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Pulmonary Atresia with Ventricular Septal Defect and Major Aortopulmonary Collaterals Associated with Left Pulmonary Artery Interruption

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A multistage plan and multidisciplinary approach are the keys to successful repair in patients with pulmonary atresia (PA) with ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCAs). In this article, we present a multidisciplinary approach adopted to treat a patient with PA with VSD and MAPCAs associated with left pulmonary artery interruption.

Key words: 1. CHD, pulmonary atresia
2. CHD, septal defect
3. Major aortopulmonary collateral arteries

Case report

An infant weighing 2.91 kg was born at a gestational age of 38+0 weeks. Prenatal echocardiography revealed pulmonary atresia (PA) with ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCAs). Echocardiography performed immediately after birth revealed a disconnected left pulmonary artery supplied by the left patent ductus arteriosus (PDA), originating from the left innominate artery, in addition to the abovementioned findings. On the computed tomography (CT) performed 1 day after birth, a diminutive central right pulmonary artery (RPA), a disconnected left pulmonary artery supplied by the left PDA, and 2 MAPCAs from the proximal descending thoracic aorta were observed (Fig. 1A–C).

We decided to allow the diminutive RPA to grow and delay the reconstruction of the central pulmonary artery until the RPA was large enough to be reliably enlarged surgically. As the first intervention, a stent was placed in the left PDA at 26 days of age. On follow-up CT scan, severe narrowing of the PDA just proximal to the stent was observed and the RPA was found to be still diminutive (diameter <1.5 mm) (Fig. 1D, E). At 41 days of age, a central shunt was placed through a midline sternotomy with a method introduced by Gates et al. [1] in 1998. Subsequently, a stent was reinserted into the proximal PDA with the stent-in-stent approach in the catheterization laboratory. The hospital course was uncomplicated, and the patient was discharged with prescriptions for warfarin, captopril, and diuretics at 65 days of age. The level of brain natriuretic peptide at discharge was 510 pg/mL.

At 5 months of age, a follow-up angiography and CT scan revealed that the left pulmonary artery had grown to a sufficient size with normal arborization

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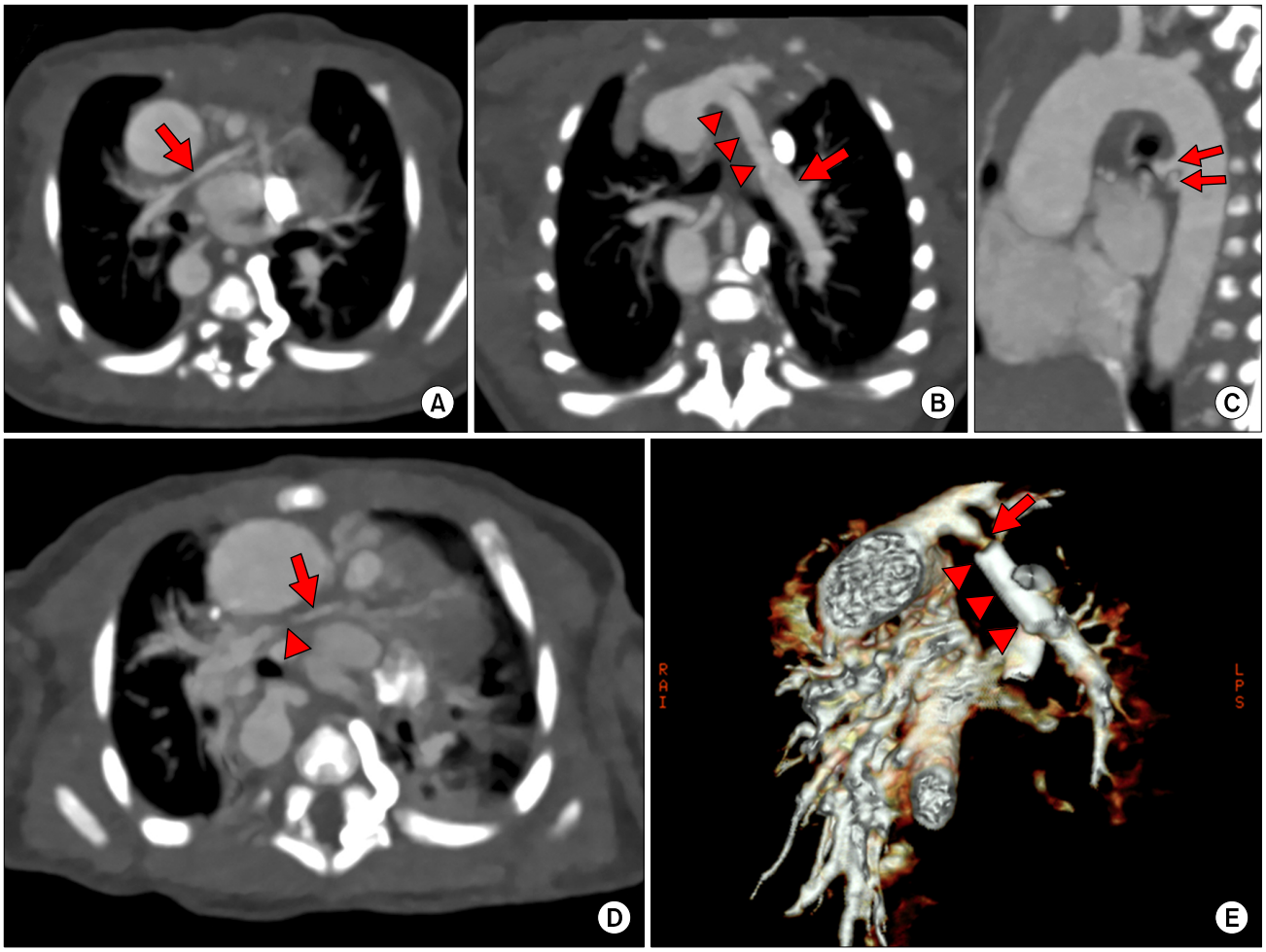

Fig. 1. Initial and post-stent follow up heart CT. Figures A–C are the initial heart CT image at one day old. (A) Small RPA (arrow). (B) The left pulmonary artery (arrow) from the left PDA (arrowhead). (C) Two major aortopulmonary collateral arteries (arrow) are shown arising from the proximal descending thoracic aorta. Figures D and E are the follow-up heart CT image following PDA stent at 34 days of age. (D) Diminutive RPA (arrow) with focal stenosis (arrowhead) at the distal portion. (E) The left pulmonary artery distal to the PDA stent (arrowhead). Severe narrowing (arrow) is seen proximal to the PDA stent. CT, computed tomography; RPA, right pulmonary artery; PDA, patent ductus arteriosus.

and that the intrapericardial RPA had grown to 3.5 mm in diameter. Two MAPCAs (C1 with communication and C2 without communication) were also identified, originating from the proximal descending aorta (Fig. 2A, B).

Surgical correction was planned; whether or not to proceed the intracardiac repair would be decided in the operating theater, depending on the result of the intraoperative pulmonary blood flow study [2,3] after the reconstruction of the central pulmonary artery. The patient's age and weight at corrective surgery were 8 months and 7.9 kg, respectively. After sternal reentry, sufficient fresh autologous pericardium was harvested for reconstruction of the central pulmonary artery. At first, central pulmonary artery reconstruction was performed under moderate hypothermic cardiopulmonary bypass and beating heart following ligation of the left PDA and division of the central shunt. A central left pulmonary artery was created by using an autologous pericardial roll, and the central RPA was reconstructed with unifocalization of the MAPCA (C2) and patch widening with fresh autologous pericardium (Fig. 2C). Under cardioplegic arrest with cold blood cardioplegia and normal ventilation, an intraoperative pulmonary blood flow study [2,3] was performed by cannulating

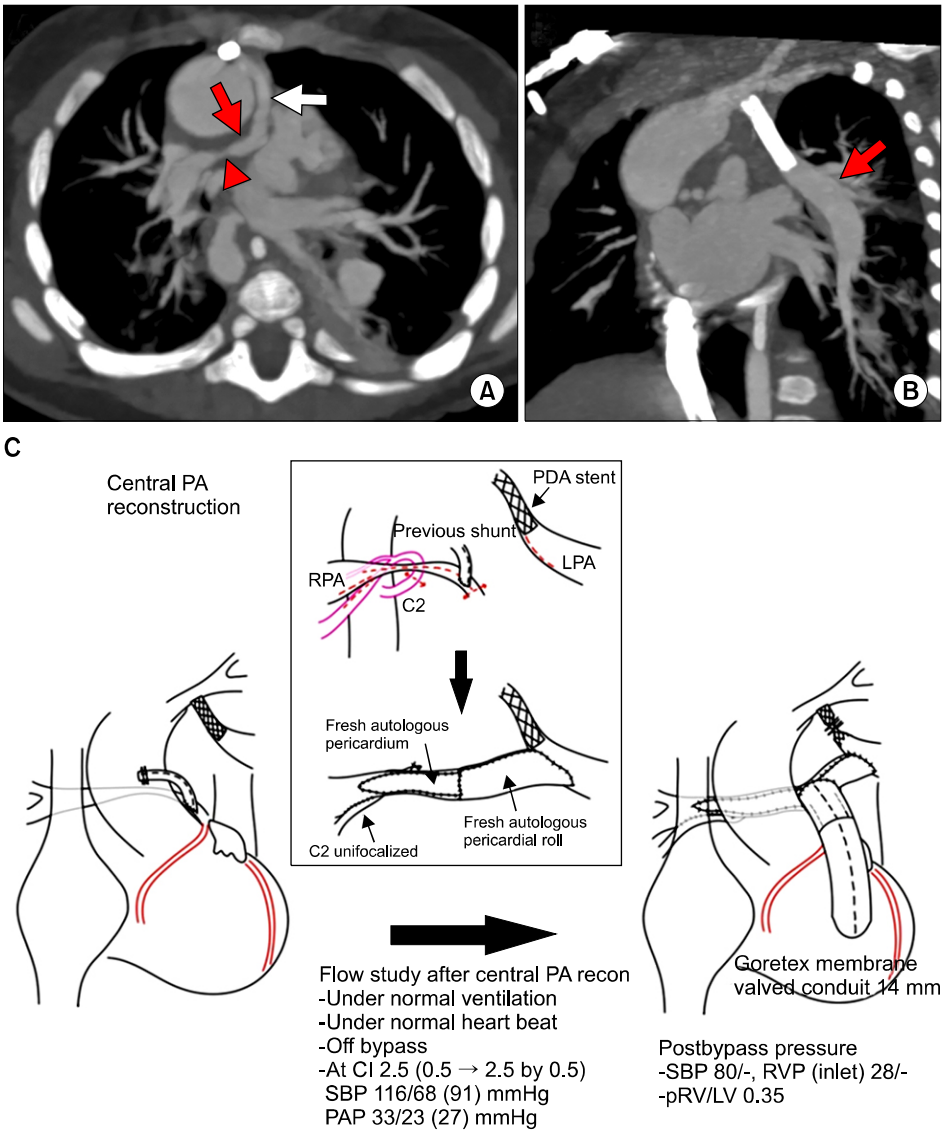

Fig. 2. Pre-repair heart CT scan at 5 months of age and the diagram of surgery. (A) Patent central shunt with nice contour (white arrow). The diameter of RPA (red arrow) increased compared to that before central shunt. Distal segmental stenosis (red arrowhead) still existed. (B) Well-grown left PA (red arrow) distal to the PDA stent. (C) Diagram of surgical correction shows methods of central PA reconstruction and RV-to-PA connection, and post-bypass profiles. C, major aortopulmonary collateral artery; CI, cardiac index; CT, computed tomography; LPA, left pulmonary artery; PA, pulmonary artery; PAP, pulmonary arterial pressure; PDA, patent ductus arteriosus; pRV/LV, pressure ratio of right ventricle to left ventricle; RPA, right pulmonary artery; RVP, right ventricular pressure; SBP, systemic blood pressure.

the reconstructed central pulmonary artery at a flow rate of 0.5 L/(min · m^2), increased incrementally up to 2.5 L/(min · m^2). Mean pulmonary artery pressure was measured during the study. Since the mean pulmonary artery pressure was 27 mmHg at a rate of 2.5 L/(min · m^2), we decided to close the VSD and establish the connection between the right ventricle (RV) and pulmonary artery. The VSD was closed with a Dacron patch through longitudinal right ventriculotomy. The connection between the RV and pulmonary artery was established with a homemade Gore-Tex membrane valved conduit, and the patent foramen ovale was left open. After separation from the bypass and completion of the modified ultrafiltration, the RV systolic pressure was 28 mmHg and the RV systolic pressure to systemic blood pressure ratio was 0.35. For the MAPCA (C1 with communication) that was not identified in the operating room, it was decided that interventional closure in the catheterization laboratory would be performed in the short term. The hospital course was uneventful, and the patient was discharged at 13 days after the surgery with a prescription for captopril. The level of brain natriuretic peptide at discharge was 87 pg/mL.

At 3 months after the repair, occlusion of the residual MAPCA (C1) was attempted in the catheterization laboratory, but failed. One month later, surgical occlusion of the residual MAPCA (C1) and con-

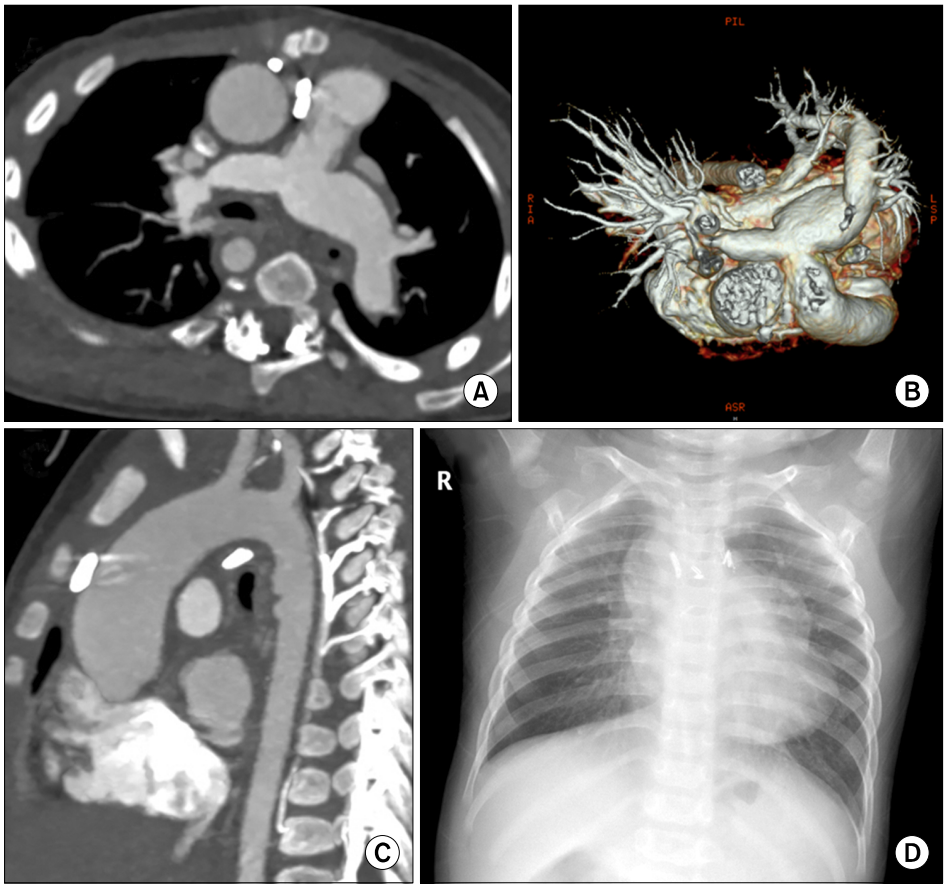

Fig. 3. Follow-up heart CT at discharge and simple chest radiography on the latest follow-up. Figures A–C are the follow-up heart CT image at discharge. (A, B) The central right pulmonary artery and left pulmonary artery look widely patent. (C) Major aortopulmonary collateral arteries originating from descending thoracic aorta have disappeared. (D) Simple chest radiography obtained at the latest follow-up shows symmetrical and normal pulmonary vascularity, and prominent contour of ascending aorta and conduit between right ventricle and pulmonary artery. Otherwise, no remarkable findings are seen. CT, computed tomography.

comitant unifocalization of the newly identified small MAPCA (C3) were performed through left thoracotomy. The patient was discharged at 1 week after the operation without complications. The follow-up CT scan obtained before discharge showed that the residual MAPCA disappeared and the reconstructed central pulmonary was widely patent (Fig. 3A–C).

The brain natriuretic peptide level at 3 months after the last surgery was 41 pg/mL. The latest follow-up echocardiogram (6 months after the operation) revealed no residual intracardiac shunt, a wide patent RV outflow tract, no pulmonary regurgitation, no evidence of RV hypertension, and excellent function of both ventricles. At the latest follow-up (7 months after the operation), the patient was in New York Heart Association functional class 1 without cardiac medication and had normal symmetric pulmonary vascularity on chest radiography (Fig. 3D).

Discussion

Two-thirds of the patients with PA are in association with MAPCA, and the survival rate in these cases is as low as 50% at 1 year of age and 8% at 10 years of age without surgery [4]. The management of PA with VSD and MAPCAs involves a multidisciplinary team approach that often requires repeated procedures, especially for those at the worst end of the spectrum [5]. The most crucial prerequisite for successful repair of PA with VSD and MAPCA is a well-prepared pulmonary artery, that is, a single-compartment, unobstructed, and low-pressure pulmonary circulation [6].

Although the central RPA was diminutive in our case, we could grow it with a central shunt, which was first introduced by Gates et al. [1]. In case of conducting a shunt anastomosis to the middle of a diminutive RPA, the shunt flow may be insufficient or blocked due to poor distal runoff, and therefore the RPA could not grow. It will be more effective for

growth of the branch RPA to anastomose the graft to the main pulmonary artery as close as possible to the ventriculoarterial junction rather than the RPA.

For the disconnected left pulmonary artery supplied by PDA, central left pulmonary artery reconstruction should have required non-vascular material, which would not guarantee its durability, and it was also difficult to establish a systemic-to-pulmonary artery shunt due to the uncertainty of the exact extent of the ductal tissue. Stent implantation through the catheter intervention could resolve these issues. Although tight narrowing occurred in the uncovered ductus, this can be effectively addressed with a stent-in-stent approach.

In patients with PA with VSD and MAPCA, the preoperative decision on a plan for intracardiac repair is always difficult due to the uncertainty about the unobstructed reconstruction of the central PA and the lack of predictability for post-repair RV pressure, which is known to be a major factor affecting long-term outcome. Honjo et al. [3] demonstrated that an intraoperative pulmonary blood flow study is better than preoperative anatomy in predicting RV pressure and physiological tolerance after intracardiac repair. Likewise, it was very useful in our case to predict post-bypass RV systolic pressure following central PA reconstruction and intracardiac repair.

Since the patient underwent 5 CT scans within a year, the radiation dosage was a concern for this growing young infant. However, the total dosage of radiation exposure during one year was 2.28 mSV, which is less than annual background radiation. This should be attributed to an effort by our radiologist to reduce the radiation exposure by applying a delicately designed and individualized CT protocol to this small child.

Even though this child had undergone many non-invasive and invasive studies—with 3 catheter interventions and 3 surgical operations in his short lifetime—he is now doing well without any medication. A lesson learned from our case is that a multidisciplinary and proactive approach is the key to success in treating patients with PA with VSD and MAPCAs.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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