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CASE REPORT

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Pulmonary root remodeling procedure for symptomatic supravalvular pulmonary stenosis in an adult patient who underwent pulmonary artery banding: a case report

Kei Mukawa[1*], Yutaka Iba[1] and Nobuyoshi Kawaharada[1]

Abstract

Background Pulmonary artery banding (PAB) is performed as a palliative surgery for congenital heart diseases. Although pulmonary stenosis is one of the complications of PAB, symptomatic supravalvular pulmonary stenosis (SVPS) in adulthood after PAB is an extremely rare condition. Further, very few studies have focused on surgical cases of SVPS in adult. Herein, we describe a case of symptomatic SVPS in adulthood after PAB that was successfully managed with surgery.

Case presentation A 55-year-old male patient had been presenting with worsening shortness of breath for 10 years. Of note, he had been diagnosed with membranous ventricular septum defect at birth and he underwent PAB at 11 months of age. The patient underwent direct closure of ventricular septum defect and pulmonary artery debanding at 3 years and 6 months old. Medical examinations showed SVPS, with a pressure gradient of 60 mmHg. We planned pulmonary artery anterior wall plasty. However, based on the operative findings, the pulmonary stenosis was located in the whole circumference just above the annulus. Hence, it was challenging to completely relieve the stenosis with a pulmonary patch, and the Yacoub remodeling technique was applied to pulmonary artery reconstruction. Intraoperative pressure gradient between the right ventricle and the distal pulmonary artery dropped to 11 mmHg. Post operative course was uneventful.

Conclusions Patient with SVPS could present with symptoms in the long term after PAB in childhood. Therefore, due to the currently increasing number of adult congenital heart disease (ACHD) cases, attention should be paid to patients who underwent PAB. In this case, pulmonary artery was degenerated in the whole circumference because of the remained banding tape and the stenosis part was just above the annulus. Therefore, we applied Yacoub remodeling technique to this reconstruction. This pulmonary root remodeling procedure was an effective option for such SVPS.

*Correspondence:
Kei Mukawa
m13112km@jichi.ac.jp

Full list of author information is available at the end of the article

Keywords Adult congenital heart disease, Supravalvular pulmonary stenosis, Supravalvular pulmonary stenosis after pulmonary artery banding, Pulmonary root remodeling procedure, Yacoub remodeling technique for pulmonary artery

Introduction

The number of adult congenital heart disease (ACHD) cases has been increasing in recent years owing to medical and surgical evolutions [1]. Pulmonary artery banding (PAB) is performed as a palliative surgery for congenital heart diseases. However, it can cause secondary supravalvular pulmonary stenosis (SVPS) [2]. In most cases, SVPS is generally repaired during pulmonary artery debanding [3]. Therefore, there are only a few cases of symptomatic SVPS in adulthood after PAB. In numerous cases, SVPS in childhood is commonly managed with surgical procedures, particularly patch augmentation into one or two sinuses [4]. To the best of our knowledge, there are no reports on adult surgical cases. Herein, we describe a case of symptomatic SVPS in adulthood after PAB that was successfully managed with surgery.

Case report

A 55-year-old male patient who had no regular medication presented to the local hospital due to shortness of breath, which was progressively getting worse within 10 years. Of note, he had been diagnosed with membranous ventricular septum defect at birth and he underwent PAB at 11 months of age. The patient underwent direct closure of ventricular septum defect and pulmonary artery debanding at 3 years and 6 months old.

The patient' vital signs were unremarkable. His blood pressure, heart rate, and oxygen saturation level on room air was 135/89 mmHg, 88 beats per minute, and 98%, respectively. Transthoracic echocardiography showed a normal ejection fraction and no right ventricular dilatation. Trivial tricuspid regurgitation, and mild pulmonary regurgitation was observed. Of note, the diameter of the supravalvular pulmonary artery was just 13.6 mm, the maximum velocity was 4.1 m/s, and the maximum pressure gradient was 69 mmHg. Three-dimensional computed tomography scan showed severe stenosis of the supravalvular pulmonary artery (Fig. 1a). Then, right ventricular angiography was performed. The pressure gradient between the right ventricle and the main pulmonary artery was 60 mmHg. Moreover, angiography showed pulmonary stenosis (Fig. 1b).

Our multidisciplinary team planned pulmonary artery anterior wall plasty with a prosthetic patch for symptomatic severe SVPS. The patient underwent redo median sternotomy (Fig. 2a), and cardiopulmonary bypass (CPB) with ascending aortic cannulation and bicaval drainage was established. After cardiac arrest, the anterior wall of the main pulmonary artery was incised longitudinally. The sinotubular junction in the pulmonary artery, which is the stenotic part, was degenerated over the whole circumferences and the posterior wall was bulging into the lumen (Fig. 2b). Further, the residual banding tape was found in the posterior wall (Fig. 2c), which had been used during PAB. Stenosis could not be relieved with anterior wall patch plasty. The stenotic part was just above the annulus, and using a 21 mm ball sizer, we confirmed the absence of valvular stenosis. The mobility and coaptation of the valve remained satisfactory. Consequently, we decided to reconstruct the three sinuses of the pulmonary artery such as the Yacoub remodeling technique. The main pulmonary artery was transected completely just above the sinotubular junction. Each sinus was incised straight down to the annulus. A 24 mm Gelweave straight graft (Vascutek, Inchinnan, Scotland) was trimmed to correspond with each sinus. The graft scallops at each nadir were anastomosed and sewed to commissures with 5−0 polypropylene (Fig. 3d). This remodeling procedure did not affect symmetric valve closure based on water test. Subsequently, the end of the main pulmonary artery was anastomosed with the graft, which the distal part was beveled (Fig. 3e). After these procedures were completed (Fig. 2d), an aortic cross-clamp was removed. CPB was weaned easily, and the absence of pulmonary regurgitation was confirmed on transesophageal echocardiography. Thereafter, right heart catheterization showed that the pressure gradient between the right ventricle and the distal pulmonary artery was 11 mmHg. The total CPB time was 142 min and the cross-clamp time was 107 min. The patient was extubated after 1 h. The postoperative course was uneventful, and postoperative computed tomography scan revealed the release of stenosis (Fig. 4). The patient was discharged 14 days after surgery.

Discussion

SVPS in childhood could occur after PAB [2]. One of the reasons for the secondary SVPS is indicated that the banding duration is long [5] and the banding tape induces local inflammation within the vascular wall [6]. Adhesions in the posterior wall of the banding site can occasionally complicate the complete removal of the tape because of inflammation. In this case, the banding tape remained in the posterior tissue of the pulmonary artery. Pathological findings revealed that abundant collagen fibers were found around the adventitia with lymphocyte and plasma cell infiltration within the pulmonary artery wall. We hypothesized that the residual banding tape can cause tissue proliferation over the whole circumferences.

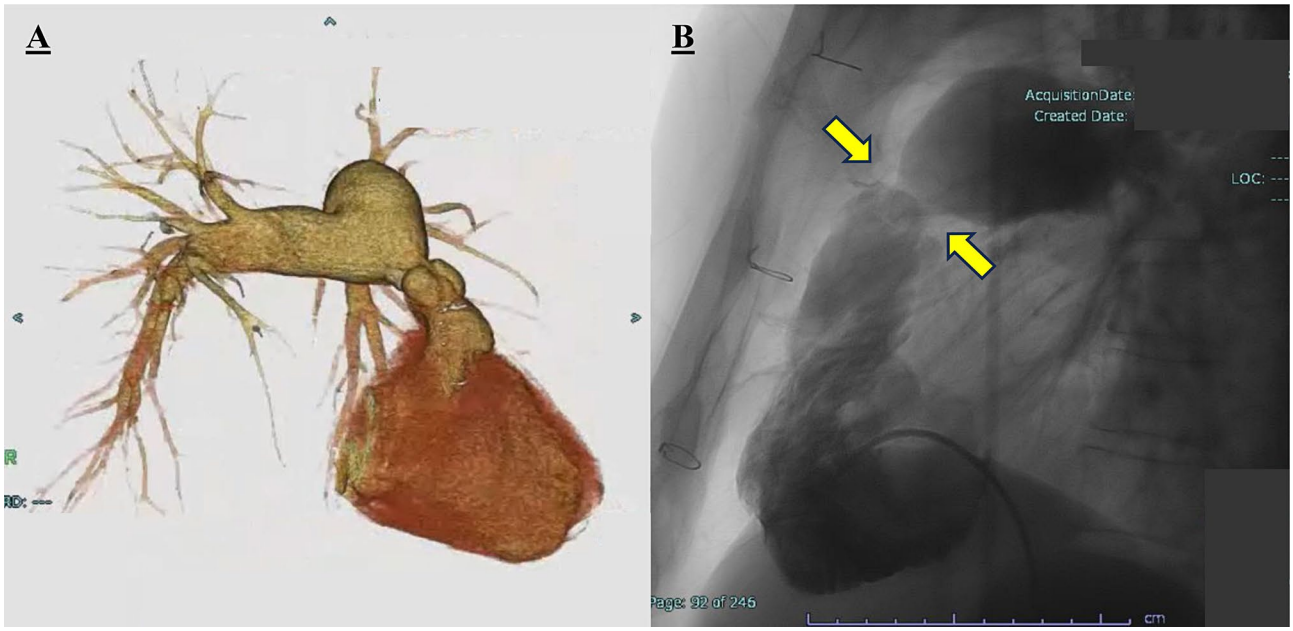

Fig. 1 (**A**) Three-dimensional computed tomography scan of the right ventricular outflow tract and pulmonary artery. (**B**) Right ventricular angiography, pulmonary stenosis (yellow arrow). The pressure gradient was 60mmHg

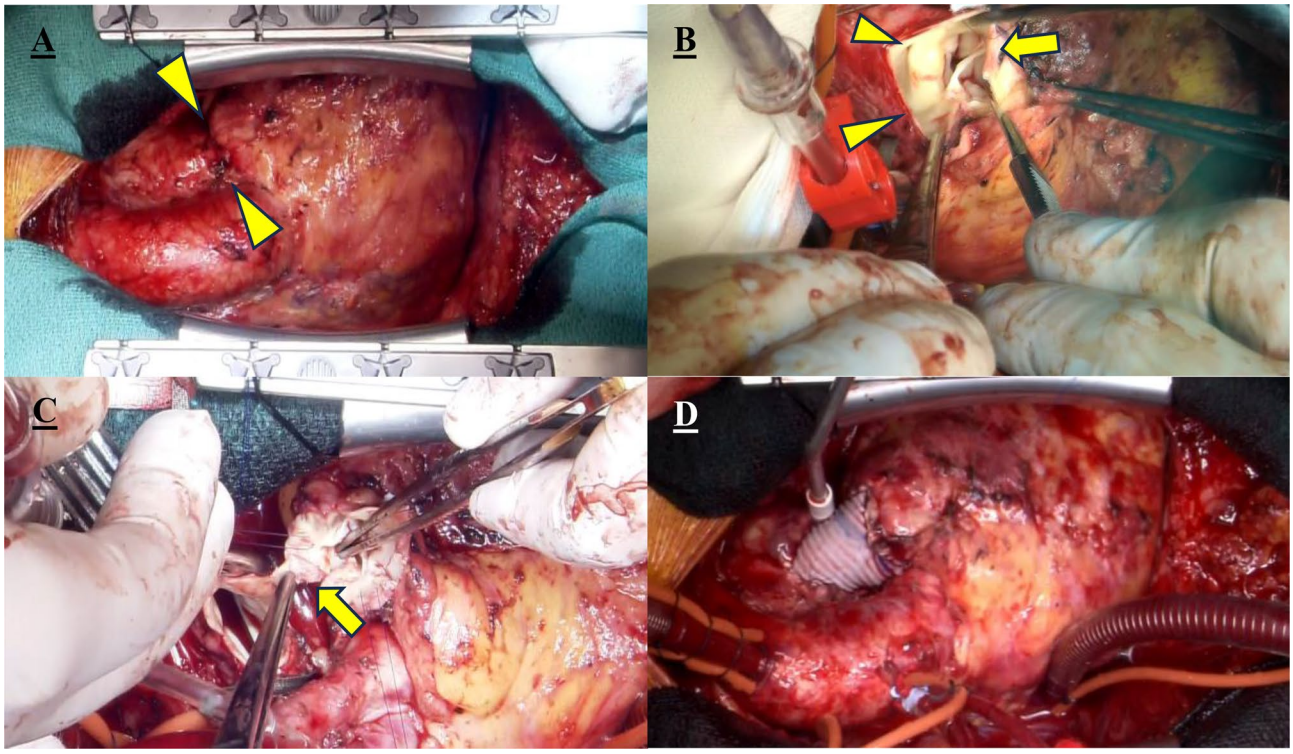

Fig. 2 (**A**) Stenosis part (yellow triangle). (**B**) Inner lumen of the pulmonary artery, protruded wall (yellow triangle), pulmonary valve (yellow arrow). (**C**) Residual band from the posterior wall, which was used at the time of PAB (yellow arrow). (**D**) Postoperative findings

According to the 2020 ECS for ACHD [1], the right ventricular outflow tract stenosis is severe if the gradient is >64 mmHg. When this severity is applied to this case, the SVPS is assessed as severe. Based on these points, severe SVPS gradually caused symptoms in the long term. Symptomatic SVPS is an extremely rare condition. Meanwhile, the number of ACHD cases is increasing owing to medical innovations, and the proportion of adult patients

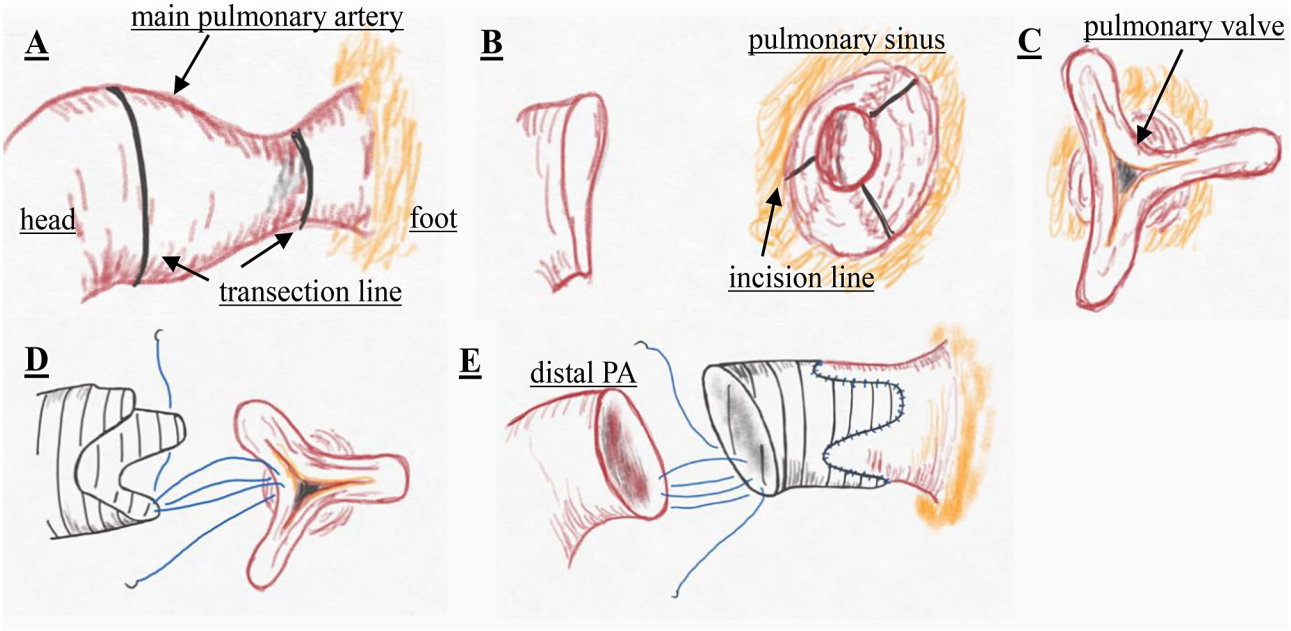

Fig. 3 Operative procedures (modified Yacoub remodeling technique) (**A**) Main pulmonary artery transection. (**B**) Sinus incision down to each annulus. (**C**) Trimmed pulmonary valve. (**D**) Proximal anastomosis with graft scallops. (**E**) Distal anastomosis

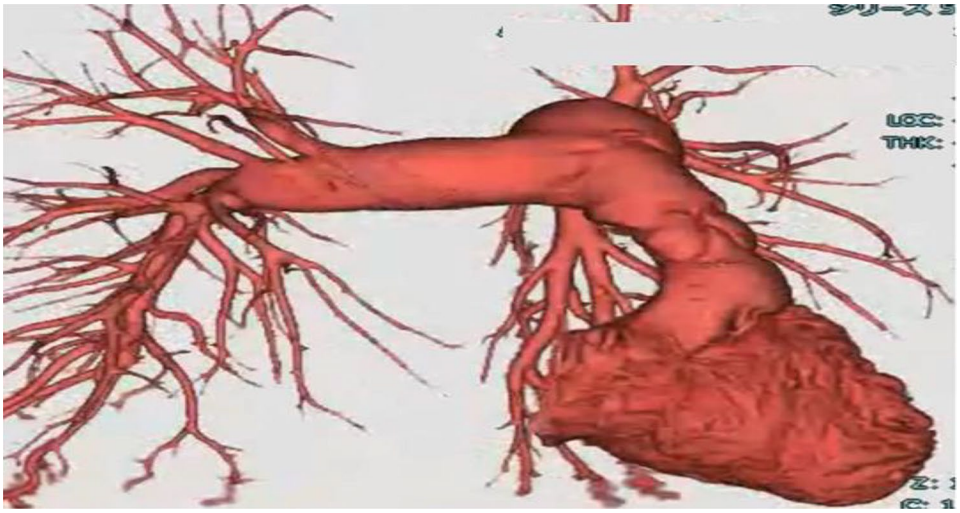

Fig. 4 Postoperative three-dimensional computed tomography scan

who underwent PAB can increase. In relation to this, special caution should be taken when managing these cases.

A previous study has reported some surgical procedures that can be used for SVPS in childhood [4]. To the best of our knowledge, there are no adult surgical cases of SVPS. Balloon dilatation/stent implantation and surgical repair are the two treatment options for SVPS. Percutaneous dilatation for childhood SVPS is reportedly less effective [7]. In this case, the patient presented with chronic adult SVPS, and he was symptomatic. Therefore, surgery was performed based on the recurrence risk of balloon dilatation. Regarding surgical procedures, single or Y-shaped patch dilatation is occasionally used for SVPS [2, 4]. In this case, we initially planned to perform prosthetic patch dilatation of the pulmonary anterior wall. However, from intraoperative findings, it was challenging to relieve the stenosis caused by the pulmonary artery, which degenerated over the whole circumferences, particularly the posterior wall. Hence, the degenerated tissue was excluded, and the pulmonary artery was completely reconstructed. The application of completely three-sinus repair on pediatric SVPS have been reported [8]. Nevertheless, autologous repair for an adult case poses challenges due to lack of tissue elasticity. The Yacoub remodeling technique provided inspiration for this reconstruction. The reconstruction in the current

case was modified and applied because the stenotic part was just above the annulus. The pressure gradient significantly decreased after the surgery. The pulmonary artery in postPAB SVPS can be degenerated over the whole circumference, and patch dilatation is generally insufficient for such a case. Therefore, this pulmonary root remodeling procedure can be an effective method particularly in cases which the stenosis is located just above the annulus unless the pulmonary valve function is abnormal.

With the greater number of ACHD, the incidence of pulmonary artery diseases can increase. A previous study has reported about the use of the Yacoub remodeling technique for pulmonary artery aneurysm [9]. Results have shown that this pulmonary root remodeling procedure can be effective against pulmonary artery disease.

Conclusions

Patients with SVPS could present with symptoms in the long term after PAB in childhood, and pulmonary root remodeling procedure was an effective .

Abbreviations

ACHD	Adult Congenital Heart Disease
CPB	Cardiopulmonary Bypass
PAB	Pulmonary Artery Banding
SVPS	Supravalvular Pulmonary Stenosis

Supplementary Information

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Supplementary Material 1

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Author contributions

K.M wrote the main manuscript text and prepared all figures. All authors reviewed the manuscript.

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Data availability

No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

A written consent for the submission and publication of this case report was obtained from the patient.

Competing interests

The authors declare no competing interests.

Author details

[1]Cardiovascular Surgery Department, Sapporo Medical University, S-1, W-16, Chuo-ku, Sapporo, Hokkaido 060-8543, Japan

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