The Heart Surgery Forum #2006-1114
10 (1), 2007 [Epub December 2006]
doi: 10.1532/HSF98.20061114

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Congenital Aorto-Left Atrial Tunnel—An Unusual Communication: A Case Report

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ABSTRACT

Congenital aortico-cameral communications are rarely seen. We present an asymptomatic patient in whom there was a congenital vascular communication rising from the aortic root and terminating in the left atrium. She had an atrial septal defect (ASD). The diagnosis was made with echocardiography and confirmed by aortography. She was treated by closing the tunnel and the ASD. The outcome was satisfactory in this extremely rare case of a congenital cardiac lesion. Coincidental diagnosis could be made during careful echocardiographic examination.

INTRODUCTION

Aortico-cameral communications are relatively uncommon. Most of them are usually acquired and associated with aneurysms of the sinus of Valsalva. They rupture as a result of bacterial endocarditis, paravalvular abscess, or trauma [Archer 1997]. The communications are rarely congenital and usually placed between the aorta and the right atrium or right ventricle [Otero Coto 1980; Rosenberg 1986; Kalangos 2000; Turkay 2003]. Aorto–left atrial communications are extremely rare in the literature [Jean 1986; Nihoyannopoulos 1987; Topcuoglu 1987].

We present a patient in whom there was a congenital vascular communication rising from the aortic root and terminating in the left atrium.

CASE REPORT

A 2-year 6-month–old girl was referred for evaluation of a heart murmur and cardiomegaly. Physical examination was normal except for a grade 1 to 2/6 systolic murmur at the level of the second intercostal space. A chest x-ray revealed a mild cardiomegaly and a slight increase in the pulmonary vascularity. A 2-D echocardiogram showed a secundum atrial septal defect (ASD) with enlargement of the right atrium and ventricle. On parasternal long-axis view, there was a vascular structure rising from the left coronary cusp of the aorta (Figure 1) and a continuous turbulent flow with high velocity (4.1 m/s) into the left atrium, close to the left pulmonary vein. This finding suggested that there was a connection between the aortic root and the left atrium, probably a corona-cameral fistula. We performed cardiac catheterization and aortography. Pulmonary artery pressure was measured as 30/24 mmHg. The pulmonary-systemic blood flow ratio was calculated as 1.55:1. On the left ventricular angiogram, the left ventricle and aorta were filled with contrast material and a vascular structure originating from the aortic root was opacified (Figure 2). Aortic root angiography revealed a large tunnel rising above the left sinus of Valsalva with a dilated sac, passing posteriorly, and terminating in the left atrium (Figure 3).

The patient underwent an operation to repair the aortico-left atrial tunnel and ASD. The heart was exposed through a midline sternotomy. The right atrium was large, and the surgeon palpated a continuous trill on the left atrium. Cardiopulmonary bypass was instituted. The aorta was cross clamped. After the right atriotomy, the left atrium was inspected through the ASD. The cardioplegic solution given into the ascending aorta appeared in the left atrial orifice of the tunnel between the mitral annulus and the left inferior pulmonary vein. Subsequently, an aortotomy was performed. The tunnel rose from the left sinus of Valsalva where the ostium of the left coronary artery should have been present. Additionally, a coronary artery origin anomaly, the left coronary artery rising from the right sinus of Valsalva, was observed. The aortic orifice of the tunnel was closed with several interrupted stitches reinforced with teflon felt. The left atrial orifice of the tunnel and ASD were closed via the right atriotomy. Postoperative aortograms showed no residual defect (Figure 4). The postoperative period was uneventful. The patient is asymptomatic in follow-up examinations.

DISCUSSION

Aortico-cameral communications are rarely described in the literature. Virtually all reported communications are

Received August 23, 2006; accepted September 12, 2006.

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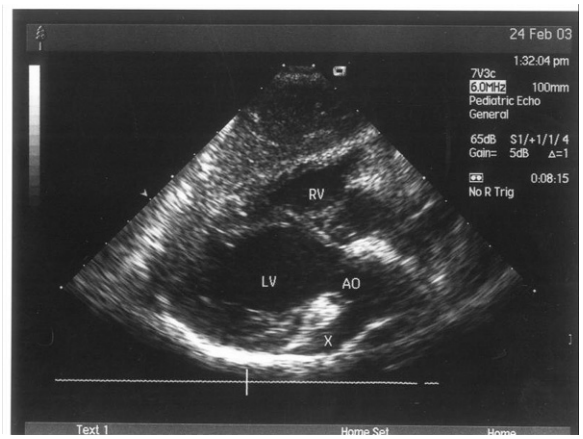

Figure 1. In the modified parasternal long axis view, a large vascular structure originates from the left coronary cusp. AO indicates aorta; LV, left ventricle; RV, right ventricle; X, vascular structure.

between the aorta and the right atrium, right ventricle or left ventricle, and quite rarely the left atrium. Most are acquired and have been causally associated with bacterial endocarditis, paravalvular abscess, ruptured aneurysm of the sinus of Valsalva, aortic dissection, or trauma [Archer 1997]. Congenital aortico-cameral communications are seen much more rarely.

Communications between the aortic root and right atrium were first reported by Otero Coto et al [1980]. Later Rosenberg et al described 4 cases with the congenital tunnel between the left sinus of Valsalva and the right atrium, and 2 of them had coronary artery abnormalities. Rosenberg et al discussed the difference between an aortico-cameral connection and a coronary-cameral connection [Rosenberg 1986]. Kalangos reported 2 patients with a congenital aorto-right atrial tunnel [Kalangos 2000]. These patients, like ours, had a large canal that originated from the left sinus of Valsalva and terminated in the roof of the right atrium.

Acquired communications occur either as varicose aneurysms or as direct anastomoses caused by trauma, operation, or

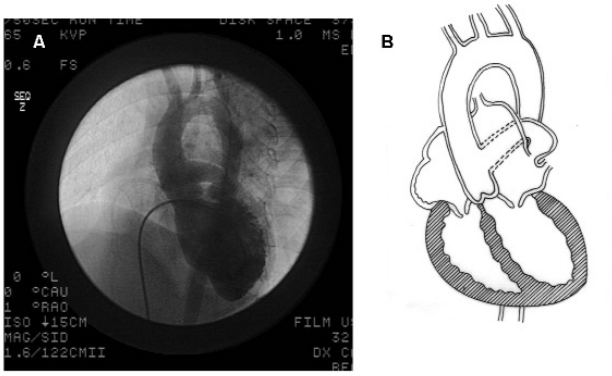

Figure 2. Photograph (A) and diagram (B) of the left ventriculography in the long axis projection. The left ventricle and aorta were filled with contrast material and a vascular structure originating from the aortic root was opacified.

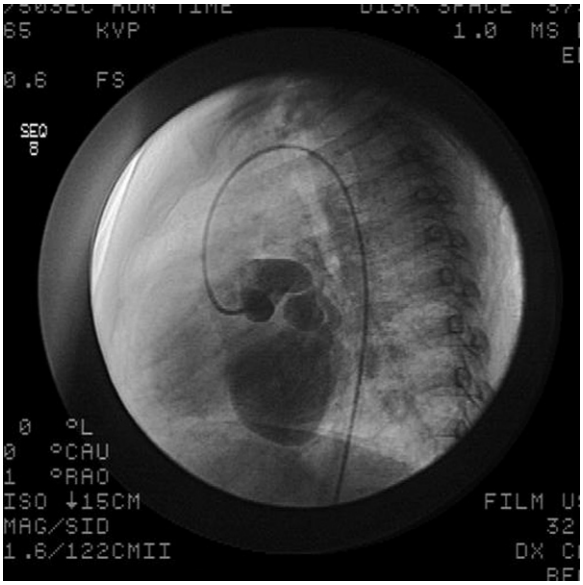

Figure 3. Injection of contrast material into the aortic root in the anterior-posterior projection revealed a large tunnel rising above the left sinus of Valsalva with a dilated sac, passing posteriorly, and terminating in the left atrium.

infection. Because of the absence of symptoms suggestive of a rupture, configuration of connection, and lack of morphologic evidence associated with aortic dissection such as Marfan's syndrome, we believe that this communication is a structural defect of congenital origin.

Jean et al presented a patient with an aorto-left atrial fistula that originated near the orifice of the left coronary artery; the patient had no coronary anomaly [Jean 1986]. This case was similar to our case. However, the fistula Jean reported had 4 large aneurysmal sacs. Our case had only a long vascular canal with no aneurysmal formation. Our patient was asymptomatic, but the risk of continued potency of communication could manifest by aneurysmal formation, volume overload of ventricles, bacterial endocarditis, or spontaneous rupture. Surgical repair was performed soon after the diagnosis. If she had not undergone the operation early, aneurysmal sacs could have developed. Ligation of the canal is sufficient, as it is simple and curative without the need for a cardiopulmonary

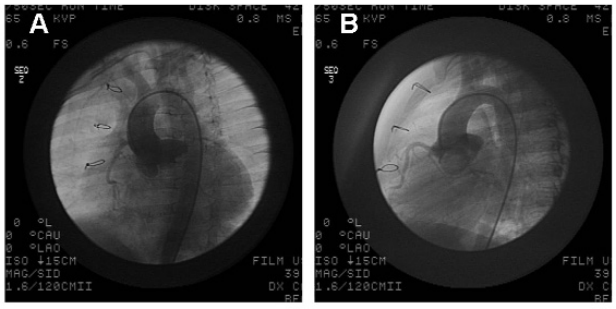

Figure 4. Postoperative aortograms in left anterior oblique (A) and lateral (B) projections show no residual defect.

bypass. A cardiopulmonary bypass could have been required if these complications had developed. In our case, surgery was performed with cardiopulmonary bypass for the closure of ASD. If the patient had not had an ASD, the surgery would have been a simple ligation without cardiopulmonary bypass. Nihoyannopoulos et al described a 4-year-old patient with a congenital fistula between the descending aorta and the left atrium [Nihoyannopoulos 1987]. They performed the operation through a left thoracotomy incision without cardiopulmonary bypass. Topcuoglu et al reported a case with aorto-left atrial fistula with bicuspid aortic valve and coronary artery origin anomaly [Topcuoglu 1987]. This case was similar to ours with the same coronary anomaly, but their patient had only a fistula formation, not a vascular structure. Our patient had a large and long tortuous vascular structure rising from the left sinus of Valsalva.

Echocardiography is a useful noninvasive diagnostic tool in patients with heart murmur. In our patient, a vascular structure rising from the left coronary cusp and a continuous turbulent flow into the left atrium could be seen in echocardiographic views. Although this anomaly can be seen with echocardiography, the diagnosis should be confirmed by cardiac catheterization and aortography.

In conclusion, aortico–left atrial communications are very rare congenital anomalies. Patients may not have any symptoms or any abnormal physical finding, as in our patient. In this particular case, 2-D echocardiography was performed because of the cardiomegaly due to an associated anomaly (ASD). Therefore, this extremely unusual anomaly was recognized merely incidentally. An incidental diagnosis can be made during careful echocardiographic examinations.

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