

Non Pharmacological Management of Cyanotic Spell in a Child with Tetralogy of Fallot Undergoing Noncardiac Surgery: A Case Report

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Abstract

Patient with Tetralogy of Fallot (TOF) may sometimes present for noncardiac surgery. These patients may have lots of other congenital anomalies, which may need repeated surgeries for their correction. Anaesthetic management in this sort of cases for noncardiac surgeries may pose a real challenge to the anaesthesiologists because of the high incidence of perioperative complications. Cyanotic spell is one among them. Cyanotic spells are usually managed by drugs. But there is report of use of non-pharmacological measures also. In this case report we present successful management of the intraoperative cyanotic spell with intravenous fluids and blood.

Keywords: Non pharmacological management of cyanotic spell; Tetralogy of Fallot; Noncardiac surgery; Paediatric patient

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Introduction

Tetralogy Of Fallot (TOF) is the commonest cyanotic heart disease [1]. It may sometimes be associated with other congenital anomalies [2]. Morbidity and mortality increase if TOF is associated with other non-cardiac surgical congenital anomalies [3]. Surgical correction of other anomalies sometimes needs to be done earlier than TOF correction depending on the nature of the anomalies. Gastrointestinal anomalies like caudal duplication and meningomyelocele are such which demand early correction before TOF. Case reports for correction of these congenital anomalies in paediatric patients with TOF have not been frequently reported in literature. Anaesthetic administration for these procedures is a challenge to the anaesthesiologist because of high incidence of perioperative mortality and morbidity [4]. Intraoperative cyanotic spells are usually managed by drugs [5-7]. But non pharmacological means used to treat cyanotic spells are rarely reported [8]. We are here presenting such a case report of the non-pharmacological management of cyanotic spell in a child with TOF scheduled for surgical correction of caudal duplication with septate bladder and meningomyelocele.

Case Report

An eighteen-month-old female child, weighing 5.4 Kg, presented with caudal duplication and lumbosacral

meningomyelocele, along with congenital heart disease. The child was found to have double anal opening and lumbosacral meningomyelocele. There was also weakness of left lower limb and deformity of right foot since birth. Interestingly, there was no history suggestive of cyanotic spells, seizures, repeated upper or lower respiratory tract infection or any other associated illness except a history of poor weight gain. On examination of the Cardiovascular System (CVS), a pan-systolic murmur in the left parasternal area and an ejection systolic murmur in the left second intercostal space were detected. Rest of the chest was clear on auscultation. Child was found to be irritable and had cyanosis as well as clubbing of fingers on general physical examination. She had a lumbosacral meningomyelocele and her motor power in lower limbs was found to be 3/5 in thigh and 0/5 in the foot respectively.

Her haemoglobin was 11.9 gm%, and the liver, renal functions as well as other investigations were within the normal limit. Echocardiography showed a large VSD (left to right shunt) with 50% overriding of aorta, ASD with left-to-right shunt and mild pulmonary artery obstruction. Her right ventricular dimension was 85 mL/m² and function was within acceptable limit. Radiograph of lumbosacral spine showed vertebral anomalies like hemi vertebrae, butterfly vertebrae and fusion of the L₂₋₃ vertebral bodies. MRI of spine and pelvis showed lipomeningomyelocele in spinal cord at L₄₋₅ with multiple segmental anomalies, agenesis of the sacrum and tethered cord. MRI also revealed agenesis of right kidney and non-visualization of ovaries. She was already on oral propranolol therapy (5 mg three times daily) at the time she presented for surgery.

The child was operated three months before for excision of duplicated colon, formation of anal opening, sigmoid colostomy and excision of meningomyelocele under general anaesthesia. There were 2-3 attacks of cyanosis. Nothing in detail was mentioned in the anaesthesia report. At the end of operation she was not extubated, but put on elective ventilation. In the postoperative period she had two attacks of cyanotic spells, which were managed with morphine and

propranolol. On the 2nd postoperative day she was extubated. Rest of her postoperative stay was uneventful.

This time she was scheduled for closure of the sigmoid colostomy and division of congenital bladder septum. There were no fresh symptoms or signs after the last operation and her haematocrit was 48%. The child was kept fasting for four hours and intravenous fluid was started concurrently with dextrose in 0.18% saline. Propranolol was continued till the day of operation. Infective endocarditis prophylaxis with intravenous ampicillin (250 mg) and gentamycin (7.5 mg) was given 30 min before the surgery. Routine noninvasive monitoring like Electrocardiogram (ECG), Noninvasive Blood Pressure (NIBP), peripheral oxygen saturation (SpO₂), End tidal Carbon Dioxide (EtCO₂), temperature and urine output were used. SpO₂ in the room air was 60-65%. An inhalational induction technique was used with oxygen and sevoflurane. Child was intubated with vecuronium (0.08 mg/kg). Oxygen saturation improved to 88% after induction of anaesthesia. Anaesthesia was maintained with oxygen in air, sevoflurane, and incremental doses of vecuronium and fentanyl. During intraoperative period child developed two episodes of hypotension (40/25 and 52/30 mmHg) and desaturation (73% and 66%), which were managed initially by providing 100% oxygen and lightening the plane of anaesthesia. But there was not much response. So we thought that hypovolemia may be the cause and gave bolus of crystalloids and transfused blood. With this, blood pressure returned to normal thereby improving the SpO₂ (82%). In the remaining period, SpO₂ varied between 75-85%. Total duration of anaesthesia was three and half hours and a total of 20 µg fentanyl were used. Total blood loss was 75 mL. As like previous operation, the child was not extubated and shifted to the Paediatric Intensive Care Unit (PICU) and put on a ventilator (PCV₂₀ RR₂₅ ASB₁₀ PEEP₄ FiO₂ 1.0). She was sedated with fentanyl infusion. The child was extubated on second postoperative day with no postoperative complications. Later she was discharged uneventfully.

Discussion

TOF is the commonest and represents about 10% among the congenital heart disease [1, 9, 10]. TOF consists of

Ventricular Septal Defect (VSD), infundibular pulmonary stenosis, over-riding of aorta and right ventricular hypertrophy [5]. Occasionally it may be associated with Atrial Septal Defect (ASD) [9, 11]. The mortality associated with TOF is very high and about one fourth of the untreated children die within first year of life [9]. Mortality increases to 50% by the age of five years and that to 70% by the 10th year in uncorrected TOF. Survival rate by the time they reach adulthood is only 3-5% [1].

Caudal duplication is a rare congenital anomaly, where the colons are usually fused with a common wall. Sometimes there may be common ileum, associated duplication of genitourinary system and spinal defects like spina bifida. It is rarely associated with extra lobular pulmonary sequestration and esophageal communication [12]. In our case the child had a double colon, septate bladder and vagina along with spina bifida and tethered cord. Congenital cyanotic heart disease patients undergoing noncardiac operative procedures are at the risk of further increase in the right to left shunt, which result in hypoxemia. This is especially high in patients with TOF where there is large VSD with overriding of aorta along with pulmonary stenosis. The relative resistances of the pulmonary and systemic vascular tracts are the major determinants of shunt flow. In these patients reduction in Systemic Vascular Resistance (SVR) or increase in right ventricular outflow tract obstruction [either by dynamic infundibular spasm or increase in Pulmonary Vascular Resistance (PVR)] increases the right to left shunts resulting in increased hypoxemia. This is called cyanotic spell. Cyanotic spell may be initiated by sympathetic stimulation, increase in PVR or decrease in SVR and hypotension (systolic blood pressure less than 60 mmHg) [6]. The treatment of this cyanotic spell consists of increasing the SVR by a adrenergic agonists like phenylephrine [5, 6], decreasing the dynamic right ventricular outflow obstruction by beta blockers like propranolol [6] or shorter acting esmolol [7]. In some of the refractory cases use of direct abdominal aortic compression has also been mentioned. Alpha-adrenergic agonists are preferred in cases of low SVR and blood pressure, while adrenergic blockers are preferred in cases of high BP and tachycardia for the treatment of cyanotic spells [8]. Interestingly

this can even be reverted by maintaining adequate circulating blood volume with crystalloids or colloids [1, 5, 11]. Some of these patients may have less number of cyanotic spells if the ventricular shunting is from the left to right direction, as in our case. These are known as “pink Tet”. Usually they become cyanotic by 1-2 years of age [5].

Goal of anaesthesia in TOF is to maintain SVR, minimize the PVR, to allow mild myocardial depression and to maintain lower heart rate. A noncompliant hypertrophied right ventricle may require higher preload for optimal functioning, as per the Starling’s law. Decrease in the preload and systemic vascular resistance in presence of hypovolaemia can increase the right to left shunt [13]. This is because the hypertrophied and outflow obstructed right ventricular output depends in preload and failure to do so may lead to reverse shunting. So maintaining the higher preload by infusion of IV crystalloids or colloids may be preferable. There is literary evidence that administration the fluids can be used as one of the means to tide over the crisis during the cyanotic spells [1, 5]. Since volume overload and congestive heart failure is very rare in TOF, fluid challenge can always be tried [1]. Maintenance of adequate cardiac output and blood pressure is very important since it is very difficult to improve the oxygenation of blood in these patients [1]. But literary search did not reveal any such report of use of fluid and blood transfusion to terminate the cyanotic spell in the intraoperative period.

In the preoperative period we started intravenous fluids for optimization of preload and to maintain adequate fluid balance. Sedative premedication was avoided in order to prevent adverse effect on SVR and PVR [14]. We also avoided the use of anticholinergics in premedication to maintain low heart rate during the intraoperative period. Though ketamine has been preferred as the induction agent in cyanotic heart disease patients, but there is a chance of further increase in PVR in a patient already having pulmonary arterial hypertension [15]. Since sevoflurane has been accepted as safe induction agent in CHD [5, 9], we preferred inhalational induction. It has been mentioned in the literature that in absence of congestive heart failure, inhalational induction can be safely used and can be

carried out with a variety of agents [1]. Though there is theoretical constraint against the use of inhalational agents, oxygen saturation actually improves after induction [16], as seen in our case. Sevoflurane lacks the significant myocardial depressant property; it has become agent of choice in pediatric patients with CHD. We did not see any fall in SpO₂ or cyanotic spell during inhalational induction. The SpO₂ actually improved compared to preoperative period. We used fentanyl for the analgesia as it has minimal effect on circulation. It also prevents pulmonary vasoconstriction induced by other stimuli [17]. Vecuronium was used for muscle relaxation as it has minimal effect on the circulation. We avoided nitrous oxide to minimize its effect on PVR [11].

For the episodes of desaturation that occurred during the operation, we firstly suspected it to be due to fall in the SVR. So we reduced sevoflurane concentration and ventilated with 100% oxygen. But as there was no improvement, we decided for fluid administration. So we transfused blood (to maintain haemoglobin on higher side as she was relatively anaemic for TOF) and crystalloids to. The blood pressure and SpO₂ improved with this. Ideally phenylephrine would have been the best agent to tide over this crisis. But unfortunately it was not available in our institute. So we proceeded with the fluid management. We electively ventilated the child after the surgery, as this was a major procedure. This had reduced the work of breathing and allowed us to use analgesia liberally.

In this patient we used standard noninvasive monitoring. Though the patient with complex heart disease like this warrants invasive monitoring like invasive blood pressure, central venous pressure monitoring. But we managed anaesthesia successfully for non-cardiac surgeries without these invasive monitors, and this is also reported in literature [9]. As in every CHD case we took strict precautions to prevent air embolism [11].

Though the significance of single case report is not much, but we decided to report it because of evidence of successful use of inhalational induction, noninvasive monitoring and non pharmacological management of cyanotic spell intraoperatively (which is not reported previously) in a patient

with TOF presented with non cardiac concurrent surgical anomalies.

Conclusion

Thus we conclude that with the adequate understanding of pathophysiology cyanotic spells can be managed effectively and non pharmacologically by infusion of crystalloid and blood.

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