Case Report

Case series: Dexmedetomidine and ketamine for anesthesia in patients with uncorrected congenital cyanotic heart disease presenting for non-cardiac surgery

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Abstract

The number of patients with uncorrected congenital cyanotic heart disease is less but at times some may present for non-cardiac surgery with a high anesthetic risk. Some of these may even be adults with compromised cardiopulmonary physiology posing greater challenges to the anesthesiologist. The authors have used a combination of dexmedetomidine and ketamine for anesthesia for non cardiac surgery in five patients with cyanotic heart disease and right to left shunt (3-Eisenmenger's syndrome, 2-Tetralogy of Fallot). The sympathoinhibitory effects of dexmedetomidine were balanced with the cardiostimulatory effects of ketamine, thereby maintaining good cardiovascular stability. The analgesia was good and there was no postoperative agitation.This drug combination was effective and safe for patients with cyanotic heart disease for non cardiac surgeries.

Key words: Congenital cyanotic heart disease, dexmedetomidine, Eisenmenger's syndrome, ketamine

Introduction

The advancement in pediatric cardiac surgery has resulted in an increasing population of young adults with grown up congenital heart disease (GUCHD).[1] Most of these adults would have undergone a palliative, reparative, or a corrective surgery in early childhood or sometime later. Those with various congenital cyanotic heart diseases (CCHD) pose a greater risk to life and are usually corrected early. However, in many developing nations there still exist a number of patients with uncorrected CCHD, both as children or young adults. Some of the acyanotic heart diseases like atrial septal defect (ASD) and patent ductus arteriosus (PDA) may be relatively asymptomatic for several years because of the balance between the systemic and pulmonary circulation and therefore the diagnosis is missed. The pulmonary blood flow is more in the initial phase along with a high pulmonary artery pressure. Gradually, the pulmonary vascular resistance (PVR) starts rising and over the years, often reaches a point where it is irreversible and then they also pose as cyanotic CHD. Eisenmenger's syndrome is thereby a common cause of GUCHD in adults with a reversed or a bidirectional shunt flow. In children however, tetralogy of Fallot (TOF) remains the commonest cause of CCHD;[2] though there are reported series of 40 years and older, many of which without even a palliative surgery earlier.[3]

These children or adults often present for noncardiac surgery, mainly emergencies and the anesthetic risk is high even for minor surgeries.[4,5] We present a series of five different cases of cyanotic heart disease with right to left (R-L) shunt where a combination of dexmedetomidine and ketamine (ketodex)[6] was used and the patients recovered well.

Case Report

Written informed consent was taken from all the five cases being presented. There were three cases of Eisenmenger's syndrome and two of TOF. Four were GUCHD while one was a child (TOF), and they were all uncorrected cyanotic heart disease with (R-L) shunt. The complete perioperative data of all the patients is shown in Table 1. Case 1-3 were managed with total intravenous (IV) anesthesia with a

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Table 1: Patient characteristics and perioperative data

Case no.	Age (year)/ gender	Diagnosis	Echo findings	Preoperative SpO_2 on room air (%)	Diagnosis/ surgery	Anesthesia technique	Dose of ketodex	Intraoperative hemodynamics			
								HR (per min)	MAP (mmHg)	SpO_2 (%)	ABG
1	22/F	PDA	8 mm PDA Bidirectional flow PAH (86 mmHg±RAP) Dilated PA 38 mm	90-92	1st trimester pregnancy/ medical termination of pregnancy	TIVA	D 1 μg/kg K 2 mg/kg over 10 min	68-70	86-90	90-94	pH 7.44 PO_2 92 mmHg PCO_2 30 mmHg HCO_3 19.6 mmol/l
2	27/M	PDA	13 mm PDA Bidirectional flow PAH (40 mmHg±RAP)	90	Right inguinal hernia/ hernioplasty	TIVA and field block (30 ml, 0.25% bupivacaine)	D 1.0 μg/kg/h K 1 mg/kg/h	78-92	74-82	88-94	pH 7.48 PO_2 56 mmHg PCO_2 34 mmHg HCO_3 24.3 mmol/l
3	17/F	ASD	32 mm large ASD 2 mm VSD 6 mm PDA (R-L) shunt PAH (88 mmHg±RAP) Dilated RA, RV	90	Fracture medial malleolus right/ open reduction and internal fixation	TIVA	D 1 μg/kg K 2 mg/kg over 10 min Maintenance D 1-1.5 μg/kg/h K 2 mg/kg/h	84-92	75-82	90-92	pH 7.32 PO_2 48 mmHg PCO_2 42 mmHg HCO_3 19.8 mmol/l
4	20/F	TOF	Large VSD Bidirectional shunt Overriding of aorta Moderate pulmonic stenosis RV hypertrophy	86	Cerebral abscess right frontal lobe with raised ICP midline shift 12 mm/ emergency craniotomy and drainage	TIVA±IPPV	D 1 μg/kg K 2 mg/kg over 10 min Maintenance D 0.7-1 μg/kg/h K 2 mg/kg/h	98-104	100-110	80-88	pH 7.465 PO_2 37.3 mmHg $PaCO_2$ 33.4 mmHg HCO_3 23.5 mmol/l
5	2/M	TOF	Large subaortic VSD Bidirectional shunt Severe pulmonic stenosis Overriding of aorta, small PA	88	Malignant right parietal lobe infarct with raised ICP/ emergency hemicraniectomy	TIVA±IPPV	D 1 μg/kg K 2 mg/kg over 10 min followed by D 1-1.5 μg/kg/h K 2-2.5 mg/kg/h	108-116	58-62	88-90	pH 7.36 PO_2 47 mmHg $PaCO_2$ 34 mmHg HCO_3 22.7 mmol/l

PDA=Patent ductus arteriosus, PAH=Pulmonary artery hypertension, PA=Pulmonary artery, ASD=Atrial septal defect, VSD=Ventricular septal defect, TOF=Tetralogy of Fallot, RA=Right atrium, RV=Right ventricular, RAP=Right atrial pressure, ICP=Intracranial pressure, TIVA=Total intravenous anesthesia, D=Dexmedetomidine, K=Ketamine, IPPV=Intermittent positive pressure ventilation, ABG=Arterial blood gas, HR=Heart rate, MAP=Mean arterial pressure

combination of dexmedetomidine 1 ∝g/kg and ketamine 2 mg/kg IV infused over 10 min. They were short duration surgeries of <90 min with spontaneous respiration, supplemented with oxygen via face mask. Case 4 and 5 required endotracheal intubation (details mentioned later). IV paracetamol was added for analgesia and the hydration was maintained with a balanced salt solution 10-15 ml/kg/hr. Infective endocarditis prophylaxis was administered to all the patients. Standard monitoring was used in all cases and all the patients were observed in the intensive care unit (ICU) for at least 24 h.

Case 4 was a 20-year-old female patient, a diagnosed case of TOF who presented with a frontal lobe abscess with a 12 mm midline shift and features of septic shock. She was posted for emergency craniotomy and drainage of the abscess. There was a history of occasional episodes of cyanotic spells since the last 1 year. However, she had not undergone any corrective or palliative surgery and was not on any medication. Preoperative evaluation revealed SpO_2 86% on room air, arterial blood gas (ABG) analysis showed PaO_2 47.8 mmHg, pH 7.465, $PaCO_2$ 50.3 mmHg, and HCO_3 30 mmol/l. She was being managed with vasopressors (noradrenaline: 0.05-0.07 μg/kg/min) to maintain the mean arterial pressure (MAP) above 65 mmHg. After preoxygenation, the patient was induced with dexmedetomidine 1 μg/kg and ketamine 2 mg/kg over 10 min. Anesthesia was maintained on controlled ventilation with vecuronium and an infusion of dexmedetomidine (0.7-1.0 μg/kg/h) and ketamine (2 mg/kg/h). Paracetamol 1 g IV was added for analgesia though no inhalational anesthetic was used for maintenance. Cardiac output and systemic vascular resistance (SVR) were monitored by Flotrac™. SVR was maintained >1,200 dyne.s/cm^5 by an infusion of noradrenaline (0.05-0.1 μg/kg/min). There was a large parietal abscess and about 100 ml of pus was drained. In view of the raised intracranial pressure (ICP) and septic shock, she was electively ventilated in the ICU.

Case 5 was a 2-year-old boy who was diagnosed as TOF with a large subaortic VSD, bidirectional shunt, severe pulmonic stenosis, and overriding of aorta with a small sized pulmonary artery on echocardiography.[7] The child had developed right hemiparesis at 1 year of age and his heart disease was detected during the work up. He recovered well over the next few months, but could not undergo any palliative surgery for TOF. He developed a malignant right parietal lobe infarct with features of raised ICP at 2 years of age. A decompressive right hemicraniectomy was done under general anesthesia. The child developed cerebrospinal fluid (CSF) leak from the surgical site and an external lumbar drain was placed 3 days later for controlled CSF drainage. During both the above procedures, dexmedetomidine (1 μg/kg and 1-1.5 μg/kg/h) and ketamine (2 mg/kg and 2-2.5 mg/kg/h), respectively were used for induction and maintenance of anesthesia. The former surgery was done with controlled ventilation using muscle relaxation and the latter procedure was carried out with the two drugs along with oxygen via face mask with spontaneous respiration. The child maintained hemodynamic stability perioperatively and recovered uneventfully.

Discussion

In patients with uncorrected CHD, some of whom may be adults, the anesthetic management is focused on preventing a further increase in the R-L shunt by maintaining SVR, controlling PVR, decreasing oxygen consumption, and prevention of arrhythmia and hypovolemia. Avoiding an increase in the R-L shunt prevents a decrease in the flow of blood to the lungs, thus preventing a further fall in the oxygenation of an already compromised circulation.

Transesophageal echocardiography is known to be very useful in real time assessment of the preload, cardiac contractility, and intracardiac shunting. We did not have it in our institution, but it should be used wherever possible even for noncardiac surgeries. Postoperative monitoring in a high dependency unit is recommended even after short procedures.[8]

Ketamine is preferred, though all commonly used IV induction agents can be used if given judiciously. It is important that the rate and dose used is adjusted so as to cause a minimal fall in the SVR in order to prevent an increase in the R-L shunt.[9] Theoretically, IV agents score over inhalation agents as the latter may prolong induction time in patients with R-L shunt.[10] Peripheral nerve blocks may be added where appropriate, but care should be taken to avoid systemic toxicity of local anesthetics.

Dexmedetomidine is a highly selective alpha-2 agonist that provides anxiolysis and cooperative sedation without respiratory depression. It decreases central nervous system sympathetic outflow in a dose-dependent manner and has analgesic effects. There is evidence that it alleviates postoperative delirium also.[11] It reduces the hyperdynamic responses (increased MAP and heart rate (HR)) mediated by the sympathetic nervous system and attenuates the cardiovascular and neuroendocrine response to surgery. The dose-dependent hypotension and bradycardia may limit the sole use for anesthesia or procedural sedation.[12]

Ketamine is a fast-acting general anesthetic which produces profound analgesia, maintains normal pharyngeal-laryngeal reflexes, and causes cardiovascular stimulation. The downside to its use is the high incidence of emergence agitation.

The sympatho-inhibitory effects of dexmedetomidine are balanced with the cardio-stimulatory effects of ketamine, thereby maintaining a stable hemodynamic profile within normal physiological range, in cases with R-L shunt. There is no significant rise in PVR or fall in SVR with this drug combination. Both the drugs offer analgesia via different modes and their effects appear additive. They are also known to preserve dose dependent airway reflexes and spontaneous breathing, thus useful in short duration surgeries where controlled ventilation can be avoided.[13] The emergence delirium likely to occur with ketamine is negated with the sedation of dexmedetomidine. Mester *et al*., used the same drug combination for cardiac catheterization in 16 children with CHD with good results.[14] There are no other reported cases of the use of these drugs in the literature for similar patients.

The authors have used this combination of ketamine and dexmedetomidine in five patients with R-L shunt who presented for emergency noncardiac surgery. The HR and blood pressure were maintained within 10% of the baseline. There was no episode of intraoperative hemodynamic instability, desaturation, arrhythmias, or heart failure. There was no requirement of any additional vasopressor to maintain SVR. The analgesia was also adequate for the surgery. Awakening from anesthesia was smooth and none of the patients had any postoperative agitation.

There are other known techniques of administration of safe anesthesia in such cases like various regional nerve blocks and general anesthesia with newer inhalational agents, which are safe and effective. However, each technique has its inherent pros and cons and it much depends on the attending anesthesiologist on what the person is most confident with.

In this series the combination of dexmedetomidine and ketamine provided excellent intraoperative hemodynamic stability. It was found to be effective and safe for anesthesia for patients with cyanotic heart disease, though larger controlled trials are recommended before any conclusion is drawn.

References

1. Daliento L, Mazzotti E, Mongillo E, Rotunda M, Dalla Volta S. Life expectancy and quality of life in adult patients with congenital heart disease. Ital Heart J 2002;3:339-47.
2. Daliento L, Somerville J, Presbitero P, Menti L, Brach-Prever S, Rizzoli G, *et al*. Eisenmenger syndrome. Factors relating to deterioration and death. Eur Heart J 1998;19:1845-55.
3. Attenhofer Jost CH, Connolly HM, Burkhart HM, Scott CG, Dearani JA, Carroll JA, *et al*. Tetralogy of Fallot repair in patients 40 years or older. Mayo Clin Proc 2010;85:1090-4.
4. Ammash NM, Connolly HM, Abel MD, Warnes CA. Noncardiac surgery in Eisenmenger syndrome. J Am Coll Cardiol 1999;33:222-7.
5. Carmosino MJ, Friesen RH, Doran A, Ivy DD. Perioperative complications in children with pulmonary hypertension undergoing noncardiac surgery or cardiac catheterization. Anesth Analg 2007;104:521-7.
6. Goyal R, Singh S, Shukla RN, Patra AK, Bhargava DV. Ketodex, a combination of dexmedetomidine and ketamine for upper gastrointestinal endoscopy in children: A preliminary report. J Anesth 2012;27:461-3.
7. Goyal R. Lumbar drainage of cerebrospinal fluid in a child with Tetralogy of Fallot and cerebral infarct. MJAFI 2012;68:63-4.
8. Khan ZH, Zeinaloo AA, Khan RH, Rasouli MR. Cardiac decompensation in a patient with Eisenmenger syndrome undergoing T5-T7 levels laminectomy in the sitting position. Turk Neurosurg 2009;19:86-90.
9. Lovell AT. Anaesthetic implications of grown-up congenital heart disease. Br J Anaesth 2004;93:129-39.
10. Huntington JH, Malviya S, Voepel-Lewis T, Lloyd TR, Massey KD. The effect of a right-to-left intracardiac shunt on the rate of rise of arterial and end-tidal halothane in children. Anesth Analg 1999;88:759-62.
11. Levanen J, Makela ML, Scheinin H. Dexmedetomidine premedication attenuates ketamine-induced cardiostimulatory effects and postanesthetic delirium. Anesthesiology. 1995;82:1117-25.
12. Mason KP, Lerman J. Review article: Dexmedetomidine in children: Current knowledge and future applications. Anesth Analg 2011;113:1129-42.
13. Tobias JD. Dexmedetomidine and ketamine: An effective alternative for procedural sedation? Pediatr Crit Care Med 2012;13:423-7.
14. Mester R, Easley RB, Brady KM, Chilson K, Tobias JD. Monitored anesthesia care with a combination of ketamine and dexmedetomidine during cardiac catheterization. Am J Ther 2008;15:24-30.

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