentration with 50% N2O. Patient was hyperventilated to maintain slightly alkalosis. Oxytocin was not used to avoid sudden increase in preload. Invasive monitoring CVP and arterial blood pressure could not be monitored in that case because of unavailability. Patient was managed properly.

Patients with tetralogy of Fallot with pregnancy need special care by a team consisting of obstetrician, cardiologist and anesthesiologist among others.

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