



# Coronary-pulmonary artery fistula with lung hypoplasia and a bicuspid aortic valve: A case report

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## Abstract

**Background:** With an incidence of less than 1%, a Coronary Artery to Pulmonary Artery fistula (CAPF) is a rare coronary anomaly that causes heart failure. It causes a left to right cardiac shunt. While guidelines favor surgical correction in symptomatic patients, we present a challenging case with multiple cardio-thoracic pathologies.

**Case Presentation:** We present a 38-year-old obese male with persistent atrial fibrillation (AF). He presented to our hospital in decompensated heart failure and AF with rapid ventricular response. He was found to have a CAPF, a bicuspid aortic valve and left lung hypoplasia in the presence of severely reduced left ventricular systolic dysfunction. The patient subsequently underwent various cardiac testing demonstrating advanced anatomical and physiologic involvement of his CAPF, including suggested coronary steal. Despite some indications for percutaneous or surgical referral, we optimized his AF and congestive heart failure in lieu of formulating a treatment strategy for his CAPF and other abnormalities.

**Conclusion:** This report illustrates a case of a young adult who presented in decompensated heart failure with newly diagnosed left ventricular systolic function and rapid AF, who had a triad of congenital defects including a CAPF, a bicuspid aortic valve and left lung hypoplasia. To the best of our knowledge, this triad of defects is unreported. This case highlights the clinical approach in the evaluation of a cardiac shunt and its management strategies in the presence of multiple cardio-thoracic comorbidities.

## Keywords

Other heart failure < Heart failure < Cardiology, Valvular heart disease < Hypertension < Cardiology, CT and MRI < Diagnostic Testing < Cardiology, Coronary imaging: angiography/ultrasound/Doppler/CC < Diagnostic Testing < Cardiology

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## Introduction

We present a case of a young adult male who presented in decompensated heart failure and atrial fibrillation (AF), who was incidentally found to have a *Coronary artery to Pulmonary artery fistula (CAPF), a bicuspid aortic valve, and left lung hypoplasia*. To the best of our knowledge this is the first report of this triad of congenital cardio-thoracic defects.

## Case presentation

A 38-year-old male presented to our emergency department with progressively worsening dyspnea for several weeks. He had no chest pain or fevers. There was no significant social or travel history. He presented with a heart

rate of 144 beats per minute, a blood pressure of 122/82 mmHg, a respiratory rate of 30 breaths per minute and an oxygen saturation of 86% on room air. Physical findings showed jugular venous distention, an apically displaced maximal impulse, no right ventricular

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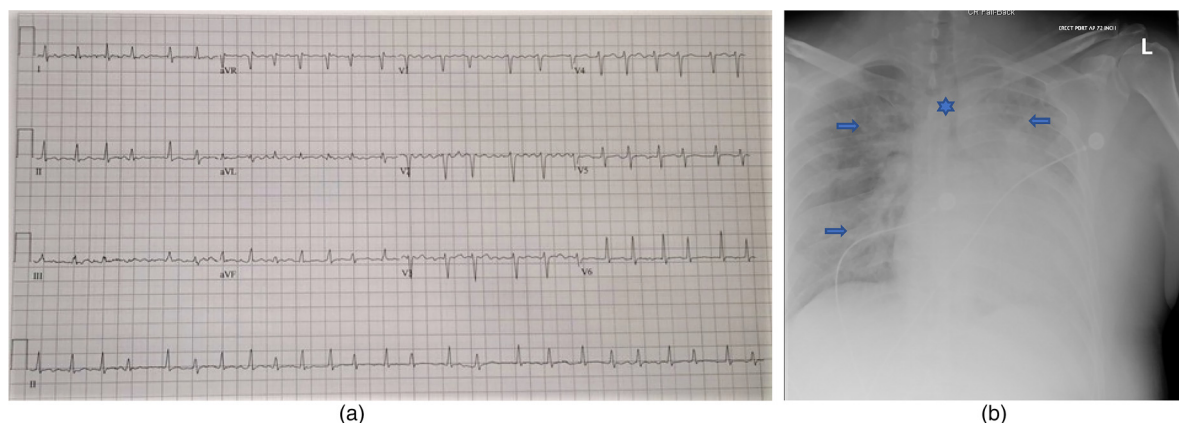
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**Fig. 1.** (a) ECG showing AF with rapid ventricular response, normal axis with low voltage. (b) Chest x-ray, shows marked central vascular congestion, bilateral opacities (arrows) with left lung volume loss, left mediastinal and tracheal shift (star) with partial volume loss of the left lung.

**Table 1.** Right heart catheterization.

	Value	Normal Range*
Right Atrium	Mean 11 mm Hg	1–5 mm Hg
Right Ventricle	40/15 mm Hg	15–30/ 1–7 mm Hg
Pulmonary Artery	Mean 26 mm Hg	9–19 mm Hg
Pulmonary Capillary Wedge	Mean 13 mm Hg,	4–12 mm Hg
Cardiac Output	4.8 L/min	4.8–7.3 L/min
Cardiac Index	1.97 L/min/m <sup>2</sup>	2.8–4.2 L/min/m <sup>2</sup> )
Systemic Vascular Resistance	1194.70 dyn. Sec. cm <sup>-5</sup>	700–1600 dyn. Sec. cm <sup>-5</sup>
Pulmonary Vascular Resistance	218.75 dyn. Sec. cm <sup>-5</sup>	20–130 dyn. Sec. cm <sup>-5</sup>

\*Kelly, C. R et al. (2013). Pulmonary-artery catheterization. *The New England journal of medicine*, 369(25), e35.<https://doi.org/10.1056/NEJMvcm1212416>.

heave, an irregularly irregular fast rhythm, bibasilar lung rales, and pedal edema. There were no significant murmurs or clicks appreciated. Initial 12-lead electrocardiogram (ECG) demonstrated AF without any ischemia or Q waves, a normal axis with low voltage (Figure 1(a)).

### Past medical history

His history is notable for hypoplasia of the left lung (diagnosed incidentally during childhood), hypertension, hyperlipidemia, obesity (Body Mass Index 38.6 Kg/m<sup>2</sup>), AF with two prior hospitalizations at an outside facility, as well as failed electrical cardioversion. He also had a reported myocardial infarction; however, this was thought to be due to demand ischemia from AF. Hence, an ischemic evaluation was not performed. He had a prior reported echocardiogram with preserved left ventricular systolic function without any reported valvular abnormalities.

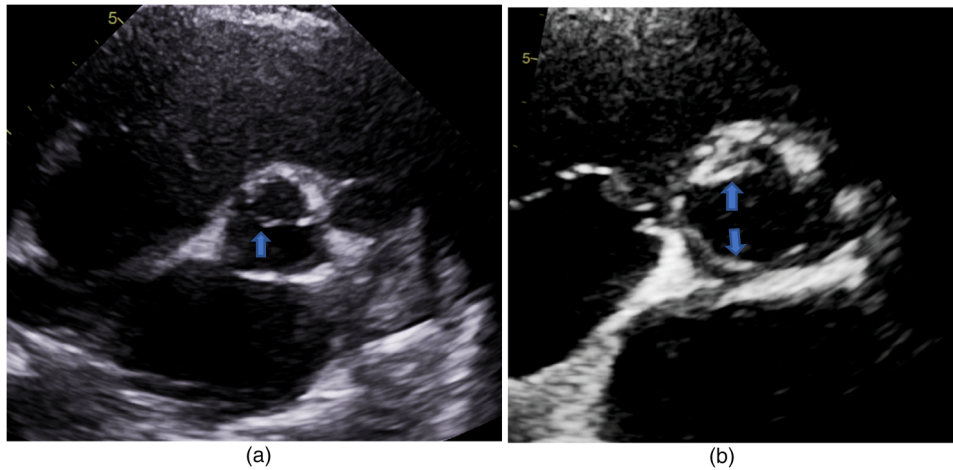
### Investigations

Laboratory investigations were significant for a normal cardiac peak Troponin I (less than 0.031 ng/mL) and

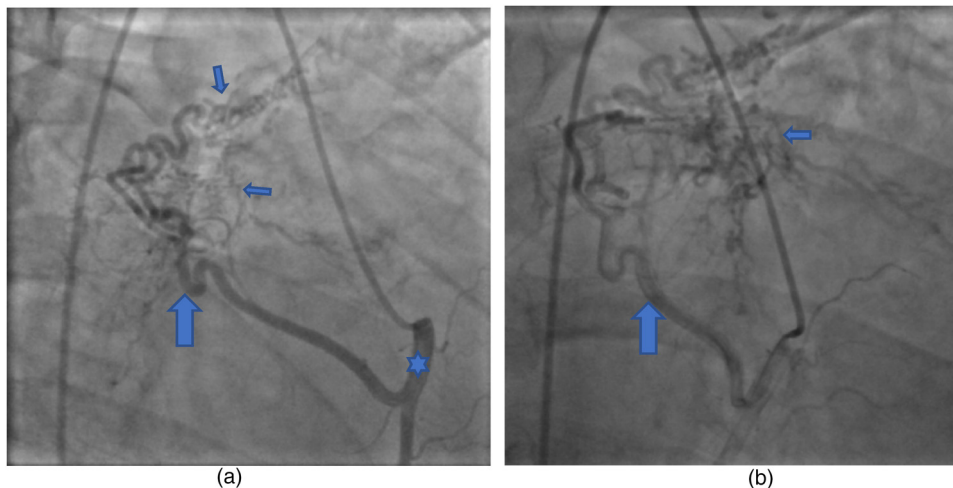
thyroid-stimulating hormone, and a brain natriuretic peptide of 819.6 pg/ml (normal less than 101.0 pg/mL). His chest x-ray is shown below (Figure 1(b)). An echocardiogram showed a moderately dilated left ventricle, with an ejection fraction between 20–25%, mildly reduced right ventricular systolic function, suspicion for a bicuspid aortic valve with fusion of the left and non-coronary leaflets (Figure 2) with no significant mitral regurgitation. Cardiac catheterization revealed patent coronary arteries and a torturous fistula from the proximal right coronary artery to the pulmonary artery (Figure 3(a, b)). Swan-Ganz catheterization is resulted in Table 1. Cardiac magnetic resonance (CMR) was performed electively, 1 month later (Figure 4–6).

### Management

In-hospital management included guideline-directed therapy for AF and decompensated heart failure.<sup>1</sup> He was scheduled to undergo transesophageal echocardiographic guided electrical cardioversion, however, this was deferred due to the presence of a left atrial appendage thrombus (image not shown). After several weeks of uninterrupted anticoagulation, he underwent successful atrial fibrillation



**Fig. 2.** Below, transthoracic echo, (a) parasternal short axis view in diastole suggesting a bicuspid aortic valve (arrow, during diastole), (b) during systole, small arrows point to valve leaflets.



**Fig. 3.** (a, b) Left heart catheterization, femoral approach, right anterior oblique view of the right coronary artery (star) showing proximal bifurcation to CAPF (arrows) with multiple small vessel arterial anastomosis (small arrows).

ablation and pulmonary vein isolation with restoration to sinus rhythm. He has since returned to work with improved symptoms.

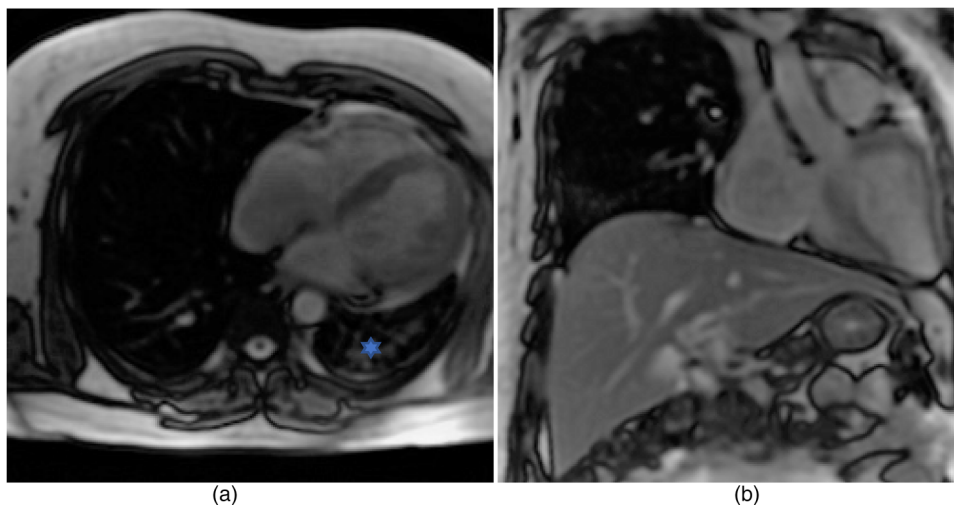
## Discussion

CAPF is a coronary artery anomaly in which an abnormal connection exists between a coronary artery and the pulmonary artery bypassing the capillary bed.<sup>2</sup> It has a reported prevalence between 0.17% to 0.68%, however the true prevalence of this abnormality is unknown as most cases remain asymptomatic due to the volume traversing the fistula being hemodynamically insignificant.<sup>2</sup> Symptomatic patients may have left and/or right sided heart failure with the development of pulmonary hypertension and/or Eisenmenger syndrome. In addition, they

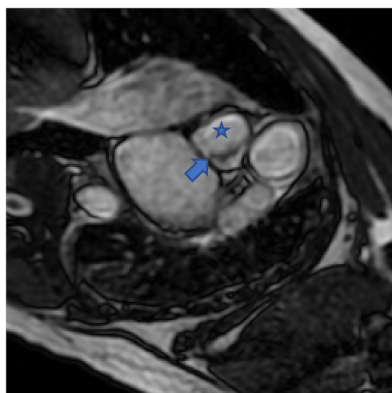
may also present with myocardial ischemia or infarction due to coronary steal phenomenon.<sup>2</sup>

CAPF may occur from embryological or acquired defects such as prior trauma or cardiac surgery.<sup>3</sup> The Hackensellner involution-persistence hypothesis states that 6 anlagen in the truncus arteriosus corresponding to the formation of the cusp of the *pulmonary* and *aortic* valves involute during embryological development, and the persistence of 2 of these anlagen result in the fistula formation.<sup>4,5</sup> Interventions are reserved for symptomatic patients or for those with a hemodynamically significant shunt and include surgical epicardial or endocardial ligation or percutaneous intravascular coil embolization.<sup>6,7</sup>

Our patient was found to have a *CAPF* with concomitant *bicuspid valve* and *left lung hypoplasia*. We feel this deserves further comment. While the presence of a bicuspid aortic valve can be explained due it's shared



**Fig. 4.** (a) CMR, General Electric, 1.5 Tesla, Gradient Echo, FIESTA, bright blood imaging, axial view, showing left lung hypoplasia (star) with Levocardia. (b) Coronal view, levocardia with left mediastinal shift.



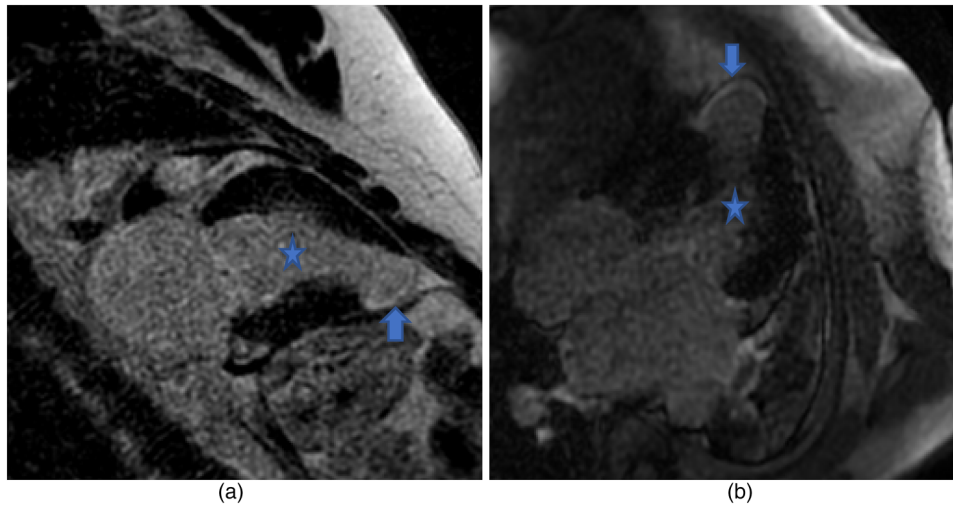
**Fig. 5.** CMR, General Electric, 1.5 Tesla, Gradient ECHO, FIESTA, showing short axis, basal view of heart showing a bicuspid aortic valve in diastole (arrow), star denotes the ascending aorta above the level of the sinus of valsalva.

embryological development with CAPF, we are unclear of the significance of this combination of findings, particularly with lung hypoplasia, and if either defect contributed to hypoxia and or heart failure. We should also note the significance of *left* lung hypoplasia. Various syndromes such as Scimitar syndrome has *right* lung hypoplasia with cardiac defects including anomalous pulmonary venous return. However, variants of these conditions with the association of *left* lung hypoplasia are even rarer.<sup>8</sup> Given his right ventricular dysfunction on echocardiogram, we initially considered group 3 pulmonary hypertension as a differential; either from his shunt, undiagnosed obesity-related sleep apnea or chronic lung disease (related to lung hypoplasia).

We performed a Swan-Ganz catheterization, which showed mild pulmonary hypertension and elevated pulmonary vascular resistance. This precluded overt frank

shunt reversal or Eisenmenger's syndrome (shunt reversal occurs when pulmonary vascular resistance significantly elevates causing forward flow to reverse) and hence did not support his CAPF being the main contributor to his hypoxia. Furthermore, his mild pulmonary hypertension conferred a good prognostic determinant for future closure, when warranted.<sup>9</sup> Given that his swan was performed after diuresis, in compensated heart failure (with normal pulmonary capillary wedge pressure), we emphasized the importance of his pulmonary vascular resistance, as his pulmonary pressures may have been lower after diuresis. Thus, this being elevated may confer early hemodynamic involvement of his CAPF. However, we felt it prudent to assimilate this data with a shunt evaluation as it would help quantify the hemodynamic significance of CAPF and differentiate other causes of elevated right sided pressures.

Overall, the reduction in his left ventricular cardiac output we felt to be from primary left heart failure and not right sided failure from his shunt. His coronary angiogram showed no evidence of coronary obstruction or embolic phenomena (arteries were widely patent) without angiographic evidence of vasospasm. Our workup was further complimented with a CMR, which confirmed a bicuspid valve and lung hypoplasia. Moreover, it showed an inferior wall ischemic scar in the same vascular territory as his CAPF (Figure 5 and 6(a)). Due to the lack of evident coronary artery obstruction, we suspected this to be from coronary steal syndrome due to competing flow of the right coronary artery and his CAPF, particularly in the prior setting of tachycardia. Notably, his infarct size (or scar burden) on CMR was small, focal and out of proportion to the degree of his LV dysfunction and hence, supported our assessment of a non-ischemic cardiomyopathy that was tachycardia mediated. Of note, this lack of significant



**Fig. 6.** CMR, General Electric, 1.5 Tesla, Fast gradient ECHO, (a) 2-chamber view of the left ventricle with apical inferior and infero-septal wall thinning and near transmural infarct with LGE (arrow); (b) 4-chamber view (stars denote the left ventricle, arrow points to LGE).

scar burden also confers a good prognosis for the recovery of LV function once medically optimized.<sup>7</sup> In addition, he also had no pathological Q waves on his ECG. We thus prioritized management of his heart failure and pursued rhythm control for AF over immediate CAPF closure.<sup>10</sup>

Our case had several limitations. His Swan-Ganz catheterization was performed before his coronary angiogram (which later detected his CAPF incidentally) and hence a shunt evaluation was not performed. We also felt that his newly diagnosed LV systolic function and tachycardia may have altered hemodynamics to accurately perform a shunt evaluation. His CMR was also limited due to gating artifact (from AF) and prolonged scan time. For similar such reasons, an electrocardiographically gated cardiac coronary tomographic angiography (Coronary CT) was deferred to prioritize medical optimization, tighter rate control and additionally was not offered at our institution.<sup>6</sup> We also felt that while Coronary CT was the preferred imaging modality in CAPF evaluation, it would not have immediately changed our management but more so complimented optimal surgical planning for the correction of his CAPF, when indicated.<sup>6</sup> There were added limitations to the availability of this test at local imaging centers due to the Covid-19 pandemic. Our Patient had a prior echocardiogram at an outside institution that did not show a bicuspid valve and we suspect this was possible due to limited image quality with obesity and tachycardia.

Now that our patient is clinically compensated, we plan to assess his CAPF with a dedicated shunt evaluation and to evaluate for anomalous pulmonary venous return, given its possible associations with lung hypoplasia.<sup>8</sup> In addition, we will repeat assessment of his left

ventricular systolic function to guide further heart failure management and may consider stress testing for ischemic burden, which if present, may suggest persistent coronary steal further warranting expedited CAPF closure. We will also refer for a sleep study, pulmonary function testing and genetic screening. Finally, due to the presence of a bicuspid aortic valve without evidence of aortic stenosis or an aortopathy, we felt it to be reasonable for close surveillance of his valve and CAPF, which may necessitate concomitant surgical correction, when warranted.

## Conclusion

We present a patient with heart failure and rapid AF who was found to have a *CAPF*, a *bicuspid aortic valve* and a *hypoplastic lung*. To the best of our knowledge, this triad of cardio-thoracic defects has not been reported. Although shunt closure is indicated in symptomatic patients, a comprehensive work-up including both anatomical and physiological aspects should be considered to determine optimal treatment strategies.

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## Abbreviations

AF	Atrial Fibrillation
CAPF	Coronary Artery to Pulmonary Artery Fistula
ECG	Electrocardiogram
CMR	Cardiac Magnetic Resonance
Coronary-CT	electrocardiographically gated cardiac coronary tomographic angiography
LGE	Late Gadolinium Enhancement