One-Stage Surgical Repair for Berry Syndrome With Preoperative Diagnosis by 3-Dimensional CT

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Abstract
Berry syndrome is a rare congenital combination of an aortopulmonary window, an aortic origin of the right pulmonary artery, an interrupted aortic arch with a patent ductus arteriosus, and an intact ventricular septum. We report a successful one-stage surgical correction of Berry syndrome. Also, we demonstrate the importance of prompt clinical recognition with echocardiography and 3-dimensional reconstruction of computed tomography (3D-CT) and timely operation for the management of this rare cardiac anomaly.

Keywords
aortopulmonary window, interrupted aortic arch, patent ductus arteriosus, right pulmonary artery, Berry syndrome

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Introduction
Berry syndrome is a very rare congenital cardiac anomaly. It consists of an aortopulmonary (AP) window, an anomalous origin of the right pulmonary artery (RPA) from the ascending aorta, an interrupted aortic arch (IAA) with a patent ductus arteriosus (PDA), and an intact ventricular septum. Since the first case was reported by Berry et al in 1982,¹ most repairs were performed in two-stage operations, but one-stage repair has been reported recently. We also experienced this rare case, and successful one-stage repair was done in a 10-day-old neonate. This is the 31st case in the literature. Recognition of all the features by echocardiography remains difficult, therefore we present the accuracy and effectiveness of preoperative 3-dimensional computed tomography (3D-CT) for diagnosis.

Presentation
A 5-day-old female neonate was referred to our hospital with respiratory deterioration and circulatory dynamics instability. The perinatal history was unremarkable and at a previous hospital she was born at 38 weeks and 4 days of gestation by scheduled cesarean section with a birth weight of 2759 g and an Apgar score of 9/9. On the fifth day postpartum she presented with severe cyanosis and congestive heart failure and was referred to us for possible surgical repair.

On physical examination, she was tachypneic with retic- tive breathing while differential O₂ saturation was noted between the upper and lower limbs (90% and 70%, respectively). No apparent cardiac murmur was detected. Her chest X-ray showed mild cardiomegaly and decrease in lung field permeability and an electrocardiogram demonstrated right ventricular hypertrophy with right axis deviation. Echocardiography revealed an AP window and IAA (type A) with a PDA but no ventricular septal defect (Figure 1). On 3D-CT, we identified further details of this anatomic feature clearly; 8-mm AP window, type A IAA (defect of arch) and we confirmed the RPA arising from the origin of the ascending aorta and the descending aorta connected with dilated PDA (Figure 2).

After admission, prostaglandin and diuretics were administered. On the 10th day of birth, a one-stage surgical correction was performed through a median sternotomy under profound hypothermia (25°C). Cardiopulmonary bypass was established by the right hemicerebral perfusion and the bicaval drainage. The right hemicerebral perfusion was obtained via expanded polytetrafluoroethylene (ePTFE) graft sewn to the right brachiocephalic artery. The aortic cross clamp was applied and cardioplegia (St Thomas blood cardioplegia) was infused.

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Aortic arch vessels were clamped at the origin of the brachiocephalic artery, the left common carotid artery, and left subclavian artery. The ascending aorta was longitudinally incised. Inspection confirmed the location of the AP window as well as aortic origin of the RPA. The left pulmonary artery (LPA) arose from the main pulmonary trunk in the normal fashion. A patch of autologous pericardium treated with glutaraldehyde was used to establish continuity between the AP window and the RPA, effectively partitioning the pulmonary arteries from the aorta (Figure 3A). After the removal of ductal...
tissue, an end-to-side anastomosis of the descending aorta to the
distal ascending aorta was performed with a continuous 7-0 poly-
propylene suture (Figure 3B). After rewarming, cardiopulmonary bypass was easily weaned with moderate inotropic support. Total bypass time was 1 hour 59 minutes and a cross clamp time was 43 minutes. Peritoneal dialysis was performed postoperatively for 2 days and electively delayed sternal closure was performed 4 days later. Subsequent hospitalization was uneventful, and she was discharged in good clinical condition and has been followed up at outpatient clinic with reasonable growth.

Discussion

Berry syndrome is a subtype of an IAA which is occasionally associated with AP window. This congenital cardiac anomaly requires early diagnosis and timely surgical repair in order to restore normal perfusion especially for the lower body and to prevent the pulmonary damage. Most neonates may not tolerate an invasive examination such as angiography; but echocardiography, used instead, has limitations for a complete diagnosis, since it often does not allow the detection of an abnormal RPA origin. In previous reports, they only found abnormal RPA origin after opening pericardium at operation.2-4 We believe that 3D-CT clearly demonstrates the positional relationship of every anomaly and helps construct a more precise surgical image. It exactly shows RPA originating from the ascending aorta as well as the AP window and IAA.

As for the surgical technique, a two-stage or one-stage repair could be adopted for this cardiac anomaly. In the two-stage approach, closure of AP window comes electively after the reconstruction of aorta with ligation of PDA,5 while one-stage surgery has all these corrections in a definitive operation.

One definitive repair is obviously superior to staged procedures if a patient’s condition is endurable for the surgery, and its results have become favorable without any critical complications these days.2-4,6-8

The one-stage repair includes reimplantation of the RPA or patch closure. We adopted the latter since the distance between RPA and LPA was relatively short. Also, considering the growth potential, drawback from autologous pericardial patch technique is obviously shrinkage to cause stenosis. Excision of the pulmonary artery from the ascending aorta and then putting a patch into the defect could reduce a potential problem. We did not use this reimplantation technique since avoiding prolonged ischemic insult in small baby is the first priority.

Careful follow-up is necessary since stenosis at the anastomosis of aorta is also a potential complication, which can be treated adequately by balloon angioplasty.

In summary, we report a case of Berry syndrome which was definitively diagnosed by 3D-CT and successfully repaired in a one-stage procedure with autologous tissue allowing future growth.

Declaration of Conflicting Interests

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