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*Brief Report*

# Multiple fistulas from the coronary arteries to the left ventricle in tricuspid atresia

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**Abstract** It is rare to find multiple fistulas arising from all three coronary arteries and draining into the left ventricle. Coronary angiography revealed this anomaly in a one-year-old girl with tricuspid atresia after conversion to the Fontan circulation. To the best of our knowledge, this is the first report of such multiple fistulas in the setting of tricuspid atresia, and also the first report in childhood.

Keywords: Absent right atrioventricular connection; fistulaous communications; congenital heart disease

CONGENITAL CORONARY ARTERY FISTULAS ARE found at cardiac catheterisation in 0.2% of cases. In more than nine-tenths of these cases, a single fistula drains into the right heart chambers or the pulmonary trunk.[1] Multiple fistulas connecting all three major coronary arteries to the left ventricle are rare.[2–8] We report, to the best of our knowledge, the first instance of these anomalies in the setting of tricuspid atresia, and also the first instance discovered in childhood.

## Case report

A female infant was born at 37 gestational weeks with a weight of 2825 g. She became cyanotic on her first day of life. The echocardiogram revealed typical findings of tricuspid atresia with normally related great arteries and pulmonary stenosis. It also revealed a persistent left superior caval vein. On the twenty-first day of life, she underwent construction of a modified Blalock-Taussig shunt. Her serial electrocardiograms since three-months-old showed inversion of the T-waves, with minor depressions of the ST-segment of 0.1 mV in the left precordial leads. A thallium-201 perfusion scan performed at rest revealed no defects. At the catheter performed at one year of age prior to construction of the Fontan circulation, left ventriculography showed a slightly enlarged left ventricle, albeit without regional abnormalities of wall motion. Aortography showed normal epicardial coronary arteries. At the age of one year and eight months age, she underwent a Fontan-type operation with construction of a lateral tunnel. A left superior caval vein was confirmed, and it was ligated. The surgeon also confirmed a normally situated orifice for the coronary sinus.

One month after the Fontan operation, precordial examination revealed a diastolic murmur, of grade two out of six, at the cardiac apex. Colour Doppler echocardiography then showed multiple flows perpendicular to the epicardial surface draining into the left ventricular cavity. Left ventriculography showed a normal-sized left ventricle, again without regional abnormalities of wall motion, and with an ejection fraction of 58%. Left ventricular end-diastolic pressure was 14 mmHg, and the mean pulmonary arterial pressure was 15 mmHg. Coronary arteriography revealed mildly dilated epicardial arteries. Shortly after injection of the contrast material, a diffuse plexus of fine vessels was seen arising from the left anterior descending, left circumflex, and right coronary arteries (Figs 1 and 2). Subsequently, the left ventricle was opacified, permitting the diagnosis of multiple fistulas from the coronary arteries to the left ventricle. The coronary sinus was not opacified. After seeing these changes, we reviewed the aortogram performed prior to the Fontan procedure. With the aid of hindsight, we were able to see the left ventricle opacifying from the epicardial coronary arteries.

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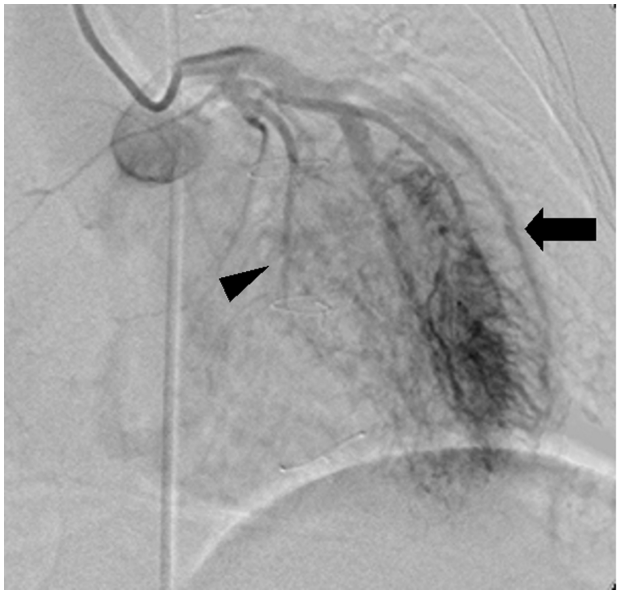

**Figure 1.**
*Selective left coronary arteriography in the right anterior oblique view showed a plexus of fine vessels arising from branches of the left anterior descending (arrow) and circumflex arteries (arrowhead).*

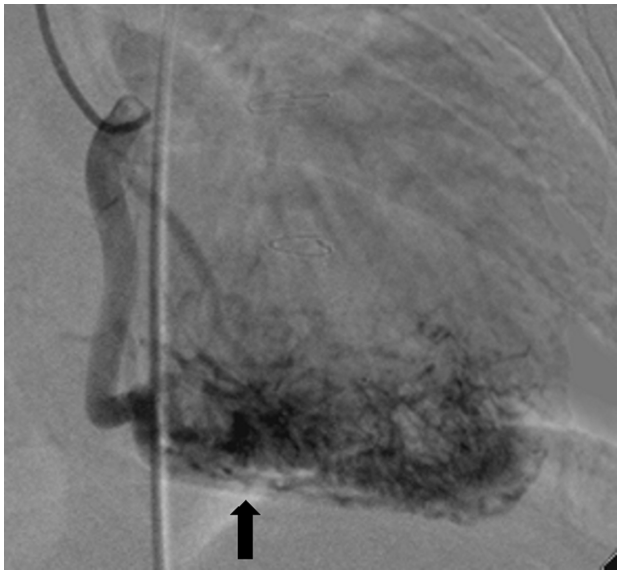

**Figure 2.**
*Selective right coronary arteriography in the right anterior oblique view showed a plexus of fine vessels arising from the inferior interventricular artery (arrow).*

## Discussion

Our case has two interesting points. First, to the best of our knowledge, this is the first case reported with multiple fistulas from the coronary arteries to the left ventricle in the setting of tricuspid atresia. As far as we know, however, there is no pathogenetic relation between the fistulas and failure of formation of the right atrioventricular connection.

Although the pathogenetic origin of such multiple fistulas has yet to be determined, some have suggested an abnormal prominence of the Thebesian system.[9] Embryonic myocardial sinusoids arise from endothelial protrusions into the intertrabecular spaces. These structures regress during normal development, leaving the Thebesian vessels of the adult heart. Interference with developmental changes may produce partial persistence of the embryonic intertrabecular vascular network and result in the morphological appearance of multiple coronary fistulas.

Tricuspid atresia is most usually the consequence of morphologic absence of one atrioventricular connection.[10] In most cases, the ventricular mass comprises a dominant left ventricle with a rudimentary right ventricle. No previous reports have described any relations between the absence of one atrioventricular connection and an abnormally prominent Thebesian system. It remains possible, nonetheless, that the coincidence of these two rare anomalies may suggest that the anomalies derive from one event in the embryonic developmental stage.

The second interesting point of our case is that it is the first case thus far reported in childhood. Our review of the previously described cases shows that patients with this anomaly usually present with chest pain in advanced adult life.[3,4] On physical examination, continuous murmurs are not usually a feature of this anomaly, in contrast to those fistulas that drain into the right heart chambers or the pulmonary trunk. The chest X-ray is usually unremarkable, while the electrocardiogram shows only nonspecific changes. The clinical syndrome is attributed to a coronary steal mechanism, with the capillaries bypassed for delivery of oxygen to the myocardium.[4,5]

Little is known about the natural history or prognosis of such multiple fistulas. Some have reported a good clinical course,[6,7] while others reported a high incidence of myocardial infarction.[5] The ST-T changes on the electrocardiogram did suggest the hazard of myocardial damage in our patient.

Because of the diffuse nature of the fistulas, surgical or transcatheter treatment is not possible. Medical treatment with nitrates, in conjunction with beta-blockers or calcium-antagonists, has proved effective in adults.[4,5] We did consider the administration of angiotensin-converting enzyme inhibitors or beta-blockers.

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