

Unexplained Syncope: Is a Negative Electrophysiology Study Good Enough?

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Type of submitter

Fellow in Training

Abstract

Introduction:

Unexplained syncope presents an often difficult diagnostic challenge. In the presence of evidence of conduction disease on an ECG, such as with bundle branch block or sinus bradycardia, an electrophysiology study (EPS) can be performed to evaluate for cardiac arrhythmias including sinus node dysfunction and advanced AV block. However, the sensitivity and specificity of this test are low.

Case presentation:

A 73-year-old woman with a history of hypertension, presented to her PCP with recurrent episodes of near syncope. She reported sudden episodes of extreme dizziness and lightheadedness associated with diaphoresis. The episodes were not associated with change in position and occurred while standing or while sitting. They lasted a few minutes. She is otherwise active and denies exertional dyspnea, chest pain, or palpitations. Her physical exam was unremarkable. Her ECG in the PCP's office revealed NSR with an atypical LBBB with a QRS of ~150 ms. She had no prior ECGs. She was referred to the ED where her vital signs were normal. She was admitted for telemetry observation. Her blood test was unremarkable. Her echocardiogram showed a structurally normal heart with no significant valve pathology. She was seen by the electrophysiology service and felt that her symptoms could be arrhythmic in origin. However, she had a similar episode of dizziness while being monitored and telemetry didn't show any significant tachy or brady-arrhythmia. Therefore, she underwent an EPS which showed a normal sinus node recovery time, a normal HV interval of 55 ms that only minimally increased to 60 ms with the infusion of Procainamide. The AV node Wenkebach cycle length was 320 ms (188 bpm). There was no evidence of infrahisian block, AH jump, or dual AV node physiology. Immediately following the EPS an injectable loop recorder was placed to further monitor the patient in the outpatient setting. She was discharged home and presented 12 days later with a sudden episode of syncope. Loop recorder interrogation showed an episode of transient complete AV block resulting in a 10 second pause and recurrent syncope. A dual chamber pacemaker was placed.

Discussion:

Although EPS accurately delineates abnormalities in patients with fixed cardiac-conduction defects, its sensitivity in identifying transient rhythm disturbances is low. The data suggest that a negative EPS in a patient with unexplained syncope does not entirely exclude a transient brady-arrhythmia as a cause of the syncope. In fact, the EPS is poor for diagnosing both sinus node dysfunction and transient AV block. Furthermore, EPS may sometimes reveal unrelated rhythm disturbances that may mistakenly be designated as the cause of the syncope.

Conclusion:

Our case proves that a normal EPS doesn't rule out intermittent brady-arrhythmias. In cases where the syncope remains unexplained and the suspicion for a transient rhythm disorder is high, further evaluation with an event monitor or implantable loop recorder is recommended. The value of an EPS in such a clinical scenario remains questionable due to low sensitivity and specificity.

Categories

2nd year Fellow: Case

Program Name

Summa Health, NEOMED

Endometrial Stromal Sarcoma Metastasis to the Tricuspid Valve without inferior vena cava involvement: The Second Documented Case in English Literature. Case report and literature review.

Mohammed Abdulrazzaq, Brian Cothorn, Theresa Ratajczak, Umbreen Hussain, Balaji Shanmugam, Abdur Rehman, Mohey Saleh, Abdelhamed Abdelhamed

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Type of submitter

Fellow in Training

Abstract

Introduction:

endometrial stromal sarcoma (ESS) is an uncommon diagnosis, with incidence of approximately 2 diagnosis per 1,000,000 women annually. Cardiac malignancies are also uncommon, with primary tumors reported incidence of 0.02% per year. Metastatic cardiac involvement is still rare, but more frequent than primary cardiac tumors. It is estimated that metastases are 20-40 times more likely than primary tumors.

Case report:

52-year-old Caucasian female with a history of recurrent ESS. She had removal of endometrial polyp in 2007 with biopsy could not rule out LGESS, which led to robotic hysterectomy with bilateral salpingo-oophorectomy. No evidence of remaining pathology was identified. She remained well until May of 2017 when she was diagnosed with right ureteral obstruction. CT scan showing concern for lymphadenopathy versus soft tissue mass resulting in obstruction at ureterovesical junction. Cystoscopy and ureteroscopy completed with evidence of extrinsic compression. A stent was placed. Lymph node and retroperitoneal mass biopsy then completed and was consistent with metastatic endometrial stromal sarcoma. Neoplastic cells were positive for vimentin, CD10, ER, and PR, but negative for AE1/AE3, CAM 5.2, Desmin, and S-100. Whole body PET scan consistent with localized disease to right pelvis. Surgical resection was elected given these findings. A new holosystolic murmur was noted by PCP at routine visit. TTE revealed a large 2.6 x 1.8 cm mass in the right ventricle attached to the tricuspid valve with prolapse into the right atrium, suspicious for thrombus, but a primary tumor could not be ruled out. LHC revealed normal coronary arteries, a normal left ventricle, and no mitral regurgitation or gradient across the aortic valve. TEE reported a large right atrial mass likely attached to the atrial surface of the tricuspid valve. This mass was partially prolapsing through the tricuspid valve with associated turbulence and tricuspid insufficiency. It was possibly attached to the right ventricle as well. She was then referred to cardiothoracic surgery. Surgical exploration showing a tricuspid valve anterior leaflet mass extending into the right ventricle and replacing tricuspid valve leaflets with extension into annulus and papillary muscles. No evidence of vena caval involvement. The mass removed, and the tricuspid valve was replaced successfully. Biopsy revealed metastatic endometrial stromal sarcoma.

Discussion:

The combination of LGESS with cardiac metastases is very rare. In 2008 Renzulli et al. reviewed 18 cases of LGESS with cardiac involvement. All of these cases described caval metastases that extended to cardiac structures. Since that time other cases have been described, but all with IVC involvement. One case reported isolated left sided involvement. It wasn't until 2015 that Shakerian et al. added their own case of LGESS metastatic to the right heart. However, this additional case was unique in that it did not have any caval involvement. It was believed to be the first documented case in English literature. Our case demonstrates a patient with cardiac involvement of LGESS to the tricuspid valve and right atrium without caval involvement, and is believed to be only the second documented case in English literature.

Categories

2nd year Fellow: Case

Program Name

Wright State University Cardiovascular Disease Fellowship Program

Coronary Artery Fistula and Pectus Excavatum Paradox

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Type of submitter

Fellow in Training

Abstract

Summary

We present the case of a middle age man with severe pectus excavatum and a coincidental finding of a large left main coronary artery to right atrium fistula. Despite a pectus index of 4.8, he had normal hemodynamic studies. Despite a very large coronary fistula, he had normal myocardial perfusion. He was asymptomatic and without functional limitation. His pectus deformity and coronary fistula had opposing hemodynamic effects, thus protecting him from severe complications of either.

Case Report

A 35-year-old man with known atrial fibrillation, severe uncorrected congenital pectus excavatum (pectus index 4.8) and congenital left main coronary artery to right atrium fistula with a prior unsuccessful coil embolization, presented with an accidental two-story fall. Pre-operative risk assessment for trauma surgery led to a coronary angiogram and right heart catheterization.

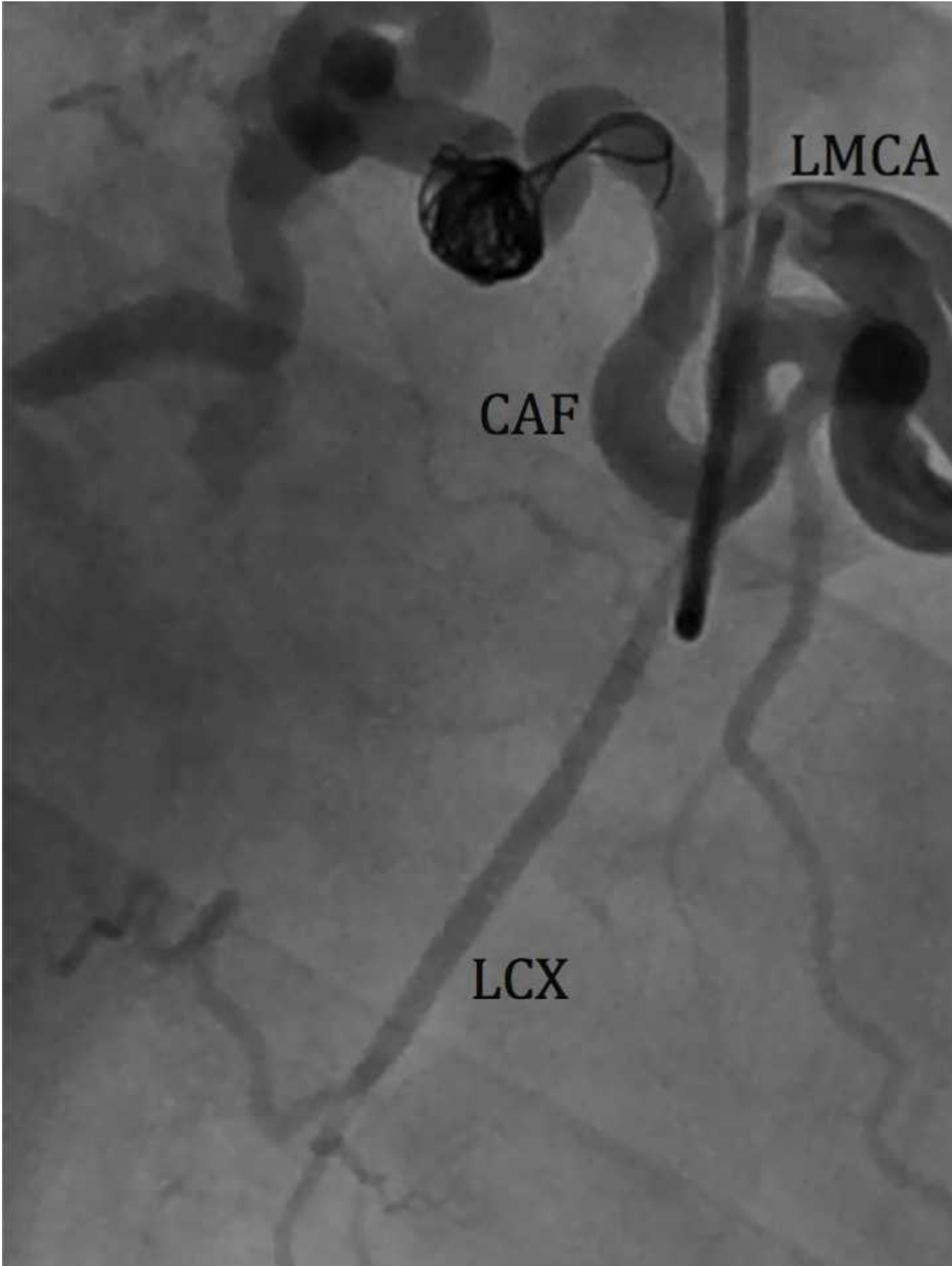
Coronary angiography showed a short left main coronary artery (LMCA) with normal bifurcation into intermediate sized left anterior descending and circumflex arteries, along with a large tortuous coronary artery fistula originating from LMCA near bifurcation and draining into the right atrium. A previously deployed coil within the fistula was also seen but did not appear to offer any meaningful impedance to the fistula flow (Figure 1). Right heart catheterization was notable for normal right ventricular and normal pulmonary artery pressures. Mixed venous oxygen saturation from inferior and superior vena cava was noted at 73% and pulmonary artery oxygen saturation at 76%, with a Qp/Qs ratio of 1.15, suggesting absence of any significant left-to-right shunt.

Discussion

Pectus excavatum, a posterior depression of the sternum and costal cartilages, is the most common congenital chest wall deformity, of likely genetic origin. Pectus Index is defined as the ratio of maximal transverse diameter to narrowest anterior posterior length of chest cavity, and is greater than 3.0 in severe cases, which results in right atrial and right ventricular compression, reduced lung volumes, decreased cardiac output and reduced exercise capacity. If identified in early years of life, such patients are deemed candidates for surgical correction before cardiopulmonary complications develop.

Large coronary artery fistulae to right heart pose a risk of myocardial ischemia via steal phenomenon, and can cause left-to-right shunt with subsequent heart failure and pulmonary hypertension. Symptoms usually develop before the age of 20. ACC and AHA guidelines recommend that coronary fistulae of large size should be treated regardless of symptoms or clinical manifestations (Class I indication).

According to the current standard of practice, surgical repair of both pectus excavatum and coronary fistula would have been indicated in our patient. Surprisingly, he was asymptomatic and had no functional limitation. It is likely that the coexistence of these two conditions resulted in a physiological and functional balance, maintaining normal right-sided hemodynamics and normal myocardial perfusion (pectus deformity providing a neutralizing effect against the development of right sided overload and preventing the steal phenomenon (Figure 2 and 3). These findings led to a carefully considered decision to allow both congenital deformities to be left untreated at this time.



LMCA

CAF

LCX

Figure 1

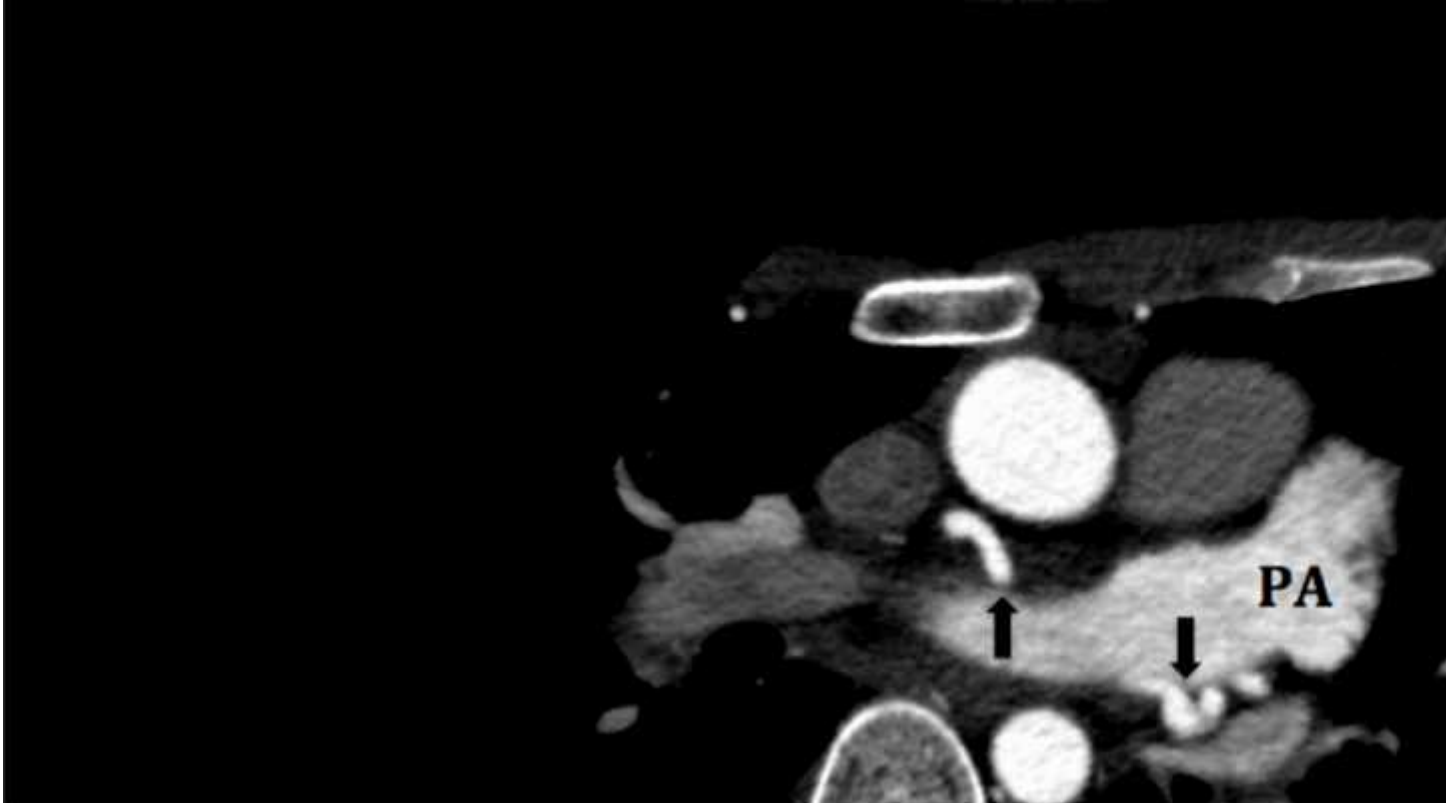
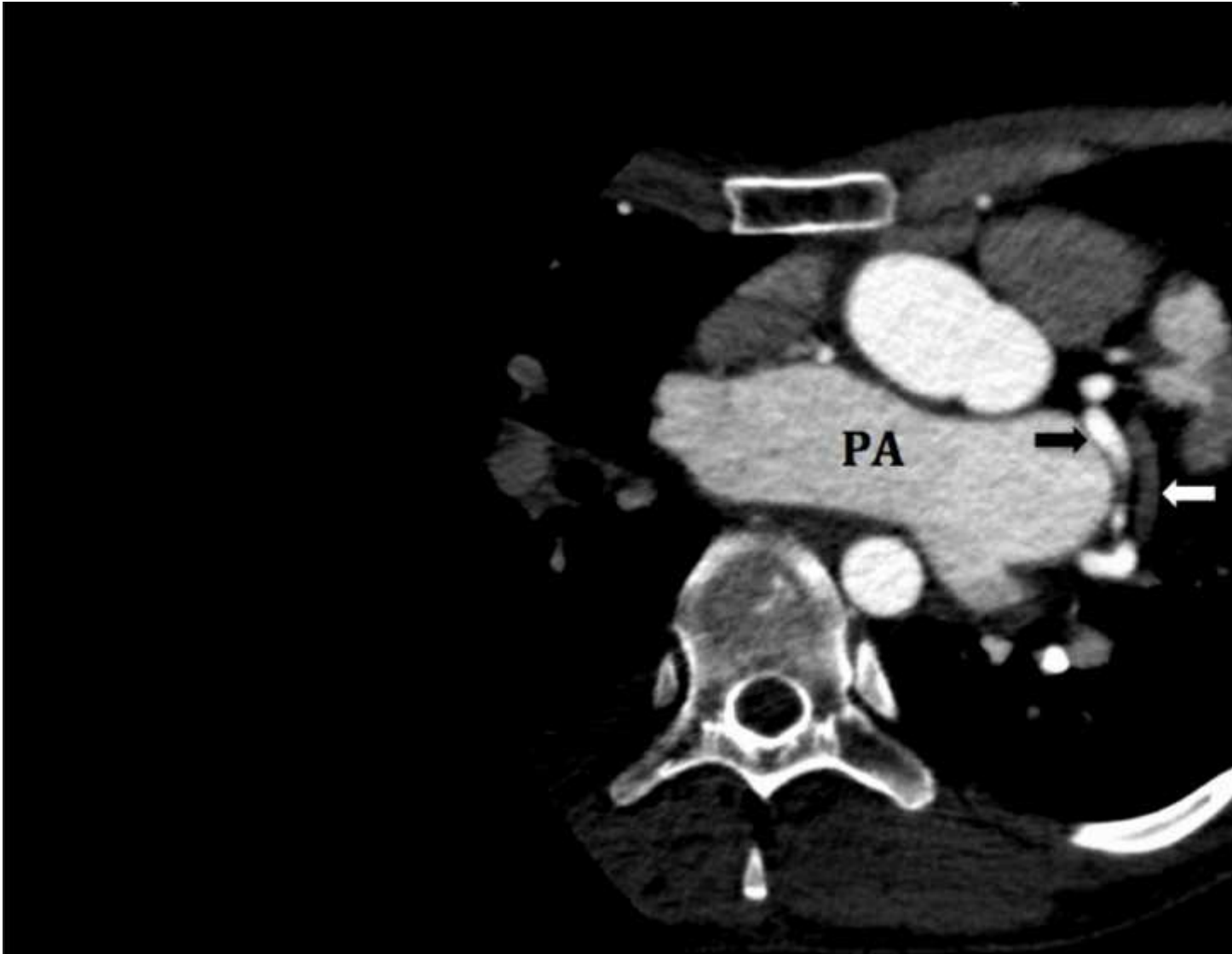


Figure 2

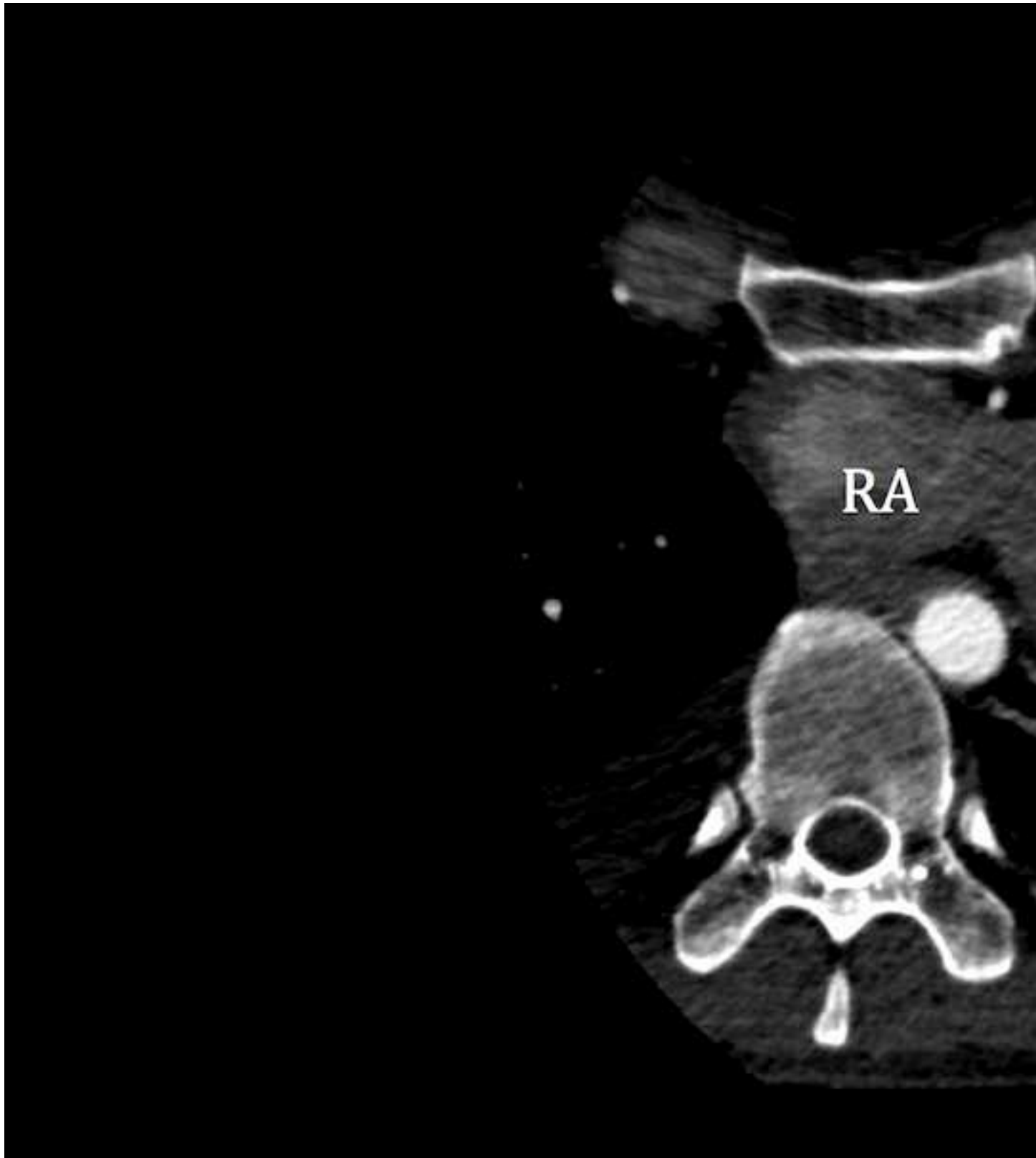


Figure 3

Categories

2nd year Fellow: Case

Program Name

University of Cincinnati Cardiovascular Fellowship Program

Mobile pedunculated ventricular thrombi in young male with non-ischemic cardiomyopathy

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Type of submitter

Fellow in Training

Abstract

Objective: We present a case of mobile pedunculated ventricular thrombi in a young male with non-ischemic cardiomyopathy that subsequently embolized requiring emergent thrombectomy.

Case: A 22-year-old male with a history of non-ischemic cardiomyopathy since early childhood, nonadherent to medications for 3 years and recent methamphetamine abuse presented with cough and chest discomfort. He was initially diagnosed with streptococcus pneumonia. He was discharged on antibiotics but returned within 14 days with worsening dyspnea, lower extremity edema, and abdominal distention. CT Chest showed dilated right and left ventricles, likely thrombus in left ventricle, right lower lobe pneumonia and abdominal ascites. A chest tube was placed for right empyema. Surface echocardiogram demonstrate ejection fraction of 10-15% and biventricular masses. At his last evaluation 3 years ago, his ejection fraction was 45-55%. Patient was transferred to our institution for further management. On arrival, he was started on milrinone for cardiogenic shock and continued on broad spectrum antibiotics for septic shock. A transthoracic echocardiogram revealed severely depressed ejection fraction as well as biventricular masses. In the left ventricle, there were two pedunculated masses. The first was 1.8 x 1.9 cm with a 2.4 cm stalk attached apically. The second was 1.2 x 0.9 cm mass with a 0.2 cm stalk attached mid-inferolaterally. In the right ventricle, there was a large apical mass 3.5cm x 1.7 cm. No valvular vegetations were seen. Patient was started on heparin infusion for thrombi. Given the pedunculated nature of the clots, there was a high concern for embolization. He was evaluated by cardiothoracic surgery and was to have surgical thrombectomy. However, a pre-procedure transesophageal echocardiogram revealed that the largest left ventricular thrombus was no longer present and the procedure was canceled. At that time, his exam was notable for cold and pulseless left lower extremity. CT angiogram brain, chest, abdomen, and pelvis with run off was obtained. He had no neurologic defects. He had pulmonary emboli and small renal infarctions bilaterally. The right anterior tibial artery and left tibial peroneal artery were occluded. Vascular surgery took the patient urgently to the OR. Exploration of the right anterior tibial artery revealed a chronic occlusion. He underwent a left popliteal thrombectomy. Surgical pathology demonstrated fragments of organizing fibrin/blood clot with clusters of neutrophils. For remainder of his eighteen-day hospital course, he underwent inotrope-assisted diuresis, his heart failure regimen was optimized, his chest tube was removed, and he was transitioned to warfarin for anticoagulation. He was discharged to a skilled nursing facility, dependent on milrinone, to complete a six-week course of antibiotics.

Discussion/Conclusion: Left ventricular thrombus can accompany severely depressed ejection fraction. Mural, flat, and immobile masses have a low risk of embolism. The risk of embolization approaches 60-80% for pedunculated thrombus. Although there is no established protocol for treatment of these thrombi, case reports and small case series generally recommend surgical thrombectomy. This case offers excellent visualization of pedunculated thrombi in a young patient with non-ischemic cardiomyopathy who unfortunately had subsequent embolization before surgical thrombectomy could be performed.

Categories

2nd year Fellow: Case

Program Name

The Ohio State University Cardiology Fellowship

The novel use of the Gore Viabahn VBX Balloon-Expandable Endoprosthesis in the congenital heart disease population: A case series

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Type of submitter

Fellow in Training

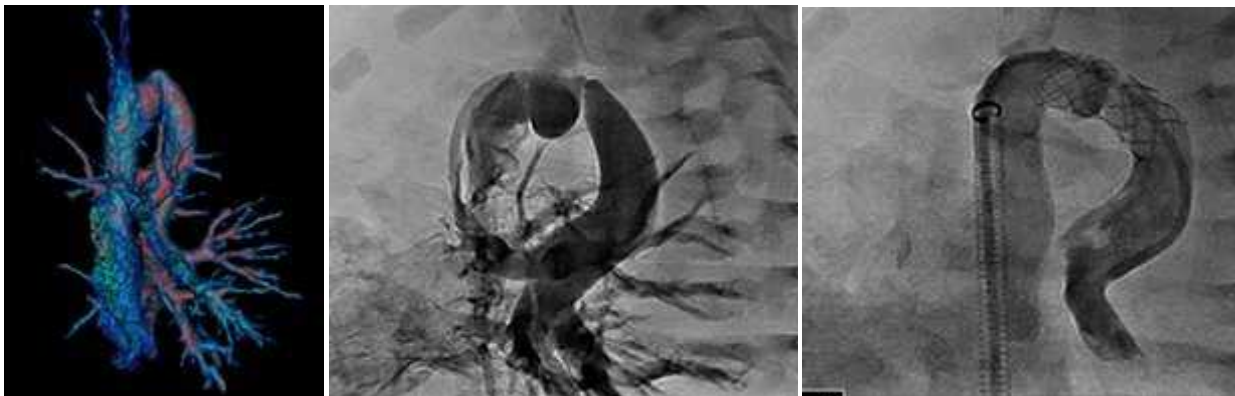
Abstract

Introduction: The purpose of this study is to report the novel use of the Gore Viabahn VBX Balloon-Expandable Endoprosthesis (VBX stent graft); (W.L. Gore & Associates, Flagstaff, AZ, USA) in three patients with congenital heart disease (CHD). The VBX stent graft utilizes optimal ring spacing and orientation, along with a thin fluoropolymer/elastomer film and percutaneous balloon delivery system that is FDA approved to treat peripheral vascular lesions in adults.



Case Presentation: Three patients with CHD had vascular lesions treated using the VBX stent graft in this case series.

Case 1: A 1 day old male with heterotaxy syndrome, dextrocardia and LV dominant atrioventricular septal defect (AVSD), pulmonary atresia with major aortopulmonary collateral arteries (MAPCAs), bilateral superior vena cavae (SVC) and obstructed supracardiac total anomalous pulmonary venous return (TAPVR) to the left sided SVC via a discretely stenotic vertical vein was brought to the interventional catheterization suite for palliative treatment. The vertical vein was stented with a 6mm x 19mm VBX stent graft, decreasing the mean gradient through the vertical vein from 20mmhg to 4mmhg.



Case 2: A 30 year old male with a history of ventricular septal defect (VSD), Type B interrupted aortic arch (IAA), and aberrant right subclavian artery (RSCA) underwent neonatal VSD patch and aortic arch repair with a 10mm Gore-Tex interposition graft and re-implantation of RSCA. Subsequently, he underwent Dacron patch repair of recurrent aortic arch stenosis and placement of a 14mm Hemashield Dacron jumpgraft from the ascending aorta to thoracic descending aorta. Both of these grafts became severely stenotic, resulting in severe recoarctation, upper extremity hypertension and lower extremity claudication. He underwent successful transcatheter reconstruction of both the interposition graft and the jumpgraft using a total of four VBX stents grafts. Initial pressure gradient of 53mmhg between the ascending aorta and distal thoracic descending aorta was reduced to 5mmhg.

Case 3: A 29 year old female with heterotaxy syndrome, RV dominant AVSD, double outlet right ventricle (DORV), pulmonary stenosis (PS), interrupted inferior vena cava (IVC), and bilateral SVC was status post Kawashima procedure and subsequent extracardiac conduit Fontan. As a result of her anatomy, she developed diffuse left-sided pulmonary arteriovenous malformations (AVMs) requiring a left axillary arteriovenous fistula. She subsequently developed a massive aneurysm (103x56mm) near the fistula, dilated LSVC-LPA junction and severely dilated aortic root. The aneurysm was excluded using a 13x50mm Gore Viabahn self-expanding endoprosthesis, and two 9x89mm Viabahn VBX stent grafts with no residual arterial or venous flow into the aneurysm following stent placement.

Discussion: Due to its significant flexibility and adequate radial strength, the Viabahn VBX stent graft has been utilized to treat complex peripheral vascular lesions in adults. In this case series, three patients ranging from a newborn to adult, with severe forms of congenital heart disease had complex vascular lesions successfully treated with the VBX stent graft.

Conclusion: This case series demonstrates the successful application of an advanced stent technology to help palliate and correct complex vascular lesions in patients with underlying congenital heart disease.

Categories

2nd year Fellow: Case

Program Name

Nationwide Children's Hospital

"Two Faced" A Case of Cor Triatriatum Sinister and Dexter

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Type of submitter

Fellow in Training

Abstract**Introduction:**

Cor triatriatum is rare congenital heart defect with an incidence of approximately 0.1% of all congenital heart disease. It is classically defined as an atrium being divided by a persistent thin membrane. The presentation of this finding can vary widely, from an asymptomatic adult patient to a neonate with severe cyanosis or low cardiac output state. We present two such cases revealing the extremes of these presentations.

Case Presentation:

The first case is a 15 year old who presented to the emergency room with a panic attack. A chest x-ray was obtained which revealed a well circumscribed mass lesion over the posterior heart border (Image 1). A chest CT was performed which was concerning for a left atrial appendage aneurysm (Image 2). He was transferred to our facility and diagnosed with cor triatriatum sinister with obstruction (Image 3). He underwent repair with resection of the membrane. His hospital course was complicated by pericardial effusion and atrial fibrillation requiring cardioversion and atenolol. He has been well and remains asymptomatic.

The second case is a newborn who presented with cyanosis. At 17 hours of life, she was found to be dusky with oxygen saturations in the 80s on 100% oxygen. She was administered prostaglandin and transferred to our facility. An echocardiogram demonstrated cor triatriatum dexter (Images 3 and 4). She was slowly weaned off of oxygen and has since been stable on room air. She has remained asymptomatic and we continue to monitor her for signs of cyanosis and restriction of the membrane.

Discussion:

Cor triatriatum was first described in 1868 by Church. In cor triatriatum sinister, failure of fusion between the common pulmonary vein and the left atrium is the proposed embryological etiology. Cor triatum dexter is a much rarer finding and is due to a failure of regression of the right valve of the embryological sinus venosus.

The clinical presentation depends on the size of the communication in the membrane between the venous chamber and the atrial chamber. If the communication is restrictive, the patient can present with cyanosis or in low cardiac output state. If the communication is not restrictive, the patient can remain asymptomatic. Due to the asymptomatic patient, there are likely a number of undiagnosed cor triatriatum cases.

Conclusion:

Cor Triatriatum is a rare diagnosis with a myriad of presentations. Therefore, a high degree of suspicion is required to include in one's differential. These two cases illustrate the difference of presentation based on the location and restrictiveness of the membrane.



Image 1: Lateral x-ray revealing aneurysmal tissue of the left atrium

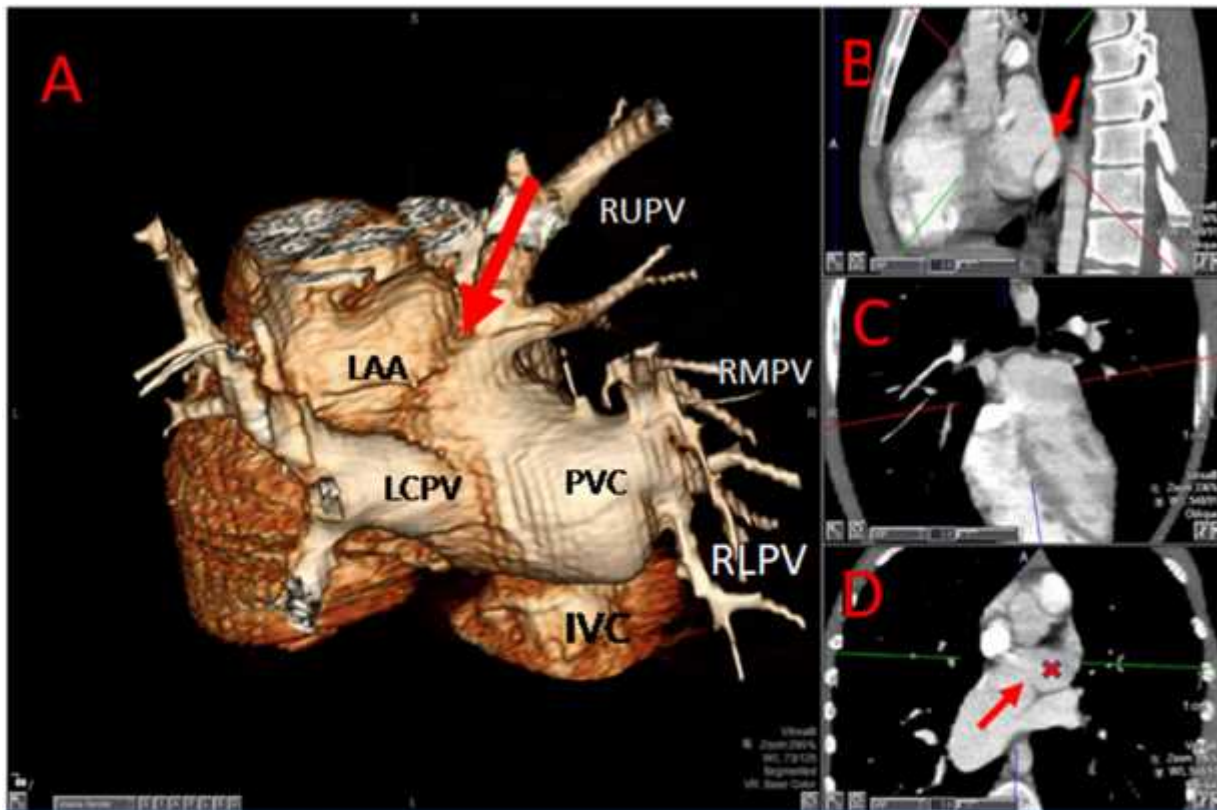


Image 2: CT scan A) LAA= Left atrial appendage, RUPV= Right upper pulmonary vein, RMPV= Right middle pulmonary vein, RLPV= Right lower pulmonary vein, PVC= Pulmonary vein confluence, LCPV= Left common pulmonary vein, IVC= Inferior Vena Cava; 3D reconstruction showing the restriction caused by the membrane between the pulmonary venous confluence and left atrium B) Lateral projection with the arrow pointing to the membrane C) Axial projection D) Coronal projection with the arrow pointing to the membrane and the X pointed to the confluence

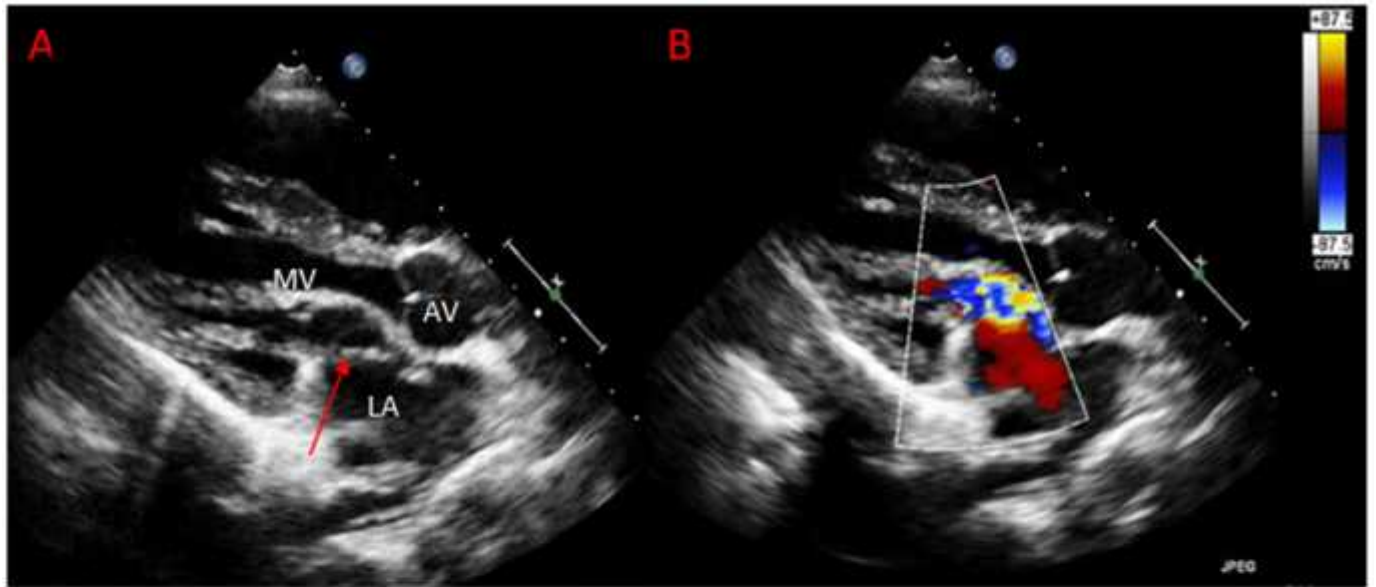


Image 3: A) Parasternal long axis(PSLX); MV= Mitral Valve, LA= Left Atrium, AV= Aortic Valve, Arrow= Membrane; Dilated Left atrium with a redundant membrane within the atrium B) Color over the mitral valve, there is aliasing through the membrane (Peak velocity of 2.1 m/s, PG 18 mm Hg and a mean of 10 mm Hg)

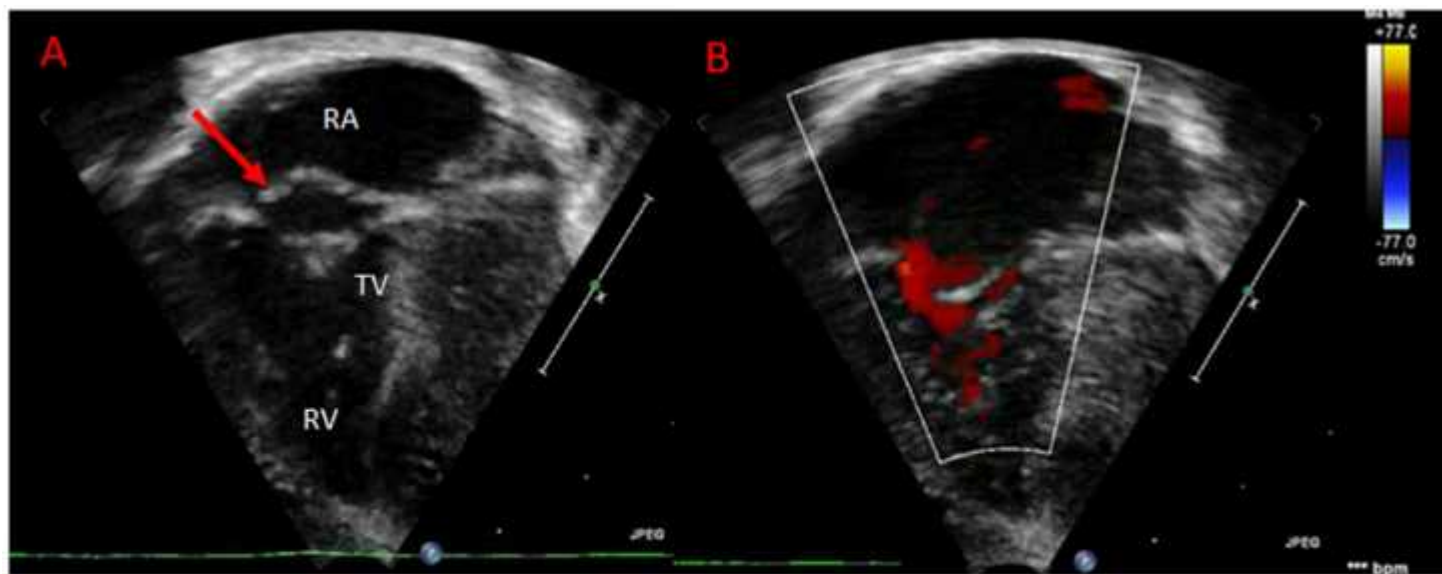


Image 5: A) Apical Four Chamber (A4CH) RA= Right atrium, TV= Tricuspid Valve, RV= Right Ventricle, Arrow= Membrane; There is a dilated right atrium with a redundant membrane above the tricuspid valve B) A4CH with color over the tricuspid valve there are two separate jets of flow through the membrane

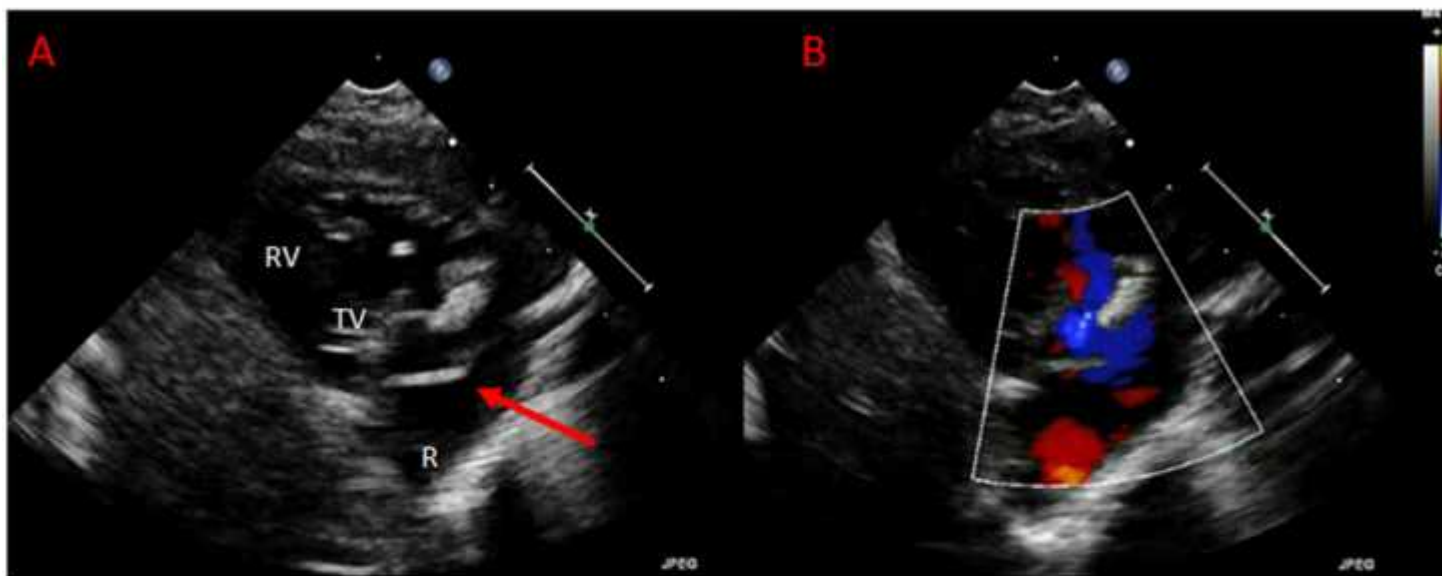


Image 4: A) (PSLX) RA= Right atrium, TV= Tricuspid Valve, RV= Right Ventricle, Arrow= Membrane; There is a dilated right atrium with a redundant membrane above the tricuspid valve B) PSLX with color over the tricuspid valve there is flow through an opening in the membrane

Categories

2nd year Fellow: Case

Program Name

Nationwide Children's Hospital

Isolated Right Ventricular Thrombus in Severe Ischemic Cardiomyopathy

Vishal Dahya, Navdeep Tandon, Justin Dunn, Mark Iler

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Type of submitter

Fellow in Training

Abstract

Intracardiac thrombus occurs most frequently in the setting of acute myocardial infarction (MI), healed infarction, and ventricular aneurysm following an acute MI¹. The first study to emphasize mural thrombi of the heart done by Garvin in 1941 found left ventricular thrombi were significantly more common than right ventricular thrombi² and a subsequent study done by Jordan et al. found left sided thrombi to be 10 times more common than right sided³ ones. Similar studies of intra-cardiac thrombus have also found that thrombi confined only to the right ventricle are a very rare complication of healed prior MI⁴⁻⁵. This case describes a patient who presented with severe ischemic cardiomyopathy and was found to have an isolated right ventricular thrombus secondary to apical RV akinesis from a prior infarction. The clinical utility of cardiac MRI was also showcased in this case study as the thrombus was diagnosed through signal characteristics.

Case History:

A 75 year old male with no significant past medical history other than tobacco abuse presented to the emergency department with one month of progressively worsening dyspnea and swelling in his abdomen and lower extremities. On physical examination, the patient was found to have significant JVD and rales throughout the lung fields bilaterally. Cardiovascular exam revealed a regular rate and rhythm with an S3 gallop. There were no audible murmurs. Abdominal exam revealed significant distention with associated anasarca and 3+ lower extremity edema. Echocardiogram was performed which revealed a severely decreased systolic function with an estimated ejection fraction of 28% with severe diffuse hypokinesis. The right ventricle was noted to have moderately decreased systolic function with a large echogenic mass 1.6 x 2.1 cm in size concerning for thrombus vs. tumor (Figure 1). Due to this finding, the patient was started on a heparin drip and underwent a CT Chest Angiogram which revealed small bilateral pulmonary emboli seen in distal branches. Transesophageal echocardiogram was then performed for further characterization of the mass which found it to be pedunculated below the right ventricular outflow tract with evidence of 2+ mitral regurgitation (Figure 2). Heart Catheterization revealed severe multi-vessel coronary artery disease with chronic total occlusions of the proximal Left Anterior Descending (LAD), Left Circumflex (LCx), and Right Coronary Artery (RCA) (Figures 3-4). All of the distal territories were supplied by collateral arteries. Patient was evaluated for coronary bypass surgery and removal of right ventricle mass and the decision was made to evaluate for myocardial viability and further characterize the mass with a Cardiac MRI. CMRI revealed a moderately dilated left ventricle with severe systolic dysfunction and EF of 26% and a moderately dilated right ventricle with severe systolic dysfunction. In the akinetic apical portion of the right ventricle there was lobulated mobile mass with signal characteristics were consistent with thrombus (Figure 5-6). Late gadolinium enhancement imaging showed total scar burden of over 40% with LAD and RCA territories appearing mostly nonviable with only LCx territory being viable. Patient was medically optimized and discharged on full anticoagulation therapy with Apixaban.

Categories

2nd year Fellow: Case

Program Name

Summa Health

An energizing etiology of abnormal ST elevations: An unusual case of pediatric ingestion

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Type of submitter

Fellow in Training

Abstract

Case presentation: An 8 year old girl with developmental delay presented to the emergency department for acute onset of stabbing, intermittent back and substernal chest pain. She had a negative troponin, a BUN of 28, and a creatinine of 0.64 mg/dL with otherwise normal labs. Initial chest x-ray at outside hospital was interpreted as normal. An electrocardiogram (ECG) was markedly abnormal with ST segment elevations and depressions raising concern for myocarditis/cardiomyopathy. She was transferred to our facility for further evaluation. At our hospital, she had intermittent, non-reproducible mid to lower sternal chest pain. Her cardiac exam was normal, but she had a palpable stool burden. An ECG was repeated with similar findings to the previous one (**Figure 1**). Abdominal and chest x-rays demonstrated a round radiolucent object in the lower esophagus concerning for a button battery. She underwent successful endoscopic retrieval and the foreign body was identified as a corroded button battery. The anesthesiologist reported that intraoperative telemetry tracings changed as the battery was removed. A post-operative echocardiogram was normal. A post-operative ECG was normal with resolution of ST changes and T-wave abnormalities (**Figure 2**). Further ECGs during admission remained normal.

Discussion: Only a few case reports that exist in the literature discuss the effects of battery ingestions on ECGs tracings. One prior case demonstrated ST changes on ECG concerning for possible myocardial infarction. All three published cases occurred in adults who ingested cylindrical batteries. This is the first reported case, to our knowledge, of pediatric button battery ingestion leading to significant ST changes on ECG, which resolved with battery removal.

Conclusion: Pediatric button battery ingestion is an emergency. Knowledge of the associated ECG changes in these cases is essential to prevent healthcare providers from delaying critical treatment of battery ingestion for further cardiac workup.

Figure 1. Initial ECG: Sinus rhythm with ST elevations in leads I, aVL, V1, and V2, and ST depression in the inferior leads

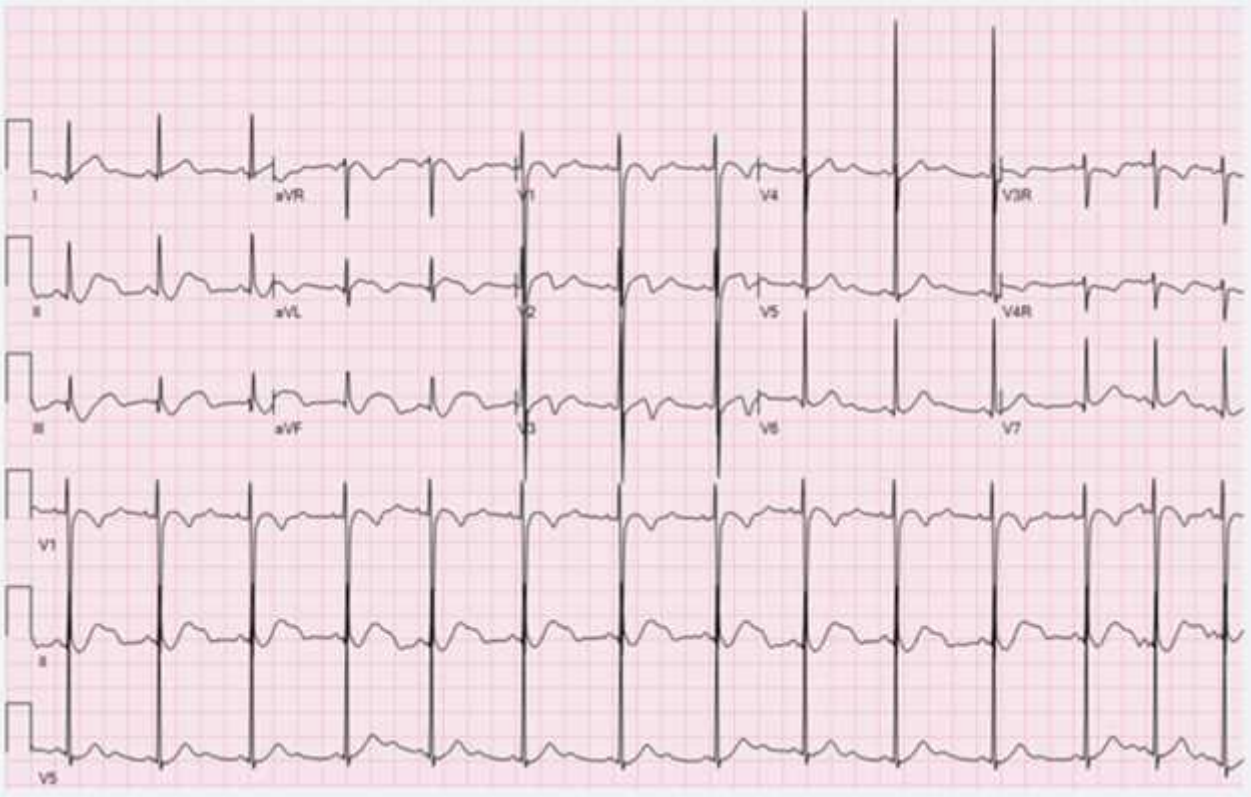
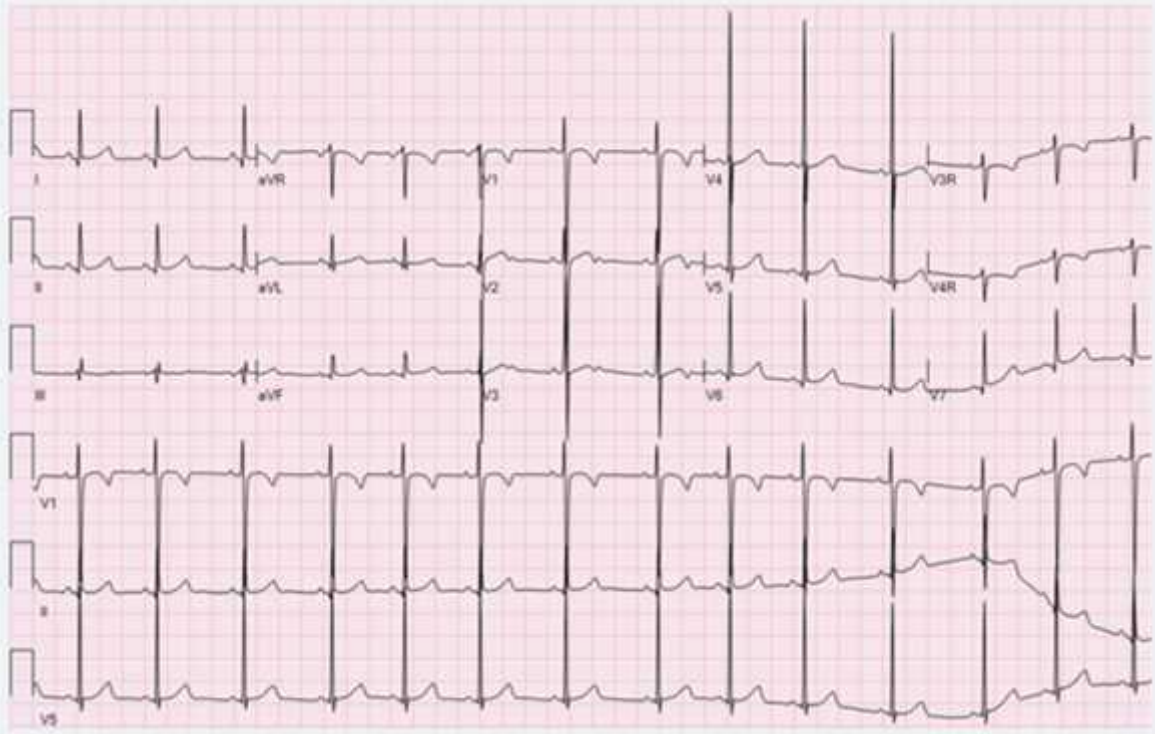


Figure 2. ECG after button battery removal: normal sinus rhythm with normal ST segments and T waves



Categories

2nd year Fellow: Case

Program Name

Pediatric Cardiology Fellowship, Nationwide Children's Hospital

Myopericarditis Leading to Diagnosis of Multiple Myeloma and Cardiac Amyloidosis

Beth Foreman, Jim Liu

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Type of submitter

Fellow in Training

Abstract

Introduction:

Amyloid light-chain (AL amyloidosis) is a systemic disorder characterized by deposition of misfolded immunoglobulin light-chain proteins, most commonly in kidneys and heart, leading to organ dysfunction. Clinical evidence of cardiac involvement occurs in ~50% of AL amyloidosis cases; however, cardiac amyloidosis can be diagnostically challenging. Increased recognition of AL amyloidosis by cardiologists may allow for earlier treatment and improved outcomes.

Case:

A 40-year-old man with no past medical history presented with fevers and pleuritic chest pain. Labs revealed leukocytosis, elevated ESR/CRP, and troponin elevation to 59 ng/mL. ECG showed diffuse concave ST-elevations with PR-depressions consistent with acute myopericarditis. Viral titers and autoimmune work-up were negative. He was started on steroids and colchicine. Echocardiogram revealed low-normal LV systolic function (EF 50%), concentric LV wall thickening of 1.4 cm, biatrial enlargement, and small pericardial effusion. Cardiac MRI (CMR) showed prominent diffuse subendocardial late gadolinium enhancement and elevated myocardial interstitial volume expansion concerning for cardiac amyloidosis. Further lab work revealed persistent mild troponin elevation, NT-pro BNP > 3000 pg/mL, renal failure and anemia, prompting further work-up for multiple myeloma. Bone marrow biopsy demonstrated IgG-kappa multiple myeloma and Congo red staining showed apple-green birefringence under polarized light, confirming amyloid deposition. Chemotherapy was initiated (bortezomib, lenalidomide, dexamethasone) for multiple myeloma and AL amyloidosis with cardiac, renal, and bone marrow involvement.

Discussion:

AL amyloidosis has an incidence of ~10.5 cases per million person-years in the US with ~10% of patients having overt multiple myeloma. Untreated, the median survival with cardiac AL amyloidosis is < 6 months once heart-failure symptoms begin. Most common manifesting symptoms are heart-failure followed by syncope. Increased wall thickness on echocardiogram in the absence of hypertension may be the first red flag for diagnosis. Other characteristic echo findings include biatrial enlargement, diastolic dysfunction often in restrictive pattern, and characteristic LV strain pattern with preservation of apex (bull's eye appearance). CMR should be considered when suspicion is high; subendocardial late gadolinium enhancement, abnormal T1 signal, and increased myocardial extracellular volume expansion are characteristic findings. Diagnosis must be confirmed by tissue biopsy detecting amyloid deposits. Endomyocardial biopsy conclusively identifies cardiac amyloidosis; however, abdominal fat pad or bone marrow are less invasive and safer approaches. Cardiac biomarkers can be used to predict prognosis in AL amyloidosis; detectable troponin and elevated BNP portend an adverse prognosis.

Conclusion:

Our case is rare as our patient presented with myopericarditis that led to diagnosing cardiac AL amyloidosis. In review of literature, only one case reported acute myocarditis associated with cardiac amyloidosis. The Amyloidosis Research Consortium estimates that up to one-third of patients visit five physicians before a diagnosis is reached. Prolonged diagnosis time is a frequent factor in poor outcomes, highlighting the need for increased physician awareness. New imaging techniques such as echo with strain imaging and CMR offer promise for early detection of cardiac amyloidosis. Cardiologists must have a high index of suspicion when reviewing non-invasive testing, especially when the diagnosis of amyloidosis has not otherwise been considered.

Categories

2nd year Fellow: Case

Program Name

Ohio State University

Shared Decision Making in an Athlete At-Risk for Sudden Death

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Type of submitter

Fellow in Training

Abstract

Introduction:

Sudden cardiac death in young people is a tragic diagnosis with multiple etiologies. Patients may be identified early when presenting with exertional syncope. Those who are at increased risk of sudden cardiac death have historically been restricted from sports participation to varying degrees given the perceived risk. Recent studies have demonstrated that participation in sports by at-risk athletes who are non-adherent to sports participation restrictions is safer than previously thought. Considering the importance of sports on the quality of life for our patients, in addition to these recent findings, the use of shared decision making is encouraged. This approach involves educating a patient about the risks of participation in certain activities, listening to their priorities, and coming to a joint decision and plan for them individually.

Case Presentation

We present a 21 year old female collegiate soccer player who collapsed without warning while sprinting at a team practice. She was cyanotic and unresponsive for one minute before awakening without recollection of the event. She was taken to a community hospital where she was reported to have a normal echocardiogram and a normal ECG (later review showed a QTc > 500 msec). She was told that she could return to play without restrictions. A concerned coach pursued further evaluation at our Pediatric Electrophysiology clinic where an ECG again demonstrated a long QT syndrome phenotype. Genetic testing confirmed a diagnosis of long QT type 1 (LQT1) with mutation of the *KCNQ1* gene. She was started on a beta blocker and was advised against further participation in competitive sports. She expressed a desire to participate in her team's final soccer game of the season. After reviewing the guidelines, and the risks of this decision, she ultimately decided to participate. We reiterated the importance of compliance with her beta blocker and confirmed availability of an AED and CPR-trained bystanders at the game. We then agreed with the patient, her family, and the school to this plan. There were no adverse events and the patient discontinued participation in competitive sports shortly thereafter.

Discussion

Shared decision making was introduced in 1982 and has been explained and interpreted in varying ways over the years. At the most basic level, it is a process of decision making shared by both the physician and the patient, neither paternalistic, nor completely autonomous as both have responsibility for the final outcome of the discussion. The use of a shared decision making model in sports participation in those at risk for sudden cardiac death is a relatively new concept among pediatric cardiologists that is more readily accepted now that recent research has demonstrated that participation in sports by at-risk athletes is safer than previously thought. Shared decision making allows for young athletes to balance safety and health with quality of life.

Conclusion

The experience of our patient demonstrates the benefits and safety of shared decision making in the lives of our high risk patients.

Categories

2nd year Fellow: Case

Program Name

Cleveland Clinic Children's