

A case of pediatric atrial fibrillation ablation resulting in unilateral pulmonary vein occlusion during long-term follow-up



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Introduction

Pulmonary vein (PV) isolation is a treatment of choice for drug-resistant atrial fibrillation (AF) in adult patients, and PV obstruction is one of the complications that may occur in less than 1.0% of patients.¹ Because AF is rare in pediatric patients, there are few reports of PV isolation in children, and its complications have not been reported. Here we report a case of PV occlusion that developed as a long-term complication of PV isolation performed for paroxysmal AF in an otherwise healthy pediatric patient.

Case report

A 10-year-old boy (height 136.0 cm, weight 37.3 kg, body mass index [BMI] 20.2) was referred to our center for the treatment of paroxysmal AF. He had no prior medical history except arrhythmia that had been detected during a fifth-grade school screening and led to the diagnosis of paroxysmal AF by a referring physician. There were no subjective symptoms, and the CHADS2 score was zero. The Holter recordings from the referring physician showed an AF burden of 71% per day. At the initial consultation, an electrocardiogram showed AF, and oral aprindine 20 mg and bisoprolol fumarate 5 mg were prescribed. There were no findings suggestive of secondary AF, such as hypertension or abnormal thyroid function. Treadmill stress testing showed repetitive bursts of AF, and Holter recordings showed an AF burden of 74% per day that lasted up to 5.5 hours and was poorly responsive to the drug therapy. The patient was admitted for ablation therapy 1 month after the first consultation. On admission, there

was a mild elevation of brain natriuretic peptide (18.7 pg/mL). No organic or functional abnormalities were found by echocardiography, and the left atrial diameter was 29.3 mm (113% of normal). On contrast-enhanced chest computed tomography (CT), the left atrial volume was 59 mL, and PV structures were normal. The longest diameter of the left superior PV was 11.2 mm, and that of the left inferior PV was 10.4 mm.

An anesthesiologist performed controlled ventilation and general anesthesia with propofol throughout the procedure. An 8.5F long sheath (Agilis; Abbott, Chicago, IL) and an 8F long sheath (SL1; Abbott) were inserted through the right femoral vein into the left atrium via the septum. A defibrillation catheter (BeeAT; Japan Lifeline, Tokyo, Japan) was inserted through the right internal jugular vein via a 7F short sheath. An esophageal temperature monitoring catheter was also placed in the esophagus (Esophastar with an alarm system at 39°C; Japan Lifeline). A circled electrophysiology catheter (Inquiry Optima; St. Jude Medical, Saint Paul, MN) was positioned in the left PV. Irregular potentials with cycle length of 120–200 ms were identified within the left superior and inferior PVs (Figure 1).

SmartTouch Surround Flow catheters (Biosense-Webster, Irvine, CA) were used for ablation. PV isolation was performed by expanding the posterior wall side to the left atrial side. Radiofrequency ablation was performed from the bottom of the left pulmonary vein (LPV) for extensive encircling isolation at 25–30 W at the anterior wall and 15–20 W at the posterior wall of the LPV (Figure 2). The current was applied over 10–30 seconds. Ablation was discontinued at the left atrial posterior wall near the esophagus, determined by the enhanced CT images, when the esophageal temperature reached 39°C. In total to the posterior wall, 5 sites were ablated at 15–20 W for a mean of 13.65 seconds (range 10.05–18.6 seconds) with a total lesion size of 13.6 mm. Sustained AF disappeared at the beginning of radiofrequency delivery, and sinus rhythm was maintained by the time LPV isolation was completed. Right PV isolation was not performed, as AF and atrial extrasystole were no longer detected after the completion of LPV isolation.

KEYWORDS Children; Paroxysmal atrial fibrillation; Pulmonary vein isolation; Complication; Pulmonary vein occlusion
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KEY TEACHING POINTS

- For lone atrial fibrillation in children, antiarrhythmic drugs should be the treatment of choice, and pulmonary vein (PV) isolation should be avoided whenever possible.
- If PV isolation is selected, long-term follow-up at shorter intervals would seem reasonable to closely monitor the patient for signs of PV stenosis.
- When complications are suspected, a more thorough examination should be performed aggressively to evaluate PV stenosis and occlusion.

Transthoracic echocardiography at the end of the procedure and on the following day showed a high-echo region consistent with postablation edema, but the PV flow rate was not elevated. The patient was discharged 3 days after the procedure and received an oral anticoagulant (edoxaban tosylate monohydrate) and potassium-competitive acid blocker (vonoprazan fumarate) for 2 months.

Electrocardiography and chest radiography findings 30 days after the procedure were unremarkable. At 99 days post-procedure, the patient consulted the referring physician for occasional pain in the back of the left chest. Left pulmonary congestion was not detected by thoracic radiography. At 135 days postprocedure, transthoracic echocardiography was performed at our center to evaluate possible PV stenosis. PV stenosis was not recognized at the time, although a retrospective

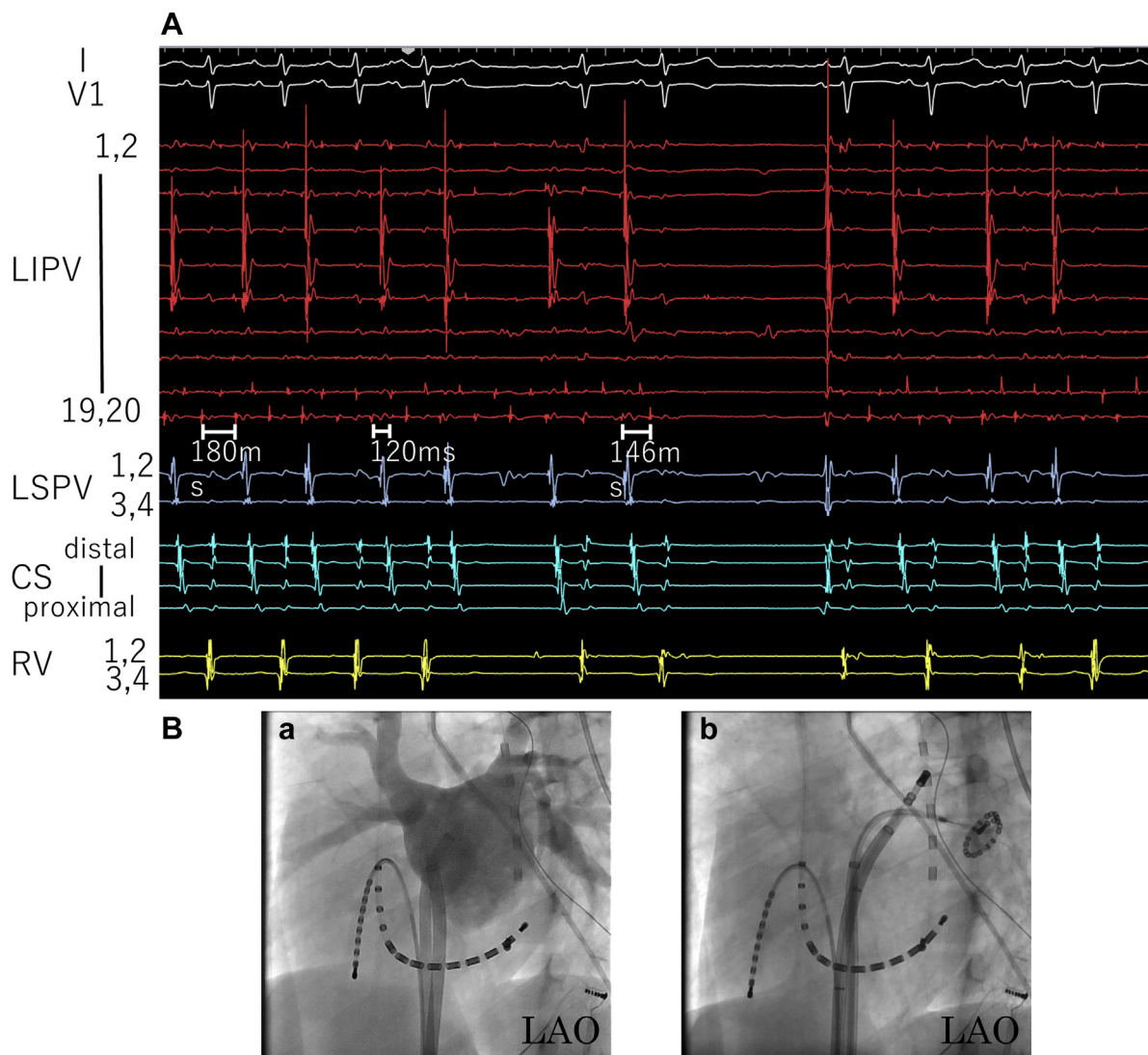


Figure 1 Intracardiac potentials and catheter placement. **A:** Left atrial potential. Atrial fibrillation potentials of cycle lengths of 120–200 ms are identified within the left pulmonary vein. **B:** Left atrial angiography (a) and catheter position (b). The Optima catheter (St. Jude Medical, Saint Paul, MN) is in the left inferior pulmonary vein (LIPV), and the ablation catheter is in the left superior pulmonary vein (LSPV). CS = coronary sinus; LAO = left anterior oblique; RV = right ventricle.

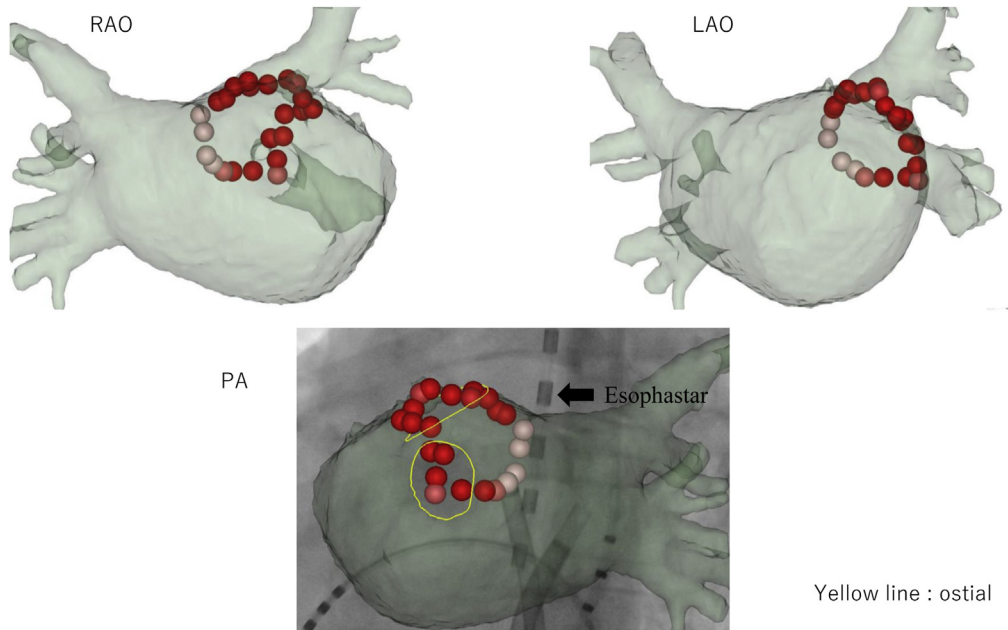


Figure 2 Ablation lesions in the left atrium. Pulmonary vein (PV) isolation was performed on the left pulmonary veins only. The tag radius was 2 mm, and the AI indicator was used for ablation. In the posterior-to-anterior (PA) view, the PV was cut to make for better visual understanding, and the ostia are shown in yellow. LAO = left anterior oblique; RAO = right anterior oblique.

review of the images revealed findings suggestive of LPV stenosis. The follow-up monitoring was continued by the referring physician.

One year and 9 months after the procedure, the patient consulted the referring physician for coughing up blood-stained sputum after exercise. Contrast-enhanced CT revealed LPV occlusion and collateral vessels around the

bronchi, and the patient was referred to our center for treatment. Vital signs were within normal ranges. Expectoration of a small amount of blood-tinged sputum was observed a few more times but resolved after 2 days. Echocardiography did not show any evidence of pulmonary hypertension, but complete LPV occlusion was confirmed by contrast-enhanced CT (Figure 3). Lung scintigraphy revealed an

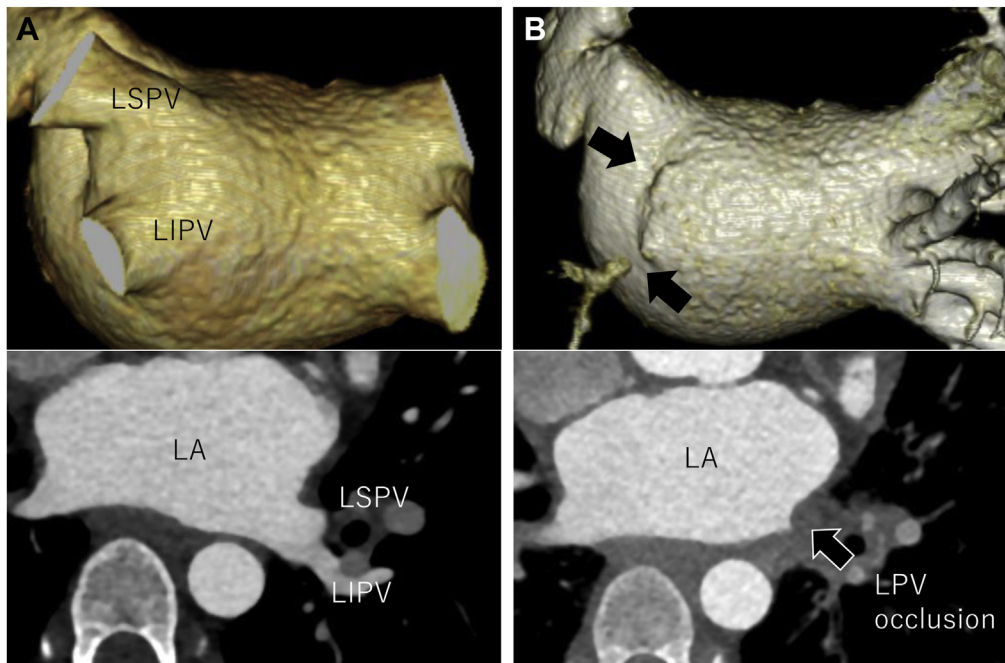


Figure 3 Contrast-enhanced computed tomography (CT) before and after ablation. **A:** Contrast-enhanced CT taken before ablation. **B:** Contrast-enhanced CT at 1 year and 9 months after ablation. Arrows indicate complete occlusion of the left pulmonary vein. LA = left atrium; LIPV = left inferior pulmonary vein; LPV = left pulmonary vein; LSPV = left superior pulmonary vein.

asymmetric perfusion pattern with 89.8% in the right lung and 10.2% in the left lung.

To evaluate the hemodynamics, cardiac catheterization was performed under general anesthesia with propofol and controlled ventilation. The left pulmonary artery pressure was 30/10/16 mm Hg, and the right pulmonary artery pressure was 25/9/16 mm Hg. Pulmonary hypertension was not indicated. The LPA wedge pressure was 15 mm Hg and higher than the right side (8 mm Hg). The pulmonary vascular resistance was $1.91 \text{ Unit} \cdot \text{m}^2$ with a pulmonary resistance to systemic resistance ratio (Rp/Rs) of 0.12. Upon dobutamine stress, the systemic systolic pressure increased to 192 mm Hg, and the mean pulmonary arterial pressure of 25 mm Hg indicated mild pulmonary hypertension. The pulmonary vascular resistance was $2.31 \text{ Unit} \cdot \text{m}^2$, and the Rp/Rs was 0.08. Angiography demonstrated complete occlusion of the LPV and collateral vessels to the bronchial arteries, which were the likely cause of hemoptysis.

After cardiac catheterization, no blood-tinged sputum or any other complications were observed. Balloon dilation, stenting, and surgical interventions were considered but thought to be challenging owing to the complete stenosis of the PV. Given the resolution of the clinical symptoms and the absence of pulmonary hypertension at rest, close monitoring of the patient was continued without any intervention or medication. At the time of this report (1 year after the first episode of hemoptysis and 2 years and 9 months after ablation), the patient is on mild exercise restriction and free from clinical symptoms, such as blood sputum and pulmonary hypertension at rest, or recurrence of AF.

Discussion

Reports of PV isolation for pediatric lone AF are infrequent, and there are no data on complications. Furst and colleagues² reported 16 cases of ablation therapy for pediatric lone AF, and PV isolation was performed in 10 patients (14–19 years old, BMI 21.9–26.5). Recurrence was experienced by 5 of the 10 patients, but complications were not reported. In another retrospective study by Mills and colleagues,³ 6 of 42 patients with lone AF (14.6–16.1 years old) underwent ablation therapy, but complications were not reported.

In our 2020 study,⁴ PV isolation was performed in 3 pediatric AF patients without congenital heart disease (10, 13, and 16 years old). The case presented in this report is one of these patients. At the time of the previous report, he did not have recurrence or complication but later developed hemoptysis. Complications were not reported in the other 2 patients, but the 13-year-old patient had a recurrence and underwent the second ablation.

In adults, the incidence of postablation PV stenosis is lower with wide circumferential ablation than with ostial PV isolation. The overall incidence has fallen to below 1%. PV stenosis or occlusion can be prevented by avoiding radiofrequency energy delivery within the PV,^{5–7} and reported preoperative predictors of this complication do not include the body size and left atrial volume.^{8,9} The patient in the

present case was 10 years old, but PV isolation was considered an option based on his body size (136 cm, 37.3 kg, and BMI of 20.2) that was close to that of a small adult woman. His left atrial volume was 59 mL. For comparison, we extracted data of 35 consecutive adult female patients who underwent ablation for AF at our center in 2019. The median (range) age, height, weight, and BMI were 75 years (47–87 years), 151.7 cm (142–167.3 cm), 56 kg (35–86.4 kg), and 24.2 (14.8–33.4), respectively. The median left atrial volume was 120 mL (range 72–215 mL). Although the height, weight, and BMI of the present case fall within or close to average (-0.7 standard deviation [SD], $+0.3$ SD, and $+1.2$ SD, respectively), his left atrial volume was much smaller than those of adult female patients (-3 SD). The left atrial volume may have played a part in the development of PV stenosis.

Although the patient received oral aprindine and bisoprolol fumarate, the AF burden did not change before and after the treatment. This was why we selected ablation, but amiodarone and flecainide were other options that should have been considered. Given these pharmacological options, we now believe that PV isolation should be avoided in children whenever possible.

Symptoms of PV stenosis include dyspnea, cough, chest pain, hemoptysis, and recurrent lung infections. In the present patient, PV stenosis may have developed as early as 99 days after ablation, when he experienced chest pain. Although the symptom was mild and resolved spontaneously, more thorough examination should have been performed to evaluate PV stenosis and occlusion.

In adult patients, a decision to treat PV stenosis is made based on clinical signs. Arentz and colleagues¹⁰ reported that 11 patients with significant stenosis or occlusion of 1 or 2 PVs did not develop pulmonary hypertension at rest during the follow-up period of 50 ± 15 months after ablation. Seven of these patients had elevated pulmonary pressures during exercise, but lesion progression was not observed even without treatment. Nonetheless, close monitoring seems essential, because the consequences of exercise-induced pulmonary hypertension on the pulmonary vasculature remain largely unknown. On the other hand, symptomatic stenosis often requires percutaneous (eg, balloon dilatation and stent placement) or surgical intervention, and stent implantation appears to be the treatment of choice, as a lower restenosis rate compared to balloon angioplasty¹¹ and good long-term outcomes¹² have been reported. Surgical intervention is considered when the percutaneous approach has failed, or irreversible pulmonary vascular remodeling has developed. However, the data on surgical intervention are still scarce, and its benefits over percutaneous intervention are yet to be determined.^{7,13} The patient in the present case developed a complete occlusion for which catheterization or surgical revascularization is not applicable, especially after such a long period of time. Pneumonectomy is another option to consider. At the time of this report, 1 year after the development of PV occlusion, the patient is being closely monitored without treatment, as he is asymptomatic with normal

pulmonary artery pressure at rest. Exercise restriction is gradually being eased.

Conclusion

We describe a rare pediatric case of PV occlusion, which was confirmed 21 months after PV isolation for paroxysmal AF. For lone AF in children, antiarrhythmic drugs should be the treatment of choice, and PV isolation should be avoided whenever possible. When this procedure is selected, long-term follow-up at shorter intervals would seem reasonable to closely monitor the patient for signs of PV stenosis.

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