Received: 13 January 2021 | Revised: 18 February 2021 | Accepted: 22 March 2021

DOI: 10.1111/jocs.15545

CASE REPORT

JOURNAL OF CARDIAC SURGERY WILEY

# Multidisciplinary management of a rare case of mixed total anomalous pulmonary venous connection

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## Abstract

**Introduction:** Mixed total anomalous pulmonary venous connection (TAPVC) is a extremely rare congenital heart disease.

**Methods:** We report the initial management of a case of Mixed total anomalous pulmonary venous connection associated to right extralobar bronchopulmonary sequestration (BPS).

**Results:** Mixed TAPVC associated to right extra-lobar BPS was diagnosed at birth in a full-term newborn. At one month of age, the patient underwent embolization of the BPS, complicated by coil entrapment in the right common iliac artery requiring urgent laparotomy. Few days later, the congenital cardiac repair was accomplished uneventfully. At 12-months follow-up, the patient did not have pulmonary hypertension, but presented a moderate stenosis of the right femoral artery, which was effectively treated with anticoagulation therapy.

**Conclusions:** The multidisciplinary approach allowed a successful treatment of these complex anomalies and the related potential complications.

KEYWORDS

amplatzer plug, bronchopulmonary sequestration, total anomalous pulmonary venous connection, transcatheter embolization

## 1 | INTRODUCTION

This paper reports the case of an infant affected by mixed total anomalous pulmonary venous connection (TAPVC), an extremely rare congenital heart disease (CHD),[1] and bronchopulmonary sequestration (BPS), representing the second most frequent lung malformations in children.[2] We described our multidisciplinary therapeutic strategy of a complication that occurred during the embolization of the anomalous BPS vessel and discussed possible different surgical approaches to correct both the TAPVC and BPS.

## 2 | CASE PRESENTATION

A full-term male newborn, with hypoxemia at birth, was diagnosed with mixed TAPVC, in which the left pulmonary veins (PVs) were draining into the coronary sinus and the right PVs were connected to the inferior vena cava (Scimitar syndrome).

The cardiac catheterization also described a right extralobar BPS, supplied by a vessel from the celiac artery (Figure 1).

A multistage elective treatment was planned which included the percutaneous embolization of the anomalous vessel at the age of

  

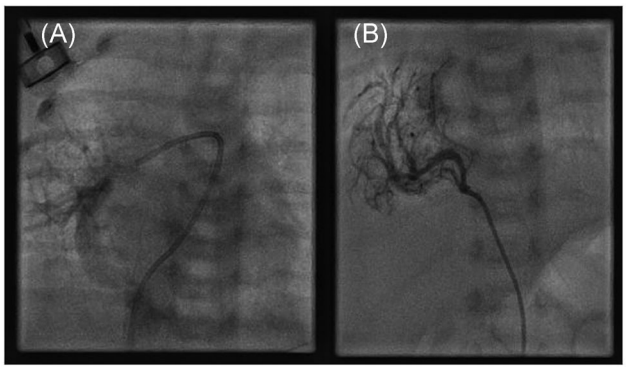


**FIGURE 1** First cardiac catheterization; right anomalous pulmonary venous connection (A); anomalous arterial vessel from the celiac artery (B)

1 month, to prevent pulmonary systemic overflow and pulmonary hypertension, followed by the repair of the CHD.

Through a 4Fr Benson® catheter, a 5-mm-Amplatzer® vascular plug was placed in the anomalous vessel. A second 5 × 5-mm-Cook® coil was inserted but remained entrapped in the celiac artery. Attempting to remove it, the coil remained blocked in the right common iliac artery (Figure 2). An urgent laparotomy was performed through a Rutherford Morrison incision. The iliac artery was incised, the coil was removed, and 2Fr Fogarty catheter angioplasty allowed to remove a femoral artery thrombus.

On POD 1, moderate stenosis of the right femoral artery was detected, due to possible clotting. Systemic urokinase was administered for 48 h, followed by continuous infusion of heparin until the next surgery.

After 7 days, the patient underwent partial cardiac repair, consisting of baffling the left anomalous pulmonary venous flow into the left atrium, unroofing the coronary sinus, and closing the coronary sinus ostium with a patch. The child was discharged on POD 26 on warfarin and metoprolol due to postoperative supraventricular tachycardia.

At 12-month follow-up, the patient is growing well, with no recurrence of arrhythmias. The femoral arterial flow was improved by collateral circles. The cardiac evaluation showed good ventricular function, with no residual pulmonary hypertension and good left pulmonary venous flow. Metoprolol and warfarin were discontinued after 6 months and the patient was scheduled for the final cardiac repair at school age.

## 3 | COMMENT

Repair of complex TAPVC is challenging and presents a high risk of postoperative pulmonary hypertension and possible occurrence of stenosis,[3] which raises the overall mortality up to 40%.[1] Mixed anomalous TAPVC with anomalous right PV in a scimitar syndrome and anomalous left vein in the coronary arteries is an even more rare CHD. Since the repair of scimitar syndrome in infancy is affected by high postoperative morbidity and risk of thrombosis,[4] a staged procedure was preferred. The first step consisted of baffling the left anomalous PV into the left atrium, unroofing the coronary sinus, and closing its exit in the right atrium with a patch. Nevertheless, the residual shunt was not clinically significant in most of the cases, as it was previously reported.[4] For this reason, the child can uneventfully grow, until the scimitar vein repair will be indicated in older age.

The extralobar BPS is normally located at the base of the left chest, and children may be asymptomatic or have important respiratory distress and severe congestive heart failure secondary to the volume-loading from a large systemic artery for the sequestered segment.[2] The thoracoscopic resection of the lesion is the standard treatment.

These anomalies require the involvement of pediatric cardiologists, pediatric surgeons, and cardiothoracic surgeons, in a tertiary-care center, who share different skills for defining the diagnostic/therapeutic approaches.[5]

The BPS embolization using the Amplatzer device was indicated to prevent postoperative pulmonary overflow, as already described.[6] However, large case series of BPS embolization showed a high rate of failure, due to incomplete embolization, and persistence of the anomalous lung tissue.[2]

A further issue concerns the observed complication. This could be resolved through an extraperitoneal approach, normally used for kidney transplantations.

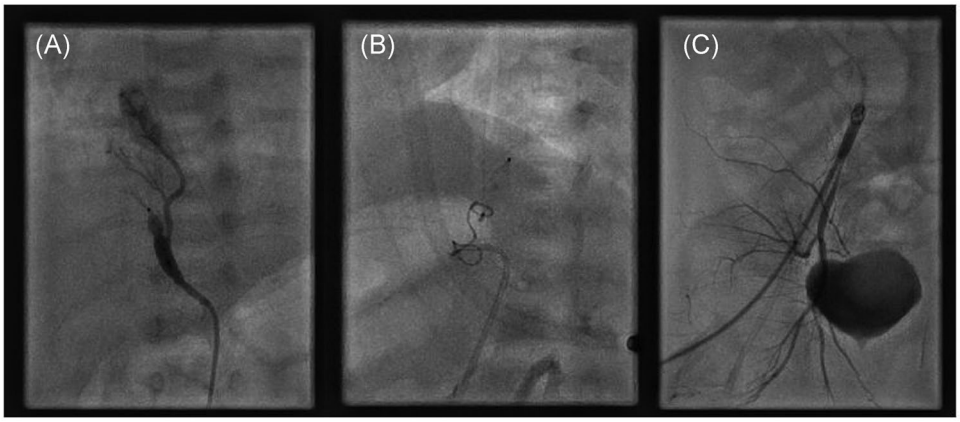


**FIGURE 2** Second cardiac catheterization; positioning of the Amplatzer plug (A and B); entrapment of the coil (C)

## 4 | CONCLUSION

The multidisciplinary approach allowed a successful treatment of these complex anomalies, despite the potential complications of BPS embolization.

The multidisciplinary team should be aware that BPS may persist after embolization, and possible alternative methods of treatment should be discussed.

### CONFLICT OF INTERESTS

The authors declare that there are no conflict of interests.

### AUTHOR CONTRIBUTIONS

*Concept/design*: Filippo Ghidini, Massimo Padalino, and Patrizia Dall'Igna. *Drafting*: Filippo Ghidini, Massimo Padalino, and Patrizia Dall'Igna. *Critical revision*: Biagio Castaldi, Massimo Padalino, and Patrizia Dall'Igna.

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**How to cite this article:** Ghidini F, Castaldi B, Padalino M, Dall'Igna P. Multidisciplinary management of a rare case of mixed total anomalous pulmonary venous connection. *J Card Surg*. 2021;1-3. https://doi.org/10.1111/jocs.15545