

Brief Report

Atresia of the orifice of the coronary sinus after surgery

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Abstract We describe a patient with a coronary arteriovenous fistula, atresia of the orifice of the coronary sinus, and persistence of the left superior caval vein. Depression of her ST segments was revealed by exercise electrocardiography long after the initial surgical reconstruction of the coronary sinus.

Keywords: Persistent left superior vena cava; coronary arteriovenous fistula; cut back method

ATRESIA OF THE ORIFICE OF THE CORONARY sinus is rare in the setting of a persistent left superior caval vein. When found, other cardiac anomalies have been present in more than half of the reported cases.^{1,2} As far as we know, however, the combination has yet to be reported in association with a coronary arteriovenous fistula. We have now followed such a patient for 10 years subsequent to surgical reconstruction of the coronary sinus.

Case report

A 5-year-old girl was referred to us with a heart murmur and cardiomegaly seen on the chest X-ray. Transthoracic echocardiography revealed dilation of the left circumflex coronary artery, with an abnormal vessel also being seen which connected to the dilated coronary sinus. Aortography confirmed the diagnosis of a coronary arteriovenous fistula, extending from the circumflex coronary artery to the coronary sinus. It also revealed atresia of the orifice of the sinus, which drained a persistent left superior caval vein. The fistula itself was dilated and tortuous. Coronary venous blood flowed into the right superior caval vein via the left vein (Fig. 1). The ratio of pulmonary-to-systemic flow was calculated as 1.6 : 1.

At operation using cardiopulmonary bypass, the atresia was discovered to be due to a membrane at the mouth of the coronary sinus. The abnormal

membrane was removed, and the orifice was cut back to produce an opening 5 mm in diameter. A fistula of 3 mm in diameter was found between the circumflex coronary artery and the coronary sinus which was closed from the venous side. The circumflex coronary artery was also ablated distal to its obtuse marginal branch because of a residual left-to-right shunt through the fistula. The postoperative course

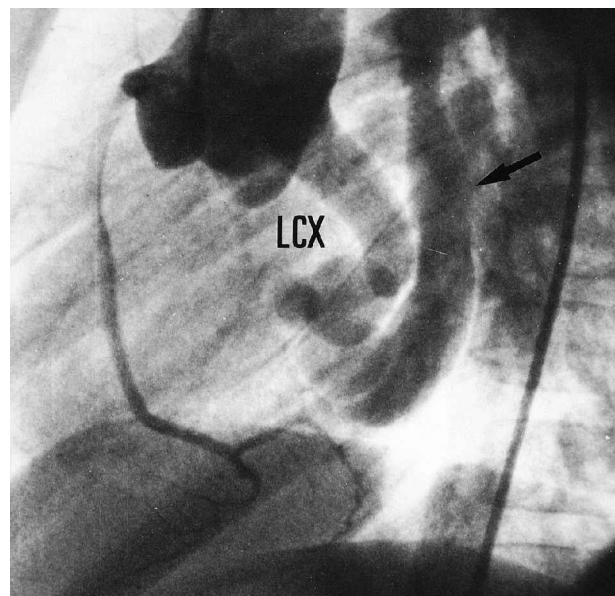


Figure 1. Lateral view of the aortogram at 5 years of age showing an arteriovenous fistula from the left circumflex coronary artery to the coronary sinus vein. Note the flow of contrast medium into the persistent left superior caval vein (arrow). LCX: left circumflex coronary artery.

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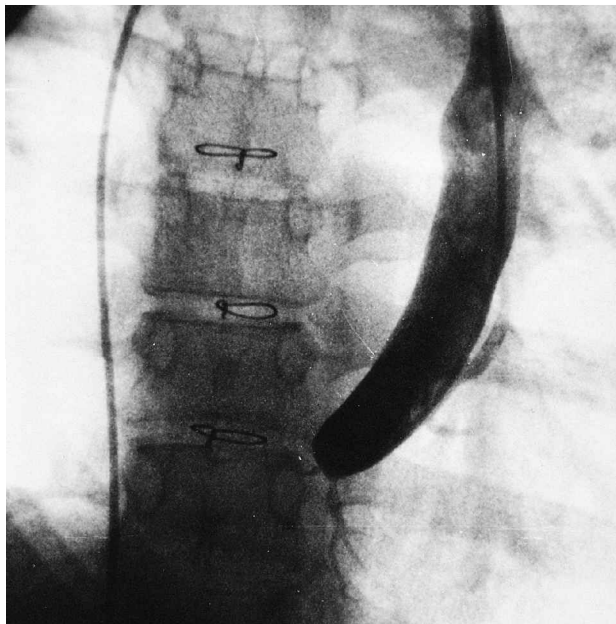


Figure 2.
Anteroposterior view of contrast injection in the left superior caval vein. The reconstructed orifice of the coronary sinus was already closed at 11 years of age.

was uneventful. Cardiac catheterization at the 23rd day after operation demonstrated no residual shunt, normal left ventricular contraction, and showed the reconstructed orifice of the coronary sinus to be patent.

Selective coronary angiography at 11 years of age, however, revealed acquired closure of the reconstructed orifice of the coronary sinus. At 15 years of age, cardiac catheterization revealed a mild pressure gradient of 3 mmHg across an area of stenosis within the left superior caval vein (Fig. 2). Pooling of contrast medium was noted at the dead end of the circumflex coronary artery proximal to the site of ligation. Exercise electrocardiography exhibited depression of slowly rising ST segments in leads II, III, aVF and V4-6. No deficiency in myocardial perfusion was detected by exercise-loading single photon emission computed tomography using technetium-99m-tetrofosmin. The patient has since been treated with anti-platelet therapy to prevent coronary thrombosis.

Discussion

The majority of reported cases with atresia of the orifice of the coronary sinus in presence of a persistent left superior caval vein have been diagnosed at post-mortem examination. In just under half the cases, the association has been found in isolation, but

the remaining cases were combined with further abnormalities, such as unroofing of the sinus or additional systemic venous anomalies.³⁻⁵ To our knowledge, this is the first patient in whom a coronary arteriovenous fistula has been found in association with atresia of the orifice of the coronary sinus and persistence of the left superior caval vein. The combination was identified by aortography prior to the surgical correction.

This is also the first report of surgical reconstruction of the orifice of an atretic coronary sinus without concomitant ligation of the left superior caval vein. Six years after the operation, however, the repaired orifice had closed, albeit without precipitating any coronary event. It is known that reconstruction of the orifice of the coronary sinus can induce critical cardiac events when the left superior caval vein has been ligated.⁶⁻⁸ In our case, the persistently patent left superior caval vein functioned as a bypass tract for the coronary venous return.

Also in our patient, exercise testing induced depression of the ST segments but without any evidence of myocardial ischemia. Such changes in the ST segments, along with, temporary bradycardia, has been reported when the left superior caval vein was obstructed by a catheter in an infant⁸ with atresia of the orifice of the coronary sinus. The fate of the stenosis in the left superior caval vein of our patient remains unknown.

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