Ebstein Anomaly in the Adult:
Management Using the 2018 AHA/ACC Clinical Practice Guidelines
Guideline Review and Case Studies

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Overview

- Spectrum of Ebstein Presentation in Adulthood
- 2018 ACC/AHA ACHD Guidelines A-P Classification
- Recent Case Presentations
- Case Guideline Classifications Recommendations
- Recent Case Current Outcomes
- ACHD Comprehensive Care Centers
- Summary and Discussion

Ebstein Anomaly in Adulthood:
Tremendous Variability

As a New Patient

- Murmur (TR/PS/ASD)
- Abnormal Routine Test
  - CXR (Cardiomegaly)
  - ECG (CRBB, WPW)
- New Arrhythmia
- New Cyanosis
- Exercise Intolerance

As an Existing Patient

- Prior Arrhythmia (SVT)
  - S/P RFA, or medication
- Prior Surgery (TV/PV/ASD)
  - Primary Repairs
  - Prosthetic Valves/Devices (ASD)
- Completely Different Disease
  - Fontan
  - 1 ½ Ventricle (Bidirectional Glenn)

NCHI-ACHD Program with currently > 30 Pts with Ebstein Anomaly

2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease (ACHD)

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Publication Information

This slide set is adapted from the 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

Published on August 16th, 2018, available at: *Circulation* and *Journal of the American College of Cardiology.*

The full-text guidelines are also available on the following websites: AHA (professional.heart.org) and ACC (www.acc.org)

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ACHA
Adult Congenital Heart Association

The 32nd Bethesda Conference on the Care of the Adult with Congenital Heart Disease—Highlights for Patients and Families

For many years the American College of Cardiology convened a yearly meeting in Bethesda, Maryland, to address important issues in cardiology. In 2000, the 32nd Bethesda Conference gathered international congenital cardiology experts to address the needs of adults living with heart defects. Below is a summary of their recommendations. You can read the full text online at www.acc.org.

Group 1—Simple Congenital Heart Disease
These patients can usually be cared for in the general medical community.

Unrepaired conditions:
Isolated small atrial septal defect (ASD)
Isolated small ventricular septal defect (VSD)
Mild pulmonic stenosis
Isolated dextrocardia—no other heart problems

Repaired conditions:
Patent ductus arteriosus (PDA)
Secundum atrial septal defect (ASD)
Isolated ventricular septal defect (VSD)

Repaired or unrepaired conditions:
Isolated aortic valve disease
Isolated mitral valve disease
Isolated patent foramen ovale (PFO)

Group 2—Congenital Heart Disease of Moderate Severity
These patients should be seen periodically at adult congenital heart disease centers.

Repaired or Unrepaired:
Anomalous Left Coronary Artery from Pulmonary Artery (ALCAPA)
Anomalous pulmonary venous drainage (partial or total)
Atrioventricular (AV) canal/septal defects (partial or complete)
Ostium primum or sinus venosus ASDs.
Coarctation of the aorta
Ebstein's anomaly
Infundibular right ventricular outflow obstruction (moderate to severe)
Pulmonary valve regurgitation (moderate to severe)
Pulmonic valve stenosis (moderate to severe)
Sinus of Valsalva fistula/aneurysm
Subvalvar or supravalvar aortic stenosis
Tetralogy of Fallot
Ventricular septal defect (VSD) with any valve problems and/or obstructions

Group 3—Congenital Heart Disease of Great Complexity
These patients should be seen regularly at adult congenital heart disease centers.

Repaired or Unrepaired:
Congenitally Corrected Transposition of the Great Arteries (ccTGA or l-TGA)
Double outlet ventricle
Mitral atresia
Pulmonary atresia (all forms)
Pulmonary vascular obstructive diseases
Shone's Syndrome
Single Ventricle—all forms (i.e., double-inlet ventricle; HLHS, HRHS, common/primitive ventricle)
Transposition of the Great Arteries (d-TGA)
Tricuspid atresia
Truncus arteriosus/hemitruncus
Other abnormalities of AV connections (i.e., crisscross heart, isomerism, heterotaxy syndromes)

All patients who have undergone the following procedures:
Arterial switch procedure
Blalock-Taussig shunt
Any Conduit(s), valved or nonvalved
Double-switch procedure
Fontan procedure
Mustard procedure
Norwood procedure
Rastelli procedure
Senning procedure

All Patients with Eisenmenger syndrome

All Patients who are cyanotic ("blue")

Not sure where your diagnosis/problem fits? Ask your cardiology team about which group above applies to you. You can read the full text of the ACC ACHD care recommendations at www.acc.org.

Norton Children's Medical Group
Affiliated with UL School of Medicine

Norton Children's Heart Institute
Affiliated with the University of Louisville

Guidelines for the Management of Adults with Congenital Heart Disease

Highlights for Patients and Families

The American College of Cardiology and the American Heart Association made rules (guidelines) for taking care of adult patients with congenital heart defects (CHD). The rules give doctors the most important information for adult congenital heart disease (ACHD). They also say it is important to have special heart centers for adults with CHD. This summary will tell you steps to take and questions you should ask to help take care of your heart. You can find the guidelines online at http://bit.ly/W8QLdu.

- If you were told you had a congenital heart defect (CHD) as a child, you should have your heart checked **at least once** at a special adult congenital heart disease (ACHD) center. This center can make sure that your heart is OK and any repairs you had are still working.
- If you were born with a more complex CHD, you should visit an ACHD center at least **every two years**. You should go even if you had surgery for your heart. *A list of all CHDs is on the other side of this page.*
- CHD heart problems are very different from "regular" adult heart problems. All staff at your ACHD center should be trained in adult CHD.
- If you do not live near an ACHD center, you will need to travel to visit an ACHD center. Your local heart team should talk to your ACHD center about your care.
- If you have a complex heart defect, all your doctors should check with your ACHD center about any medications or treatments. In most cases all medical procedures should only be done at hospitals that have an ACHD center.
- If you have complex CHD, the following should be done only by your ACHD center:
  - Echocardiograms
  - Pacemaker/ICD implantation
  - Cardiac MRI
  - Catheterizations – diagnostic and interventional
  - Cardiac CT/MUGA
  - Cardiac surgery
- Ask your ACHD center to teach you the name(s) of your CHD and any surgeries you've had. Know what you may be at risk for and the signs of new heart problems in the future.
- Your ACHD center should ask you about your emotional and social needs. They should offer non-medical help. This can include mental health support and social services like insurance and employment resources.
- Get an exercise prescription from your ACHD cardiologist. Your exercise prescription should be updated regularly. It is important to live a heart-healthy lifestyle because you can still get other forms of heart disease. Exercising every day can help strengthen your heart and maintain a healthy weight.
- If you need emergency care, make sure your ACHD center knows about it. Even if the emergency is not related to your heart, all emergency care providers need to talk with your ACHD center. Your center can decide if it is safe for you to continue your care locally.
- All women with complex CHD should check with their ACHD center **before** becoming pregnant. With the right care, most women with complex CHD can have a baby. An ACHD center should review the risks of pregnancy to both mother and baby. In some cases more heart treatment should occur before pregnancy begins.
- Certain forms of birth control can be risky for women with complex CHD. You should talk to your ACHD center to find a choice that is safe and effective for you.

ACHA Adult Congenital Heart Association www.achaheart.org

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How often should I visit an ACHD center?

SIMPLE CONGENITAL HEART DISEASE

Periodic heart checks should occur. General cardiologists can usually oversee care. One visit to an ACHD program to verify diagnosis and health status is recommended. ACHD care is usually not needed unless new problems occur.

Repaired conditions:
- Patent ductus arteriosus (PDA)
- Secundum atrial septal defect (ASD)
- Isolated ventricular septal defect (VSD)

Repaired or unrepaired conditions:
- Isolated aortic valve disease
- Isolated mitral valve disease
- Isolated patent foramen ovale (PFO)

Unrepaired conditions:
- Isolated small atrial septal defect (ASD)
- Isolated small ventricular septal defect (VSD)
- Mild pulmonic stenosis
- Isolated dextrocardia, no other heart problems

MODERATELY COMPLEX CONGENITAL HEART DISEASE

These patients should be seen every two years or more frequently at an adult congenital heart program.

Repaired or unrepaired conditions:
- Anomalous Left Coronary Artery from Pulmonary Artery (ALCAPA)
- Anomalous pulmonary venous drainage (partial or total)
- Atrioventricular (AV) canal/septal defects (partial or complete)
- Ostium primum or sinus venosus ASDs
- Coarctation of the aorta
- Ebstein's anomaly
- Infundibular right ventricular outflow obstruction (moderate to severe)
- Pulmonary valve regurgitation (moderate to severe)
- Pulmonic valve stenosis (moderate to severe)
- Sinus of Valsalva fistula/aneurysm
- Subvalvar or supravalvar aortic stenosis
- Tetralogy of Fallot
- Ventricular septal defect (VSD) with any valve problems and/or obstructions

HIGHLY COMPLEX CONGENITAL HEART DISEASE

These patients should be seen every year or more frequently at an adult congenital heart program.

Repaired or unrepaired conditions:
- Congenitally Corrected Transposition of the Great Arteries (ccTGA or L-TGA)
- Double outlet ventricle
- Mitral atresia
- Pulmonary atresia (all forms)
- Pulmonary arterial hypertension
- Shone's syndrome
- Single ventricle – all forms (*i.e., double-inlet ventricle; HLHS, HRHS, common/primitive ventricle*)
- Transposition of the Great Arteries (d-TGA)
- Tricuspid atresia
- Truncus arteriosus/hemitruncus
- Other abnormalities of AV connections (*i.e., criss-cross heart, isomerism, heterotaxy syndromes*)

All patients who have undergone any of the following procedures:
- Arterial switch procedure
- Blalock-Taussig shunt
- Any conduit(s), valved or nonvalved
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- Mustard procedure
- Norwood procedure
- Rastelli procedure
- Senning procedure

All patients with Eisenmenger syndrome

All patients who are cyanotic ("blue")

2018 ACHD AP CLASSIFICATION

(CHD Anatomy + Physiological Stage = ACHD AP Classification)

CHD Anatomy

I: Simple

Native disease

- Isolated small ASD
- Isolated small VSD
- Mild isolated pulmonic stenosis

Repaired conditions

- Previously ligated or occluded ductus arteriosus
- Repaired secundum ASD or sinus venosus defect without significant residual shunt or chamber enlargement
- Repaired VSD without significant residual shunt or chamber enlargement

II: Moderate Complexity

Repaired or unrepaired conditions

- Aorto-left ventricular fistula
- Anomalous pulmonary venous connection, partial or total
- Anomalous coronary artery arising from the pulmonary artery
- Anomalous aortic origin of a coronary artery from the opposite sinus
- AVSD (partial or complete, including primum ASD)
- Congenital aortic valve disease
- Congenital mitral valve disease
- Coarctation of the aorta
- Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations)
- Infundibular right ventricular outflow obstruction
- Ostium primum ASD
- Moderate and large unrepaired secundum ASD
- Moderate and large persistently patent ductus arteriosus
- Pulmonary valve regurgitation (moderate or greater)
- Pulmonary valve stenosis (moderate or greater)
- Peripheral pulmonary stenosis
- Sinus of Valsalva fistula/aneurysm
- Sinus venosus defect
- Subvalvar aortic stenosis (excluding HCM; HCM not addressed in these guidelines)
- Supravalvar aortic stenosis
- Straddling atrioventricular valve
- Repaired tetralogy of Fallot
- VSD with associated abnormality and/or moderate or greater shunt

III: Great Complexity (or Complex)

- Cyanotic congenital heart defect (unrepaired or palliated, all forms)
- Double-outlet ventricle
- Fontan procedure
- Interrupted aortic arch
- Mitral atresia
- Single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)
- Pulmonary atresia (all forms)
- TGA (classic or d-TGA; CCTGA or l-TGA)
- Truncus arteriosus
- Other abnormalities of atrioventricular and ventriculoarterial connection (i.e., crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

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2018 ACHD AP CLASSIFICATION

(CHD Anatomy + Physiological Stage = ACHD AP Classification)

CHD Anatomy

I: Simple

Native disease

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- Isolated small VSD
- Mild isolated pulmonic stenosis

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2018 ACHD AP CLASSIFICATION

(CHD Anatomy + Physiological Stage = ACHD AP Classification)

Physiological State

A
• NYHA FC I symptoms • No **hemodynamic or anatomic sequelae** • No **arrhythmias** • Normal **exercise capacity** • Normal **renal/hepatic/pulmonary** function

B
• NYHA FC II symptoms • Mild hemodynamic sequelae (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction) • Mild valvular disease • Trivial or small shunt (not hemodynamically significant) • Arrhythmia not requiring treatment • Abnormal objective cardiac limitation to exercise

C
• NYHA FC III symptoms • Significant (moderate or greater) valvular disease; moderate or greater ventricular dysfunction (systemic, pulmonic, or both) • Moderate aortic enlargement • Venous or arterial stenosis • Mild or moderate hypoxemia/cyanosis • Hemodynamically significant shunt • Arrhythmias controlled with treatment • Pulmonary hypertension (less than severe) • End-organ dysfunction responsive to therapy

D
• NYHA FC IV symptoms • Severe aortic enlargement • Arrhythmias refractory to treatment • Severe hypoxemia (almost always associated with cyanosis) • Severe pulmonary hypertension • Eisenmenger syndrome • Refractory end-organ dysfunction

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Rationale for ACHD A-P Classifications

Recognizes the high degree of variability in any particular CHD

- Status of the “Repair”
- Importance of residual, secondary or concomitant defects
- Considers both cardiac and non-cardiac “Co-morbidities”

Provides specific intervals for evaluations and interventions

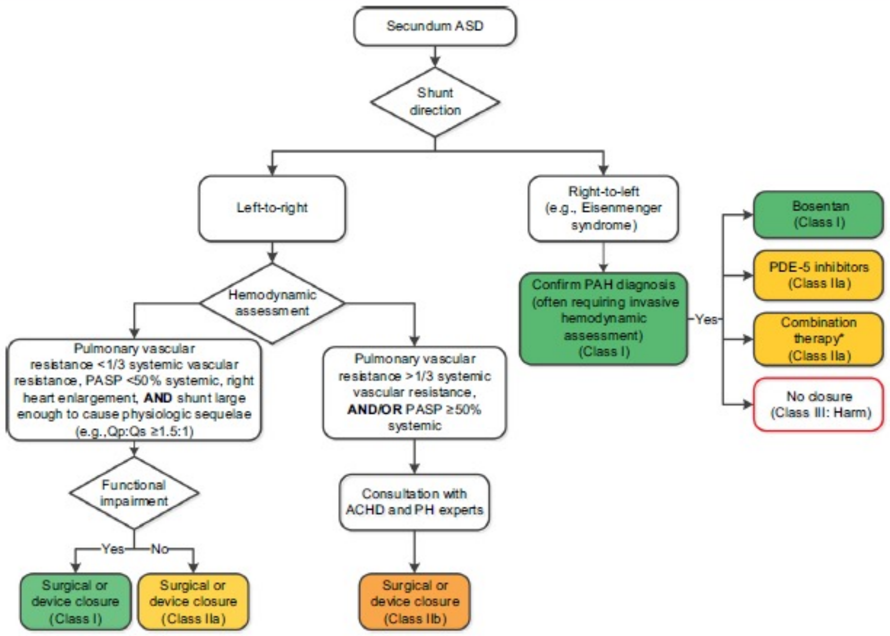
- Supportive information for those awkward “Peer to Peer” calls!

Example: A-P Guideline Directed Management and Therapy (GDMT) for Secundum ASD

Recommendations for Atrial Septal Defect
Referenced studies that support recommendations are summarized in Online Data Supplement 26 and the systematic review report (S4.1.1-1).

COR	LOE	RECOMMENDATIONS
		Diagnostic
I	C-EO	1. Pulse oximetry at rest and during exercise is recommended for evaluation of adults with unrepaired or repaired ASD with residual shunt to determine the direction and magnitude of the shunt.
I	B-NR	2. CMR, CCT, and/or TEE are useful to evaluate pulmonary venous connections in adults with ASD (S4.1.1-2–S4.1.1-4).
I	B-NR	3. Echocardiographic imaging is recommended to guide percutaneous ASD closure (S4.1.1-5, S4.1.1-6).
		Therapeutic
I	B-NR[SR]	4. In adults with isolated secundum ASD causing impaired functional capacity, right atrial and/or RV enlargement, and net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., pulmonary-systemic blood flow ratio [Qp:Qs] ≥1.5:1) without cyanosis at rest or during exercise, transcatheter or surgical closure to reduce RV volume and improve exercise tolerance is recommended, provided that systolic PA pressure is less than 50% of systolic systemic pressure and pulmonary vascular resistance is less than one third of the systemic vascular resistance (S4.1.1-7–S4.1.1-12).
I	B-NR	5. Adults with primum ASD, sinus venosus defect or coronary sinus defect causing impaired functional capacity, right atrial and/or RV enlargement and net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs ≥1.5:1) without cyanosis at rest or during exercise, should be surgically repaired unless precluded by comorbidities, provided that systolic PA pressure is less than 50% of systemic pressure and pulmonary vascular resistance is less than one third of the systemic vascular resistance (S4.1.1-13, S4.1.1-14).
IIa	C-LD[SR]	6. In asymptomatic adults with isolated secundum ASD, right atrial and RV enlargement, and net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs 1.5:1 or greater), without cyanosis at rest or during exercise, transcatheter or surgical closure is reasonable to reduce RV volume and/or improve functional capacity, provided that systolic PA pressure is less than 50% of systemic pressure and pulmonary vascular resistance is less than one third systemic resistance (S4.1.1-7–S4.1.1-10, S4.1.1-12).
IIa	C-LD	7. Surgical closure of a secundum ASD in adults is reasonable when a concomitant surgical procedure is being performed and there is a net left-to-right shunt sufficiently large to cause physiological sequelae (e.g., Qp:Qs 1.5:1 or greater) and right atrial and RV enlargement without cyanosis at rest or during exercise (S4.1.1-15–S4.1.1-18).
IIb	B-NR	8. Percutaneous or surgical closure may be considered for adults with ASD when net left-to-right shunt (Qp:Qs) is 1.5:1 or greater, PA systolic pressure is 50% or more of systemic arterial systolic pressure, and/or pulmonary vascular resistance is greater than one third of the systemic resistance (S4.1.1-19, S4.1.1-20).
III: Harm	C-LD	9. ASD closure should not be performed in adults with PA systolic pressure greater than two thirds systemic, pulmonary vascular resistance greater than two thirds systemic, and/or a net right-to-left shunt (S4.1.1-21, S4.1.1-22).

FIGURE 1 Secundum ASD



*Combination therapy with bosentan and PDE-5 inhibitor if symptomatic improvement does not occur with either alone. ACHD indicates adult congenital heart disease; ASD, atrial septal defect; PAH, pulmonary artery hypertension; PASP, pulmonary artery systolic pressure; PDE-5, phosphodiesterase type-5 inhibitors; PH, pulmonary hypertension; and Qp:Qs, pulmonary-systemic blood flow ratio.

TABLE 13 ASD: Routine Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	36-60	24	6-12	3-6
ECG	36-60	24	12	12
TTE	36-60	24	12	12
Pulse oximetry	As needed	As needed	Each visit	Each visit
Exercise test†	As needed	As needed	12-24	6-12

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Ebstein Anomaly: Presentation as an ACHD Patient

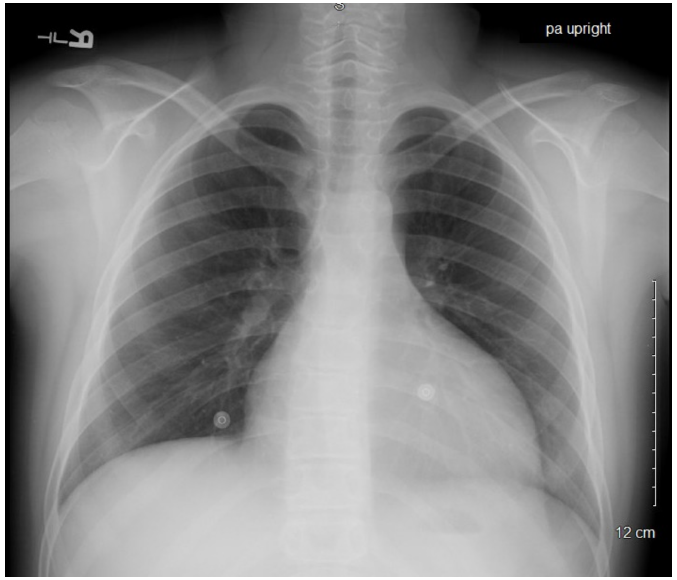
Case Study #1

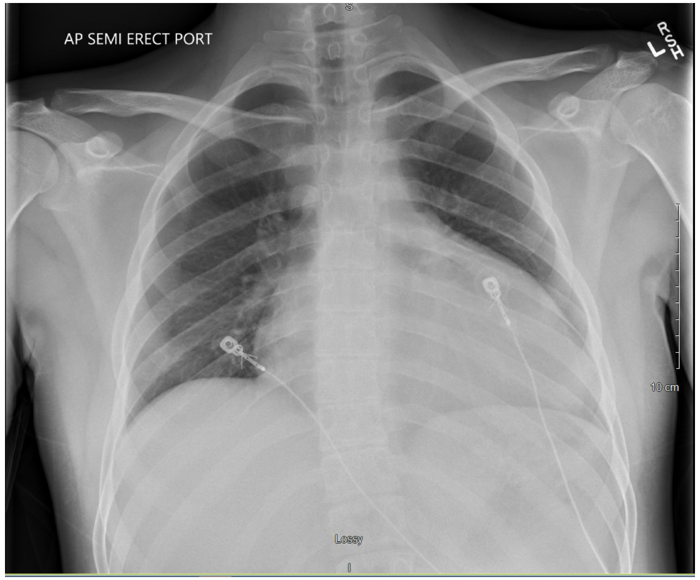
-WW is a 19 year old male with Ebsteins Anomaly of the Tricuspid Valve and no known surgical intervention to date (4/2022).

-Followed by local cardiologist until 2016 at which time echo, ecg, and exam were all stable

-No cardiac medications

-Co-morbidities: NONE





Ebstein Anomaly:
Presentation as an ACHD Patient

Case Study #2

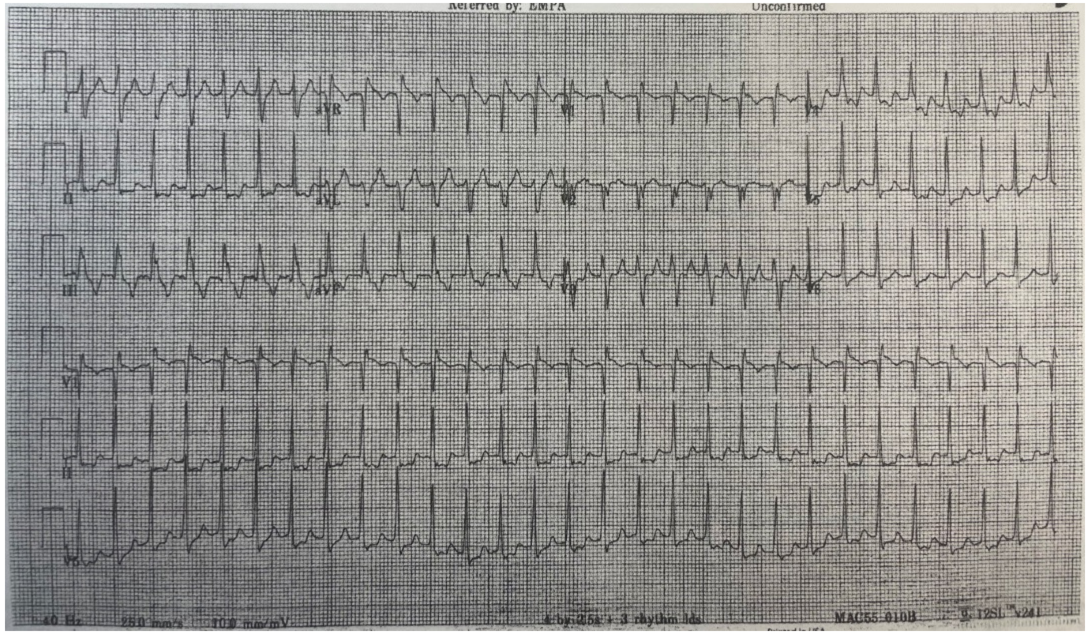
-LC is a 27 year old male diagnosed with Ebstein's Anomaly and ASD at 18 months of age.

-No interventions, no cardiac medications.

-Presented to ED in Sept 2018 with c/o palpitations.

-Recurrent tachycardia with at least 2 ED visits.

-Co-morbidities: HTN (HCTZ), migraines.



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2018 AHA/ACC Guidelines for the Management of Patients with Ebstein Anomaly

Recommendations for Ebstein Anomaly		
COR	LOE	Recommendations
Diagnostic		
IIa	B-NR	In adults with Ebstein anomaly, CMR can be useful to determine anatomy, RV dimensions, and systolic function .
IIa	B-NR	In adults with Ebstein anomaly, TEE can be useful for surgical planning if TTE images are inadequate to evaluate tricuspid valve morphology and function.
IIa	B-NR	Electrophysiological study with or without catheter ablation can be useful in the diagnostic evaluation of adults with Ebstein anomaly and ventricular preexcitation but without supraventricular tachycardia.
IIa	B-NR	In adults with Ebstein anomaly, electrophysiological study (and catheter ablation, if needed) is reasonable before surgical intervention on the tricuspid valve even in the absence of preexcitation or supraventricular tachycardia.

2018 AHA/ACC Guidelines for the Management of patients with Ebstein Anomaly

Therapeutic		
I	B-NR	Surgical repair or reoperation for adults with Ebstein anomaly and significant TR is recommended when one or more of the following are present: HF symptoms, objective evidence of worsening exercise capacity, progressive RV systolic dysfunction by echocardiography or CMR.
I	C-LD	Catheter ablation is recommended for adults with Ebstein anomaly and high-risk pathway conduction or multiple accessory pathways.
IIa	B-NR	Surgical repair or reoperation for adults with Ebstein anomaly and significant TR can be beneficial in the presence of progressive RV enlargement, systemic desaturation from right-to-left atrial shunt, paradoxical embolism, and/or atrial tachyarrhythmias.
IIb	B-NR	Bidirectional superior cavopulmonary (Glenn) anastomosis at time of Ebstein anomaly repair may be considered for adults when severe RV dilation or severe RV systolic dysfunction is present, LV function is preserved, and left atrial pressure and LV end diastolic pressure are not elevated.

(Con't.)

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Ebstein Anomaly

TABLE 26 Ebstein Anomaly: Routine and Follow-Up and Testing Intervals

Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12–24	12	6–12	3–6
ECG	12–24	12	12	12
CXR	As needed	As needed	12–24	12–24
TTE†	12–24	12–24	12	12
Pulse oximetry	24	12	Each visit	Each visit
Holter monitor	As needed	As needed	24	12–24
CMR‡/CCT§	60	36	24–36	12–24
Exercise test‖	36	24–36	24	12

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GDMT Case Management and Outcome

Case Study #1 WW

ACHD AP Classification: IIC

-Met indications for surgical intervention

-Class I

1) mod-sev TR and depressed RV function
2) Progressive RV systolic dysfunction by Echo and cMRI

-Class II

1) reduced systemic saturations due to R->L shunt.

-Met indication for cardiac ablation

-Class IIa

1) Electrophysiological study (and catheter ablation, if needed) is reasonable before surgical intervention on the tricuspid valve even in the absence of preexcitation or supraventricular tachycardia

GDMT Case Management and Outcome

Case Study #1 WW

- Underwent pre op EPS/ABL: radiofrequency ablation of the slow pathway for typical AVNRT.

GDMT Case Management and Outcome

Case Study #1 WW

-Underwent Cone procedure, right ventricular reduction with plication, right atrial reduction with excision, primary closure of ASD, and modified right atrial Maze on 8/22/22.

-Immediate post operative period complicated by hypovolemia, transaminitis, and thrombocytopenia as well as frequent NSVT on telemetry.

-Post-op TTE revealed no TS and traceTR, moderate to severely depressed RV function. No residual atrial level shunt.

GDMT Case Management and Outcome

Case Study #1 WW

-iNO transitioned to Sildenafil

-Transaminitis and resolved

-Telemetry stable on Toprol XL 25mg daily

-Most recent TTE with no TS, trace TR, improvement in RV function

-Discharged home on post op Day 12

GDMT Case Management and Outcome

Case Study #2

ACHD AP Classification: IID

-Met indications for surgical intervention
- -Class I
  1) mod-sev TR and low normal RV systolic function
  2) mod reduced functional capacity by MST
- -Class IIa
  1) reduced systemic saturations due to R->L ASD shunt.
  2) recurrent SVT (multiple ED visits)

-Met indication for EPS
- -Class IIa

-Met indication for cardiac ablation
- -Class I

GDMT Case Management and Outcome

Case Study #2

-Underwent pre-op EPS/ABL (Feb 2019): AVRT induced and multiple accessory pathways were targeted for radiofrequency and cryoablation.

-Recurrence of SVT thereafter and was restarted on BB therapy.

GDMT Case Management and Outcome

Case Study #2

-Underwent Cone procedure (Mar 2019) with tricuspid anuloplasty (34 mm anuloplasty ring), right atrial MAZE procedure and primary closure of ASD.

-Post-op TTE revealed no TS and trivial TR, with stable low normal RV systolic function. No residual ASD shunting.

-Post-op ventricular ectopy (PVCs, NSVT), rare second degree, Mobitz type 2 AVB.

-Discharged home off BB therapy. ASA 81 mg daily (valve patency).

GDMT Case Management and Outcome

Case Study #2

-Most recent OV (May 2022)

-TTE with no significant TS, stable mild TR with normal RVP. Low normal RV systolic function.

-MST with pVO2 27.9 mL/kg/min with mildly reduced functional capacity.

-HM predominately SR. No significant ectopy or tachyarrhythmia.

-Continues working as a firefighter with minimal symptoms. Recently bought a farm and is frequently completing remodeling projects. He and his wife are expecting their second child.

ACHA
Adult Congenital Heart Association

Adult Congenital Heart Association

New Logo

- Founded in 1998, the Adult Congenital Heart Association is an organization begun by and dedicated to supporting individuals and families living with congenital heart disease and advancing the care and treatment available to our community.

- The mission of the Adult Congenital Heart Association is to improve and extend the lives of the millions born with heart defects through education, advocacy and research.

ACHA-ACHD
Comprehensive Care Centers

Recent Studies Demonstrate:

- Improved Outcomes in ACHD Centers
- Improved Outcomes in Pediatric Hospitals

- Currently 46 Centers in 27 States
  - NCHI only one in Kentucky
  - UK-Cincinnati Affiliation
- **<u>UofL/NCHI ACHD Program</u>**
  - Approved August 2021
  - ACHD Local Community Support
    - 16 Outreach Clinics
      - KY: Murray, Paducah, Madisonville, Owensboro, Bowling Green, Elizabethtown, Campbellsville, Mt Sterling, Corbin, Mt Washington, Shelbyville
      - IN: New Albany, Corydon, Jasper, Scottsburg, Madison
    - 31 Tele-Echo sites
    - Daily Louisville ACHD visits
    - Daily ACHD Tele-health Visits

- Instituted 2016
- 19 Services area requirements
- A. ACHD Cardiologist
- B. ACHD Medical Program Director
- C. Advanced Practice Nurse/Physician Assistant
- D. Registered Nurse
- E. Cardiothoracic Surgery and Cardiothoracic Intensive Care Unit
- F. Heart Failure, Heart Transplant, Heart/Lung Transplantation
- G. Interventional Cardiac Catheterization
- H. Interventional Electrophysiology
- I. Inpatient Services
- J. Outpatient Services
- K. Transitional Services
- L. Patient-Centered Care
- M. Echocardiography
- N. Cardiac Magnetic Resonance Imaging
- O. Cardiac Computed Tomography
- P. Pulmonary Arterial Hypertension
- Q. Exercise Testing and Cardiac Rehabilitation
- R. Reproductive Services
- S. Psychology and Social Work

Summary:
Ebstein Anomaly in Adulthood

- Complex, Highly Diverse Condition
- Presents with or Progresses to Involve
    - Valvular function
    - Ventricular function
    - Atrial Arrhythmias
- Best Outcomes Require Coordination
    - Experienced Surgical Expertize
    - Experienced EP/Pacemaker Expertize
    - Specialized Multi-disciplinary long-term ACHD Care

Questions and Discussion

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To Find an ACHD Center:
Website: ACHAheart.org