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Anesthetic management for severe aortic regurgitation in an infant repaired by Ross procedure

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Abstract
We report the anesthetic management of a 7-month-old male infant with severe aortic regurgitation (AR) scheduled for the Ross procedure. To the best of our knowledge, this is the first report from the viewpoint of anesthetic management for the Ross procedure performed in an infant. He had been suffering from severe AR that occurred suddenly when he was 5 months old. The cause of the AR was considered to be spontaneous rupture of a fenestrated aortic valve, owing to congenital tissue defect in part of the aortic valvular leaflet. The Ross procedure was scheduled to be performed under general anesthesia using deep hypothermic cardiopulmonary bypass (CPB). Continuous infusion of nitroglycerin was started during CPB and continued after CPB to dilate the newly implanted coronary arteries for the prevention of myocardial ischemia and to decrease afterload and pulmonary vascular resistance. Weaning from CPB was performed without difficulty, but after the prolonged CPB he had a bleeding tendency that needed transfusion and a hemostatic drug. Monitoring with transesophageal echocardiography was very useful for evaluating myocardial ischemia, and for assessing the procedure and the completion of surgical repair. His postoperative course was uneventful and he was discharged on the 25th postoperative day.

Key words Anesthetic management · Ross procedure · Aortic regurgitation · Infant

Introduction

The Ross procedure, which consists of the replacement of a diseased aortic valve with a pulmonary autograft [1], is an alternative for aortic valve replacement in children. Although there are a few case reports about the Ross procedure performed in infants [2–4], we know of no reports from the viewpoint of anesthetic management. We report a case of anesthetic management for the Ross procedure scheduled for a 7-month-old infant.

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Case report

The male patient was born at 37 weeks without fetal distress and his body weight was 2710 g. No serious abnormalities were pointed out. At 5 months, he was admitted to hospital because of a fever of 38.0°C owing to pharyngitis, and cardiac murmur was pointed out. Later, his illness was diagnosed as severe AR by transthoracic echocardiography (TTE). Blood cultures were taken repeatedly for the purpose of excluding infectious endocarditis. However, all of the cultures were negative for the bacteria. Although he received medical treatment for 1 month, left ventricular volume overload owing to AR could not be improved. He was therefore referred to our university hospital for surgical intervention.

On admission to our hospital at 7 months (weighing 7680 g), his chest roentgenogram showed cardiomegaly, in which the cardiothoracic ratio was 60% (Fig. 1). Blood pressure (BP) was 80/37 mmHg, and heart rate (HR) was 128 beats·min^{-1}. Respiratory rate was 40 breaths·min^{-1}, and oxygen saturation of peripheral artery (S_{PO_2}) was 98% under room air. TTE showed severe AR and moderate mitral regurgitation (MR). Left ventricular end-diastolic dimension was 35.9 mm (140.6% of the normal value) and the MR was thought to be due to annular dilation. Fractional shortening was 29%. Accordingly, the Ross procedure was scheduled, using deep hypothermic (23°C, rectal temperature) cardiopulmonary bypass (CPB).

General anesthesia was induced with intravenous injection of fentanyl 10 μg, midazolam 1 mg, vecuronium 2 mg, and atropine 0.08 mg and maintained with oxygen 2 l·min^{-1}, air 2 l·min^{-1}, sevoflurane 1%–3%, remifentanil 0.2 μg·kg^{-1}·min^{-1}, and bolus injection of fentanyl,

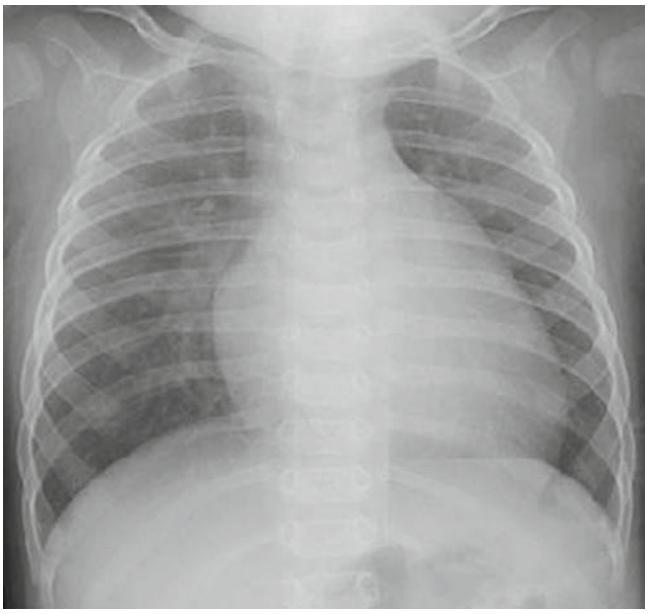

Fig. 1. On admission at 7 months, the patient's chest roentgenogram showed cardiomegaly, in which the cardiothoracic ratio was 60%

vecuronium, and midazolam. Tracheal intubation (internal diameter, 4.0 mm) was performed, and a central venous catheter, a femoral arterial catheter, and a probe for biplanar transesophageal echocardiography (TEE; outer diameter of the shaft, 6.8 mm; Aloka, Tokyo, Japan) were inserted. TEE showed severe AR and left ventricular dilatation, and mild MR (Fig. 2A–C). After the induction of anesthesia, arterial blood pressure (ABP) was 55/15 mmHg (diastolic hypotension due to AR); HR, 115 beats·min^{-1}; and central venous pressure (CVP), 5 mmHg. Arterial blood gas analysis (ABGA) showed pH, 7.43; Pa_{O_2}, 273 mmHg (fraction of inspired oxygen [$F_{I_{O_2}}$] = 0.6); Pa_{CO_2}, 38 mmHg; base excess (BE), +0.9 mEq·l^{-1}; and hematocrit (Ht,) 29.4%. The baseline level of activated clotting time (ACT) was 139 s.

The operation was started with median sternotomy. After the intravenous injection of heparin 2300 units, CPB was instituted using aortic and bicaval cannulation. Sufficient antegrade cardioplegia was not attained because of AR. TEE showed left ventricular expansion with regurgitated cardioplegia solution. Then, additional selective antegrade cardioplegia solution was administered through a 6-Fr (outer diameter, 2.0 mm) enteral feeding tube, as there are no ready-to-use cannulae for selective cardioplegia for infants. The aortic valve had three cusps, but the left coronary cusp had a large defect with thin tissue. The defect was thought to be the result of spontaneous rupture of the fibrous strand of the fenestrated aortic valve, owing to congenital tissue defect in part of the aortic valvular leaflet. The aortic valve was replaced with the infant's normal anatomical pulmonary valve. The left and right coronary arteries were reimplanted to the autograft. The right ventricular outflow tract was reconstructed with a 16-mm bulging sinus, expanded polytetrafluoroethylene (ePTFE) valved conduit. From the start of CPB, chlorpromazine (CPZ) 0.1 mg·kg^{-1}·h^{-1} was infused continuously to add to the sedative effect and to decrease afterload. Continuous infusion of nitroglycerin (NTG) 1 μg·kg^{-1}·min^{-1} was also started to decrease afterload and pulmonary vascular resistance, and to dilate the newly implanted coronary arteries.

Aortic cross-clamp time was 160 min and CPB time was 290 min. Antegrade cardioplegia solution was infused twice and selective cardioplegia was performed three times in total (395 ml). After his rectal temperature had returned to 37°C, modified ultrafiltration was done and he was weaned from CPB with CPZ 0.1 mg·kg^{-1}·h^{-1}, NTG 1 μg·kg^{-1}·min^{-1}, dopamine (DOA) 7 μg·kg^{-1}·min^{-1}, and dobutamine (DOB) 7 μg·kg^{-1}·min^{-1}. ABP was 60/35 mmHg; CVP, 11 mmHg; left atrial pressure (LAP), 6 mmHg; and HR, 150 beats·min^{-1} with normal sinus rhythm when weaning was completed. An electrocardiogram (ECG) did not show any ST-T changes. TEE showed trivial aortic (pulmonary autograft) regurgitation (Fig. 3A); it showed no right ventricular outflow tract obstruction (Fig. 3B) and no regional wall motion abnormality. We could not assess the function of the new ePTFE pulmonary valve by TEE. In order not to generate ePTFE pulmonary valvular regurgitation, we avoided hypoxemia, hypercapnea, and acidosis, and we raised the dose of NTG (to 5 μg·kg^{-1}·min^{-1}) to decrease pulmonary vascular resistance. We were able to taper DOA and DOB as TEE showed good left ventricular contraction and his urine output was satisfactory. He had a bleeding tendency in the surgical field after the CPB, although the ACT was 154 s after the administration of protamine 23 mg. Transfusion of 470 ml (packed red cells, 230 ml; fresh frozen plasma, 140 ml; platelets, 100 ml) and infusion of tranexamic acid 500 mg were performed after the CPB, and then the bleeding tendency was attenuated.

The operation time was 497 min and blood loss was 1131 g. Urine output was 218 ml, and in-out balance during anesthesia was –379 ml. When he was brought to the intensive care unit, his ABP was 70/40 mmHg; CVP, 10 mmHg; LAP, 4 mmHg; and HR, 125 beats·min^{-1} with normal sinus rhythm by intravenous injection of CPZ 0.1 mg·kg^{-1}·h^{-1}, NTG 5 μg·kg^{-1}·min^{-1}, DOA 1 μg·kg^{-1}·min^{-1}, and DOB 1 μg·kg^{-1}·min^{-1}. ABGA showed pH 7.34; Pa_{O_2}, 276 mmHg ($F_{I_{O_2}}$ = 0.6); Pa_{CO_2}, 41 mmHg; BE, –3.0 mEq·l^{-1}; and Ht, 30.1%.

His trachea was extubated 2 days after the surgery and he was discharged from the intensive care unit 5 days after the surgery. His postoperative course was almost uneventful. Postoperative TTE showed mild AR

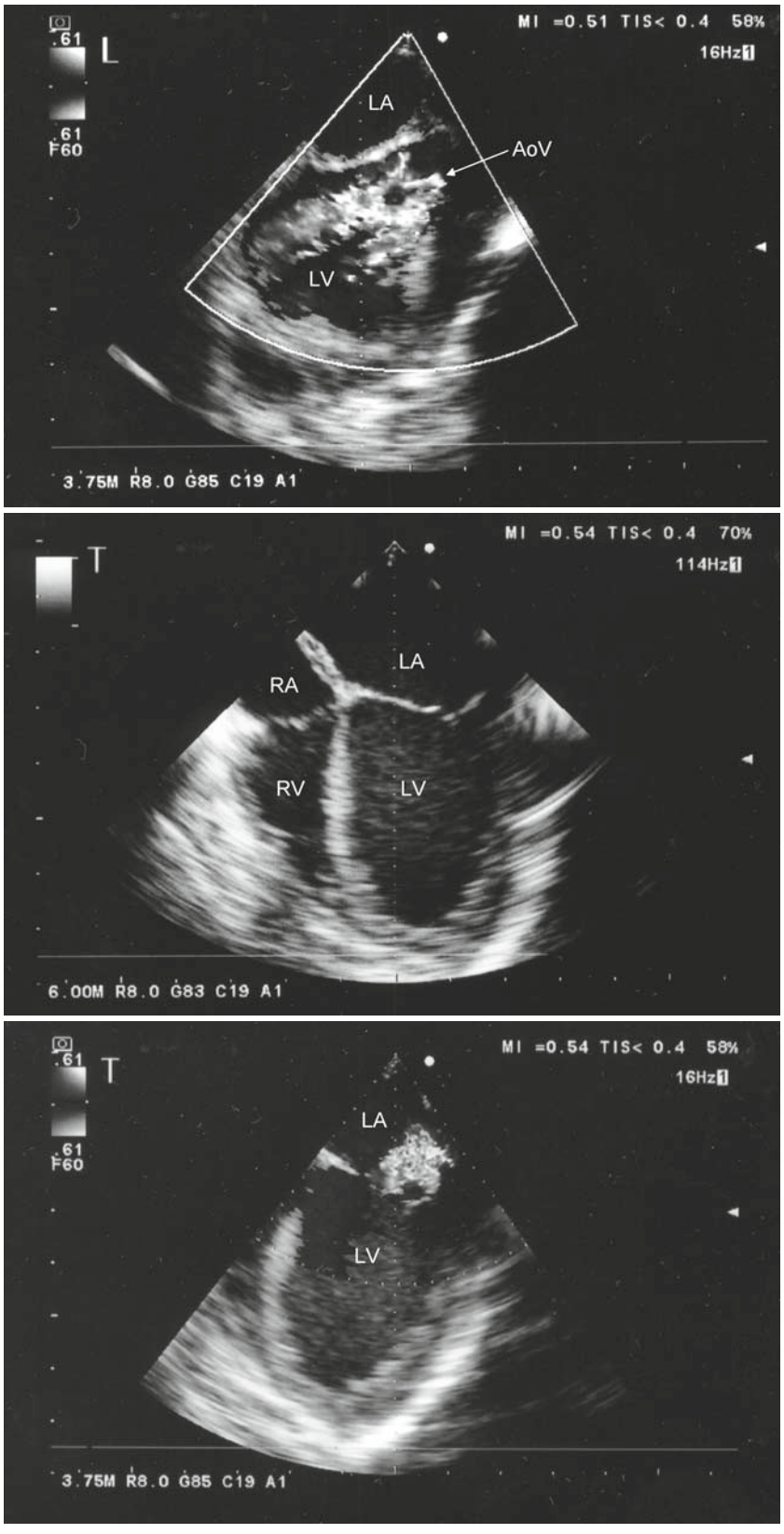

Fig. 2A–C. Preoperative transesophageal echocardiography (TEE). **A** Severe aortic regurgitation during diastole. **B** Left ventricular dilatation. **C** Mild mitral regurgitation during systole. *AoV*, Aortic valve; *LA*, left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle

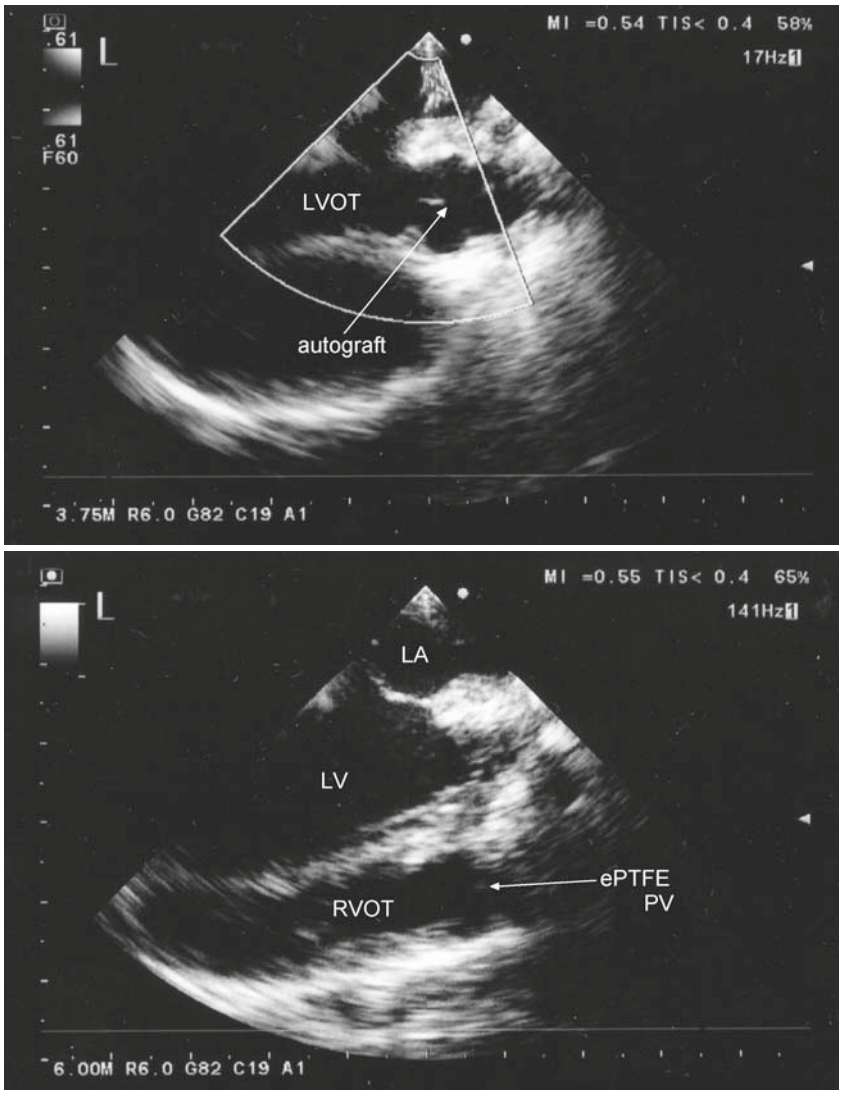

Fig. 3A,B. Transesophageal echocardiography (TEE) after the Ross procedure. **A** Trivial autograft regurgitation during diastole; **B** no right ventricular outflow tract (*RVOT*) obstruction. *LVOT*, Left ventricular outflow tract; *ePTFE PV*, expanded polytetrafluoroethylene pulmonary valve

and mild pulmonary regurgitation, and mild MR. He was medicated with an angiotensin-converting enzyme inhibitor and diuretics, and discharged from our hospital 25 days after the surgery.

Discussion

The patient in our report had been suffering from severe AR since he had had pharyngitis and a fever at 5 months after birth. Intraoperative findings showed that the left coronary cusp had a large defect. Aortic valve fenestration, owing to defect of the valvular tissue, is a common anatomical finding that is present more than 70% of adult intact hearts [5,6]. It is usually not associated with clinical symptoms. However, in rare settings, symptomatic AR develops as a consequence of rupture of the connecting tissue bands and enlargement of the defect, which is associated with hypertension, high fever, severe anemia, and other hyperkinetic circulatory states [5]. We supposed, therefore, that our patient had had a congenitally fenestrated aortic valve and the hyperdynamic state associated with fever had caused sudden rupture of the connecting tissue band of the aortic valve, resulting in severe AR, when he was 5 months old. Although we found some case reports of symptomatic AR due to rupture of a fenestrated valve [7–12], all of them were about adults. Accordingly, we considered that this was an extremely rare infant case.

In adult cases of aortic valve fenestration, surgical intervention for the AR includes repair of the valve [7,11] or aortic valve replacement [12]. The Ross procedure is the alternative for aortic valve replacement for young individuals. Replacement with a prosthetic valve is generally avoided due to the small size of the annulus, the necessity for a second surgery after the child grows, and the requirement for anticoagulation. The Ross procedure does not require anticoagulation and the autograft has excellent hemodynamics and potential for future growth as the child grows, and the procedure has

shown excellent mid-term results [13,14]. There are some case reports of the Ross procedure performed in infants for congenital aortic stenosis [2,3] and myxomatous degeneration of the aortic valve [4], but we could not find any reports from the viewpoint of anesthetic management.

We consider that one of the most serious anesthetic problems with the Ross procedure in infants is myocardial ischemia, for two reasons. First, the procedure involves the reimplantation of coronary arteries. Secondly, CPB and aortic cross-clamp time are generally prolonged due to the technical difficulties of the procedure, and it is accepted that the duration of CPB and aortic cross-clamp time correlates with myocardial damage [15]. Moreover, in infants, cardioplegia is not easy because the diameters of the coronary orifices are so small that there are no ready-to-use cannulae for selective cardioplegia. What was even worse in our patient was that he could not receive effective antegrade cardioplegia because of the severe AR. Therefore, we made efforts to prevent and monitor myocardial ischemia during the anesthetic management. To prevent myocardial ischemia, we used deep hypothermic CPB and administered nitroglycerin intravenously to decrease the afterload and to dilate the newly implanted coronary arteries during and after CPB. To monitor myocardial ischemia, we used TEE and ECG.

The monitoring with TEE is considered to be very important in the Ross procedure, especially in infants. With this modality, we can not only detect myocardial ischemia but we can also assess the presence of neoaortic valve insufficiency after the operation [16, 17]. Moreover, in our patient, we were able to avoid severe myocardial damage by detecting ineffective antegrade cardioplegia. On the other hand, the TEE probe is difficult to insert in infants and several complications may occur, including injury of the digestive tract and airway trouble. However, we were able to advance the probe to the stomach without any resistance and there was no ventilatory trouble or malpositioning of the tracheal tube.

Another serious anesthetic problem with the Ross procedure is a bleeding tendency, owing to prolonged hypothermic CPB. In our patient, deep hypothermic (23°C) CPB was performed for the purpose of protecting the myocardium and other organs against ischemia. The CPB time extended to 290 min (including deep hypothermic CPB 120 min) and blood loss after CPB reached 1131 g. Although he was weaned from CPB after his rectal temperature returned to 37°C, we had to transfuse fresh frozen plasma and platelets and infuse tranexamic acid to alleviate the bleeding tendency.

In conclusion, we report a case of anesthetic management for the Ross procedure for severe AR in an infant. Important anesthetic problems are protecting the myocardium against ischemia and attenuating the bleeding tendency. Monitoring with TEE was very useful for evaluating myocardial ischemia, and for assessing the procedure and the completion of surgical repair.

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