

Staged Repair of Truncus Arteriosus Associated with Complete Atrioventricular Septal Defect

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We report a case of successful repair of truncus arteriosus (TA) associated with complete atrioventricular septal defect (c-AVSD) using a staged approach. TA associated with c-AVSD is a very rare congenital cardiac anomaly. No report of successful staged repair in South Korea has yet been published. We performed bilateral pulmonary artery banding when the patient was 33 days old, and total correction using an extracardiac conduit was performed at the age of 18 months. The patient recovered uneventfully and is doing well.

Key words: 1. Truncus arteriosus
2. Complete atrioventricular septal defect
3. Staged operation

Case report

A male infant was born after 41 weeks of gestation, weighing 3.69 kg. Prenatal echocardiography showed truncus arteriosus (TA) and a balanced complete atrioventricular septal defect (c-AVSD). He had cyanosis and did not cry initially after birth; his Apgar scores were 4 at 1 minute after birth and 6 at 5 minutes. The patient underwent endotracheal intubation and ventilator care at a neonatal intensive care unit, and was extubated from the ventilator the following day. His oxygen saturation on room air was around 90%. The patient was diagnosed with CHARGE syndrome (coloboma, heart defect, atresia choanae, retarded growth and development, genital hypoplasia, ear anomalies/deafness) accompanied by abnormalities of the ear, urinary tract, and heart. A

transthoracic echocardiogram (TTE) and computed tomography (CT) angiography confirmed that the patient had type III TA with a truncal valve arising mainly from the right ventricle (RV) and a balanced c-AVSD with an unrestrictive ventricular septal defect (VSD) (Fig. 1A, Fig. 2A). CT angiography also showed a right aortic arch and aberrant left subclavian artery. At 33 days after birth, bilateral pulmonary artery (PA) banding was performed using a strip cut from a 3.5-mm polytetrafluoroethylene (PTFE, Gore-Tex; W. L. Gore and Associates, Flagstaff, AZ, USA) tube graft. The infant's systolic blood pressure increased from 61 to 80 mm Hg. On the second day after PA banding, he was successfully weaned from the ventilator.

Second-stage total correction was initially scheduled between the ages of 6 months and 1 year.

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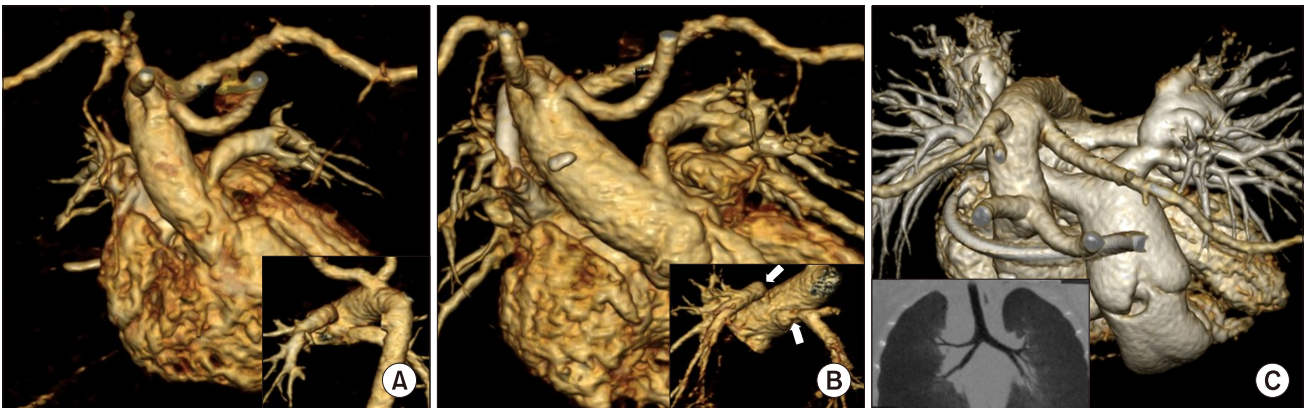


Fig. 1. CT angiography scans shows initial truncus arteriosus type III (small box, posterior view of the great arteries) (A), findings after bilateral pulmonary artery banding (small box, posterior view of the great arteries; white arrows indicate the stenotic area of the pulmonary arteries) (B), and postoperative findings without significant kinking or malposition of the inserted conduit (small box, a patent airway after the operation) (C). CT, computed tomography.

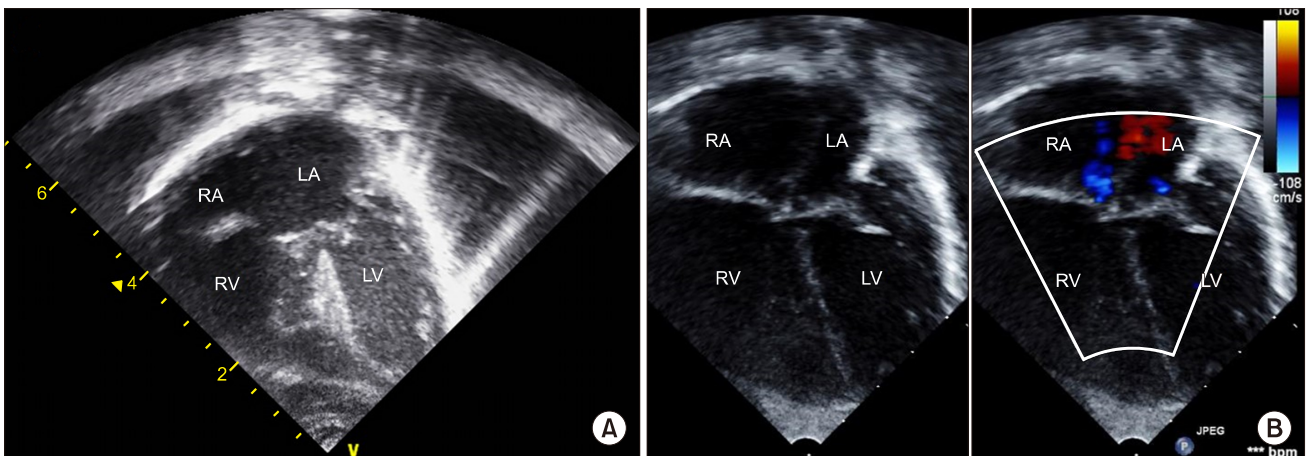


Fig. 2. A preoperative 2-dimensional echocardiogram demonstrates a common atrioventricular valve with a balanced ventricle and an unrestrictive ventricular septal defect (A). After total correction, there are no residual ventricular septal defect and no other problems except trivial left atrioventricular valvular regurgitation (B). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

However, the operation was delayed and performed at 17 months after PA banding due to recurrent upper respiratory infections and thrombocytopenia, of which we were not able to identify any special cause. At admission, his percutaneous oxygen saturation was 81%. TTE and CT angiography revealed stenosis of both PA banding sites (Fig. 1B). His body weight was 8.03 kg, which was below normal for his age. The sternum was reopened and both PAs were extensively mobilized from the truncus. Division and patch angioplasty of both pulmonary arteries were performed under cardiopulmonary bypass with a beating heart. After temporary snaring of the branch

PAs, the truncal artery was partially clamped using a side-bite clamp, and the right and left PAs were divided from the truncal artery to achieve PA confluence. The PTFE tapes used for PA banding were removed, and the stenotic banding sites were dilated with a balloon. After completion of the balloon angioplasty, we made incisions at the anterior surfaces of both pulmonary arteries, from the sites of division to near the branching portion of the PAs, crossing the stenotic sites. The posterior vessel walls of the 2 divided PAs were directly anastomosed to each other, and the anterior incisions of both PAs were covered with a long bovine pericardial patch.

After creation of the confluence and patch angioplasty of the PAs, we made a longitudinal incision on the bovine pericardial patch and connected a 14-mm Contegra valved conduit (Medtronic Inc., Minneapolis, MN, USA) to the incision for right ventricular outflow tract (RVOT) reconstruction with a beating heart. The remaining truncal artery was clamped and cardioplegic solution was infused. An oblique right atrial incision and a longitudinal right ventricular incision were made along the atrioventricular (AV) groove and at the RV free wall (approximately 5 mm below the truncal valve, 15 mm in length), respectively. The c-AVSD was a Rastelli type C defect with large inter-ventricular communication. The 2-patch technique was applied for intracardiac repair of the c-AVSD. Closure of the VSD, including intraventricular tunneling to the aorta, was performed using both atrial and ventricular approaches. A guiding stitch was made at the superior and inferior bridging leaflets. The VSD was located just below the superior bridging leaflet, and the truncal valve arose exclusively from the RV. The VSD closure and intraventricular tunneling were performed using a large comma-shaped bovine pericardial patch through the AV valve and right ventriculotomy site. The division of common AV valve with closure of interatrial communication was then performed. The coronary sinus was not definitely identified. The cleft of the left AV valve created was repaired with multiple interrupted sutures. After the division of the common AV valve and intraventricular tunneling were completed, the other end of the adequately trimmed Contegra valved conduit was anastomosed to the right ventriculotomy. Trans-esophageal echocardiography showed no leakage from the VSD, trivial left AV valve regurgitation, and no RVOT obstruction. The cardiopulmonary bypass time was 302 minutes, and the aortic cross-clamp time was 157 minutes.

On the second day after the operation, the patient was successfully weaned from the ventilator and subsequently discharged on the 10th postoperative day without postoperative complications. No PA obstruction was evident on a CT scan obtained 1 month after the operation (Fig. 1C).

After total correction surgery, the patient is doing well, with no need for additional surgery or hospitalization because of heart problems. TTE has confirmed good ventricle contractility, and no apparent

problems other than trivial left AV and aortic valvar regurgitations remain (Fig. 2B).

Discussion

The coexistence of both AV and ventriculoarterial junction abnormalities is rare [1,2]. The combination of TA and c-AVSD probably results from a failure of the embryologic process of septation at both the AV and ventriculoarterial junctions [2]. Only 13 patients with both deformities have been reported [3]. To date, no report of successful repair in South Korea has appeared, and only 2 operative survivors have been described worldwide [4,5].

In patients with TA and c-AVSD, neonatal management is difficult; it is essential to balance systemic and pulmonary circulation [2]. To overcome this problem, total correction in the neonatal period should be considered. Sousa-Uva reported successful complete repair of this anomaly in the neonatal stage [5]. However, single-stage total correction for this anomaly is technically demanding. When considering neonatal complete repair, we have to face a very friable AV valve and the need of a small extracardiac conduit to achieve competent RVOT valve function which is critical for TA and c-AVSD repair. We thought that it would be too difficult to achieve a competent AV valve function from a friable AV valve and to get a small homograft or other small valved conduit for the small heart in our country. Thus, we chose a staged repair strategy.

c-AVSD repair is generally not recommended during the neonatal period unless a patient is experiencing severe congestive heart failure because of the fragility of the neonatal AV valve. Additionally, the competence of the RVOT conduit is important in the repair of this anomaly because there is a possibility of residual right AV valve regurgitation after c-AVSD repair, which imposes further RV volume loading. Furthermore, bilateral PA banding is commonly used to palliate hypoplastic left heart syndrome. We were able to adequately control pulmonary blood flow using bilateral PA banding in the neonatal period, allowing total correction to be safely delayed, and we successfully performed the total correction later.

Conflict of interest

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

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