

Cesarean section in a patient with non-compaction cardiomyopathy managed with ECMO “stand-by”

A.A. Koster¹, F. Pappalardo², S. Silveti², U. Schirmer¹, J. U. Lueth¹, R. Dummler³, M. Emmerich³, M. Schmitt⁴, G. Kirchner⁵, D. Kececioglu⁵, E. Sandica⁶

¹Institute of Anaesthesiology, Heart and Diabetes Centre North Rhine-Westphalia, Bad Oeynhausen, Ruhr-University Bochum, Germany; ²Department of Anesthesia and Intensive Care, San Raffaele Scientific Institute, Milan, Italy; ³Department of Anaesthesiology, Hospital of Bad Oeynhausen, Germany; ⁴Department of Gynaecology and Obstetrics, Hospital of Bad Oeynhausen, Germany; ⁵Clinic for Congenital Heart Disease, Heart and Diabetes Centre North Rhine-Westphalia, Bad Oeynhausen, Ruhr-University Bochum, Germany; ⁶Department of Surgery of Congenital Heart Disease, Heart and Diabetes Centre North Rhine-Westphalia, Ruhr-University Bochum, Bad Oeynhausen, Germany

Heart, Lung and Vessels. 2013; 5(3): 183-186

ABSTRACT

Isolated ventricular non-compaction is a rare cardiomyopathy associated with left heart failure, severe arrhythmias and thromboembolism. We report about our interdisciplinary strategy in a patient with severe isolated ventricular non-compaction cardiomyopathy scheduled for caesarean section in general anaesthesia. Monitoring included placement of an arterial line, a central venous catheter and a pulmonary artery catheter with pacing option. Small introducer gates were placed in the femoral artery and vein to facilitate quick percutaneous institution of extracorporeal life support via extracorporeal membrane oxygenation in case of acute cardiac failure refractory to medical treatment. Inotropic pharmacological therapy with 3 µg/kg/min dobutamine and 0.25 mg/kg/min milrinone was started before surgery. Induction of general anaesthesia and rapid sequence intubation was performed with an analgesic dose of 0.5 mg/kg S ketamine, 0.25 mg/kg etomidate and 5 mg rocuronium followed by 1.5 mg/kg succinylcholine. This regimen provided completely stable hemodynamics in this critical period until delivery of the child and continuation of anaesthesia with continuous infusion of propofol and remifentanyl. The current strategies, particularly the preparation for femoro-femoral extracorporeal membrane oxygenation, may be considered in similar cases with a high risk of acute cardiac decompensation which may be refractory to medical treatment. Anaesthesiologist involved in performing caesarean section in women with complex cardiac disease, should encompass extracorporeal membrane oxygenation standby in management of the perioperative period.

Keywords: ECMO, heart failure, setio.

INTRODUCTION

Heart disease and thromboembolism are the leading indirect causes of maternal death during pregnancy (1, 2). Isolated ventricular non-compaction cardiomyopathy

(IVNC) is a relatively rare (incidence of approximately 0.05 % in adult population), but thought to be underdiagnosed disease, caused by intrauterine arrest of compaction of the myocardial fibers and meshwork (3, 4). The patho-anatomy is characterized by the presence of deep intertrabecular recesses in the hypertrophic and hypokinetic segments of the left ventricle (3, 4). The diagnosis of IVNC is performed by echocardiography, which apart from the mas-

Corresponding author:

Federico Pappalardo
Department of Anesthesia and Intensive Care
San Raffaele Scientific Institute
Via Olgettina, 60 - 20132 Milan, Italy
e-mail: pappalardo.federico@hsr.it

sive myocardial trabeculations, shows the characteristic myocardial 2-layers structure consisting of a thin compacted epicardial and a thicker non-compacted endocardial layer (3, 4). Leading clinical symptoms of IVNC are signs of heart failure, systemic thromboembolism and arrhythmias.

We herein report about our considerations and management of a patient with IVNC and severely impaired left ventricular function, who underwent a caesarean section at our institution.

CASE REPORT

The 39-year old, 68 kg and 165 cm patient, mother of a 20 year old son, was referred to our institution during the 33rd week of pregnancy. One year before, she had experienced multiple transitory ischemic attacks, she was in NYHA (New York Heart Association) class IV and IVNC was diagnosed based on echocardiography. Besides warfarin, which during pregnancy was switched to low molecular weight heparin, pharmacological therapy with an angiotensin converting enzyme inhibitor had been established. Although severe ventricular arrhythmias had been diagnosed, the patient refused implantation of an internal defibrillator. The transthoracic echocardiography (TTE) at admission showed the typical picture of IVNC: a massively dilated left ventricle (end-diastolic diameter of 80 mm - normal value < 50 mm), a mitral valve regurgitation grade I, a severely impaired left ventricular ejection fraction (LVEF) of 25 % (normal value > 55 %) and a semi-systemic systolic pulmonary artery pressure (PAP) of approximately 60 mmHg. Due to the inherent risk of cardiac decompensation in the late pregnancy, the patient was admitted to our heart centre one week later and elective caesarean section scheduled for the 35th week of gestation. Due to the risk

of sudden life-threatening decompensation during surgery, it was planned to perform the operation under general anaesthesia and “stand by” of veno-arterial extracorporeal membrane oxygenation (ECMO) for extracorporeal life support. On the day before surgery, TTE revealed further decrease of the LVEF to 20 %, increase of the mitral valve regurgitation to grade II and increase of the systolic PAP to 2/3rds of the systemic value of approximately 80 mmHg.

The patient received 150 mg of ranitidine 12 and 2 hours before the operation. After having arrived in the anaesthesia induction room in nearly sitting position, 30 ml of sodium citrate (0.3 molar) was administered orally. Standard monitoring with electrocardiogram and pulseoxymetry was established. In addition, paddle leads for external defibrillation were placed. Thereafter, an arterial line was placed into the left radial artery; a 4 lumen central venous catheter and a 9 gauge introducer were placed into the right jugular internal vein in local anaesthesia. A pulmonary artery (PA) catheter with atrial and ventricular pacing option (Baxter International, Illinois, USA) was inserted into the introducer and placed into the PA.

At this time the heart rate was 110/min with multiple polytopic ventricular extra-systolic (VES) beats. In the nearly sitting position, the arterial blood pressure was 110/70/90 mmHg and the PAP 75/30/50 mmHg with a pulmonary capillary wedge pressure of 30 mmHg. Pharmacologic therapy with a continuous infusion of dobutamine 3 µg/kg/min and milrinone 0.25/kg/min was started. Additionally, two 6 French introducer catheters were placed under local anaesthesia into the right femoral artery and right femoral vein. These catheters were inserted to facilitate quick percutaneous access to the vessels for emergency installation of ECMO for extracorporeal life support in case of acute cardiac decompensation.

After these preparations, the patient was transferred into the operating room and preparations for surgery were performed. After pre-oxygenation, anaesthesia was induced with 5 mg of rocuronium, 15 mg of etomidate, 35 mg of S ketamine and 100 mg of succinylcholine and a rapid sequence intubation was performed. Three minutes later, a healthy 2800 g child was delivered by classic longitudinal caesarean section (APGAR values of 9-10-10). Thereafter, anaesthesia was continued with a bolus of 30 mcg of sufentanil and 35 mg of rocuronium and a continuous infusion of remifentanil (2 µg/kg/min) and propofol (0.1 mg/kg/min) without using Target Controlled Infusion (TCI). Additionally, 1 g of paracetamol and 1 g of novamin-sulfate and 4 mg of ondansetron were infused. At the end of the procedure residual muscle relaxation was antagonized with 300 mg of sugammadex.

During the entire period of anesthesia, hemodynamics remained completely stable, the PAP decreased to 1/3rd systemic values of 50/30/20 mmHg while, however, polytopic VES were still observed. The completely awake and orientated patient was extubated on the surgical room. The patient was transferred to the intensive care unit (ICU) and she remained here for 5 days. Therapy with milrinone and dobutamine was continued during ICU stay. The patient was discharged uneventfully from hospital 14 days after surgery.

DISCUSSION

There are only a few case reports published regarding the management of patients with severe heart failure undergoing caesarean section. However, suggestions for performance of the anaesthetic procedure and medications used for induction and maintenance of anaesthesia vary largely (5, 6).

Additionally, reports about acute management strategies in case of refractory cardiac decompensation are limited to one publication in which ECMO for extracorporeal life support was used after acute decompensation in the early postpartum period (7).

In the current case we decided to use general anaesthesia, because we believe that it provides the best control of ventilation and hemodynamics. Additionally, the option to install an ECMO system further excluded the use of regional anaesthetic procedures for the concomitant need for systemic anticoagulation.

Patients with IVNC are at high risk for developing severe arrhythmias, which may be increased during surgical stress. However, life threatening arrhythmias may be resistant to defibrillation and antiarrhythmics. Possible side-effects of these drugs are bradycardia and atrio-ventricular (AV) blocks, which can be detrimental in a patient with borderline compensated cardiac function, or cardiac arrest. Therefore, it is necessary to provide an option for pacing and transcutaneous defibrillation, by means of pacing Swan-Ganz catheter and external paddles. Finally, we had to face the possibility of acute cardiac failure refractory to pharmacological treatment. Therefore, we decided that extracorporeal life support with an arterio-venous ECMO should be used as an early strategy for such an event. Having inserted the small introducer gates into the femoral vessels, it would have been easy to introduce the guidewire for the ECMO cannulas and insert these via Seldinger technique percutaneously. Thereby, ECMO would have been established quickly as an acute rescue manoeuvre and bridge to recovery or bridge to implantation of a left ventricular assist device and heart transplantation.

If refractory cardiac decompensation or cardiac arrest ensue, the ECMO strategy needs to be planned beforehand as the feasibility

of access to the groins and successful cannulation are extremely difficult due to the large abdomen and uterus which displace the anatomical landmarks; moreover, the surgical site might be at risk for bacterial contamination. On top of that if ECMO is instituted surgical hemostasis needs to be meticulous.

Fortunately, the current case was without severe complications. However, we think that our preparations and precautions would have facilitated the management of any severe decompensation in emergency with relative minimal invasiveness.

We believe these precautions are worth considering as a management strategy for teams which have to face a similarly complex case.

REFERENCES

1. Cantwell R, Clutton-Brock T, Cooper G, et al. Saving mothers' life: reviewing maternal death to make motherhood safer: 2006-2008. The eighth report of the confidential enquiries into maternal death in the United Kingdom. *BJOG* 2011; 118 (Suppl. 1): 1-203.
2. Ray P, Murphy GJ, Shutt LE. Recognition and management of maternal cardiac disease in pregnancy. *Br J Anesth* 2004; 93: 428-39.
3. Jenni R, Oechslin EN, van der Loo B. Isolated ventricular non-compaction of the myocardium in adults. *Heart* 2007; 93: 11-5.
4. Schwartzberg S, Sherez J, Wexler D, et al. Isolated ventricular non-compaction: an underdiagnosed cause of congestive heart failure. *IMAJ* 2009; 11: 426-9.
5. Zangrillo A, Landoni G, Pappalardo F, et al. Different anesthesiological management strategies in two high risk pregnant women with heart failure undergoing emergency cesarean section. *Minerva Anestsiol* 2005; 71: 227-36.
6. Pryn A, Bryden F, Reeve W, et al. Cardiomyopathy in pregnancy and caesarean section: four case reports. *Int J Obstet Anesth* 2007; 16: 68-73.
7. Jo YY, Park S, Choi YS. Extracorporeal membrane oxygenation in a patient with stress-induced cardiomyopathy after caesarean section. *Anaesth Intensive Care* 2011; 39: 954-7.

Cite this article as: Koster AA, Pappalardo F, Silvetti S, Schirmer U, Lueth JU, Dummler R, Emmerich M, Schmitt M, Kirchner G, Kececioglu D, Sandica E. Cesarean section in a patient with non-compaction cardiomyopathy managed with ECMO "stand-by". *Heart, Lung and Vessels*. 2013; 5(3): 183-186.

Source of Support: Nil. **Disclosures:** None declared.