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

Abstract
Among the congenital heart diseases causing intra cardiac R-L shunts Tetralogy of Fellot 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 fellot (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 malformations. 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) hypertrophy. 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 years and With complete repair >85% survive to adulthood.

Women with uncorrected TOF do poorly during pregnancy and maternal mortality approaches 10%. 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%.

Stillbirth rates of 14% and fetal growth retardation of 36% of pregnancies in women with cyanotic heart disease has been reported. 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 life. 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 death.

CASE REPORT
A 28 year female primigravida was posted for emergency cesarean section, having complaints of dyspnoea, early fatigue 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 (propanolol) 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 $SpO_2$ was 85-86%. Clubbing and murmur was present. Her ABG showed PaO$_2$-61mmHg, PaCO$_2$-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 $SpO_2$ 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 µg/ml 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 µg given but
saturation was not improved and it was diagnosed as Tet
spell and Propanolol 1mg was given after which she gained
SpO₂ 90%. Fentanyl 2µg/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 cases. The classical and most
commonly encountered (10% of all CHD) congenital cardiac
lesion in pregnancy is the malalignment VSD of TOF (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 conditions.

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). 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 anulus, 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
repair.

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
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
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repair.

Chronic hypoxemia in such patients leads to adaptations to provide adequate tissue oxygenation i.e. polycythemia,
increased blood viscosity, vasodilatation, hyperventilation
and chronic respiratory alkalosis. Such adaptive mechanisms
may limit cardiac reserve and O₂ delivery during stress.
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
viscosity.

Both general and regional techniques have been employed
successfully in parturient with TOF. Regarding cesarean section, general anesthesia (GA) is probably the technique of choice. 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 sympathomimetics 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 PVR. 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 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.

References
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