Abstract A 2-year-old girl with isolated unilateral absence of right pulmonary artery is described. Catheterization at 5 months demonstrated hypoplastic right pulmonary artery by pulmonary venous wedge angiography, and the patient underwent right Blalock–Taussig shunt and angioplasty of right pulmonary artery with autologous pericardial roll as an initial step. At 2 years, she underwent anastomosis of right pulmonary artery to main pulmonary artery with an autologous pericardial tube. Postoperative computed tomography showed a patent reconstructed right pulmonary artery.

Key words Pericardium · Pulmonary artery · Surgical anastomosis · Congenital heart defect

Introduction

Congenital isolated unilateral absence of a pulmonary artery (PA) without any associated cardiac anomalies is a rare disease.1 We report here our experience of the staged surgical correction including Blalock–Taussig shunt and PA angioplasty at 7 months of age, and PA reconstruction with an autologous pericardial tube at 2 years of age.

Case report

The patient was a 2-year-old girl who was transferred to our hospital at 5 months of age. Cardiac catheterization demonstrated only the left pulmonary artery from the right ventricle without intracardiac disorder, while the hypoplastic lower lobar branch of the right PA with a diameter of 4 mm was visualized by right pulmonary venous wedge angiography (Fig. 1a,b). The right-sided ductal diverticulum originated from the base of the innominate artery (Fig. 1c). Left pulmonary arterial pressure in systole (PAP) was 27 mm Hg and the pressure ratio of the right to left ventricle was 0.28. Staged surgical correction was planned, and a right Blalock–Taussig shunt using a 5-mm polytetrafluoroethylene graft was created with simultaneous angioplasty of the right PA at 7 months of age as an initial step to stimulate growth of the hypoplastic right PA. At the time of the first surgery, it was noted that ligamentum tissue of the right patent ductal artery was connected to the proximal side of the right PA. At 2 years of age, cardiac catheterization revealed the created pericardial roll connected to the right PA as measuring 10 mm in diameter, with left PAP of 23 mm Hg and pressure ratio of the right to left ventricle of 0.27 (Fig. 2). Subsequently the patient underwent definitive surgery. Median sternotomy was carried out and the right hilar region was dissected. An autologous pericardium was harvested and an autologous pericardial tube, 10 mm in diameter, was made. Cardiopulmonary bypass was established and the right modified Blalock–Taussig shunt was ligated first. Under cardiac beating, the anastomosis of the previous autologous pericardial roll connecting the right PA to the main PA was replaced with the newly prepared autologous pericardial tube. We placed this tube behind the superior vena cava and ascending aorta in the anatomical pathway to avoid possible kinking or compression. The postoperative course was smooth, and a three-dimensional computed tomography (CT) scan 8 months after the definitive surgery showed a patent reconstructed right PA without stenosis at the anastomotic site (Fig. 3).

Discussion

Congenital isolated unilateral absence of a pulmonary artery without any associated cardiac anomalies is a rare
As the peripheral pulmonary artery exists, it is also named absence of the proximal pulmonary artery. The hypoplastic right pulmonary artery is connected to the right-sided patent ductus arteriosus (PDA) or its ligament. Unilateral absence of the PA has also been considered to be equivalent to the distal form of PA originating artery from the aorta. Reconstruction of the PA in the proximal form is accomplished by direct anastomosis, with satisfactory results. Welch et al. reported two cases of direct anastomosis of right PA to the main PA under the ascending aorta in a neonate and a 3-month-old infant. Direct anastomosis could be possible in the neonatal period or early infancy, and should be recommended if possible. However, it is frequently impossible in the distal form, because the right ductal tissue is involved at the proximal side of the aberrant PA, sufficient excision of the ductal tissue is necessary, and a prosthetic vascular graft is required, which may carry the risk of occlusion or infection. Because a size mismatch between the graft and native PA develops as the child grows, follow-up surgery for revision and/or catheter intervention can be anticipated. The reconstruction using the autologous tissue or autologous pericardium has been reported, and it may have potential to grow or could at least dilate.

Moreno-Cabral et al. reported a reconstruction case using an autologous pericardial tube in 1991, but they placed the pericardial tube in front of the superior vena cava and ascending aorta. Although a postoperative pulmonary angiogram 4 years later showed a patent new right PA that appeared to have grown with the child, the anastomotic site to the main PA appeared stenotic, probably due to compression of the ascending aorta. We placed the autologous pericardial tube behind the superior vena cava and ascending aorta and a postoperative three-dimensional CT scan showed no stenosis at the anastomotic site, which suggests that an anatomical pathway should be selected to avoid excessive tension at the anastomosis and compression by the ascending aorta. However, the long-term outcome of this procedure is unclear, and careful observation is needed in the future.