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Case Report

Non-operating Room Anesthesia: Anesthetic Management of Pediatric Patients with William Syndrome for Cardiac Magnetic Resonance Imaging

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Abstract

William syndrome is a chromosomal anomaly characterized by developmental abnormalities, cardiac involvement, and craniofacial dysmorphic features. These set of pediatric patients present for various non-operating room anesthesia requiring imaging or procedure. This poses a unique challenge to anesthesiologists. Here, we report a case of anesthetic management of a 4-year-old child with William syndrome requiring cardiac magnetic resonance imaging. A thorough systemic review and anticipation of difficult airway ensured an uneventful anesthesia course for the child.

Key words: Magnetic resonance imaging, Non-operating room anesthesia, Pediatric, William syndrome

INTRODUCTION

Williams-Beuren syndrome is a genetic autosomal dominant disorder associated with *de novo* deletion in the long arm of chromosome 7 (7q11.23), occurring 1:10.000 live births. This syndrome is characterized by developmental abnormalities, cardiac involvement, and craniofacial dysmorphic features. The William syndrome phenotype is characterized by dysmorphic facial features, intellectual disability, elastin arteriopathy, short stature, connective tissue abnormalities, infantile hypercalcemia, and a unique personality and cognitive profile. These set of pediatric patients present for various non-operating room anesthesia (NORA) requiring imaging or procedure. This poses challenge to anesthesiologist in three aspects, complex nature of the disease, pediatric age group, and unfavorable anesthesia environment compared to operating room. Structural cardiovascular abnormalities are present in majority of these children and may be responsible for sudden cardiac arrest in this subset of patients. Here, the authors report a case of anesthetic management of William syndrome requiring cardiac magnetic resonance imaging (MRI).

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CASE REPORT

A 4-year-old male child was incidentally detected to have a cardiac murmur during a routine school health check-up as informed by mother. The child was apparently asymptomatic giving no history of recurrent chest infections or failure to thrive. On detailed evaluation, the child was found to have elfin facies, small chin, prominent lip, depressed nasal bridge, and global developmental delay. On general physical examination, the child was poorly nourished with weight of 11 kg. Vitals were stable. Subsequent findings revealed a suprasternal thrill and a Grade III/IV systolic murmur which radiated to the neck and entire chest. His review of systems was otherwise negative. Laboratory investigations were unremarkable. Chest X-ray showed cardiomegaly with normal lung fields. Electrocardiogram showed normal sinus rhythm with no ST-T changes or QT prolongation. Two-dimensional echocardiography revealed supravalvular aortic stenosis (SVAS) with peak pressure gradient of 105 and mean pressure gradient of 54, mild right pulmonary artery stenosis, biventricular hypertrophy, good biventricular function, and dilated left main coronary artery measuring 4 mm.

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Anesthetic Management

The procedure and the risks involved were explained to the parents and written informed consent was obtained. The child was premedicated with 1 mg of oral midazolam 30 min before the procedure. The child was shifted to the MRI suite accompanied by the mother.

Difficult airway cart was kept ready anticipating a difficult intubation. Inhalational induction of anesthesia was conducted with sevoflurane in graded manner after which a wide bore intravenous (IV) access was established. Midazolam 0.5 mg and fentanyl 20 mcg was administered, and the child was paralyzed with atracurium 5 mg. The child was ventilated with 100% oxygen for 3 min following which he was intubated with size 5 cuffless endotracheal tube (ET) and was mechanically ventilated with pressure controlled mode to deliver adequate tidal volume. The child was maintained on isoflurane air and oxygen mixture. The child was constantly monitored for hemodynamic fluctuations and arrhythmias.

The procedure was uneventful. At the end of the imaging, the child was reversed and extubated after administration of calculated dose of neostigmine and glycopyrrolate and after full recovery from muscle relaxant and regaining full consciousness.

DISCUSSION

Williams syndrome was first described by Williams, Barratt–Boyes, and Lowe in 1961. Williams syndrome is caused by hemizygous microdeletion on chromosome 7 which code for the elastin gene (ELN); the estimated prevalence varies from 1:20,000 to 1:50,000 live births. It is characterized by cardiovascular disease, distinctive facies and personality, mild intellectual disability, and connective tissue growth and endocrine abnormalities.[1] The ELN gene encodes tropoelastin, which is involved in elastic vascular wall fiber assembly. Reduced elastin formation results in widespread arteriopathy. In addition to SVAS, patients with WS can develop supravalvar pulmonic stenosis, obstructive coronary disease, and diffuse aortic stenosis.[2]

Even though patients with WS have a known risk of adverse events with sedative and anesthetic drugs, they often need anesthesia for various procedures throughout their lives. Many patients with WS have some central nervous system involvement and have an average IQ of 41-80, developmental delay, and significant procedural anxiety, which can sometimes make even painless procedures (e.g. transthoracic echocardiograms, computed tomography scans, and dental procedures) unattainable without sedation. In addition, the workup for surgical management of severe cardiac disease often requires more invasive imaging techniques such as cardiac MRI or cardiac catheterization, which usually require prolonged sedation or general anesthesia for pediatric patients. Therefore, thorough pre-operative workup and risk assessment are essential for proper management to reduce risk.

The MRI suite is a challenging environment for the anesthesiologists and carries inherent risks. Several factors account for this, including the remote location, the unique features of the MRI scanner, and patient-related factors.[3]

The challenges faced by anesthesiologist in a NORA are added up by the extremes of age, especially pediatric population and the complicating nature of the disease for which they are undergoing the imaging. In WS, sudden death has been described as a very common complication associated with anesthesia, surgery, and procedures in this population. Anatomical abnormalities associated with the heart pre-dispose these individuals to sudden death. In addition, high prestenotic pressure can be transmitted to the coronary arteries and may result in severe dysplasia, in the form of intimal fibrosis and muscular hypertrophy, leading to narrowing of these arteries.

In addition to a sudden and rapid downhill course, lack of response to resuscitation is another significant feature seen in these patients.[4]

Anesthesia management of a child with William syndrome may be challenging and at times catastrophic in view of nature of the disease as well due to the non-operating room environment. A thorough pre-anesthetic evaluation is mandatory with optimization of systems for administering anesthesia. The MRI suit should be assessed completely for the MRI compatible anesthesia workstation, working suction apparatus, and all the equipment and medications required for securing the airway and for emergency situation.

Parents or the caretakers should be well sensitized about the nature of the disease and the adverse events associated with the administration of anesthesia.

Monitoring in these patients should include standard ASA monitors and consideration of invasive arterial blood pressure monitoring given that many of the arrests are preceded by hypotension and/or bradycardia. Intraoperative esophageal echocardiography has been suggested, especially during the management of complex surgical procedures.[5]

Inhalation induction is preferred to IV induction to avoid the stress response associated with pain on securing the

IV access. A graded sevoflurane induction was done in this case keeping a watch on blood pressure, so as not to let a significant fall in systemic vascular resistance, which is deleterious in a case of aortic stenosis. The use of inhalational anesthetic agents (sevoflurane or desflurane) as the sole agent is debatable as they may result in peripheral vasodilatation with a decrease of pre-load and after-load, thereby reducing blood pressure and myocardial perfusion. Sevoflurane offers the advantage of less vasodilatation and a decrease in heart rate which may preserve the myocardial oxygen supply-demand ratio better than desflurane which may result in tachycardia and vasodilatation.[6]

Once deep plane of anesthesia is established with sevoflurane, IV access is established and 3-4 mcg/kg of fentanyl is administered in titrated dose to the response. Fentanyl is cost effective and brings many other advantages. As a member of the opioid family, Fentanyl is a μ-opioid receptor agonist that is characterized by high potency, rapid onset, and short duration of action and produces no histamine release, thus avoiding the negative cardiovascular effects caused by such a response.[7] Furthermore, combining fentanyl with anesthetic agents is known to reduce the amount of anesthetic necessary to induce unconsciousness.[8] Although ketamine may provide hemodynamic stability through the release of endogenous catecholamine, it may induce tachycardia thereby altering the balance between myocardial oxygen supply and demand. In the above-mentioned case muscle relaxation with atracurium, a non-depolarizing muscle relaxant (NMDA) was chosen in view of its advantage of Hoffman elimination and minimal effect on hemodynamic. In this population, there is generally some degree of associated muscular-skeletal involvement including joint laxity that may lead to contractures and muscular weakness. Lipid storage in muscles and increased variability in fiber size has also been reported.[9] Given the choice, neuromuscular monitoring is suggested but limited by the availability of MRI compatibility of the equipment. The use of succinylcholine and other NMDAs has not been fully studied limited by number of literature in similar population.

ET intubation in this case was carried out with appropriate size ET tube after full muscle relaxation and mechanical ventilation initiated with pressure-controlled ventilation to deliver adequate amount of tidal volume. Brief periods of apnea were provided for particular MRI sequences. Hence, during the apneic duration, careful monitoring is necessary; desaturation, hemodynamic fluctuations, and arrhythmias should be anticipated, promptly identified and treated, if required.

REFERENCES

1. Kandasamy S, Saxena D, Kishore Y, Phadke SR. Williams syndrome: A case series. Indian Pediatr 2014;51:411-2.
2. Matisoff AJ, Olivieri L, Schwartz JM, Deutsch N. Risk assessment and anesthetic management of patients with Williams syndrome: A comprehensive review. Paediatr Anaesth 2015;25:1207-15.
3. Serafini G, Ongaro L, Mori A, Rossi C, Cavalloro F, Tagliaferri C, *et al.* Anesthesia for MRI in the paediatric patient. Minerva Anestesiol 2005;71:361-6.
4. Black JA, Carter RE. Association between aortic stenosis and facies of severe infantile hypercalcaemia. Lancet 1963;2:745-9.
5. Andrzejowski J, Mundy J. Anaesthesia for MRI angiography in a patient with Williams syndrome. Anaesthesia 2000;55:97-8.
6. Lashkari A, Smith AK, Graham JM Jr. Williams-Beuren syndrome: An update and review for the primary physician. Clin Pediatr (Phila) 1999;38:189-208.
7. Bowdle TA. Adverse effects of opioid agonists and agonist-antagonists in anaesthesia. Drug Saf 1998;19:173-89.
8. Kodaka M, Okamoto Y, Handa F, Kawasaki J, Miyao H. Relation between fentanyl dose and predicted EC50 of propofol for laryngeal mask insertion. Br J Anaesth 2004;92:238-41.
9. Baldinelli F, DeCarli A, Accinelli G. Sedation for magnetic resonance imaging in a child affected with Williams syndrome. Minerva Anestesiol 2003;69:169-73, 173-75.

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