Anaesthetic management of a patient with pentalogy of Fallot: a case report

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Pentalogy of Fallot (POF) is a form of congenital heart disease having tetralogy of Fallot with an atrial septal defect. It is characterised by right to left intracardiac shunt with a decrease in pulmonary blood flow and the development of arterial hypoxaemia. Arterial hypoxaemia depends on the magnitude of shunting and leads to erythrocytosis which in turn poses a risk of thromboembolism. We report on the anaesthetic management of a rare case of a 4-year-old male child with pentalogy of Fallot for total correction.

Keywords:
anesthetic management, congivital heart disease (CHD), pentalogy of fallout (POF)

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Introduction

The pentalogy of Fallot (POF) is a rare cyanotic congenital heart disease. It consists of atrial septal defect (ASD) with tetralogy of Fallot (TOF) [1]. TOF consists of interventricular septal defect (VSD), obstruction of the outflow tract of the right ventricle (RV), right ventricular hypertrophy (RVH), and biventricular origin of the aortic valve. In children, the infundibulum is prone to spasm leading to hypoxaemia (Tet spells). In Tet spells acute severe obstruction of the RV tract occurs which in turn leads to syncope. This is precipitated by crying, agitation, pain, injury, or fright, that is, conditions that increase sympathetic activity and cardiac contractility, leading to infundibular spasm. Induction of anaesthesia is challenging and hazardous if intravenous access is not obtained before. In a nonrestrictive VSD there is equalisation of pressures in both the ventricles and the degree of cyanosis/shunting is dependent on systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR). A fall in the SVR as seen in hypovolemia, acidosis, and hypoxia causes an increase in PVR (infundibular spasm) which in turn causes right to left shunting and worsening of cyanosis. Associated modalities with risk in these congenital heart diseases are infective endocarditis, coagulation defects due to deficiency of vitamin K-dependent clotting factors in the liver and defective platelet aggregation and brain abscess. Anomalies which occur with TOF include right aortic arch in 25% of cases, ASD (POF) in 15% of patients and coronary anomalies in the rest. It is very challenging for the anaesthetist to manage such cases because of haemodynamic variations intraoperatively.

Case report

A 4-year-old, male child weighing 10 kg was scheduled for cardiac surgery. The patient had a history of cyanosis and breathlessness during exertion and crying since birth. He had a history of difficulty in feeding and failure to gain weight.

On preoperative examination, the patient had no history of any upper respiratory tract illness, any other chronic illness, any drug allergy, or previous surgery. The patient was on tablet propranolol. On general physical examination the patient was thin built and poorly nourished. He had central cyanosis and clubbing. Bilaterally equal air entry was present and no added sound was present on auscultation. A prominent apex impulse was seen in fourth intercostal space. On auscultation S1 and S2 was present with systolic ejection murmur at the upper left sternal edge.

In investigations ECG showed features of right axis deviation and RVH. Chest radiograph showed left-sided aortic arch and oligemic lung fields. Complete blood count revealed haemoglobin 16.3 g% with normal coagulation profile. Echocardiography showed a 14 mm large subaortic perimembranous ventricular septal defect, 15 mm large ostium secundum ASD, predominant left to right shunt, and severe valvular and subvalvular infundibular pulmonary stenosis with a pressure gradient of 100 mmHg and a thickened pulmonary valve, trivial tricuspid regurgitation with less than 50% aortic override, dilated and hypertrophied RV, dilated right atrium (RA), and left ventricular ejecton fraction (LVEF) of 55%. Computed tomography angiography revealed a small main pulmonary artery

in the subpulmonic region (diameter, 4.5 mm); the size of the right pulmonary artery was 6.1 mm and the left pulmonary artery was 8.6 mm with an overriding aorta with normal ascending and descending aorta.

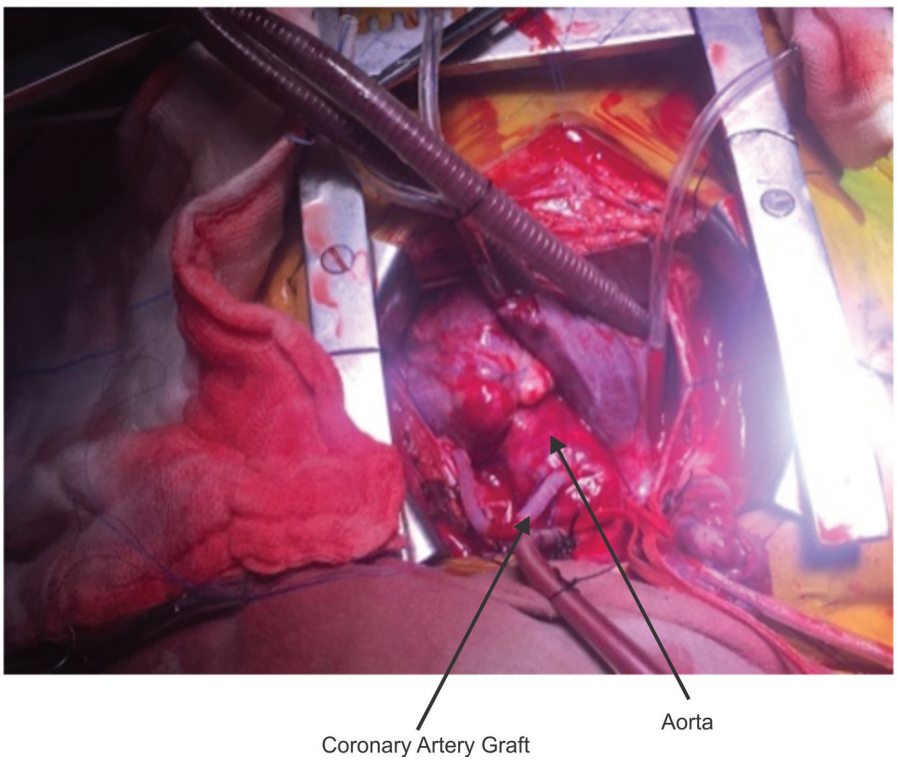
Anaesthetic management

Preoperative vitals were: pulse 102 beats/min, blood pressure 106/54 mmHg, SpO_2 was 78%; after intravenous access the patient was preoxygenated with 100% oxygen for 5 min. Premedication included midazolam 0.4 mg, fentanyl 20 µg given intravenously. Induction was done with ketamine 20 mg and rocuronium 10 mg intravenously with intermittent positive pressure ventilation. The patient was intubated with a 4 mm cuffed EndoTracheal Tube (ETT). Bilateral air entry was checked and the endotracheal tube was fixed at 12 mark at the angle of mouth. Central venous cannulation in the right internal jugular vein and arterial cannulation in right femoral artery were performed. Maintenance of anaesthesia was achieved with 100% oxygen; inhalational sevoflurane and fentanyl 10 µg intermittently and vecuronium 0.4 mg and midazolam 0.08 mg intravenously were given. After incision, the surgery started. Ventilation was stopped transiently during sternotomy. A dose of 40 mg heparin was given intravenously before aortic cannulation. When the activated clotting time (ACT) was above 500 s, the patient was taken on cardiopulmonary bypass and ventilation stopped after achieving full flows.

Myocardial protection was done by using cold cardioplegia solution into the aorta at a pump flow rate of 250 ml/kg/min using a membrane oxygenator after priming the pump with albumin, and repeating half of the cardioplegia solution every 20 min after the initial dose.

Iatrogenic left coronary artery got injured because of its abnormal route. A venous graft was anastomosed between the left coronary artery and the aorta (Fig. 1). The right ventricular outflow tract (RVOT) was exposed through the RA and tricuspid valve (TV) and resection of the infundibulum was carried out. Corrective repair of TOF included the VSD closure with Dacron synthetic patch (which would direct the blood flow from the left ventricle to the aorta). The stenosis of the pulmonary valve and the RV outflow tract was enlarged by resecting the fibres/bands. The ASD defect was repaired with a pericardial patch. The patient was tried to wean off from cardiopulmonary bypass. But the blood pressure was unable to reach above 64/42 mmHg. RV pressure measurement was done and it was 72/48

Figure 1



Sephenous graft anastomosis between the coronary artery and the aorta.

mmHg. As the RV/LV ratio was more than 0.7, the patient was again taken on Cardiopulmonary Bypass (CPB) on full flows. The facility of transesophageal echocardiography is not available in our institute. So we had this option only of comparing the pressures between the two ventricles. Transannular pericardial patch was put and the repair was extended to the main pulmonary artery. Then again trial was attempted to wean off from the bypass; this time the pressure was 68/44 mmHg. Then an injection of dopamine 5 μg/kg/min was started along with an injection of 0.05 μg/kg/min. After that the patient's blood pressure was about 80/35 mmHg; the patient was gradually weaned off from CPB with the additional support of adrenaline in a low dose of 0.01–0.03 μg/kg/min. Protamine was given and an ACT of 98 s was obtained. The patient was shifted to the ICU on ventilator with a pulse rate of 98 beats/min and blood pressure of 96/48 mmHg.

Discussion

TOF is associated with various abnormalities such as that of the right aortic arch, ASD (POF), and anomalous coronary arteries. An additional ASD is seen in 3–5% of cases of TOF. Ostium secundum type is the most common type of ASD in TOF, followed by ostium primum type and rarely sinus venosus type [1]. Coronary artery anomalies are also present in 5–12% patients with TOF. Of the greatest concern are those anomalies in which the aberrant artery courses anteriorly across the RV outflow tract. In these cases while performing right ventriculotomy for TOF repair, inadvertent transection of such a vessel adds considerably to surgical mortality and morbidity [2]. In these patients, alternative surgical techniques such as the transatrial-transpulmonary approach [3], modified right ventriculotomy, and an extracardiac RV to pulmonary artery conduit should be used [4]. Although multidetector coronary computed tomography and MR angiography can be used as alternatives to catheter angiography for evaluating the coronary anatomy in patients with congenital heart disease [5].

Anaesthesia can be induced with a combination of ketamine and fentanyl and maintained with a volatile agent. The anaesthetic goals include the following:

(1) The maintenance of adequate SVR to limit the R-L shunting through the VSD. Sevoflurane is a good choice as it has the least effect on SVR [6]. Isoflurane is a poor choice as it causes vasodilatation and tachycardia [7]. Ketamine and sevoflurane were used to maintain the SVR before bypass in our case.

(2) Treatment of right ventricular dysfunction which might result as a consequence of transannular incision extended down along the RV free wall was managed by fluid loading to higher filling pressures, inotropic support, and reduction of RV after load. Dopamine 5 μg/kg/min was started and milrinone was added to help RV function and reduce PVR. Other means of reducing PVR is by using nitric oxide. Due to RVH and diastolic dysfunction milrinone is needed to maintain the cardiac output. An RA and /or an Left Atrial (LA) pressure line might be useful in optimising the preload. Ventilation is adjusted to reduce PVR prior to weaning.
The role of dopamine has been described in a research by Flanagan *et al.* [8]. Chronic renal failure hypoxaemia usually causes changes in the glomeruli. In these patients, polycythemia occurs because of hypoxaemia which in turn leads to increased blood viscosity which is a risk factor for thrombosis. Platelets are decreased in these patients making them more prone to intraoperative bleeding. Doses of dopamine 3–10 μg/kg/min have inotropic action, being adequate for patients with TOF. One should avoid doses higher than 15 μg/kg/min because the marked increase in vasoconstriction and chronotropic properties. We used dopamine from the renal protection point of view.
Adrenaline in low doses (0.01–0.03 μg) was used as an inotropic support with milrinone and dopamine as the patient was maintaining a pressure of 80/35 mmHg, which increased to 96/48 mmHg. So we did not increase the dose of adrenaline even after the injury to the coronary artery as to avoid tachycardia. Doses of 0.01–0.03 μg/kg/min provide a low-dose adrenaline effect in which the beta-adrenergic receptor agonist effects predominate. At higher doses of up to 0.1 μg/kg/min, vasoconstrictive effect of alpha-adrenergic receptors predominates.

(3) Arrhythmias and heart block are common after VSD repairs because of the close proximity to the conduction system. Epicardial pacing might be needed to accomplish weaning from CPB. In our case, these effects were not seen. Heart block is usually transient due to oedema around the VSD patch for which prophylactically steroid-like dexamethasone was given.

(4) Post CPB bleeding is due to extreme haemodilution and the effects of CPB on platelet dysfunction for which transfusion of

multiple components of blood products and use of antifibrinolytic agents such as e-aminocaproic acid was given.

Limitation of the current report

Immediate adequacy of repair is assessed by intraoperative Transesophageal Echo (TEE). We did not have the facility of intraoperative TEE. TEE is very useful for seeing residual VSDs, assessing the ventricular function and valves and seeing the gradient across RVOT. Owing to its lack we had to go via the old method of measuring the pressure by asking the surgeon to put a needle in the RV and connecting it to the transducer via a pressure monitoring line. RV : LV pressure ratio which should be less than 0.7 and blood gas measurement from the vena cava and the PA can also be used to detect residual shunts [9,10]. Our Arterial Blood Gas (ABG) reporting was within normal limits.

Conclusion

A basic understanding of the pathophysiology and knowledge of the various inotropes and their applications in various cardiac anomalies can take out the anaesthetist from a difficult situation.

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Conflicts of interest

There are no conflicts of interest.

References

1 Dabizzi RP, Teodori G, Barletta GA, Caprioli G, Baldrighi G, Baldrighi V. Associated coronary and cardiac anomalies in the tetralogy of Fallot. An angiographic study. Eur Heart J 1990; 11:692–704.

2 Bonchek LI. A method of outflow tract reconstruction in tetralogy of Fallot with anomalous anterior descending coronary artery. Ann Thorac Surg 1976; 21:451–453.

3 Hudspeth AS, Cordell AR, Johnston FR. Transatrial approach to total correction of tetralogy of Fallot. Circulation 1963; 27:796–800.

4 Hurwitz RA, Smith W, King H, Girod DA, Caldwell RL. Tetralogy of Fallot with abnormal coronary artery: 1967 to 1977. J Thorac Cardiovasc Surg 1980; 80:129–134.

5 Gilkeson RC, Ciancibello L, Zahka K. Multi-detector CT evaluation of congenital heart disease in pediatric and adult patients. Am J Roentgenol 2003; 180:973–980.

6 Rivenes SM, Lewin MB, Stayer SA, Bent ST, Schoenig HM, McKenzie ED, Fraser CD, Andropoulos DB. Cardiovascular effects of sevoflurane, isoflurane, halothane, & fentanyl – midazolam in children with congenital heart disease: An echocardiographic study of myocardial contractility & hemodynamics. Anaesthesiology 2001; 94:223–229.

7 Tugrul M, Camci E, Pembeci K, Telci L, Akpir K. Ketamine infusion versus isoflurane for the maintenance of anaesthesia in the prebypass period in children with tetralogy of Fallot. J Cardiothorac Vasc Anaesth 2000; 14:557–561.

8 Flanagan MF, Hourihan M, Keane JF. Incidence of renal dysfunction in adults with cyanotic congrnital heart disease. Am J Cardiol 1991; 68:403–406.

9 Joyce JJ, Hwang EY, Wiles HB, Kline CH, Bradley SM, Crawford FA. Reliability of intra operative transesophageal echocardiography during tetralogy of Fallot repair. Echocardiography 2000; 17:319–321.

10 Kaushal SK, Radhakrishanan S, Dagar KS, Iyer PU, Girotra S, Shrivastava S, Iyer KS. Significant intra-operative right ventricular outflow tract gradients after repair for tetralogy of Fallot: to revise or not to revise? Ann Thorac Surg 1999; 68:1705–1713.