Double-outlet right ventricle with complete atrioventricular canal
Michiaki Imamura, Jonathan J. Drummond-Webb, George E. Sarris, Daniel J. Murphy, Jr and Roger B.B. Mee

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A female infant was transferred to our hospital for evaluation at a few hours of age. The infant presented with severe congestive heart failure, a murmur, and mild cyanosis. Echocardiography revealed DORV with subpulmonary VSD, unbalanced complete AV canal, large patent ductus arteriosus, severe AV valve regurgitation, a hypoplastic aortic arch, and coarctation of the aorta. The left ventricle/right ventricle cross-sectional area ratio was 1.86, and the left ventricle/right ventricle area ratio from an apical four-chamber view was 1.40 [4]. Administration of prostaglandin E1, and dobutamine was commenced. At this time, right ventricular hypoplasia appeared to preclude biventricular repair.

When the patient was 4 days of age, a palliative procedure was performed via midline sternotomy under cardiopulmonary bypass with profound hypothermia and circulatory arrest. Aortic arch repair, atrial septectomy, common AV valve regurgitation repair, and pulmonary artery banding were performed. Operative assessment documented a patent foramen ovale and a partially formed septum primum, and all pulmonary veins entering the left atrium to the left of a normal looking interatrial septum. The anatomy of the triangle of Koch and position of the coronary sinus appeared normal in its relationship with the septum primum and patent foramen ovale. The common AV valve arose entirely from the right atrium. The nomenclature is difficult. From the AV ring downward the AV valve looked exactly like a Rastelli type C AV canal. Some may wish to call this a straddling tricuspid valve (Fig 1). The aorta, which was anterior and to the right of the main pulmonary artery, was remote from the VSD and both the aorta and pulmonary artery originated from the right ventricle. The superior and inferior common leaflets were sewn together in the middle of the common AV valve with interrupted sutures and a thin polytetrafluoroethylene strip was sutured across the common AV valve to the annulus superiorly and inferiorly with pledged horizontal mattress sutures. Postoperative recovery was complicated by oliguria requiring 24 hours of peritoneal dialysis. Postoperative echocardiography showed moderate to severe AV valve regurgitation. Because of heart failure gastrointestinal intolerance precluded successful oral feeding, and she was discharged with nasogastric tube feeding 23 days after operation.

Repeat evaluation at age 14 months by cardiac catheterization and echocardiography showed a DORV with anterior aorta, complete AV canal, subpulmonary VSD, no subaortic stenosis, no residual coarctation, moderately severe AV valve insufficiency, adequately sized right and left ventricle with good biventricular function, and moderate proximal left pulmonary artery stenosis (Fig 2). The left ventricle/right ventricle cross-sectional area ratio was 1.40, and the left ventricle/right ventricle area ratio from arch, and coarctation of the aorta. After neonatal palliation, successful biventricular repair involving two-patch complete AV canal repair and arterial switch operation for DORV were successfully performed when the patient was 14 months of age.

References


Double-Outlet Right Ventricle With Complete Atrioventricular Canal

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A case of double-outlet right ventricle, unbalanced complete atrioventricular canal, hypoplastic aortic arch, coarctation of the aorta, and atrioventricular valve regurgitation was treated with a staged approach. At 4 days of life the patient underwent aortic arch repair, atrial septectomy, common atrioventricular valve regurgitation repair, and pulmonary artery banding. When she was 14 months of age a biventricular repair was accomplished by two-patch complete atrioventricular canal repair and arterial switch procedure.

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Double-outlet right ventricle (DORV) can be associated with many other congenital heart defects [1, 2]. Double-outlet right ventricle with complete atrioventricular (AV) canal is an uncommon congenital heart defect, and the ventricular septal defect (VSD) in this instance is usually subaortic [3]. We present a case of DORV and subpulmonary VSD in combination with complete AV canal with AV valve regurgitation, hypoplastic aortic arch, and coarctation of the aorta. After neonatal palliation, successful biventricular repair involving two-patch complete AV canal repair and arterial switch operation for DORV were successfully performed when the patient was 14 months of age.
the apical four-chamber view was 1.23 [4]. At this time surgical options included a bidirectional Glenn shunt with a left pulmonary artery repair and further AV valve repair, or a biventricular repair depending on the feasibility of channeling left ventricle blood to the pulmonary artery. After a repeat sternotomy and hypothermic cardiopulmonary bypass with cardioplegic arrest, the right atrium was opened obliquely and the strip of polytetrafluoroethylene that had been used previously to join the two bridging leaflets of the common AV valve was removed. The anatomy was Rastelli type C. The absence of chordal attachments to the superior edge of the VSD allowed the performance of a biventricular repair. The VSD, which had prominent outlet extension, was closed with a comma-shaped knitted Dacron patch, which was sewn around the pulmonary valve annulus and along the septal crest with interrupted, pledgeted horizontal mattress sutures (Fig 3). The patch was trimmed in line with the plane of the common AV valve, and horizontal mattress sutures were placed to sew the patch to the superior and inferior common leaflets. The cleft of the left-sided AV valve was closed with interrupted sutures. The now almost common atrium was septated with a polytetrafluoroethylene patch, with the plane of the original interatrial septum lying entirely to the left of the patch. Inspection of the coronary artery anatomy revealed that the right coronary artery arose from the right sinus (sinus 2) and the left anterior descending coronary and circumflex arteries were found arising from the left sinus (sinus 1). Both great arteries were transected and the right and the left coronary arteries were translocated to the proximal main pulmonary artery. The Lecompte maneuver was carried out and end-to-end anastomosis of proximal main pulmonary artery to distal ascending aorta was performed. The aortic sinus defects were repaired with two separate autologous pericardial patches. The proximal aorta was anastomosed end-to-end to the distal main pulmonary artery, with a pericardial patch used to repair the stenotic origin of the left pulmonary artery. Postoperative recovery was uncomplicated. Echocardiography before hospital discharge on postoperative day 6 showed only mild left- and right-sided AV valve regurgitation and good biventricular function. The patient remains well 18 months after the operation.
Comment

Double-outlet right ventricle with complete AV canal is a rare congenital cardiac anomaly [1, 2]. Bharati and associates [3] reported 34 cases of DORV in 507 cases of complete AV canal (6.7%). In the Royal Children’s Hospital experience, DORV was observed in 3.1% of patients with complete AV canal and, conversely, complete AV canal was observed 4.3% of patients with DORV [5].

Recently, several groups have reported that concomitant repair of the aortic arch and the associated lesions is feasible and gives better results [6, 7]. One-stage biventricular repair of DORV especially in the presence of aortic arch abnormalities is advocated. In this case the small right ventricle precluded initial biventricular repair, and it seemed likely that this patient would have been best staged toward a Fontan-type procedure.

Several centers have reported repair of DORV with complete AV canal with use of a patch sewn into place to close the interventricular communication in such a way as to conduct left ventricular blood to the aorta [5, 8]. In these reports, the diagnosis was DORV with subaortic VSD. In this case the VSD was subpulmonic, and blood flow from the left ventricle could not be directed to the aorta through the VSD. The VSD was channeled to connect to the pulmonary artery, and a concomitant arterial switch operation was performed. In Bharati and associates’ [3] series of 34 cases of DORV with complete AV canal, only 1 case (3%) had a subpulmonic VSD.

At the first staged operation bivalvaton with bridging for common AV valve regurgitation was performed [9]; however, moderate to severe AV valve regurgitation persisted. At reoperation, biventricular repair was feasible, and AV valve regurgitation was largely eliminated, the two patches for atrial and ventricular septal defect providing secure support for the inferior and superior bridging leaflets.

References


Surgical Resection of Lung Cancer Originating in a Tracheal Bronchus

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We experienced a case of lung cancer that developed from a tracheal bronchus in an 80-year-old man. The tumor was completely resected by right upper lobectomy and resection of the tracheal bronchus as well as dissection of the mediastinal lymph nodes. Postsurgical pathologic staging was stage IB (T2 N0 M0) adenocarcinoma.

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Tracheal bronchus (also called “eparterial bronchus”) is an aberrant bronchus that usually arises from the right lateral wall of the trachea [1]. Although other congenital anomalies, such as laryngeal web or hypoplastic or fused first and second thoracic ribs, are occasionally associated with the tracheal bronchus [2], most of the cases are asymptomatic and detected incidentally by bronchoscopy or radiologic examination. We could find only 2 case reports [3, 4] of malignant tumor that developed from the tracheal bronchus, neither of which was resected. We are reporting a case of adenocarcinoma that developed from the tracheal bronchus.

An 80-year-old man was referred to our hospital for the evaluation of incidentally detected right upper lung mass. He complained of productive cough, mild dyspnea, and weight loss of 4 kg for 6 months. He had a history of pulmonary tuberculosis and hepatitis when he was young. He was a smoker (half pack a day) for more than 20 years, but quit smoking 30 years ago.

A simple chest roentgenogram showed increased density in the right upper lung field. Computed tomography revealed a 6-cm lobulating mass in the right upper lobe near the right tracheal border. Linear translucency in the mass, which was posterolateral to the trachea, suggested
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