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Anesthetic management of bidirectional cavopulmonary shunt in a patient with pulmonary atresia with intact ventricular septum associated with sinusoidal communications

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Abstract
Pulmonary atresia with intact ventricular septum (PAIVS) is sometimes associated with coronary artery anomalies, including right ventricle (RV)-to-coronary artery fistulas (sinusoidal communications), coronary artery stenoses, and coronary artery occlusions. In some cases, the coronary circulation depends entirely or partly on the desaturated systemic venous blood supply from the RV. Under these circumstances, decompression of the RV can result in fatal myocardial ischemia. A 6-month-old boy, diagnosed with PAIVS associated with sinusoidal communications, underwent a bidirectional cavopulmonary shunt procedure under venoarterial cardiopulmonary bypass (CPB). During CPB, to prevent RV decompression, we maintained right atrial pressure above 5 mmHg and used a pump perfusion rate of 30%–40% of the calculated value based on body surface area. Although electrocardiography showed slight ST depression and bradycardia, myocardial contractility after weaning from CPB was adequate to maintain the circulation with the administration of dobutamine and atrial pacing. In patients with PAIVS and RV-dependent coronary circulation, it is important to maintain coronary artery perfusion throughout the period of anesthesia.

Key words Pulmonary atresia with intact ventricular septum · Sinusoidal communication · Bidirectional cavopulmonary shunt · Anesthesia

Introduction

Sinusoidal communications (right ventricle [RV]-to-coronary artery fistulas) have been reported in 31%–69% of patients with pulmonary atresia with intact ventricular septum (PAIVS) [1–3]. The dominant sinusoidal blood flow is usually at high pressure from the RV to the coronary arteries, and the coronary circulation depends wholly or partly on the desaturated systemic venous blood supply from the RV. Under these circumstances, decompression of the RV may result in fatal myocardial ischemia. Thus, coronary artery pathology is one of the determinants of the surgical strategy in patients with PAIVS. To our knowledge, the anesthetic management of PAIVS with sinusoidal communications has not been reported. Herein, we describe the anesthetic management of a 6-month-old boy, diagnosed with PAIVS with sinusoidal communications, undergoing a bidirectional cavopulmonary shunt procedure.

Case report

A 6-month-old boy (3.1 kg) with a prenatal diagnosis of PAIVS was delivered normally (39 weeks + 5 days; birth weight, 2316 g). He underwent balloon atrial septostomy at 3 days of life, placement of a right Blalock-Taussig shunt at 67 days, and patent ductus arteriosus ligation at 87 days. At 6 months, because of poor body weight gain, he was scheduled for a bidirectional cavopulmonary shunt procedure. Preoperative angiography showed that the RV was severely hypoplastic, that the RV end-diastolic volume was 28.6% of normal, and that there were multiple sinusoidal communications between the RV and the right coronary artery (RCA), the left circumflex artery (LCA), and the left anterior descending artery (LAD; Fig. 1). Flow to the RCA, LCA, and LAD was from the RV through the fistulas, posing a risk of RV-dependent coronary circulation.

In the operating room, the patient was prepared for blood pressure (noninvasive), oxygen saturation, and electrocardiographic monitoring. Anesthesia was induced with intravenous midazolam (0.5 mg), pancuronium (0.5 mg), and fentanyl (10 μg). After tracheal intubation, an arterial catheter and central venous

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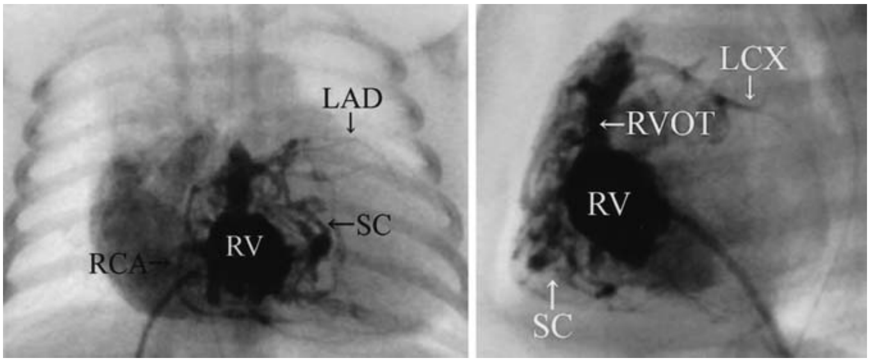

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Fig. 1A,B. Right ventriculogram, showing severe hypoplasia of the right ventricle (*RV*), and sinusoidal communications (*SC*) to the coronary artery. **A** anterior-posterior view; **B** lateral view. *RCA*, right coronary artery; *LAD*, left anterior descending artery; *LCX*, left circumflex artery; *RVOT*, right ventricular outflow tract

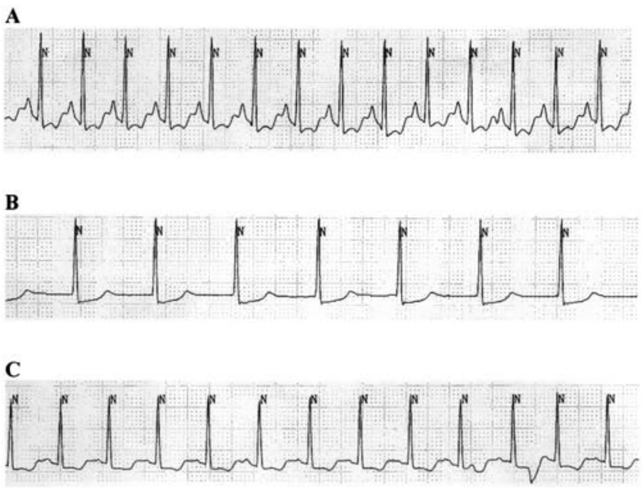

Fig. 2A–C. Electrocardiograms obtained **A** before the initiation of cardiopulmonary bypass, **B** 10 min after the initiation of cardiopulmonary bypass, and **C** after weaning from cardiopulmonary bypass. *10 mm*, 1 mV

catheter were placed in the left radial artery and left internal juglar vein, respectively. Anesthesia was maintained with intravenous midazolam (2 mg·kg^{-1}), fentanyl (70 μg·kg^{-1}), and pancuronium. After median sternotomy, an arterial perfusion cannula was inserted into the ascending aorta, and a venous cannula was inserted into the superior vena cava. To prevent RV decompression, a pressure line was inserted into the right atrium, and right atrial pressure was monitored. After partial cardiopulmonary bypass (CPB) was established, right atrial pressure was maintained above 5 mmHg, and we used a pump perfusion rate of 30%–40% of the calculated value based on body surface area; we continued mechanical ventilation with a tidal volume of approximately 25 ml at a rate of 10 breaths·min^{-1}. Although no ST change was seen on the electrocardiogram before the establishment of CPB (Fig. 2A), slight ST depression and bradycardia (80 beats·min^{-1}) were seen 10 mins after the institution of CPB (Fig. 2B). Atrial pacing, at a rate of 120 beats·min^{-1} was applied, but the ST depression continued throughout the period of CPB. With the administration of dobutamine (6 μg·kg^{-1}·min^{-1}), nitroglycerin (1 μg·kg^{-1}·min^{-1}), and nitric oxide (10 ppm), weaning from CPB was smooth, and myocardial contractility was adequate to maintain the circulation. Although slight ST depression continued (Fig. 2C) until the end of surgery, the anesthetic course was uneventful. On the first postoperative day, the electrocardiogram showed no ST change, and the patient was extubated.

Discussion

Sinusoidal communications (RV-to-coronary artery fistulas) are reported in 31%–69% of patients with PAIVS [1–3]. In patients with RV-to-coronary fistulas with coronary stenoses or coronary occlusion proximal to the fistulas, a large part of the coronary blood supply depends on the RV. This RV-dependent coronary circulation (RVDCC) is a major risk factor limiting subsequent surgical management, because surgery-related RV decompression can lead to myocardial ischemia. RVDCC was previously reported in 5%–34% of patients with PAIVS [2–5] and can contribute to significant ischemia [6–8]. Although RV decompression in patients with PAIVS is necessary for biventricular repair, RV decompression is generally contraindicated in the presence of RVDCC. The usual management strategy in patients with RVDCC is a palliative systemic-to-pulmonary arterial shunt procedure and subsequent univentricular repair [2,5]. Therefore, it is essential to recognize any coronary artery pathology preoperatively and to prevent fatal myocardial ischemia intraoperatively. In our patient, we could not evaluate the coronary artery proximal to the fistulas because we were limited in regard to the total amount of contrast medium we could administer because of the risk of renal dysfunction. However, we considered the coronary circulation to be RV-dependent due to the existence of multiple fistulas, and myocardial ischemia (ST depression) was actually recognized during anesthesia.

CPB unloads and decompresses the RV; therefore, attention must be paid to the coronary circulation during CPB. In the present patient, it was technically difficult to monitor RV pressure, due to very small RV cavity, so we monitored right atrial pressure instead of RV pressure. We maintained right atrial pressure above 5 mmHg. However, considering the ST depression during the surgical procedure, it is possible that it would have been better to set up right atrial pressure higher than 5 mmHg. On the other hand, we used venoarterial CPB. In a previous report, venovenous bypass was shown to be useful to prevent myocardial ischemia during a right heart bypass operation in patients with PAIVS and RVDCC [9]. It is possible that venovenous bypass, draining systemic venous blood through cannulas inserted into the superior vena cava and inferior vena cava and perfusing the right atrium with oxygenated blood, can supply more oxygen to the coronary artery system than venoarterial bypass can supply. Another surgical option is an off-pump technique. However, there are questions about the safety of the nonbypass approach.

In conclusion, in patients with PAIVS and RV-dependent coronary circulation, RV decompression can lead to myocardial ischemia. Therefore, it is important to maintain coronary artery perfusion throughout the period of anesthesia, including the period of CPB.

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