Anaesthetic Management for Emergency Caesarean Section of Patient with Unrepaired Tetralogy of Fallot with Absent Pulmonary Valve (Severe Annular Stenosis) - A Case Report

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Abstract
As tetralogy of fallot (TOF) is one of the most common congenital cyanotic cardiac disease with combinations of abnormalities like ventricular septal defect (VSD), Pulmonary stenosis, overriding of aorta and right ventricular hypertrophy (RVH)¹. Uncorrected TOF results in increase incidence of complications and morbidity. Management of such cases are always challenging.

We are presenting a case report of emergency caesarean section in a patient of TOF with absent pulmonary valve due to severe annular stenosis and not on regular medication. She was diagnosed case of TOF since birth.

Keywords- tetralogy of fallot (TOF), LSCS, General anaesthesia, severe pulmonary stenosis

CASE REPORT
A 20 yr old 40 kg female was posted for emergency caesarean section. She was diagnosed case of TOF since birth and advised correction surgery. But surgery was postponed seven years back due to low haemoglobin level. After that she was not on regular treatment, also not went for surgery. She had continued pregnancy against gynaecologist advice and had undergone prenatal check up at peripheral hospital. She had history of breathlessness on minimal work and paroxysmal nocturnal dyspnoea. She was not on any cardiac treatment. She was posted for emergency caesarean section for failure to progress.
Her preanesthetic examination revealed she was conscious, oriented, dyspnoeic, nil by mouth since 6 hours. She had pallor without cyanosis or clubbing. All peripheral pulses felt. Her pulse rate was 134 /min. regular. Blood pressure was 130/80 mm hg in right upper limb in supine position. Her oxygen saturation was 95-96% on air. Her respiratory rate 36-40/min. on auscultation, respiratory system normal. On cardiovascular system, harsh systolic murmur heard all over precordium. Neurological examination normal. Airway modified mallampatti score 1.

Her investigations showed- haemoglobin 8.8gm%, PCV-27, platelet 2.31,000/cmm, Peripheral smear showed microcytic hypochromic RBCs. International normalised ratio-1.92. Serum electrolytes normal. 2D echo done in first trimester showed tetralogy of fallot with absent pulmonary valve, large subaortic VSD with bidirectional shunt, overriding of aorta >50%,grossly dilated pulmonary artery and branches, severe pulmonary valve annular stenosis with gradient 120 mm Hg, free pulmonary regurgitation present, left ventricular ejection fraction-60%.

Anaesthetic management- General anaesthesia was planned after taking written valid informed consent. Prophylaxis for infective endocarditic was given. Monitors for ECG, SPO₂, NIBP, ETCO₂ were attached. Patient was premedicated intravenously with inj.ranitidine 50mg, inj.metoclopramide 10mg, inj.glycopyrolate 0.2 mg IV. After painting and draping of surgical field, patient was induced with 1.25% inj thiopentone 5mg/kg slowly and inj. suxamethonium 2mg/kg IV. Patient was intubated with portex cuffed endotracheal tube of 7 mm. After confirmation of bilaterally equal air entry, endotracheal tube fixed. The patient was maintained on 50% O2:50% N20:Sevoflurane and intermittent dose of suxamethonium (0.4mg/kg slowly) .After delivery of baby, inj.midazolam 1mg and inj. Fentanyl 1.5mg/kg and inj. oxytocin 10mg (slowly as infusion) was given to patient. Her uterus was well contracted. After induction heart rate of patient came down from 140 to 90/min and was remained between 80-90/min. also blood pressure maintained between 110/76 to 136/82 mmhg and SPO₂ >96% during procedure. After confirmation of haemostasis, skin wound was closed. The procedure was completed in 35 min. and was uneventful. The total blood loss was 300 -400 ml approximately and urine output was 50 ml. The baby cried immediately after birth and Apgar score was7/10.

After fulfilment of all extubation criteria, gentle oral suction was done and patient was extubated uneventfully. Her pulse rate was 92/ min regular, blood pressure 130/74mmhg and Spo₂ 97% with O₂ supplementation on mask 2l/min.

She was observed surgical intensive care unit. Post operative analgesia was maintained with inj. tramadol. In postoperative period patient received 350 ml of packed RBC. Fluid management according to urine output.

Patient was discharged after seven days and advice her follow up in cardiothoracic and cardiology department.
DISCUSSION

Tetralogy of fallot is congenital heart disease due to anatomical defect in heart structure at birth. It involves four heart defects-

1. A LARGE VENTRICULAR SEPTAL DEFECT (VSD)
   Defect or hole in the septum separate right and left ventricle. This hole allows oxygen rich blood from left ventricle to mix with oxygen poor blood of right ventricle.

2. PULMONARY STENOSIS (PS)
   This defect is due to narrowing of pulmonary valve leading to obstruction to blood flows from right ventricle to pulmonary artery. The severity of pulmonary stenosis or right ventricular outflow tract (RVOT) obstruction graded by peak gradient across RVOT such as-TRIVIAL (gradient <25%), MILD (25-49%), MODERATE (50-79%) and SEVERE PS (>79%).
   Absence of pulmonary valve to due severe annular stenosis leads to pulmonary regurgitation which is often associated with massive dilation of pulmonary artery.

3. OVERRIDING OF AORTA-
   In healthy normal heart, aorta is attached to left ventricle allowing only oxygen rich blood to flow to body. In TOF, the aorta is between left and right ventricle, directly over VSD. As a result oxygen poor blood from right ventricle flows directly into aorta instead of pulmonary artery.

4. RIGHT VENTRICULAR HYPER-TROPHY (RVH)-RV
   thickening occurs as heart has to pump harder due to narrow pulmonary valve.

PREGNANCY IN UNCORRECTED TOF PATIENT

During pregnancy, many physiological changes occurred in body. As per the cardiovascular system is concerned, there is a rise in stroke volume and cardiac output by 30-50%, rise in blood volume 40-50%, heart rate rise by 10-15/min. (4)

All these changes aggravate the symptoms of pulmonary stenosis and pulmonary hypertension. Also, shunt reversal chances increases. This leads in increase in haematocrit and coagulopathy, cyanosis worsens. Changes in haemoglobin and oxygen saturation are most important factor for fetal outcome. So while managing such patients, complications like coagulopathy, chronic hypoxia, congestive heart failure, embolism, polycythemia, acid-base disturbances, pulmonary vasoconstriction may be present. (2,3)

During anaesthesia management of such patients, it is important to understand anatomical defect and changes in cardiovascular physiology to avoid adverse maternal fetal outcome.

The main aim during anaesthesia management is to avoid or decrease shunting(R→L) by maintaining systemic vascular resistance, avoid decrease in peripheral vascular resistance and increase in pulmonary vascular resistance and also avoid hypotension. Monitoring of oxygen saturation, blood pressure, ECG is essential (4). All emergency drugs should be available specially phenylephrine, beta blockers. Beta blockers are useful for treatment of cyanotic spell by reducing infundibular spasm of pulmonary valve and
RVOT obstruction, decrease in heart rate. Intraoperative hyperventilation helps in preventing hypoxia, hypercarbia and acidosis to avoid increase in pulmonary vascular resistance. Inj. Ketamine can also be used for induction so as to maintain systemic vascular resistance. In our case patient already had tachycardia so we avoided it. Postoperative analgesia and fluid management is essential.

CONCLUSION
Unrepaired TOF with absent pulmonary valve due to severe annular stenosis are rarely survive till child bearing age and also complete full term antenatal period without any medication. Management of such cases are always challenging as they carry high risk of morbidity and mortality, such cases needs integrated team approach which includes obstetrician, anaesthesiologist, cardiologist and intensivist at fully equipped centres.

ACKNOWLEDGEMENT
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