Aortico-Left Ventricular Tunnel

Aortico–left ventricular tunnel is a congenital extracardiac channel connecting the ascending aorta above the sino-tubular junction to the left or —less common— right ventricular cavity. (1)

It is extremely rare, and the exact incidence is unknown. Estimates range from 0.5% of fetal cardiac malformations to less than 0.1% of congenitally malformed hearts in clinico-pathological series.

To date, approximately 130 cases have been reported in the literature, about twice as many cases in males as in females. (2, 3) Associated defects, usually involving the proximal coronary arteries, or the aortic or pulmonary valves, are present in nearly half the cases. (4)

Some patients have asymptomatic heart murmur, but most of the patients develop symptoms of heart failure during the first year of life.

The onset, severity and progression of heart failure vary and ranges from in-utero fetal death to asymptomatic adulthood. Onset of heart failure depends on the cross-sectional area of the tunnel and the amount of aortic regurgitation. Chronic preload due to regurgitation leading to left ventricular (LV) dilatation is seen in asymptomatic grown-up patients. (2, 3)

The etiology is uncertain; it appears to result from a combination of maldevelopment of the cushions which give rise to the pulmonary and aortic roots, and abnormal separation of these structures. (1, 2)

Early diagnosis and surgical correction are essential to prevent irreversible myocardial dysfunction and heart failure. Doppler echocardiography is the diagnostic method of choice, and after correction, all patients require lifelong follow-up because of the risk of tunnel recurrence, aortic valve incompetence, left ventricular dysfunction and aneurysmal enlargement of the ascending aorta. (6)

We report the case of a 2-year-old male patient, referred by his pediatrician due to auscultation of a murmur during clinical monitoring. His parents reported that he had episodes of dyspnea associated with exercise, with no other symptoms.

Physical examination showed a weight of 12.5 kg (60th percentile) and a height of 88 cm (50th percentile). On auscultation, normal S1 and S2, no S3, and a systolic-diastolic murmur in the mitral-aortic area were detected.

ECG revealed sinus rhythm, indeterminate QRS axis and signs of left ventricular hypertrophy with negative T-waves in V1 to V3 precordial leads.

Echocardiography showed dilated sinus of Valsalva and flow in diastole from the right coronary sinus to the left ventricle outside the aortic annulus. Pressure half time of 153 msec. Reverse flow in the abdominal aorta. Left ventricular enlargement, with left ventricular diastolic diameter of 45 mm (SD Z Score +5.7). Mitral valve with restricted anterior leaflet opening secondary to regurgitant jet; mean gradient 8 mmHg. Mild mitral regurgitation. Patent ductus arteriosus of 1.5 mm with gradient of 29 mmHg. Normal coronary arteries.

The case was discussed in a grand round, and surgical closure of the congenital aortico-left ventricular tunnel was decided (Figure 2).

A median sternotomy was performed, after aortic and bicaval cannulation, cardiopulmonary bypass was performed at hypothermia (30 °C). The aorta was

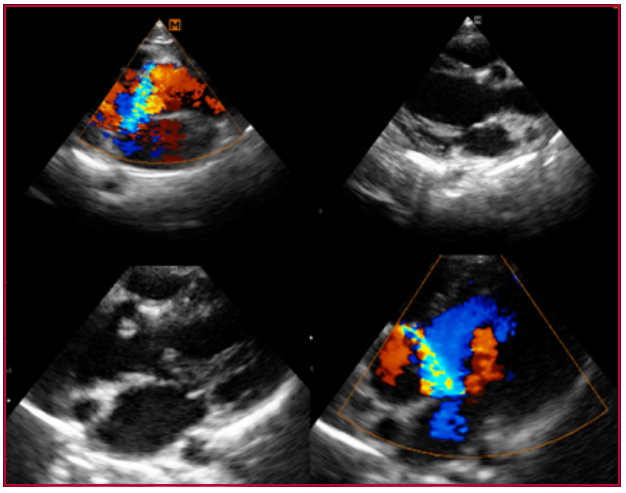

Fig. 1. Transthoracic echocardiography: LV long axis and 5 chambers, showing regurgitant jet outside the aortic annulus.

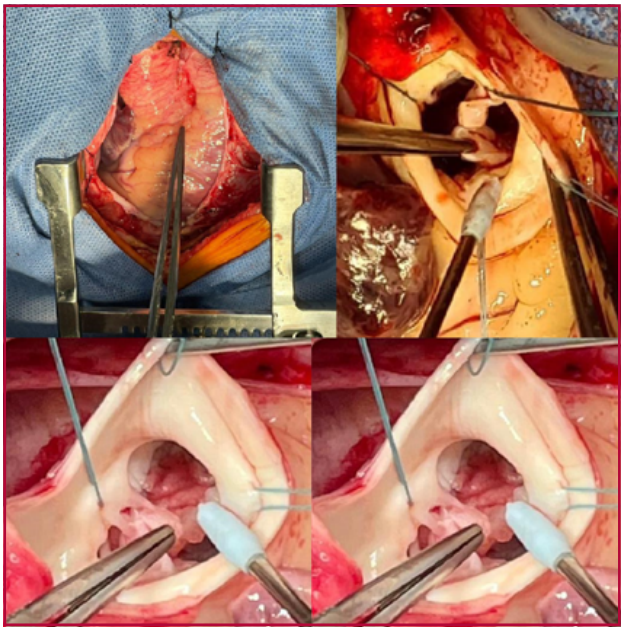

Fig. 2. Aorta, prior to aortotomy. Open aorta showing the defect/tunnel. Tunnel closed with pericardial patch

clamped and antegrade cardioplegia was performed. Transverse aortotomy and cardioplegia were performed through both coronary ostia. Aortico-left ventricular tunnel was identified in the right coronary sinus, with unsupported right coronary leaflet. The tunnel orifice was closed with an autologous pericardial patch treated with a nadir of the right coronary leaflet. Aortorrhaphy, purging and aortic unclamping were performed. Bypass output, decannulation and chest closure without complications.

The patient made good postoperative progress, and was extubated within 6 h of the postoperative period on inotropic drugs in usual doses, which were stopped at 24 h. Mediastinal drainage was removed. The patient was discharged at 72 h, and continued with outpatient follow-up.

Postoperative echocardiography showed tunnel closure and no residual shunt. Free left ventricular outflow tract, velocity 1.07 m/sec. Normal aorta. Wider mitral valve opening, slightly redundant, mean gradient 0.7 mmHg. Enlarged left ventricle —improved in comparison to preoperative study—, with acceptable function and hypokinetic, rectified interventricular septum. Shortening fraction 30%. Mild tricuspid regurgitation. No pericardial effusion.

The aortico-left ventricular tunnel is a rare entity and constitutes a challenge for pediatricians and pediatric cardiologists. Once diagnosed, it should be fixed early, mainly due to aortic annulus enlargement and to different degrees of valve incompetence that will result in mid- and longer-term poor prognosis.

While several techniques have been described, double-patch or single-patch closure of the aortic end are the most commonly used. Closure with an occluder device is another treatment option, but with higher rates of complications; therefore, cardiopulmonary bypass surgery continues to be the gold standard with a favorable mid- and long-term prognosis, when timely treated.

Conflicts of interest

None declared.

(See authors' conflict of interests forms on the web/Additional material.)

Ethical considerations

Not applicable.

María Florencia Núñez, Evangelina Papini, Martín Irazusta, Francisco González, Marcela Vyera, Pablo García Delucis

María Florencia Núñez, MD
Hospital de Niños Dr. Orlando Alassia
Santa Fe - Argentina
E-mail: maflorencianunez.91@gmail.com

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Rev Argent Cardiol 2022;90:300-301.
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Intracranial Hemorrhage and Symptomatic Coronary Artery Disease as a Form of Presentation of Moyamoya Disease in Adult Patients

We present the case of a 42-year-old male patient with a history of hypertension, smoking, dyslipidemia, and physical inactivity, who was admitted for headache, 72 h-history of unsteady walking, and seizures. On physical examination, the patient was lucid and tolerated supine position; bilateral blood pressure: 170/100mmHg. Cardiovascular system: normal, regular S1 and S2, no S3, no cardiac murmurs or lower limb edema. Preserved peripheral pulses; neurologically, increased wide-based gait.

ECG: sinus rhythm, heart rate 96 bpm. Left ventricular overload.

Chest X-ray: cardiothoracic ratio < 0.5.

Lab tests (blood count, liver function and kidney function tests) with normal values.

After evaluation by the Department of Neurology, the brain CT scan reported bilateral subarachnoid hemorrhage with ventricular dump; therefore, a digital angiography was performed, revealing (Figure 1) severe lesion in the origin of the right internal carotid artery and decreased distal flow, consistent with carotid dissection. Terminal branches, anterior and middle cerebral arteries, appear to be filled by collateral circulation from the external carotid artery, with multiple wall irregularities, abnormal arborization and multiple anastomoses; left common carotid artery with a mild distal lesion; reduced diameter of the left internal carotid artery, affecting the entire length of the vessel. Terminal branches, anterior and middle cerebral arteries, appear to be filled by collateral circulation from the external carotid artery, with multiple wall irregularities, abnormal arborization and multiple anastomoses. A basilar apex aneurysm with an apparent narrow neck is observed. Terminal branches show multiple wall irregularities, abnormal arborization and multiple anastomoses.

The patient was submitted to endovascular treatment with embolization of the basilar artery. Postoperative follow-up brain CT scan at 72 h showed partial ventricular dump absorption. Modified Fisher scale, 4; Hunt and Hess scale, 1. In the Intensive Care Unit, the patient referred precordial pain; ECG showed sinus rhythm, anterior negative T-wave (Figure 2); troponin was positive. Non-ST segment elevation acute coronary syndrome (NSTE-ACS) was diagnosed. Coronary angiography revealed left main coronary artery and right and circumflex coronary arteries without significant lesions, and severe lesion in the middle third of the left anterior descending artery (LAD); percutaneous coronary intervention (PCI) with conventional stent of 2.75 x 16 mm was performed (Figure 3A & B). The patient was discharged asymptomatic on enalapril, phenytoin, atorvastatin, aspirin and clopidogrel.

Four months after angioplasty, Doppler echocardiography showed left ventricular diastolic diameter of 44mm and systolic diameter of 23 mm; septum 8 mm, left atrium 34 mm, and LVEF (left ventricular ejection fraction) 78%; normal motility, and LV diastolic dysfunction. Tc-99m gated SPECT revealed apical and anterior ischemia from apex to base, LVEF 54%, septal hypokinesis with preserved systolic thickening. Stress test reported normal ST-T, pseudonormalization of T-wave, 10 METs, and double product 22500. Follow-up coronary angiography (Figure 4) showed restenosis of the LAD due to in-stent occlusion in the middle third portion, and obtuse marginal lesion of 80%. PCI to LAD using a drug-eluting stent (Sirolimus 2.75 x 24 mm) and with conventional stent of 2.75 x 16 mm was performed. The patient was discharged asymptomatic, on aspirin, bisoprolol, clopidogrel, atorvastatin, and phenytoin. The follow-up stress test 5 months after the second PCI was negative for

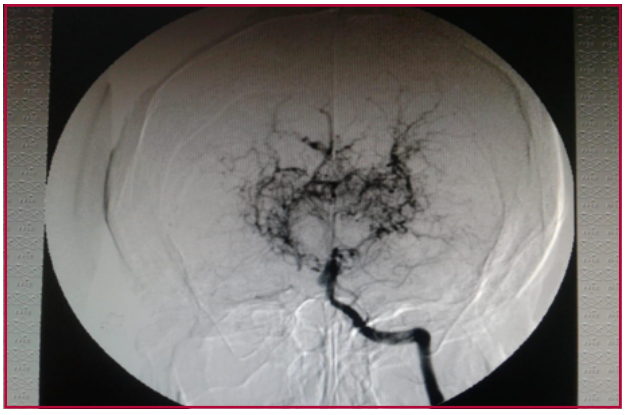

Fig. 1. Digital angiography: Right internal carotid artery : Severe lesion in its origin, with decreased distal flow, consistent with carotid dissection; terminal branches, anterior and middle cerebral arteries, appear to be filled by collateral circulation from the external carotid artery, with multiple wall irregularities, abnormal arborization and multiple anastomoses

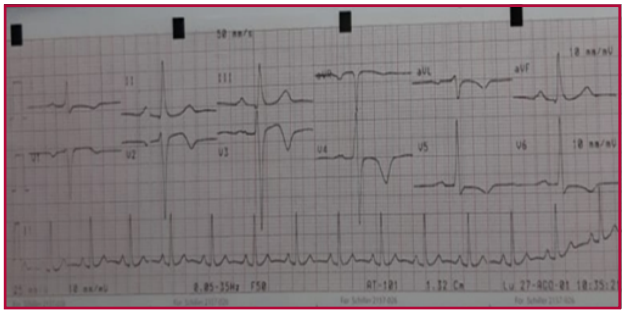

Fig. 2. ECG: Sinus rhythm, anterior negative T-wave.

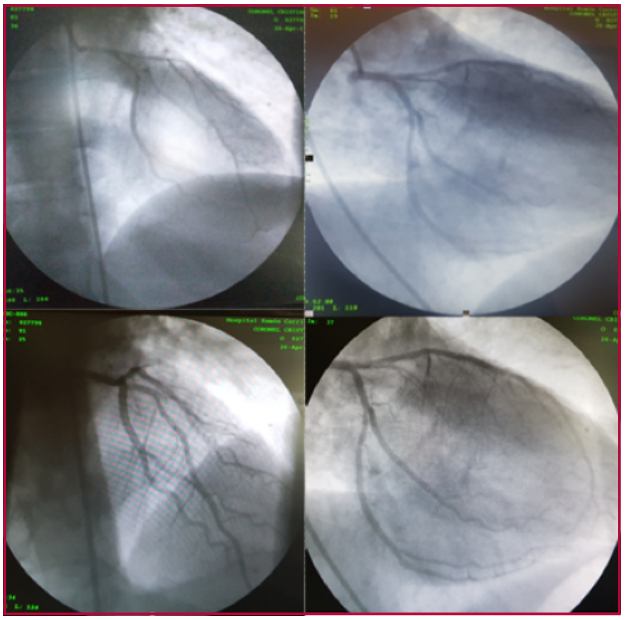

Fig. 3. Coronary angiography: Severe lesion in the middle third of the left anterior descending artery (LAD) (Top Figure). Coronary angioplasty of LAD with conventional stent of 2.75 x 16 mm (Bottom Figure).

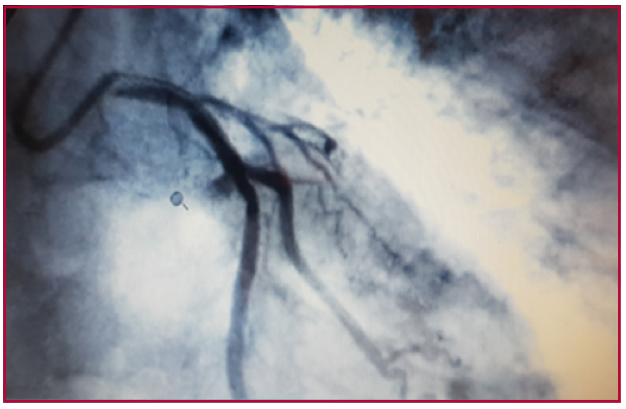

Fig. 4. Percutaneous coronary angioplasty: Left anterior descending artery (LAD) with restenosis due to in-stent occlusion in the middle third portion

angina and ST-T, reaching 9 METs. A year later, the patient referred precordial pain; a follow-up perfusion study was performed, revealing apical and anterior-apical ischemia. Stress test reported ST-T elevation, 8.6 METs, double product 20460. Coronary angiography revealed occluded stent origin (Mehran Score IV restenosis) with distal recanalization by hetero-coronary collateral circulation, Cohen grade 3. Given the angiographic characteristics (J-CTO Score 2, very difficult) the patient was not eligible for revascularization; medical treatment was optimized and cardiac rehabilitation was indicated.

Moyamoya disease (MMD) is a rare, progressive cerebrovascular disease of unknown etiology, characterized by progressive stenosis/occlusion of the distal portion of both internal carotid arteries or branches, compensated by small collateral vascular formation, which may or may not be accompanied by a defect or anomaly of the anterior and middle cerebral arteries associated with abnormal vascular network of vessels called moyamoya. The vessels get through the basal ganglia and thalamus, and provide collateral flow to distal hypoperfused areas (diagnostic criteria for MMD, Research Committee on Spontaneous Occlusion of the Circle of Willis, Japan) (1); pathogenically, MMD has been associated with genetic and environmental factors; the familial form of the disease accounts for 15% of cases.

Isolated cases of coronary artery disease have been described in MMD. A study on the prevalence of MMD on 456 patients showed that 4.6% had symptomatic coronary heart disease. The associating mechanism could be related to genetic factors or the result of atherosclerosis due to underlying endothelial proliferation. (2) Other authors found that, in male and female patients, MMD can present as diffuse, multivessel variant angina and infarction, and stenotic lesions in the epicardial coronary arteries remained undetected by coronary angiography. (3) The stenotic lesions in MMD are different from those in patients with atherosclerotic coronary artery disease in that they are composed of fibrous thickened tissue of the intima and soft intimal proliferation with minimum lipid pooling, but without any calcium deposition.

Another study used intravascular ultrasound and virtual histology to evaluate a coronary lesion in a patient with MMD, and revealed homogeneous, eccentric, echogenic intimal thickening composed of fibrous tissue; the mid and distal portions showed no intimal thickening. (4) A genetic study found that the p. R4810K variant in RNF213 appears to be significantly associated with coronary artery disease in the Japanese population, with no difference by sex. (5) In a report of a patient with MMD and NSTE-ACS, coronary angiography showed several areas in which the vessel resembles a beaded necklace —previously described— in peripheral pulmonary, renal and carotid arteries, as a direct result of intimal thickening.

Table 1. Staging System of Suzuki and Takaku

Grade	Vascular lesion
1	Narrowing of the ICA apex
2	Incipient MM collateral vessels
3	Progressive stenosis of the ICA and intensification of collateral vessels
4	Development of collaterals from the ECA
5	Intensification of ECA collaterals and reduction of moyamoya vessels
6	Total ICA occlusion and disappearance of collaterals, with supply from ECA

ICA: internal carotid artery. MM: moyamoya. ECA: external carotid artery.

Suzuki and Takaku described this —usually bilateral— entity, but there are also unilateral cases. (6) Its incidence in the Asian population is 0.35-0.94/100 000 inhabitants, with a higher prevalence in women (1.8:1). It occurs between the ages of 5-9 and 45 years; it may be asymptomatic or manifested by ischemic or hemorrhagic stroke, epilepsy, cognitive impairment or severe neurological deficit. Mortality rate is 5% in adults.

MMD is described as a bilateral process, without associations; Quasi-Moyamoya is a unilateral and/or bilateral process with associations, and Probable Moyamoya is the presence of a unilateral process, without associations; this semantic/regional classification does not affect clinical management.

When MMD is associated with sickle cell anemia, Down syndrome, neurofibromatosis type 1, autoimmune disease, cranial radiation, etc., it is called moyamoya syndrome. Cerebral angiography is the diagnostic tool. Pharmacological or surgical treatment does not reverse the primary disease process, but may prevent secondary cerebrovascular events and improve cerebral blood flow; options include low-molecular weight heparin, cilostazol, or clopidogrel. The AHA and Stroke Guidelines (AHJ/2008) recommend aspirin use in children with MMD after revascularization or in asymptomatic patients with delayed surgery. Revascularization surgery is effective in the treatment of moyamoya syndrome. The Japan Adult Moyamoya Trial identifies the temporo-sylvian bypass as the treatment of choice in adults, and indirect revascularization in the pediatric population.

According to Suzuki, the disease is classified into 6 progressive stages (Table1).

During follow-up of patients with MMD, detecting extracranial vessel involvement should be considered —as in the case presented— so that tests for coronary artery disease diagnosis are necessary.

Conflicts of interest

None declared.

(See authors' conflicts of interest forms on the website/ Supplementary material).

Ethical considerations

Not applicable.

Jorge L. Goral[1-MTSAC], Osvaldo A. Barbosa[1], Alejandra M. Gómez[1], Felipe D. Olivieri[1], Carlos A. Mallama[1], Roberto Farfan[1]

[1] Centro de Atención Primaria para la Salud CAPS 10. Tres de Febrero

Jorge Luis Goral. Department of Cardiology

Centro de Atención Primaria para la Salud CAPS N.º 10. Caseros

Tres de Febrero, (1678) Buenos Aires, Argentina

jgoral@intramed.net

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