Successful Blade Atrial Septostomy in a Patient With Severe Primary Pulmonary Hypertension

— A Case Report —

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Blade atrial septostomy (BAS) for pulmonary hypertension has increased long-term survival and is an effective and palliative preliminary to heart and/or lung transplantation. We treated an 18-year-old woman with severe pulmonary primary hypertension whose symptoms had worsened as a result of low cardiac output. The patient's right ventricular pressure was 150/23 mmHg, cardiac index (CI) 1.0 L/min per m², and she showed signs and symptoms of severe primary pulmonary hypertension. We performed BAS successfully, paying particular attention to the following points. To maintain pulmonary blood flow after creating an atrial right-to-left shunt, the patient was infused intravenously with packed red blood cells and volume expander. Oxygen delivery was also increased by the transfusion of packed red blood cells. To avoid unacceptable hypoxemia immediately after the procedure, the atrial septum was initially incised with a very small-blade catheter. Nine months after the BAS, catheterization revealed a decrease in mean pulmonary arterial pressure to 73 mmHg and an increase in CI to 2.5 L/min per m². Thirteen months after the BAS, the patient died as a result of progressive worsening of right-sided heart failure. We concluded that BAS could be successful in patients with severe pulmonary hypertension providing attention is paid to the patient's condition and that BAS is an effective therapy for prolonging survival.

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Key Words: Primary pulmonary hypertension; Blade atrial septostomy; Park blade catheter

PPrimary pulmonary hypertension (PPH) is a disease with a poor prognosis characterized by a progressive increase in pulmonary arterial pressure with consequent right-sided heart failure. Patients who show severe right-sided heart failure and low cardiac output are often refractory to conventional therapy¹ and additional vasodilator therapy may sometimes cause worsening of symptoms.

Recently, it has been reported that blade atrial septostomy (BAS) can result in clinical improvement in patients with PPH and serve as a palliative bridge to heart/lung transplantation^{2–4}. In this report we describe the first successful use of BAS in Japan for the treatment of severe PPH.

Case Report

The patient was an 18-year-old woman who had been followed since 14 years of age for primary pulmonary hypertension (PPH). Her symptoms deteriorated despite oxygen and vasodilator therapy in 1995. The patient presented with dyspnea and cyanosis at admission to the hospital on 31 May 1995. Her height was 151 cm and weight 51 kg. Systemic blood pressure was 86/62 mmHg and pulse rate 82 beats/min. Her respiratory rate was 48/min. Arterial oxygen saturation was 88%. Her condition was classified New York Heart Association (NYHA) class IV. Diastolic murmur was audible at 2 LSB and systolic murmur at 4 LSB. The pulmonary component of the second heart sound was markedly accentuated. Chest radiography

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Table 1 The Results of Cardiac Catheterizations in our Patient

	August 1994	June 1995		March 1996
		Before BAS	After BAS	
PAP (mmHg)	114/50 (m74)	150/edp23 (RVP)	165/edp20 (RVP)	104/65 (m73)
FAP (mmHg)	114/80 (m85)	108/75 (m85)	150/100 (m116)	104/75 (m90)
mRAP (mmHg)	6	15	14	17
PVR (units·min m ²)	24.5			47.9
SaO ₂ (%)	91	88	72	73
CI (l/min per m ²)	2.2	1.0	1.2	2.5

Serial catheterizations revealed the progression of pulmonary hypertension. After BAS, blood pressure increased from 108/75 to 150/100 mmHg and CI increased from 1.0 to 1.2 L/min per m². SaO₂ fell from 88% to 72%. Nine months after the procedure, mean pulmonary arterial pressure had decreased to 73 mmHg and CI had increased to 2.5 L/min per m². PAP, pulmonary arterial pressure; FAP, femoral arterial pressure; mRAP, mean right atrial pressure; PVR, pulmonary vascular resistance; SaO₂, arterial oxygen saturation; CI, cardiac index; RVP, right ventricular pressure.

Fig 1. Clinical course in our patient. Pulmonary hypertension progressed markedly and CI decreased to 1.0 L/min per m². However, after BAS, blood pressure increased, respiratory distress gradually improved, and the amount of urine produced increased. CI increased to 2.2 L/min per m² 1 month after the procedure. BAS, blade atrial septostomy; DOA, dopamine (maximum 3 µg/kg per min); DOB, dobutamine (maximum 5 µg/kg per min); lipo-PGE₁, alprostadil (maximum 8 ng/kg per min); SpO₂, percutaneous arterial oxygen saturation; SBP, systemic blood pressure; CVP, central venous pressure; CI, cardiac index.

revealed an increased cardiothoracic ratio of 58%. Echocardiography demonstrated a marked increase in right ventricular pressure and severe regurgitation of the tricuspid valve and the pulmonary valve. Fractional shortening of the left ventricle was 0.44. Compared

with the previous catheterization, the data obtained in catheterization revealed progressive worsening of pulmonary hypertension (Table 1).

After admission, symptoms caused by low cardiac output continued to worsen in spite of vigorous

Fig 2. Femoral arterial pressure at BAS. Because the first incision was made using a 10-mm Park blade catheter, blood pressure increased from 108/75 mmHg to 150/100 mmHg. FA, femoral arterial pressure.

Fig 3. Histology of the small pulmonary artery ($\times 40$, hematoxylin-eosin stain). Severe intimal proliferation with medial hypertrophy and near-complete luminal obstruction is apparent.

medical treatment: the administration of oxygen, dopamine, dobutamine, and diuretics (Fig 1). Use of a vasodilator, ie, intravenous infusion of lipoprostaglandin E_1 , only caused further deterioration by lowering systemic blood pressure. In addition, the patient's family hoped that she could undergo a heart/lung transplantation, although this was later canceled for medical and other reasons.

Thus, we performed blade atrial septostomy on 8 June to relieve the patient's symptoms after obtaining informed consent from her and her family. Before the procedure, packed red blood cells and volume expander were infused to maintain the pulmonary blood flow. Central venous pressure ranged from 15 to 18 mmHg. During the catheterization, the patient remained in a sitting position because she complained of respiratory distress. After placement of an arterial catheter to monitor systemic arterial pressure, a trans-septal punc-

ture was performed with a Brockenbrough needle through a 9-French Mullins long sheath. The process was continuously monitored by echocardiography to avoid perforation of the posterior wall of the enlarged right atrium. A 10-mm Park blade catheter was placed through the long sheath into the left atrium after removing the needle and the dilator. The long sheath was withdrawn to the level of the inferior vena cava. The blade, extended to an angle of approximately 45° and directed inferiorly, was withdrawn slowly across the right atrium. The expected diameter of the hole produced by the blade was about 6 mm. As the incision was predicted to produce a marked decrease in arterial oxygen saturation (SaO_2), 200 ml of packed red blood cells was rapidly transfused. Immediately after the incision, the blood pressure increased from 108/75 to 150/100 mmHg, SaO_2 decreased from 88% to 72% and cardiac index (CI), measured by the Fick

Table 2 Comparison of Preoperative Data in our Patient and Those of Kerstein's et al

	mRAP	mPAP (mmHg)	mSAP	PVR (units·m ²)	CI (L/min per m ²)	SaO ₂ (%)
<i>Kerstein's et al</i>						
Successfully treated patients (n=13)	11.2±7.1	68.8±22.1	91.9±14.2	30.6±10.2	2.2±0.6	97.4±1.7
Intraoperation deaths (n=2)						
Patient 1	5	105	73	62	1.7	100
Patient 2	26	98	75	64	1.4	93
Our patient	15	150/edp23 (RVP)	86		1.0	88

Two patients who died during the operation had more serious pulmonary hypertension than the successfully treated patients. In our patient, pulmonary resistance was expected to exceed 60 units m² and SaO₂ decreased to 88% at rest. Our patient's condition was as severe as the 2 patients in Kerstein et al's series who died intraoperatively. mRAP, mean right atrial pressure; mPAP, mean pulmonary arterial pressure; mSAP, mean systemic arterial pressure; RVP, right ventricular pressure.

method, increased from 1.0 to 1.2 L/min per m² (Table 1). As the blood pressure was elevated sufficiently, we discontinued the procedure after only 1 incision (Fig 2).

After the catheterization, the patient suffered from respiratory distress due to hypoxemia for 2 weeks. As the systemic blood pressure was sufficiently elevated, we were able to keep her under sedation during this period. The symptoms caused by low cardiac output gradually improved. One month later respiratory distress had almost disappeared, and catecholamines and diuretics were discontinued. CI increased to 2.2 L/min per m² (Fig 1). The patient was not discharged from the hospital, but she was able to enjoy eating and speaking.

Catheterization in March 1996, 9 months after the BAS, showed that the mean pulmonary arterial pressure had decreased to 73 mmHg and CI had increased to 2.5 L/min per m² (Table 1). Only a 5-mm balloon could pass through the atrial septal defect created.

Thirteen months after admission, the patient died as a result of progressive worsening of the right-side heart failure. At the postmortem examination, the atrial septal defect created was recognized at the fossa ovalis and was 4 mm×3 mm in size. The right ventricle was markedly enlarged and hypertrophied. Severe intimal proliferation with medial hypertrophy and almost complete luminal obstruction of pulmonary arteries was found (Fig 3).

Discussion

The NIH Registry on PPH has documented a mean survival of 2.8 years in a large cohort study.¹ Patients who have syncope or right-sided heart failure have an especially poor prognosis.⁵ Many patients do not

respond to medical treatment, such as the use of vasodilators and anticoagulants.^{6,7} However, other options for the treatment of severe PPH are now available, such as continuous intravenous infusion of prostacyclin, inhalation of nitric oxide, blade atrial septostomy (BAS), and heart/lung transplantation.

Patients with PPH and a patent foramen ovale do not suffer from recurrent syncope and survive longer than patients without a patent foramen ovale. This is because the right-to-left shunt through the patent foramen ovale maintains cardiac output and reduces right ventricular overload.^{8,9} However, in the first report of atrial septostomy for pulmonary hypertension,¹⁰ the patient experienced marked hypoxemia and blood pressure fell after the operation, leading to death. On the other hand, Kerstein et al² have recently performed BAS in 15 patients with PPH. They reported that BAS was successful in 11 patients and clearly increased the 3-year survival rate. They concluded that BAS was effective as a palliative procedure until heart/lung transplantation. They considered as candidates for BAS patients who experienced recurrent syncope despite maximal medical therapy, whose systemic arterial saturation was greater than 92% at rest, and in whom left ventricular function was normal. Among the patients who met their criteria for BAS, they were able to reduce postoperative hypoxemia to a minimum level and achieved a considerable success rate.

The preoperative data of our patient are compared with those of the patients who underwent BAS in Kerstein's report in Table 2. In the successful cases of Kerstein's report, the mean pulmonary arterial pressure ranged from 42 to 126 mmHg, averaging 68.8 mmHg, and the pulmonary vascular resistance ranged from 16 to 49 units·m², averaging 30.6 units·m². In 2

patients who died during the operation, the preoperative findings were poorer than in the successfully treated patients, pulmonary vascular resistance being over 60 units·m² and CI under 2.0 L/min per m². In the patient described here, pulmonary vascular resistance was expected to exceed 60 units·m² and SaO₂ was 88% at rest. Thus, the condition of our patient was as severe as in the 2 patients reported by Kerstein et al who died intraoperatively. We therefore paid particular attention to the following factors when performing the BAS.

1. To maintain pulmonary blood flow after creating the right-to-left shunt, packed red blood cells were transfused, a volume expander was infused, and diuretics were stopped 12 h before the catheterization. We maintained central venous pressure in the range of 15–18 mmHg.
2. To increase oxygen delivery, the hematocrit was increased by transfusion of packed red blood cells.
3. To avoid unacceptable hypoxemia due to a massive right-to-left shunt, the atrial septum was initially incised with a small-blade catheter. We used a 10-mm Park blade catheter for the first incision. In previous reports, large Park blade catheters, between 14 mm and 21 mm, were used, which may have been a possible cause of the excessive hypoxemia. We believe that it is extremely important to carry out the first incision carefully using the smallest blade catheter possible, especially in patients with severe PPH.

We performed the BAS as an emergency measure and the patient was thought to be at the outer limits of suitability as a candidate for BAS. Kerstein et al² stated that patients with markedly elevated pulmonary vascular resistance do not tolerate BAS. Nihill et al³ also recommended that the criterion for performing BAS in patients with pulmonary hypertension should be pulmonary vascular resistance of less than 50 units·m².³ Of course, BAS is not indicated in patients with deep cyanosis due to a right-to-left shunt or left ventricle dysfunction, but it may be indicated in patients who have elevated pulmonary artery resistance if we pay special attention to the factors mentioned above.

At the catheterization 9 months after the BAS, CI had increased and both mean pulmonary arterial pressure and pulmonary vascular resistance had decreased (Table 1). Even such a small defect as was shown in the postmortem examination may maintain cardiac output for a long time. In addition, the long-term reduction in right ventricular overload caused by the right-to-left shunt appears to prevent the worsening of pulmonary hypertension.

Recently, a continuous infusion of prostacyclin was reported to produce symptomatic and hemodynamic improvement, as well as improved survival, in patients with PPH.¹¹ In future, if this drug is approved in Japan, intravenous infusion of prostacyclin may become an important treatment for PPH. However, for patients who do not react to vasodilators, BAS seems to be one of the best therapies for prolonging survival.

Our patient experienced respiratory distress and intense anxiety for 2 weeks until she adapted to hypoxemia. Patients with severe symptoms of PPH are expected to develop severe postoperative hypoxemia. Thus, it is necessary to evaluate each patient carefully on the basis of risk-benefit considerations.

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