

# Acute pulmonary artery dissection in an adult with chronic pulmonary hypertension secondary to congenital heart disease: a case report

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## Background

D-Transposition of the great arteries (d-TGA) is characterized by the aorta positioned above the right ventricle and the pulmonary artery above the left ventricle. Acute pulmonary artery dissection (PAD) is a rare and often lethal condition. We present a case report of acute PAD in an adult with d-TGA and pulmonary hypertension.

## Case summary

A 49-year-old male with history of d-TGA palliated with an atrial switch (Mustard) operation, pulmonary venous baffle stenosis treated percutaneously, chronic pulmonary hypertension (mixed group 1 and 2), and severe dilatation of pulmonary arteries (pulmonary trunk of 75 mm) presented to the emergency department with chest pain and acute respiratory failure. Blood pressure was 106/78 mmHg, heart rate 93 b.p.m., and oxygen saturation 88% on room air. A computed tomography (CT) scan showed acute right PAD. He was not considered suitable for surgery nor percutaneous procedure. Epoprostenol was initiated to reduce parietal stress, but after initial stabilization, pulmonary venous stent baffle gradient increased and acute pulmonary oedema occurred. Epoprostenol was withdrawn, and furosemide was initiated, with good clinical response. A follow-up CT scan showed dissection morphological stabilization and false lumen thrombosis, and the patient was discharged.

## Discussion

Pulmonary hypertension and previous pulmonary artery dilatation are reported as the main underlying conditions leading to PAD. No previous cases of PAD are described in patients with history of d-TGA and atrial switch procedure. Evidence regarding the best treatment of PAD is lacking, but it seems reasonable to reduce parietal stress using pulmonary vasodilators. Nevertheless, in patients with post-capillary pulmonary hypertension, pulmonary vasodilatation may cause important pulmonary congestion.

## Keywords

Acute pulmonary artery dissection • D-Transposition of the great arteries • Congenital heart disease • Pulmonary hypertension • Case report

## ESC curriculum

2.4 Cardiac computed tomography • 9.7 Adult congenital heart disease • 9.6 Pulmonary hypertension • 7.3 Critically ill cardiac patient

## Learning points

- Acute pulmonary artery dissection can be a complication of pulmonary hypertension and pulmonary artery dilatation.
- Pulmonary vasodilation in patients with post-capillary pulmonary hypertension may worsen pulmonary congestion.

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## Introduction

D-Transposition of the great arteries (d-TGA) is a congenital heart disease characterized by atrioventricular concordance and ventriculo-arterial discordance, with the aorta positioned above the right ventricle and the pulmonary artery above the left ventricle.<sup>1</sup>

The current standard for restoring physiological circulation in patients with d-TGA is the arterial switch operation, also known as the Jatene procedure, which consists of arterial trunk transection and re-anastomosis to the contralateral root and re-implantation of the coronary arteries in the neo-aorta.<sup>2</sup>

However, the majority of older adults with d-TGA underwent atrial switch surgery, which was the treatment of choice until a few decades ago. In atrial switch procedures, connection between the systemic and pulmonary circulation is restored by the construction of a systemic venous baffle that redirects blood to the subpulmonary left ventricle and a pulmonary baffle that redirects blood to the systemic right ventricle. The baffle is made of Dacron or pericardium in the Mustard operation and of atrial flaps in the Senning operation.<sup>3,4</sup>

Systemic right ventricular systolic dysfunction with increased filling pressures and consequently pulmonary hypertension have been described as late-onset complications of the atrial switch procedure, even though the presence of predominant pre-capillary pulmonary hypertension is less frequent.<sup>5,6</sup>

Pulmonary artery dissection (PAD) is a very rare entity, most frequently described post mortem.<sup>7</sup> In this case report, we describe severe combined post- and pre-capillary hypertension and pulmonary artery dilatation in a patient with d-TGA and atrial switch palliation as possible risk factors for acute PAD.

## Summary figure

## Case presentation

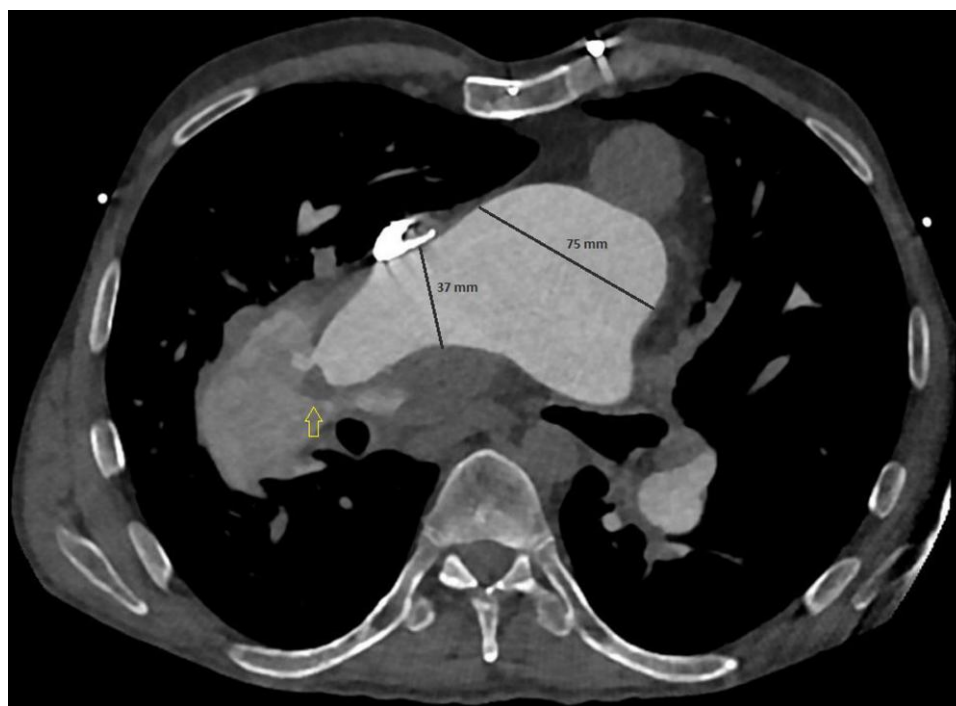
A 49-year-old male presented to our Spanish tertiary centre with general malaise, cough, chest pain, mild haemoptysis, and progressive shortness of breath over the last week.

His medical history was significant for d-TGA palliated with a Mustard operation at 15 months old and pulmonary venous baffle stenosis treated with percutaneous transcatheter stent placement at 35 years old. Over the years, he developed pulmonary hypertension, and in a more recent follow-up, he was found to have severe pulmonary hypertension of mixed group 1 and 2, with a mean pulmonary pressure of 81 mmHg, a pulmonary arterial wedge pressure of 19 mmHg, and a pulmonary vascular resistance of 10 Wood units.<sup>8</sup> The patient suffered severe biventricular dilatation with moderate dysfunction and severe dilatation of the pulmonary trunk (75 mm) and pulmonary arteries (Figure 1), with a growth rate of 2 mm/year over the previous 4 years. Furthermore, the patient had suffered isthmus atrial flutter successfully ablated and a subcutaneous cardioverter-defibrillator had been implanted for a primary prevention indication due to the systemic right ventricular dysfunction, a QRS amplitude of 140 ms, and World Health Organization functional class (WHO-FC) II–III. The patient was in the process of a heart–lung transplant evaluation.

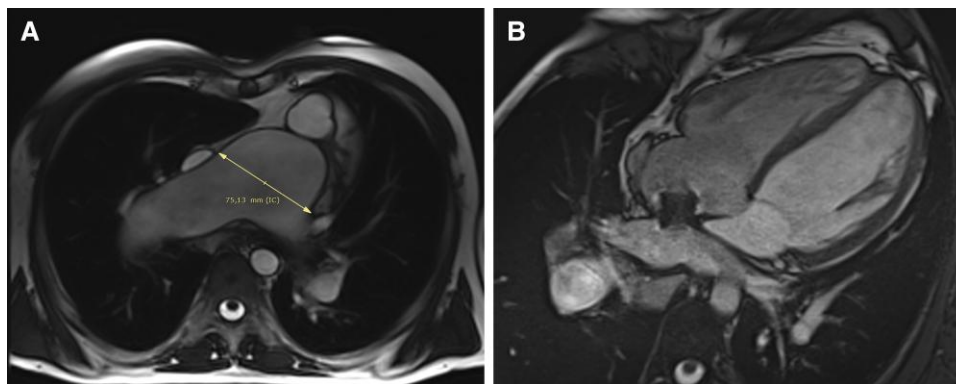
Outpatient medication included sildenafil 40 mg three times daily, macitentan 10 mg once daily, spironolactone 25 mg once daily, sacubitril-valsartan 24–26 mg twice daily, torsemide 5 mg once daily, amiodarone 200 mg once daily, and a vitamin K antagonist.

At admission, body temperature was 37.1°C, with a blood pressure of 109/78 mmHg, a heart rate of 93 b.p.m., oxygen saturation of 88% on room air, and a respiratory rate of 26 per min.

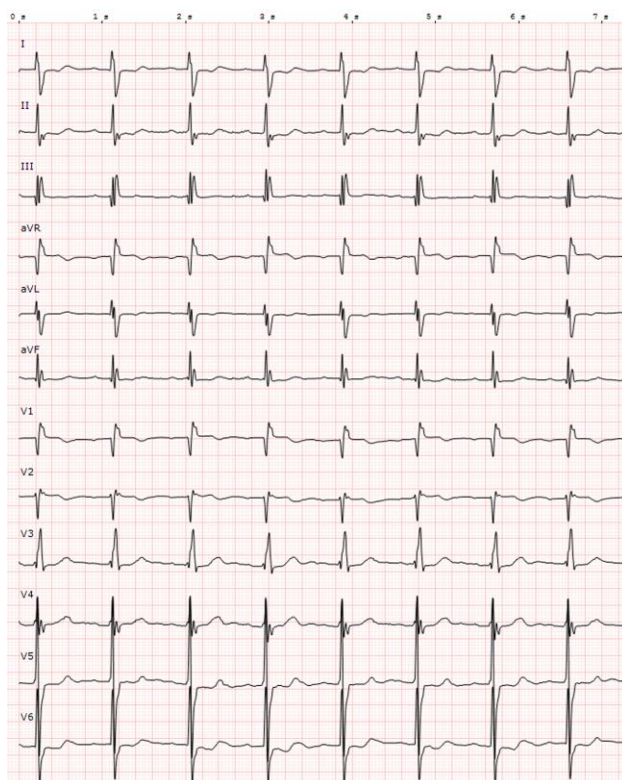
Physical examination showed mild cyanosis; heart sounds were rhythmic, with an accentuated second sound and a grade 2/4 pulmonary diastolic murmur; vesicular breath sounds were preserved bilaterally,



Follow-up CT scan at 6 months since acute dissection of a severely dilated right pulmonary artery (37 mm). The yellow arrow indicates false lumen thrombosis. Also, severe dilatation of the pulmonary trunk can be appreciated (75 mm).



**Figure 1** Previous magnetic resonance showing severe dilatation of the pulmonary trunk (75 mm) and pulmonary arteries (A) and severe biventricular dilatation (B).



**Figure 2** Electrocardiogram at admission showing right axial deviation and right bundle branch.

with wet crackles audible in the right lung. Furthermore, there was hepatomegaly of 4 cm below the costal margin, with no jugular vein distension or oedema.

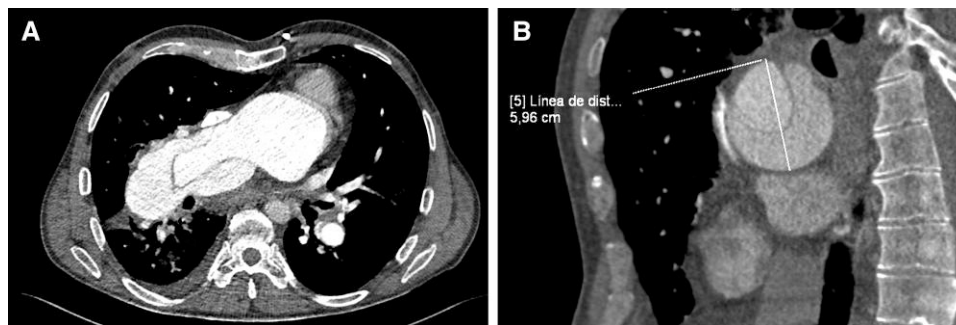
Electrocardiogram (ECG) demonstrated sinus rhythm at 67 b.p.m., PR 230 ms, QRS 140 ms with a right bundle branch block, and axis +90 degrees (Figure 2). No difference from previous ECG could be noted.

Arterial blood gas test showed pH 7.45 (normal range 7.38–7.42), carbon dioxide partial pressure 31 mmHg (normal range 38–42), oxygen partial pressure 59 mmHg (normal range 75–100), oxygen

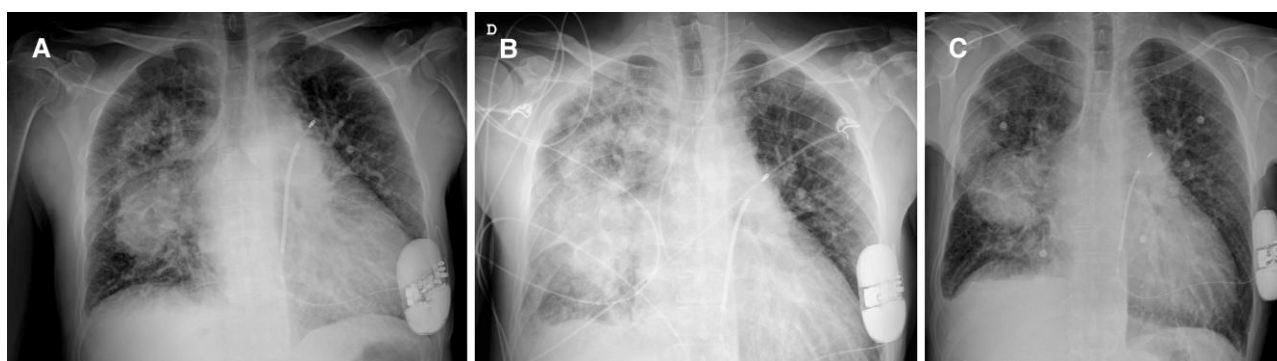
saturation 93% (normal range 94–100%), HCO<sub>3</sub> 21.5 mEq/L (normal range 22–28), and lactate 0.5 mmol/L (normal value < 2).

Additional investigations supported congestive heart failure and respiratory failure, with N-terminal prohormone of brain natriuretic peptide (NT-proBNP) measuring 3800 ng/L (normal range ≤125 ng/L and previous outpatient values of 600–700 ng/L), high-sensitivity troponin I 16 ng/L (normal range <45 ng/L), and D-dimer 673 ng/mL (normal range ≤280 ng/L).

Chest X-ray was significant for an enlarged cardiac silhouette with a dense pulmonary infiltration, especially on the right fields.



**Figure 3** Urgent computed tomography scan showing acute right pulmonary artery dissection: transversal (A) and sagittal (B) sections.



**Figure 4** Chest radiography at admission (A), after epoprostenol initiation (B), and after epoprostenol withdrawal and furosemide initiation (C).

Transthoracic echocardiogram (TTE) revealed moderate-to-severe dysfunction of the subpulmonary left ventricle, moderate dysfunction of the systemic ventricle, and mean pulmonary baffle gradient of 5 mmHg, with a dilated inferior vena cava.

Supplemental oxygen with high flow nasal cannula at oxygen fraction of 60% and 60 L/min was initiated. Differential diagnosis of acute respiratory failure included acute ventricular failure, pulmonary hypertensive crisis, or thromboembolic pulmonary disease.

An urgent CT scan showed severe dilatation of pulmonary arterial circulation (trunk measuring  $74 \times 63$  mm and right pulmonary branch measuring  $60 \times 56$  mm), right PAD with two points of intimal rupture of 0.5 and 1.5 mm, and presence of perivascular fat reticulation and liquid that was suggestive of imminent rupture (Figure 3).

Surgical risk was estimated to be extremely high. Moreover, he was not considered suitable for extracorporeal membrane oxygenation and circulatory support due to very high bleeding risk and uncertain destination therapy.

Outpatient pulmonary vasodilators were continued (sildenafil 40 mg every 8 h and macitentan 10 mg once daily), and intravenous epoprostenol was initiated and titrated up to 9 ng/kg/min in order to try to reduce pulmonary artery parietal stress.

After stabilization during the first 48 h, respiratory failure worsened significantly, chest X-ray was significant for acute pulmonary oedema (Figure 4), and NT-proBNP increased up to 7000 ng/L. Transthoracic echocardiogram showed an increase of mean baffle gradient up to 13 mmHg (previously 5 mmHg); this effect was attributed to pulmonary vasodilatation and subsequent increase of pulmonary blood flow

caused by epoprostenol. Epoprostenol was gradually discontinued, and intravenous continuous furosemide was initiated, obtaining negative fluid balance.

Respiratory failure responded well to negative fluid balance, and supplemental oxygen therapy could be withdrawn after 21 days. A follow-up CT scan before discharge showed morphologic stabilization of the PAD (Figure 5A).

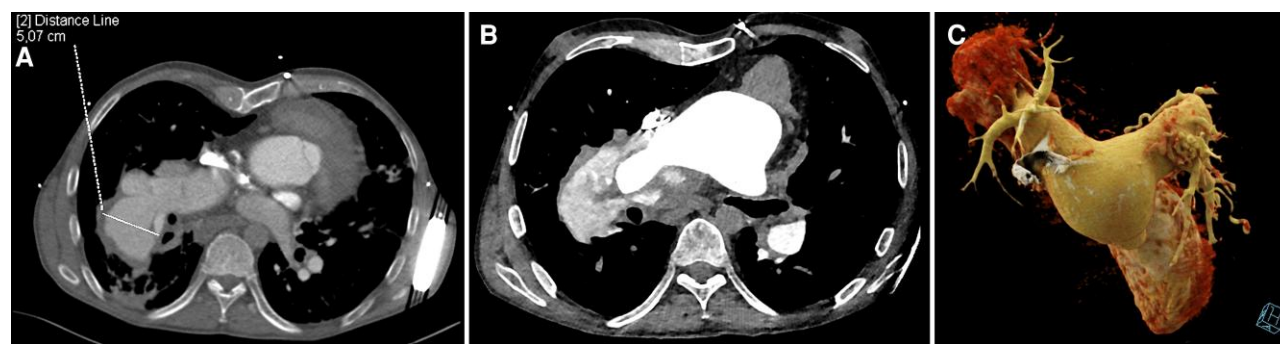
After stabilization, the patient was sent for evaluation in a heart–lung transplant centre, being eventually refused for extremely high perioperative bleeding risk due to abundant bilateral pulmonary neovascularization. He was discharged home in WHO-FC III.

Treatment was optimized by adding oral furosemide. Anticoagulation was not re-started for high bleeding risk. Outpatient CT scan (Figure 5B and 5C) showed morphological stabilization of the PAD, with a diameter of  $60 \times 59$  mm and with false lumen thrombosis. Follow-up TTE showed only mild biventricular dysfunction, without pulmonary baffle obstruction. Six months after the acute event, he was stable and recovered his previous WHO-FC II–III.

## Discussion

Pulmonary artery dissection is a very rare and potentially lethal complication of pulmonary hypertension, with few cases described ante mortem.<sup>9</sup> Pulmonary hypertension and previous pulmonary artery dilatation are reported as the main underlying conditions leading to PAD.





**Figure 5** Follow-up computed tomography scans: at 21 days, showing morphological stabilization (A), and at 6 months, showing false lumen thrombosis (B and C, with three-dimensional reconstruction).

Although pulmonary artery dilatation may be a prognostic factor in patients with pulmonary hypertension,<sup>10</sup> it has not shown prognostic significance in adults with congenital heart disease.<sup>11</sup> There are no formal recommendations on radiological monitoring of patients with congenital heart disease and pulmonary artery dilatation or severe pulmonary hypertension; based on our experience, we recommend baseline imaging, as well as periodic follow-up every 3–5 years or in case of symptoms that suggest progression, such as hoarseness, haemoptysis, or worsening dyspnoea.

Pulmonary artery dissection, as a complication of congenital heart disease, has been described in patients with a patent ductus arteriosus and with Eisenmenger syndrome, but not in patients with history of d-TGA and Mustard procedure.<sup>12</sup>

Even though surgical treatment of acute PAD is reported in literature, strong evidence regarding the best treatment is lacking.<sup>7</sup>

Epoprostenol is a strong pulmonary vasodilator,<sup>13</sup> and in this patient with d-TGA and atrial switch correction, with post-capillary pulmonary hypertension, epoprostenol administration was followed by an increase of venous pulmonary baffle gradients and severe pulmonary congestion, which was eventually treated with negative fluid balance.

In patients with a component of post-capillary pulmonary hypertension, pulmonary vasodilation with drugs such as epoprostenol can cause important pulmonary congestion, and it should be avoided in the presence of a high pulmonary wedge pressure.

## Lead author biography



Marco Tomasino is a medical doctor from Italy (University of Palermo). Since 2020, he has been training as a cardiology resident in Vall d'Hebron Hospital (Barcelona, Spain). He has a special interest in adult congenital heart disease and in acute cardiovascular care.

**Consent:** The authors confirm that written informed consent for submission and publication of this case report including images and

associated text has been obtained from the patient in line with the Committee on Publication Ethics (COPE) guidance.

**Conflict of interest:** None declared.

**Funding:** None declared.

## Data availability

The data underlying this article are available in the article and in its online supplementary material.

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