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Editorial

Surgical management of giant coronary aneurysms in Noonan syndrome

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A 19-year-old patient with Noonan syndrome with previously diagnosed severe pulmonary stenosis was admitted to our institution for chest pain both at rest and on exertion.

Electrocardiogram showed sinus tachycardia with complete right bundle-branch block and right axis deviation.

Troponin Ic was elevated at 3 ng/mL (normal value < 0.02) with no other abnormalities on blood tests.

Echocardiography (Fig. 1) showed a severely dilated and hypertrophied right ventricle with severe pulmonary stenosis (peak transpulmonary pressure gradient 104 mm Hg), mitral prolapse with mild mitral regurgitation. Left ventricular systolic function was normal with no wall motion abnormality. The exam also showed a dilated circumflex artery (Cx) on parasternal and apical 4 chamber views.

CT coronary angiogram (Fig. 2A) was performed, followed by *coronary angiography* (Fig. 2B), and revealed diffuse dilatation of coronary vessels (>15 mm) — classified as type 1 ectasia [1], and anomalous origination of the left anterior descending (LAD) from the right sinus. Both LAD and right coronary artery (RCA) were proximally occluded: LAD at its origin from an ectatic left main stem, and RCA at the genu superius after the origin of conus artery and right marginal branch. Collateral vessels issued from ectatic right branches and supplied mid-LAD and distal RCA. Circumflex artery (Cx) also presented fusiform ectasia, with slow flow but no thrombosis.

After multidisciplinary discussion with the heart team, the patient underwent heart surgery, consisting in a pulmonary valve commissurotomy and a double coronary bypass (left internal thoracic artery grafted to LAD and left radial artery grafted to RCA) (Fig. 2C).

The patient was discharged after surgery and followed up in the outpatient clinic. Two months after the intervention, clinical status

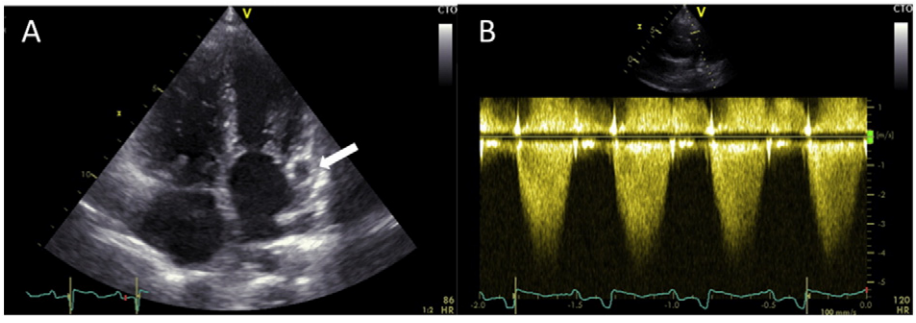

Fig. 1. Echocardiography. A: Apical 4 chamber view showing right ventricular dilatation and circumflex artery aneurysm (arrow). B: Continuous Doppler of pulmonary valve ejection flow.

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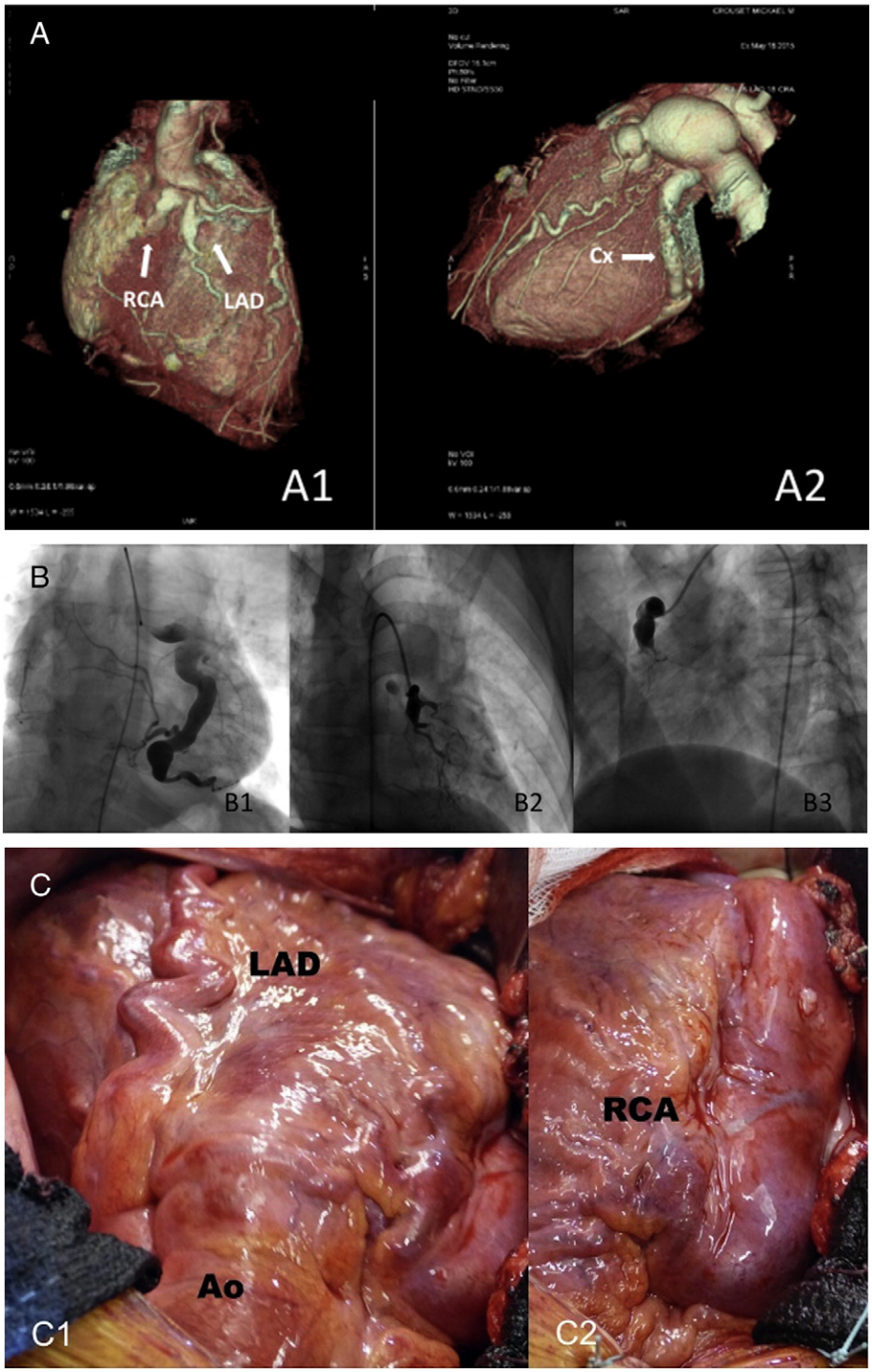

Fig. 2. A. CT coronary angiogram. A1: Left anterior descending (LAD) and right coronary artery (RCA) dilated at their origin and occluded at their proximal segment. A2: Aneurysmal circumflex artery (Cx). B. Invasive coronary angiography. B1: Aneurysmal circumflex artery (Cx). B2: Left anterior descending artery (LAD) ectatic at its origin and occluded at its proximal segment. B3: Right coronary artery (RCA) with a dilated origin and an occluded proximal segment. C. Intraoperative view of ectatic coronary arteries. C1: Left anterior descending artery (LAD). C2: Right coronary artery (RCA).

was good, and mild to moderate residual pulmonary stenosis was found on echocardiography (peak transpulmonary gradient 29 mm Hg).

Noonan syndrome is an autosomal dominant genetic syndrome characterized by the association of short stature, facial dysmorphy and congenital cardiac malformations. It was first described by Jacqueline Noonan in 1963, and its prevalence is 1 in 1000 to 1 in 2500. Most common cardiac abnormalities are pulmonary stenosis and hypertrophic cardiomyopathy. Giant coronary aneurysms have been seldomly described in Noonan syndrome [2–4] but may result in potentially life-threatening complications. Multimodality cardiac imaging can provide an accurate assessment of coronary anatomy in order to implement appropriate screening and therapeutic strategies for these complex patients. The present case report suggests that CABG may be an appropriate therapeutic option in case of coronary aneurysms-related ischemic complications.

Conflict of interest

The authors do not have any conflict of interest to declare.

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