

Persistent postoperative hypercyanotic spells in an adult with surgically untreated tetralogy of Fallot: Use of ketamine infusion

Sir,
We read with interest the article by Goyal et al.\textsuperscript{[1]} in which the authors use dexmedetomidine and ketamine infusions for total intravenous anesthesia in patients with tetralogy of Fallot (TOF) for noncardiac surgery. We report the use of ketamine infusion only for management of persistent postoperative hypercyanotic spells in a 45-year-old male with surgically untreated TOF operated for open reduction and internal fixation of the humerus.

The patient had a history of dyspnea on exertion and cyanosis for the past 10 years. An ejection systolic murmur was heard in the pulmonary area. Results of biochemical investigations reported hemoglobin of 20 g/dL (hematocrit = 61%), platelet count of 95,000/mm\textsuperscript{3} but a normal leucocyte count, renal function test, serum electrolyte and coagulation profile. Cardiomegaly was seen on chest X-ray and right ventricular hypertrophy, tall R waves (V1-V3 leads), right axis deviation was seen in the electrocardiogram (ECG). Echocardiography reported a ventricular septal defect (VSD) of 22 mm, 40% of aortic override, severe right ventricular outflow tract obstruction (valvular and infundibular), severe tricuspid regurgitation, concentric left ventricular hypertrophy/ right ventricular hypertrophy, 62% ejection fraction and right ventricular systolic pressure of 136 mmHg above right atrial pressures. Arterial blood gas analysis reported PaO\textsubscript{2} of 50 mmHg and PaCO\textsubscript{2} of 36 mmHg. Standard American Society of Anesthesiologists monitoring (noninvasive blood pressure, pulse oximetry and ECG) was attached, and right radial artery was cannulated for invasive blood pressure monitoring. Baseline oxygen saturation of hemoglobin (SpO\textsubscript{2}) was 82%. Nerve stimulator guided supraclavicular brachial plexus block was given, and local anesthetic was injected (15 ml of 0.5% bupivacaine + 15 ml of 2% lignocaine with adrenaline + 10 ml normal saline). However, complete sensory and motor block could not be accomplished, and patient had to be administered general anesthesia. Morphine (7.5 mg), ketamine (100 mg) and succinylcholine (100 mg) was used for induction and intubation. Anesthesia was maintained with O\textsubscript{2}/air/halothane and vecuronium. Intraoperative period was uneventful and the patient was extubated. Patient had saturation of 88% at the time of shifting to the intensive care unit.

One hour after surgery, SpO\textsubscript{2} decreased to 40% with heart rate of 88/min and blood pressure of 100/60 mmHg. Morphine (5 mg), propanolol (0.1 mg/kg over a period of 10 min), sodium bicarbonate (80 mEq + 80 mEq + 80 mEq after every 10 min) and phenylephrine infusion were used to manage the tet spell. After 12 h the patient again had five episodes of acute fall in SpO\textsubscript{2}, managed with the above-mentioned drugs plus ketamine; initial bolus of 0.5 mg/kg, followed by infusion of 0.1 mg/kg/h for 12 h. Patient did not have any spell after that.

Due to limited cardiac surgical centers and inadequate medical treatment, adult patients with untreated TOF are not uncommon in developing countries. Anesthetic considerations in adults with grown-up congenital heart disease are tailored on the basis of the anatomy and physiology of the cardiovascular system.\textsuperscript{[2]} Prevalence of congestive heart failure and cardiomegaly are more common in adults with untreated TOF. Longevity in natural survivors with unoperated TOF...
is attributed to either of the following:\[3\]
a. Presence of left ventricular hypertrophy leading to a delay of shunting of blood from the right-to-left ventricle.

b. Presence of extra-cardiac shunts: Patent ductus arteriosus, aorto-pulmonary collaterals (systemic to pulmonary artery shunting) which contribute blood to the pulmonary circulation.

c. A hypoplastic pulmonary artery with slow development of sub-pulmonary obstruction.

Hypercyanotic spells or “tet spells” are paroxysmal episodes of acute worsening of cyanosis and are common in children with TOF. Decreased pulmonary blood flow and increased right-to-left shunting due to an imbalance between pulmonary and systemic vascular resistance (SVR) lead to a tet spell.\[4\] Risk increases with increasing severity of resting cyanosis and hematocrit. Factors precipitating tet spells are summarized in Table 1.

Ketamine increases SVR, decreases right-to-left shunting via VSD and improves oxygenation due to increased pulmonary blood flow.\[5\] Goyal et al.\[6\] have administered it at as a bolus of 2 mg/kg over 10 min, followed by an infusion at 2 mg/kg/h in combination with dexmedetomidine as a bolus of 1 µg/kg followed by infusion at 0.7-1 µg/kg/h (1-1.5 µg/kg/h for pediatric patient). The rationale behind the combination is the additive analgesic action, ability of dexmedetomidine to negate emergence delirium secondary to use of ketamine and the balance of sympatho-inhibitory effect of dexmedetomidine with cardio-stimulatory effect of ketamine with an aim to provide surgical anesthesia. We in our case have successfully managed the tet spell with low dose ketamine only and do not report any hemodynamic perturbation, respiratory compromise or emergence delirium with its use.

**Table 1: Factors causing increase right-to-left shunting**

| Pathophysiology            | Etiology                                                                 |
|----------------------------|--------------------------------------------------------------------------|
| Increase in PVR            | Sympathetic stimulation due to pain, light anesthesia, tachycardia, acidemia, hypoxemia, hypercarbia, hypothermia and increase in intrathoracic pressure |
| Dynamic outflow obstruction| Tachycardia, hypovolemia and increased myocardial contractility can cause infundibular spasm and dynamic outflow obstruction |
| Decrease in SVR            | Volatile anesthetic agents, histamine releasing drugs, ganglionic blockade and alpha adrenergic blockade decrease SVR   |

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Conflict of interest
There are no conflicts of interest.

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