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Case Report
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Anesthesia for Laparoscopic Cholecystectomy in a Patient with Eisenmenger's Syndrome and Down's Syndrome

*Claire V Chatelain[1], Océane J Jaquet[1], Olivier Detry[2], Julien Guiot[3] and Jean L Joris[1]**

[1]Department of Anesthesia and Reanimation, CHU Liège, University of Liège, Belgium

[2]Service of Digestive Surgery, CHU Liège, University of Liège, Belgium

[3]Service of Pneumology, CHU Liège, University of Liège, Belgium

Abstract

Eisenmenger Syndrome (ES) may be defined in pathophysiologic terms as pulmonary arterial hypertension caused by a high pulmonary vascular resistance, with reversed or bidirectional shunt at aortopulmonary, ventricular, or atrial level. Perioperative management of patients with ES is challenging. Anesthetic management aims at avoiding factors that decrease systemic vascular resistance or increase PVR which would increase right to left shunt and hypoxemia. We report the case of a 45 years man with Eisenmenger's syndrome and Down's syndrome scheduled for laparoscopic cholecystectomy.

Introduction

Eisenmenger Syndrome (ES) may be defined in pathophysiologic terms as Pulmonary Arterial Hypertension (PAH) caused by a high Pulmonary Vascular Resistance (PVR), with reversed or bidirectional shunt at aortopulmonary, ventricular, or atrial level [1,2]. Perioperative management of patients with ES is challenging. Non-cardiac surgery is associated with increased risk of cardiovascular complications, such as atrial or ventricular arrhythmias, major bleeding, thromboembolic events, hypoxemia, and death (5% to 25%) [1-3]. Hemodynamic changes related to anesthesia, ventilation, fluid shift, or surgery itself have to be monitored to maintain a balance between pulmonary and systemic blood flow. Anesthetic management aims at avoiding factors that decrease Systemic Vascular Resistance (SVR) or increase PVR which would increase right to left shunt and hypoxemia. These factors are commonly hypovolemia, hypoxemia, hypercarbia, acidosis, and various anesthetic agents and techniques [1-4]. In previous reports, a variety of anesthesia techniques have been used for patients with Eisenmenger syndrome for non-cardiac surgery. However, most of the published papers focus either on the parturient [5] or children, particularly with Down's syndrome [6]. The prevalence of cholelithiasis and asymptomatic gallstones is significantly high in patients with cyanotic congenital heart disease [7]. Laparoscopic cholecystectomy classically proposed to treat this pathology requires CO_2-pneumoperitoneum which results in hypercarbia, increased PVR, and systemic hemodynamic changes potentially deleterious in patients with Eisenmenger's syndrome [8,9]. We report the case of a 45 years man with Eisenmenger's syndrome and Down's syndrome scheduled for laparoscopic cholecystectomy. His legal representative gave informed consent for this case report.

Case Presentation

A 42-year-old male (68 kg, 160 cm) with a history of Eisenmenger's syndrome due to a peri-membranous Ventricular Septal Defect (VSD), PAH, and right to left shunt was admitted for elective laparoscopic cholecystectomy for symptomatic cholelithiasis. Over the previous 5 years he had experienced recurrent increasing shortness of breath on exertion and one episode of pulmonary embolism. The chronic treatment for PAH consisted of macitentan 10 mg (an endothelin-1 receptors antagonist), tadalafil 40 mg (a phosphodiesterase-5 inhibitor), and dabigatran 110 mg twice a day. He is also treated intermittently with oxygen 2 $L.min^{-1}$ at home. Thanks to the optimization of his treatment of PAH he could climb a staircase of 15 steps without stopping and without oxygen (NYHA II dyspnea). He had no history of syncope. On examination we found cyanosis, marked clubbing, clear lungs, and no ankle edema. The electrocardiogram demonstrated right axis deviation and a right ventricular hypertrophy. Preoperative echocardiography demonstrated a high VSD with right to left flow, a dilated right heart and right ventricle hypertrophy with preserved function. Transtricuspid gradient was 83 mmHg. Left ventricle function was reported to be normal. Preoperative blood tests

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Correspondence:
Jean L Joris, Department of Anesthesia and Reanimation, CHU Liège, University of Liège, Domaine du Sart Tilman, Avenue de l'Hôpital, B-35, B-4000 Liège, Belgium, Tel: +32-4-3667180; Fax +32-4-3667636; E-mail: jean.joris@chuliege.be

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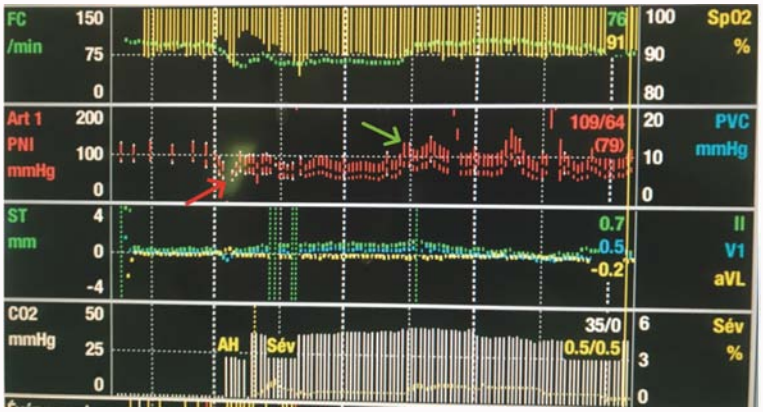

Figure 1: Hemodynamic parameters, SpO_2, $PETCO_2$, and end-tidal concentration of sevoflurane.
Art.: Invasive Arterial Pressure; PNI: Non-Invasive Blood Pressure; CO_2: $PETCO_2$; Sév: End-Tidal Concentration of Sevoflurane
Red arrow = induction of anesthesia; green arrow = creation of pneumoperitoneum

Table 1: Intraoperative arterial blood gas analyses.

	Post-induction	15 min after PNP	40 min after PNP
pH	7.39	7.29	7.30
$PaCO_2$: mmHg	42	53	49
$P_{ET}CO_2$: mmHg	35	38	34
PaO_2: mmHg	71	67	67
Base excess: mmol.L^{-1}	0.3	-1.9	-2.8
HCO_3 std: mmol.L^{-1}	24.9	23.1	22.4
Lactate: mg.dL^{-1}	14	8	7
Tidal volume: mL	350	350	400
Respiratory rate: min^{-1}	12	18	18
F_IO_2	0.55	0.55	0.55

PNP: Creation of Pneumoperitoneum

showed hemoglobin: 19.3 g.dL^{-1}; hematocrit: 61%; platelets: 137,000 L^{-1}; leukocytes: 4,400 L^{-1}; normal renal function and hepatic tests; and NT-Pro-BNP (N-terminal Pro-Brain Natriuretic Peptide) = 16 U.L^{-1} (normal value <300). The secondary erythrocytosis was regularly treated by venesection when hemoglobin concentration increased above 20 g.dL^{-1}. Peripheral Oxygen Saturation (SpO_2) was 88% in sitting position and 82% in recumbent position breathing room air.

Anesthetic and surgical management

The patient calm and cooperative were given no premedication. Antibiotic prophylaxis for endocarditis consisted of cefazoline 2 g. On arrival in the operating room, SpO_2 was 89% in recumbent position while breathing oxygen 4 L.min^{-1}. Pre-warming and prevention of hypothermia were achieved using forced warm air blanket. An infusion of noradrenaline was ready to treat potential hypotension. Target-controlled infusion of remifentanil 2.5 ng.mL^{-1} (Minto's model), propofol 50 mg, and ketamine 50 mg were used to induce anesthesia and neuromuscular block was achieved with rocuronium. Despite a Mallampati-1 score and a correct mouth opening, we used a video-laryngoscope for tracheal intubation to avoid multiple laryngoscopies and its associated hemodynamic changes. Anesthesia was maintained with target-controlled infusion of remifentanil 2.0 ng.mL^{-1} and sevoflurane at a 0.5% to 1.0% end-tidal concentration. Oxygen enriched air (F_IO_2 0.5-0.6) was titrated to maintain SpO_2 around 90 %. During induction of anesthesia, arterial pressure decreased from 123/80 mmHg to 69/43 mmHg and heart rate from 92 bpm to 64 bpm with a decrease in oxygen saturation to 84% (Figure 1). Infusion of noradrenaline was started to maintain systemic pressure. As expected, there was a direct correlation between arterial systemic pressure and oxygen saturation and it was necessary to maintain systolic arterial pressure at 100 mmHg to 110 mmHg to keep oxygen saturation around 90%. Midazolam 2 mg was given to deepen anesthesia while reducing the need for sevoflurane and its associated risk of hypotension. Before and during surgery, low doses of noradrenaline (range 0.025 µg kg^{-1} min^{-1} to 0.10 µg kg^{-1} min^{-1}) were continuously titrated to maintain systolic arterial pressure above 100 mmHg. Inhalation of 10 ppm of NO tested before surgery during 5 min while patient in stable condition had no effect on SpO_2. Pneumoperitoneum was created after intravenous infusion of 500 ml of lactated Ringer solution to prevent significant reduction of venous return secondary to the increased intra-abdominal pressure. Intraabdominal pressure was set at 8 mmHg and reached slowly using a CO_2 insufflation rate of 1 L.min^{-1}. Pulse pressure variation remained always inferior to 10%. Adequate surgical working conditions were ensured thanks to deep muscle relaxation (no response to the train of four). Creation of pneumoperitoneum resulted in a slight increase in arterial pressure (135/76 mmHg) with no drop in SpO_2 and corrected by increasing end-tidal sevoflurane concentration to 1.0% (Figure 1). The increase in end-tidal PCO_2 ($PETCO_2$) during CO_2-pneumoperitoneum was countered by increasing the respiratory rate rather than the tidal volume to limit the increase in intrathoracic pressure. A head-up tilt of 15° with lower limbs raised was allowed. Repeated blood gas analyses were performed to adjust ventilation and avoid hypercapnia (Table 1). The surgeon complained of abnormal bleeding and oozing in the bladder bed, which was treated by 1000 mg intravenous tranexamic acid. Postoperative multimodal analgesia was provided with 1 g paracetamol, 40 mg parecoxib, and 100 mg tramadol intravenously combined with infiltration of the different trocarts sites with levobupivacaine. Surgery was uneventful and on completion neuromuscular blocking agents were antagonized with sugammadex to allow quick return of normal spontaneous ventilation. After removal of the endotracheal tube, the patient breathing oxygen 6 L.min^{-1} was transferred to the recovery room for postoperative monitoring. The noradrenaline infusion was weaned gradually and was stopped within 1 h. The patient was discharged to the surgical ward 3 h after the end of surgery. Postoperative recovery was uneventful. Patient was allowed to drink 2 h and eat 4 h after surgery. Ambulation supervised by a physiotherapist occurred 4 h postoperatively. Patient complained of only minor pain relieved by

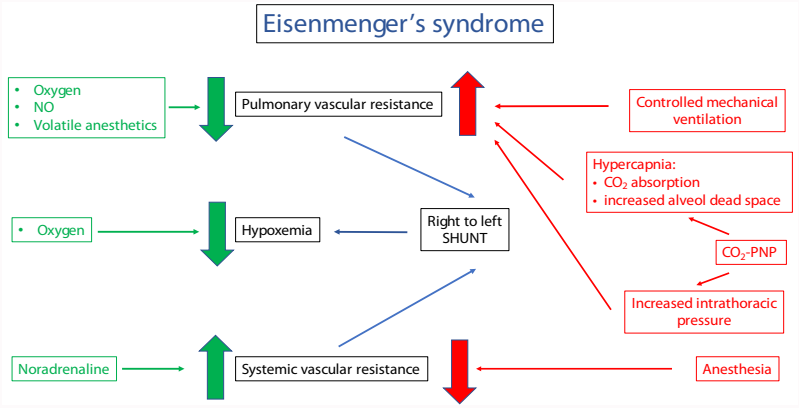

Figure 2: Schematic representation of the pathophysiologic changes during laparoscopy and CO_2 pneumoperitoneum in patient with Eisenmenger's syndrome. PNP: Pneumoperitoneum

paracetamol and left the hospital the day after surgery.

Discussion

The global risk of anesthesia-related complications in patients with Eisenmenger's syndrome is high and needs to be evaluated by a dedicated multidisciplinary team used to deal with patients suffering from PAH. The main acute complication results from a decrease in systemic vascular resistance and subsequent increase in right to left shunt and hypoxemia. Careful management of systemic vascular resistance is therefore required [1-3]. In this context, continuous invasive arterial monitoring upon induction of anesthesia is mandatory. A pulmonary artery flotation catheter was not used because we considered the risks of misdirection and cardiac arrhythmias outweighed the potential benefits in this patient given the good biventricular function demonstrated by the preoperative echocardiography. We decided not to use intraoperative transesophageal echocardiography because of the general condition of the patient, his good biventricular function, and the minimally invasive nature of the surgical procedure. Despite good biventricular function and the administration of a small dose of propofol, a drop of arterial blood pressure occurred associated with a decrease in SpO_2 during the induction of anesthesia. To correct this almost inevitable decrease in systemic vascular resistance and hypotension at induction, we immediately initiated an infusion of small dose of noradrenaline which provided easy and rapid control of arterial pressure. No premedication was given to avoid the risk of preoperative respiratory depression, hypoventilation and hypoxemia. On arrival in the operating room, SpO_2 was 88% to 90% while breathing nasal oxygen with a flow of 4 $L.min^{-1}$. Hypoxemia caused by chronic right to left shunt is usually weakly reversible by oxygen therapy. A potential vasodilator response of the pulmonary vasculature to oxygen reducing PVR associated with an improvement in pulmonary arterial flow and decreased flow across the shunt [10] is not expected in these chronic conditions. Likewise, no improvement of SpO_2 was observed in our patient during NO inhalation suggesting the absence of pulmonary vascular reversibility of his long-lasting PAH. Maintaining systemic arterial pressure allowed to avoid worsening of hypoxemia after tracheal intubation and positive pressure ventilation. To reduce the impact of mechanical controlled ventilation on pulmonary hypertension, lungs were ventilated with low tidal volume (6 $mL.kg^{-1}$), no positive end expiratory pressure, and a respiratory rate adjusted to avoid hypercapnia. After surgery, trachea was quickly extubated to remove the potential deleterious effect of positive pressure ventilation. Complications associated with anesthesia for laparoscopic procedures in patients with Eisenmenger's syndrome are manifold. CO_2-pneumoperitoneum causes an increase in $PaCO_2$ and $PETCO_2$ secondary to absorption of CO_2 from the peritoneal cavity [8]. Furthermore, in patient suffering from pulmonary disease, the rise in intra-abdominal pressure can increase alveolar dead space evidenced by an enlargement of the gradient between $PaCO_2$ and $PETCO_2$ as described in Table 1 during our procedure [8]. Acidosis, arrhythmias and hypotension may follow, all of which can precipitate a shunt crisis. Repetitive arterial blood gas analyses allowed us to adjust patient ventilation and avoid hypercapnia. Hyperventilation to correct hypercapnia as well as the increase in intrathoracic pressure secondary to pneumoperitoneum further increase pulmonary vascular resistance [8,9]. To limit the impact of controlled ventilation on pulmonary pressure, hyperventilation was achieved more by an increase of the respiratory rate rather than an increase of tidal volume. Furthermore, the used of low intra-abdominal pressure (8 mmHg) allowed to attenuate the respiratory and ventilatory consequences as well as the hemodynamic changes secondary to CO_2-pneumoperitoneum [8]. Indeed, only a slight increase in arterial blood pressure was observed at the creation of the pneumoperitoneum. Preventing intraoperative hypothermia and ensuring adequate postoperative analgesia are important concerns to avoid an increase of pulmonary vascular resistance. Finally, the risk of gas embolism and paradoxical embolism secondary to the right to left shunt should deserve a special attention. The potential pathophysiologic changes secondary to laparoscopy and CO_2-pneumoperitoneum in patient with Eisenmenger's syndrome are represented in Figure 2. Arterial and venous thromboembolic complications secondary to blood stasis, erythrocytosis, dilated heart chambers, endothelial dysfunction, and atrial arrhythmia are common in these patients. Our patient had a history of pulmonary embolism. However, patients with Eisenmenger's syndrome are also at increased risk of bleeding. Hemostatic abnormalities are attributed both to platelet disorders (thrombocytopenia and thrombasthenia) and abnormalities in coagulation pathways, and increased hematocrit has been correlated with impaired fibrinogen function [2]. Likewise, our surgeon complained of abnormal bleeding in the bladder bed.

In conclusion, anesthetic management of patient with Eisenmenger's syndrome for laparoscopic cholecystectomy is challenging. This management should avoid decreases in systemic vascular resistance and subsequent increase in right to left shunt and hypoxemia. Maintenance of arterial pressure is therefore required

using noradrenaline infusion if necessary. Increase in pulmonary vascular resistance occurs during CO_2-pneumoperitoneum because of the hyperventilation necessary to prevent hypercapnia associated with CO_2-intraperioneal insufflation and the increased intrathoracic pressure secondary to the increased intra-abdominal pressure. These deleterious effects can be attenuated by using low intra-abdominal pressure.

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