Aorto-Right Ventricular Tunnel: An Uncommon Problem with a Common Solution

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Aorto-ventricular tunnel is a rare congenital malformation, and aorto-right ventricular tunnel (ARVT) is an even less common entity. Here, we report the case of a 3-month-old female who underwent successful surgical closure of ARVT. The origin of the right coronary artery was proximal to the ostium of the tunnel.


Case report

A 3-month-old female child weighing 4.6 kg was referred to us with the provisional diagnosis of a ventricular septal defect in the status of congestive heart failure under medication with furosemide (2.8 mg three times a day), spironolactone (6.25 mg once a day), and digitalis (10 mcg twice a day). A harsh continuous murmur was heard over the left precordium. The echocardiogram showed a tunnel connecting the aorta and the right ventricle, called the aorto-right ventricular tunnel (ARVT) (Fig. 1). The ostium of the right coronary artery was in close proximity to the opening of the tunnel; therefore, interventional management was not feasible.

During surgery, after opening the pericardium, we could see the tunnel on the anterior aspect and on the left of the aorta (Fig. 2A). The patent ductus arteriosus was ligated, cardiopulmonary bypass was instituted, and then, the aorta was promptly cross-clamped. Via a standard oblique aortotomy and distal to the anomaly, a slit-like opening was identified originating above the upper border of the right sinus of Valsalva, approximately 2 mm to the left and cephalad to the right coronary ostium. Both coronary arteries were normal in origin. The passage of a probe through the tunnel and into the right ventricular infundibulum confirmed the diagnosis. Identification of the ventricular end was not possible, and the aortic orifice was closed with a patch of bovine pericardium by using multiple interrupted 7-0 Prolene sutures (Fig. 2B, C). Intraoperative transesophageal echocardiography showed complete closure of the tunnel with no aortic regurgitation. The postoperative course was uneventful, and the patient was discharged from the hospital on postoperative day 9. At the...
24-month follow-up, she was thriving well and an echocardiographic examination showed no residual abnormalities and a competent aortic valve.

**Discussion**

Aorto-ventricular tunnel is a rare congenital malformation and is defined as the presence of an extracardiac channel that connects the ascending aorta above the sinotubular junction to the cavity of the left or the right ventricle [1]. ARVT is rarer [2]. Here, we report the case of a successful surgical closure of ARVT, in which the origin of the right coronary artery was proximal to the ostium of the tunnel.

Only 18 cases of ARVT have been reported thus far in the English literature [2-7]. A differential diagnosis of this malformation includes ventricular septal defect, aortic insufficiency, aortopulmonary window, coronary artery fistula, patent ductus arteriosus, and ruptured sinus of the Valsalva aneurysm. The difference between aorto-ventricular tunnel and ARVT is that in the case of the former, the vascular orifice lies in the tubular aorta and not in a sinus of the aortic valve and runs outside the heart between the muscular subpulmonary infundibulum and the aortic valvar sinuses, as in our case [1]. The possible anomalies in coronary anatomy should be recognized in order to plan a therapeutic strategy. When the entire coronary system originates from a single ostium located at the left sinus, for example, interventional closure by a device may be feasible [6,7]. However, in cases where the tunnel is in close proximity to the
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coronary ostia or aortic valve leaflets, surgical management is imperative. During surgery, the patent ductus arteriosus should be ligated before cardiopulmonary bypass institution and an aortic cross-clamp should be applied promptly in order to prevent right ventricular distention. The aortic opening of most tunnels lies above the right coronary sinus of Valsalva and rarely above the left sinus of Valsalva or the intercoronary commissure. Surgical repair of ARVT consists of closing the tunnel in such a way that the aortic valve is supported, the coronary circulation is not compromised, and the right ventricular outflow tract obstruction is prevented. We prefer patch closure of the aortic end as it is a safer method for preventing recurrence or progressive aortic regurgitation [8]. Ventricular end closure in aorto-left ventricular tunnels is necessary to prevent blind-ending pouch formation and subsequent compression of the right ventricle. However, under the low-pressure conditions of ARVTs, when a ventricular orifice is not identifiable, ventricular end closure can be omitted to avoid the consequences of a right ventricular incision. When the ostium of a coronary artery is proximal to the tunnel, the patch should be deviated distally in order to maintain the perfusion from the aorta. Moreover, re-attachment of the orifice or patch angioplasty is considered an alternative technique in these cases [1].

ARVT repair should be conducted as early in life as possible in order to minimize damage to the right ventricle and to avoid pulmonary vasculature obstruction. Careful preoperative work-up is necessary to choose the most appropriate management.

Conflict of interest

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

References