Clinical Reports

Anaesthetic induction in a child with end-stage cardiomyopathy

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The purpose of this clinical report is to discuss the anaesthetic management of a patient with end-stage cardiomyopathy. An eight-year-old boy sustained electro-mechanical dissociation following a "standard anaesthetic induction" but was subsequently successfully anaesthetized using a combination of etomidate, midazolam, lidocaine, vecuronium and glycopyrrolate. Anaesthetic techniques and their relationship to the specific category of cardiomyopathy (dilated, hypertrophic or restrictive) are discussed. The potential detrimental effects of anaesthetic sympatholysis, myocardial depression and negative chronotropy on cardiac output are explained.

L'objectif de cette observation est de discuter la gestion anesthésique d'un malade porteur d'un cardiomyopathie en phase terminale. Une dissociation électro-mécanique survient chez un enfant de huit ans au cours d'une induction anesthésique standard; par la suite, il est anesthésié avec succès avec une association d'étomidate, midazolam, lidocaïne, vécuronium et glycopyrrolate. Les techniques anesthésiques et leur rapport avec la catégorie de cardiomyopathie spécifique (congestive, hypertrophique et restrictive) sont discutés. Les effets potentiellement nocifs de la lyse sympathique causée par l'anesthésie, la dépression myocardique et l'effet chronotrope négatif sur le débit cardiaque sont expliqués.

The diagnosis of cardiomyopathy in childhood is being made with increasing frequency¹ and, because of advances in medical care, many of these patients are surviving

to an older age. Increasingly, children with end-stage cardiomyopathies are coming to the operating room for brief surgical procedures such as percutaneous endoscopic gastrostomy, central line placement, dental rehabilitation, diagnostic endoscopy or biopsy. The provision of anaesthesia for these critically ill patients can be difficult. For many, regional or local anaesthesia are not appropriate due to technical, age, or behavioural factors. Similarly, general anaesthetic techniques, such as high-dose opioid techniques, frequently administered to patients with haemodynamic instability, can make postoperative ventilation obligatory which may become prolonged. In this group of patients, postoperative ventilation may affect long-term outcome by increasing the risk of iatrogenic complications and also increase the economic burden on health care by necessitating intensive care management during a portion of the waiting period.² The provision of a safe anaesthetic to such critically ill patients that will enable tracheal extubation at the conclusion of the procedure is difficult. In this case report we discuss the anaesthetic induction of such a patient and the three functional categories of cardiomyopathy that the anaesthetist may encounter in paediatric patients.

Case report

An eight-year-old boy with a diagnosis of cardiomyopathy, skeletal myopathy and hepatic fibrosis secondary to juvenile polysaccharidosis presented for percutaneous placement of a gastrostomy tube for nutritional support.

He was evaluated initially at six years of age when he developed congestive heart failure. At that time echocardiographic evaluation revealed a dilated left ventricle with increased ventricular mass and global hypokinesis. The shortening fraction was 15% (normal >34%). Electrocardiogram (ECG) revealed sinus rhythm of 100 beats per minute, first degree atrio-ventricular block, left bundle branch block and ST-T segment abnormalities in the lateral precordial leads. Cardiac catheterization demonstrated a left ventricular end diastolic pressure of 24 mmHg, pulmonary capillary wedge pressure of

Key words

ANAESTHESIA: paediatric;

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24 mmHg and a mean pulmonary artery pressure of 30 mmHg.

Initial treatment consisted of digoxin, furosemide, and captopril, but the patient remained severely restricted in terms of exercise tolerance. At eight years of age he experienced an episode of ventricular tachycardia. Electrophysiological studies demonstrated dual AV-nodal pathways and easily inducible, non-sustained ventricular tachycardia. Haemodynamically, pulmonary artery pressure was 45 mmHg, right ventricular end diastolic pressure was 12 mmHg, and wedge pressure was 29 mmHg. The cardiac index was $3.0 \text{ L} \cdot \text{m}^{-2} \cdot \text{min}^{-1}$. Amiodarone was added to the therapeutic regimen, with improvement.

At this time the patient was scheduled for percutaneous endoscopic placement of a gastrostomy tube (PEG). General anaesthesia was chosen because of technical and behavioural considerations. In order to optimize ventricular function for the procedure the patient was to receive a ten-day course of dobutamine.

Initial anaesthetic

The patient was transferred to the operating room receiving an infusion of dobutamine ($7.5 \mu\text{g} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$). The initial heart rate was 115 bpm and blood pressure 95/60 mmHg. Induction of anaesthesia was commenced with slow titration of midazolam ($0.1 \text{ mg} \cdot \text{kg}^{-1}$) ketamine ($1.5 \text{ mg} \cdot \text{kg}^{-1}$), vecuronium ($0.15 \text{ mg} \cdot \text{kg}^{-1}$) and alfentanil ($30 \mu\text{g} \cdot \text{kg}^{-1}$) *iv*. The patient became rapidly hypotensive. Systolic blood pressure decreased to 45 mmHg and was followed by loss of heart tones and pulses despite easy ventilation. Electro-mechanical dissociation with an ECG rate of 40 was diagnosed and cardiopulmonary resuscitation was instituted. Following the administration of volume, phenylephrine, atropine, epinephrine, and dopamine, perfusion was re-established. The procedure was deferred and the patient transferred to the Intensive Care Unit. The trachea was extubated later that evening. Neurological status was unchanged from the preanaesthetic condition. The patient continued to receive dobutamine by infusion and was returned to the operating room four weeks later for the same procedure.

Subsequent anaesthetic

The patient was brought to the operating room receiving an infusion of dobutamine ($2 \mu\text{g} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$). This was increased to $4 \mu\text{g} \cdot \text{kg}^{-1} \cdot \text{min}^{-1}$ approximately half an hour before the induction sequence. This anaesthetic was induced with etomidate ($0.3 \text{ mg} \cdot \text{kg}^{-1}$) after midazolam ($0.15 \text{ mg} \cdot \text{kg}^{-1}$), lidocaine ($1 \text{ mg} \cdot \text{kg}^{-1}$), vecuronium ($0.01 \text{ mg} \cdot \text{kg}^{-1}$) and glycopyrrolate (0.1 mg) *iv*. Neuromuscular blockade was established with vecuronium ($0.2 \text{ mg} \cdot \text{kg}^{-1}$) *iv*. Haemodynamically, there was no change in heart rate or blood pressure throughout the induction

sequence. Anaesthesia was maintained with fentanyl *iv* administered incrementally to a total dose of $8 \mu\text{g} \cdot \text{kg}^{-1}$ and with 0.25% isoflurane. Following completion of the PEG, the isoflurane was discontinued, neuromuscular blockade was reversed with neostigmine and glycopyrrolate and the trachea was extubated without adverse sequelae.

Discussion

Anaesthesia for patients with cardiomyopathy presents considerable challenges to the clinician. Cardiomyopathies are classified as either primary or secondary. *Primary cardiomyopathies* result from diseases in which there is an inability of the cell either to create or to use energy effectively or from diseases in which substances are not degraded normally by lysosomal enzyme systems. These substances accumulate and lead to organ dysfunction. *Secondary cardiomyopathies* may be associated with a multiplicity of disease states such as infection, endocrine abnormalities, nutritional deficiency states, immunologic disorders, neuromuscular diseases and poisons.

Functionally there are three types of cardiomyopathies: dilated cardiomyopathies (DCM), hypertrophic cardiomyopathies (HCM) and restrictive cardiomyopathies (RCM).³ Although all three are dependent upon preload and maintenance of sinus rhythm for cardiac output, differences in pathophysiology dictate important anaesthetic management considerations.

Dilated cardiomyopathies are associated with depressed ventricular *systolic* function⁴ and large cavity dimension. Increased ventricular volume and pressure will preserve stroke volume, to a limit by the Starling mechanism. The high left ventricular end diastolic pressure impairs pulmonary venous return, increasing pulmonary venous pressure producing interstitial pulmonary oedema.

Hypertrophic cardiomyopathies are characterized by *diastolic* dysfunction; systolic function is preserved. Ventricular chamber size is usually normal but may, over time, be decreased. Muscle mass may be thickened globally (concentric hypertrophy) or segmentally (asymmetric hypertrophy).^{1,5} Dynamic obstruction to left ventricular outflow may occur during systole if asymmetric septal hypertrophy is present.

High left ventricular pressures may limit pulmonary venous drainage and contribute to pulmonary oedema. Myocardial ischaemia develops because increased muscle mass and increased ventricular diastolic pressures impairs subendocardial blood flow. Other reported abnormalities include: abnormal intramural coronary arteries,¹ dysrhythmias, syncope and sudden death.⁶

The restrictive cardiomyopathies, seen associated with some storage diseases such as Gaucher's disease,⁷ are rare

in children and differ from the dilated and hypertrophic cardiomyopathies in that they have a small chamber volume which limits diastolic filling. Systolic and diastolic dysfunction may occur concomitantly. The ventricle is stiff and poorly functional and cardiac output is highly rate and preload dependent.

The DCM, seen in this patient, is progressive in nature, and myocardial function becomes increasingly impaired. Adequate heart rate and preload is needed to maintain cardiac output. In patients with systolic dysfunction, that is not ischaemic in origin, drugs that are vagotonic, such as alfentanil, sufentanil and fentanyl, may induce bradycardia and decrease cardiac output considerably if not administered with a vagolytic agent.

Ketamine is a useful agent for patients with congenital heart disease or acute shock states. However, it is a primary myocardial depressant⁸ and its negative inotropic effects may predominate in patients who have had chronic illness associated with a prolonged hyperdynamic state or patients who have been given drugs, such as dopamine, both of which are known to deplete catecholamine stores.⁹⁻¹¹

The combination of even small doses of opioids and benzodiazepines may result in severe sympatholysis, decreased systemic vascular resistance and increased venous capacitance that will limit ventricular filling.^{12,13} Since patients with cardiomyopathies are exquisitely preload dependent, the decrease in preload caused by sympatholysis may severely limit cardiac output.

Etomidate is pure sedative/hypnotic devoid of analgesic properties when used in conventional induction doses. Certain properties of etomidate make its use attractive in patients with reduced myocardial function or reserve. In studies of its cardiovascular properties, no changes were noted in inotropic state, preload or afterload when used for induction of anaesthesia; a less than ten percent change was noted in mean arterial, pulmonary capillary wedge and central venous pressures.^{14,15} Stroke volume index, cardiac index and systemic vascular resistance index changes were similar.¹⁶ The myocardial oxygen supply-demand ratio is well maintained with a concomitant decrease in coronary artery blood flow, oxygen consumption and an increase in coronary sinus oxygen saturation.¹⁷ Although etomidate has been used in patients with pulmonary hypertension with success, there are few data concerning pulmonary artery pressure responses during induction of anaesthesia in either children or adults. Etomidate also produces minimal respiratory depression, has cerebral protective effects including attenuation of the rise in intracranial pressure observed during tracheal intubation, and demonstrates rapid recovery,¹⁸ characteristics that make it attractive for brief procedures.

However, etomidate does have disadvantages including pain on injection, thrombophlebitis, myoclonus, a high incidence of nausea and vomiting postoperatively, and inhibition of steroid biosynthesis.¹⁸ Because of these inhibitory effects on steroidogenesis and an increased mortality of intensive care patients sedated with etomidate, the use of a continuous infusion for the maintenance of anaesthesia or for sedation is not recommended.^{15,19} Lidocaine injected intravenously has been used to attenuate the painful response to intravenous injection of etomidate.²⁰ Similarly, small doses of a benzodiazepine or opioid may be used to eliminate myoclonus.²¹

When general anaesthesia is indicated for a short surgical procedure in a patient with a known cardiomyopathy, the patient should be in optimal medical condition before administration of anaesthesia; dysrhythmias and other treatable causes of decreased myocardial function must be addressed. Volume status must be carefully assessed. Consideration should be given to providing intravenous dextrose to patients whose myopathic disease prevents effective utilization of energy from cellular sources. Infusions of inotropic agents should be continued throughout the induction. If a patient with a DCM is not receiving infusion of an inotropic agent, consideration should be given to starting an agent such as dobutamine perioperatively; haemodynamic improvements as well as a decrease in chronically elevated levels of atrial natriuretic factor and cyclic guanosine monophosphate levels may be seen within 30 min.^{22,23} Dobutamine also has coronary artery dilating effects that may be advantageous to the patient with a cardiomyopathy.²³⁻²⁶ Phosphodiesterase III inhibitors such as amrinone or milrinone may be particularly useful in patients with chronic ventricular dysfunction and cardiac beta receptor down regulation.²⁷⁻²⁹ This combination has also been shown to have beneficial effects on ventricular performance without an adverse elevation in myocardial oxygen consumption or lactate production.³⁰ Amrinone, however, has an elimination half-life of approximately four hours and therefore the dose cannot be titrated as rapidly as dobutamine to achieve a haemodynamic response. Patients receiving amrinone may develop a thrombocytopenia of which the anaesthetist must be aware. Positive-pressure ventilation should be initiated with care since it may have unpredictable effects on ventricular filling and performance.

Possible causes of this patient's inability to tolerate the initial anaesthetic include primary myocardial depression following ketamine, sympatholysis from the combination of opioid and benzodiazepine with consequent decreased left ventricular filling, or bradycardia from alfentanil administration decreasing cardiac output. Also, when cardiac beta receptors are chronically stimulated a decrease in their density and sensitivity can occur.³¹ This down

regulation results in reduced beta agonist-induced contractility. Therefore, the effects of exogenously administered dobutamine may have been diminished in our patient. Combining the down regulation with the patient's underlying disease increases the likelihood of ventricular dysfunction with an attenuated response to certain inotropic agents.^{32,33} To accommodate this possibility the dobutamine infusion was lowered prior to the second anaesthetic and was increased immediately before anaesthetic induction.

In summary, anaesthetists are being called upon more frequently to anaesthetize patients with severe cardiomyopathies for brief procedures. These patients require special preparation and care. Commonly used drugs used in combination or without a clear understanding of the haemodynamic actions in this population of patients may have disastrous results.

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