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Berry Syndrome
—— Two Cases of Successful Surgical Repair ——

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Berry syndrome is a very rare congenital cardiovascular anomaly that consists of a distal aortopulmonary window, aortic origin of the right pulmonary artery, an intact ventricular septum, a patent ductus arteriosus, and an interrupted aortic arch. Two cases of Berry syndrome are presented. A one-stage surgical correction for this complex anomaly was successfully performed, and a 1-year follow-up demonstrated favorable outcomes. (*Circ J* 2008; **72:** 492–495)

Key Words: Aortopulmonary window; Berry syndrome; Congenital heart disease; Interrupted aortic arch; Surgery

Berry syndrome is a rare congenital cardiac anomaly that consists of a distal aortopulmonary (AP) window, aortic origin of the right pulmonary artery (RPA), an intact ventricular septum, and an interrupted aortic arch with a patent ductus arteriosus (PDA). Only a few surgical cases have been reported since the first case was described by Berry et al[1] in 1982. Here, we report 2 cases of a successful 1-stage surgical correction of Berry syndrome and describe the patients' mid-term results.

Case Reports

Case 1

A 4-month-old male infant weighing 3.8 kg presented for a routine clinical work-up. On physical examination, tachypnea was noted as well as a cardiac murmur. On chest X-ray, the cardiac silhouette was slightly enlarged, and the pulmonary vasculature was engorged. An electrocardiogram demonstrated normal sinus rhythm with right ventricular hypertrophy and right-axis deviation. Echocardiography revealed a type A interrupted aortic arch, an AP window, and a PDA. It also identified the RPA arising from the right side of the ascending aorta. This anatomic feature was confirmed on chest computerized tomography (CT) scan (Fig 1).

A 1-stage surgical correction was performed through a median sternotomy. Under deep hypothermic circulatory arrest, the ascending aorta was transversely incised at the level of the AP widow, and the pulmonary arteries were disconnected from the aorta. The posterior wall of the ascending aorta was retained in order to form the confluence between the RPA and the main pulmonary artery (MPA). The anterior half of the RPA was reconstructed with a patch of autologous pericardium. Then, after removal of ductal tissue, the descending aorta was anastomosed to the aortic arch. The ascending aorta, which was divided in order to separate the pulmonary artery, was repaired with an end-to-end anastomosis (Figs 2A,B). After re-warming, the patient was easily weaned from cardiopulmonary bypass. Electively-delayed sternal closure was performed 7 days later. The subsequent hospitalization was uneventful, and the patient was discharged in good clinical condition on postoperative day 19. One year later, echocardiography and cardiac catheterization showed a satisfactory arch reconstruction. However, it also revealed stenosis of the RPA junction, with a pressure gradient of 20 mmHg. This was successfully relieved with percutaneous balloon angioplasty, and the residual pressure gradient dropped to 5 mmHg (Fig 3). As of the 18-month follow-up, the patient was growing satisfactory and has remained asymptomatic.

Case 2

A female neonate weighing 3.2 kg was born full-term to a healthy mother. The infant was referred to us because of respiratory distress. Echocardiography and a CT scan revealed the anatomic characteristics of Berry syndrome: a type A interrupted aortic arch, an AP window, and an RPA arising from ascending aorta (Fig 4). Surgery was performed on the 8th day after birth, using the same surgical technique as in Case 1 to reconstruct the interrupted aortic arch and ascending aorta. For the RPA reconstruction, the posterior wall of the ascending aorta was also used to form the posterior portion of the RPA, but in this case, the defect on the anterior aspect of the RPA was repaired by directly suturing the edges with a vertical suture line, without using any patching material (Fig 2C). The postoperative course was uncomplicated, and the baby was discharged in good clinical condition on postoperative day 27. Echocardiography at 1 year after the operation demonstrated a satisfactory aortic arch and pulmonary artery configuration without a significant pressure gradient.

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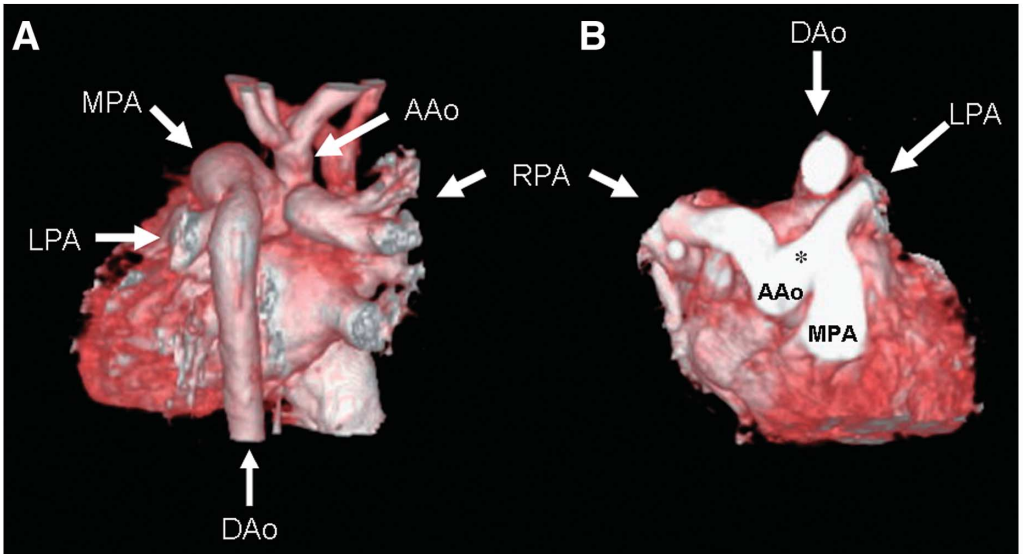

Fig 1. Three-dimensional reconstruction of the preoperative chest computerized tomography scan of Case 1. (A) Posterior view; (B) superior view with transsection at the level of the aortopulmonary window (*). The aortic arch was interrupted distal to the left subclavian artery, and the right pulmonary artery arose from the right side of the ascending aorta.

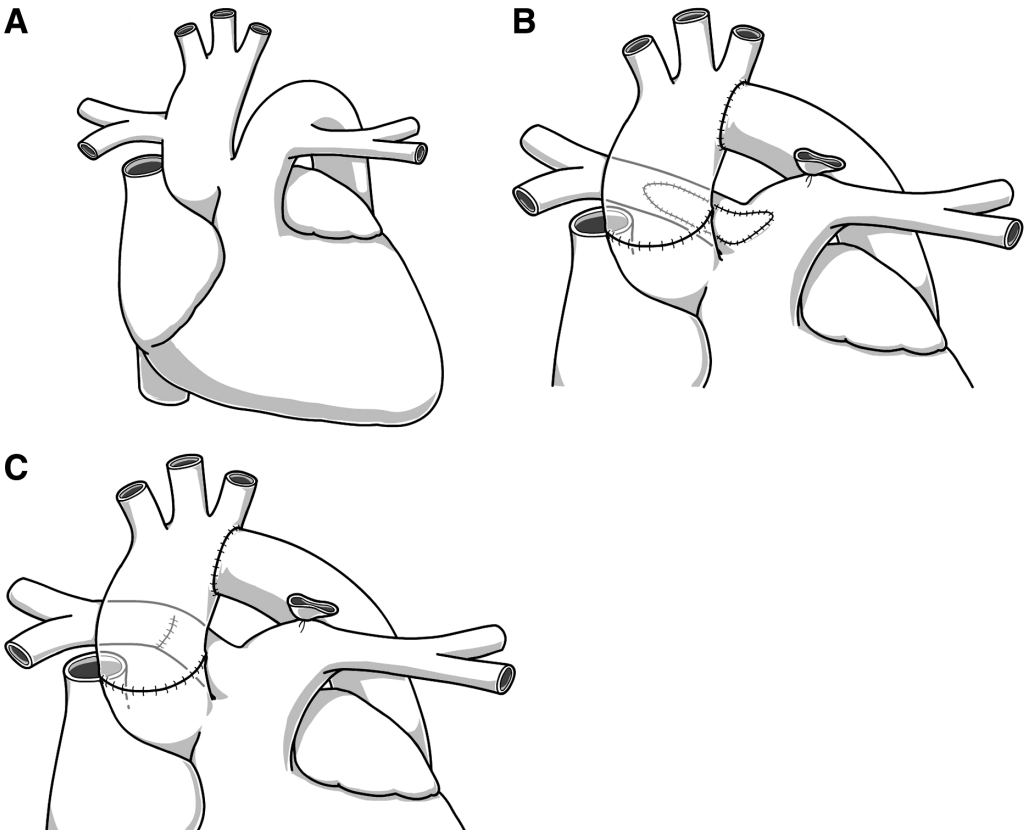

Fig 2. Diagram of the surgery. The aortic arch was reconstructed with an end-to-end anastomosis. The ascending aorta was divided, leaving its posterior wall in order to form the posterior wall of the right pulmonary artery. The anterior defect was repaired with an autologous pericardial patch in Case 1 (B), and by direct closure in Case 2 (C). The aortic arch and ascending aorta were reconstructed with an end-to-end anastomosis.

Discussion

Berry syndrome is a rare congenital cardiovascular anomaly that is amenable to surgical repair. Usually, neonates presenting with this complex anomaly are critically ill and may need stabilization and urgent surgery. Previously, surgical repair was attempted using the left subclavian artery or a synthetic graft for aortic arch repair! In some patients, surgical correction involved a staged approach[2,3] but the results have been unsatisfactory. Recently, a few

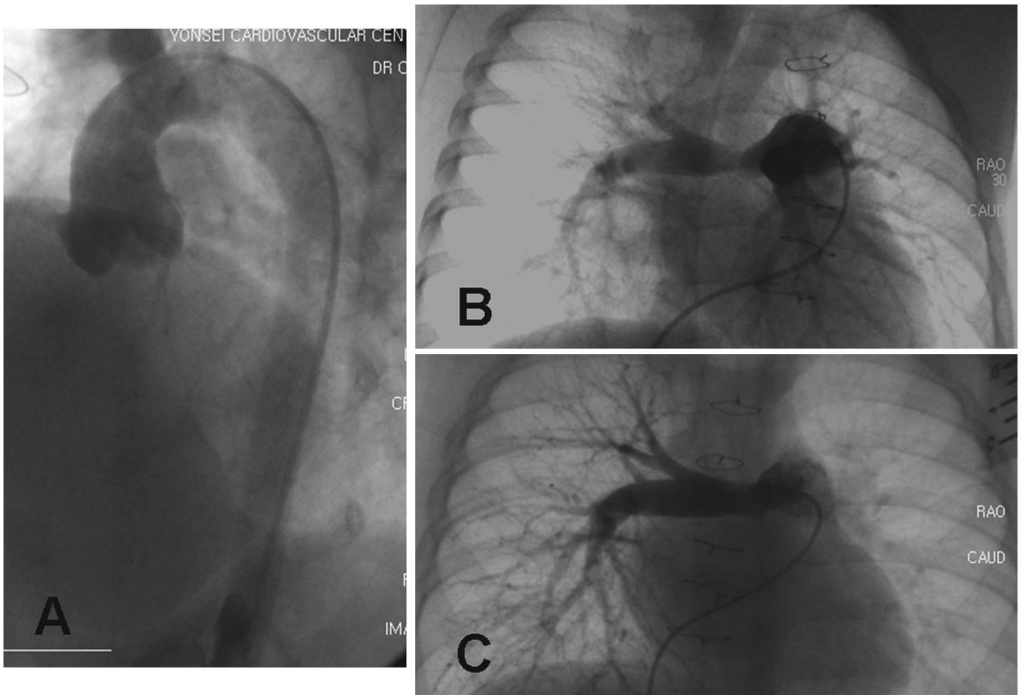

Fig 3. Aortogram (A) and pulmonary angiogram (B, C) at 1 year after surgical repair in Case 1. The ascending aorta and aortic arch demonstrate a satisfactory contour without stenosis (A). Stenosis can be seen at the right pulmonary arterial junction (B) and was successfully relieved by percutaneous balloon angioplasty (C).

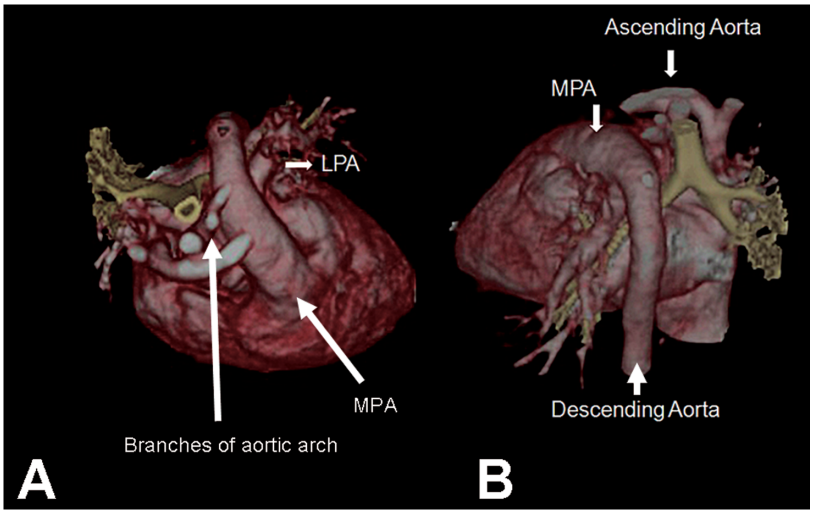

Fig 4. Three-dimensional reconstruction of the preoperative chest computerized tomography scan of Case 2. (A) Superior view; (B) Posterior view.

cases involving a 1-stage surgical correction have been reported, and the results have been favorable.[4–6] For optimal surgical outcome, accurate preoperative recognition of the complex anatomic features is mandatory. Tension-free, nonstenotic reconstruction of the aortic arch and the RPA is also essential during surgery. The aortic arch can be reconstructed by direct end-to-end anastomosis of the aortic arch and the descending aorta. The continuity between the RPA and MPA can be obtained by excising the RPA from the ascending aorta and attaching it to the MPA.[7] In the present cases, the posterior wall of the ascending aorta was used to form the posterior portion of the confluence between the RPA and the MPA. In Case 1 the anterior half of the RPA was repaired with an autologous pericardial patch, which was not needed in Case 2. Considering the growth potential, we believe that our method has advantages over the reimplantation method, because the circumferential suture lines and tension on the anastomosis can be avoided using our technique. At the 1-year follow-up, each of these cases showed just a mild pressure gradient across the RPA junction (20 mmHg for Case 1, and 7 mmHg for Case 2). The repaired aortas showed no stenosis on follow-up.

In summary, Berry syndrome can be successfully corrected without using any patching material. Careful follow-up is mandatory because stenosis at the site of the aortic reconstruction and the RPA is a potential problem. It is possible to treat pulmonary artery branchial stenosis adequately by percutaneous balloon angioplasty.

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