Aortico-left ventricular tunnel: two new cases with a long-term follow-up

Carlo Pace Napoleone, Gaetano Gargiulo, Angelo Pierangeli

Department of Cardiac Surgery, University of Bologna, Bologna, Italy

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Aortico-left ventricular tunnel is a rare congenital communication between the ascending aorta and the left ventricle. Its hemodynamic effect is severe aortic incompetence. Surgery is the only treatment and should be performed before aortic incompetence or ventricular dilation develops.

Two neonates with aortico-left ventricular tunnel were operated on at our institution, with closure of the aortic end of the tunnel with a Gore-Tex patch. The 2 patients were discharged in good conditions, and at 112 and 42-month follow-up respectively they are in good health, without medication and with a normal echocardiographic pattern.

Aortico-left ventricular tunnel should be treated surgically as soon as possible in order to prevent any damage to the aortic valve and the left ventricle. The operative risk is not low, but results are very encouraging.

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Address:

Dr. Carlo Pace Napoleone

Dipartimento di Cardiochirurgia
Università degli Studi
Policlinico S. Orsola-Malpighi
Via Massarenti, 9
40138 Bologna
E-mail: pace@orsola-malpighi.med.unibo.it

Aortico-left-ventricular tunnel is a rare anomaly first described by Levy et al.[1] in 1963 as an abnormal paravalvular communication between the ascending aorta and the left ventricle that bypasses the aortic valve. A long-term follow-up of a previously described case[2] and a new patient successfully submitted to surgery are described.

Description of cases

Case 1. A 2-day-old baby weighing 3200 g arrived to our observation with clinical signs of congestive heart failure. Physical examination revealed a 5/6L systo-diastolic murmur. Echocardiographic evaluation performed at 28 weeks of gestation showed aortic valve dysplasia with severe incompetence and progressive hypertrophy and dilation of the left ventricle.

The electrocardiogram (ECG) revealed left ventricular hypertrophy with overload and the chest roentgenogram showed marked cardiomegaly. Two-dimensional echocardiography confirmed the severe left ventricular hypertrophy and dilation with a tunnel-like structure between the aortic valve and the left ventricle. Cardiac catheterization confirmed the diagnosis of aortico-left ventricular tunnel with massive diastolic regurgitation. The patient underwent surgery on the second day of life. A bulge was present in the ascending aorta in the region opposite to the right infundibulum. The valve was bicuspid and an orifice of 10 × 15 mm, directly opening into the left ventricle, was present anterior to and to the left of the non-coronary cusp. The origin of the right coronary artery (RCA) was displaced down into the tunnel. The origin of the tunnel was closed with a Gore-Tex patch introduced via the coronary ostium. The closure of the sternum was delayed to the fifth postoperative day. The patient was extubated on the seventh postoperative day. Echocardiography showed an improvement in contractility with only trivial aortic incompetence. At the last follow-up, 112 months after the operation, the patient is well and presents with normal growth and without medications. The cardiothoracic ratio is normal and echocardiography shows a normal left ventricular contractility without valve incompetence.

Case 2. The patient was born on the 34th week of gestation, weighing 3300 g, and arrived to our observation for a grade 4/6 systo-diastolic murmur. The ECG showed repolarization anomalies with overloading of the right ventricle. Echocardiography showed a bicuspid aortic valve, slightly dysplastic but continent. The ascending aorta was dilated (12 mm in diameter) and a tunnel-like communication between the dilated origin of the RCA (5 mm in diameter) and the left ventricle was highlighted. The hemodynamic effect was similar to

that of severe aortic valve incompetence. The left ventricular contractility was normal. Cardiac catheterization confirmed the diagnosis, showing the run-off of contrast from the dilated ascending aorta through a tunnel into the left ventricular outflow tract. The RCA originated from the dilated portion of the tunnel, with the distal part being perfused by an anomalous vessel from the left coronary artery. Even the latter was dilated up to the origin of the anomalous artery (Fig. 1). The cardiac index was 2.5 l/min/m^2. Rest myocardial scintigraphy showed a big captation defect of the distal part of the antero-lateral wall and apex.

The patient underwent surgery on the fifth day of life. Intraoperative inspection confirmed the presence of a bulge in the anterior portion of the ascending aorta. The aneurysmatic dilation of the first portion of the RCA, with a big vessel interposed between it and the left anterior descending coronary artery, was evidenced. The ascending aorta was opened and a 4 mm ostium in the right sinus of Valsalva was visualized. Owing to the difficult access to the tunnel via this small ostium, the roof of the enlarged portion of the tunnel was incised. The origin of the RCA had been dislocated and seemed to originate from the middle portion of the tunnel (Fig. 2). The aortic and ventricular openings of the tunnel were evidenced. Its examination showed that it did not penetrate the ventricular musculature, confirming the diagnosis in accordance with Ho et al.[3]. A Gore-Tex patch was sutured with interrupted stitches to close the origin of the tunnel, trying to avoid stenosis of the RCA origin. The roof of the tunnel, now functionally RCA, was then closed with a 7/0 poly-dioxanone suture. The patient was admitted in the Intensive Care Unit with a closed sternum and was extubated on the first postoperative day. Rest myocardial scintigraphy showed a marked improvement in the antero-lateral wall and apex perfusion. Echocardiography revealed a normal left ventricular contractility with mild aortic valve incompetence. Forty-two months later the patient is well and echocardiography shows only trivial aortic incompetence with normal contractility.

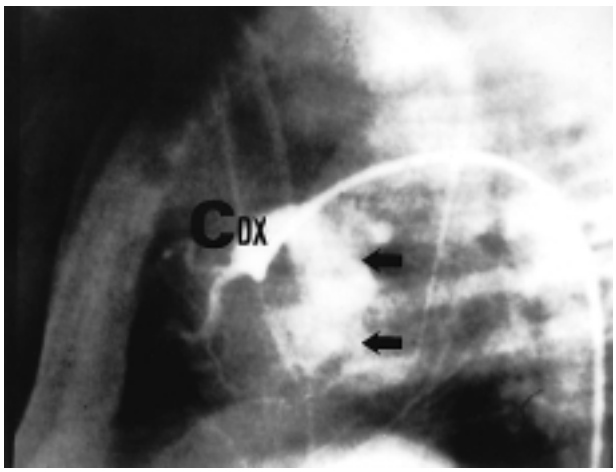

Figure 1. *Angiography showing the dilated origin of the right coronary artery (CDX) with aortico-left ventricular tunnel.* Arrows *indicate aortico-left-ventricular tunnel with contrast injection in the left ventricle.*

Figure 2. *Aortico-left ventricular tunnel with dislocation of the origin of the right coronary artery.*

Discussion

Since the initial description by Levy et al.[1] in 1963, only a few cases of this rare pathology have been reported in the literature. It is characterized by an abnormal paravalvular communication between the aorta and the left ventricle. The aortic opening is above the RCA ostium in 40% of cases, below it in 25% of cases and at the level of the RCA in 16% of cases[4]. A bulge is present on the aortic wall in correspondence of the aortico-left ventricular tunnel. Then the tunnel passes downward beside the aortic valve and through the infundibular septum before entering the left ventricle. Hovaguimian et al.[4] classified four types of aortico-left ventricular tunnel:

1. a simple tunnel with a slit-like opening in the aortic end and no valve distortion;
2. a large extracardiac aortic wall aneurysm of the tunnel with an oval opening at the aortic end, with or without valvular distortion;
3. intracardiac aneurysm of the septal portion of the tunnel, with or without right ventricular outflow tract obstruction;
4. types 2 and 3 combined.

Both our cases had an aortic origin of the tunnel at the level of the RCA origin, and they can be referred to type 2 of Hovaguimian classification[4] (Fig. 2).

The evidence of aortic incompetence at prenatal echocardiography is a reliable sign of the congenital origin of this pathology. There is no agreement about the pathogenesis of aortico-left ventricular tunnel. Levy et al.[1] proposed that the tunnel was an abnormal coronary artery. Spooner et al.[5] and Turley et al.[6] considered the tunnel as an early or intrauterine rupture of a sinus of Valsalva aneurysm. In our second case there was a common origin of the tunnel and the RCA (Fig. 2).

A big collateral vessel originating from the left coronary artery perfused the distal portion of the RCA. A very similar case was described by the group of Turina et al.[7]. In agreement with Levy et al.[1] and the group of Turina et al.[7], we think that the tunnel is the result of the abnormal development of a RCA, while the dilation is acquired. We do not know whether the proximal portion of the RCA is of tunnel[7] or of coronary[1] pertinence, or of both. The operative aspect was not that of an aneurysmatic dilation of the sinus of Valsalva dislocating the origin of the RCA and then breaking into the left ventricle, as proposed in some theories[5,6,8]. The surgical techniques proposed are direct closure of the aortic end of the tunnel, the use of patch material to close the aortic end of the tunnel, and the closure of both aortic and ventricular end of the tunnel with its obliteration[8]. The first two techniques had an incidence of aortic incompetence at follow-up of 23.5 and 25%, while for the third one there has been no long-term follow-up[4]. We chose to close the origin of the tunnel from its proximal side using a Gore-Tex patch that twists the RCA and the aortic valve less[2]. To us it seems that reimplantation of the RCA on the ascending aorta is too difficult[7]. In our cases they were dislocated too much, and reimplantation without excessive tension on the anastomosis would have been impossible. The Gore-Tex patch also allows for a more stable support to the aortic annulus, thus preventing aortic incompetence at follow-up[2].

In conclusion, in the cases in which the coronary circulation is altered by the presence of big abnormal vessels, surgical correction of this pathology is mandatory and it must be performed as soon as possible in order to prevent left ventricular dilation or coronary ischemia. The operative risk is not low, but results are very encouraging.

References

1. Levy MJ, Lillehei CW, Anderson MD, Amplatz K, Edwards JE. Aortico-left ventricular tunnel. Circulation 1963; 27: 841.
2. Zannini L, Gargiulo G, Albanese SB, et al. Successful surgical repair of an aortico-left ventricular tunnel in a two day old child. J Cardiovasc Surg 1992; 33: 295-7.
3. Ho SY, Muriago M, Cook AC, Thiene G, Anderson RH. Surgical anatomy of aorto-left ventricular tunnel. Ann Thorac Surg 1998; 65: 509-14.
4. Hovaguimian H, Cobanoglu A, Starr A. Aortico-left ventricular tunnel: a clinical review and new surgical classification. Ann Thorac Surg 1988; 45: 106-12.
5. Spooner EW, Dunn JM, Behrendt DM. Aortico-left ventricular tunnel and sinus of Valsalva aneurysm. Case report and operative repair. J Thorac Cardiovasc Surg 1978; 75: 232-6.
6. Turley K, Silverman NH, Teitel D, Mavroudis C, Snider R, Rudolph A. Repair of aortico-left ventricular tunnel in the neonate: surgical, anatomic and echocardiographic considerations. Circulation 1982; 65: 1015-20.
7. Grunenfelder J, Zund G, Pretre R, Schmidli J, Vogt PR, Turina MI. Right coronary artery from aorto-left ventricular tunnel: case report of a new surgical approach. J Thorac Cardiovasc Surg 1998; 116: 363-5.
8. Myers JL, Mehta SM. Congenital Heart Surgery Nomenclature and Database Project: aortico-left ventricular tunnel. Ann Thorac Surg 2000; 69: S164-S169.