


# The Appearance of an Underrecognized Congenital Heart Disease in a Patient With COVID-19 Pneumonia

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## Abstract

A 60-year-old woman presented to the emergency department with worsening shortness of breath and non-productive cough for 1 week, which was preceding a recent COVID-19 infection. At the time the patient thought this was part of the constellation of symptoms of COVID-19, so she stayed home until her symptoms worsened to the point of needing hospitalization. The patient was found to have a rare and complex congenital heart disease that led her to develop acute heart failure precipitated by COVID-19 pneumonia. Medical management and surgical repair were essential in this patient given the late presentation.

## Keywords

congenital heart disease, late onset Shone complex, hypertension, COVID, cardiology

## Introduction

During this pandemic, a multi-disciplinary approach is essential for many patients to provide the best care. This report describes the management of a woman thought to be presenting of acute respiratory failure due to COVID-19 pneumonia who was incidentally found to have a rare adult congenital heart disease (CHD) which required extensive collaborative medical and surgical management.

Shone complex was first described by Shone et al, in 1963 and is found to be a very rare CHD consisting of sub-aortic stenosis, supra-ventricular mitral ring, parachute mitral valve, and coarctation of aorta.<sup>1</sup> Most cases are diagnosed, and associated lesions are managed early in childhood. Late presentations have also been reported in the literature as either complete or incomplete variants of the syndrome.

Here, we present a case of a 60-year-old woman with a variant of Shone complex that was found after an acute onset of heart failure believed to be precipitated by her recent COVID-19 pneumonia. The effect of COVID-19 on patients with complex CHD remains to date unclear.

## Case Presentation

A 60-year-old woman with history of diabetes mellitus, hypertension, and hyperlipidemia presented to the emergency department with worsening shortness of breath and

non-productive cough for 1 week. Approximately 2 weeks prior, she was found to have COVID-19 pneumonia and self-quarantined. She thought her shortness of breath was a symptom of COVID-19 and waited until she eventually required hospitalization.

On examination, she was hypertensive with a blood pressure of 164/69 mm Hg, heart rate 64 bpm, and SpO<sub>2</sub> 83% on room air. On physical examination, she had mild bibasilar crackles and a harsh systolic murmur which became musical at the apex suggestive of Gallavardin phenomenon. She appeared near-euvolemic, jugular venous pressure was not elevated, and the patient had no lower extremity edema.

Footnote: Gallavardin phenomenon is a physical examination finding in patients with aortic stenosis (AS). It is characterized as a murmur that radiates to the apex and sometimes presents with a musical quality due to high frequency vibrations.

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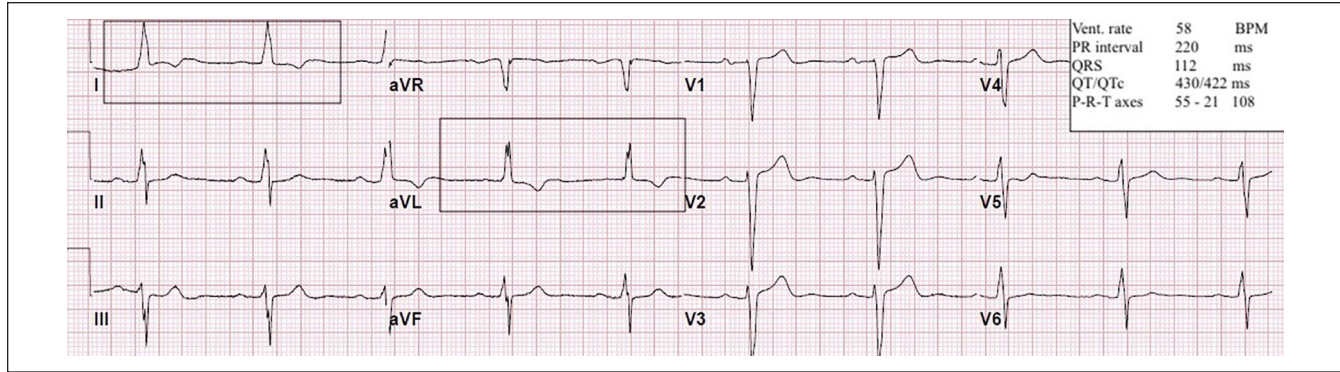
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**Figure 1.** ECG on admission: First degree atrioventricular (AV) block (PR interval 220 ms) with T wave abnormalities. Minimal voltage criteria for LVH.

Abbreviation: ECG = electrocardiogram.

**Table 1.** Initial Labs on Presentation.

Lab	Result	Reference
White blood count (WBC)	7.93	3.80-10.50 K/uL
Hemoglobin (HGB)	8.6	11.5-15.5 g/dL
Hematocrit (HCT)	28.2	34.5-45%
Blood urea nitrogen (BUN)	22	7-23 mg/dL
Creatine (Cr)	0.8	0.50-1.30 mg/dL
Potassium (K)	5.2	3.5-5.3 mmol/L
Magnesium (Mg)	1.40	1.3-1.9 mEq/L
Troponin	0.135	≤0.010 ng/mL
Beta Natriuretic Peptide (BNP)	183	0-100 pg/mL
D-Dimer Quant	665	0-243 ng/mL DDU

## Investigations

Initial work up was significant for sinus rhythm with first degree AV block and T wave abnormalities (Figure 1). Significant laboratory results on admission are found in Table 1. Computed tomography (CT) angiography of the chest (Figure 2) was obtained and revealed an ecstatic and atherosclerotic thoracic aorta, mild fusiform dilatation of the descending thoracic aorta as well as patchy ground glass attenuation within both lungs with areas of coalescing air-space consolidation. Patient was admitted with the diagnosis of acute hypercapnic respiratory failure secondary to COVID-19 pneumonia.

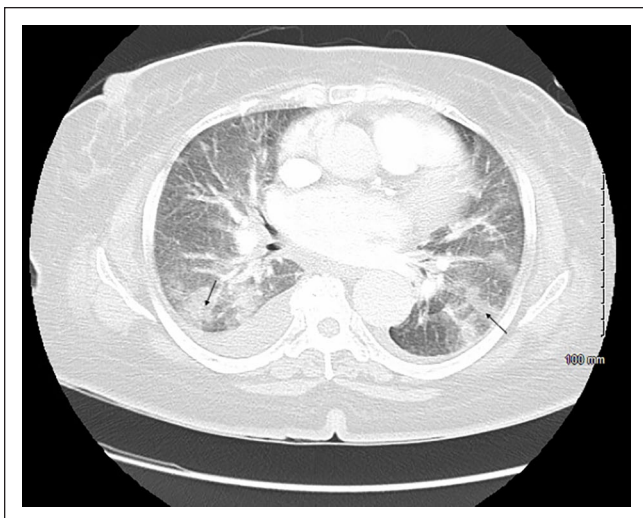
Follow-up transthoracic echocardiography (TTE) was obtained to evaluate the murmur heard on physical examination and revealed severe AS with preserved ejection fraction of 65%, grade III left ventricular diastolic dysfunction, and severely dilated left atrium (Supplemental Video 1). Aortic valve area was measured as 0.73 cm<sup>2</sup> with a mean valve gradient of 36 mm Hg and Peak valve gradient of 70 mm Hg (Figure 3). TTE also demonstrated an elevated velocity in the descending aortic arch likely due to AS, but there was concern for coarctation as well as mild mitral stenosis with parachute mitral valve. Magnetic resonance (MR) angiography

(Supplemental Videos 2 and 3) of the chest showed discrete coarctation of the aorta just distal to the origin of the left subclavian artery, in addition to bicuspid aortic valve with severe stenosis and right adrenal mass. The patient underwent bare metal stent placement for the coarctation. Then was taken for both left and right heart catheterization and required coronary artery bypass graft with aortic valve replacement (Supplemental Video 4).

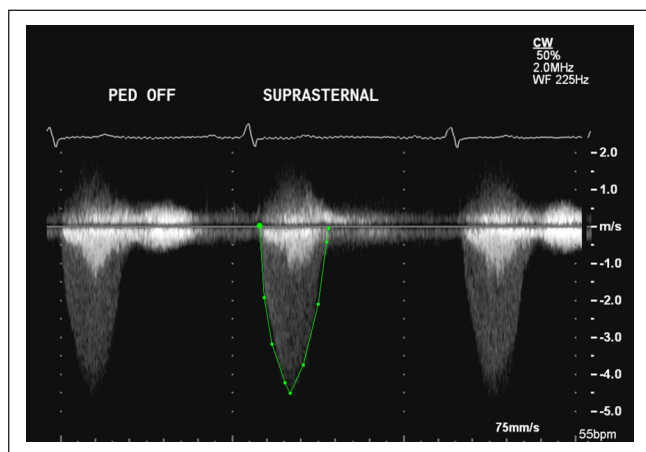
## Discussion

This case report reveals the uncommon constellation of findings in a woman who was found to have severe AS with Gallavardin phenomenon. On further evaluation, she was found to have both aortic coarctation (CoA) and mitral valve parachute. In this case, the patient most likely had a bicuspid aortic valve (BAV). The BAV and CoA are commonly associated in 85% of cases.<sup>2</sup> The CoA is not an uncommon congenital heart anomaly found in childhood, with an incidence ranging from 5% to 8% of all congenital cardiac defects.<sup>3</sup> It is very rare to diagnose CoA beyond the fourth decade of life because the survival of patients with a non-corrected severe CoA is impaired, with mortality rates as high as 75% in that decade of life and 90% at the end of the fifth decade of life.<sup>4</sup> The combination of AS with a parachute mitral valve and coarctation of aorta is known as a variant of Shone complex. Shone complex has been well recognized in children, yet only a few case reports were reported in adults. Variant forms of Shone complex to our knowledge have never been reported in patients of the age of 60 and older.

When it comes to CoA, the most common presenting sign in adults who have unrepaired CoA is hypertension as seen in this patient. CoA tends to be commonly overlooked as a cause of secondary hypertension. This patient in this case has had uncontrolled hypertension for several years and has been on 4 classes of antihypertensives. She has never been screened for coarctation but was worked up for all other



**Figure 2.** CT angiography of the chest with evidence of multifocal patchy ground glass attenuation (see arrows) within both lungs and areas of coalescing airspace consolidation. Abbreviation: CT = computed tomography.



**Figure 3.** Doppler echocardiography: Continuous-wave measurement of aortic valve Vmax from the apical echo window.

causes of secondary hypertension. Her blood pressure remained controlled for several years until she became ill with COVID-19. The COVID-19 infection has been shown to cause life threatening illnesses in patients with cardiovascular risk factors. The impact on adults with CHD affected by COVID-19 remains unknown to date as well as the challenges associated with the care of this specific population. It is unclear if this patient decompensated after developing COVID-19. Typically, individuals with CHD are younger than those with acquired disease, and less likely to have comorbidities associated with severe COVID-19 infection; but it is unclear if having a CHD is also associated with infection severity.

Very few case reports in the literature were described with a late diagnosis of Shone complex and unrepaired CoA. Shone complex must be diagnosed or suspected by echo; however, multimodality imaging allows better evaluation of intra and extra-cardiac obstructive.<sup>5,6</sup> If either component of Shone complex is detected, a comprehensive work up is essential and should be performed to proceed with proper surgical and medical management. Management of patients with Shone complex is usually performed in stages. The first surgical intervention according to the literature is CoA repair early in life allowing patients to remain asymptomatic for a long period of time until new worsening of valvular lesions.<sup>7</sup> In this patient, as she has not had any repairs during her childhood, she initially underwent stent placement for CoA, followed by coronary bypass graft (CABG) with aortic valve repair. The CABG was performed after the patient was found to have 90% occlusion of the mLAD on left heart catheterization.

With further literature review, the genetic underpinnings of Shone complex are essential in understanding the pathophysiology as well as in improving management options. What is emerging is an improved understanding of how certain underlying genetic factors play a role in clinical outcomes. It is important to consider these factors when assessing the effectiveness of interventions and new treatment approaches.<sup>8</sup> The MYH6 (a myosin heavy chain 6) is commonly seen in autosomal dominant familial atrial septal defect (ASD) and sporadic cases of complex congenital heart disease, including Shone complex and Hypoplastic left heart syndrome (HLHS).<sup>9</sup>

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### Declaration of Conflicting Interests

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### Ethics Approval

Our institution does not require ethical approval for reporting individual cases. This is an observational case report describing a patient's clinical course. We confirm that the manuscript has been read and approved by all named authors.

### Informed Consent

Verbal informed consent was obtained from the patient for their anonymized information to be published in this article.

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**Supplemental Material**

Supplemental material for this article is available online.

**References**

1. Shone JD, Sellers RD, Anderson RC, Adams P Jr, Lillehei CW, Edwards JE. The developmental complex of “parachute mitral valve,” supravulvular ring of left atrium, subaortic stenosis, and coarctation of aorta. *Am J Cardiol.* 1963;11:714-725.
2. Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease. *Eur Heart J.* 2010;31:2915-2957. doi:10.1093/eurheartj/ehq249.
3. Teo LL, Cannell T, Babu-Narayan SV, Hughes M, Mohiaddin RH. Prevalence of associated cardiovascular abnormalities in 500 patients with aortic coarctation referred for cardiovascular magnetic resonance imaging to a tertiary center. *Pediatr Cardiol.* 2011;32:1120-1127.
4. Bhatt AB, Foster E, Kuehl K, et al. Congenital heart disease in the older adult: a scientific statement from the American Heart Association. *Circulation.* 2015;131:1884-1931.
5. Lorenzo N, Claver E, Alió J, Aguilar R. Incomplete Shone’s complex in the sixth decade of life: echo and cardiac magnetic resonance imaging assessment. *Eur Heart J.* 2017;38(6):459.
6. Aslam S, Khairy P, Shohoudi A, et al. Shone complex: an under-recognized congenital heart disease with substantial morbidity in adulthood. *Can J Cardiol.* 2017;33(2):253-259. doi:10.1093/eurheartj/ehw390.
7. Grimaldi A, Vermi AC, Ho SY, et al. Surgical outcome of partial Shone complex. *Interact Cardiovasc Thorac Surg.* 2012;14(4):440-444.
8. Russell MW, Chung WK, Kaltman JR, Miller TA. Advances in the understanding of the genetic determinants of congenital heart disease and their impact on clinical outcomes. *J Am Heart Assoc.* 2018;7:e006906.
9. Jin S, Homsy J, Zaidi S, et al. Contribution of rare inherited and de novo variants in 2,871 congenital heart disease probands. *Nat Genet* 2017;49:1593-1601.