

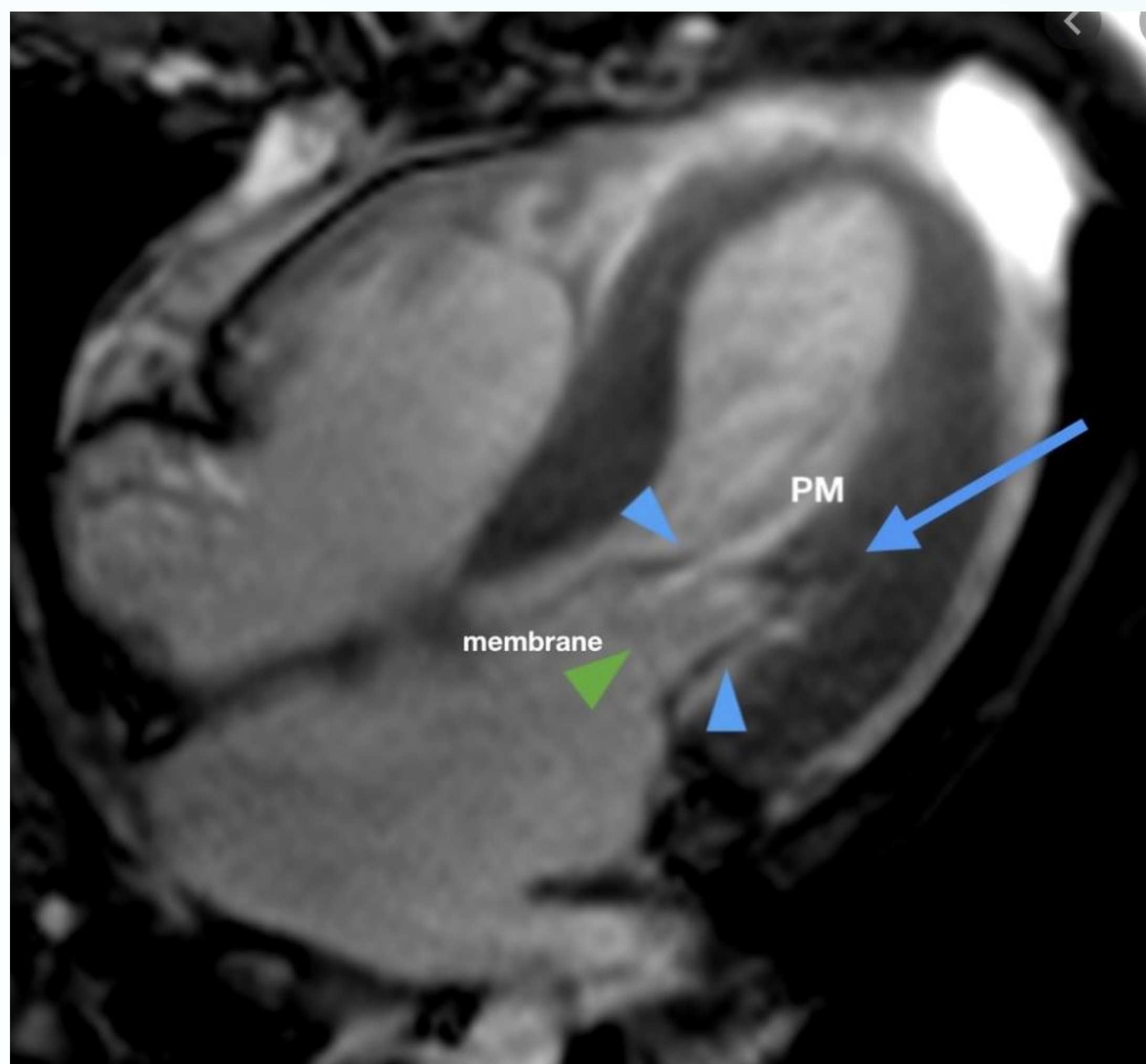
Severe Pulmonary Hypertension and Biventricular Heart Failure in an Adult Patient with Shone's Complex

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Introduction

Shone's Complex is a form of congenital heart disease that consists of a constellation of findings including coarctation of the aorta, valvular and subvalvular aortic stenosis, a supralvalvular mitral membrane, and a "parachute" mitral valve. We present an adult patient with Shone's Complex who developed severe pulmonary hypertension and biventricular failure as a result of her congenital anomaly.



•Image 2- reference 2

- blue arrowheads and arrow: chordae tendineae attached mainly to a single papillary muscle (parachute mitral valve)
- green arrowhead: thin mitral membrane

Case Report

A 59 year-old female with history of Shone's complex, heart failure with preserved ejection fraction with mild right ventricular failure, mixed pre and post capillary pulmonary hypertension, chronic atrial fibrillation, was admitted for worsening dyspnea and weight gain, despite being on furosemide and sildenafil. She has a history of multiple cardiac surgeries for her congenital heart disease including surgery to repair her coarctation of the aorta at age 7, mitral valve replacement for supra mitral ring and concomitant parachute mitral valve at age 9, transverse aortic arch replacement with multibranch grafting, and reconstruction of her left common carotid and subclavian at the age of 48. During admission, she had an echocardiogram which showed left ventricular ejection fraction of 50-60%, severe tricuspid regurgitation, moderate aortic stenosis, and severe pulmonary hypertension with a pulmonary artery systolic pressure of 70mmHg. She also underwent a right heart catheterization which showed a cardiac output of 2.98 L/min, cardiac index of 1.78 L/min/m², right atrial pressure of 13mmHg, PA pressure of 85/50mmHg with mean of 61mmHg, and pulmonary capillary wedge pressure of 25mmHg. The aforementioned findings were consistent with severe biventricular failure in the setting of severe pulmonary arterial hypertension. The patient was diuresed with furosemide during her admission which resulted in symptomatic relief. Subsequently upon stabilization, she was recommended to undergo a cardiac MRI for evaluation of her anatomy and for consideration of a combination heart-lung transplant for definitive therapy.

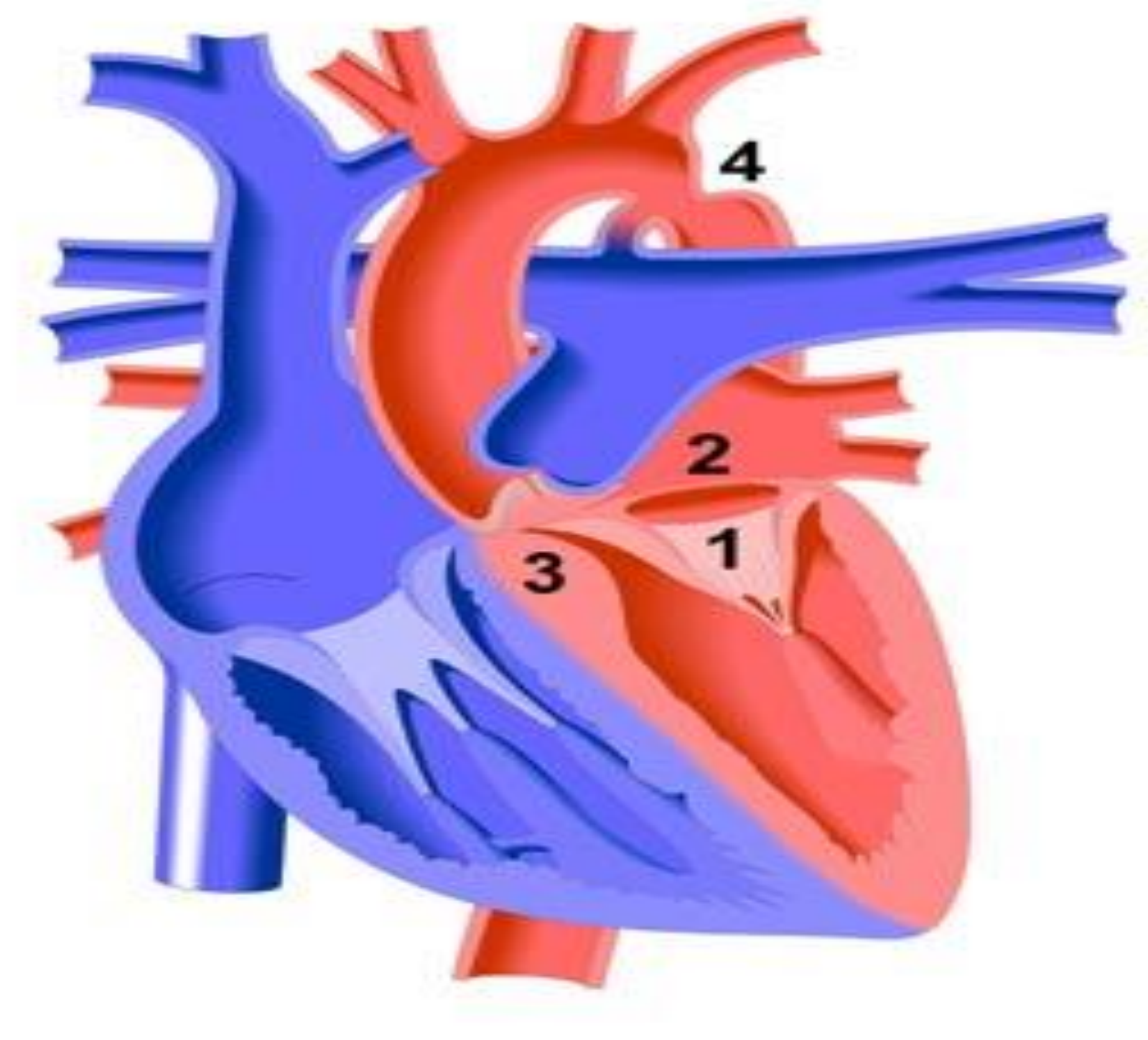


Image -1- reference 1

1. parachute mitral valve
- 2 – supralvalvar mitral membrane
- 3 – subaortic stenosis
- 4 – coarctation of the aorta

Discussion

Shone's Complex is a rare clinical entity with an incidence of <1% of all congenital heart diseases. It consists of an array of obstructive abnormalities of the left side of the heart, including a parachute mitral valve, supralvalvular mitral membrane, subaortic stenosis, and coarctation of the aorta. These abnormalities often predispose a patient to develop heart failure, arrhythmias, or require cardiovascular surgeries. Severe biventricular failure can result from impaired ventricular filling secondary to the modified right to left ventricular interaction in the setting of severe pulmonary arterial hypertension..

Conclusion

Early surgical intervention is often indicated in order to prevent the development of pulmonary arterial hypertension and its associated sequelae in patients with Shone's Complex. Recognition of pulmonary hypertension in Shone's Complex in the adult population is crucial since this is a late complication with limited therapeutic options, primarily focusing on transplantation in select candidates

References

1. Burns, Paul Burns and Jasper. "Shone's Complex: Congenital Heart Disease - Cove Point Foundation: Johns Hopkins Children's Hospital." *Shone's Complex | Congenital Heart Disease - Cove Point Foundation | Johns Hopkins Children's Hospital*, www.pted.org/?id=shones1. -Image -1
2. Cuevas, David. "Shone Complex: Radiology Case." *Radiopaedia Blog RSS*, Radiopaedia, radiopaedia.org/cases/shone-complex. Image 2