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CASE REPORTS/CASE SERIES

Laparoscopic cholecystectomy in a patient with Fontan circulation

Cholécystectomie par laparoscopie chez un patient avec circulation de Fontan

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Abstract

Background *Fontan circulation is created when a baby is born with only one functioning cardiac ventricle. A series of surgeries are performed to allow the ventricle to provide oxygenated blood to the systemic circulation and to create passive flow of venous blood to the pulmonary circulation via a conduit. Laparoscopic surgery poses several hemodynamic challenges to a patient with Fontan physiology attributable to carbon dioxide insufflation, positive pressure ventilation, and reverse Trendelenburg positioning.*

Clinical features *A 39-yr-old male with a Fontan physiology was referred to our tertiary care centre because of repeated bouts of cholecystitis requiring a percutaneous drain and now elective laparoscopic cholecystectomy. Because of repeated cardiac surgeries, the patient also had complete heart block and was pacemaker dependent. We placed an arterial catheter prior to induction of general anesthesia with tracheal intubation. Transesophageal echocardiography allowed for real-time intraoperative assessment of venous blood flow through the patient's extracardiac diversion system throughout the surgery. This information was used to guide management and determine circulation tolerance during the various stages of laparoscopy. Inhaled milrinone resulted in the shunt fraction returning to the patient's baseline. Intraperitoneal pressure was kept below 10 mm Hg, and systemic blood pressure was supported with a low-dose norepinephrine infusion.*

Conclusions *Intraoperative transesophageal echocardiography is a useful monitoring device during laparoscopic surgery when a patient has Fontan circulation. Knowing how to administer inhaled milrinone is a useful skill to decrease the shunt fraction through a patient's conduit, increasing pulmonary blood flow while avoiding hypotension.*

Résumé

Contexte *La circulation de Fontan est créée lorsqu'un bébé naît avec un seul ventricule cardiaque fonctionnel. Une série d'interventions chirurgicales est pratiquée pour permettre au ventricule de fournir du sang oxygéné à la circulation systémique et de créer un flux passif de sang veineux vers la circulation pulmonaire via un conduit. La chirurgie par laparoscopie pose plusieurs défis hémodynamiques à une personne présentant une physiologie de Fontan, attribuables à l'insufflation de dioxyde de carbone, à la ventilation par pression positive et au positionnement en Trendelenburg inversé.*

Caractéristiques cliniques *Un homme de 39 ans présentant une physiologie de Fontan a été référé à notre centre de soins tertiaires en raison d'épisodes répétés de cholécystite nécessitant un drainage percutané puis une cholécystectomie non urgente par laparoscopie. En raison de chirurgies cardiaques répétées, le patient avait également un bloc cardiaque complet et dépendait d'un stimulateur cardiaque. Nous avons placé un cathéter artériel avant l'induction de l'anesthésie générale avec intubation trachéale. L'échocardiographie transœsophagienne a permis d'évaluer en temps réel la*

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circulation sanguine veineuse à travers le système de dérivation extracardiaque du patient tout au long de la chirurgie. Cette information a été utilisée pour guider la prise en charge et déterminer la tolérance à la circulation au cours des différentes étapes de la laparoscopie. L'inhalation de milrinone a entraîné le retour de la fraction de dérivation aux valeurs de base du patient. La pression intrapéritonéale a été maintenue en dessous de 10 mm Hg, et la tension artérielle systémique a été soutenue par une perfusion de noradrénaline à faible dose.
Conclusion *L'échocardiographie transœsophagienne peropératoire est un dispositif de monitorage utile lors d'une chirurgie laparoscopique lorsqu'un patient a une circulation de Fontan. Savoir comment administrer la milrinone par inhalation est une compétence utile pour diminuer la fraction de dérivation à travers le conduit d'un patient, augmentant ainsi la circulation sanguine pulmonaire tout en évitant l'hypotension.*

Keywords case report · Fontan circulation · laparoscopic surgery · transesophageal echocardiography

Background

First described in 1971, the Fontan procedure was introduced as a palliative correction for tricuspid atresia.[1] Conceptually, a Fontan procedure is any surgical technique that diverts systemic venous return to the pulmonary circulation via a conduit instead of passing through a ventricle. The conduit can be a prosthetic tube outside the heart (an extracardiac shunt) or via the atria (an intracardiac shunt). Anything that affects the pressure differential through the conduit will change blood flow to the lungs, oxygenation, and ventilation and pulmonary pressures. To protect the pulmonary circulation from being exposed to high venous pressures, a "pop off" system (conduit) is also created; increased venous return pressure will result in shunting of deoxygenated blood through the conduit and into the systemic circulation, bypassing the lungs.

Laparoscopic surgery poses unique challenges to patients with Fontan circulation. Reverse Trendelenburg position and positive pressure ventilation both decrease systemic venous return.[2] Insufflation of the abdomen with carbon dioxide (CO_2) both increases the systemic venous partial pressure of carbon dioxide (pCO_2) and intrathoracic pressure, potentially increasing pulmonary vascular resistance (PVR).[2,3] Combined, these changes can result in a simultaneous decrease in venous flow into the heart with proportionally more of this deoxygenated blood directly entering the systemic circulation via the conduit.

There is limited literature describing the anesthetic management in patients with Fontan circulation undergoing laparoscopic procedures.[4–6] The use of point-of-care ultrasound and/or transesophageal echocardiography (TEE) as potential monitoring tools to assess, in real time, the hemodynamic changes associated with laparoscopic surgery also remains unknown.

The evolution of the Fontan procedure has resulted in more patients surviving into adulthood. As a result, anesthesiologists may encounter patients with Fontan circulation presenting for noncardiac surgery. This case report describes an adult patient with Fontan physiology undergoing laparoscopic cholecystectomy and the use of intraoperative transesophageal echocardiography.

Case presentation

The patient consented to the publication of this case report. The patient was a 39-yr-old man, who presented for elective laparoscopic cholecystectomy at The Ottawa Hospital (Ottawa, ON, Canada). When he initially presented elsewhere with acute cholecystitis and sepsis, he had developed episodes of nonsustained ventricular tachycardia. These resolved with treatment of his sepsis and the insertion of a percutaneous drain.

The patient had a history of a univentricular heart with a morphologic right systemic ventricle, mitral atresia, and pulmonic atresia. Flow of systemic venous return to the pulmonary circulation was via an extracardiac bicaval conduit. Also present was a fenestration connecting the extracardiac conduit to the common atrium. Should pulmonary pressure increase, this fenestration allows venous blood to flow into the lower pressure common atrium, thereby maintaining systemic circulation (albeit with venous blood). Pulmonary veins, supplying oxygenated blood from his lungs, are connected to a common atrium. The common atrium is connected to a single morphologically right ventricle responsible for his systemic circulation.

During his first admission with acute cholecystitis, the patient was managed by a multidisciplinary team involving cardiologists, infectious disease specialists, gastroenterologists, and general surgeons. In view of his complex medical history and elevated perioperative risk, the decision was made to treat him conservatively. The patient responded well to antibiotics, marked by resolving fever and abdominal pain. Laparoscopic cholecystectomy was arranged electively for a later date at our centre, a tertiary care facility equipped with appropriate resources including intensive care access and an anesthesiologist certified in advanced TEE.

Despite his cardiac complexity and multiple cardiac surgeries resulting in complete heart block and pacer dependency, the patient had excellent functional capacity including working fulltime as a property manager. On preoperative assessment, he was noted to be mildly cyanotic with digital clubbing. His peripheral oxygen saturation on room air was 82% with a blood pressure of 126/89 mm Hg. The patient stated that these findings were normal for him. Preoperative transthoracic echocardiography showed a moderately reduced systolic function, mild to moderate atrioventricular valve regurgitation, and a patent conduit. His hemoglobin was 170 $g \cdot dL^{-1}$; the remainder of his laboratory results were unremarkable.

The patient discontinued his chronic warfarin medication five days prior to surgery, with anticoagulation being continued perioperatively with subcutaneous dalteparin. On the day of surgery, his pacemaker was programmed to an asynchronous ventricular pacing mode with a rate of 80 beats per minute. The surgeon's goal was to maintain a low intra-abdominal pressure (10–12 mm Hg).[6] Procedural termination criteria, communicated among the team members, included but were not limited to deteriorating hemodynamic parameters and unstable arrhythmias.

We inserted a preinduction arterial catheter and placed a five-lead electrocardiogram, peripheral nerve stimulator, and monitors according to the standard Canadian Anesthesiologists' Society guidelines.[7] External defibrillation/pacing pads were also placed. General anesthesia was induced with fentanyl (200 µg *iv*), lidocaine (100 mg *iv*), propofol (slowly titrated to 300 mg *iv*), and rocuronium (50 mg *iv*). The dose of propofol was surprisingly large; however, this was required to achieve loss of voice response and eyelash reflex. No significant hypotension occurred despite the required propofol dose. Sevoflurane and an intravenous remifentanil infusion were used for maintenance. A mean arterial pressure of $\geq$ 65 mm Hg was maintained with a low-dose intravenous norepinephrine infusion.[8] Inhaled milrinone (5 mg) was available via nebulization via the tracheal tube in case of an increase in PVR. This was diagnosed indirectly by an increase in venous return through the conduit and into the systemic circulation. The setup of inhaled milrinone is described in a separate section. We used positive pressure ventilation with a maximum airway pressure of 23 cm H_2O, and we maintained pneumoperitoneum at 10 mm Hg. An initial arterial blood gas prior to abdominal insufflation showed respiratory acidosis (pH, 7.27; pCO_2, 56 mm Hg; HCO_3, 26 $mEq \cdot L^{-1}$). Although some permissive hypercapnia was acceptable, minute ventilation was adjusted such that pCO_2 was kept in the mid-40s. One-thousand millilitres of intravenous crystalloid was given and minimal blood loss was observed.

Intraoperative TEE showed a univentricular heart with mild global dysfunction (ejection fraction, 41–51%). The extracardiac conduit was visualized connecting the inferior vena cava to the main pulmonary artery with fenestration to the common atrium. There was intermittent flow of desaturated blood through the conduit into the common atrium and, from there, to the systemic circulation. Conduit flow was seen to increase approximately 10 min after abdominal insufflation. Conduit flow decreased after 5 mg of milrinone was nebulized into the respiratory circuit. Administering milrinone via the respiratory circuit decreases the PVR while limiting systemic hypotension, allowing more venous return through the lungs and reducing flow through the conduit. A wall-hugging jet was identified at the atrioventricular valve, indicating moderate to severe atrioventricular valve regurgitation. Spontaneous echo contrast was visualized in the hepatic veins, consistent with slow systemic venous return. Figures 1 and 2 show the anatomy of the patient's Fontan circulation. A clip showing flow through the fenestration can be found in the Electronic Supplementary Material eVideo.

The surgery proceeded uneventfully. Following neuromuscular reversal with sugammadex, the patient was easily extubated and was monitored in the postanesthesia care unit overnight. There were no postoperative complications, and the patient was discharged the following morning.

Intraoperative inhaled milrinone setup

Anesthesia assistants at The Ottawa Hospital are responsible for assembling the delivery device for patients requiring inhaled milrinone in the operating room. The device setup for ventilated patients is shown in Fig. 3. As can be seen, various connectors and corrugated tubing are assembled and a low flow jet nebulizer is used. Pressurized oxygen is delivered via oxygen tubing at 2 $L \cdot min^{-1}$. It is important to have the nebulizer positioned between the patient and the heat and moisture exchanger (HME); otherwise, the majority of the inhaled milrinone will get trapped in the HME. It is also necessary that the nebulizer remains upright for it to function correctly. The nebulizer shown in Fig. 3 is the MiniHeart® Lo-Flo Nebulizer (Westmed, Inc., Tuscon, AZ, USA; part number, WES100611). Its reservoir holds a maximum of 30 mL of fluid. This low-flow nebulizer creates particle sizes of 2–3 µm, which allows for proper deposition in the lungs. The nebulizer has an intravenous port where the initial and sequential doses of milrinone can be injected.

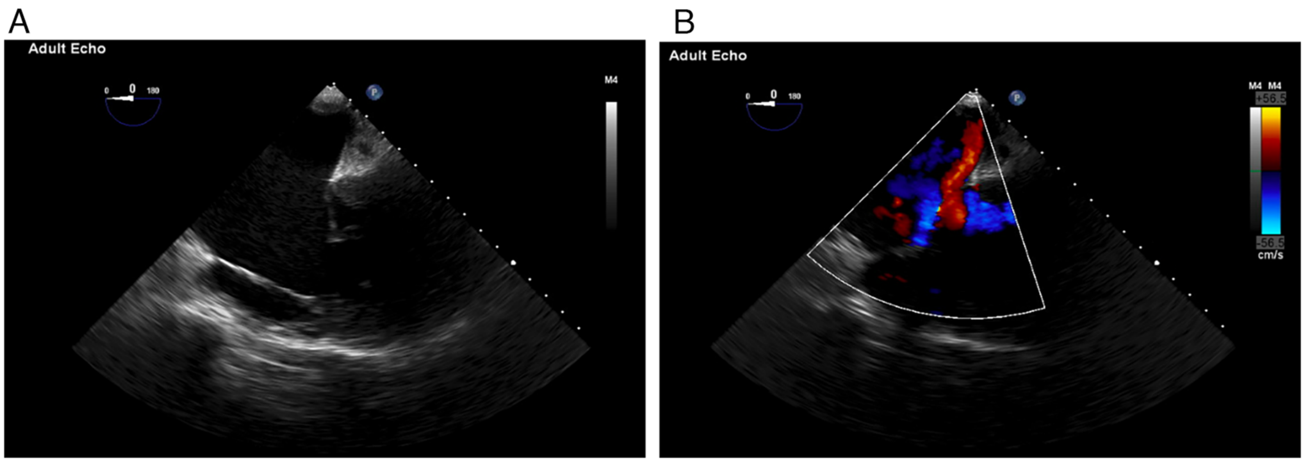

Fig. 1 Transesophageal echocardiography (TEE) in the midesophageal four-chamber view. (A) Two-dimensional TEE shows a common atrium and a single ventricle with an atrioventricular valve. (B) Colour Doppler TEE shows a wall-hugging jet indicating moderate to severe atrioventricular valve regurgitation.

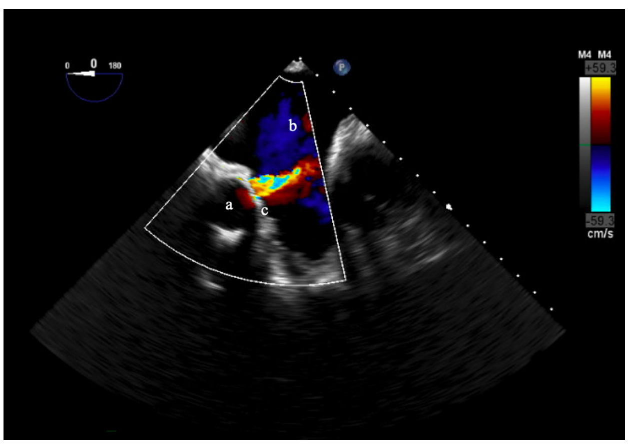

Fig. 2 A shunt flow is seen across an (a) extracardiac conduit into a (b) common atrium through a (c) fenestration

Discussion

Physiologic challenges associated with laparoscopic cholecystectomy are mainly attributed to intra-abdominal CO_2 insufflation with reverse Trendelenburg positioning.[3] Mechanical and neuroendocrine effects of CO_2 insufflation and reverse Trendelenburg positioning can result in impaired venous return, an increase in mean arterial pressure and systemic vascular resistance, and a possible decrease in cardiac output.[3,9] Carbon dioxide absorption also increases PVR, which can lead to increased venous blood flow into the systemic circulation via the patient's conduit. With respect to respiratory mechanics, pneumoperitoneum decreases lung compliance and functional residual capacity, leading to an increase in airway pressure resulting in decreased pulmonary blood flow.[3] As successful Fontan circulation relies on the maintenance of low pulmonary vascular resistance, these physiologic disturbances can lead to life- and organ-threatening consequences such as decreased cardiac output in combination with much of this cardiac output being deoxygenated venous blood.[4,10]

The hemodynamic goals in individuals with a Fontan circulation centres around preserving the systemic venous flow to the pulmonary circulation by ensuring adequate preload, avoiding an elevation in PVR while preserving sinus rhythm (if present) and cardiac contractility.[10] Before induction, the present patient was given 500 mL of intravenous crystalloid. To attenuate the effects of the pneumoperitoneum decreasing preload, intra-abdominal pressure was kept at or below 10 mm Hg. Although the conventional view is that pneumoperitoneum causes impaired venous return, various reports showed that cardiac output does not decrease if insufflation pressure is below 12 mm Hg in healthy individuals.[2,11] In fact, one study has suggested that venous return may be augmented if the intra-abdominal pressure is lower than the central venous pressure (CVP).[12] A number of case reports have shown that people with Fontan circulation may tolerate an intra-abdominal pressure of 10–12 mm Hg as this will not be higher than their chronically high CVP.[4–6] We found this to be the case in our patient. Arterial blood gas analysis was used to monitor the level of pCO_2 as end-tidal CO_2 might not be reliable in patients with Fontan circulation.[13] Other precautionary measures included deairing all of the lines to prevent the risk of paradoxical air embolism and limiting the extent of reverse Trendelenburg positioning.

Intravenous milrinone is widely used for the treatment of pulmonary hypertension because of its inodilator

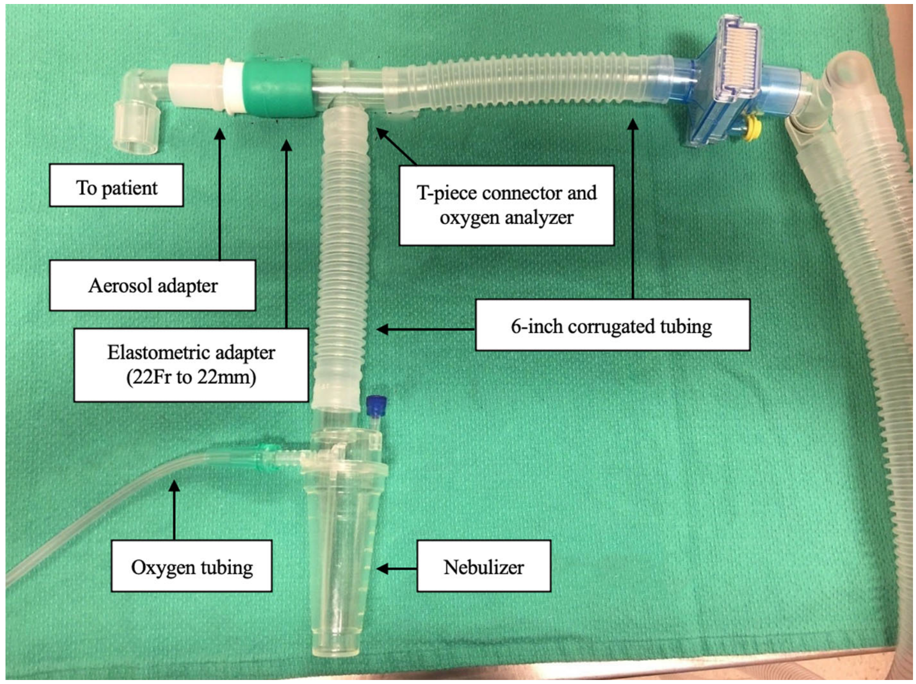

Fig. 3 Assembly of intraoperative inhaled milrinone via the MiniHeart® Lo-Flo Nebulizer (Westmed, Inc., Tuscon, AZ, USA; part number, WES100611) in the anesthesia breathing circuit. Oxygen tubing delivering pressurized oxygen at 2 $L \cdot min^{-1}$ is connected to the low-flow jet nebulizer, which contains 5 mg of milrinone (1 $mg \cdot mL^{-1}$). The jet nebulizer is connected to the anesthesia circuit using six-inch corrugated tubing and a T-piece connector as shown. The elastometric adapter (22F to 22 mm) is used in conjunction with the aerosol tubing adapter to connect the T-piece connector to the patient.

properties.[14] Due to the potential for systemic hypotension, we used inhaled milrinone as an alternative to intravenous administration.[15] A recent multicentre study in cardiac patients found that inhaled milrinone was associated with increased cardiac output and decreased systolic pulmonary artery pressures without causing systemic hypotension.[15] We found this to be the case in our patient in conjunction with a decrease in the shunt fraction through his extracardiac conduit into his systemic circulation.

Intraoperative TEE provided useful information regarding the anatomy, function, and changes in conduit flow with the physiologic challenges associated with laparoscopic cholecystectomy. Real-time assessment of systemic and pulmonary circulation during surgical insufflation and positioning provided invaluable guidance for patient management, including the timing of inhaled milrinone administration.

Conclusion

Laparoscopic cholecystectomy presented unique hemodynamic challenges for an adult patient with Fontan circulation. Intraoperative TEE provided real-time feedback for hemodynamic management. Inhaled milrinone to promote pulmonary blood flow without systemic hypotension was very useful and relatively easy to administer. Coordinated communication between anesthesiology, surgery, cardiology, and the patient led to the formulation of a care strategy well before the day of surgery. Referral to a tertiary care centre that could provide such a robust care strategy provided the patient with a high standard of care.

Author contributions All authors were involved in the management of the patient. *Rattanaporn Tankul* obtained consent from the patient, conducted the literature review, prepared the figures, and authored the first draft of the manuscript. *Becky Rodrigues* conducted the literature review, contributed to the first draft of the manuscript, and prepared a figure. *Laura Duggan* conducted the literature review and contributed significantly to the revisions of the manuscript. All authors were involved in the revision process.

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