(ePTFE) graft. The straddling mitral papillary muscle was incorporated in the left side of the rerouting patch without translocation by suturing the patch onto the RV free wall over the thin anterior margin of the VSD and around the right side of the straddling papillary muscle. The large ASD was closed using an ePTFE patch. After the aortic clamp was released, pulmonary valve commissurotomy and leaflet shaving were performed, and the pulmonary trunk was reconstructed using an end-to-end anastomosis. The gap between the RV outflow hole and the translocated pulmonary valve was supplemented with a cut-open ePTFE graft. The patient was weaned from cardiopulmonary bypass and the chest was closed primarily. Total pump time was 375 minutes and cross-clamp time was 255 minutes. His postoperative course was uneventful. Postoperative echocardiography and computed tomography showed laminar flow through both the LVOT and right ventricular outflow tract (RVOT) without obstruction. The patient has been followed for 20 months and is doing well. Echocardiography and catheterization at 10 months after operation showed normal ventricular function, trivial aortic regurgitation, mild PR, mild mitral regurgitation, no LVOT or RVOT obstruction, and normal RV pressure.

Comment

The Rastelli procedure [1] has been widely applied to biventricular repair of TGA with VSD and PS or DORV with TGA and PS. However, its long-term results have not been satisfactory because of LVOT obstruction, conduit obstruction, arrhythmias, and a surprisingly low long-term survival [3]. Moreover, the Rastelli procedure is contraindicated in the presence of a straddling mitral valve, for which univentricular repair is usually performed [2, 4]. In addition, a distant aorta resulting from DORV results in a long intracardiac tunnel. In these cases, aortic translocation procedures [5] are good alternative techniques for biventricular repair because of a relatively small RV volume reduction. A high incidence of serious complications such as coronary insufficiency and late RVOT obstruction [6] were reported for the original aortic translocation procedure by Nikaidoh [5]. To solve these problems, half-turned truncal switch and en bloc truncal switch procedures have recently been reported, with good early results [4, 7, 8]. Our patient had mild pulmonary valvular and subvalvular stenosis with anormalized annulus. It was possible to use the pulmonary valve for the RVOT with commissurotomy and leaflet shaving, thereby preserving valvular function and growth potential. We believe the en bloc truncal switch procedure is the best option to accomplish biventricular repair in cases of DORV with TGA, straddling mitral valve, and mild PS with a normalized-sized pulmonary annulus.

Reference


Surgical Repair of Aortoventricular Tunnel Connected to the Apex of the Right Ventricle in a Neonate

Dmitry Bobylev, MD, Masamichi Ono, MD, PhD, Anneke Neumann, MD, Harald Bertram, MD, and Alexander Horke, MD

Divisions of Cardiothoracic, Transplantation, and Vascular Surgery and Pediatric Cardiology and Intensive Care Medicine, Hannover Medical School, Hannover, Germany

We describe a neonate in whom early symptoms of heart failure developed because of a rare aorta–right ventricular tunnel connected to the apex of the right ventricle. Successful immediate surgical repair was carried out with closure of both ends of the tunnel.


Aortoventricular tunnel is a rare congenital malformation, which can lead to heart failure in neonates. This extracardiac channel may connect the ascending aorta above the sinotubular junction to the cavity of the left or right ventricle. Only 17 of cases of aorta–right ventricular tunnel (ARVT) have been reported in the

Accepted for publication June 10, 2013.

Address correspondence to Dr Bobylev, Division of Cardiothoracic, Transplantation, and Vascular Surgery, Hannover Medical School, Carl-Neuberg-Str. 1, 30625 Hannover, Germany; e-mail: bobylev.dmitry@ mh-hannover.de.

© 2014 by The Society of Thoracic Surgeons Published by Elsevier Inc
literature. In most cases of ARVT, the tunnel entered the outflow of the right ventricle. We present a case of ARVT connected to the apex of the right ventricle, which was successfully repaired in a neonate.

A 3-day-old girl weighing 2,370 g, without a complicated antenatal history, was admitted with a murmur and tachypnea present since birth. Cardiac auscultation revealed a troubled precordium with grade 3/6 holosystolic murmur at the upper left sternal border. Chest roentgenography showed severe cardiomegaly and pulmonary plethora. Transthoracic echocardiography showed a large fistulous tunnel arising from the left coronary sinus suggesting AVRT (Fig 1). A small muscular ventricular septal defect and a large atrial septal defect were also verified. An ostium of the right coronary artery was identified. However, the origin of the left coronary artery was not visualized. Continuous positive airway pressure ventilation and diuretic therapy were initiated.

To confirm the exact coronary artery, cardiac catheterization was performed; retrograde aortography (Figs 2A, 2B) and selective injection of contrast material proximal to the orifice of the tunnel showed a tunnel with a length of 42 mm and a width of up to 12 mm connected to the apex of the right ventricle (Figs 2C, 2D). A dominant right coronary artery was seen, but the left coronary artery could not be clearly visualized.

Immediate surgical intervention was performed through a median sternotomy. The right atrium and right ventricle were enlarged. Aortic and bicaval cannulation was performed, and cardiopulmonary bypass with moderate hypothermia (28°C) was established. A giant communication in diverticulum form was noted arising from the dilated aortic root in the left coronary artery position to the apex of the heart (Fig 3A). After cross-clamping the aorta, myocardial protection was achieved with cold blood cardioplegia through the aortic root. A transverse aortotomy was performed. The aortic cusps had a normal structure and configuration. The right coronary artery orifice was in the normal location; the left coronary artery orifice could not be identified. The left coronary sinus was markedly dilated and had multiple fenestrations (Fig 3B). After transverse incision of the pulmonary artery, we could follow the “neck” of the tunnel to the aneurysmal sac, and the tunnel was found to open into the largest area. A coronary guide was introduced through the hole in the left coronary sinus, and we identified communication between the aorta and the aneurysmal sac (Fig 3C). The distal end of the tunnel that connected to the apex of the right ventricle was easily identified. This tunnel did not have any additional holes that would have suggested the presence of the coronary ostium. After this, we performed closure of the proximal and distal parts with polytetrafluoroethylene patches (Fig 3D). The rest of the tunnel was obliterated with a running suture. The entrance to the tunnel in the left coronary sinus was also secured with a single suture external to the aortic polytetrafluoroethylene patch. Because the left coronary artery orifice was composed of multiple fenestrations in the left coronary artery sinus, retrograde cardioplegia was administered through the coronary sinus, with good flow seen in the coronary arteries. In addition, a large atrial septal defect was closed directly, but a small muscular ventricular septal defect was not closed. The aorta and pulmonary artery were reanastomosed and the right atriotomy incision was sutured. Weaning from bypass was performed without any problems, and no changes in the electrocardiogram were encountered. The patient was extubated on postoperative day 2 and discharged from the hospital when completely recovered.

Comment

ARVT is a rare entity thought to represent abnormal formation of the arterial valvular sinuses and leaflets during embryonic development of the outflow tracts [1], with only a few cases reported in the literature [2–7]. The cause of aortoventricular tunnel is unknown. Histologically, the arterial end of the tunnel resembles the aorta with fibrous tissue, elastic fibers, and smooth muscle cells [5]. Within the tunnel itself, there may be a well-defined junction between ventricular and arterial components in addition to cystic or membranous structures of valve leaflets [4]. The ostium of the coronary artery may lie within an aortoventricular tunnel, and absence of the origin of the left coronary artery has been observed with this anomaly [2]. In our patient, abnormal development of the left coronary artery and absence of the left coronary ostium were seen. For a successful operation, identification of the exact anatomy of the coronary arteries is critical.

The most remarkable fact regarding this case is that the tunnel had a long extension into the apex of the right ventricle and an atypical muscular sac formation. In most ARVT cases in the literature, the tunnels were relatively short and entered the right ventricle infundibulum. Vargas and colleagues [3] described a case of ARVT connected in the trabecular portion of the right ventricle.
close to the apex [3]. However, in this case, the long tunnel consisted of the small vessel-like part and the aneurysmal muscular sac part, which connected to the apex of the right ventricle. The tunnel appeared to extend beyond the ventriculoarterial junction and penetrate the ventricular septal structures. However, the embryology of this tunnel is unclear.

Corrective operations should be undertaken in the early neonatal period or rapidly after symptoms emerged. Repair consists of closing the tunnel. In most cases in the

---

**Fig 2.** Still images of angiographic procedure. (A) Retrograde aortography is performed with selected contrast injection into (B) right coronary artery and into (C, D) tunnel. White arrow shows an aneurysmal communication between the aorta and right ventricle. (Ao = aorta; RCA = right coronary artery; RV = right ventricle.)

**Fig 3.** Intraoperative photographs as seen from the surgeon’s view. (A) After establishment of cardiopulmonary bypass, heart was rotated and tunnel was identified. (B) Left coronary sinus had multiple fenestrations. Tunnel was opened. (C) White arrow shows coronary guide, which was introduced through the entrance of the tunnel in the left coronary sinus. (D) Closure of proximal and distal parts of tunnel with polytetrafluoroethylene patches. (Ao = aorta; PA = pulmonary artery; RV = right ventricle; * = tunnel.)
literature, this has been performed by closure of both ends of the tunnel by transaortic and transventricular access. However, closure of just the ventricular end is an option to maintain coronary perfusion [6].

In conclusion, we present a neonate with a rare aorta-right ventricle tunnel connected to the apex of the right ventricle. Surgical closure of both ends of the ARVT was successfully performed.

The authors thank Mr Vi Tran (Institute of Cardiovascular Science, UCL, London) for his comments on the writing of this article.

References


Tailored Strategies for Staged Management of Complex Congenital Cardiac Lesions

Yishay Orr, MBBS, PhD, and Richard B. Chard, BDS, MBBS

The Heart Centre for Children, Westmead, New South Wales, Australia

Although many complex congenital cardiac lesions can be primarily repaired, there remain circumstances where a staged approach may carry lower risk than a primary repair. We present 4 clinical cases where stable intermediate stages allowed somatic growth and facilitated successful biventricular repair or univentricular palliation. (Ann Thorac Surg 2014;97:1436–8) © 2014 by The Society of Thoracic Surgeons

The choice of complete primary repair for complex congenital cardiac disease versus a staging strategy is usually straightforward; most biventricular lesions are now repaired primarily. In some cases, subtleties of lesion complexity such as hypoplastic pulmonary arteries or concomitant noncardiac lesions such as tracheal stenosis increase perioperative risk [1, 2]. A staged approach that addresses immediate physiologic and anatomic concerns to allow a period of growth and stability is more appropriate in these cases and allows subsequent complete repair at a more acceptable risk. Univentricular lesions with issues such as transposition streaming and high pulmonary vascular resistance need optimal conditions for pulmonary vascular resistance to decrease. The selection and application of staging strategies is case specific. We present 4 cases where interlim palliative strategies were designed to enable somatic growth to mitigate unfavorable anatomy and physiology.

Case Reports

Patient 1

A male infant presented with the following univentricular anatomy: d-transposition of the great arteries (d-TGA), right ventricular hypoplasia, straddling right atrioventricular valve, nonrestrictive ventricular septal defect (VSD), and excess pulmonary blood flow. He underwent neonatal balloon atrial septostomy and pulmonary artery banding at age 3 months in another institution. Cyanosis limited band tightening due to transposition streaming; consequently, pulmonary blood flow remained excessive. At age 8 months, his pulmonary vascular resistance index was 10 Wood units m2 excluding Fontan candidacy. He was then referred to our unit. We opted to perform atrial septectomy, palliative arterial switch operation to correct the transposition streaming, and restricted pulmonary blood flow by placing a 6-mm polytetrafluoroethylene interposition graft between the neo-pulmonary root and the distal pulmonary artery as a modified banding technique. The postoperative course was uncomplicated and systemic saturations were 75% to 85% during the intervening months. Six months later his pulmonary vascular resistance index was 2.7 Wood unit m2. He proceeded to creation of a bidirectional superior cavopulmonary anastomosis at age 20 months and subsequently completed his total cavopulmonary connection at age 3 years. He remains asymptomatic at 3 years follow-up.

Patient 2

A 3-kg female presented with d-TGA and multiple VSDs, including a large outflow malalignment defect, 2 apical muscular defects, and a large mid-muscular defect. As the left ventricle was at systemic pressure we performed an elective arterial switch operation at age 6 weeks but considered the risks of heart block, residual VSDs, and ventricular dysfunction to be substantial if VSD closure was attempted at this stage. We therefore...