J Anesth (2002) 16:169–172

Journal of
Anesthesia

Perioperative management of Fontan operation for two patients with arrhythmogenic right ventricular dysplasia

Kiyoshi Morita[1], Mamoru Takeuchi[1], Katsunori Oe[1], Tatsuo Iwasaki[1], Naoyuki Taga[1], Masahisa Hirakawa[1], and Shunji Sano[2]

[1] Department of Anesthesiology, Okayama University School of Medicine, 2-5-1 Shikata-cho, Okayama 700-8558, Japan
[2] Department of Cardiovascular Surgery, Okayama University School of Medicine, Okayama, Okayama 700-8558, Japan

Key words Arrhythmogenic right ventricular dysplasia · Fontan operation · Pulmonary vascular resistance

Introduction

Arrhythmogenic right ventricular dysplasia (ARVD) is an idiopathic progressive disease characterized by replacement of the right ventricular cardiac muscle by adipose/fibrous tissues and the presence of frequent ventricular tachycardia. Treatment aims at prevention of persistent ventricular tachycardia and consists of administration of antiarrhythmic agents and catheter ablation, although no standard treatment has yet been established. We have successfully treated two patients with refractory ARVD by the Fontan operation. In this report, we describe the anesthetic and perioperative management in these two cases.

Case reports

Case 1

The patient was a 27-year-old man weighing 70 kg, who was incidentally diagnosed at the age of 16 years with a right bundle branch block on a routine health checkup. The patient was healthy at that time and had no symptoms. In December 1995, the patient was transferred to a local hospital after an attack of syncope. Examination showed frequent ventricular tachycardia (VT), which was later confirmed by clinical and laboratory evidence to be associated with ARVD. The condition was successfully controlled at that stage with antiarrhythmic drugs. However, in February 1996, VT became resistant to drug therapy and failed to respond to catheter ablation. In June 1996, the patient was admitted to the intensive care unit (ICU) of Okayama University Medical School for further management. On admission, frequent VT had caused marked right heart failure, and the central venous pressure (CVP) was approximately 20 mmHg. The patient was immediately treated with flecainide acetate, cibenzoline succinate, and metoprolol tartrate. This was followed by transjugular ventricular pacing. However, the treatment was ineffective in reducing the frequency of VT. Subsequently, VT worsened, as manifested by the appearance of five types of VT of different origin and the persistent presence of VT 9 days after admission to the ICU. The patient developed marked right heart failure on day 10, which was treated by mechanical ventilation following intratracheal intubation. However, heart failure did not improve, and there was no increase in urine volume. Deterioration of right and left heart function was probably due to persistent VT and administration of antiarrhythmic agents. Therefore, 13 days after admission, intraaortic balloon pumping (IABP) was initiated. On the next day, percutaneous cardiopulmonary support (PCPS) was implemented. PCPS produced a rapid increase in urine volume and improved respiration but did not result in amelioration of VT. Accordingly, 16 days after admission, an emergency Fontan operation was performed. The patient was transferred to the operating theater, while IABP and PCPS were being applied. Anesthesia was mainly maintained with 43 $\mu g \cdot kg^{-1}$ fentanyl. When the large right atrium and right ventricle were resected under cardiopulmonary bypass, VT stopped and sinus rhythm appeared. A total cavopulmonary connection (TCPC)-type Fontan operation was performed (Fig. 1). The duration of cardiopulmonary bypass was 3 h 19 min. Weaning from cardiopulmonary bypass was readily achieved by inducing mild hyperventilation and lowering arterial PCO_2 to approximately 35 mmHg. At the same time, dopamine, dobutamine

Address correspondence to: K. Morita
Received: December 27, 1999 / Accepted: October 28, 2001

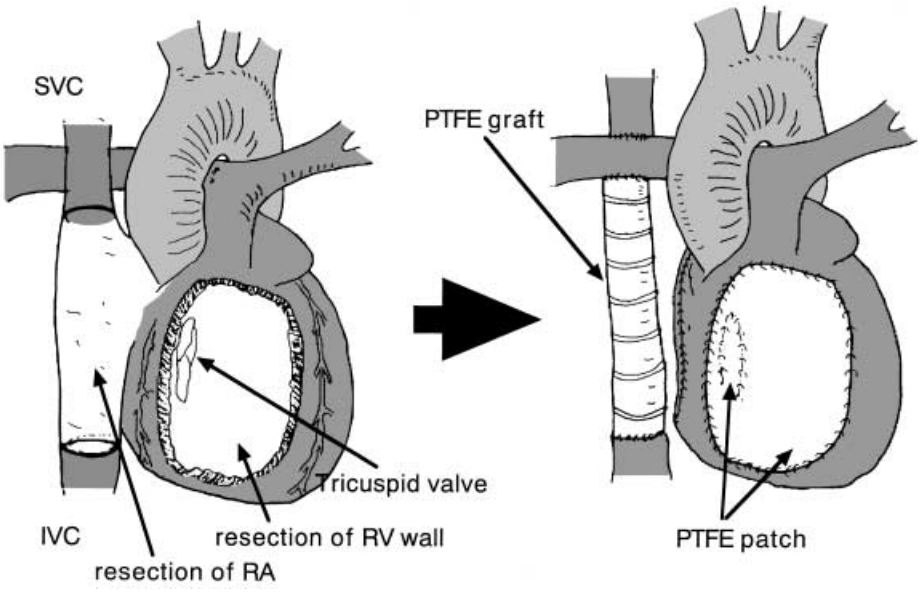

Fig. 1. Surgical procedure for total cavopulmonary connection-type Fontan operation. *IVC*, Inferior vena cava; *PTFE*, polytetrafluoroethylene; *RA*, right atrium; *RV*, right ventricle; *SVC*, superior vena cava

($5\,\mu g \cdot kg^{-1} \cdot min$, each), nitroglycerin ($1\,\mu g \cdot kg^{-1} \cdot min^{-1}$), and prostaglandin E_1 (PGE_1, $0.1\,\mu g \cdot kg^{-1} \cdot min^{-1}$) were also administered. Postoperatively, CVP increased slightly to approximately 25 mmHg, and left atrial pressure (LAP) was 7 mmHg. Weaning was achieved without IABP and PCPS. The total duration of surgery was 10 h 55 min, and the duration of anesthesia was 13 h 50 min. The volume of intraoperative blood loss was approximately 1800 ml. The urine volume was 2500 ml. Following the return to ICU, massive vasodilatory therapy, the same as that administered in the operating theater, resulted in stable respiratory and circulatory kinetics. About 19 h after surgery, the patient was extubated. This was associated with a gradual fall in CVP to 12 mmHg immediately after extubation. The follow-up period was uneventful and no VT was noted. The patient was discharged from the ICU 10 days after surgery and from the hospital 76 days after surgery.

Case 2

The patient was a 44-year-old woman weighing 52 kg. At the age of 8 years, the patient underwent repair surgery for atrial septal defect (ASD). At the age of 36 years, tricuspid annuloplasty (TAP) was performed to treat tricuspid regurgitation (TR). Two years later, she developed VT attacks, which were controlled by drug therapy. In October 1995, a ventricle-ventricle-inhibited (VVI) pacemaker was implanted, which controlled further VT attacks. However, in late August 1997, frequent VT attacks developed again. She was admitted to our hospital and treated with direct current (DC) conversion followed by drug therapy. However, in October of the same year, right heart failure was exacerbated, and echocardiography showed marked enlargement of the right heart system and grade-3 tricuspid regurgitation (TR). Therefore, we considered that the Fontan operation was indicated based on our experience in case 1. In January, 1998, elective TCPC-type fenestrated Fontan operation was performed. Anesthesia was induced with $10\,\mu g \cdot kg^{-1}$ fentanyl and maintained with total $20\,\mu g \cdot kg^{-1}$ of fentanyl and below 1.5% isoflurane. The duration of cardiac arrest was 21 min, and the duration of cardiopulmonary bypass was 3 h 55 min. Weaning from cardiopulmonary bypass was readily achieved by administering dopamine ($5\,\mu g \cdot kg^{-1} \cdot min^{-1}$), dobutamine ($5\,\mu g \cdot kg^{-1} \cdot min^{-1}$) and nitroglycerin ($1.6\,\mu g \cdot kg^{-1} \cdot min^{-1}$). After administration of protamine sulfate, $0.05\,\mu g \cdot kg^{-1} \cdot min^{-1}$ of PGE_1 was added to reduce pulmonary vascular resistance. At the end of surgery, CVP was 17 mmHg and LAP was 12 mmHg. After confirmation with a thoracic radiogram, muscle paralysis was reversed and the patient was extubated in the operating room and then moved to the ICU. The total duration of surgery was 9 h 20 min, and the duration of anesthesia was 11 h 10 min. The volume of intraoperative blood loss was 1200 ml, and the volume of collected urine was 1150 ml. The immediate postoperative period in the ICU was uneventful, and the patient was transferred 13 days after surgery to the general ward. The postoperative course was smooth, and the patient was discharged from the hospital 71 days after surgery.

Discussion

ARVD was first reported by Fontaine et al. in 1977. ARVD is an idiopathic disease characterized by local or

diffuse replacement of the right ventricular cardiac muscle by adipose/fibrous tissues and life-threatening ventricular tachycardia. ARVD is more common in adult men [1] (mean age, 39 years; male : female ratio, 2.7 : 1). Antiarrhythmic agents [2] and catheter ablation have been used for the treatment of persistent ventricular tachycardia, although no standard therapy has been established. Several studies have reported that surgical techniques, such as use of an automatic implantable cardioverter-defibrillator (AICD), ventricular incision, and cryopexy were effective [3–5].

In the present report, two patients underwent a Fontan operation for ARVD. Patient 1 developed marked right heart failure related to persistent ventricular tachycardia, as well as left heart failure mainly caused by administration of excessive doses of antiarrhythmic agents. As a life-saving measure, an emergency Fontan operation was performed, although the procedure had not been planned previously as part of the management of the case. In case 2, an elective Fontan operation was performed, because right heart failure was relatively severe, although left heart function was maintained. The decision to perform surgery in the second patient was based on our experience in case 1.

With regard to the management during and after Fontan operation in patients with ARVD, the most important step is perhaps to reduce pulmonary vascular resistance, as conducted during and after the Fontan operation for congenital heart disease. At our institution, the Fontan operation is performed in about 15 patients with congenital heart disease every year. Our team has gained good experience in intraoperative and postoperative management, which was advantageous for the two patients reported in this study. In patients with congenital heart disease, the indication criteria for the Fontan operation include low pulmonary vascular resistance, low pulmonary arterial pressure, favorable left heart function, sinus rhythm, atrioventricular valve without regurgitation, and pulmonary artery with sufficient growth [6]. With regard to preoperative status in case 1, the pulmonary condition, which was considered the most important, was poor, and PCPS was required. Furthermore, left heart failure related to the side effects of antiarrhythmic agents and compression by RV cardiomyopathy-like large right heart system [7] was detected. Persistent refractory VT was present, and a poor general condition was observed. Despite these poor conditions, weaning from cardiopulmonary bypass was successfully achieved without returning to assisted circulation, which had been required preoperatively. This may have been associated with the following factors: surgery resulted in recovery of sinus rhythm; compression by the large right ventricle was relieved, and there was a lack of abnormal interventricular septal wall movement; a decrease in blood concentrations of the antiarrhythmic agent improved heart function; and the patient was acclimatized to high CVP caused by right heart failure before surgery. In addition, the preoperative use of IABP and PCPS achieved diuresis and, to some extent, improved ventilation. Pulmonary vascular resistance was reduced as much as possible by massive vasodilatory therapy and hyperventilation after weaning from cardiopulmonary bypass. These management procedures may have contributed to saving the patient's life. Early extubation should be performed soon after Fontan operation to reduce pulmonary vascular resistance. However, in case 1, extubation was impossible on the day of surgery, because preoperative bilateral heart failure was too severe. Instead, extubation was performed 19 h after surgery.

With regard to the preoperative status in case 2, right heart failure was related to frequent VT and marked TR. However, left heart function was maintained, and there was no pulmonary circulatory disorder. The Fontan operation was indicated while the patient was in a relatively good condition. Therefore, it was possible to markedly reduce pulmonary vascular resistance by massive vasodilatory therapy after weaning from cardiopulmonary bypass, mild hyperventilation, and extubation in the operating room. This may have contributed to the excellent postoperative recovery.

At present, the two patients are evaluated as grade 1 according to the New York Heart Association (NYHA) classification criteria for cardiac patients, and they have no cyanosis or arrhythmia. In addition, they are not being treated with any medications other than anticoagulants. Therefore, the Fontan operation is a very effective surgical treatment that should be considered for the treatment of refractory ARVD.

Perioperative management is similar to that provided for patients with congenital heart disease treated by the Fontan operation, but more strict management should be done when preoperative left heart failure is severe. The important step is the reduction of pulmonary vascular resistance as much as possible. Furthermore, massive vasodilatory therapy, hyperventilation, and early extubation are essential.

Summary

Our report indicates that the Fontan operation is very effective for ARVD and should be considered in the treatment of such patients, particularly those with refractory tachycardia. Concerning postoperative management, massive vasodilatory therapy and early extubation should be used to reduce pulmonary vascular resistance as much as possible, as in the Fontan operation in patients with congenital heart disease.

References

1. Marcus F, Fontaine G, Guiraudon G, Frank R, Laurenceau J, Malergue C, Grosgogeat Y (1982) Right ventricular dysplasia: a report of 24 adult cases. Circulation 65:384–398
2. Lemery R, Brugada P, Janssen J, Cheriex E, Dugernier T, Wellens J (1989) Nonischemic sustained ventricular tachycardia: clinical outcome in 12 patients with arrhythmogenic right ventricular dysplasia. J Am Coll Cardiol 14:96–105
3. Guiraudon G, Fontaine G, Frank R, Leandri R, Berra J, Cabol C (1981) Surgical treatment of ventricular tachycardia guided by ventricular mapping in 23 patients without coronary artery disease. Ann Thorac Surg 32:439–450
4. Cox J, Bardy G, Damiano R, German L, Feder J, Kisslo J, Packer D, Gallagher J (1985) Right ventricular isolation procedures for nonischemic ventricular tachycardia. J Thorac Cardiovasc Surg 90:212–224
5. Isobe F, Fujita T, Kaku K, Sudo Y, Ohe T, Shimomura K (1986) Surgical treatment of arrhythmogenic right ventricular dysplasia with multiple refractory ventricular tachycardias. Jpn Thorac Surg 34:111–116.
6. Shemin R, Merrill W, Pfeifer J, Conkle D, Morrow A, Bethesda M (1979) Evaluation of right atrial-pulmonary artery conduits for tricuspid atresia. J Thorac Cardiovasc Surg 77:685–690
7. Fontaine G, Fontaliran F, Frank R (1998) Arrhythmogenic right ventricular cardiomyopathies: clinical forms and main differential diagnosis. Circulation 97:1532–1535