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Brief Report

Subneoaortic stenosis complicating the arterial switch operation

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Abstract We report obstruction of the reconstructed left ventricular outflow tract diagnosed 18 months after an arterial switch operation that included closure of a ventricular septal defect. We speculate that either turbulences created by the patch used to close the ventricular septal defect, or postoperative modifications of ventricular geometry, were responsible for producing this unusual complication.

Keywords: Transposition; discordant ventriculo-arterial connections; left ventricular outflow tract obstruction

SUPRAVALVAR STENOSIS, AORTIC VALVAR insufficiency, and stenosis at the origins of the transplanted coronary arteries, are the complications most frequently encountered after the arterial switch operation.[1] We have now encountered a patient who developed stenosis within the neoartic left ventricular outflow tract 18 months after the arterial switch procedure. Our patient had concordant atrioventricular and discordant ventriculo-arterial connections, together with a ventricular septal defect. Although obstruction in the new subpulmonary outflow tract is well recognised, as far as we are aware it is most unusual to encounter obstruction within the left ventricular outflow tract after the arterial switch procedure.

Case report

An 18-month-old girl underwent an arterial switch operation, with closure of a co-existing ventricular septal defect, in the neonatal period. Preoperatively, we had diagnosed usual atrial arrangement, normal venoatrial and atrioventricular connections, and discordant ventriculo-arterial connections. The aorta and pulmonary trunk were positioned side-by-side, with the aorta to the right. The aortic arch was left-sided, and there was a so-called "inverted" origin of the coronary arteries from the facing aortic sinuses, with the right coronary artery arising from the right hand sinus (#1), and the main stem of the left coronary artery arising from the left hand sinus (#2). The left coronary artery then took a retropulmonary course, before dividing into the circumflex and anterior interventricular arteries. There was also a large perimembranous ventricular septal defect. Subsequent to an uneventful arterial switch procedure, and closure of the ventricular septal defect, the post-operative period was also uneventful. Echocardiography at the time of discharge from hospital revealed good biventricular function, no gradient across the left ventricular outflow tract, no aortic insufficiency, no residual ventricular septal defect, and a small gradient of 15 millimetres of mercury across the newly reconstructed subpulmonary infundibulum. At follow-up, the child was active and growing normally, but a systolic murmur was noticed. Echocardiography now revealed marked left ventricular hypertrophy. Further echocardiography, performed one year after the operation, confirmed left ventricular hypertrophy, and now showed a gradient of 70 millimetres of mercury in subaortic position related to a discrete subvalvar fibrous ridge. There was no increase in the gradient previously noted across the right ventricular outflow tract, and the patch placed to close the ventricular septum was intact. Cardiac catheterisation performed at the same time showed a widely patent right ventricular outflow tract, with a normal pulmonary trunk and branches.

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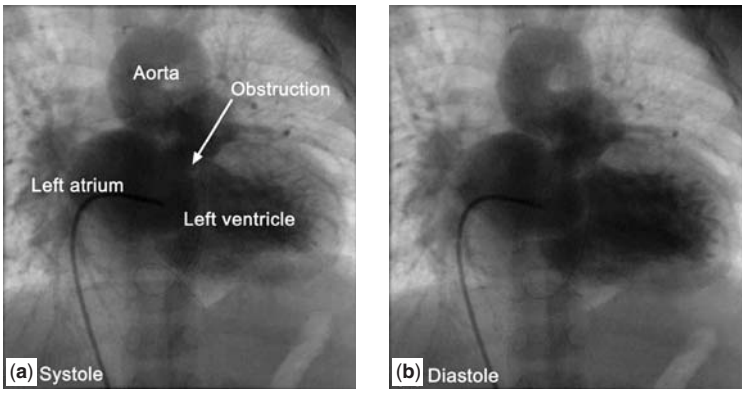


Figure 1.
*Angiography of the left ventricle in (**a**) systole and (**b**) diastole, showing the narrow left ventricular outflow tract.*

The injection in the left ventricle showed a narrowing in the subaortic region (Fig. 1), with haemodynamic measurements confirming the systolic gradient of 70 millimetres of mercury. The patch placed to close the ventricular septal defect was intact. The aortogram showed a normal neoaortic valve, with good coronary arterial anastomoses.

A second open heart operation was performed through an aortotomy above the pulmonary arteries. A fibrous diaphragm was found producing discrete subaortic stenosis. In addition, the muscular ventricular septum was protruding into the left ventricular outflow tract, but the neoaortic valve was normal. The fibrous shelf was completely excised by sharp and blunt dissection through the aortic valve. In addition, a myectomy was made underneath the zone of apposition between the right and left coronary aortic leaflets. The post-operative period was uneventful, with a residual gradient of 30 millimetres of mercury measured across the left ventricular outflow tract at the time of discharge.

Discussion

Repair of transposition associated with ventricular septal defect using the arterial switch procedure during the neonatal period is now recognised as a safe surgical method that offers good results in the early and midterm. Although most children repaired in this fashion have normal psychomotor development and cardiac function, a few do develop early or late complications, and may require reoperation.[2] The most common complication recognised as requiring treatment is subvalvar stenosis of the newly created outlet from the morphologically right ventricle to the pulmonary trunk, an unwanted occurrence in about one-sixth of surviving patients. Residual ventricular septal defects, and coronary arterial stenoses, are much less frequent, being found in less than one-twentieth of patients.[3] Problems are also known to occur during reconstruction of the left ventricular outflow tract and incorporation of the neoaortic systemic valve, and include neoaortic valvar insufficiency and stenosis of the aortic arch.

To our knowledge, however, newly developed subaortic stenosis has yet to be described subsequent to the arterial switch procedure. Turbulent subaortic flow, and geometric modifications of the left ventricle, are two potential causes to explain the occurrence of this complication in diseases other than transposition.[4] We had reviewed the development of subaortic obstruction many years ago in a few patients with ventricular septal defects in the setting of concordant ventriculo-arterial connections.[5] In 1974, Rosenthal et al.[6] described subaortic stenosis developing after banding of the pulmonary trunk in two patients with transposition and interruption of the aortic arch. This, of course, was due to stenosis developing in the outflow tract of the morphologically right ventricle, and is well explained on the basis of hypertrophy of the completely muscular subaortic infundibulum in patients not undergoing the arterial switch procedure.

Freed et al.[7] have also reported documented subaortic stenosis occurring after banding of the pulmonary trunk in patients with functionally univentricular hearts with concordant ventriculo-arterial connections. They proposed leftward deviation and hypertrophy of the muscular outlet septum encroaching on the left ventricular outflow tract as a possible cause for this obstruction. In the setting of functionally univentricular hearts with discordant ventriculo-arterial connections, fibrous tissue can proliferate about the margins of the ventricular septal defect, further compromising aortic flow and producing subaortic stenosis, which can also occur after the Fontan operation, but this is clearly a different mechanism from that encountered in our patient. In our case, the mechanism of subaortic stenosis is related to the underlying morphology of the dominant ventricular morphology, and is not related to prior banding of the pulmonary trunk as seen in some patients undergoing the Fontan operation.[8]

Our patient had not undergone banding of the pulmonary trunk prior to the arterial switch operation.

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In our case, we think it was turbulences created across the newly patched ventricular septal defect that were probably responsible for generating the subaortic stenosis. This mechanism has already been described after intraventricular baffling of the left ventricle to the aorta,[9] with the obstruction being shown by angiography to be located proximally at the site of the ventricular septal crest, and with post-operative echocardiographic findings revealing a contracted ventricle with thickened walls and a small end-diastolic cavity. Such a "contraction phenomenon" has also been reported in the left ventricle after closure of a ventricular septal defect in a patient with functionally biventricular circulations.[10] These authors[9,10] suggested that this complication could develop in any patients undergoing repair of congenital heart disease which involved the acute removal of a chronic volume load. Our experience attests to the validity of their suggestion.

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