ACHD and Heart Failure: Problem Solving 101

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UVA Heart Failure for the Advanced Practice Provider Symposium
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Disclosures

- I have no disclosures

Objectives

- Introduce adult congenital lesions susceptible to heart failure
- Discuss framework to start problem solving CHF in ACHD
- Understand heart failure in various scenarios
  - Left heart failure
  - Right heart failure
  - Systemic right ventricle heart failure
  - Single ventricle heart failure
  - Eisenmenger's syndrome

When I think of congenital heart disease…

Instead… Think Positive

https://www.superheroheartrun.com/

CHF Questions (Gatzoulis)

1. Basic ABC of resuscitation if appropriate? (may not have DNR or advanced care planning)
2. What is the anatomy and what does it mean?
3. Is there a new or worsening hemodynamic lesion that needs intervention? (valve stenosis, chordal rupture, valve regurgitation, arterial or venous stenoses)
4. Is the patient in sinus rhythm?
5. Is there evidence of infective endocarditis or systemic infection?
6. Does the patient have a Fontan repair? (or palliated single ventricle or Eisenmenger's)
7. What is the site of the problem? LV? RV? Both?
8. What is the main problem: poor cardiac output (L), peripheral edema (R), pulmonary edema (L)? (systolic or diastolic?)
9. What are the disadvantages of giving a drug on systemic output, filling pressures, pulmonary vascular resistance, kidneys, etc?
10. Where will this line end up if I attempt to gain access? (Fontan, Glenn, L SVC, Mustard)

Seek expert advice as soon as possible.

Gatzoulis, M. "Diagnosis and Management of Adult Congenital Heart Disease." 3rd Edition

Statistics

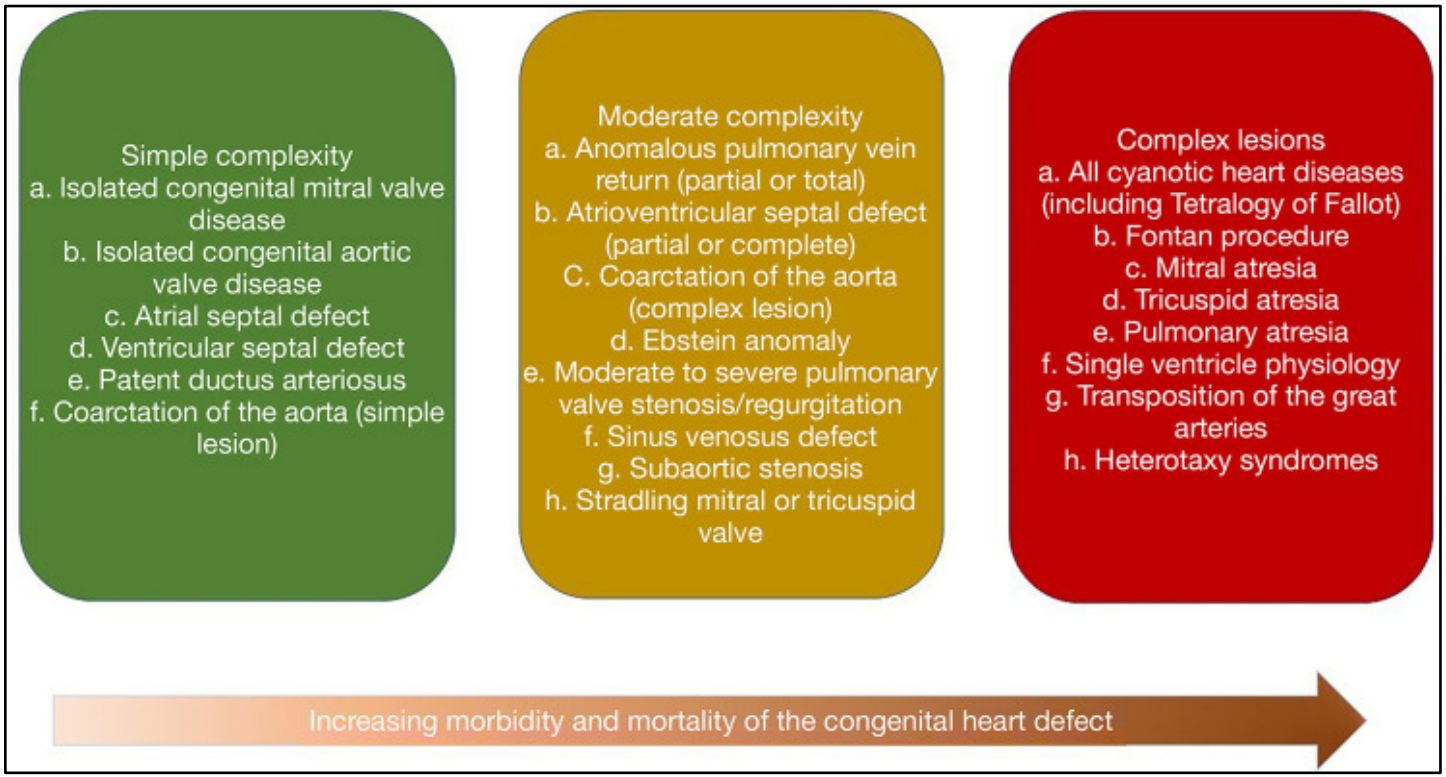
- CHD is 1% of the general population (“we are the real 1%”)
- Improved survival, 88% make it to adulthood
  - Better fetal screening and disease identification
  - Newer surgical/cath techniques and earlier interventions
- CHF becoming increasing cause of hospitalization in ACHD patients
- CHF hospitalization associated with increased mortality rate
- Leading cause of death in ACHD
- Number of patients being assessed for transplant has increased
- Not everyone will be able to get a heart transplant (anatomy, organs needed – lungs/kidneys/liver, volume of patients, how to list)

Classifying Lesions

ACHD AP Classification

1) Anatomic
   -Simple
   -Moderate
   -Complex

2) Functional Class
   -NYHA 1
   -NYHA 2
   -NYHA 3
   -NYHA 4



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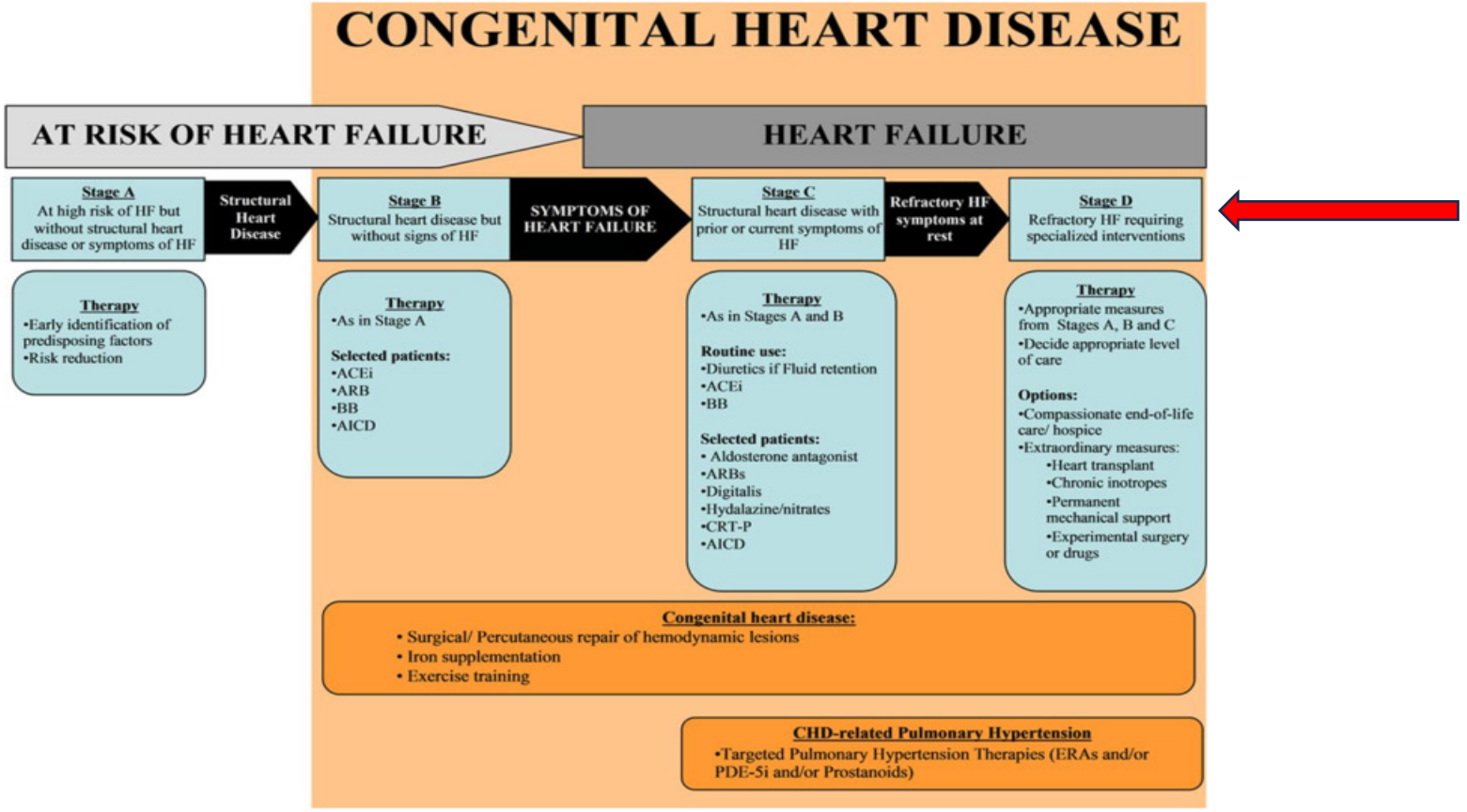
Heart Failure: ACHD Stage C + D



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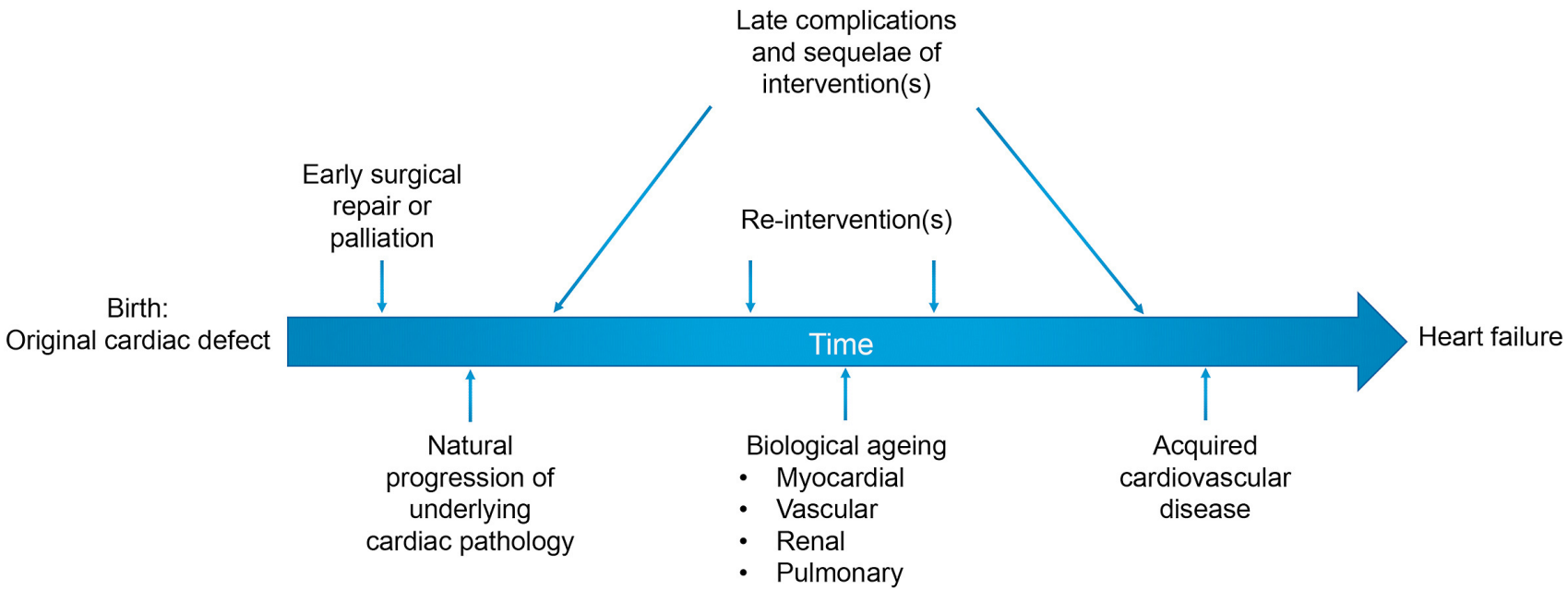
Rush, C J. "The emerging burden of heart failure in adults with congenital heart disease." International Journal of Cardiology CHD, 2021 Aug; Volume 4. https://www.sciencedirect.com/science/article/pii/S2666668521000951

Risk Factors for Heart Failure in ACHD



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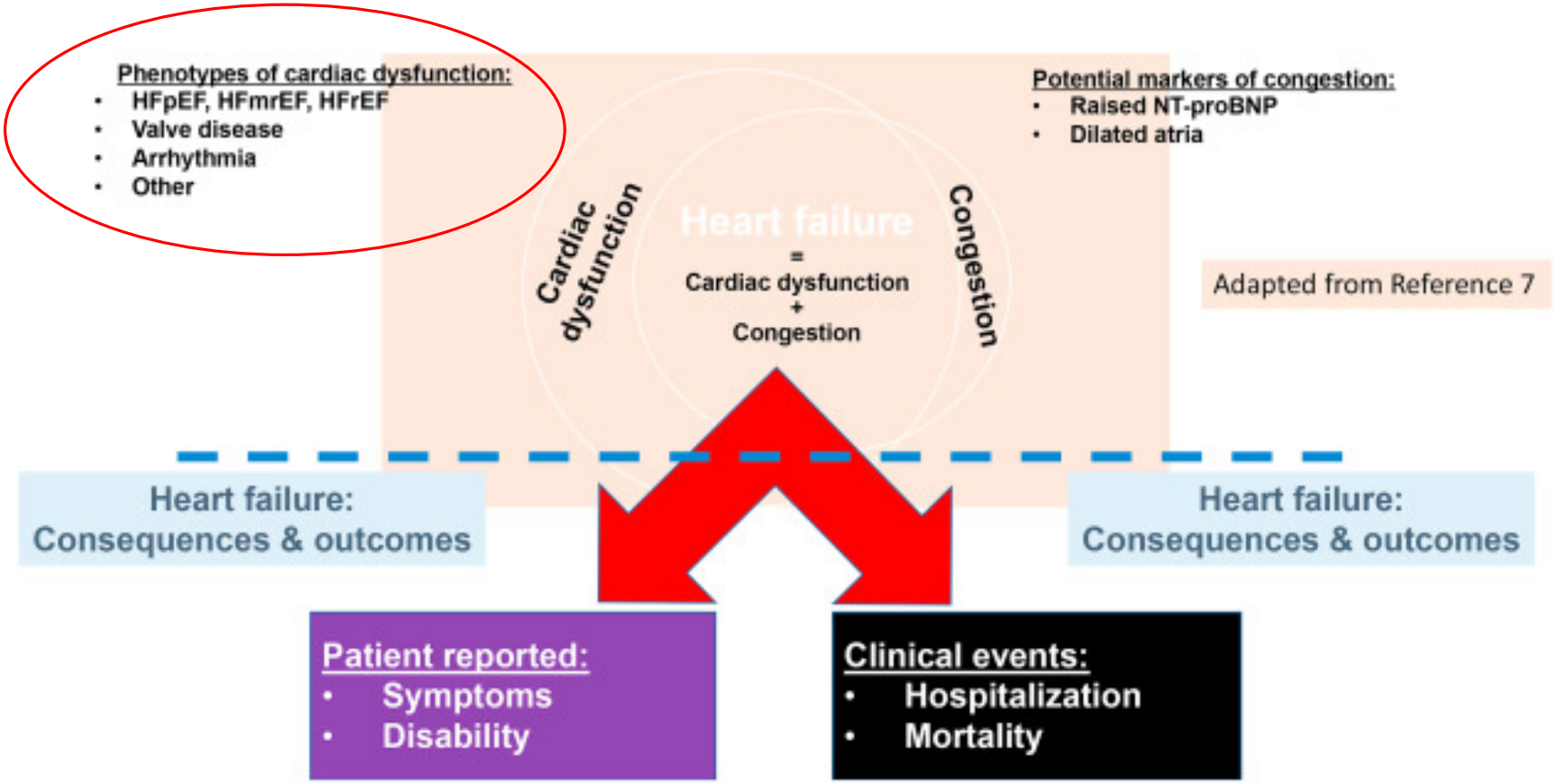
Risk Factors for Heart Failure in ACHD

- Multiple pump runs and surgeries
- Chronic cyanosis when younger (causing fibrosis, ischemic changes)
- Coronary anomalies (congenital) (Ross, sinusoids, DTGA, etc)
- Acquired Coronary artery disease (ICM)
- Typical adult issues (HTN, DM2, renal failure, etc)
- Reconstructed ventricles (VSD patch material does not contract)
- Arrhythmias
- Sinus node dysfunction and heart block
- Valve disease
- Genetic issues (HCM, NICM, Duchenne's, noncompaction)
- Pulmonary hypertension
- Restrictive lung disease
- Other special needs or systemic diseases

CHF Phenotypes in ACHD



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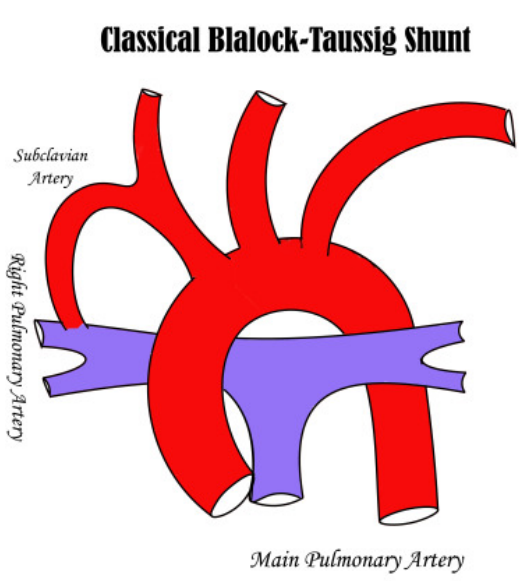
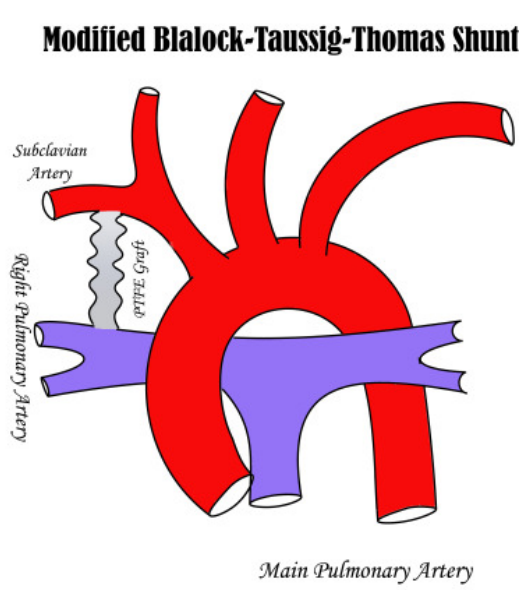
Key History and Exam Pearls

- Look at their scars! (median sternotomy vs lateral thoracotomy vs clamshell, where is pacemaker?)
- Check their age (may give clues to era surgery was done in)
- Check to see if they have all their pulses
  - BTT shunts
  - Aortic repairs
  - Multiple femoral caths
- Figure out their baseline O2 saturation
- If cyanotic/hypoxic check their labs (Hgb, iron)
- Check for clubbing, jaundice, edema, varicosities, liver
- Correlate murmur with imaging (pts with multiple murmurs)
- Pulse gradients (aortic repairs)
- Rhythm/pacing (AV synchrony?)
- Look up their anatomy (pted.org, achaheart.org) / understand the physiology / ask for help
- Ask the patient what they know, it may be more than you realize!
- Include family in the discussion
- Screen for/recognize mental health or cognitive barriers to care (Guardianship? IQ? Functional level?)

What is the Anatomy?

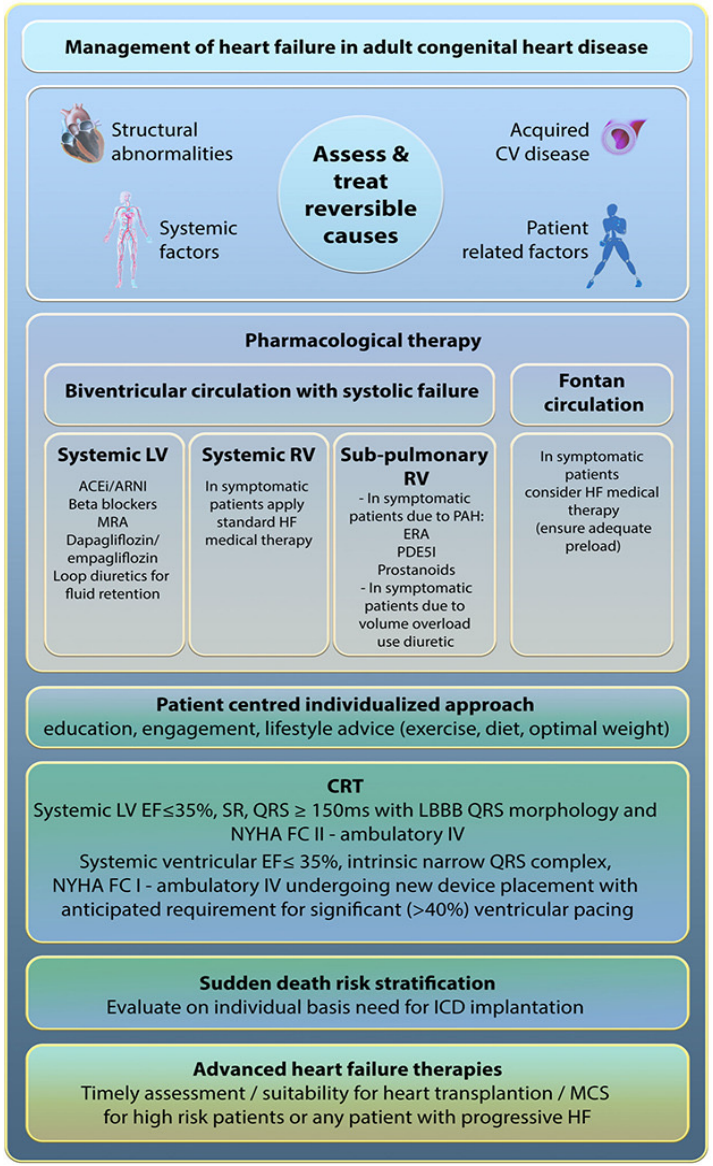
- May need to look up their anatomy
- Could be in patient's chart as a scanned image from cardiac cath
- References
  - www.pted.org
  - www.achaheart.org
  - Cincinnati Children's app
  - Boston Children's
    (Multimedia Library)
  - CHOP
  - Mayo Clinic



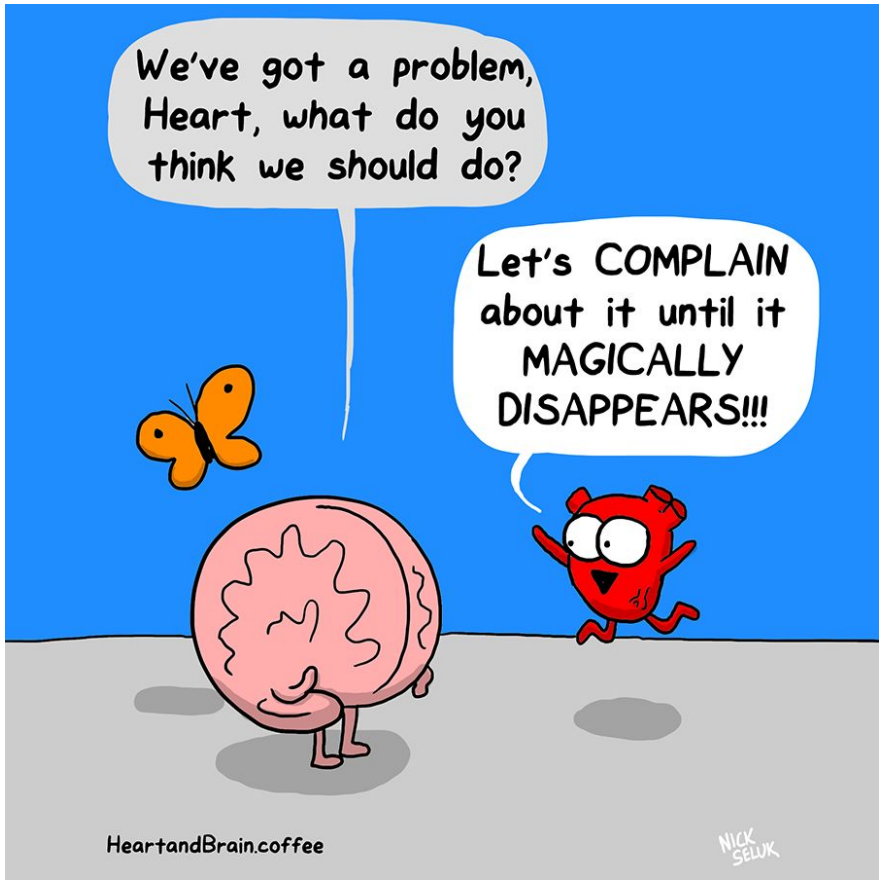


May not have pulse in arm that subclavian artery was used from

ACHD CHF Treatment



Cases and Problem Solving



Left Heart Failure

- We are experts in this in adult cardiology
- Dealing with lesions that are typical 4 chamber hearts
- Systolic dysfunction we have clear GDMT
- Diastolic dysfunction is a harder problem
- Can lead to pulmonary hypertension and RV failure
- Some ACHD lesions are already a set up for RV dysfunction (TOF, pulmonary stenosis)
- In ACHD do not forget about lesions with residual pathology that may need intervention (sub AS, coarctation for example)

Left Heart Lesions

- Sub aortic stenosis
- Coarctation of the aorta
- Congenital aortic stenosis
- Congenital mitral stenosis
- Mitral or left AV valve regurgitation
- Cor triatriatum
- Acquired heart disease issues

Typical CHF Management Options

- These may not work for all patients with CHD
- MCS/acute options
  - IABP
  - Impella
  - LVAD
  - TAH
- Systolic
  - Afterload reduction - ACEi/ARB (hydralazine)
  - NH blockade/remodeling – spironolactone, Farxiga/Jardiance, beta blocker
  - Digoxin (end stage, decrease hospitalization benefit)
  - Diuretics
  - SGLT2i
  - MCS/AHFT
  - Transplant
  - Pacing?
- Diastolic
  - SGLT2i
  - Diuretics
  - Transplant

Coarctation

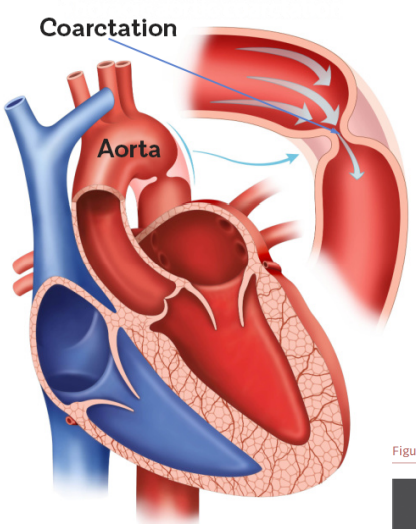
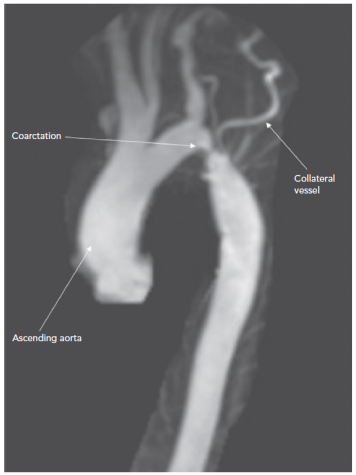
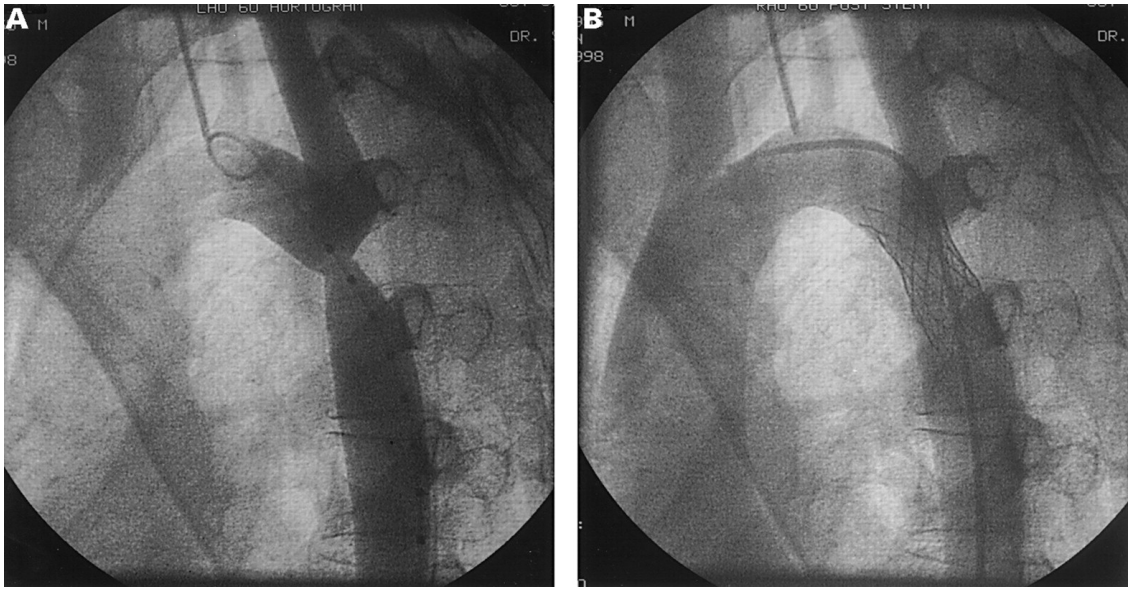


Figure 1: Coarctation Magnetic Resonance Image



Cardiac MRI of a 53-year-old patient with severe coarctation and evidence of collateral flow.



Intervention with stenting for re-coarctation

Case 1

- 40 year old man with coarctation s/p repair, jump graft at aortic aneurysm site at 16 yo
- Increased fatigue, dizziness, elevated BP, started on ACEi, CCB, and finally b blocker, bradycardia to 50, near syncope
- Echo showed LVEF 40%, LVH, diastolic dysfunction, severe coarctation gradient 30 mmHg (over 20 mmHg needs intervention)
- Sent for cardiac cath and stenting, s/p coarctation stent
- Improved BP but needed close management inpatient due to rebound HTN afterwards
- With afterload reduction and coarctation intervention, LVEF improved to 55% at 2 months post cath

Case 2

- 20 year old man with Shone's complex (multiple left sided lesions), bicuspid aortic valve, moderate aortic regurgitation, coarctation of the aorta s/p repair, mild mitral stenosis
- Establishes with new cardiologist after last provider retires
- Echo shows LVEF 40 to 45%, mild 18 to 20 point descending aortic gradient, mildly dilated and hypertrophied LV, moderate AR
- TEE done to make sure AR is not worse than what was on TTE
- Sent for cardiac cath, showed 25 point gradient in descending aorta, s/p stenting and started on ACE inhibitor for cardiac remodeling
- At 3 months post stent, his LVEF improved to 60% again and he noted increased endurance and improved activity tolerance

Case 3

- 25 year old lady with ASD and VSD s/p repair, PDA s/p ligation, mildly narrow aorta, isolated LL pulmonary vein stenosis s/p stent
- When younger had CHB and dual chamber pacemaker implant
- Got older and LVEF started to drop
- Became more fatigued with decreased exertion
- CT imaging showed only mild residual aortic narrowing, no significant LVH on echo, normal arm/leg gradient
- EKG did show QRS > 150 ms
- Decision to upgrade to biV pacing
- LVEF improved without need for aortic intervention

Right Heart Failure

- Tetralogy of Fallot
- Pulmonary atresia/intact ventricular septum
- Pulmonary stenosis
- Tricuspid stenosis
- Left heart failure
- Pulmonary hypertension patients including Eisenmenger’s

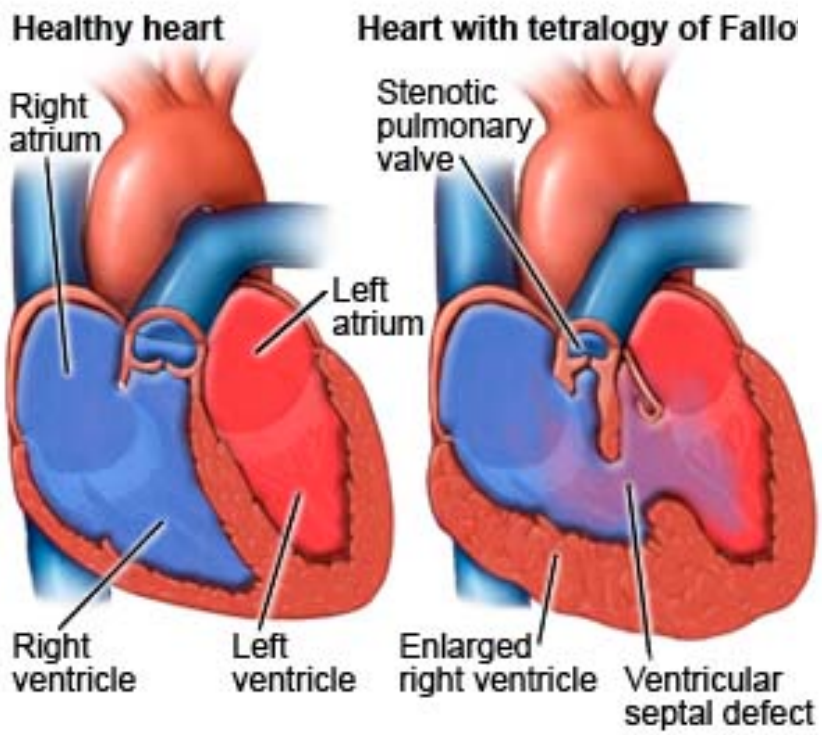
Right Heart Failure treatment

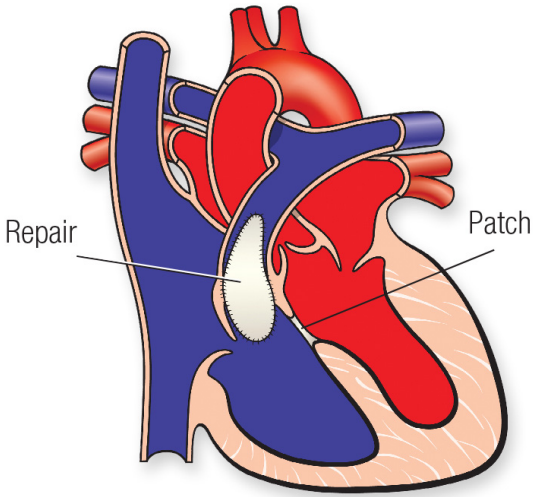
- Supportive care
- Diuretics
- RVAD limited options
- RV Impella is only short term
- Management of left heart failure
- Treat OSA and HTN
- PAH treatment
- Don’t forget valve replacement in TOF
- Important to maintain AV synchrony for euvolemia

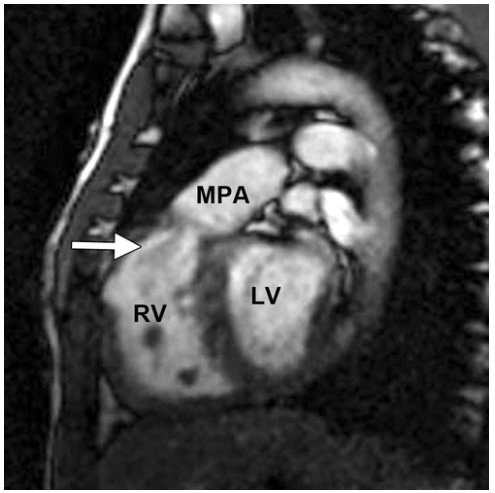
Case 4

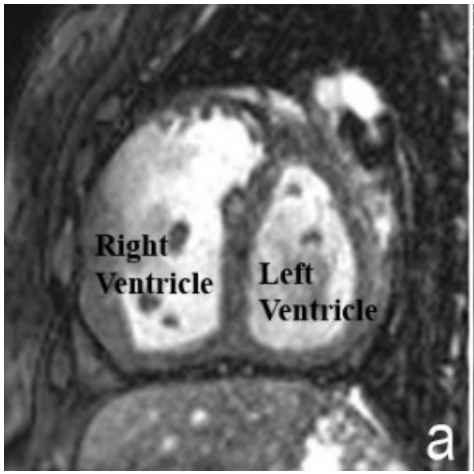
- 65 yo F presented feeling weak, found to be in atrial flutter with RVR, history of Tetralogy of Fallot s/p repair as a toddler in 1960s
- Echo shows severe pulmonary regurgitation, moderately decreased right ventricular function, mild LV diastolic dysfunction
- After cardioversion, tele shows sinus rhythm, short runs of nonsustained ventricular tachycardia (she does not feel them), started on amiodarone
- Mild diuresis with Lasix inpatient, now euvolemic
- We are not done, arrhythmia “tipped her over” but underlying issue is severe pulmonary regurgitation
- Options: surgical vs catheter pulmonary valve replacement
- Had TPVR, follow up improved biV EF and improved symptoms, weaned off amio post TPVR and is doing well

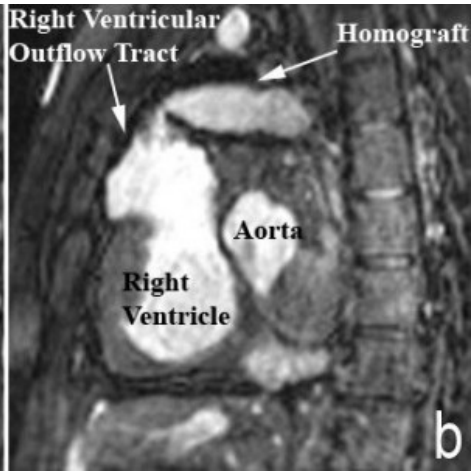
Tetralogy of Fallot









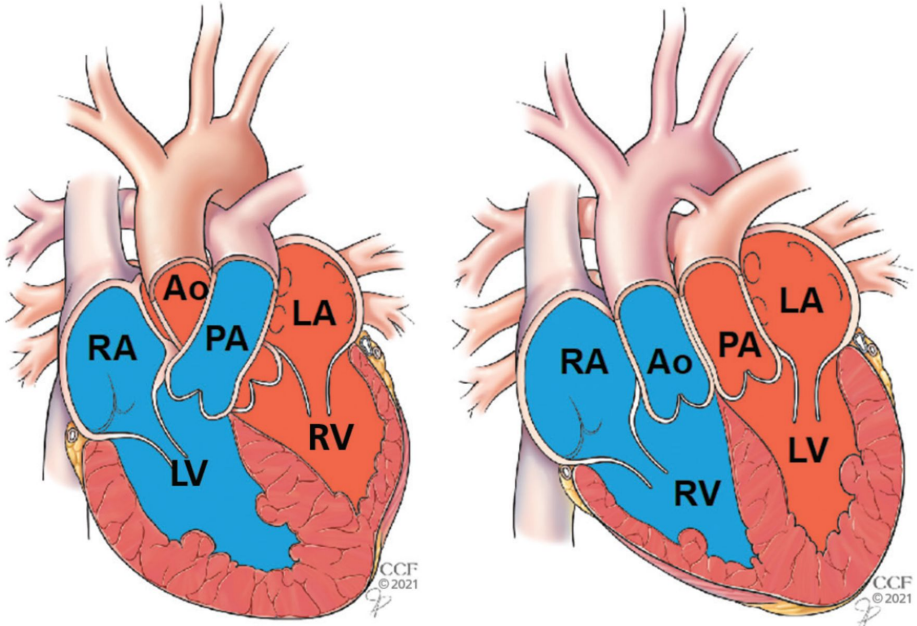


Case 5

- 40 yo M with Tetralogy of Fallot s/p most recent PVR in 2021, SSS s/p pacemaker, atrial fib on sotalol and apixaban, currently sinus at last clinic visit
- Presents with fever, hypotension, DIC, septic shock, in atrial fib with RVR, early signs of cardiogenic shock, mild Cr elevation, needs pressors and inotropes and volume resuscitation
- Echo shows new severe pulmonary regurgitation, mildly decreased RV and LV function, and TEE shows pulmonary valve vegetation, blood cultures are positive
- Treated with IV antibiotics and IR vacuum aspiration of vegetation, started on heart failure therapy outpatient
- Presented back after 3 months for surgical pulmonary valve replacement

Systemic Right Ventricles

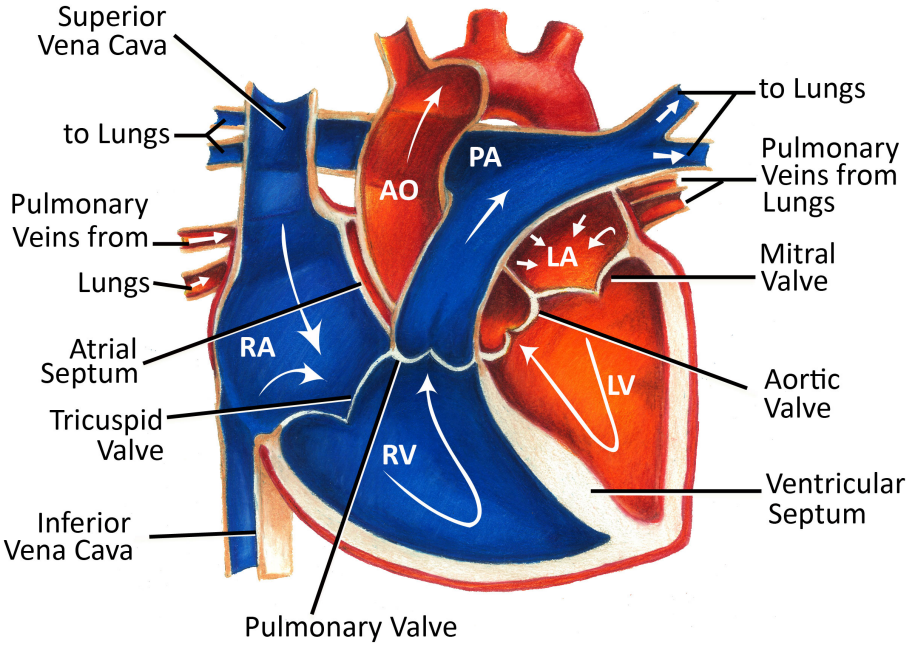
- DTGA s/p Mustard or Senning Palliation
- LTGA (ventricular inversion) (CCTGA)

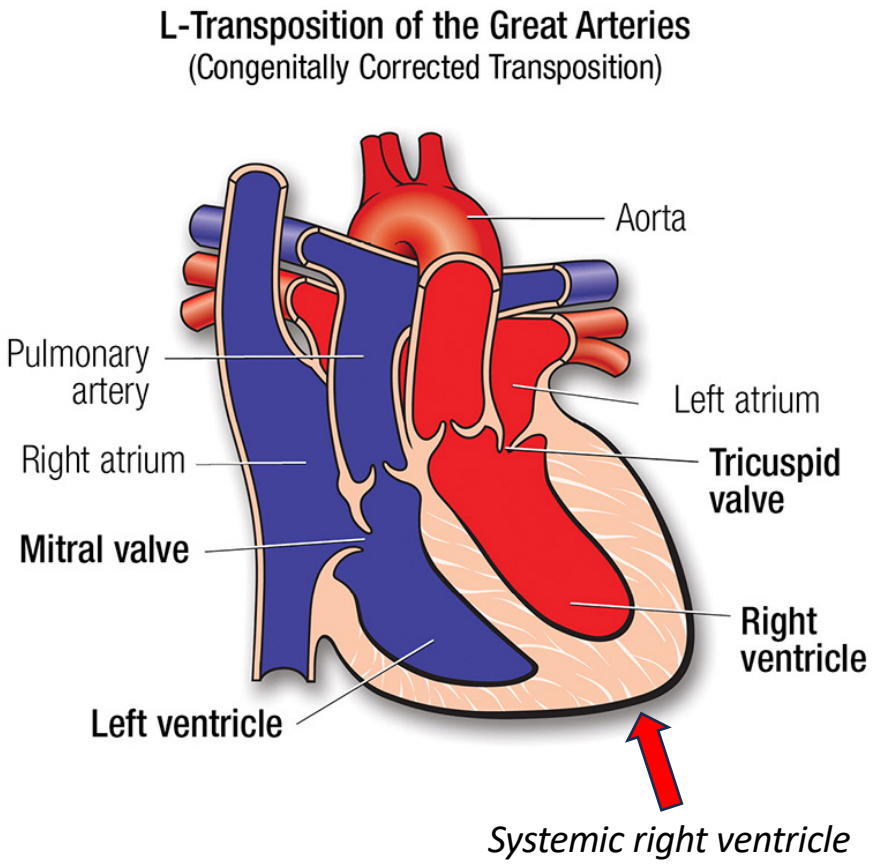


CC-TGA D-TGA

LTGA (CCTGA)

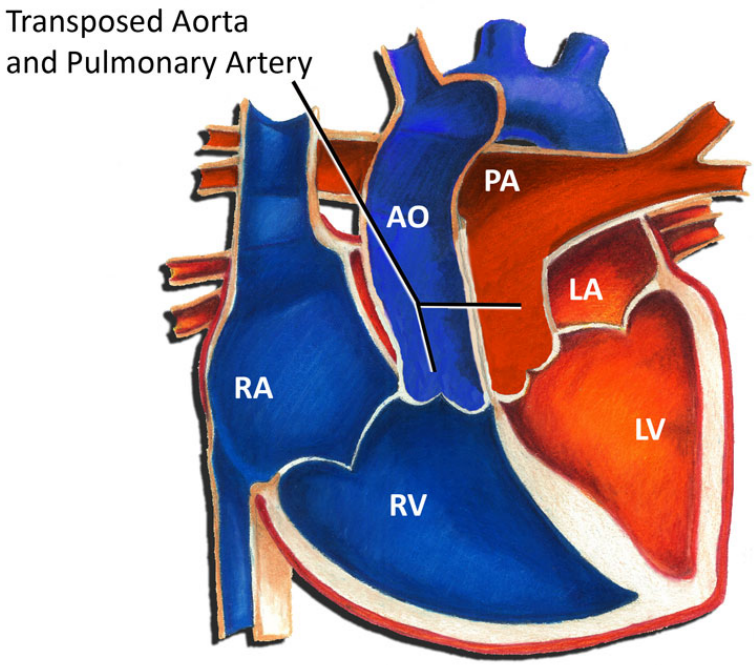
Normal Heart

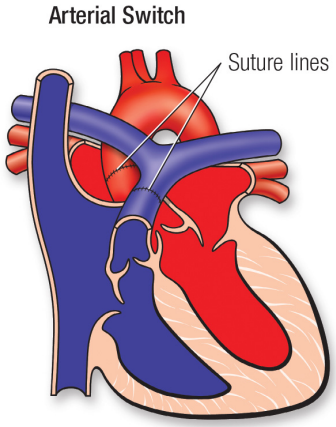




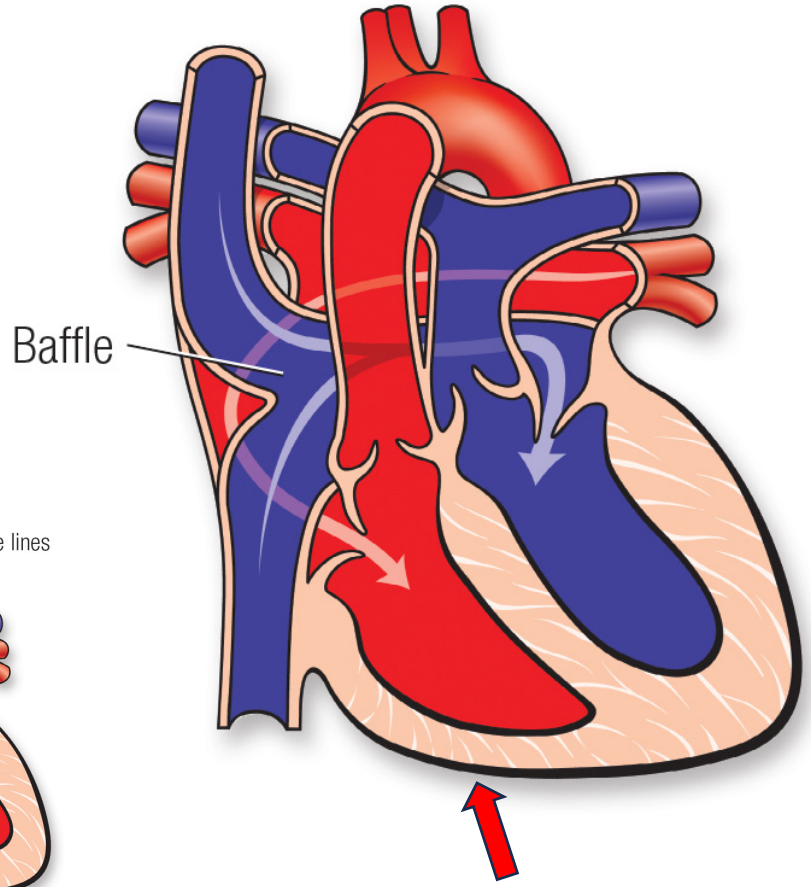
DTGA

Transposition of the Great Arteries





Intra-Atrial Baffle (Mustard or Senning Procedure)



Why does systemic RV differ from LV?

- Different ventricular anatomy
- LV is conical shaped, muscle fibers “squeeze” and wring blood out with force, and LV myocardium is compacted
- RV is tripartite shape, more like a reservoir (inflow, outflow, middle portion) and tissue is not as compact, more boggy myocardium
- Systemic RV will tend to dilate as well as hypertrophy, changing the ventricular geometry

Case 6

- 60 yo M with DTGA s/p Mustard palliation, systemic right ventricle with systolic dysfunction, atrial arrhythmias, sinus node dysfunction s/p dual chamber pacemaker
- Starts to develop worsening systolic heart failure with exertional intolerance, edema, oral diuretics, worsening VO2 max
- Off label decision for biventricular pacemaker upgrade, no data for this in systemic right ventricle, but extrapolated from adult literature
- Per literature, traditional CHF meds have no impact on EF (ACE inhibitors, beta blockers); can consider starting spironolactone, diuretics, SGLT2 inhibitors

Few Studies on Systemic RV

- Small patient population
- Variable surgical interventions (no patient is alike, time cyanotic, age at surgery, surgical technique was not as advanced for DTGA)
- Study about ACEi showed no benefit
- Arrhythmia is often a cause of death, intra atrial reentry tachycardia that will conduct 1:1 to ventricles causing decreased cardiac output
- No data to support AICD implantation for low EF like in LV systolic heart failure

Case 7

- 67 year old man with L transposition of the great arteries (ie, “congenitally corrected transposition), AV block s/p dual chamber pacemaker, presenting with chest pain
- Cocaine positive tox screen, clean coronaries on prior cath
- CXR with mild pulmonary edema
- EKG shows 3:1 atrial flutter, no ventricular rhythms on device check
- Has outpatient cardioversion, returns to sinus rhythm, hesitant to start beta blocker, does not desire amiodarone, started on anticoagulation
- Stayed out of arrhythmia short time, recurrent shortness of breath, found to be in AF again, has gained 10 lbs of volume
- Attempted repeat cardioversion, eventually ablation, went back in to atrial flutter, upper limit set on pacemaker, and volume status controlled with Lasix daily and anticoagulated
- Eventually has worsening shortness of breath around 70 years old, mental health issues, paranoid of going to hospital, taken by family, had biventricular failure likely from heart attack, not improving, suffered stroke, decision to withdraw care

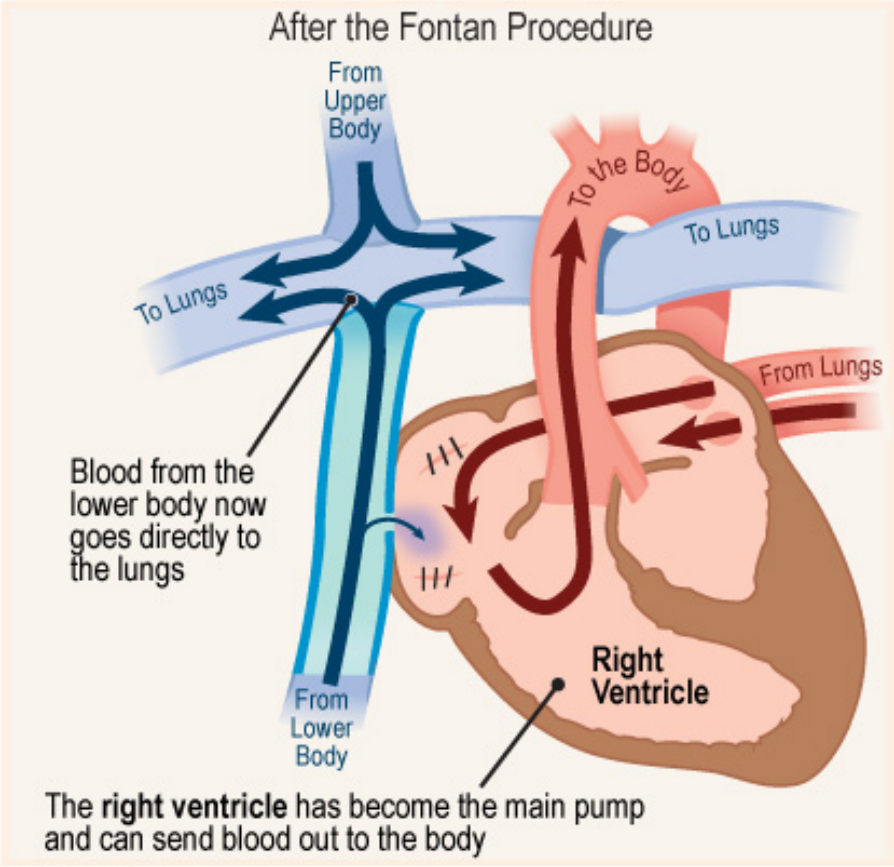
Treatment Options

- Beta blockers could worsen sinus node dysfunction in DTGA, careful monitoring when starting but will not change outcome, but if patient is able to mount good sinus response less concern for starting b-blocker
- Afterload reduction has not been shown to be helpful, start ACEi and ARB if you are treating hypertension, but not as GDMT
- Spironolactone might have some benefit but not much
- New studies ongoing about Farxiga/Jardiance currently, have started on patients but may not change much
- Diuretics
- Pacing
- Surgical interventions before ventricle fails (TR – which is like MR)
- Transplant

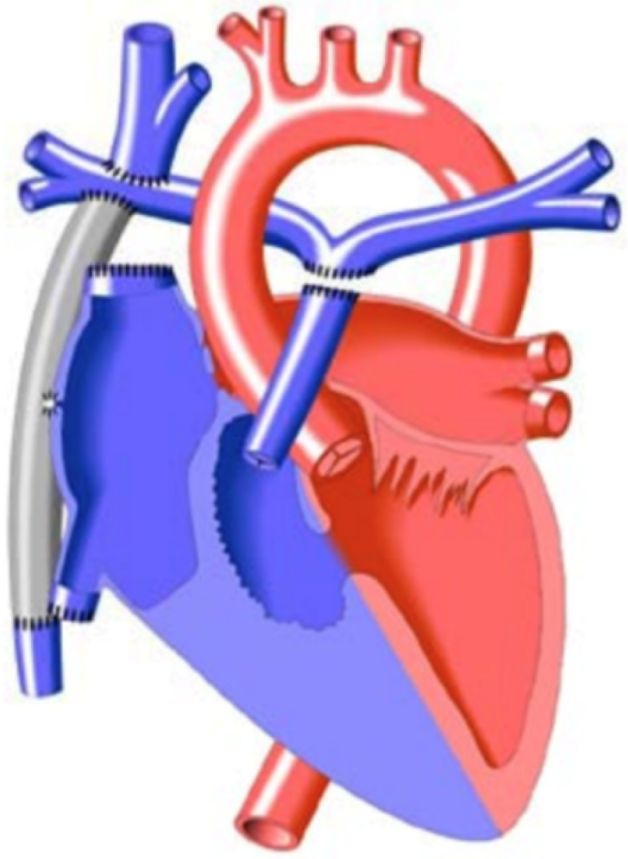
Single Ventricle Heart Failure

- Let's talk about the Fontan circulation
- The "right heart" is now a total cavopulmonary anastomosis
- This circulation is much more sensitive to PVR and EDP changes
- Need to maintain AV synchrony
- Inotropes may be needed! *Do not hesitate to add milrinone early!*
- If HR stuck at 100 without R to R variation, may be in IART
- Other oddities: older palliated single ventricle patients living off of an aorto pulmonary (BTT) shunt or Glenn circulation
- Single ventricles can include: tricuspid/pulmonary atresia, double inlet left ventricle, unbalanced atrioventricular canal defect, aortic/mitral atresia, double outlet right ventricle
- Fontan patients have liver disease from passive congestion (Fontan associated liver disease)
- Decisions regarding interventions are made in case conference discussions
- Do not do paracentesis! Will only cause patient to get worse, they already have dilated lymphatic in abdomen and draining abdomen will only cause re-accumulation and hasten the end of life

Fontan Physiology



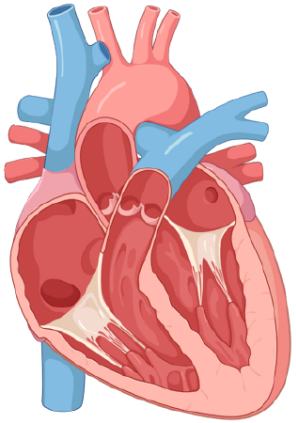
Dominant RV



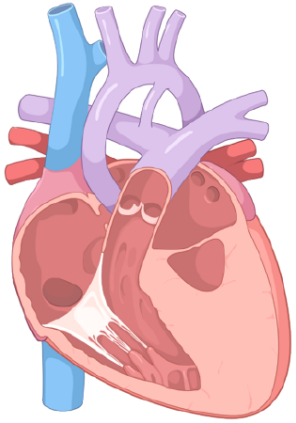
Dominant LV

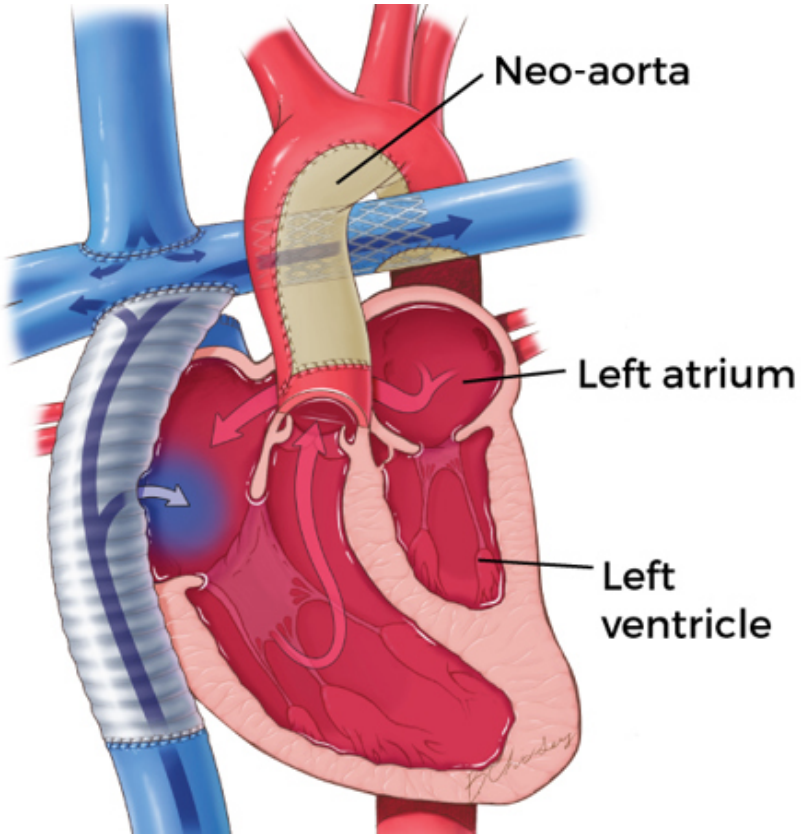
Hypoplastic Left Heart Syndrome

Normal Heart



HLHS Heart





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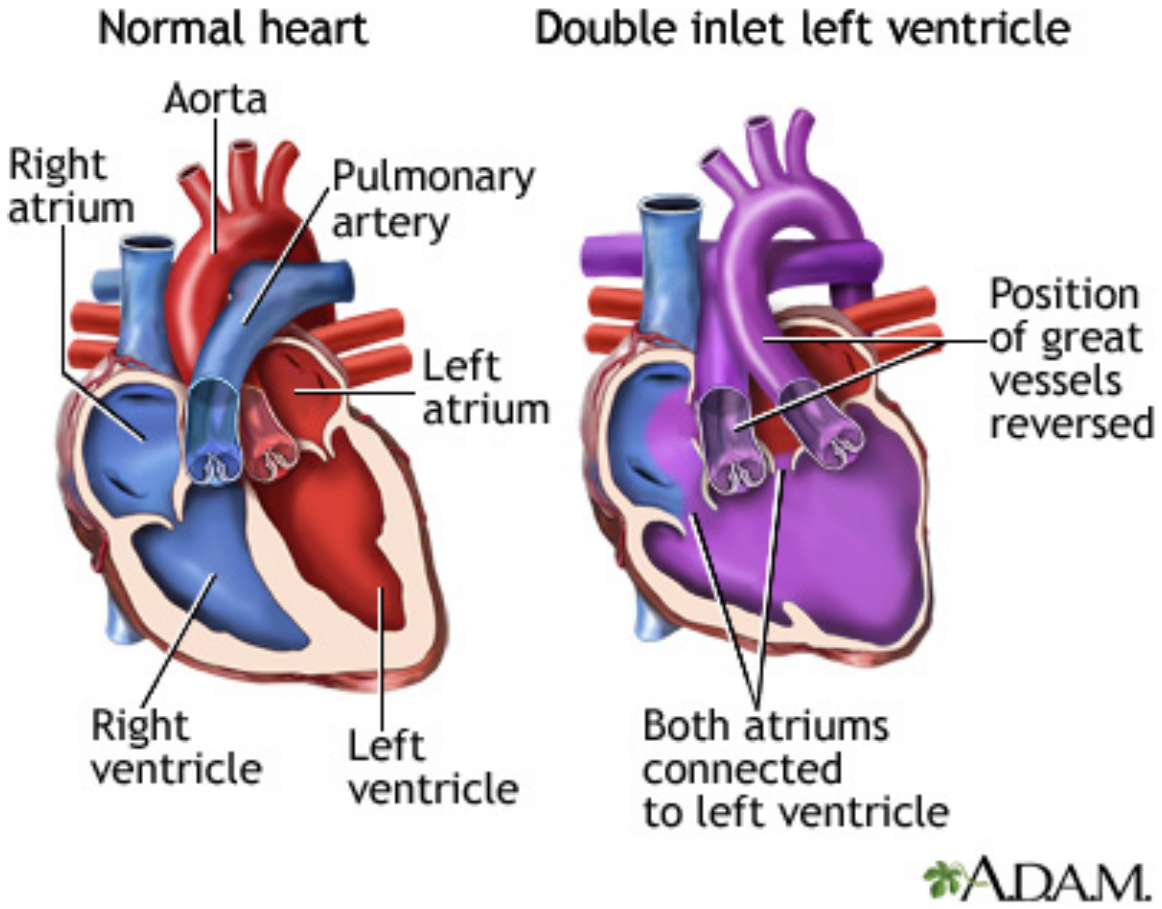
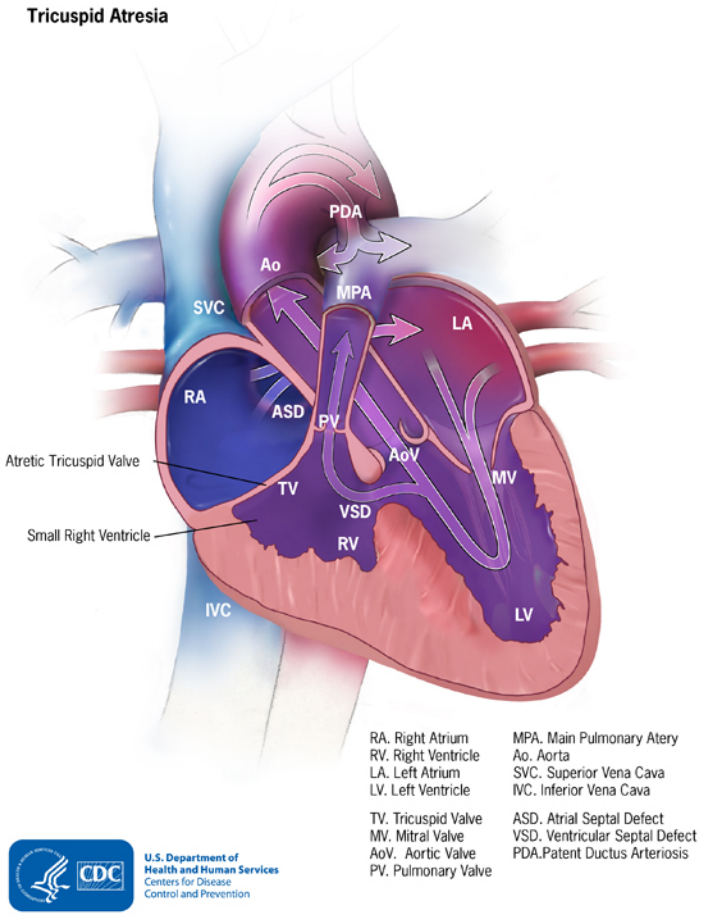

Normal heart
Double inlet left ventricle
Aorta
Right atrium
Pulmonary artery
Left atrium
Right ventricle
Left ventricle
Position of great vessels reversed
Both atriums connected to left ventricle
A.D.A.M.

Tricuspid Atresia



Case 8

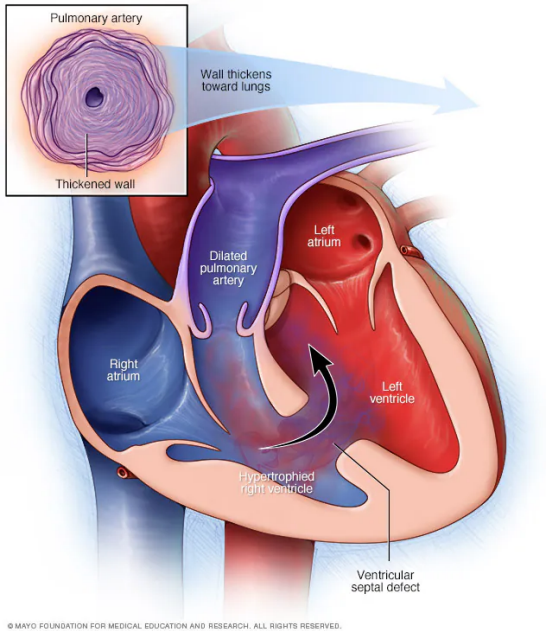
- 41 yo M with double inlet left ventricle s/p staged Fontan palliation (more than 1 surgery), pacemaker for sinus node dysfunction, got covid 8 months ago, started to get worsening fatigue
- Went to PCP, saw acute renal injury (Cr up to 1.8), edema in legs and mildly in abdomen, started Lasix
- Still accumulating volume, needed hospital admission for diuretics, did not need inotropes because no worsening AKI or liver issues
- RHC demonstrated increased Fontan pressures (22 mmHg like an “RA” pressure, in IVC, SVC, PA), normal PVR, increased LVEDP in the 20s
- Diagnosed with diastolic heart failure
- Put on diuretic regimen, renal function too poor for Entresto, tried SGLT2i but did not help, worsening edema despite addition of metolazone
- Discussed at other centers, now starting work up for transplant

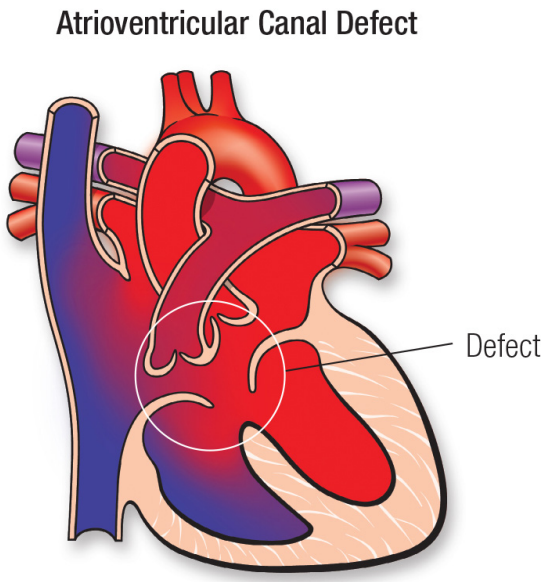
Case 9

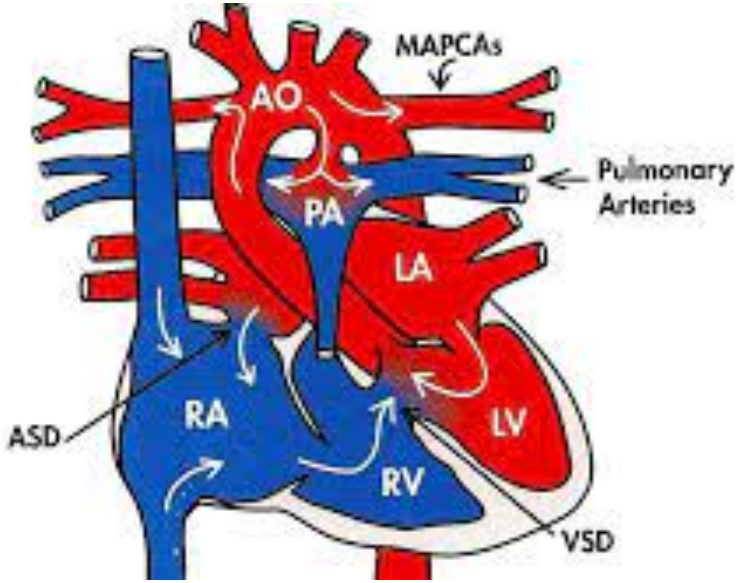
- 45 year old man with double inlet left ventricle, atretric tricuspid valve, s/p pacemaker for SSS, atrial fib, chronic anticoagulation, on carvedilol, Lasix, ramapril, spironolactone, Fontan associated liver disease (congestive hepatopathy and cirrhosis) presents with sudden shortness of breath, NYHA 4
- Severe edema in legs and abdomen on exam (baseline was NYHA 1)
- Echo demonstrates new acute mitral valve chordae rupture and severe mitral valve regurgitation
- Attempt diuresis with IV Bumex, worsening AKI, liver function
- Addition of inotropes with milrinone provides lucitropy and contractility and helps diurese, however not maintaining volume status on po Bumex
- Evaluated for surgical MVR but not candidate due to liver disease
- Novel approach for TEER (Mitraclip) x 3 by ACHD/structural cath team
- Improved MR to moderate, stable on oral diuretics outpatient, now NYHA 2

Eisenmenger's / Cyanotic Cases

- Unrepaired ASD, VSD, AV Canal
- Unrepaired TOF
- Unrepaired cyanotic disease
- Eisenmenger (shunt reversal due to PAH)







TOF with MAPCAS

Case 10

- 55 year old woman with unrepaired Tetralogy of Fallot with pulmonary atresia and MAPCAS, and thalidomide toxicity
- Starts developing worsening edema after trip in Spain where she did end up eating out and having some salt
- Admitted for IV diuresis, echo shows diastolic dysfunction
- Recurrent admissions for diuresis, oral diuretics, had renal impairment, attempted peritoneal dialysis for short time, no good transplant options (?heart lung), eventual discussion regarding palliative care and withdrawal of care inpatient due to acute heart failure

Case 11

- 64 year old man with unrepaired VSD and Eisenmenger, travelled from Michigan to Seattle by car to see family, ended up eating lots of salt, gaining 20 lbs on the way and had mild shortness of breath
- Admitted, started on milrinone gtt, got IV diuresis (care not to over diurese, need a little preload)
- Had RHC to check PA pressures and PVR, were elevated
- Started pulmonary hypertension medication, able to wean milrinone, and get on oral diuretic regimen
- Returned home without issues and followed back up with his cardiologist at Mayo Clinic

Case 12

- 44 year old man with single ventricle, tricuspid atresia, pulmonary hypertension, unable to have Fontan due to PAH, s/p bilateral BTT shunts (aorta to pulmonary artery shunt), with baseline cyanosis and SpO2 around 80%
- Develops worsening shortness of breath, exercise intolerance, decreased O2 saturation, echo shows LVEF now 40% from 60% last year
- Cardiac cath shows elevated PVR
- Started on pulmonary hypertension therapy
- Improved cardiac function and O2 saturation
- Eventually admitted with atrial fib/flutter with RVR, intubated for hypoxia, cardioverted in field, transferred here, started on amiodarone
- Better rhythm control helped improve cardiac status as well

CHF ACHD Caveats Summary

- ACHD patients are difficult to manage when unwell – think multi-facted cardiac system issues (rhythm, valves, function, diastology, etc)
- Understand the anatomy, check the scars, pulses, etc
- Think through the physiology (be aware they can have delicate physiology)
- Ask how will this medicine or treatment help the physiologic state?
- Consider inotropes in Fontan patients and avoid paracentesis
- Often dealing with low cardiac output states, not traditional systolic ventricular failure (HFpEF, HFrEF, low CO state, RV failure, LV failure, both)
- May not have classic signs of heart failure like increased JVP, crackles, extremity edema due to lack of L heart failure symptoms
- Do not delay interventions if needed (valves, stenting, etc)
- Higher level care should be given in ACHA accredited center
- Co management with CHF and ACHD is appropriate and helpful
- Not often related to coronary disease due to age, but still important to screen for based on other risk factors
- Traditional left heart failure meds may not work for systemic RV
- Consider palliative care discussions when needed in ACHD

Resources

- www.achaheart.org
- www.pted.org
- 2018 ACC ACHD guidelines
- ACC heart failure guidelines
- HRS/PACES guidelines
- ACC valve guidelines
- M Gatzoulis, *Diagnosis and Management of ACHD*

The End


"I'm the doctor who brings
the cards. I'm a cardiologist."
GET
WELL