

## Pseudoaneurysm of the Mitral-Aortic Intervalvular Fibrosa: Two Cases of a Rare Complication of Aortic Valve Surgery

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### Type of submitter

Fellow in Training

### Abstract

#### INTRODUCTION

Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (P-MAIVF) is defined as an outpouching of fibrous tissue in the area between the aortic and mitral annular rings, in particular dividing the left (noncoronary aortic) cusp and the anterior mitral valve leaflet. Disruption of the fibrous skeleton by mainly aortic valve surgery or infectious endocarditis may result in pseudoaneurysm formation and, thus, potential risk for rupture into the pericardial space, coronary compression, fistula formation with adjacent structures, or thromboembolism.

#### CASE PRESENTATION

Case #1: An 84-year-old male with a remote history of severe aortic stenosis status post bioprosthetic aortic valve replacement underwent follow up transthoracic echocardiogram (TTE,) which was notable for mild to moderate prosthetic aortic valve stenosis and mild aortic insufficiency with two jets, one in the coaptation zone and the other close to the commissure. A transesophageal echocardiogram (TEE) was performed to further characterize the aortic insufficiency, which showed a 1.2 - 1.3 cm pseudoaneurysm of the MAIVF. Cardiac MRI confirmed a 2.5 cm in length (from the base of the anterior mitral leaflet) and 1.5 cm in width pseudoaneurysm of the MAIVF measured in systole. Patient declined surgical intervention and later succumbed to urosepsis.

Case #2: A 60-year-old male with two prior aortic valve replacements, the first for bicuspid valve with severe aortic stenosis followed by repeat two years later for paravalvular leak and severe aortic insufficiency, was admitted for evaluation of progressive dyspnea on exertion and hemolytic anemia. TEE was performed to rule out mechanical hemolysis which was notable for P-MAIVF with mural thrombus, prosthetic aortic stenosis, and paravalvular leak. He was referred to a valve center where he successfully underwent 3rd redo sternotomy, aortic root replacement with reimplantation of the coronary arteries, and ascending aortic repair of the paravalvular leak.





## DISCUSSION

The avascular nature of MAIVF makes it prone to infection and injury. The most common causative factors for P-MAIVF are aortic valve surgery and endocarditis. Potential complications include rupture, fistulous communications with adjacent structures, and thrombosis. High risk features for progression of P-MAIVF include active endocarditis, P-MAIVF large than 3cm, bicuspid aortic valve, aortic regurgitation, presence of fistula, thrombus, and compression of adjacent structures (coronary or pulmonary arteries.) Diagnosis using echocardiography is made by visualization of an echo-free space with systolic expansion and diastolic collapse. CT and MRI have been assessed as alternative, noninvasive methods for diagnosing P-MAIVF that may avoid limitations of echocardiography. Recent innovations in percutaneous methods for correction of structural cardiac abnormalities may allow for non-surgical intervention of P-MAIVF in prohibitive risk patients, however surgical repair has been the preferred treatment method.

## CONCLUSION:

P-MAIVF is a rare complication of prior surgical trauma or infective endocarditis. Although TTE may detect P-MAIVF, TEE is the study of choice with cardiac CT/MRI as an optional noninvasive method when diagnosis is ambiguous. The natural course of P-MAIVF is not clear, nor is recommended long term management; however, given potential risk for potentially devastating complications as described in prior case reports, repair is usually pursued.

## Categories

2nd year Fellow: Case

## Program Name

Wright State University Boonshoft School of Medicine

## Adrenal Insufficiency: A Rare Cause of Complete Heart Block

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### Type of submitter

Fellow in Training

### Abstract

#### Introduction

Adrenal insufficiency is a rare disease that affects 1 in 100,000 people. We report a case of complete heart block due to secondary adrenal insufficiency.

#### Case Description

A 55-year-old man with history of COPD, hypertension and CKD-3, presented with syncope. He was sitting in a chair at home and started to feel dizzy before passing out. In the ED, he was hypotensive with a blood pressure of 74/54 mmHg and pulse of 32/min. 12-Lead ECG revealed third-degree AV block with junctional escape rhythm at a rate of 35/min [Figure 1].

His initial labs including CBC, CMP, troponin-T, and TSH were within normal limit. Urine toxicology was positive for cocaine and benzodiazepines.

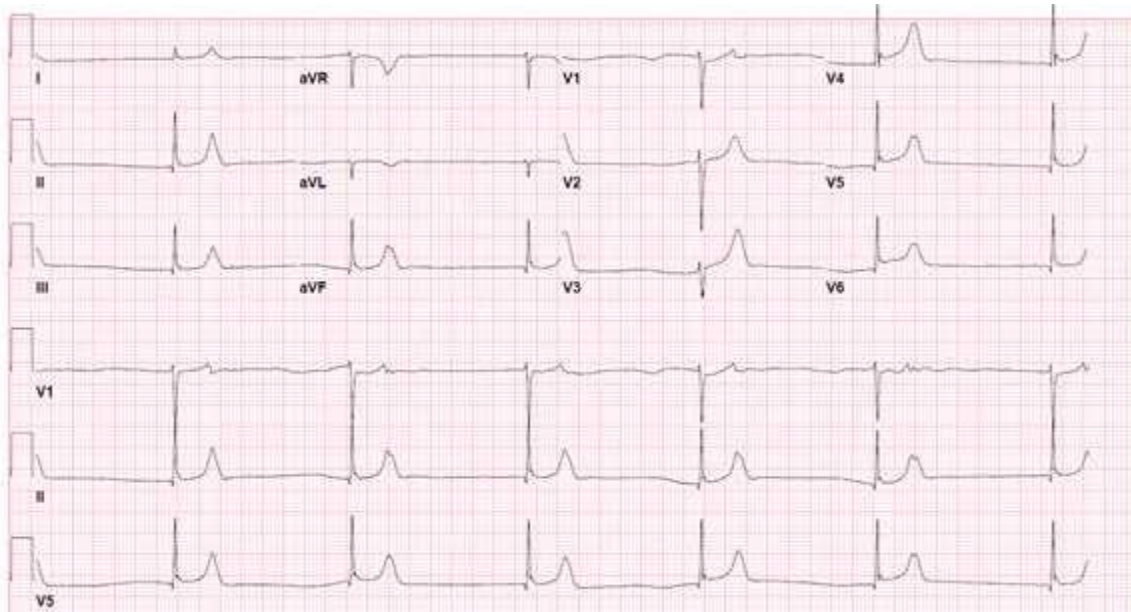


Figure 1. A 12-lead ECG showing complete heart block with junctional escape rhythm

He was given 1 mg of IV Atropine without any improvement. An emergent temporary transvenous pacemaker was placed. The pacemaker rate was set at 90/min due to hypotension. He continued to remain hypotensive with MAP less than 60 mmHg, and required multiple vasopressors. Given refractory shock, cortisol level was ordered 6 hours after presentation, and was very low at 1.6 ug/dL (normal 6.0 - 18.4 ug/dL). His ACTH level was low at 6 pg/mL (normal 7 - 69 pg/mL), but his aldosterone level was normal at 8.1 ng/dL (normal 4.0 - 31.0 ng/dL). One dose of dexamethasone IV was administered and IV hydrocortisone 100 mg every 8 hours was initiated.

Within 12 hours of corticosteroid replacement therapy, he was weaned off vasopressors. The temporary pacer was successfully weaned to 60/min. A repeat 12-lead ECG showed normal sinus rhythm at a rate of 80/min with 1<sup>st</sup> degree AV block [Figure 2]. The temporary pacemaker was removed. His heart rhythm and rate remained stable. Upon further questioning, he acknowledged taking prednisone frequently over the last 12 months for COPD exacerbations. The remainder of his hospital course was uneventful. He was discharged on a slow tapering dose of glucocorticoids with planned follow-up with an endocrinologist.

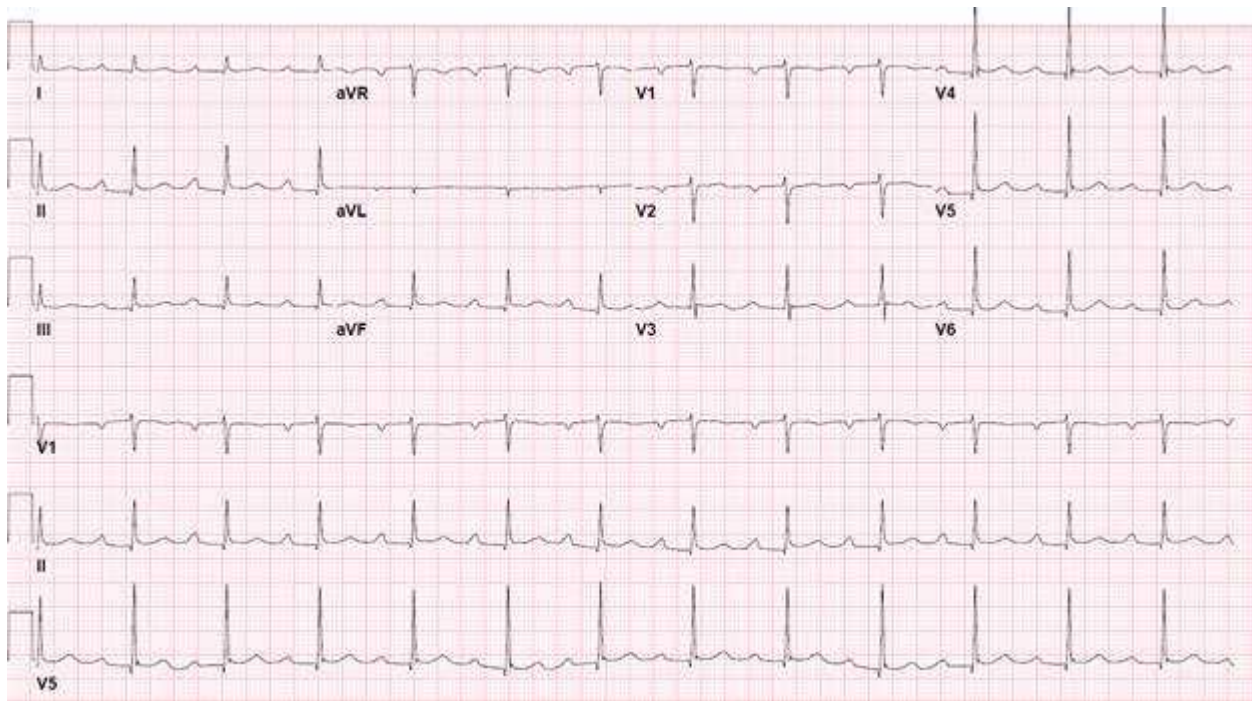


Figure 2. A 12-lead ECG showing normal sinus rhythm with 1<sup>st</sup> degree AV block.

### Discussion

Adrenal insufficiency is classified as primary (Addison's disease), secondary, or tertiary. It arises from hypothalamic–pituitary–adrenal axis malfunction. Addison's disease results in low cortisol and aldosterone with high ACTH, while secondary adrenal insufficiency results in low cortisol and ACTH, but normal aldosterone.

Our patient received frequent prescriptions of glucocorticoids. His ACTH level was low indicating that he had secondary adrenal insufficiency caused by iatrogenic suppression due to repeated steroid doses.

He showed rapid improvement in his blood pressure and heart rate with steroid replacement. Other more common causes of complete heart block, including thyroid disease, infection, hypoxia, acidosis, medications, and malignancy, were ruled out.

### Conclusion

This case highlights another metabolic cause that should be considered as part of the differential diagnosis of patients presenting with complete heart block. Adrenal insufficiency can manifest with cardiac rhythm disturbances, such as complete heart block.

### Categories

2nd year Fellow: Case

### Program Name

Mercy St. Vincent Medical Center

## A Case of an Aberrant Right Subclavian Artery arising from the Right Pulmonary Artery in a Patient with Complete Transposition of the Great Arteries

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### Type of submitter

Fellow in Training

### Abstract

#### Introduction

While an aberrant right subclavian artery (RSCA) is the most common congenital anomaly of the aortic arch, it is still relatively uncommon, only occurring in 0.7-2% of the general population. Even more rare, there is the entity of the right subclavian artery arising from the right pulmonary artery (RPA), also known as 'isolation' since there is no longer an aortic-subclavian connection. We report an echocardiographic diagnosis of this anomaly in a newborn with Complete Transposition of the Great Arteries (TGA) and its implications.

#### Case Presentation

We present a case of a former 34 week low birth weight infant male born to a mother with an antenatal history of gestational diabetes. He had significant cyanosis at birth and was postnatally diagnosed with TGA with an intact ventricular septum. Specifically, his initial echocardiogram showed situs solitus, a restrictive atrial septum, atrio-ventricular concordance, ventricular-arterial discordance, a left sided aortic arch with a suspected aberrant right subclavian artery, and a patent ductus arteriosus (PDA). Interestingly, his aortic valve was anterior and *leftward* in relation to pulmonary valve, as opposed to being anterior and *rightward* in most cases of complete transposition and therefore usually referred to us as D-TGA (Image # 1). The patient remained profoundly hypoxemic despite adequate ductal patency with Prostaglandin E infusion and oxygen supplementation, so a balloon atrial septostomy was performed. After septostomy, the patient revealed a very interesting clinical finding; the oxygen saturation in his right upper extremity was >95% which was higher than all his other extremities. Further echocardiographic imaging showed that the RSCA was not arising anomalously from the upper descending aorta, but rather anomalously from the RPA, therefore explaining the higher oxygen saturation in the context of complete transposition (Image #2).

The intraoperative findings confirmed the origin of the RSCA arising from the RPA. Arterial switch was performed with re-anastomosis of the RSCA to the proximal descending aorta. The post-operative course was uneventful.

#### Discussion

An anomalous connection between the RSCA from the RPA is a rare finding, especially in TGA. Embryologically, isolation of RSCA is theorized to be from the remnant of the right sided PDA maintaining its connection to the RPA. Diagnosis is made by clinical findings and imaging techniques such as trans-thoracic echo, computed tomography arteriography, magnetic resonance arteriography, or diagnostic angiography under cardiac catheterization. In this case, early diagnosis of this anomaly allowed us to surgically address it at the time of the arterial switch and avoid the complications of post-operative hypoxemia in the right arm, as well as subclavian steal as pulmonary pressures decreased post-operatively.

#### Conclusion

Anomalous RSCA from the RPA is a very rare entity that can occur in TGA. Timely diagnosis allows for early surgical intervention to avoid the complications of hypoxemia in the right arm as well as subclavian steal as pulmonary pressures decrease after arterial switch.

Images

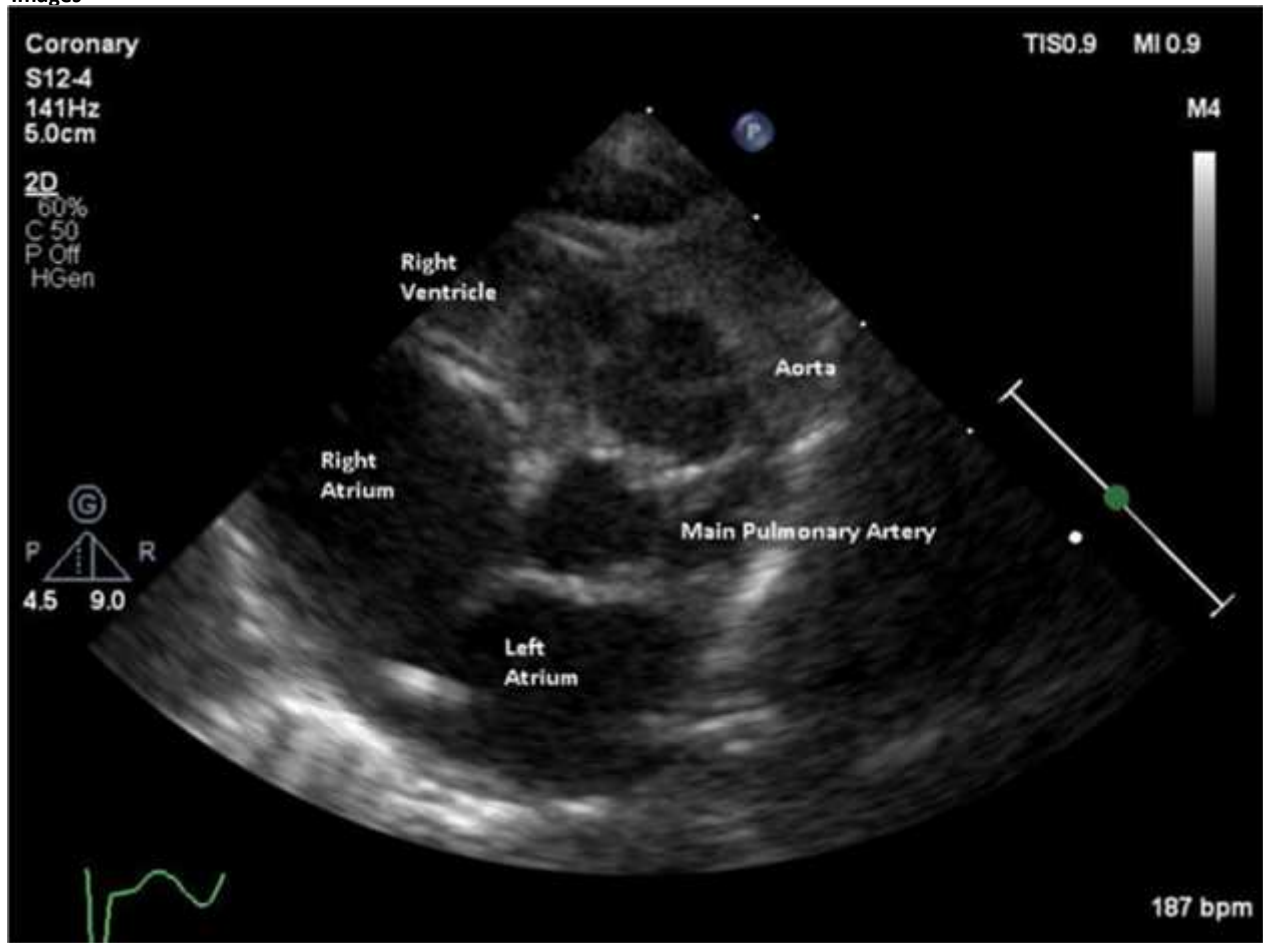


Image #1: The Aorta is more anterior and leftward in relation to the Main Pulmonary Artery

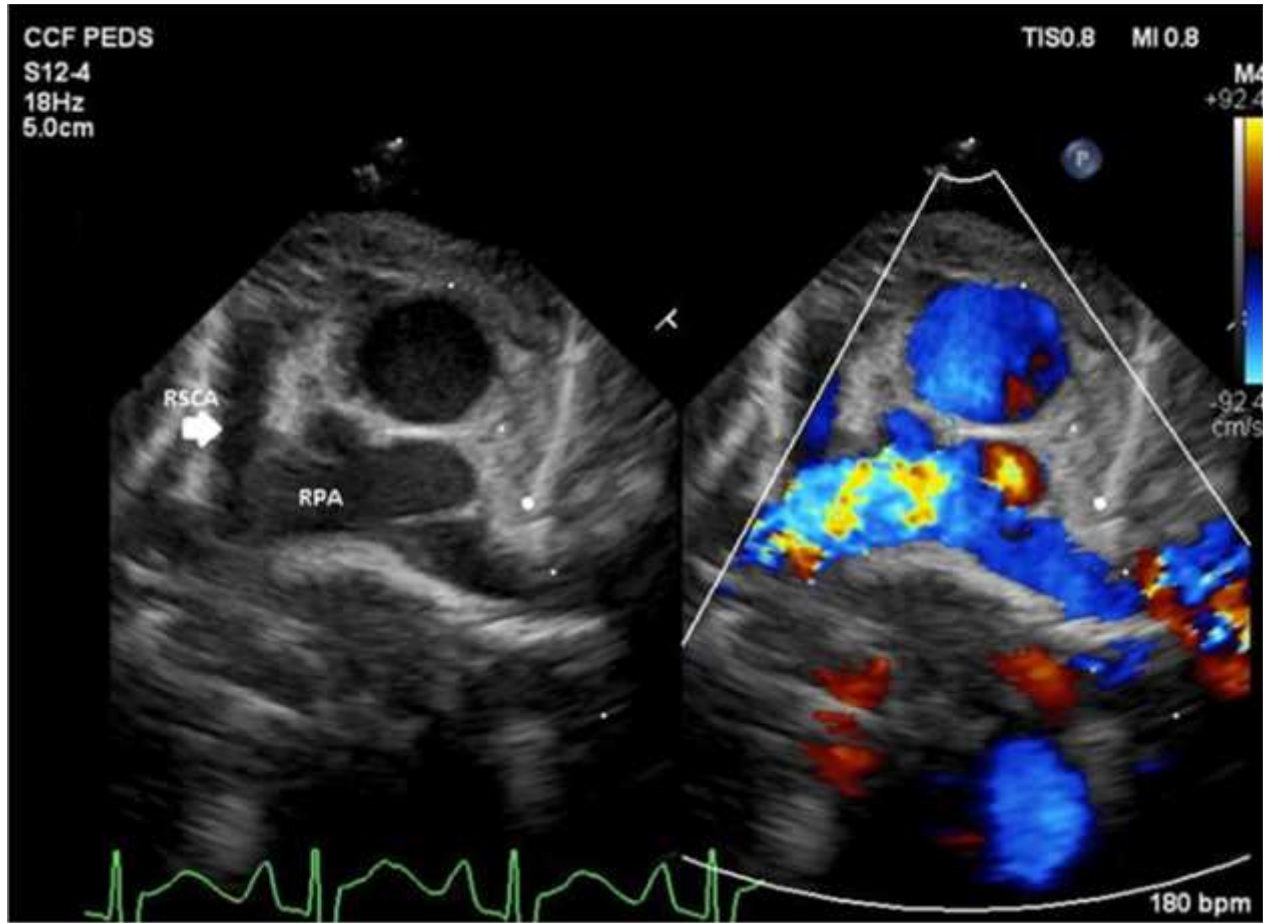


Image #2: The Right Subclavian Artery is arising from the Right Pulmonary Artery

**Categories**

2nd year Fellow: Case

**Program Name**

Cleveland Clinic Children's

## A Case for Caution: The Use of Estrogen Contraception and Hypertriglyceridemia

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### Type of submitter

Fellow in Training

### Abstract

#### Introduction/objective

Mild hypertriglyceridemia is common lipid disorder with a prevalence of 33% of the US population. Moderate hypertriglyceridemia (500-1000 mg/dL) is much less common, in only 1.7% of patient in the US and has a strong association with atherosclerosis and increased cardiovascular events. We present a case of presumed familial hypertriglyceridemia for several years duration being driven by oral estrogen contraception.

#### Case presentation

A 34 year old woman with past medical history of polycystic ovarian syndrome (PCOS), obesity (BMI 33 kg/m<sup>2</sup>) and presumed familial hypertriglyceridemia presented to our clinic after a four year history of abnormal lipid panels. In 2014 she had total cholesterol 222, triglycerides 284, LDL 100. At this time she was also recently started on combined oral contraception (COC) using norgestimate/ethinyl estradiol for symptom control of her PCOS. Over the next several years she continued to have abnormal lipid profiles including serum triglycerides of 800-1000, for which she was referred to our cardiology clinic. For one year, she was treated for her hypertriglyceridemia with Icosapent Ethyl (Vascepa) 2gm PO twice daily for one year without significant change in her triglyceride levels. However, this was discontinued due to medication cost. Given her metabolic profile and lab values it was concerning that the COC could be exacerbating her lipid profile. Her COC was discontinued without adverse effects and she was started on low dose Rosuvastatin 10 mg and fenofibrate 160 mg daily. Her lipid panel improved dramatically at 4 month follow up from total cholesterol 346 to 98; and triglycerides 966 to 141.

#### Discussion

Both oral and transdermal estrogen replacement therapies have been associated with hyperlipidemia. Oral based formulations and higher strengths of ethinyl estradiol have shown greater effect on triglyceride levels specifically. Medication discontinuation typically reverses the effects.

Those with a genetic predisposition to hypertriglyceridemia typically have only mild elevations in serum triglycerides. However marked elevations often occur when a secondary insult is also present. Other medications which adversely effect triglyceride levels are tamoxifen, HIV antiretrovirals, glucocorticoids, thiazide diuretics, beta blockers, retinoids and some atypical antipsychotics.

It is not clear if our patient also has an additional underlying genetic predisposition. The addition of low dose Rosuvastatin and fenofibrate alone would be expected to decrease serum triglycerides by at most 30-50%, whereas our patient had a striking 85% decrease suggesting a possible significant contribution from her COC pills. We predict that her lipid panel will remain normal after discontinuation of statin and fenofibrate therapies which are currently being withheld.

#### Conclusion

While COCs are known to cause mild elevation in triglyceride level, they can be associated with severe hypertriglyceridemia in some patients. Discontinuation of COCs results in normalization of TG levels. Recognizing such uncommon side effect of COCs is very important in management of patients with hypertriglyceridemia to avoid unnecessary testing, medical therapies and to decrease risk of adverse outcomes.

#### Categories

2nd year Fellow: Case

#### Program Name

The Ohio State University

Sudden cardiac arrest in an adult due to anomalous origin of the left main coronary artery from pulmonary artery Introduction (ALCAPA): A rare presentation of a rare disorder

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#### **Type of submitter**

Fellow in Training

#### **Abstract**

##### **Introduction**

Anomalous origin of left main coronary artery from pulmonary artery (ALCAPA) is rare and potentially fatal congenital cardiac anomaly that affects 1 in every 300,000 live births. It accounts for 0.24–0.46% of all congenital heart diseases.

##### **Case Discussion**

38-year-old male with no previous medical history was admitted after he sustained a sudden cardiac arrest during sexual intercourse. On arrival of paramedical staff, he was noted to be in V-Fib. He was successfully resuscitated after multiple rounds of defibrillations and CPR before return of spontaneous circulation after 30 minutes. His initial ECG showed sinus rhythm and left ventricular hypertrophy with mild repolarization changes. Troponin-T were elevated at 0.03, 0.47 and subsequently 1.34. CT head was negative for any acute intracranial abnormality. He was placed on mechanical ventilation and he underwent therapeutic hypothermia for 24 hours. He was also started on standard medical therapy with aspirin, atorvastatin and IV heparin. 2D Echocardiogram revealed severely reduced LV Systolic function with EF 20% and severe global hypo-kinesis. On day 3 of admission, he was successfully weaned off ventilator and extubated.

Coronary angiography was then performed which showed an anomalous origin of left main bifurcating into LAD and LCX from the main pulmonary artery. This was detected on selective injection into a large right coronary artery supplying collaterals to entire left coronary circulation. There was no discrete obstructive lesion in the coronaries. Coronary CT confirmed the finding of an anomalous left main artery originating from the main pulmonary trunk.

Cardiothoracic surgery team was consulted and after discussion, patient was transferred to an advanced center where he underwent open surgical repair with re-implantation of left coronary system into the aorta after placement of an interposition graft in the main Pulmonary artery to prevent compression of the coronary pathway. Post op echo showed improvement in ejection fraction up to 48% with mid and apical septum hypo-kinesis. Post op cardiac MRI was done which showed dilated left coronary artery with net antegrade flow on phase contrast imaging. Patient was later discharged home on optimized medical therapy in a stable condition.

##### **Discussion**

ALCAPA is well tolerated in fetal life but reduction in the pulmonary artery pressure after birth leads to significant myocardial ischemia in the LCA territory due to steal phenomenon. In pediatric population, it presents with congestive heart failure. Those who survive into adulthood can present with signs of myocardial infarction, heart failure, mitral regurgitation, severe pulmonary hypertension and sudden cardiac death. ACC/AHA guidelines recommend the reconstruction of dual coronary artery with re-implantation of coronary artery to aorta as preferred option of management. Patients should be followed every 3 to 5 years with Echocardiogram post-surgery.

##### **Conclusion**

ALCAPA is rare congenital coronary anomaly which can remain undiagnosed until late adulthood and can cause sudden cardiac arrest.

**Categories**

2nd year Fellow: Case

**Program Name**

Mercy St Vincent Medical Center

21

Acute Cardiogenic Shock Associated with Influenza and Bacterial Superinfection

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**Type of submitter**

Fellow in Training

**Abstract**

**Introduction:** Influenza with bacterial superinfection triggers a cascade of protective as well as deleterious infiltrating immune cells to the myocardium. Evidence shows that the extent of cardiomyocyte damage can be dependent on the balance of the immune cellular response to disease. As such, cardiac function can be rapidly compromised, leading to acute cardiogenic shock. Cardiogenic shock refers to the inadequacy of end-organ perfusion due to cardiac dysfunction. The role of influenza on the development of cardiogenic shock is rarely reported and unclear, with contributions related to sepsis and the immune response. We describe a young male with influenza and bacterial superinfection with cardiomyopathy that deteriorated to acute cardiogenic shock.

**Case Description:** A 23 year old male presented to the ER with worsening dyspnea. Days prior to arrival he complained of symptoms consistent with a viral prodrome and tested positive for influenza at an Urgent Care center. Imaging showed multilobar pneumonia and he was transferred to intensive care while started on antibiotics. He then became hemodynamically unstable, requiring endotracheal intubation and multiple vasopressors. After a 2D echocardiogram showed severely reduced ejection fraction, patient was immediately transferred to the cardiac catheterization laboratory due to concern for cardiogenic shock. A temporary mechanical left ventricular assist device and a Swan-Ganz catheter were placed and there was no evidence for coronary artery disease. Viral PCR and blood cultures were negative, however sputum cultures showed growth of multi-resistant staphylococcus aureus. His ejection fraction had recovered to normal in four days. The patient was eventually transferred to long-term acute care.

**Discussion:** There is data that shows that staphylococcus aureus exotoxin toxic shock syndrome toxin 1 (TSST-1) is associated with reversible cardiomyopathy. This case shows that with cardiogenic shock associated with sepsis, intense hemodynamic support as well as aggressive treatment of sepsis can result in favorable outcomes.

**Categories**

2nd year Fellow: Case

**Program Name**

Grandview Medical Center

## Cardiac Tamponade as Initial Presentation of Peripheral T-Cell Lymphoma

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### Type of submitter

Fellow in Training

### Abstract

#### Introduction

Cardiac tamponade as an initial presentation of lymphoma is extremely infrequent.

#### Case presentation

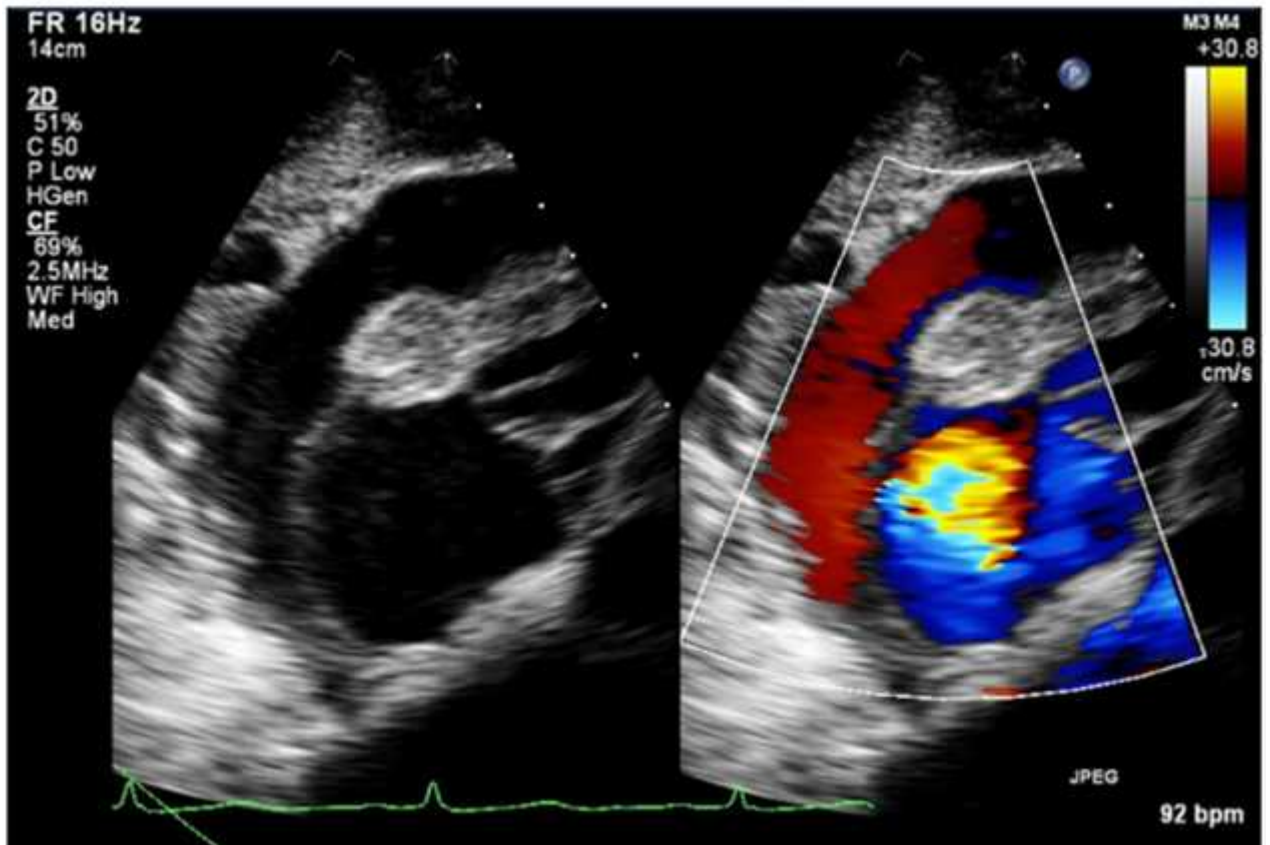
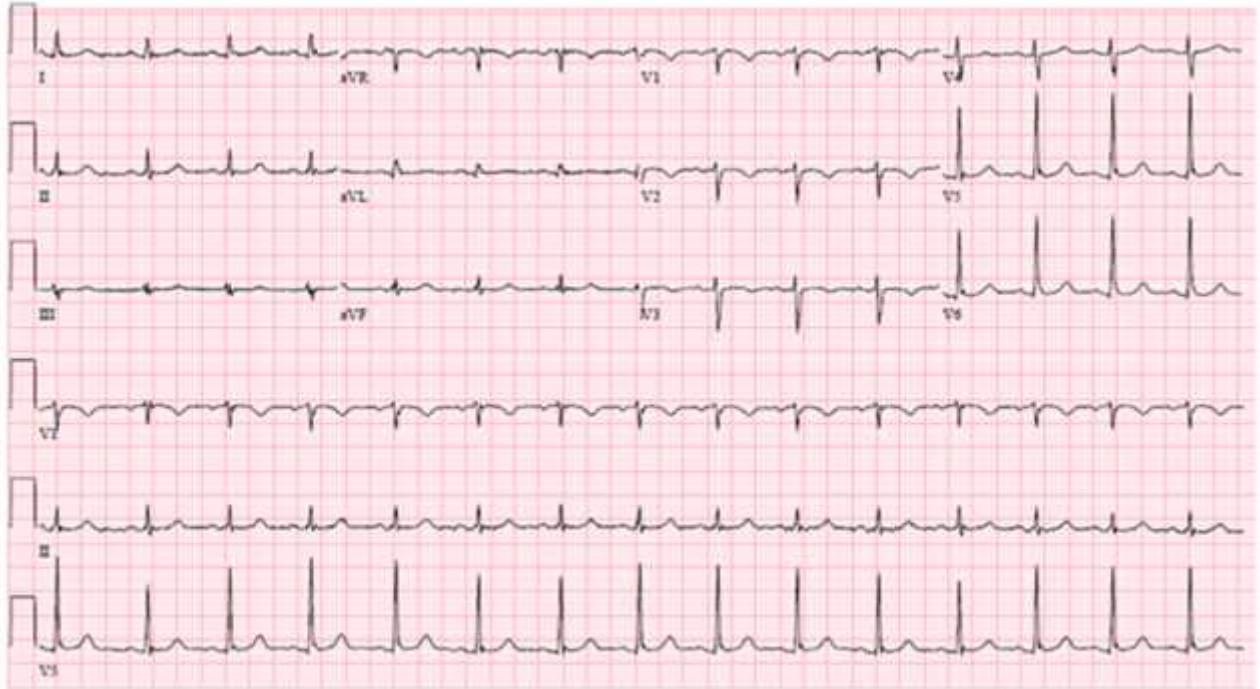
41-year-old male with history of drug and tobacco abuse who presented with a five-day history of progressive dyspnea, orthopnea, bilateral lower extremity edema, abdominal fullness and weight gain. No fever, chills or night sweats. On physical examination, blood pressure was 120/73 with pulsus paradoxus of 30mmHg, respiratory rate 18, heart rate 92 and SatO<sub>2</sub> 99% on room air. He was in moderate distress, there was jugular venous distention up to the angle of mandible, regular heart sounds, bilateral rales in the lung bases, abdominal distension with shifting dullness and bilateral pitting lower extremity edema. Pertinent laboratory studies included leukocytosis of 21.5 K/uL with 82% lymphocytes. LDH was elevated as well as aspartate aminotransferase and he tested positive for chronic hepatitis B. Hepatitis C and HIV serology was negative. Brain natriuretic peptide, troponin, renal and thyroid function tests were normal. An electrocardiogram showed low voltage in the limb leads and a chest x-ray showed cardiomegaly. An echocardiogram showed a normal left ventricular ejection fraction at 60% and a large pericardial effusion (2.8cm) with right atrial and ventricular collapse. In addition, there was a 25mm by 23mm vascular mass seen in the right ventricular AV groove. The patient then had an emergent pericardiocentesis in the setting of impending cardiac tamponade. Opening pericardial pressure was 30mmHg and 950cc of serous fluid was removed and a pericardial drain was left in place. Cytology of the fluid showed lymphocytic effusion with atypical lymphoid cells. A cardiac magnetic resonance showed a heterogeneous mass infiltrating the atrial wall and the basal right and left ventricular walls. The patient was then referred for a pericardial window since there was persistent pericardial fluid drainage. Pathology of the pericardial tissue and a bone marrow biopsy confirmed a diagnosis of Stage IV PTCL. Chemotherapy with CHOEP (Cyclophosphamide, Hydroxydaunorubicin, Oncovin, Etoposide, Prednisone) was started and he has received 3 cycles to date. His current condition is stable.

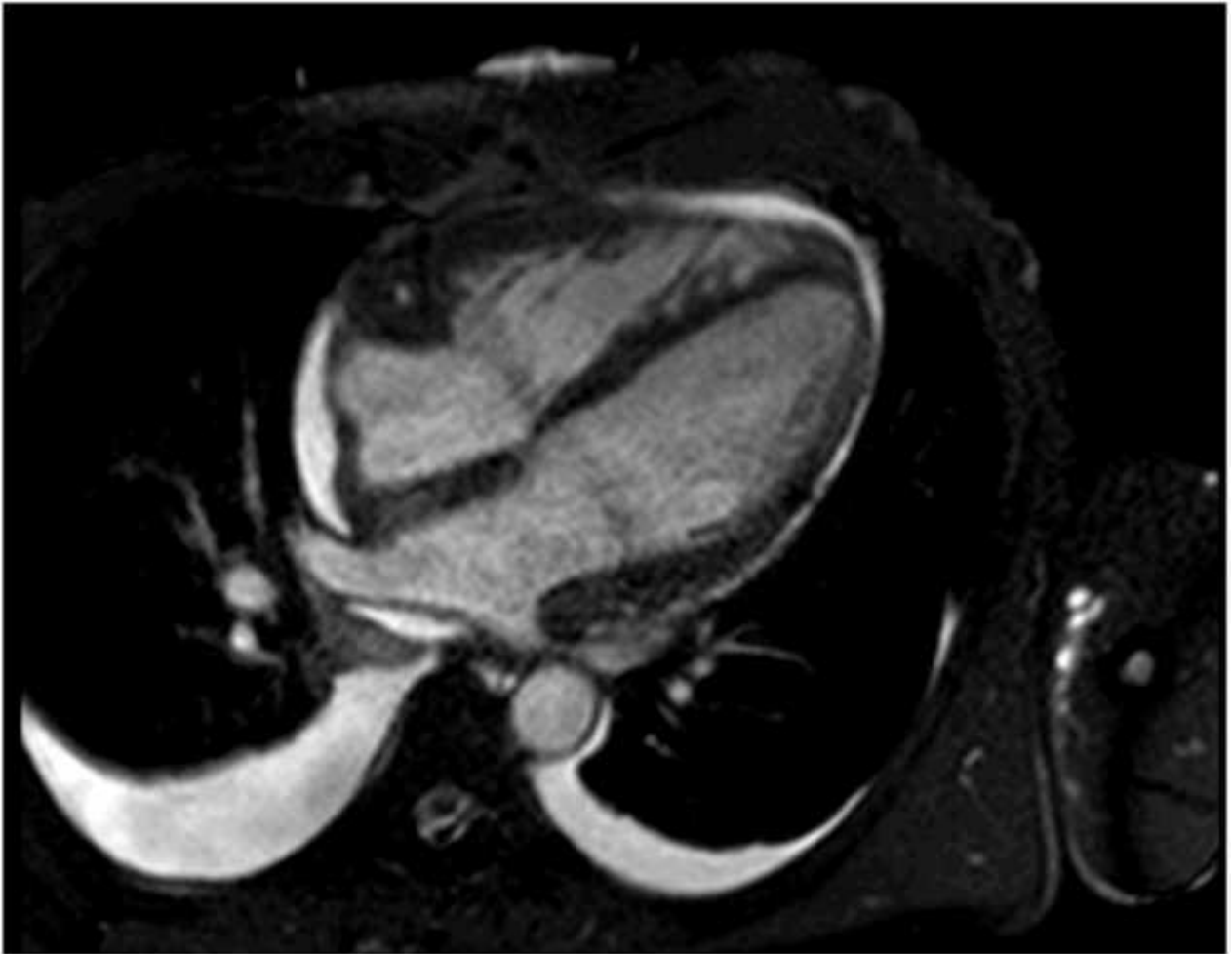
#### Discussion

Malignancy constitutes one of the most common causes of pericardial effusions leading to cardiac tamponade. While malignant tumors compose 15% of primary cardiac tumors, secondary cardiac involvement is common. Lung cancer followed by breast and esophageal cancer, melanoma, lymphoma, and leukemia are some of the most frequent metastatic tumors that invade the pericardium. About 18% of patients with non-Hodgkin lymphoma and 16% of those with non-Hodgkin lymphoma have cardiac involvement occurring at a median of 20 months after initial diagnosis. Although pericardiocentesis is an effective treatment for pericardial effusions and cardiac tamponade, fluid reaccumulation can occur in up to 60 percent of cases. In these cases, definite treatment with a surgical pericardial window should be considered to prevent recurrences.

#### Conclusion

Cardiac tamponade is a life-threatening emergency that needs emergent pericardiocentesis. Only a small percentage of patients with lymphoma have cardiac involvement. Analysis of the pericardial fluid, including cytology, echocardiography, CT and cardiac MRI can aid in the diagnosis.





**Categories**

2nd year Fellow: Case

**Program Name**

MetroHealth Medical Center

## Fluoroless His Bundle Pacemaker Implantation with Selective Capture

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Fellow in Training

**Abstract**

## Background:

Fluoroless techniques are still novel in the Electrophysiology (EP) laboratory. This case illustrates the selective His Bundle capture during pacemaker implantation without the use of fluoroscopy. It is known that radiation is not devoid of its side effects and lifetime risks of malignancy and also carries orthopedic complications from those donning lead aprons. Fluoroless techniques have been something in the making in the EP laboratory and becoming more of routine practice EP procedures.

## Case Presentation:

In this case we present a pleasant 87 year old male with past medical history including typical atrial flutter, hypertension, and coronary bypass history who presented as an elective atrial flutter ablation. Our case ties in with the sinus node dysfunction that was noted post bidirectional flutter block requiring ventricular pacing. Patient likely has underlying sick sinus node dysfunction and this manifested post flutter line. At that point patient was emergently consented for pacemaker placement and preferentially with His Bundle pacing as this is more physiologic. Patient was already on the catheterization table and had received two grams of ancef for antibiotic prophylaxis. Using the modified seldinger technique the subclavian vein was accessed through an infraclavicular approach. Two sheaths were placed over guidewires and the ventricular lead was placed into the region of the His Bundle with 3D electroanatomic guidance. Lead impedance, amplitude and pacing threshold were measured and selective capture of the His was noted with high degree of certainty. Atrial lead was then implanted and parameters described above were tested and noted to be successful. Both leads were hooked up to a pulse generator. Thresholds were once again test and the pocket was closed. During this whole process no fluoroscopy was used to confirm catheter placement.

## Conclusions:

Less is more in the use of fluoroscopy is the new topic of discussion and practice in the EP arena. And even the absence of fluoroscopy in implantation of devices is of more interest. The minimalist technique reduces many adverse side effects not only for the patient but even the operator and staff. This case highlighting successful results of His Bundle Pacemaker Implantation with high degree of selective capture. It also shows that access can be achieved with ultrasound technique removing fluoroscopy use during this time as well. Fluoroless EP procedures are the new future in regards to all patients, not only pregnant, obese or pediatric patients.

**Categories**

2nd year Fellow: Case

**Program Name**

Doctors Hospital Cardiology Fellowship

## Computed Tomographic 3-Dimensional Virtual Dissection Aiding Surgical Planning in a Rare Pediatric Case of Bicuspid Aortic Valve with Ascending Aortic Pseudoaneurysm

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### Type of submitter

Fellow in Training

### Abstract

#### Introduction/Objectives:

Aortic wall injury in bicuspid aortic valves (BAV) with aortic dilation is a rare complication. We review a case of an ascending aortic (AAo) pseudoaneurysm in a patient with BAV and mildly dilated aortic root, highlighting the benefits of 3D reconstruction techniques in pre-operative planning.

#### Case Presentation:

A 17-year-old female with history of severe idiopathic pulmonary hypertension and BAV (right/left fusion) had bilateral lung transplantation 3 years prior, complicated by systemic hypertension post-operatively. Subsequent transthoracic echocardiograms (TTE) have demonstrated slowly increasing aortic root size, within the mild range with normal valve function. During recent follow-up, she continued to be clinically well, however in addition to continued progression of root dilation, now measuring 3.8 cm (Z-score +3.7), TTE was suspicious for an aortic wall injury with para-aortic thrombus. CT confirmed a large pseudoaneurysm with thick "thrombotic shell" arising from the rightward wall of the proximal AAo with a small circular entrance (8 x 7 mm). There was compression of surrounding structures (right atrium, superior caval vein and right pulmonary artery) and inferior displacement of the right coronary artery. Surgical planning was assisted by 3D CT reconstructions utilizing both volume-rendering and more novel surface-rendering "virtual dissection" techniques (Figures). The patient underwent surgical resection of the pseudoaneurysm, with resection of the AAo due to its thin and friable appearance, and primary repair of the aortic root to proximal aortic arch. Aortic tissue was sent for pathology returning no evidence for infectious etiology. The post-surgical period was uneventful.

#### Discussion:

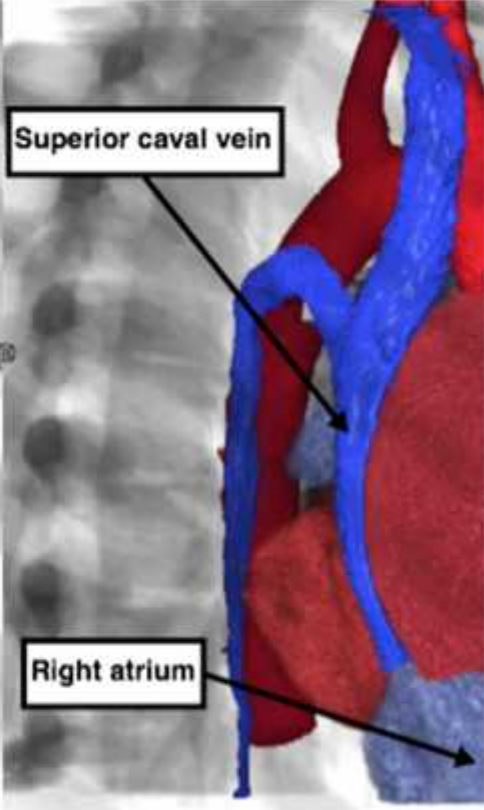
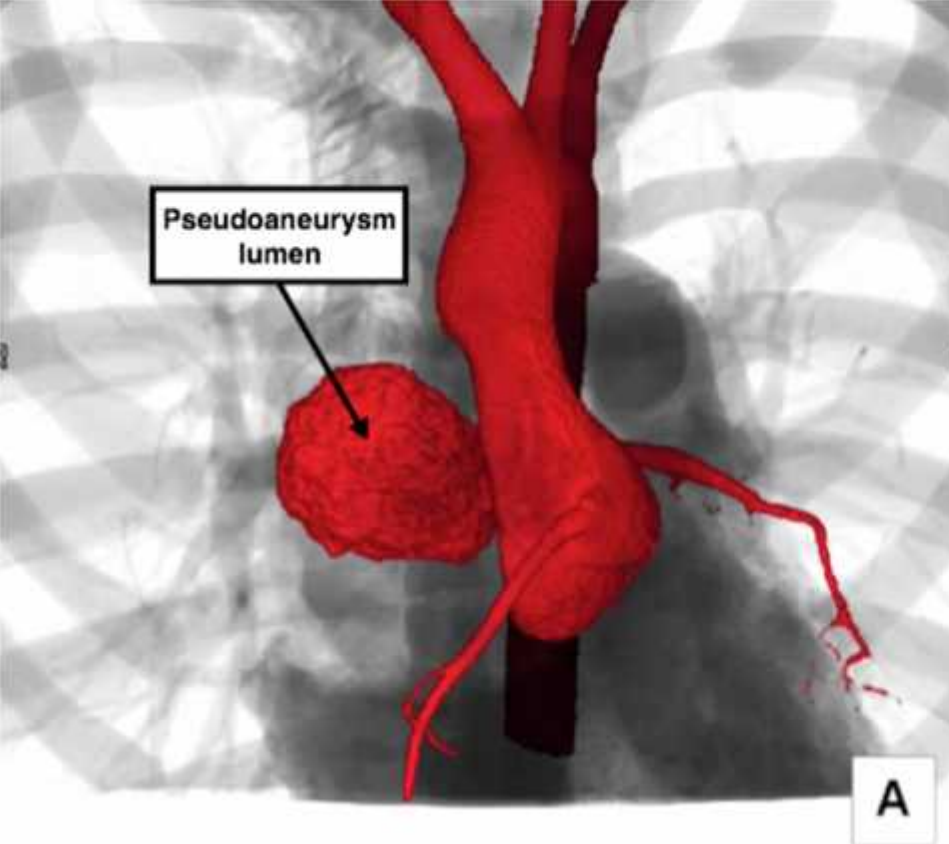
BAVs are the most common form of aortopathy. Although there is a high incidence of dilation of either the aortic root or AAo,(1) the incidence of more serious aortic wall injury (pseudoaneurysm, dissection or rupture) is extremely rare when compared to other forms of aortopathy.(2,3)

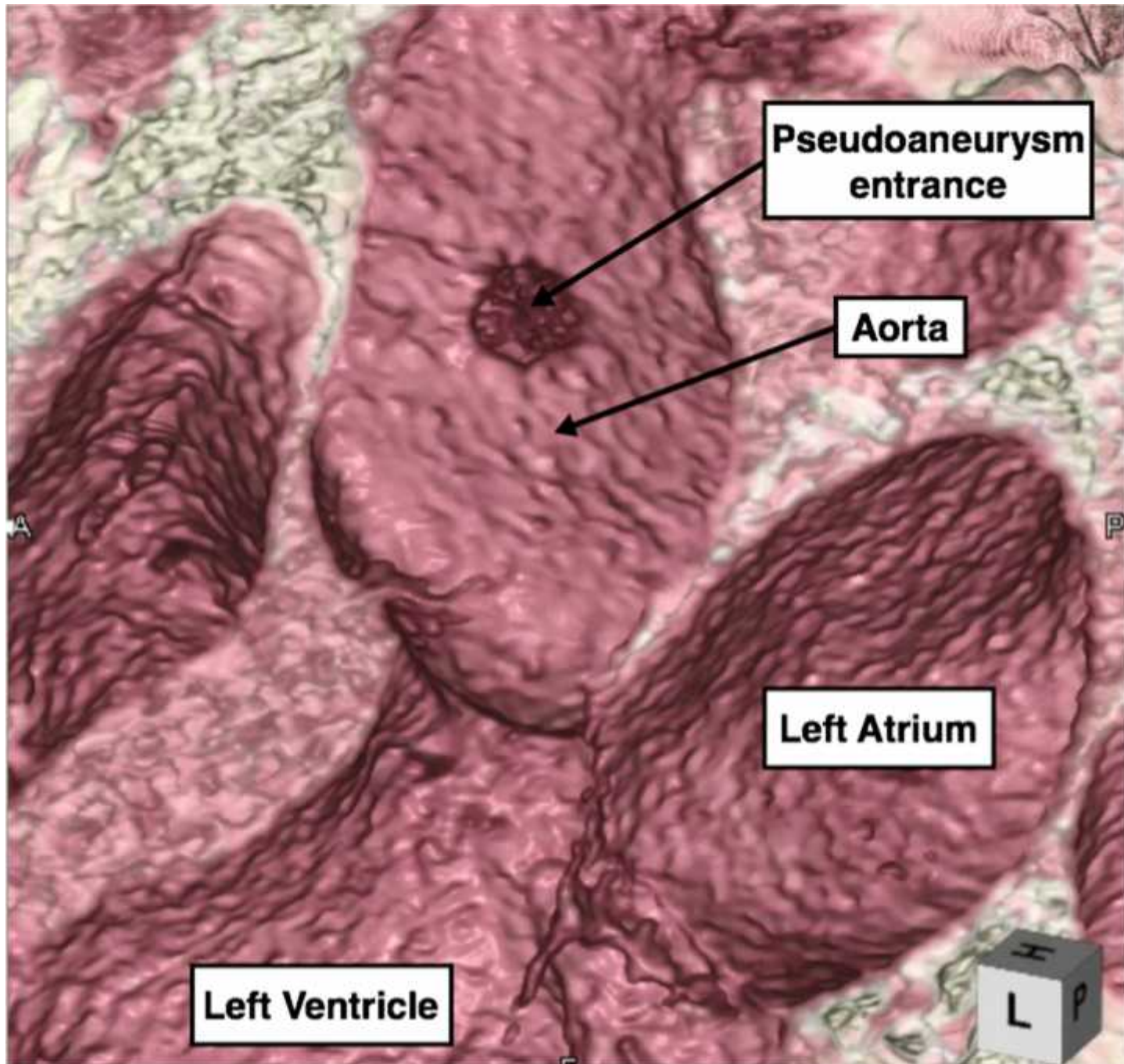
Our patient is therefore unique in that she presented in her teenage years with a large AAo pseudoaneurysm in the setting of mild aortic root dilation. We speculate that the inciting factor of her aortic wall injury is in some way related to her systemic hypertension following bilateral lung transplantation correlating with the onset and progression of aortic root dilation. Additionally, limited data links immunosuppression to acceleration of aortic dilation. Injury following aortic cannulation during her lung transplant was considered, however not consistent with the pseudoaneurysm location. Infection was considered, however with no supporting evidence.

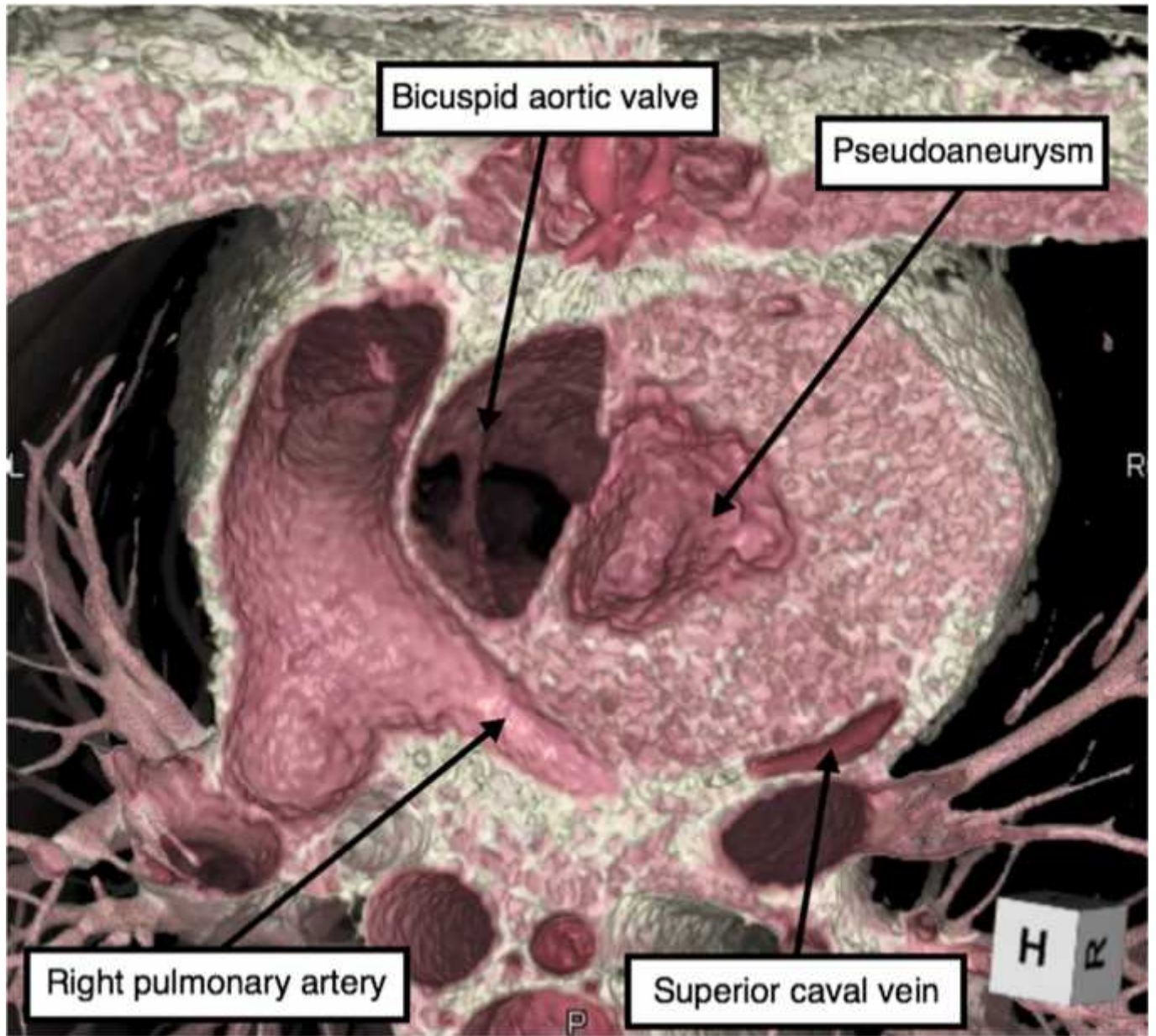
The illustrative figures in this case highlight the additional benefit of both surface-rendering and volume-rendering virtual dissection 3D CT reconstructions in understanding the relevant anatomy of the pseudoaneurysm and surrounding structures. This additional information past the standard 2D imaging left no surprises during this patient's surgical intervention.

#### Conclusion:

Aortic pseudoaneurysm is rare in pediatric patients with BAVs. Potential inciting etiology in this case include systemic hypertension and immunosuppression. The addition of 3D CT reconstruction images to the pre-operative planning left no questions going into the operating room.







**Categories**

2nd year Fellow: Case

**Program Name**

The Heart Institute, Cincinnati Children's Hospital

## Purulent Bacterial Pericarditis and Tamponade in the Setting of Acquired Immunodeficiency

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**Type of submitter**

Fellow in Training

**Abstract**

**Introduction:** Pericarditis and cardiac tamponade are commonly encountered conditions in clinical cardiology practice. While the vast majority of cases of pericarditis are due to viral, inflammatory, or malignant causes, physicians must be cognizant of rare etiologies that require unique management. We present an unusual case of purulent bacterial pericarditis and tamponade caused by *Haemophilus influenzae* in a patient with acquired hypogammaglobulinemia from prior rituximab treatment.

**Case description:** A 52 year old man with a history of lymphoma in remission presented with 3 weeks of shortness of breath, cough, and fever. His blood pressure was 102/67 mmHg, heart rate was 132 BPM, respiratory rate was 33 per minute, oxygen saturation was 98 percent, and temperature was 38.1 degrees Celsius. Physical examination was notable for a diaphoretic man with distant heart sounds, jugular venous distension, and labored breathing. Laboratory examination revealed leukocytosis, lactic acidosis, elevated creatinine and transaminases. Electrocardiogram revealed sinus tachycardia, diffuse ST segment elevations, PR segment depression, and electrical alternans. Troponin was 0.02 and ESR was 40. Bedside echocardiogram revealed a large circumferential pericardial effusion, diastolic collapse of the right ventricle, IVC dilation with loss of respiratory variation, and mitral flow respiratory variation greater than 25 percent. Left ventricular ejection fraction was 40 percent. The patient underwent pericardiocentesis and 400 ml of purulent fluid was drained. Pericardial and blood cultures were positive for *Haemophilus influenzae*. A CT of the chest revealed an empyema. Given the patient's history of rituximab use and profound infection, immunoglobulin levels were drawn and found to be undetectable. The patient received 6 weeks of intravenous antibiotics with IVIG infusions. Colchicine and aspirin were started for treatment of pericarditis. Lisinopril and metoprolol were initiated for his cardiomyopathy. Two months later, a cardiac MRI was performed, which revealed resolution of the pericardial effusion and cardiomyopathy. No evidence of constrictive pericarditis was noted, but continued pericardial enhancement was observed. Colchicine was continued for an additional 3 months. The patient made a full recovery and remains asymptomatic in follow up.

**Discussion:**

Purulent pericarditis is rare, accounting for less than 1% of cases of pericarditis. Risk factors for development of bacterial pericarditis include bacteremia, immunocompromised state, and thoracic surgery. While gram positive organisms are the most common organisms causing purulent pericarditis, infection with gram negative organisms such as *Haemophilus influenzae* remains very rare. Only 9 cases of purulent pericarditis secondary to *Haemophilus influenzae* have been reported. Our patient developed septic shock due to an acquired immunodeficiency from treatment with rituximab for lymphoma 7 years prior. Direct extension from pneumonia or empyema are frequent routes of infection into the pericardial space. Pericardiocentesis is indicated if there is a suspicion of bacterial infection as purulent pericarditis carries a 30% mortality rate. Bedside echocardiography played a critical role in the prompt diagnosis of pericardial tamponade and treatment with pericardiocentesis, resulting in an excellent patient outcome in this case.

**Conclusion:**

Early recognition and prompt treatment of purulent bacterial pericarditis is essential in order to improve patient outcomes and avoid the development of constrictive pericarditis.

**Categories**

2nd year Fellow: Case

**Program Name**

University of Cincinnati