

# Cardiogenic shock in the context of newly diagnosed anomalous origin of the right coronary artery originating from the pulmonary artery: a case report

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## Background

Anomalous right coronary artery (RCA) from the pulmonary artery (ARCAPA) is a rare congenital heart abnormality with varying clinical presentations, for which multiple imaging modalities are often required for diagnosis.

## Case summary

We present a case of a 76-year-old female presenting with 2 weeks of palpitations and shortness of breath who was found to be in rapid atrial fibrillation (AF) with congestive heart failure. Despite initial medical management, the patient developed cardiogenic shock with anuric renal failure. Emergent right and left heart catheterization did not demonstrate any significant obstructive coronary artery disease but showed severe right ventricular (RV) failure and raised the possibility of an ARCAPA. This diagnosis was further corroborated by findings on a subsequent transoesophageal echocardiogram. In view of profound decline and limited anticipated improvement, the patient ultimately decided to pursue comfort measures in a hospice setting.

## Discussion

We postulate that the underlying aetiology of our patient's shock state was multifactorial, notably progressive RCA-territory ischaemia and RV failure, sepsis, and new-onset uncontrolled AF. In adults, unrecognized congenital heart disease can uncommonly cause cardiogenic shock. In our case, echocardiography and invasive angiography were integrated for the diagnosis of ARCAPA given the clinical circumstances that limited the use of cardiac computed tomography angiography.

## Keywords

Congenital heart defect • Atrial fibrillation • Coronary vessel anomaly • Coronary circulation • Hemodynamics • Coronary angiography • Case report

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## Learning points

- Anomalous right coronary artery from the pulmonary artery is a rare condition with wide variability in presenting age and can be easily missed by echocardiography.
- Undiagnosed congenital heart disease is an important consideration in adults presenting with unexplained cardiogenic shock.
- Integration of multiple modalities of cardiac imaging (echocardiography, invasive angiography, magnetic resonance imaging, computed tomography) should be considered when initial investigations fail to yield a conclusive diagnosis.

## Introduction

Anomalous right coronary artery from the pulmonary artery (ARCAPA) is a rare and usually benign condition<sup>1</sup> which can be easily missed by echocardiography. It has an estimated incidence of 0.002% although this is likely underestimated given the variability in presenting age and the high proportion of asymptomatic patients.<sup>1</sup>

In contrast, anomalous origin of the left coronary artery from the pulmonary artery is more common with an incidence of 0.008% and is usually discovered early in life given its symptomatic nature,<sup>2</sup> with signs of ischaemia and heart failure as pulmonary vascular resistance normalizes during infancy, creating a left-to-right shunt and coronary steal physiology.<sup>3</sup> Prior to 1965, ARCAPA was uniformly diagnosed during surgery or autopsy.<sup>4</sup> Since then, just over 100 cases have been documented worldwide, with varying clinical presentations, epidemiology, and imaging modalities required to make the diagnosis.<sup>5</sup>

## Timeline

Day 0	Admitted to hospital for decompensated heart failure.
Day 1	Intubated for emergent cardiac catheterization. No flow-limiting stenoses were identified. The right coronary artery (RCA) appeared to be emptying into the pulmonary artery/right ventricular (RV) outflow tract. Transthoracic echocardiogram showed severely reduced RV systolic function, severe right atrial dilatation, and severe tricuspid regurgitation.
Day 2	Positive cultures for <i>Streptococcus bovis</i> .
Day 7	Intermittent haemodialysis initiated.
Day 9	Transoesophageal echocardiogram revealed a dilated and tortuous left main coronary artery and branches, as well as a dilated and tortuous vessel (likely the RCA) draining into the pulmonary artery. No vegetations were found. This corroborated a diagnosis of anomalous right coronary artery from the pulmonary artery. The patient was reviewed at interdisciplinary heart team rounds and turned down for surgical intervention.
Day 40	Transferred to hospice care.
Day 75	Deceased.

## Case presentation

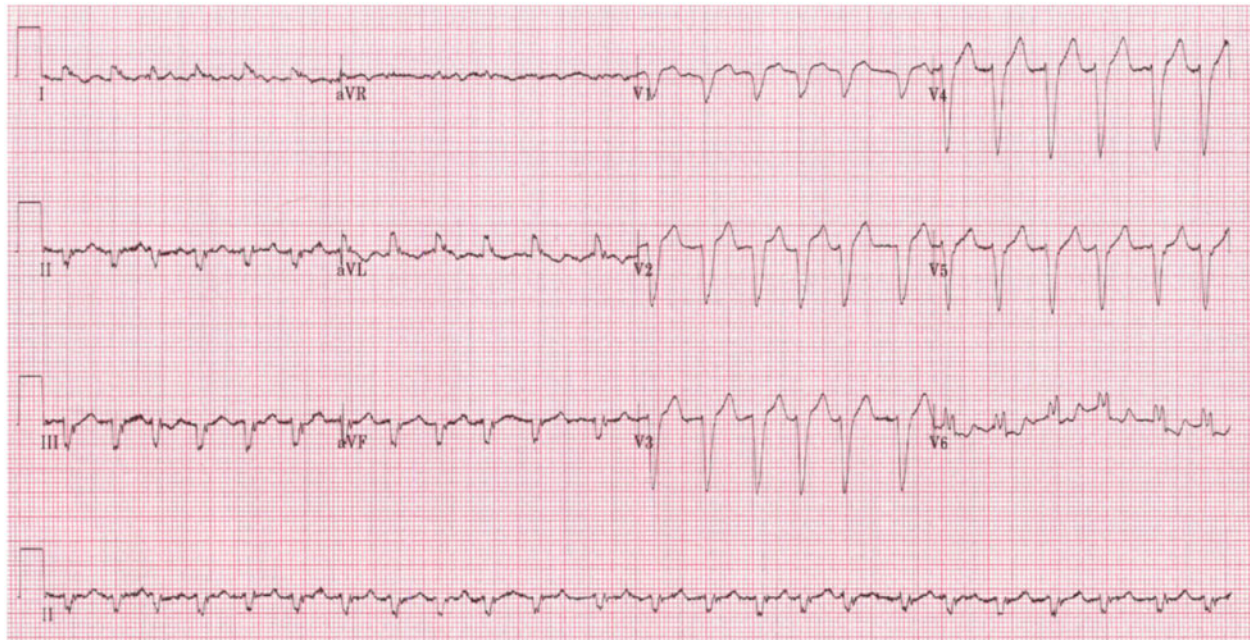
A 76-year-old Caucasian female with known hypertension and dyslipidaemia presented with 2 weeks of intermittent palpitations, orthopnoea, and New York Heart Association functional class III–IV shortness of breath. On exam, the patient was not in acute distress; however, extremities were cool and clammy. Heart auscultation revealed normal heart sounds, no murmurs, rubs, or gallops but the rhythm was irregularly irregular at ~100 b.p.m. The jugular venous pressure was elevated at the angle of the jaw and there was a positive abdominojugular reflux sign. Auscultation of the lungs revealed bilateral crackles up to mid chest with decreased air entry at the bases. The abdomen was soft, non-tender, and non-distended.

In the emergency department, she was in uncontrolled atrial fibrillation (AF) with a left bundle branch block (*Figure 1*) with clinical evidence of decompensated heart failure and her chest X-ray confirming evidence of pulmonary oedema with bilateral pleural effusions. There was no prior electrocardiogram for comparison. She was admitted for intravenous diuresis [initially with Furosemide 80 mg intravenously (IV) once then Furosemide infusion 20 mg/h] and rate control (initially with metoprolol 5 mg IV once then transitioned to oral metoprolol with an intravenous digoxin load). Over the next few hours, she became increasingly diaphoretic, tachycardic, and hypotensive at 60/30 mmHg. High-sensitivity troponin increased to 2040 ng/L (reference value  $\leq 30$  ng/L) from 13 ng/L, and serum lactate was 7.1 mmol/L (reference value 0.5–2.2 mmol/L). She also developed anuric acute kidney injury with an increase in serum creatinine from 58 to 122  $\mu\text{mol/L}$  (reference value 50–98  $\mu\text{mol/L}$ ). Other admission labs are shown in *Table 1*. She was started on dual antiplatelet therapy (ASA 81 mg orally daily, clopidogrel 75 mg orally daily, and fondaparinux 2.5 mg subcutaneously daily) for suspected acute coronary syndrome and vasopressors to maintain perfusion. Point-of-care ultrasound demonstrated significant biventricular systolic dysfunction, a plethoric non-collapsible inferior vena cava, and diastolic flattening of the interventricular septum suggesting volume overload of the right ventricle.

The following differential diagnoses were considered at that time:

- (1) Acute coronary syndrome and possible mechanical complication with right ventricular (RV) infarction.
- (2) Underlying cardiomyopathy and/or pulmonary hypertension and superimposed acute decompensation.
- (3) Massive pulmonary embolism.

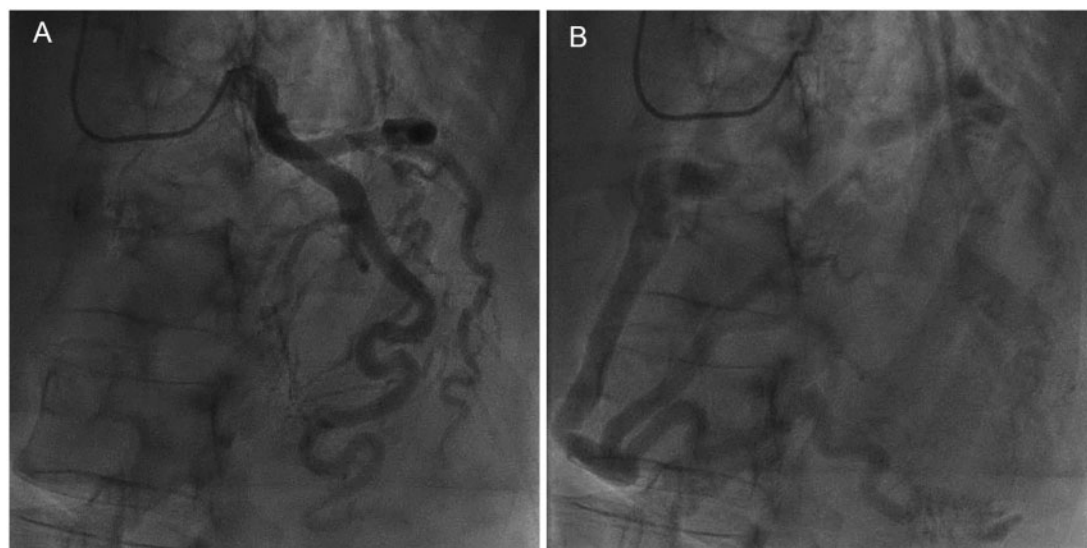
As the patient was in extremis, she was intubated for emergent cardiac catheterization. She was initiated on epinephrine, norepinephrine, and vasopressin prior to arrival to the catheterization lab. Angiographically, the left main coronary artery (LMCA), left anterior descending (LAD), and left circumflex (LCx) arteries were very large,



**Figure 1** Initial 12-lead electrocardiogram demonstrating rapid atrial fibrillation at 144 b.p.m., left axis deviation, and left bundle branch block.

**Table 1** Admission labwork

	Admission	8 h after admission	Reference range
Glucose (mmol/L)	6.4	2.7	3.8–11.0
Urea (mmol/L)	4.8	7.3	3.5–7.2
Creatinine ( $\mu\text{mol/L}$ )	58	122	50–98
Sodium (mmol/L)	131	127	135–145
Potassium (mmol/L)	4.9	5.1	3.5–5.0
Phosphate (mmol/L)	1.64	2.19	0.80–1.45
Magnesium (mmol/L)	0.96	0.92	0.66–1.07
Albumin (g/L)	33	31	35–50
Troponin (ng/L)	13	3610	$\leq 30$
pH	7.12	7.01	7.35–7.45
pCO <sub>2</sub> (mmHg)	26	41	35–45
Bicarbonate (mmol/L)	9	10	22–26
Lactate (mmol/L)	7.1	12.6	0.5–2.2
Leucocytes ( $\times 10^9/\text{L}$ )	8.3	16.9	4.0–11.0
Hemoglobin (g/L)	126	120	115–165
Platelet ( $\times 10^9/\text{L}$ )	220	103	150–400
Ferritin (g/L)		4361	
D-Dimer ( $\mu\text{g/L}$ )		>4000	<500
Aspartate Aminotransferase (AST; U/L)		4012	<35
Alanine Aminotransferase (ALT; U/L)		3301	<28
Gamma-glutamyl transferase (GGT; U/L)		181	<37
Alkaline phosphatase (ALP; U/L)		103	40–120



**Figure 2** Contrast injection of left main coronary artery in anterior-posterior cranial projection. (A) Dilated ectatic left main coronary artery and branches. (B) Delayed retrograde filling of the right coronary artery via collateral vessels back to the ostium into the pulmonary artery.

**Table 2** Haemodynamic parameters obtained during left and right heart catheterization

Left heart haemodynamics	
Aorta	150/80 mmHg
Left ventricle	150/0 mmHg
Left ventricular end-diastolic pressure	8 mmHg
Right heart haemodynamics	
Right atrium	Mean 25 mmHg
Right ventricle:	37/7 mmHg (mean 25 mmHg)
Pulmonary artery:	32/15 mmHg (mean 22 mmHg)
Pulmonary capillary wedge:	Mean 11 mmHg
Pulmonary vascular resistance	0.87 Wood units
Systemic vascular resistance	6.39 Wood units
Cardiac output	12.7 L/min
Cardiac index	7.4 L/min/m <sup>2</sup>

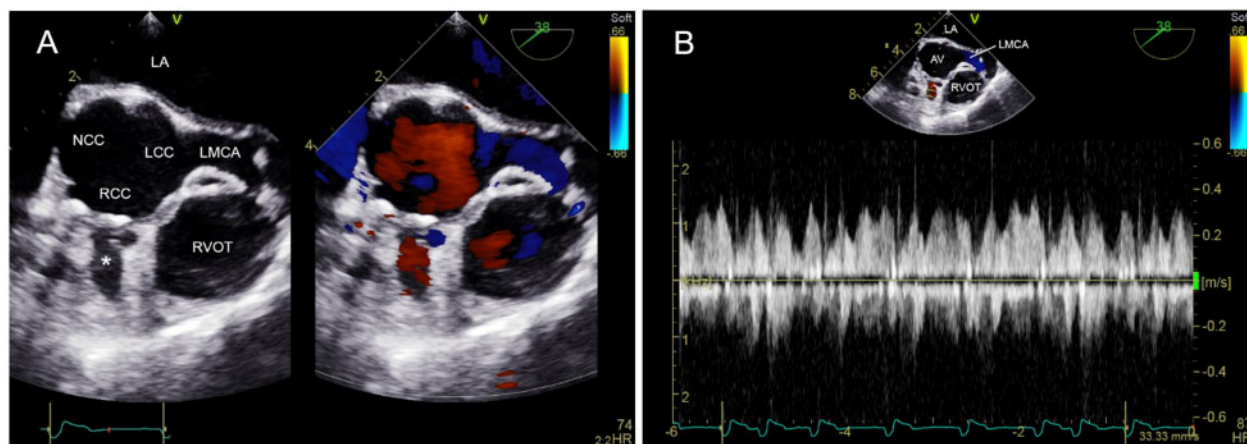
ectatic vessels with no flow-limiting stenosis. The right coronary artery (RCA) ostium could not be engaged from the aorta despite multiple attempts. Contrast injection into the left coronary system showed what appeared to be retrograde filling of the RCA via multiple left-to-right collateral vessels (Figure 2 and Video 1A and B). On further review, it was felt that the RCA was emptying back into the pulmonary artery/right ventricular outflow tract (RVOT).

The possibility of an anomalous coronary artery (ACA) as the underlying aetiology of the patient's shock prompted an *ad hoc* right heart catheterization for haemodynamic assessment. Measurements are shown in Table 2. An oximetry run did not suggest the presence of an intra-cardiac left-to-right shunt. Pulmonary artery angiography

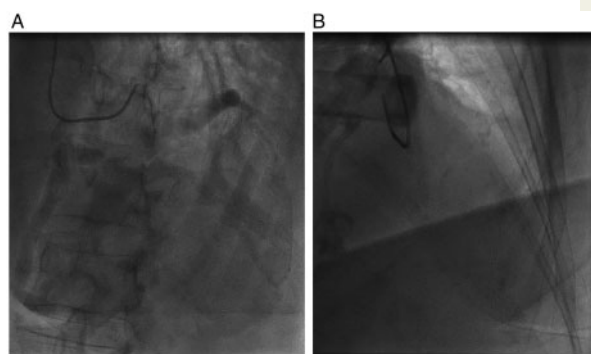
did not identify any pulmonary embolus or fistula involving the RVOT. The pulmonary vascular resistance was 0.87 Wood units and the systemic vascular resistance was 6.39 Wood units. An RV angiogram demonstrated significant tricuspid regurgitation (TR) with a dilated and severely dysfunctional right ventricle. It was felt that the cause of the patient's cardiogenic shock was due to underlying severe RV dysfunction with either AF or other catecholaminergic state triggering decompensation. The elevated cardiac index prompted consideration of sepsis on the differential diagnosis and the patient accordingly received empiric antibiotics (Piperacillin/Tazobactam 4.5 g IV every 6 h, Vancomycin 1 g IV every 12 h, and Doxycycline 100 mg orally twice daily). She returned to the cardiovascular intensive care unit (CVICU) for further physiologic support.

Formal echocardiography while in the CVICU demonstrated severely reduced RV systolic function, right atrial dilatation, and severe TR. The overall left ventricular systolic function was low-normal with an ejection fraction between 50% and 55% and no regional wall motion abnormalities. A transoesophageal echocardiogram ruled out vegetations but corroborated findings from the patient's cardiac catheterization including a dilated and tortuous LMCA and branches, as well as a dilated and tortuous vessel draining into the pulmonary artery. This vessel was felt to be likely the RCA. Overall, these findings were consistent with a diagnosis of ARCAPA (see Figure 3 and Video 2).

In the CVICU, the patient was maintained on inotropes and vasopressors. High-sensitivity troponin plateaued at 26 040 ng/L and this was felt to be consistent with an RV infarction. In the setting of concomitant volume overload and anuric acute kidney injury, continuous renal replacement therapy was initiated. She was subsequently found to be bacteremic with positive blood cultures for *Streptococcus bovis* but not stable enough to pursue a colonoscopy to rule out a



**Figure 3** Transoesophageal echocardiogram short-axis images at level of the aortic valve from the mid-oesophageal window. Colour Doppler (A) and spectral pulse-wave Doppler (B) images demonstrated a dilated vessel (\*) draining into the right ventricular outflow tract predominantly during diastole. This was consistent with anomalous origin of the right coronary artery.



**Video 1** Contrast injection of left main coronary artery in anterior-posterior (AP) cranial (A) and right anterior oblique (RAO) cranial (B) projections. The left main coronary artery and its branches are dilated and ectatic vessels. There is delayed retrograde filling of a large dominant right coronary artery via collateral vessels back to its ostium into the pulmonary artery.

gastrointestinal malignancy; management consisted of antimicrobial therapy (ceftriaxone/metronidazole) with consultation from infectious diseases.

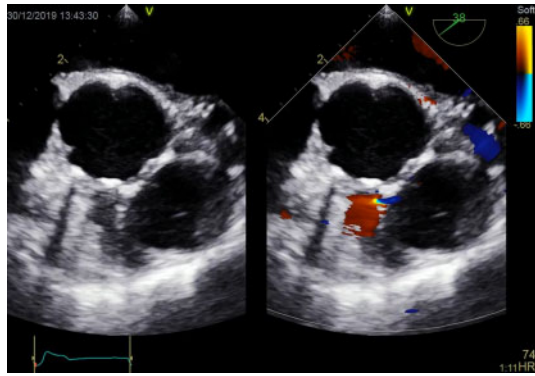
The patient's clinical status eventually stabilized enough to be extubated and transferred to the cardiology ward, although she remained dependent on intermittent haemodialysis without expected renal recovery. Potential therapeutic options for ARCAPA were discussed at interdisciplinary heart team rounds but unfortunately, she was felt not to be a surgical candidate in view of her age and clinical status. The patient decided to pursue comfort measures and requested cessation of dialysis. She was transferred to a hospice for palliative care and passed away just over a month later.

## Discussion

To the best of our knowledge, this is the first reported case where ARCAPA was diagnosed in the context of cardiogenic shock and new-onset AF. We postulate that the underlying aetiology of our patient's shock state was multifactorial, notably progressive RCA-territory ischaemia and RV failure, sepsis, and new-onset uncontrolled AF. The patient was already clinically in shock prior to arrival to the cardiac catheterization lab with tachycardia, refractory hypotension requiring inotropes/vasopressors, cool extremities, and end-organ dysfunction (oliguria, lactate of 17 mmol/L).

Pertaining to the patient's haemodynamic data, the elevated mean right atrial pressure could have been due to either chronic TR and/or RV dysfunction, while the increased cardiac index could have reflected an additional catecholaminergic contributor (e.g. sepsis, inotropes/vasopressors, tachyarrhythmia) to the patient's shock state. On balance, we felt that based on the patient's clinical presentation (cool extremities, cardiac biomarkers supporting acute RV infarction and moderate-to-severe RV systolic dysfunction on echocardiogram) the underlying aetiology of the patient's presentation was most consistent with right-sided cardiogenic shock with haemodynamic deterioration precipitated by sepsis and uncontrolled AF.

In our case, echocardiography and invasive angiography were integrated for the diagnosis of ARCAPA. Although gated cardiac computed tomography angiography is considered the gold standard investigation to confirm the diagnosis of an ACA, the patient's clinical circumstances (i.e. uncontrolled AF, renal failure requiring dialysis) did not permit this investigation. Definitive treatment involves surgical re-anastomosis of the abnormal coronary ostia back onto the aorta to normalize myocardial perfusion, after which progressive remodelling and improvement in the affected ventricle's function may be seen. If this is not feasible, an intrapulmonary aortocoronary tunnel may sometimes be implemented.<sup>6</sup> In our patient, surgical intervention was considered but felt not to be



**Video 2** Mid-oesophageal short axis images at the level of the aortic valve. Adjacent to the right coronary cusp, colour doppler images demonstrated an abnormal dilated vessel with colour flow (shown in red) suggesting drainage into the right ventricular outflow tract. This was consistent with an anomalous origin of the right coronary artery.

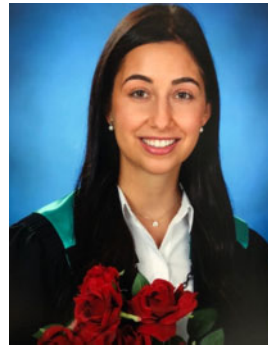
an option due to age, critical status, and perceived lack of reversibility of her disease.

Cardiogenic shock carries a broad differential diagnosis for which the most common cause is acute myocardial infarction. In the post SHOCK trial era, prompt recognition, urgent cardiac catheterization, and revascularization have demonstrated clear evidence of mortality benefit.<sup>7</sup> In our patient, urgent cardiac catheterization was diagnostic, but in a surprisingly different way than expected; no flow-limiting stenoses were identified but contrast injection into the left coronary system showed retrograde filling of the RCA via left-to-right collateral vessels. The RCA appeared to be emptying into the pulmonary artery/RVOT, providing a plausible explanation for RCA-territory ischaemia and cardiogenic shock. This case highlights the rare but important consideration of undiagnosed congenital heart disease (in this case, a rare coronary artery anomaly—which itself may not manifest on echocardiogram) in the adult presenting with unexplained cardiogenic shock.

## Conclusions

In summary, we present the case of an elderly patient presenting with right-sided cardiogenic shock and rapid AF, subsequently found to have ARCAPA, a rare coronary anomaly not easily visualized on transthoracic echocardiography. This case illustrates the importance of recognizing undiagnosed congenital heart disease as an uncommon cause of cardiogenic shock in adults and the need for multiple imaging modalities for workup when initial investigations fail to yield a conclusive diagnosis.

## Lead author biography



Dr Coralea Kappel is a 2nd-year internal medicine resident currently training at McMaster University in Hamilton, ON, Canada. Prior to this, she completed her Medical School at the University of Ottawa.

## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE best practice guidelines.

**Conflict of interest:** None declared.

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