Objective: Describe a case that highlights the ongoing need for caution when interpreting genetic test results for inherited arrhythmia syndromes and cardiomyopathies and demonstrate that when faced with marked genotypic/phenotypic discordance, it is best to view the phenotype as the most important.
Methods: N/A
Results: This is a case of a 16-year-old male with out of hospital sudden cardiac arrest secondary to ventricular fibrillation (VF) initially attributed to a maternally-inherited pathogenic/likely pathogenic frameshift variant annotated as c.1961dup; p.T655NfxX49 in *LMNA*. However, cardiac imaging demonstrated a hemodynamically and structurally normal heart in the patient. Furthermore, while three first-degree relatives had this variant, their cardiac evaluations were normal. The family was recommended to receive prophylactic implantable cardiac defibrillators and thus sought out a second opinion. Since the clinical and genetic information was incongruent with the diagnosis, subsequent electrophysiology (EP) study revealed premature ventricular contraction (PVC)-triggered VF that was ablated successfully.
Conclusion: This case presents an interesting conundrum in which a pathogenic/likely pathogenic frame-shift variant was identified in *LMNA*, a gene that when mutated is normally thought to be associated with high penetrance and substantial risk of SCA. However, the phenotype was consistent with a non-genetic etiology and the pathogenic variant deemed to be a coincidental but irrelevant finding. Careful clinical assessment and genetic evaluation allowed the correct diagnosis to be made and unnecessary ICD implants in the family members to be avoided. In a patient with a pathogenic *LMNA* variant, LMNA-mediated sudden cardiac arrest in the setting of a pristine cardiac evaluation is unlikely. Therefore, before rendering such a conclusion, search for alternative explanations such as non-genetic, PVC-triggered VF.

B-PO05-209

PERICARDIAL EFFUSION AFTER LEFT ATRIAL APPENDAGE OCCLUSION RESULTING FROM AN AORTIC DISSECTION

Kara Stout MPH and Andrew Goldsweig

Background: Atrial fibrillation (AF) affects 2.3 million Americans. In those unfit for long-term anticoagulation, left atrial appendage occlusion (LAAO) is an alternative for stroke prophylaxis.
Objective: N/A
Methods: N/A
Results: A 68-year-old female with a history of AF, 4.3 cm ascending aortic aneurysm, hemoptysis on anticoagulation, and an aspirin allergy successfully underwent a LAAO with a 27 mm Watchman device. Postoperatively, she developed a moderate pericardial effusion without tamponade physiology. The effusion and her 3.5 cm aortic root (Fig. 1A) remained stable after anticoagulation reversal and over 48-hour observation. Warfarin and clopidogrel were restarted at discharge. The patient was readmitted 48 hours later with back pain. She was diagnosed with a Stanford type A dissection from the aortic root to the renal arteries, with extension into the pericardial space without evidence of tamponade (Fig. 1B, C). Unlike previously reported post-LAAO effusions, this effusion was not caused by the LAAO device. It is most likely that the new initiation of anticoagulation and possible injury during catheter manipulation potentiated the propagation of a pre-existing aortic dissection. Cardiothoracic surgery was emergently consulted, but she suffered a cardiac arrest and expired prior to intervention.
Conclusion: When evaluating a pericardial effusion after LAAO, the differential should extend past a procedural complication to include other life-threatening conditions, like an aortic dissection which was uniquely presented in this case. Peri-procedural echocardiograms should include an appraisal of the aortic root and ascending aorta to rule out acute pathology.

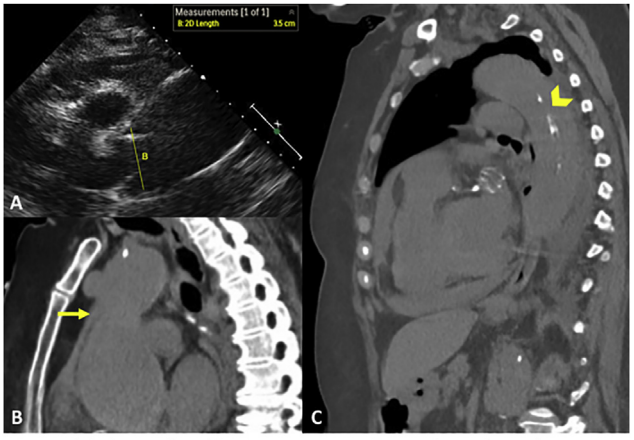


Figure 1. (A) A historically stable 3.5 cm aortic root without evidence of acute dissection. (B-C) Stanford type A aortic dissection (chevron) with extension into the pericardial space (arrow).

B-PO05-210

PREMATURE ATRIAL DEPOLARIZATION WITH ABERRANT CONDUCTION TRIGGERING VENTRICULAR FIBRILLATION

Mahmoud Elsayed, George Shaw MD, FHRS, John G. Chenarides MD, Amit J. Thosani MD, FHRS, Emerson Liu MD, William A. Belden MD and Mati Friehling MD

Background: Premature atrial contractions are common and usually benign. However, they are rarely documented to trigger ventricular fibrillation (VF).
Objective: 1. Describe a case of a patient with recurrent VF triggered by premature atrial contractions (PACs). 2. Describe our management approach.
Methods: N/A
Results: We present a 77 year old male with history of inferior myocardial infarction, coronary artery bypass grafting, and dual chamber implantable cardiac defibrillator (ICD) who presented with an episode of presyncope. Vital statistics and basic labs were normal in the emergency department. ECG showed an atrial-paced rhythm, inferior Q waves but otherwise normal QRS duration and normal QTc. Frequent PACs were noted, each with left bundle branch block aberrancy (LBBA). ICD interrogation revealed multiple episodes of defibrillation-terminated VF, each beginning with a PAC. Clinical PACs had the same ventricular aberrancy ICD EGM morphology as those triggering VF. His home Coreg dose was increased without improvement in PAC frequency. After discussion with the patient, we decided to proceed with EP study and ablation. In the EP lab, sedated, paced PACs from different locations demonstrated LBBA but did not trigger VF. PACs mapped

to the anterior right atrium. Ablation in and around this region successfully eliminated the PACs. (figure 1A, B, C and D). No further PACs were observed during his hospitalization and no further VF episodes have occurred since discharge.
Conclusion: We hypothesize that in our case, VF was triggered by aberrantly conducted PAC which induced bundle branch reentry and degenerated into VF, in the presence of arrhythmogenic substrate.

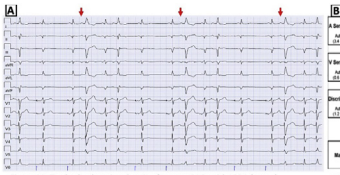


Figure 1A: Twelve lead tracing showing frequent PACs with LBBA (arrow).

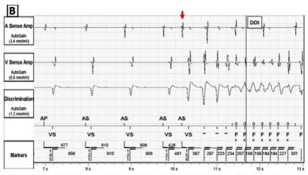


Figure 1B: Device interrogation EGM showing PAC initiation of VF (arrow).

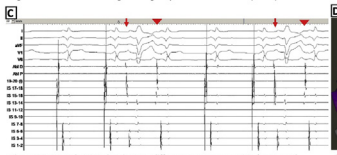


Figure 1C: EP study EGM showing two different spontaneous PACs (arrow and arrowhead).

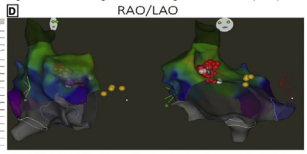


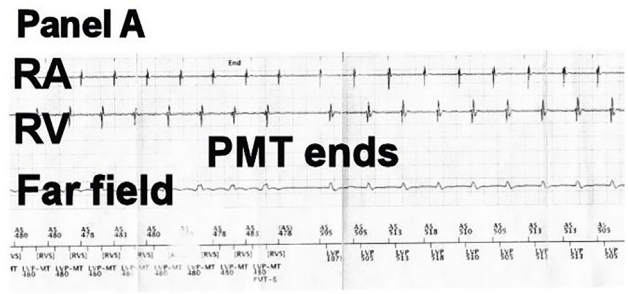
Figure 1D: 3D mapping of the right atrium showing ablation site (red dots) and His bundle (yellow dots).

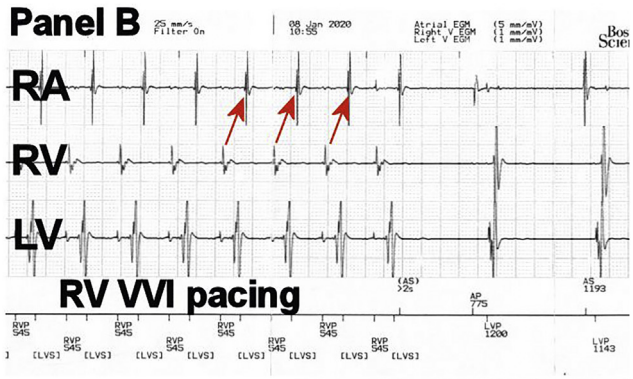
B-PO05-211

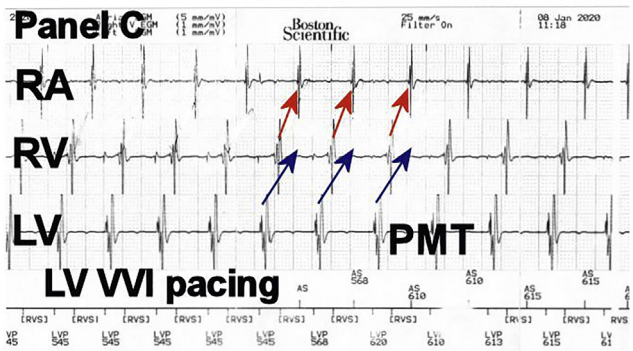
PROGRAMMING CHALLENGES DURING LEFT VENTRICULAR PACING. A CASE OF PACEMAKER-MEDIATED TACHYCARDIA

Karoly Kaszala MD, FHRS, Meredith L. Moyes MSN, FNP, CCDS, Alex Y. Tan MD, Kenneth A. Ellenbogen MD, FHRS and Jose F. Huizar MD, FHRS

Background: Multichamber pacing may be associated with complex interactions between pacing timing cycles.
Objective: Review of a case of pacemaker mediated tachycardia (PMT) during LV pacing.
Methods: Data was collected by retrospective review.
Results: A patient with a history of coronary artery disease and bypass surgery, heart failure and primary prophylaxis dual chamber defibrillator (ICD) underwent upgrade to a biventricular ICD (Boston Scientific Vigilant X4 G247) due to new left bundle branch block and heart failure symptoms. Postoperative programming was identical (DDDR 50/130/130 bpm; PVARP 240-280 ms) except for shortened AV delay with simultaneous RV/LV pacing. During an admission to an outside hospital, AV delay was shortened (SAV: 100 ms, PAV 150 ms) and pacing mode was changed to LV only. In follow-up, numerous PMT episodes were noted in the device memory (Panel A) and also during device interrogation but only with LV-only pacing. Retrograde P wave during LV pacing fell outside the programmed PVARP because retrograde left bundle branch block caused transseptal retrograde conduction via the right bundle branch with delay in LV to RA interval. Timing from RV apex to RA (Panel B, red arrow, 240 ms) was the same during RV pacing and during LV pacing (panel C) but because PVARP is initiated with the first paced ventricular chamber, the V to A time prolonged with LV-only pacing (Panel C, blue arrow, 420 ms) beyond PVARP. Programming very long PVARP limits upper tracking rate so in this case we switched to biventricular pacing which shortened the QRS duration and also eliminated PMT episodes.
Conclusion: In CRT devices, PVARP should be reassessed when LV only pacing is programmed.







B-PO05-212

RECURRENT VENTRICULAR TACHYCARDIA INITIATED BY WENCKEBACH PHYSIOLOGY DURING RAPID ATRIAL RATES

Rajan Ganesh, Alexander Hattoum MD, Kevin F. Kwaku MD, PhD and Evan Grove MD, MHS

Background: VT can be initiated by ventricular paced depolarization at the peak of the Twave, abrupt and long changes in ventricular cycle lengths or asynchronous pacing in competition with native AV conduction. It has not been described in the setting of Wenckebach physiology during sinus tachycardia.
Objective: We describe a case of VT preceded by Wenckebach physiology during sinus tachycardia.
Methods: N/A
Results: A 63 year old man presented to hospital with syncope while bicycling. Two years prior, he had a dual chamber pacemaker implanted for sinus node and infra-Hisian conduction system disease. Pacemaker interrogation demonstrated three