

# Anesthetic Management for Transverse Colostomy in a Neonate with Anorectal Malformation and Uncorrected Tetralogy of Fallot

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## ABSTRACT

Tetralogy of fallot (TOF) is a cyanotic congenital heart disease (CHD) characterized by aortic overriding, right ventricular outflow tract (RVOT) obstruction, pulmonary stenosis, and ventricular septal defect (VSD). We report a case of a neonate with TOF and anorectal malformation posted for a transverse colostomy. During this procedure, our objectives were to prevent cyanotic spells, balance pulmonary vascular resistance, and systemic vascular resistance.

**Keywords:** Anesthesia, Colostomy, Tetralogy of Fallot.

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## INTRODUCTION

Tetralogy of fallot (TOF) is a cyanotic congenital heart disease (CHD) with an incidence of 3 in 10,000 births characterized by aortic overriding, right ventricular outflow tract (RVOT) obstruction, pulmonary stenosis, and ventricular septal defect (VSD).<sup>1</sup>

Among them, 30% require surgery during the first year of life for extracardiac abnormalities, which are grouped as VACTERL (V, vertebral defects; A, anal atresia; C, cardiac anomalies; T, tracheo-esophageal fistula; R, renal anomalies; and L, limb abnormalities) or CHARGE syndrome (C, coloboma; H, heart defects; A, atresia of choanae; R, retarded growth; G, genital abnormalities; and E, ear abnormalities). Hence the presence of one anomaly necessitates to rule out all other associated anomalies. The most common structural malformation associated with congenital heart disease is gastrointestinal anomalies. As the neonates and infants with CHD are associated with a twofold increase in mortality from a noncardiac surgery, it is a challenge for the anesthesiologist to handle such patients.

## CASE DESCRIPTION

A 3-day-old male weighing 3 kg was diagnosed with TOF and imperforate anus and was posted for a transverse colostomy. On examination, the baby had a pansystolic murmur, peripheral cyanosis, absent anal opening, distended abdomen, and oxygen saturation of 80% on O<sub>2</sub> with nasal prongs. Hemoglobin was 14 g/dL and platelet count was 2 lakhs/cubic millimeter. Two-dimensional echocardiography (2D echo) revealed a hypoplastic right ventricle, large perimembranous VSD with a bidirectional shunt, infundibular valvular stenosis, and a small patent ductus arteriosus (PDA) with a left to right shunt. Rest of the routine investigations were normal.

The patient was premedicated with midazolam 0.05 mg/kg and glycopyrrolate 4 µg/kg intravenously (IV). The patient was induced with sevoflurane, intubated with size 3 uncuffed endotracheal tube and maintained on oxygen, sevoflurane, and atracurium. Caudal analgesia with 1 mL of 0.5% bupivacaine + 0.5 mL of 2% lignocaine + 1.5 µg fentanyl to a total volume of 3 mL was given. Intraoperative monitoring was done with noninvasive

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blood pressure (NIBP), electrocardiogram (ECG), end tidal carbon dioxide (EtCO<sub>2</sub>), and oxygen saturation (SpO<sub>2</sub>). The child began to take good spontaneous respiration at the end of surgery and was extubated while being awake. The postoperative period was uneventful.

## DISCUSSION

The anesthetic management for a noncardiac surgery in a child with TOF necessitates the understanding of its pathophysiology, which is very vital. The severity of disease depends on the degree of right ventricular outflow tract obstruction and VSD. Chronic hypoxia might lead to polycythemia, which may in turn lead to coagulopathy, intracranial abscess, and stroke.<sup>2</sup>

Hours of nil by mouth (NBM) should not be prolonged and it is also important to maintain adequate hydration. Premedication with anxiolytics and sedatives will help us prevent agitation in the child, which will promote a right-to-left shunt and henceforth cyanosis. In case if cyanotic spells occur, phenylephrine 5–10 µg/kg IV can be given to increase systemic vascular resistance (SVR). Dexmedetomidine 0.2 µg/kg/minute can also be used as it produces analgesia and sedation with less respiratory depression, peripheral vasoconstriction, and also inhibits the release of catecholamines.<sup>3</sup> Esmolol 0.5 mg/kg bolus IV followed by 50–300 µg/kg/minute helps

reduce infundibular spasm and thereby is used for both prophylaxis and treatment of cyanotic spells.<sup>4</sup> Treatment of metabolic acidosis, volume resuscitation, and manual compression of aorta are also to keep in mind during episodes of cyanotic spells.

As TOF is associated with CHARGE 22, VACTERL Syndromes, a difficult intubation should be anticipated. Intravenous or inhalational induction can be done but with careful dose titration. In general, intravenous induction agents are usually preferred in patients with a right-to-left shunt while inhalational agents are used for left-to-right shunts, but it is not a hard-and-fast rule and this is why we need to understand its pathophysiology. In patients with TOF, owing to RVOT obstruction and a right-to-left shunt, which results in decreased pulmonary blood flow, intravenous induction agents are preferred for faster induction.

Ketamine is the most commonly used intravenous induction agent as it increases systemic vascular resistance and propofol is best avoided.<sup>5</sup> Ketamine in combination with fentanyl can also be used.<sup>4</sup> But in this case, sevoflurane was preferred as the 2D echo of the child revealed a patent PDA and there was significant pulmonary blood flow. But careful titration is required to prevent a fall in SVR.<sup>7</sup>

Atracurium is preferred in neonates since the metabolic system is not well established in them, but must be used with caution in view of decreased SVR. Vecuronium can also be used in older children.<sup>6</sup> Nitrous oxide must be avoided if cardiac function is severely compromised as it increases pulmonary vascular resistance (PVR).

Increase in PVR will promote a left-to-right shunt, while a fall in SVR will decrease blood pressure. Hence a balance between them is essential.<sup>7</sup> Factors increasing PVR such as hypoxia, hypercapnea, hyperthermia, inadequate depth of anesthesia, acidosis, and positive end expiratory pressure should be avoided.

Not all patients with TOF experience cyanotic spells. There are some mild cases referred to as pink tet in which RVOT obstruction and VSD is not severe or there may be a patent ductus arteriosus promoting pulmonary flow. Hence it is important in those cases to keep the duct patent. NSAIDs that will promote the closure of PDA should be avoided. Intravenous prostaglandin E1 0.05–0.2 µg/kg/minute will prevent the closure of ductus arteriosus.<sup>8</sup>

Pain is one more factor that can aggravate cyanotic spells postoperatively. Hence, postoperative analgesia is necessary.

In this case, caudal analgesia with opioid was given. Regional blocks appropriate for the case can be preferred.<sup>9</sup>

Complications such as air embolism, arrhythmia, cyanotic attack, congestive cardiac failure, and right ventricular dysfunction must be anticipated and should be treated accordingly.

## CONCLUSION

Successful anesthetic management for a noncardiac surgery in patients with TOF depends on the knowledge of pathophysiology of TOF, effective evaluation, appropriate preparation, and management of the case.

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