

# Unilateral Pulmonary Artery Banding for Critically ill Neonate with Absent Pulmonary Valve Syndrome: A Case Report

Junichi Koizumi (✉ [jkoizumi@iwate-med.ac.jp](mailto:jkoizumi@iwate-med.ac.jp))

Iwate Medical University Hospital <https://orcid.org/0000-0002-3940-8957>

Shigeto Tsuji

Iwate Medical University Hospital

Tomoyuki Iwase

Iwate Medical University Hospital

Azuma Tabayashi

Iwate Medical University Hospital

Hajime Kin

Iwate Medical University Hospital

Akio Ikai

Mt. Fuji Shizuoka Children's Hospital

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## Case report

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# Abstract

**Background** Absent pulmonary valve syndrome (APVS) is a rare congenital heart disease that is challenging to manage when it presents with severe symptoms at an early age.

We describe a critically ill neonate with tetralogy of Fallot (TOF) with APVS in whom main pulmonary artery (PA) banding failed, but alternative left PA banding and PA plication and open chest strategy effectively stabilized the patient's condition and enabled elective cardiac repair 6 days later.

**Case presentation** A 3.2-kg baby girl with prenatal diagnosis of TOF and APVS was electively delivered by cesarean section. She was intubated soon after birth, however her condition deteriorated with respiratory and metabolic acidosis even with maximum ventilator settings and inotropic support. On the second day of life, she developed bloody stool suggesting necrotizing enterocolitis, indicating the need for a palliative procedure without cardiopulmonary bypass. First, we attempted main PA banding but abandoned it due to suspected myocardial ischemia caused by left main coronary artery compression. As an alternative procedure, we performed left PA banding, which improved oxygen saturation and increased blood pressure; the chest was left open. Postoperatively, the patient's condition improved dramatically within 48 hours. On the eighth day of life, we performed an elective intracardiac repair on her under stable preoperative condition. At the 5-year follow-up, she was in a stable condition at home with home oxygen therapy.

**Conclusion** Unilateral PA banding and open chest strategy may be an alternative treatment of choice for critically ill neonates with TOF with APVS.

## Introduction

The management of symptomatic neonates with tetralogy of Fallot (TOF) with absent pulmonary valve syndrome (APVS) is challenging. Currently, primary cardiac repair with pulmonary artery (PA) plication is the surgical treatment of choice. However, critical illness often precludes the use of cardiopulmonary bypass and subsequent intracardiac repair with systemic heparinization. In this situation, main pulmonary artery banding or ligation with or without Blalock-Taussig shunt may be used as a palliative procedure. We describe a critically ill neonate with TOF with APVS in whom main PA banding failed, but alternative left PA banding and PA plication and open chest strategy effectively stabilized the patient's condition and enabled elective cardiac repair 6 days later.

## Case Presentaion

A fetus at 26 weeks gestation was diagnosed with TOF with APVS, with marked dilation of the PA. At 39 weeks gestation, a 3.2-kg baby girl was electively delivered by cesarean section in our hospital. She was intubated soon after birth due to profound cyanosis and respiratory distress. Echocardiography and computed tomography (CT) (Fig. 1a, b, c) showed TOF with APVS and a huge central PA aneurysm with a short main PA segment. The left PA was especially dilated, with a diameter of 30 mm up to the hilum. Her

condition gradually deteriorated with respiratory and metabolic acidosis even with maximum ventilator settings (FiO<sub>2</sub> 1.0, SIMV 45, PIP 22 cmH<sub>2</sub>O, PEEP 5 cmH<sub>2</sub>O, inhalation of nitric oxide 40 ppm) and inotropic support (dopamine 5 mcg/kg/min, dobutamine 6 mcg/kg/min). Her blood pressure was 55/41 mmHg, and pulse rate was 160 beats per minute. Her oxygen saturation was between 75% and 80%. Her blood gas analysis revealed the following: pH 7.29, PCO<sub>2</sub> 50.0 mmHg, PO<sub>2</sub> 44.6 mmHg, lactate 1.7 mmol/L, and base excess - 3.8 mEq/L. Urgent intracardiac repair using cardiopulmonary bypass was scheduled on the second day of her life. However, she developed bloody stool suggesting necrotizing enterocolitis, indicating the need for a palliative procedure without cardiopulmonary bypass.

After median sternotomy, main PA banding to a circumference of 30 mm was applied. Oxygen saturation and blood pressure temporarily improved; however, electrocardiography showed evidence of ST depression. Epicardial echocardiography revealed deterioration of left ventricular contraction probably due to compression of the left main coronary artery by the banding tape or main PA. Loosening the band did not resolve the problem. Therefore, instead of banding the main PA, the left PA, which was significantly dilated, was banded. Initially, a left PA band with a circumference of 20 mm was applied; however, the band was believed to be too tight when oxygen saturation dropped to around 70%. Instead of 20 mm banding, 30 mm banding was successfully performed with an improvement in oxygen saturation to around 90% and blood pressure around 70/50 mmHg. Epicardial echocardiography revealed decreased pulmonary regurgitation at the pulmonary valve annulus and improvement of right ventricular dilatation. The distal portion of the enlarged left pulmonary artery was plicated with plegetted 5 - 0 polypropylene interrupted sutures. Subsequently, we also tried to perform right pulmonary artery banding; however, we abandoned it because of hemodynamic deterioration with increased pulmonary regurgitation due to possible increased main PA wall tension and dilated pulmonary valve annulus. The chest was left open to prevent tracheobronchial compression. Postoperatively, the patient's condition improved dramatically within 48 hours. Her blood pressure was 75/46 mmHg, and pulse rate was 140 beats per minute. Her saturation was 90% and blood gas analysis revealed the following: pH 7.38, PCO<sub>2</sub> 43.2 mmHg, PO<sub>2</sub> 52.1 mmHg, lactate 1.7 mmol/L, and base excess + 0.1 mEq/L on minimal inotropes (dopamine 5 mcg/kg/min, epinephrine 0.02 mcg/kg/min, milrinone 0.3 mcg/kg/min) and normal ventilator settings (FiO<sub>2</sub> 0.21, SIMV 19, PIP 20 cmH<sub>2</sub>O, PEEP 3 cmH<sub>2</sub>O) without nitric oxide inhalation. On the eighth day of life, bilateral PA plication and intracardiac repair with a reconstruction of the right ventricular outflow tract (RVOT) using a handmade bicuspid valved expanded polytetrafluoroethylene conduit were performed. The PA was transferred anteriorly to release the tracheobronchial compression. At 5 months of age, she suffered tracheobronchial compression again due to re-dilatation of the left PA. Therefore, a second repair, including PA wall re-plication and aortopulmonary-pxy, was performed. CT revealed improved tracheobronchial compression and decreased left PA dilatation (Fig. 2a, b). She was weaned from the ventilator at the age of 6 months. At the 5-year follow-up, she was in a stable condition at home with home oxygen therapy.

## Discussion

Intracardiac repair with PA plication and competent RVOT reconstruction with or without pulmonary artery anterior translocation is considered to be the first treatment of choice for TOF with APVS. However, critically ill state such as necrotizing enterocolitis often precludes the use of cardiopulmonary bypass, which may cause fatal events. In such cases, palliative salvage surgery should be considered. Historically, main PA banding or main PA ligation with modified Blalock-Taussig shunt, with or without pulmonary artery plication, has been reported with successful outcomes. [1, 2, 3] Its mechanism might involve control of pulmonary regurgitation resulting in efficient pulmonary artery forward flow. Further, it resulted in right ventricular volume unloading and improved left ventricular filling. In our case, main PA banding resulted in ST depression and left ventricular dysfunction despite relatively loose banding, which might have been due to left main coronary artery compression. Thus, instead of the main PA banding, we banded the left PA, which was markedly dilated compared with the right PA. The mechanism of severe pulmonary regurgitation in APVS might be not only a dysplastic pulmonary valve but also a dilatation of pulmonary valve annulus due to increased blood volume in the PA. It can be inferred that in conditions with high pulmonary vascular resistance and severe pulmonary regurgitation, the larger pulmonary artery holds more blood and thus more regurgitation. Therefore, the regulation of blood flow in the larger PA may have a greater effect on hemodynamics. Intraoperative echocardiography revealed a decrease in the diameter of the pulmonary valve annulus and a reduction in pulmonary regurgitation. It is speculated that left PA banding reduces left pulmonary regurgitation resulting in the reduction of main PA wall tension and pulmonary valve annular size—which decreases total PA regurgitation and increases forward PA blood flow. Concerning the worse effect of adding a right PA banding, it is hypothesized that adding a contralateral PA banding raises a main PA wall tension, resulting in dilatation of the pulmonary valve annulus, which increases PA regurgitation. It is difficult to determine the appropriate length of the unilateral PA banding. Initially, we performed left PA banding with a 20 mm circumference because we speculated that it might result in a near-normal branch PA circumference; however, it was deemed to be too tight. In usual PA banding, for the simple left to right shunt lesion, the aim is to create stenosis and reduce the forward PA blood flow. However, in TOF with APVS, we hypothesize that PA banding is performed not to create stenosis but to reduce the regurgitation. This is compatible with the looser banding and is appropriate for TOF with APVS. Epicardial echocardiography should be an effective measure to determine a suitable level of band tightness.

Hanley reported the efficacy of sternotomy for relieving the airway obstruction in the neonate with TOF and APVS. [4] We believe that open chest strategy also contributed to the improvement of respiratory and circulatory status of critically sick patient.

## Conclusion

If main PA banding fails to palliate sick neonates with TOF with APVS, especially those with a very short main PA, unilateral PA banding of the larger artery and open chest strategy may be an alternative treatment of choice to reduce pulmonary regurgitation and stabilize the sick neonate.

# Abbreviations

APVS: Absent Pulmonary Valve Syndrome

TOF: Tetralogy of Fallot

PA: Pulmonary Artery

CT: Computed Tomography

FiO<sub>2</sub>: Fraction of Inspired Oxygen

SIMV: Synchronized Intermittent Mandatory Ventilation

PIP: Peak Inspiratory Pressure

PEEP: Positive End-Expiratory Pressure

RVOT: Right Ventricular Outflow Tract

# Declarations

## **Ethics approval and consent to participate**

Not applicable.

## **Consent for publication**

Parents had signed consent for publication.

## **Availability of data and materials**

Please contact corresponding author for data requests.

## **Competing interests**

All authors declare that they have no competing interests.

## **Funding**

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## **Authors' contributions**

JK is the main and corresponding author for this manuscript. All authors read and approved the final manuscript.

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## Figures

Fig. 1A

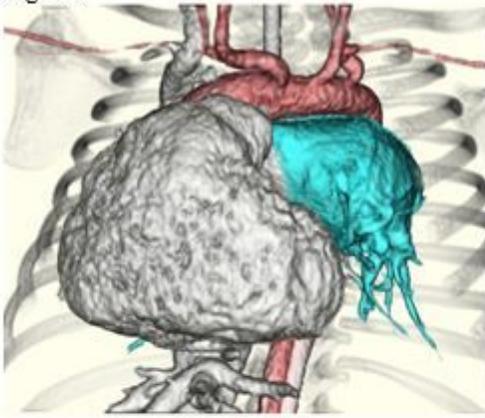


Fig. 1B

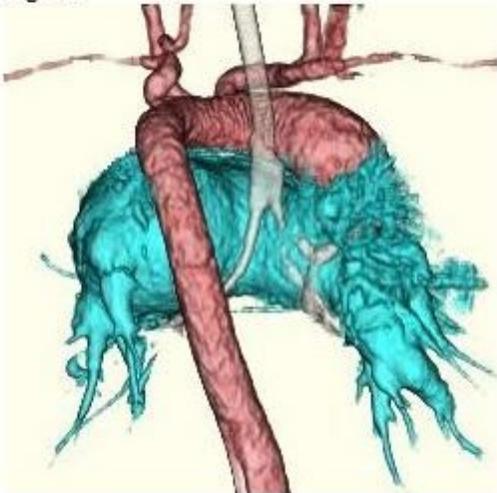


Fig. 1C



**Figure 1**

Computed tomography before left pulmonary artery banding. Left pulmonary artery is remarkably dilated (a,c), by which tracheobronchus is compressed (b,c)

Fig. 2A

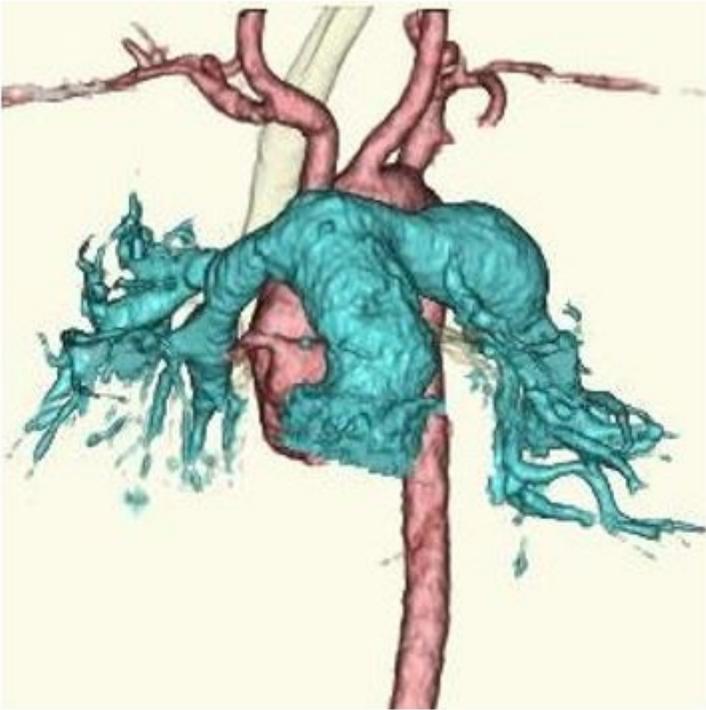
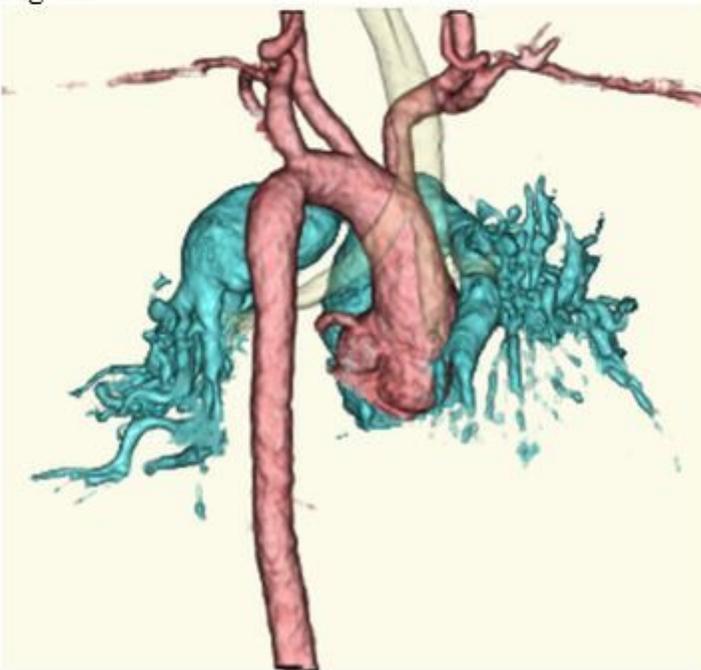


Fig. 2B



**Figure 2**

Computed tomography after intracardiac repair. Pulmonary artery is anteriorly translocated and downsized (a). Tracheobronchial compression is improved (b).

## Supplementary Files

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