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An unusual cause of pulmonary hypertension in the elderly

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Summary

In patients with suspected pulmonary hypertension, echocardiography is the primary imaging tool to assess the probability of pulmonary hypertension and to identify the underlying haemodynamic mechanism. However, for a definite diagnosis of pulmonary hypertension and its classification, right heart catheterisation is mandatory. We present the case of a 72-year-old male patient with many typical features of postcapillary pulmonary hypertension in the context of left heart disease in whom a congenital left-to-right shunt was found.

Keywords: *pulmonary hypertension, heart failure, shunt, echocardiography, right heart catheterisation*

Introduction

In patients with exertional dyspnoea, pulmonary hypertension (PH) is a differential diagnosis to consider. Echocardiography is the primary imaging tool to assess the probability of PH and to evaluate the underlying mechanism [1]. The most common form by far is PH in the context of diseases of the left heart (group 2 PH) [2]. In these patients, chronic left atrial pressure elevation due to left ventricular systolic and/or diastolic dysfunction, left atrial dysfunction, valve disease, pericardial disease and other mechanisms leads to an increase in the pulmonary artery wedge pressure and thereby pulmonary artery pressure [2]. Patients with echocardiographic evidence of PH and a high likelihood of group 2 PH most often do not need right heart catheterisation [2]. A trial of heart failure therapy and non-invasive follow-up is often appropriate as the primary approach in this context. However, in ambiguous situations a clear diagnosis should be established by means of cardiac catheterisation and additional imaging as a basis for an appropriate therapy. We herein present an elderly patient with many features of group 2 PH but some uncommon aspects and in whom a decision in favour of an invasive work-up was made, which led to the diagnosis of a congenital left-to-right shunt.

Case report

A 72-year-old male with shortness of breath on mild exertion and intermittent left-sided chest pain at rest without radiation was referred for further evaluation. His medical history was remarkable for diabetes, hypertension and recurrent ischaemic strokes due to paroxysmal atrial fibrillation. His regular medication included a non-vitamin K oral anticoagulant, a beta-blocker, a loop-diuretic, an angiotensin converting enzyme inhibitor and a statin. Clinical examination was unremarkable.

Transthoracic echocardiography showed a normal-sized left ventricle with concentric hypertrophy and an ejection fraction of 65%, evidence of diastolic dysfunction (ratio of peak early transmitral velocity [E] to peak early diastolic mitral annular velocity [e′] averaged from the septal and lateral annulus [E/e′]: 12), mild mitral regurgitation and a dilated left atrium. The right ventricle and the right atrium were dilated (fig. 1, panels A and B), right ventricular function was mildly impaired (tricuspid annular plane systolic excursion 16 mm, peak systolic tricuspid annular velocity by tissue Doppler 8 cm/s) and there was moderate tricuspid regurgitation. The peak tricuspid regurgitant velocity was 3.7 m/s (fig. 1, panel C) and the central venous pressure was estimated as 10–15 mm Hg (fig. 1, panel D). Pulmonary function tests showed normal static lung volumes and a normal diffusion capacity.

Given the dilatation of the right-sided cavities with a larger right than left atrium, the relatively high peak tricuspid regurgitant velocity and evidence of an elevated central venous pressure left and right heart catheterisation was performed revealing normal coronary arteries but confirming the presence of PH with a mean pulmonary artery pressure (mPAP) of 29 mm Hg. The mean pulmonary artery wedge pressure (mPAWP) was 16 mm Hg, and the left ventricular end-diastolic pressure was also 16 mm Hg. The full haemodynamics are shown in table 1. Surprisingly, the oxygen saturation in the pulmonary artery was 80% and therefore an oxygen run was performed, which showed a step-up in oxygen saturation between the superior vena cava and the right atrium (fig. 2). A left-to-right shunt with a ratio of pulmonary to systemic flow (Qp/Qs) of approximately 1.7:1 was calculated. The absolute values for pulmonary and systemic blood flow calculated on the basis of the indirect Fick method were 7.5 and 4.4 l/min (table 1).

Based on these findings additional diagnostic tests were performed. Transoesophageal echocardiography ruled out an atrial septal defect. Computed tomography revealed an anomalous pulmonary venous connection of the right up-

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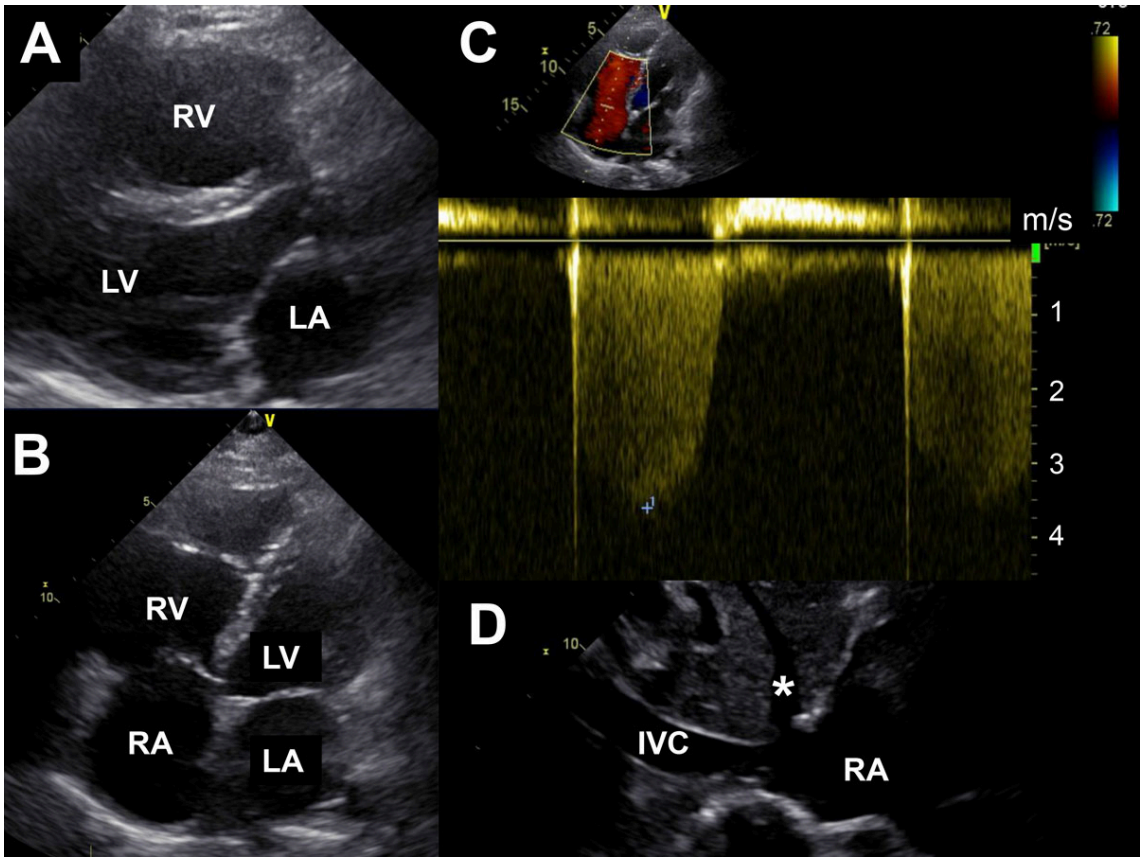
Figure 1: Echocardiography. Panels A and B: parasternal long-axis view (A) and apical four-chamber view (B) showing dilatation of the right ventricle (RV) and right atrium (RA). Panel C: Continuous wave Doppler revealing an elevated peak tricuspid regurgitation velocity. Panel D: Dilated inferior vena cava (IVC) and prominent liver veins (asterisk) suggesting elevated central venous pressure. LV: left ventricle; LA: left atrium.



per pulmonary vein to the superior vena cava (fig. 3). Medical management including symptomatic diuretic therapy (a loop diuretic and spironolactone) was continued. In the following 2 years the patient had two episodes of right heart failure precipitated by a non-cardiac operation and use of non-steroidal anti-inflammatory drugs, respectively, which could be managed by fluid restriction and transient intensification of diuretic therapy.

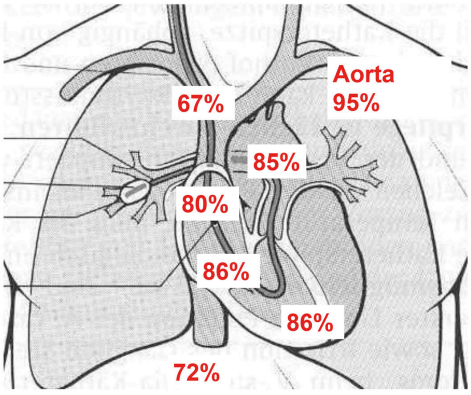
Table 1: Invasive haemodynamics.

Right atrial pressure (mean/v wave, mm Hg)		9/13
Right ventricular pressure (systolic–end-diastolic, mm Hg)		59–9
Pulmonary artery pressure (systolic/diastolic–mean, mm Hg)		59/15–29
Pulmonary artery wedge pressure (mean/v wave, mm Hg)		16/29
Aorta (systolic/diastolic–mean, mm Hg)		166/71–107
Left ventricular end-diastolic pressure (mm Hg)		16
Pulmonary blood flow (l/min)		7.5
Indexed pulmonary blood flow (l/min/m^2)		4.0
Pulmonary vascular resistance (Wood units)		1.7
Systemic blood flow (l/min)		4.4
Indexed systemic blood flow (l/min/m^2)		2.3
Systemic vascular resistance (Wood units)		22.3
Oxygen saturations	Aorta (%)	95
	Pulmonary artery	80
	Right ventricle	80
	Right atrium	80
	Superior vena cava	70
	Inferior vena cava	68
Ratio of pulmonary to systemic blood flow (Qp/Qs)		1.7:1.0

Discussion

Pulmonary hypertension is currently defined as an mPAP ≥25mm Hg at rest assessed by means of right heart catheterisation [1]. Importantly, despite being the key imaging tool for the evaluation of a patient with possible PH, echocardiography cannot establish a definite diagnosis of PH, but can only estimate the likelihood of PH based on the peak tricuspid regurgitation velocity and indirect signs of significant PH such as right ventricular dilatation, ab-

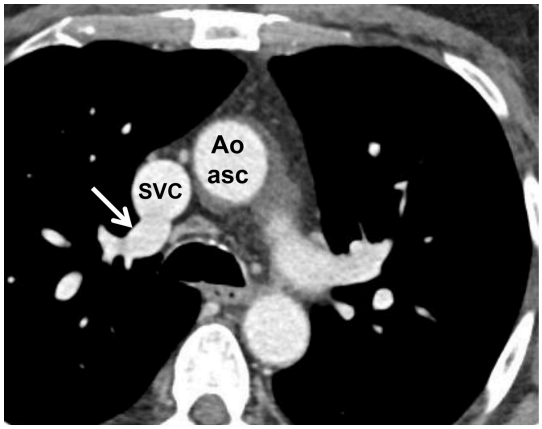
Figure 2: Results of the oxygen run. For details see text.



normal motion or shape of the interventricular septum, and dilatation of the right atrium and inferior vena cava [1].

In our patient there were (i) a high probability of PH and (ii) many features suggesting group 2 PH in the context of heart failure with preserved ejection fraction, such as older age, hypertension and atrial fibrillation. However, the extent of the dilatation of the right-sided cavities in relation to the extent of left ventricular disease and left atrial dilatation was disproportional, and although estimation of the systolic pulmonary artery pressure with the Bernoulli equation as an exact number is discouraged [1], the pulmonary artery pressure appeared to be higher than expected in this context. Therefore, a multifactorial cause of PH was possible and right heart catheterisation was performed to clarify the situation. The primary intention was to differentiate between isolated postcapillary PH (defined as mPAWP >15 mm Hg but pulmonary vascular resistance [PVR] ≤3 Wood units; group 2 PH), combined pre- and postcapillary PH (mPAWP >15 mm Hg but PVR >3 Wood units; also group 2 PH), and precapillary PH (mPAWP ≤15 mm Hg; including pulmonary arterial hypertension [PAH, group 1], PH in the context of chronic hypoxaemia / lung diseases [group 3], chronic thromboembolic PH [group 4], and rare forms of PH [group 5]) [1, 2]. The mPAWP of 16 mm Hg was mildly elevated and fulfilled the guideline criterion [1] for postcapillary PH, which was in line with the clinical impression. However, the oxygen saturation in the pulmonary artery of 80% did not fit this picture: normally, the oxygen saturation in the pulmonary artery (the mixed-venous oxygen saturation in patients without a shunt) does not exceed 70–75% and is inversely related to the cardiac output. In patients with heart failure (i.e., left heart failure), the oxygen saturation in the pulmonary artery is usually in the range of 50–65% and sometimes even lower, as a reflection of a reduced cardiac output. The situation in our patient clearly suggested a left-to-right shunt, which was finally found by additional noninvasive imaging. The transpulmonary gradient (mPAP−mPAWP) was relatively high (13 mm Hg), which did not, however, reflect the presence of a pulmonary vascular component of PH but rather a high pulmonary blood flow. The PVR (transpulmonary gradient divided by pulmonary blood flow) was absolutely normal. Notably, the PVR was so low that this patient with an only mildly elevated mPAWP would not even have met the definition of PH in absence of a shunt (mPAWP of 16 mm Hg plus a transpulmonary gradient of approximately 8 mm Hg [assuming the same PVR but a normal pulmonary flow] ≈24 mm Hg). One must be aware of the limitations of cardiac output assessment by use of the indirect Fick method (estimation rather than measurement of oxygen consumption). Thus, the data must be looked at in a qualitative rather than an absolutely quantitative manner, although they are plausible as they stand (table 1).

Figure 3: Computed tomography showing drainage of the right upper pulmonary vein into the superior vena cava (SVC). Ao asc: ascending aorta.



In a younger patient with a high probability of congenital heart disease, a cardiac magnetic imaging study would have been an appropriate radiation-free option before cardiac catheterisation. In a patient with an echocardiogram suggesting a predominantly precapillary form of PH (high probability of PH but little evidence of a left heart pathology) or in presence of invasively proven precapillary PH, a V/Q scan would have been performed for the evaluation of chronic thromboembolic pulmonary hypertension.

Partial abnormal pulmonary venous return is estimated to affect 0.1–0.2% of the adult population. A diagnosis in patients over 50 years is uncommon, but several reports are available in the literature (e.g., Sears et al. [3]). In symptomatic, typically young patients with relatively large shunts (Qp/Qs ≥1.5:1) and absence of significant pulmonary vascular disease (PVR less than one third of systemic vascular resistance) surgical correction is recommended because of the risk of progressive right ventricular dilatation and dysfunction and, rarely, pulmonary vascular remodelling with an increase in PVR [4]. Partial abnormal pulmonary venous return is often associated with an atrial septal defect (not in the present case). In this situation, a high PVR can result in shunt reversal and thereby central cyanosis (Eisenmenger syndrome) [5].

Indications for surgery in older adult patients such as this one are not well defined, since it is difficult to differentiate of exact contribution of the shunt to the patient's symptoms. The patient had an elevated mPAWP at rest, and a further rise during exercise in the context of left ventricular diastolic and left atrial dysfunction and episodes of atrial fibrillation was likely. This concept could have been explored further by use of a fluid challenge or exercise. However, given the detection of a shunt explaining the echocardiographic findings this was not done. Given the patient's age, the fact that the shunt obviously had not been large enough to stimulate an increase in PVR over decades and the fact that both episodes with decompensation were explained by "accidents", conservative management was chosen. Still, a repeat right heart catheterisation to look for an increase in PVR would have been instructive after these episodes of decompensation. This was discussed with the patient but refused.

In summary, this case highlights that in a patient with suspected PH, echocardiography is the key noninvasive tool to estimate the probability of PH, and to get an idea of the haemodynamic constellation and the underlying mechanisms, but that only right heart catheterisation can definitely establish the diagnosis of PH and discriminate between postcapillary and precapillary PH, which can then guide the selection of additional tests to classify PH (groups 1 to 5). In patients with ambiguous noninvasive findings and equivocal constellations, right heart catheterisation should be performed to clarify the situation. Sometime, this leads

to unexpected findings, such as in the present patient. This may prevent from the use of nonindicated, expensive and potentially dangerous therapies (e.g., specific PAH therapy in patients with left heart disease).

Disclosure statement
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