

## The Importance of the Pulmonic Valve Morphology in Pulmonary Hypertension

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**Received:** June 19, 2019; **Published:** October 21, 2019

### Abstract

Pulmonic valve prolapse has been described in association with the myxomatous “floppy” mitral valve syndrome and in congenital heart disease associated with pulmonary hypertension. A literature search suggests this finding of an isolated myxomatous pulmonary valve with prolapse in adults with pulmonary hypertension is primarily or due to left sided heart conditions.

This case report describes an isolated myxomatous “floppy” pulmonic valve in a patient with primary pulmonary hypertension. Prolapse of the pulmonic valve is an echocardiographic qualitative hint of pulmonary hypertension. It is plausible to explain the morphological changes of the pulmonary valve as due to exaggeration of pulmonic valve excursion due to severely elevated pulmonary artery diastolic pressure.

**Keywords:** *Pulmonic Valve Morphology; Pulmonic Valve Prolapse; Myxomatous Pulmonic Valve; Pulmonary Hypertension*

### Learning Objectives:

- Recognition of the association between pulmonary valve prolapse and pulmonary hypertension
- The importance of careful and dedicated imaging of the pulmonary valve.
- Recognize the sensitivity, specificity and positive predictive value of isolated myxomatous pulmonic valve in patients with pulmonary hypertension have not been established but the association is real.

### Introduction

Pulmonic valve prolapse has been described in association with the myxomatous “floppy” mitral valve syndrome and in congenital heart disease associated with pulmonary hypertension.

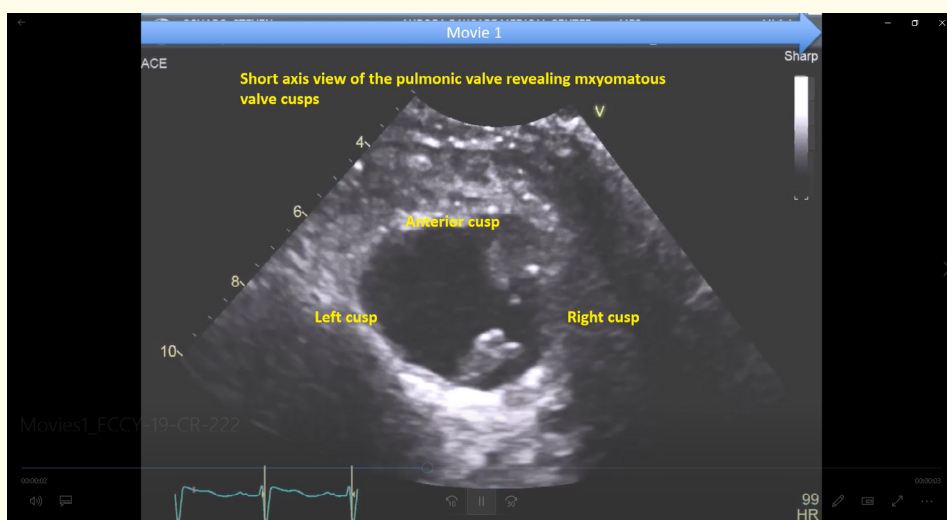
### Case Report

A 40-year-old male was admitted to the hospital with pleuritic chest pain and progressively increasing shortness of breath. He had been experiencing racing heart sensation for the previous four weeks. Past medical history was significant for hypertension and elevated body mass index. He denied pre-syncope or loss of consciousness.

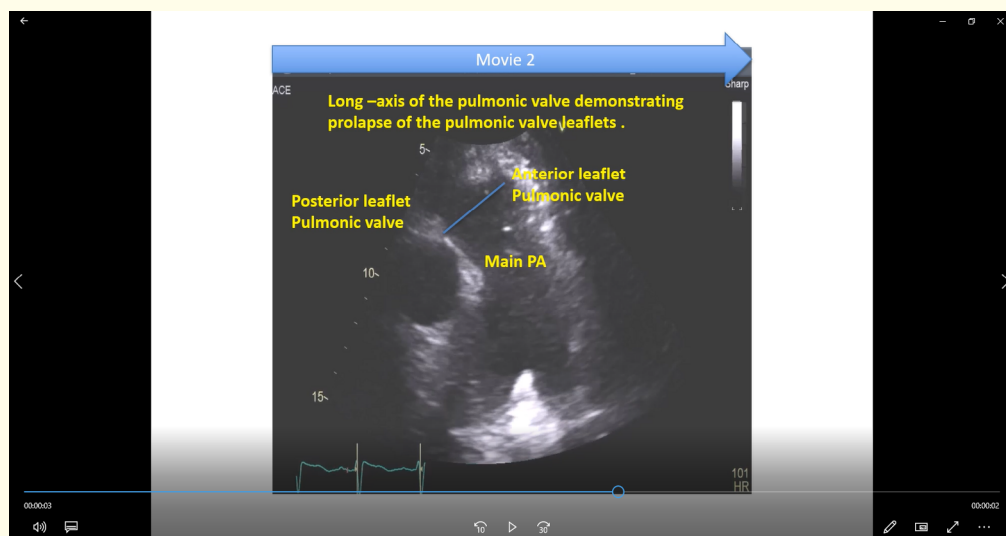
Physical examination revealed an obese male (BSA ~ 2.4), with a blood pressure of 180/100 and pulse of 98 bpm. Heart tones were distant without ejection click or systolic or diastolic heart murmur. Crackles were appreciated in lower lung fields. There was pitting edema in the lower extremities.

The chest X-ray revealed small bilateral pleural effusion and enlarged pulmonary arteries. Urgent chest computed tomography revealed no evidence of pulmonary emboli but commented on enlarged pulmonary arteries.

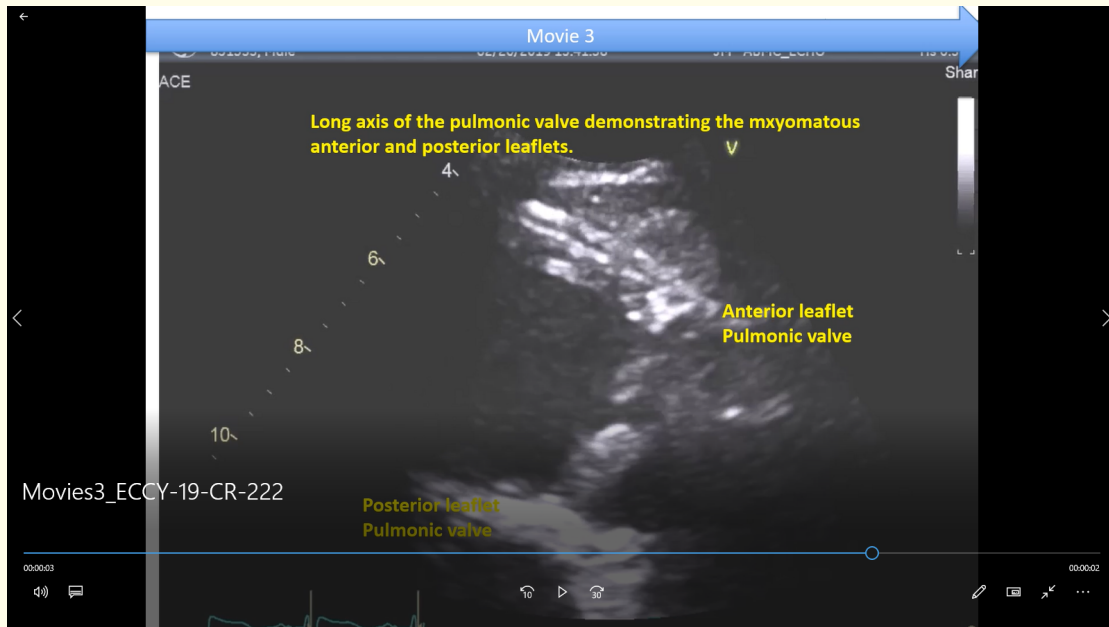
Echocardiography revealed unusual pulmonary valve morphology with a myxomatous pulmonary valve with pulmonic valve prolapse (Movie 1-3) and moderate pulmonary regurgitation (Movie 4). Echo Doppler interrogation revealed elevated pulmonary regurgitation end-diastolic velocity suggesting elevated pulmonary artery end diastolic pressure. The right ventricle was remodeled with severe enlargement and depressed right ventricular function. The inferior vena cava and hepatic veins were plethoric (Movies 5 and 6) and the tricuspid regurgitation jet revealed severe pulmonary hypertension (Movie 7). Right heart cardiac catheterization revealed severe elevation of pulmonary artery diastolic and systolic pressure concordant with the echocardiographic findings. Blood cultures were negative. CBC, CHEM-7, sedimentation rate and urinalysis were normal. The pulmonary hypertension team was consulted.



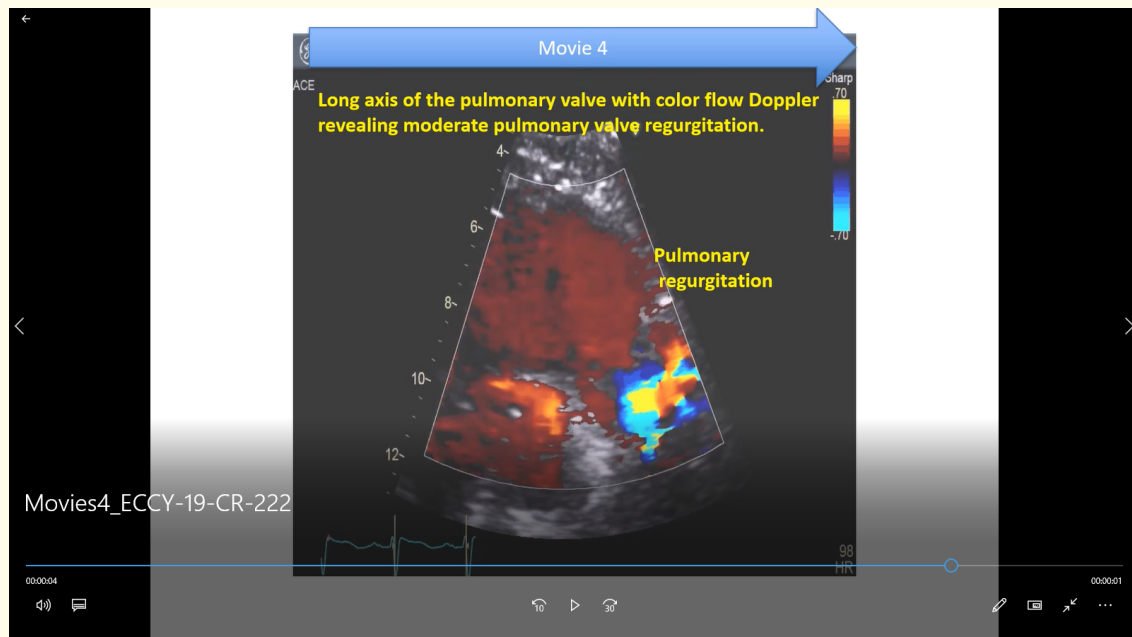
Movie 1: Short axis view of the pulmonic valve revealing myxomatous valve cusps.



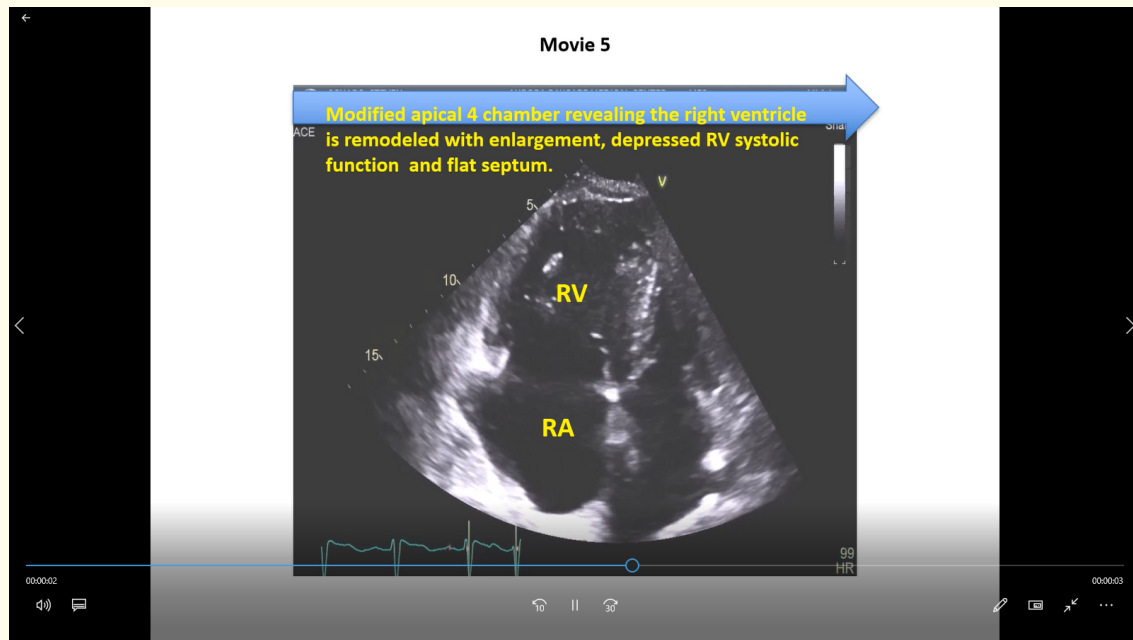
Movie 2: Long axis of the pulmonic valve demonstrating: prolapse of the pulmonic valve leaflets.



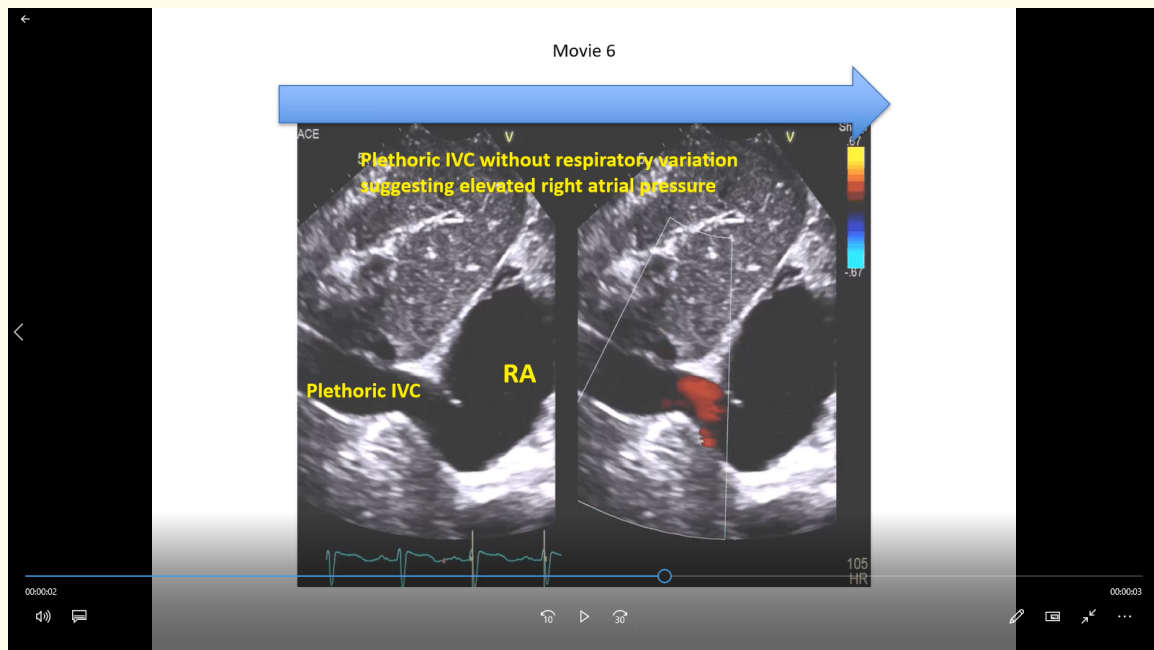
**Movie 3:** Long axis of the pulmonic valve demonstrating: the myxomatous anterior and posterior leaflets.



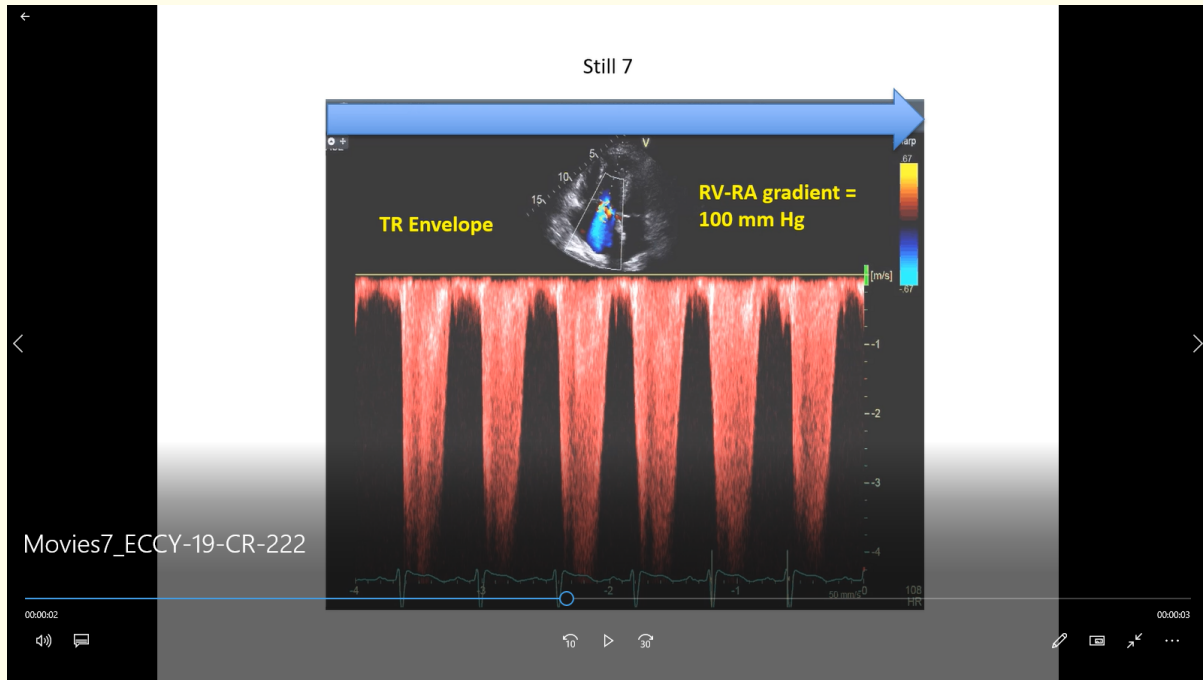
**Movie 4:** Long axis of the pulmonary valve with color flow: Doppler revealing moderate pulmonary valve regurgitation.



**Movie 5:** Modified apical 4 chamber revealing: the right ventricle is remodeled with enlargement, depressed RV systolic function and flat septum.



**Movie 6:** Plethoric IVC without respiratory variation suggesting elevated right atrial pressure.



*Movie 7: Tricuspid Valve regurgitation demonstrating RV - RA = 100 mm Hg.*

## Bibliography

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Volume 6 Issue 11 November 2019

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