Sequential Correction of Wolff-Parkinson-White Syndrome and Perforated Mitral Valve in an Active Duty Service Member

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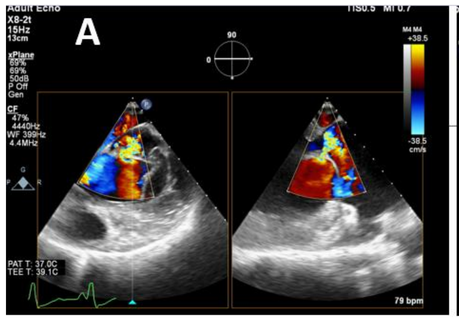
INTRODUCTION

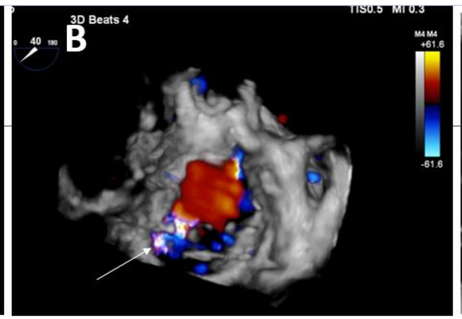
- Wolff-Parkinson-White (WPW) Syndrome is a well described pre-excitation syndrome involving the activation of the ventricles via an accessory pathway from the atria, bypassing the atrioventricular node
  - Symptoms include palpitations, syncope, and in rare cases, sudden cardiac death
- The estimated prevalence is approximately 1-3 in 1000 individuals with a slight increased incidence amongst individuals with a first degree relative with WPW
- It is estimated at 65% of adolescents and 40% of adults over 30 years of age with WPW are asymptomatic
- WPW may be associated with other, typically right-sided, cardiac abnormalities such as Ebstein's Anomaly
- Primary Mitral Valve Regurgitation in young, otherwise healthy individuals, is generally secondary to congenital malformation, rheumatic disease, or infective endocarditis
- Current guidelines and literature discuss the work-up and treatment of asymptomatic WPW, but do not typically address treatment in those undergoing concomitant cardiac surgery.

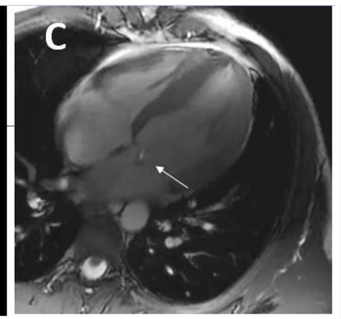
CASE

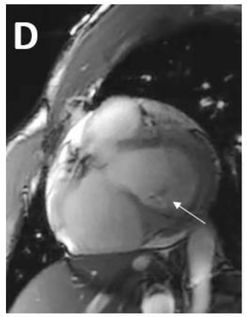
A 21 year old Active Duty male presented to Cardiology clinic following outside hospital evaluation revealing moderate to severe mitral regurgitation and WPW on EKG following emergency department evaluation for gastroenteritis.

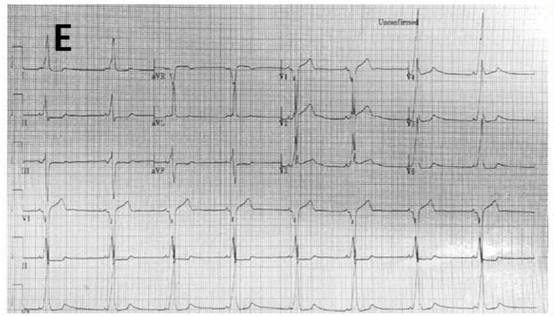
- Previously asymptomatic, the patient had noted recent increase in dyspnea on exertion, coupled with fatigue.
- Physical Exam would reveal, loud, blowing holosystolic murmur best heard at the apex with radiation to the axila
- ECG obtained demonstrated shortened PR interval with evidence of pre-excitation.
- Work-up was repeated, exercise testing showed persistence of pre-excitatory pathway.
- Transthoracic echocardiogram illustrated moderate to severe mitral regurgitation with left ventricular dilation
- Cardiac MRI was obtained that supported diagnosis of severe mitral regurgitation with dilated left ventricle
- Transesophageal echocardiogram (TEE) showed defect in the A3 position of the anterior mitral leaflet, initially thought to be a cleft

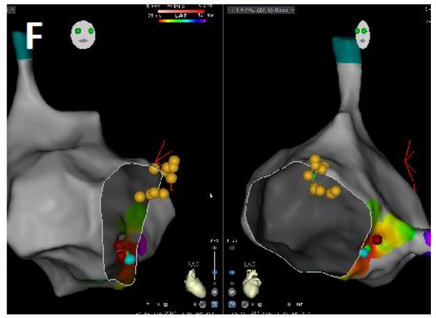












IMAGES

Images (A-F): A-TEE demonstrating a defect within the A3 portion of the anterior mitral valve leaflet. B-3D rendering of TEE showing defect (arrow) with regurgitant flow. C-Cardiac MRI four chamber view illustrating focal regurgitant flow jet. D-Short axis of cardiac MRI again showing jet; LV EDd and ESd would measure at 73mm and 47mm, respectively; with regurgitant volume of 64mL. E-Pre-excitation demonstrated on surface electrocardiogram most consistent with right posterior septal pathway. F-Posterior septal tricuspid annular region identified as source of accessory pathway in electrophysiology lab and post ablation.

TIMELINE-2020

Jan:	Feb-Mar:	Jul-Aug:	Aug -Sep:	Sep:	Late Sep:	Oct:
Eval in ED for gastroenteritis	Civilian Cardiologist work-up	Worsening symptoms	WRNMMC Cardiology Eval and work-up	Heart Team discussion—proceed to WPW ablation	WPW ablation	Mitral Repair via CT Surgery

REFERENCES

Pediatric and Congenital Electrophysiology Society (PACES), Heart Rhythm Society (HRS), American College of Cardiology Foundation (ACCF), et al. Paces/hrs expert consensus statement on the management of the asymptomatic young patient with a wolff-parkinson-white (WPW, ventricular preexcitation) electrocardiographic pattern: developed in partnership between the pediatric and congenital electrophysiology society (PACES) and the heart rhythm society (HRS). Heart Rhythm. 2012;9(6):1006-1024.

Additional References available upon request

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INTERVENTION

- Given findings consistent with severe mitral regurgitation with dilated LV, the patient was discussed multidisciplinary Heart Team Conference
- Ultimate determination was to proceed with WPW ablation prior to mitral valve repair due to high risk findings and the risk of unmitigated intraoperative accessory pathway conduction.
- Patient was taken to EP lab with and pathway was successfully ablated on the posterior septal tricuspid annulus with CARTO guidance. Post ablation there was central decremental VA conduction, with AV and VA block following adenosine administration.
- Patient proceeded to operating room, intraoperative findings were most consistent with mitral valve perforation secondary to prior native valve endocarditis.
- Right Mini mitral repair (thoracotomy) with annuloplasty band and anterior leaflet patch repair performed by cardiothoracic surgery.

DISCUSSION

- Initially thought to be a congenital malformation causing a cleft mitral valve in this otherwise healthy 21 year old male, intraoperative findings revealed a well-healed perforation consistent with prior undiagnosed endocarditis.
- Endocardial cushion abnormalities, as seen in congenital mitral valve clefts, may predispose a young patient to accessory pathway conduction, but it is likely in this case that perforation of the mitral valve was independent, and clinically distinct, from the finding of WPW
- Literature reviewed could not uncover any association between cleft or perforated mitral valve and WPW
- Although no clear practice guidelines exist, this case was performed in its sequence due to the risk, albeit low, of surgical AV nodal block, leading to unimpeded conduction along accessory pathway, and to reduce the likelihood of post-operative atrial fibrillation
- We propose that in any instance of planned cardiovascular surgery that an accessory pathway be addressed in the electrophysiology lab prior to surgical intervention.

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