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Brief Report

Childhood angina due to congenital atresia of the coronary sinus

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Abstract A young child with exertional chest pain, and an electrocardiographic pattern suggesting reversible ischaemia of the anterior ventricular wall documented by Cardiolyte stress-testing, underwent cardiac catheterization and selective coronary angiography. Although the coronary arteries were entirely normal, the recirculation phase demonstrated marked dilation of the coronary sinus, with atresia of its mouth. At surgery, the patient was confirmed to have muscular atresia at the mouth of the coronary sinus, and underwent unroofing of the coronary sinus to the left atrium, with ligation of a persistent left superior caval vein. Post-operatively, the patient continued to have persistent chest pain, albeit without inducible ischaemia on stress-testing.

Keywords: Chest pain; persistent left superior caval vein; congenital heart disease

Chest pain is a common reason for referral of a child to a pediatric cardiologist. Although parental concern frequently focuses on the possibility of angina, this is rarely the case. When the pain becomes exertional, nonetheless, or occurs shortly after peak exercise, the possibility of myocardial ischaemia must be entertained. We describe the case of an 11-year-old male who was discovered to have atresia of the mouth of the coronary sinus that resulted in impaired myocardial perfusion during exercise, with reversible myocardial ischaemia.

Case report

An 11-year-old male was referred for evaluation because of exertional dyspnea of new onset associated with dull, non-radiating, mid-sternal chest pain. There was no associated nausea, vomiting, palpitations, tachycardia, presyncope or syncope, but he did experience dizziness with exertion. An echocardiogram performed at 2 years of age during evaluation of a murmur was said to be "normal", and the past medical history was otherwise unremarkable.

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On examination the patient had a height of 148 cm, and weighed 49.8 kg. The pulse was counted at 80 beats/mm, with a respiratory rate of 18 breaths/min, and blood pressure of 116/58 mmHg. The entire physical examination was normal. Electrocardiography showed left ventricular hypertrophy, and a non-specific intraventricular conduction delay. Echocardiography demonstrated a structurally normal heart, with trivial mitral and mild tricuspid valvar regurgitation, with an estimated right ventricular systolic pressure of 35 mmHg. There was no ventricular hypertrophy, and the anatomy of the coronary arteries appeared normal. A treadmill-cardiolyte stress test demonstrated no electrocardiographic evidence of ischaemia, but a stress-induced reversible abnormality in perfusion of the anterior wall was seen. The left ventricular ejection fraction was 67%. Pulmonary function studies were normal.

Cardiac catheterization demonstrated normal oximetric and hemodynamic data. The cardiac index, estimated by the Fick method, was 4.35 l/min/m^2. There was a probe-patent oval foramen, but with no left atrial desaturation, ruling out unroofing of the coronary sinus. Angiography demonstrated normal left ventricular systolic function without mitral regurgitation. Selective coronary angiography demonstrated normal coronary arteries without stenosis, myocardial bridging, aneurysm, or dissection. The coronary sinus was dilated secondary to atresia at its mouth,

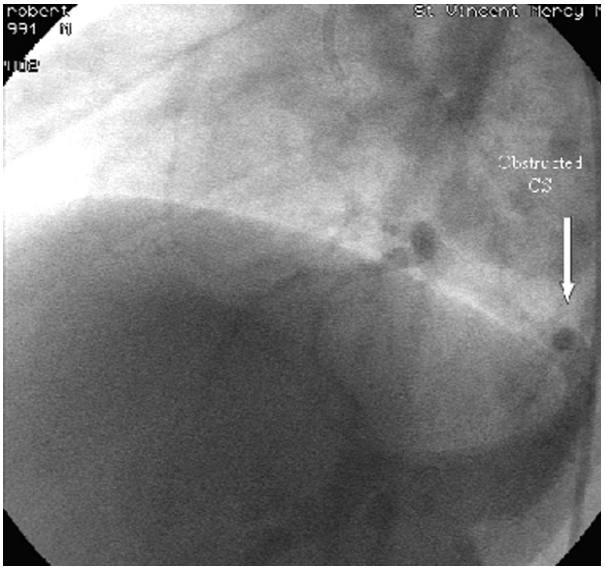

Figure 1.
The recirculation phase after selective angiography in the left coronary artery demonstrating a dilated and patulous coronary sinus consistent with obstruction to its outflow.

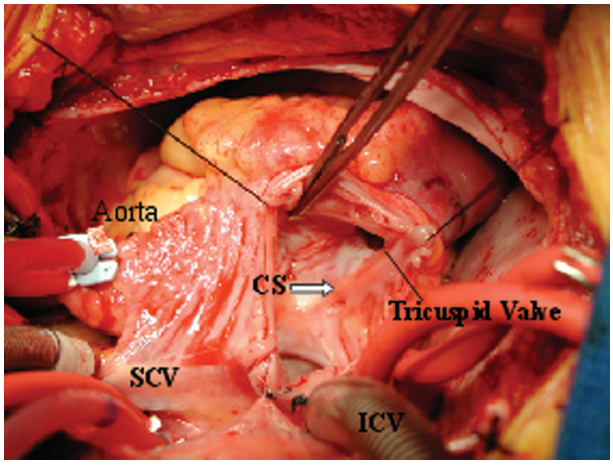

Figure 2.
Intraoperative right atrial view showing muscular atresia of the mouth of the coronary sinus. SCV: superior caval vein; ICV: inferior caval vein; CS: coronary sinus.

with venous collaterals emptying directly into the right atrium and right ventricle (Fig. 1). During surgery, atresia at the mouth of the coronary sinus was confirmed (Fig. 2). Externally, the coronary sinus was markedly dilated, and was directly entered via a small incision along the inferior atrioventricular groove. The atrial septum was opened, and the walls separating the left atrium from the coronary sinus were located by palpating a probe introduced directly into the coronary sinus. The coronary sinus was opened along much of its length to allow free drainage to the left atrium. The atrial septum and coronary sinus were closed directly. A small persistent left superior caval vein was identified and ligated to prevent the development of a significant right-to-left shunt. The post-operative course was complicated by the development of atrial flutter, requiring cardioversion and antiarrhythmic therapy. Echocardiography showed non-restrictive flow from the coronary sinus into the left atrium, with normal myocardial function.

Long-term follow-up was remarkable for persistent chest pain, both at rest and with exertion. Repeat cardiolite stress-testing showed mildly decreased exercise tolerance, no arrhythmias, and no evidence of ischaemia. Coronary perfusion and systolic function were normal. Repeat pulmonary function tests demonstrated mild obstruction of the airways. His atrial arrhythmia was effectively controlled with amiodarone, which was discontinued after 7 months.

Discussion

Chest pain is a common complaint among children. True angina, however, is extremely rare but very serious. Atresia of the mouth of the coronary sinus in the absence of complex congenital heart disease is also extremely rare, with only 11 case reports in the literature dating back to 1738.[1,2] In such cases, the coronary veins typically drain through fenestrations into the right atrium, right ventricle, or left atrium. Frequently a persistent left superior caval vein may allow for decompression into the brachiocephalic vein and prevent symptoms. Among all the cases described in the literature, seven-tenths were found at autopsy.[1]

It has been estimated that up to one-fifth of the blood entering the right coronary artery returns to the coronary sinus and enters the right atrium. The remainder is returned to the right ventricle by the anterior cardiac veins. Of the blood flowing through the left coronary artery, three-quarters is received by the coronary sinus. Thus, the left coronary arterial territory may be more influenced by obstruction to return via the coronary sinus.[3–5] Ischaemia during exercise may be related to increased right heart pressure leading to decreased coronary venous outflow and impeded coronary arterial flow, or altered myocardial metabolism. In our patient, the persistent left superior caval vein was small, and may have led to worsening obstruction as he grew older. Thus, in contrast to the report of Adatia et al.,[6] our case shows that isolated atresia of the coronary sinus may be associated with significant morbidity and possible mortality if not diagnosed expeditiously.

Surgical repair involved "unroofing" the coronary sinus to the left atrium, closing a small patent oval foramen, and ligating the small left superior caval vein. Although prior reports have advocated leaving

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patent the left superior caval vein, we felt that this could promote the development of an increased right-to-left shunt because the obstruction to the coronary venous return to the left atrium was removed.[4,7]

Repeat Cardiolyte stress-testing failed to demonstrate any evidence of inducible ischaemia and no fixed perfusion defects. Due to persistent non-exertional chest pain, a repeat cardiac catheterization was performed. The coronary sinus was still mildly dilated and patulous, but the egress of contrast appeared unobstructed. The latter was demonstrated by Doppler echocardiography. No clear etiology of his chest pain was uncovered. It may be that chronic venous obstruction resulted in changes to the coronary arterial capillary bed leading to abnormal myocardial metabolism or altered coronary arterial capillary resistance.

This very rare case demonstrates the importance of considering isolated abnormalities of the coronary sinus in patients with exertional ischaemia and "no obvious congenital heart disease". Cardiac catherization with coronary angiography demonstrating the recirculation venous phase is typically diagnostic. Parents must also be informed about the potential that cardiac symptoms may persist despite surgical repair.

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