Cardiology in the Young

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Dealing with kinked and swirled pulmonary vessels: surgical treatment of arterial tortuosity syndrome: a case report

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Abstract

We present the case of a child with arterial tortuosity syndrome, describing the operative findings and our surgical technique to address pulmonary arteries stenosis.

Brief Report

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Arterial tortuosity syndrome is a rare autosomal recessive connective tissue disorder with distinctive vascular features[1] and a mortality rate up to 40% before the age of 5.[2,3]

Glucose transporter 10 (GLUT10) deficiency, owing to mutations in SLC2A10 gene, induces the up-regulation of TGF-beta pathway in the arterial wall and a distinguishing angiopathy, with tortuosity and elongation of elastic vessels, aneurysms, dissection, and pulmonary arteries stenoses.

The incidence of this latter condition is around 60%,[4] revealing a localised or diffuse pattern, this one coupled with early presentation and right ventricular hypertension.[5]

The management options include catheterisation and/or surgery.[4]

To date, outcomes and natural history after stenoses' repairs remain poorly investigated.[3] Therefore, the treatment choice is based on expert opinion.[6]

Materials and methods

We present the case of a 4-year-old child with a confirmed genetic diagnosis, diffuse pulmonary arteries obstructions, history of hiatal and bilateral inguinal hernia surgical repairs, neither cardiac nor neurological symptoms. Echocardiogram findings included: dilated right chambers, right ventricular hypertension, preserved ejection fraction, systolic flattening of interventricular septum, laminar flow in right ventricular outflow tract, mild tricuspid valve regurgitation (transvalvular gradient 90 mmHg), no shunts at the level of atria or ventricles.

CT scan showed tortuosity, kinking and elongation of pulmonary arteries' bifurcation (Figs 1 and 2). Right pulmonary artery diameter range was 14–5 mm. Left pulmonary artery exhibited a truncated aspect in the hilus; the upper lobe artery origin appeared obstructive, the lower lobe artery diameter was 3 mm at segmental level.

Supra-aortic vessels and the largest cerebral arteries revealed prominent tortuosity.

The heart catheterisation recorded the following pressures: 96/3 mmHg (right ventricle), 84/26 mmHg – 29/12 mmHg (proximal – distal right pulmonary artery), 92/28 mmHg (proximal left pulmonary artery).

Right ventricular hypertension formed the main indication for intervention within Heart Team discussion. In view of the distinctive anatomic arrangement, surgical management of the largest pulmonary branches and delayed stenting of the tight left pulmonary artery origin were originally agreed upon.

Surgery was conducted through a median sternotomy. Intraoperative inspection was in agreement with preoperative findings.

Dissection of the main and branching pulmonary arteries was carried into the hili bilaterally, performing vessels' reconstruction on cardiopulmonary bypass with mild hypothermia (30°C), aortic cross-clamping and arrested heart. The arterial wall appeared thicker and firmer than expected for a connective tissue disorder. The segment of left pulmonary artery close to the insertion of ligamentum arteriosum (previously transected) was resected. A sliding technique was preferred to provide a wide anastomosis after the repair, adopting the same method on the right side and resecting of a long and narrow portion of the right pulmonary artery (Fig 3).

The left upper lobe branch stenosis was left untouched, according to the preoperative plan.

No sample was sent to the histopathology, since the diagnosis had been already confirmed by genetic tests.

The patient was weaned from cardiopulmonary bypass uneventfully.

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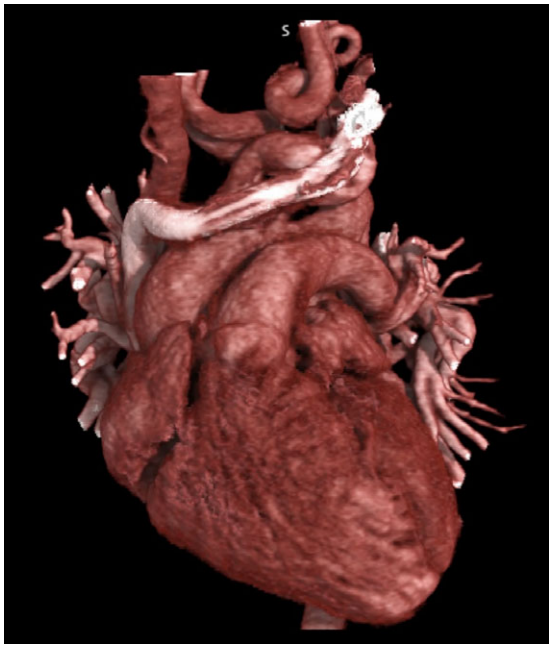

Figure 1. CT scan reconstruction showing pronounced tortuosity and kinking of systemic and pulmonary vessels.

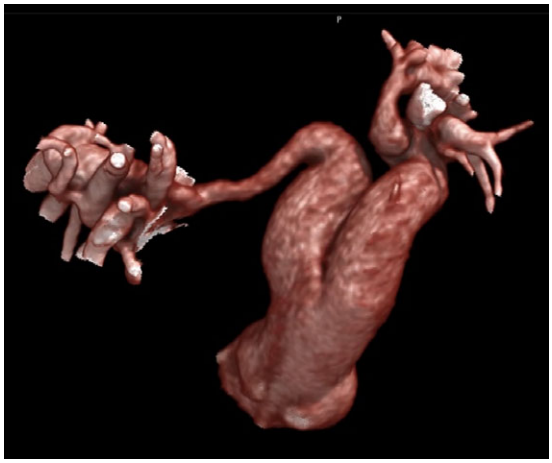

Figure 2. CT scan reconstruction showing pronounced tortuosity and kinking of the pulmonary vessels.

Results

Diameters and final position of pulmonary arteries seemed satisfying after the procedure. The right ventricular pressure dropped to about 60% of the systolic pressure with normalisation of septal systolic flattening.

The patient was discharged home on the 7th post-operative day without anticoagulation therapy, following an uneventful clinical course.

Five months later, the patient underwent the planned heart catheterisation, measuring these pressures: non-invasive blood pressure 70/33 mmHg, 5/3 mmHg (right atrium), 50/-2 (mean 17) mmHg (right ventricle), 46/6 (23) mmHg – 14/8 (12) mmHg (proximal – distal right pulmonary artery), 48/9 (25) mmHg (main pulmonary artery), 47/7 (23) mmHg – 11/7 (9) mmHg (proximal – distal left pulmonary artery). The agreed stenting procedure was not deemed possible due to the unexpected extension and kinking in the distal course of the branching left pulmonary artery (Fig 4).

Discussion

The distinguish angiopathy of pulmonary arteries in this syndrome depends upon a disproportionate growth of one segment of the involved vessels compared to the adjacent ones.

The yielded vascular wall tissue excess[7] is the cornerstone both surgeons and interventionists have to consider when discussing how to address pulmonary arteries' stenoses.

In our patient, the anatomic arrangement was deemed unsuitable for surgical repair or stent angioplasty alone, originally fitting best to a combined surgical/interventional cardiology strategy.

Santoro et al. previously described a hybrid approach for a similar clinical setting.[8] Proximal pulmonary arteries redundancy was again the primary source of obstruction, carrying a substantial discrepancy between the elongated proximal vessels and mediastinum. In such configuration, surgery or angioplasty alone would provide an incomplete result, moving further the stenosed sites and boosting kinking and weakness, this eventually endorsing onward vessels' elongation and tortuosity.

In this report,[8] the stenting procedure was performed in the cathlab at the time of surgery, realising a hybrid strategy. Yet, we decided to defer the interventional procedure, considering age and growth potential after the surgery should be carefully evaluated while selecting the best management for these patients. Indeed, being this syndrome by definition progressive, we judged a combined surgical/interventional cardiology approach the most effective: while surgery would have provided geometric realignment of main branches, stents would have been reliable for smaller vessels and dilatable to the adult size, whenever clinical conditions warranted them. Moreover, since segmental and subsegmental branching pulmonary arteries are extremely thin and fragile, we rather avoided their surgical manipulation. Likewise, to prevent the risk of right ventricular decompensation associated with right ventricular hypertension while handling these branches, a cardiopulmonary bypass strategy was favored. The need to hinder a suboptimal result guided the preference for aortic cross-clamping.

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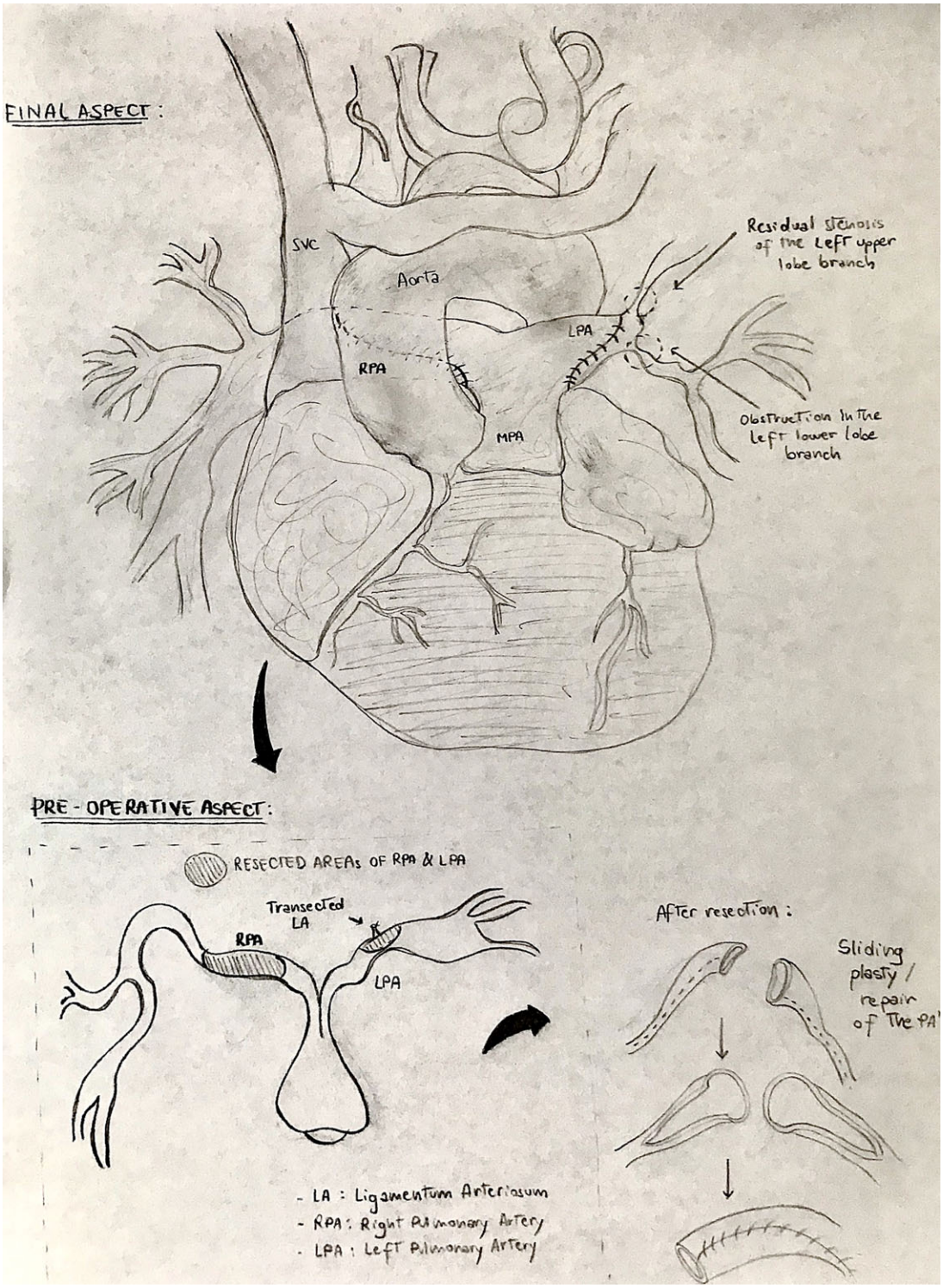

Figure 3. Personal drawing of the final aspect and technical details of the procedure.

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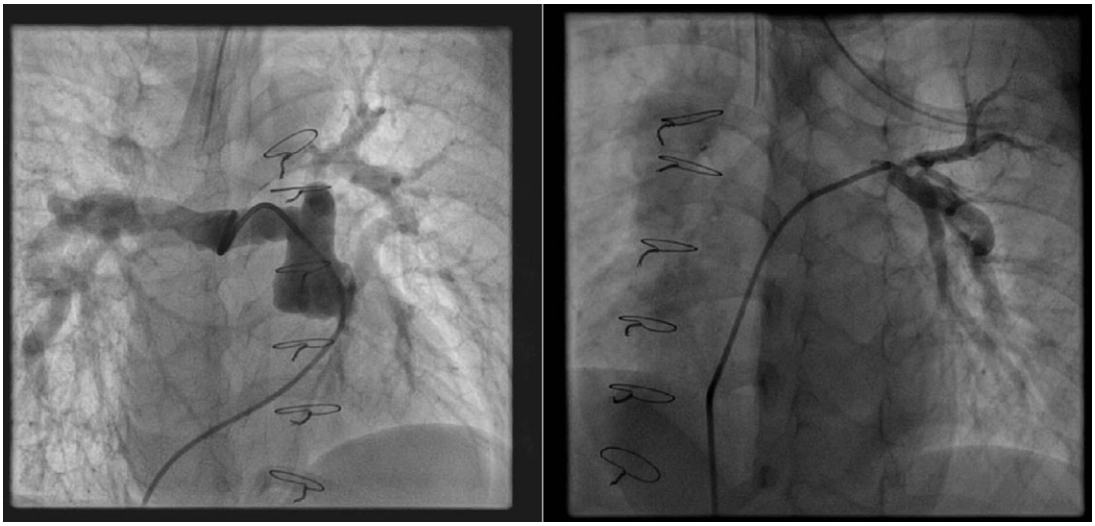

Figure 4. HC pictures showing RPA and LPA 5 months after surgery.

The considerable extension and kinking in the distal course of the branching left pulmonary artery perceived during the post-operative catheterisation made the planned stenting and/or ballooning procedure not possible. Furthermore, we considered acceptable a measured right ventricular pressure of about 2/3 of the systemic one.

In conclusion, even if little is known about the natural history and evolution after pulmonary arteries' repair in this disease,[7] early diagnosis and multimodal evaluation of the unique pattern of vascular lesions in the individual patient is key to achieve the proper management and monitoring.

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Conflicts of interest. None.

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