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Brief Report

Aortic-right atrial tunnel

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Abstract We describe an asymptomatic 1-year-old boy who presented a continuous murmur, and was found to have a tunnel between the right atrium and the aorta. The definitive diagnosis was established by both echocardiography and aortography, and surgical interruption of the tunnel was successful. We emphasise the rarity of this condition, and discuss its major features.

Keywords: Congenital cardiac malformations; aortic-cameral fistula; surgery

A TUNNEL CONNECTING THE AORTA TO THE RIGHT atrium is a rare form of aortic-cameral communication. Since the first description in 1980,[1] to the best of our knowledge seven cases have been reported.[1–3] It is the rarity of this unusual communication that has prompted us to discuss our case.

Case report

An asymptomatic 1-year-old boy was initially seen in our institution for evaluation of a heart murmur. Physical examination revealed a well-developed child. Cardiac assessment showed brisk peripheral pulses, and the blood pressure was 70/50 mmHg. The precordium was quiet. A grade 3/6 continuous murmur was best heard along the right upper sternal border. The second heart sound was loud. Chest X-rays showed mild cardiomegaly, with moderately increased pulmonary vascularity. The electrocardiogram was normal. The echocardiogram demonstrated a large vascular structure in continuity with the aorta. Doppler evaluation of this structure revealed con-tinuous flow within it. Cardiac catheterisation was performed, and a hemodynamic study revealed a step-up in the saturations of oxygen at the right atrium. The other findings were unremarkable. Injection of contrast into the aortic root showed the filling of a large and tortuous vessel, which passed rightward (Fig. 1a), with the contrast material then opacifying the right atrium (Fig. 1b). The coronary arteries were normal. The child underwent surgical treatment, through a median sternotomy, on August 29, 2002. The heart was enlarged, and a thrill was palpable over the right atrium. Conventional cardiopulmonary bypass was instituted at 32°C, and the myocardium was protected with cold cardioplegia and topical hypothermia. A large vessel, 1 cm in diameter, was identified arising from the posterior wall of the aorta (Fig. 2a). This tortuous vessel passed rightward, posterior to the superior caval vein, and entered the right atrium. The vessel was dissected near its origin, and encircled with a tape. The ascending aorta was opened, allowing the identification of the proximal orifice of the tunnel, which originated in a moderately dilated non-coronary sinus of Valsalva. The atrial end of the tunnel was confirmed through a right atriotomy, being located on the lateral aspect of the right atrium. The tunnel was interrupted by closing its atrial orifice with a 5-0 monofilament running suture, and also by placing a double 0 silk ligature round the vessel just after its origin from the aorta (Fig. 2b). The child was weaned from cardiopulmonary bypass with excellent hemodynamics and no thrill was palpable over the heart. The postoperative course was uneventful. One year after the operation, the child is well, no murmur is heard, and the echocardiogram is normal.

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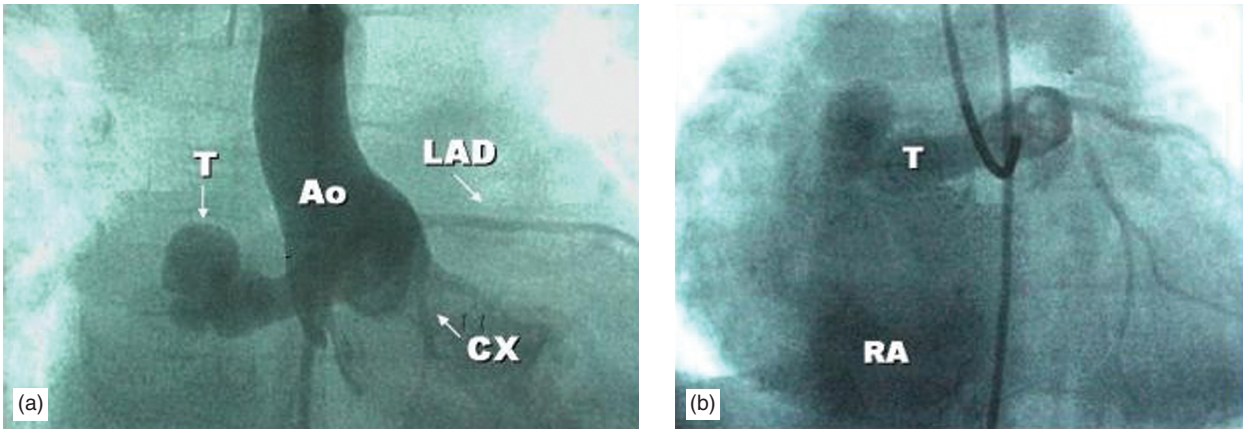


Figure 1.
The retrograde aortogram shows a large tortuous vascular tunnel (T) coursing from the aorta (Ao) to the right atrium (RA). The circumflex (CX) and left anterior descending (LAD) coronary arteries are normal.

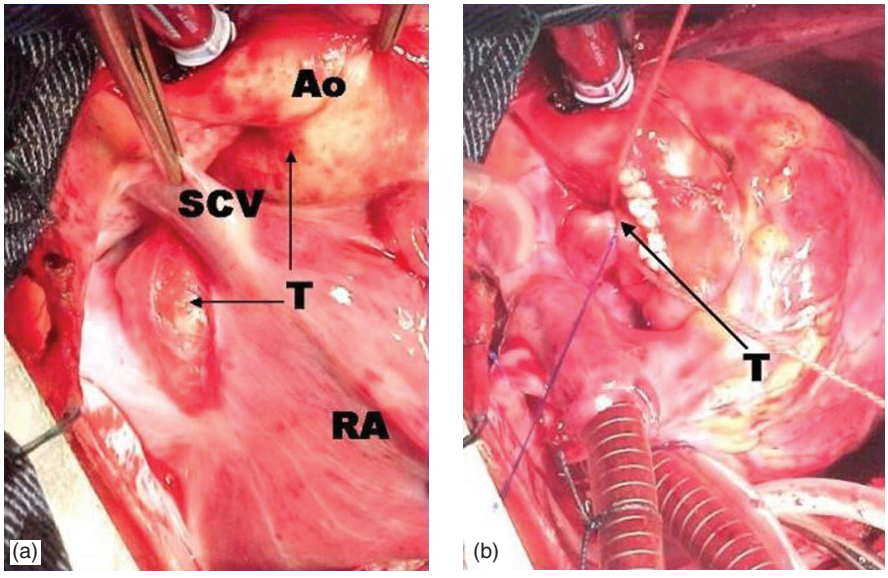


Figure 2.
An operative view (a) showing the vascular tunnel (T) originating from the non-coronary sinus of Valsalva, coursing rightward and behind the superior caval vein (SCV), to terminate in the right atrium (RA). We interrupted the tunnel externally (b) by means of a double ligature. Note also the suture line for closure of the aortotomy adjacent to the ligated tunnel.

Discussion

The aortic-right atrial tunnel is a rare pathological condition, which should be differentiated from coronary arterial fistulas since the tunnel originates in the aorta independently of the coronary arteries. As far as we can establish, the literature in the English language concerning this anomaly is limited to seven patients.[1–3] In the first reported case, an unusual aneurysmal communication was discovered in a 25-year-old man who also had absence of the right superior caval vein.[1] This patient presented because of discovery of a mediastinal mass on chest X-rays, and was submitted to an exploratory thoracotomy, which revealed an aneurysm with a continuous thrill juxtaposed to the right atrium. Further cardiac catheterisation demonstrated the communication between the aorta and the right atrium, which was closed with the use of cardiopulmonary bypass.

The other six reported cases, as well as our patient, show a remarkable anatomical and clinical similarity. Morphologically, the tunnel consists of a large and tortuous vessel, originating from either the left or the non-coronary sinus of Valsalva, and coursing rightward behind the superior caval vein to terminate in the right atrium.[2,3] The opening of the tunnel in the right atrium, however, is variable. The tunnel has been found entering through the superior caval vein, the atrial septum, or the superior or lateral aspects of the right atrium. No small coronary arterial branches arising from the tunnel have been described in any of the cases. The main coronary arteries, nonetheless, arose from the tunnel in three

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of the reported patients.[2] This finding is surgically important, because ligation of the tunnel close to the aorta, as in our patient, should only be performed after the anatomy of the lesion has been clarified by external dissection of the vessel and inspection of the inside of the ascending aorta. Absence of an identifiable sinus nodal artery has also been described in association with the malformation,[3] but we could not define such an anatomic detail in our case. Histologic examination of this vascular channel has demonstrated similarity with the normal aortic structure in two patients,[2] and the presence of myxomatous tissue in one.[3]

An important morphologic feature observed in all patients described thus far is dilation of the sinus of Valsalva from which the tunnel takes origin. The importance of this finding in the long-term follow-up is uncertain but, in the two oldest reported patients, aneurysmal expansion of the tunnel,[1] and aortic regurgitation,[3] were observed. We wonder whether ligation of the tunnel close to its origin, rather than simply suturing its atrial orifice, may prevent any further dilation of the sinus of Valsalva? To this end, surgical repair early in life may also be of value. Only long-term follow-up of the operated patients will give a definitive answer to this intriguing problem.

Clinically, all patients were essentially asymptomatic, and came to medical attention due to the presence of a continuous murmur. The tunnel was identified by cross-sectional echocardiography, with definitive diagnosis easily made by aortography.[2,3]

Surgical closure of the tunnel is indicated even in asymptomatic patients, because the tunnel produces volume overload of the ventricles, and increases the flow of blood to the lungs. Furthermore, there is a potential risk of infectious endocarditis and aneurysmal formation. On the other hand, the operative risk is low.[2,3] Our patient, like the other six previously described cases, underwent a successful operation, which did not pose any special technical problems. We interrupted the tunnel by suturing its end atrial orifice, and also ligated it close to its aortic origin.

Aortic-right atrial tunnel, therefore, is a rare condition in which clinical differential diagnosis should include coronary artery fistulas and ruptured aneurysm of the sinus of Valsalva. Definitive diagnosis and detailed morphological definition may be obtained by either aortography or magnetic resonance angiography. Surgical closure should be recommended to avoid potential complications even in asymptomatic patients.

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