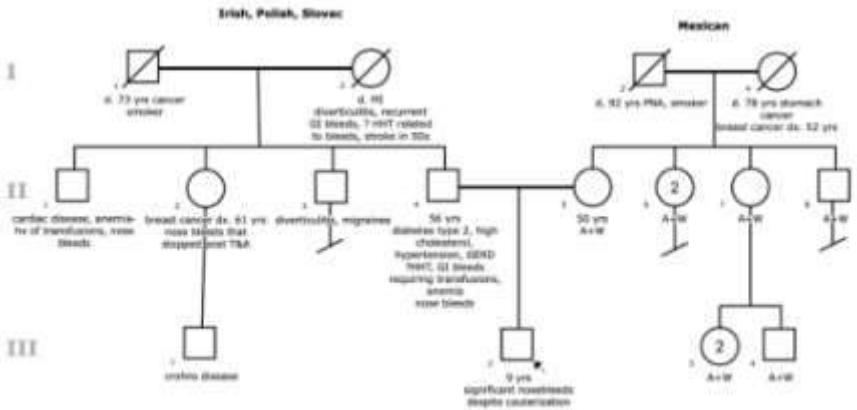
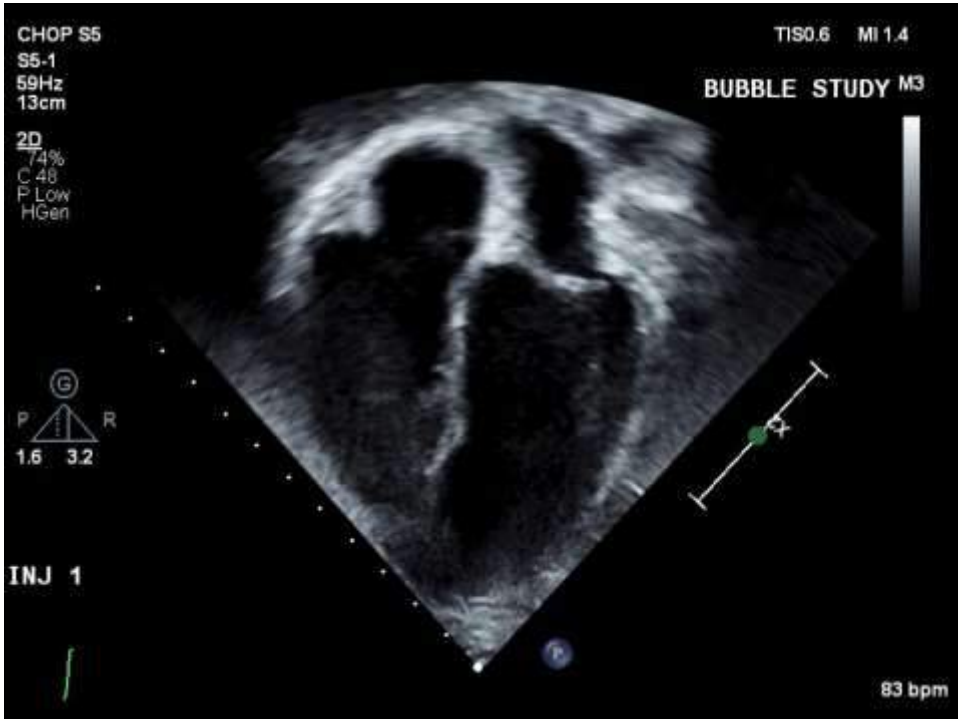
CASE PRESENTATIONS

- Patient JD:
  - Referred with new ENG-related HHT
  - Started with nosebleeds at 4 yrs but have gotten worse, now 1-3x/week
  - Possible telangiectasia
  - Father had severe GI bleeding warranting transfusions and colonoscopy but never tested for HHT or screened for other AVM.
  - JD had no cardiorespiratory sxs
  - Pulse Ox 100%
  - TTCE performed: management?

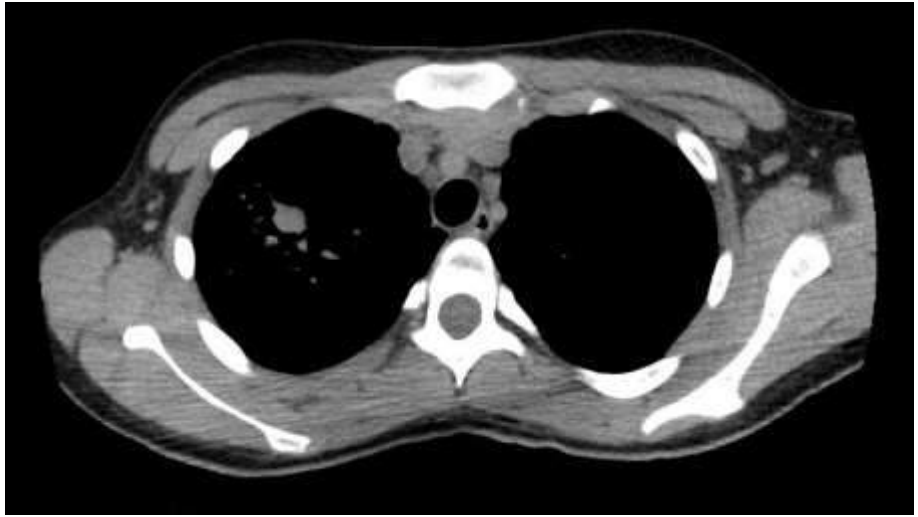


CASE PRESENTATIONS



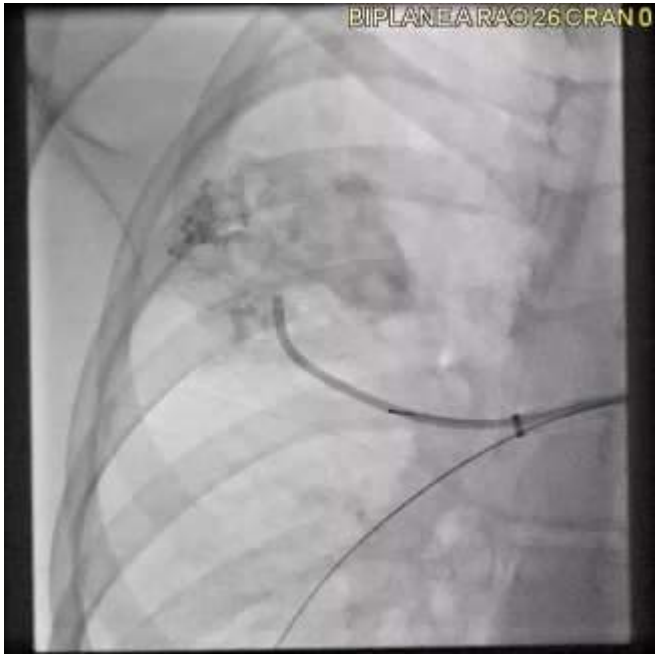
CASE PRESENTATIONS

- Patient VR
  - Presented with Covid and hypoxia (87%) but never resolved (90%)
  - CT scan performed
  - Occasional epistaxis
  - No other HHT symptoms
  - In retrospect, extensive FHx but…
    - Mother and MGM asymptomatic
    - Maternal GGM epistaxis and two strokes
    - Maternal great aunts had early strokes
  - Genetic testing: ENG variant
  - Management?



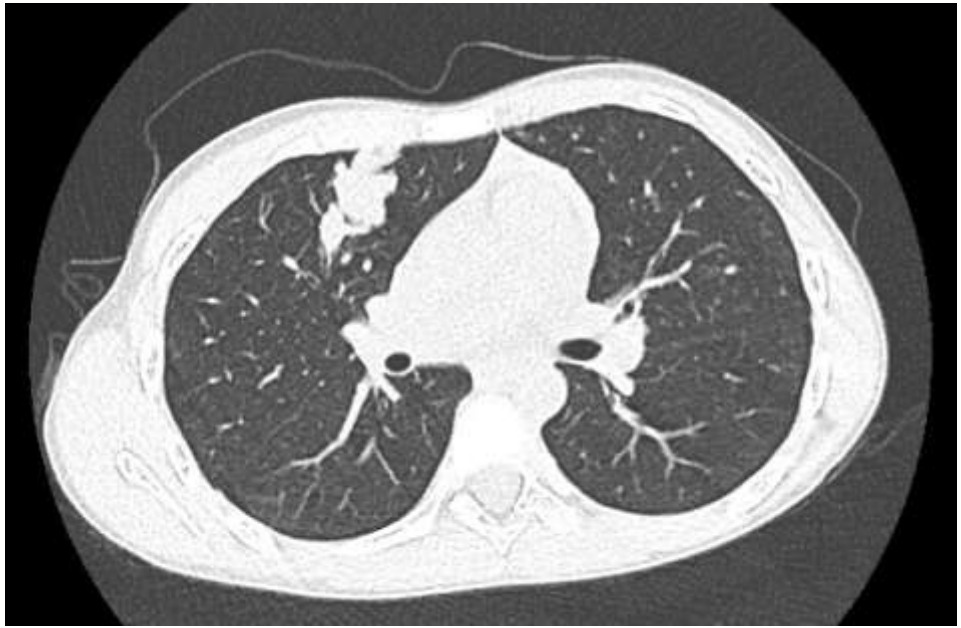
CASE PRESENTATIONS

- Patient VR



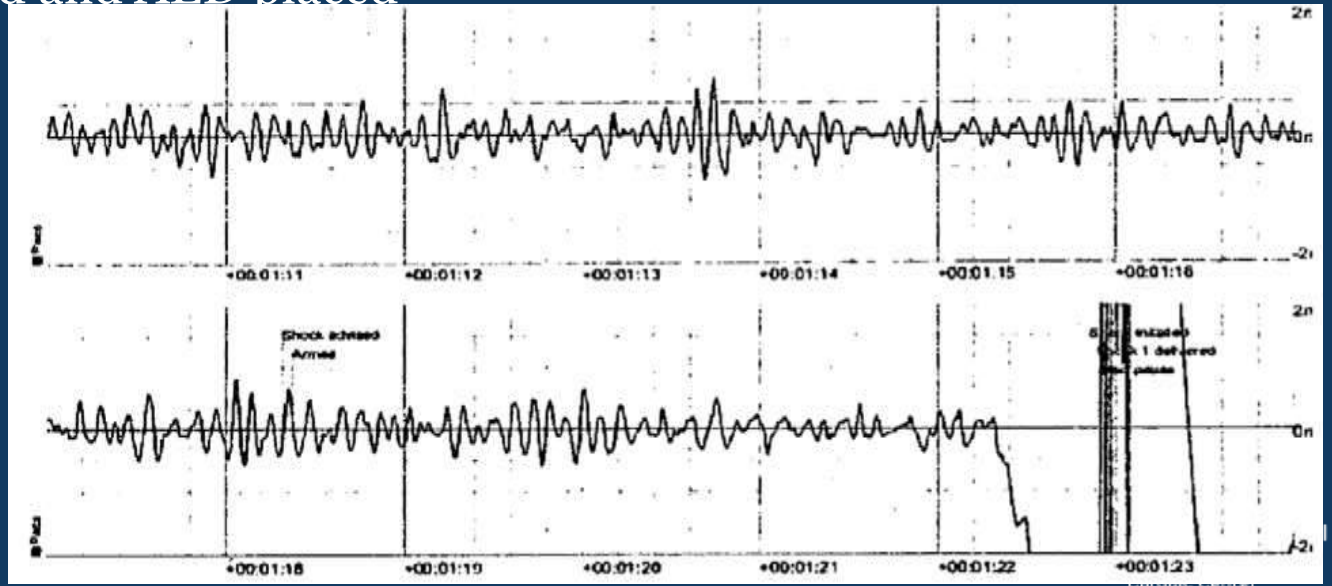
CASE PRESENTATION

- Patient FIO
  - Presented with hypoxia and clubbing with Covid
  - CT scan revealed PAVM
  - Epistaxis 1-2/month
  - No other HHT symptoms
  - Genetic testing: VUS in ENG
  - Mother also tested positive and found to have epistaxis and telangiectasia
  - Variant reclassified to likely pathologic
  - Management?



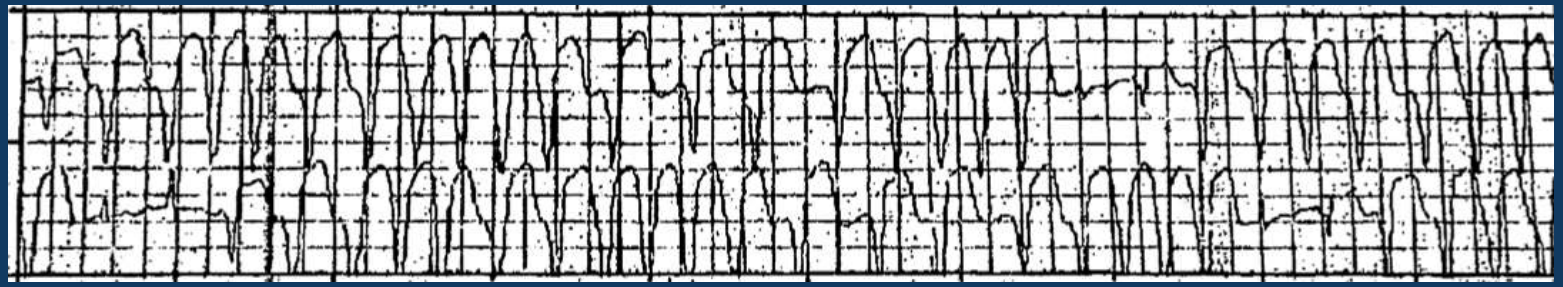
LVNC Case Presentation

- 14 year old previously healthy male presented after collapsing in school
- CPR initiated and AED placed



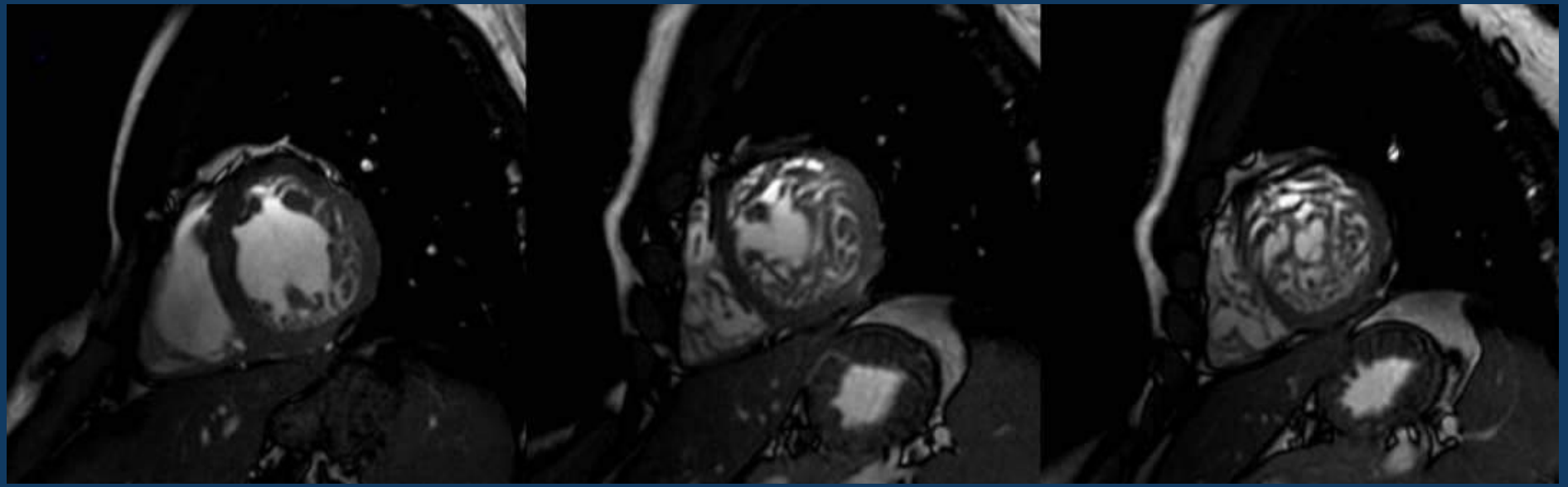
LVNC Case Presentation

- En route, noted to have both wide complex tachycardia and narrow complex tachycardia which converted with adenosine x2
- Recurrent narrow complex tachycardia (SVT) which degenerated into atrial fibrillation with rapid conduction requiring cardioversion

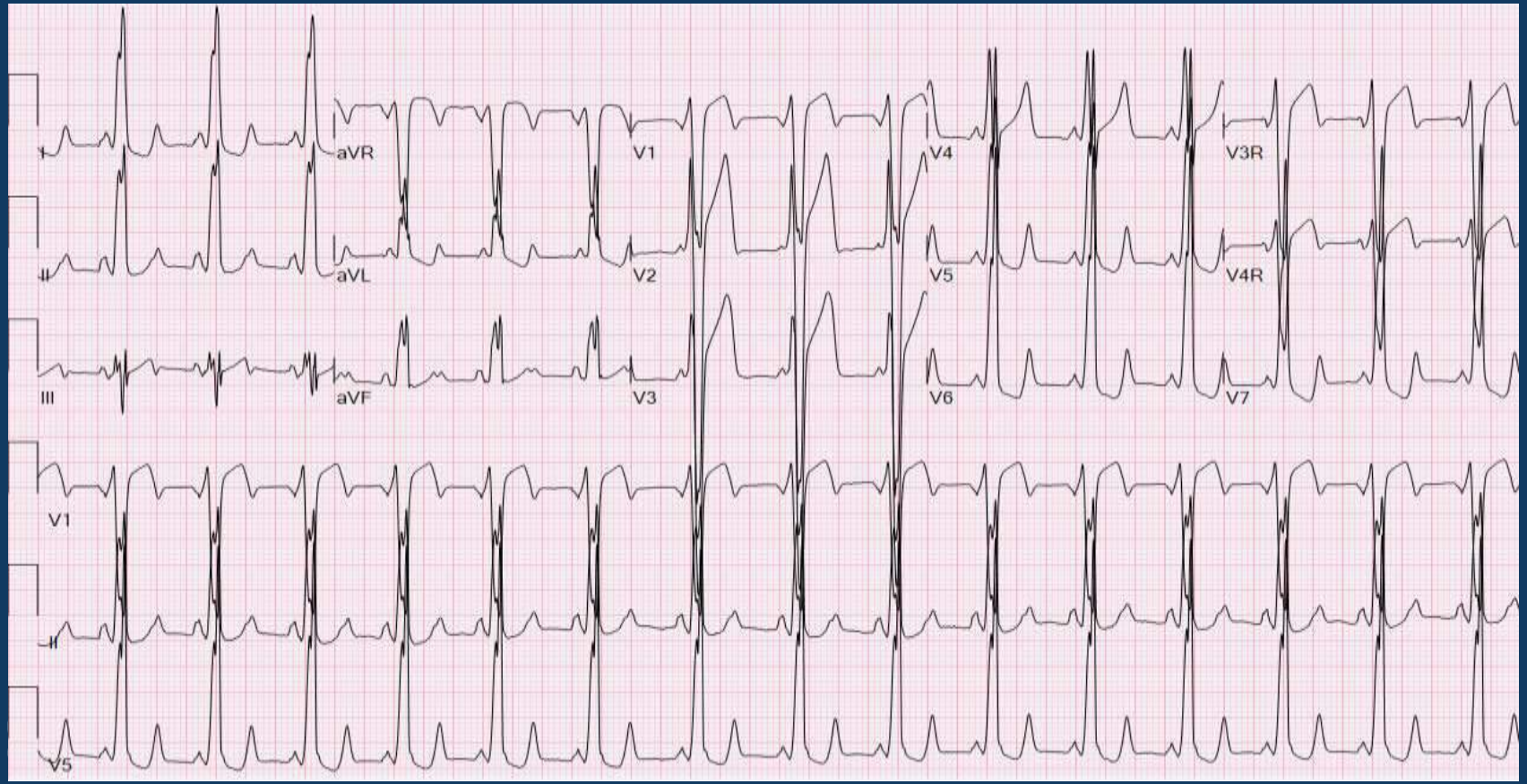


- Echocardiogram on presentation showed prominent trabeculations with normal biventricular size and mildly depressed function

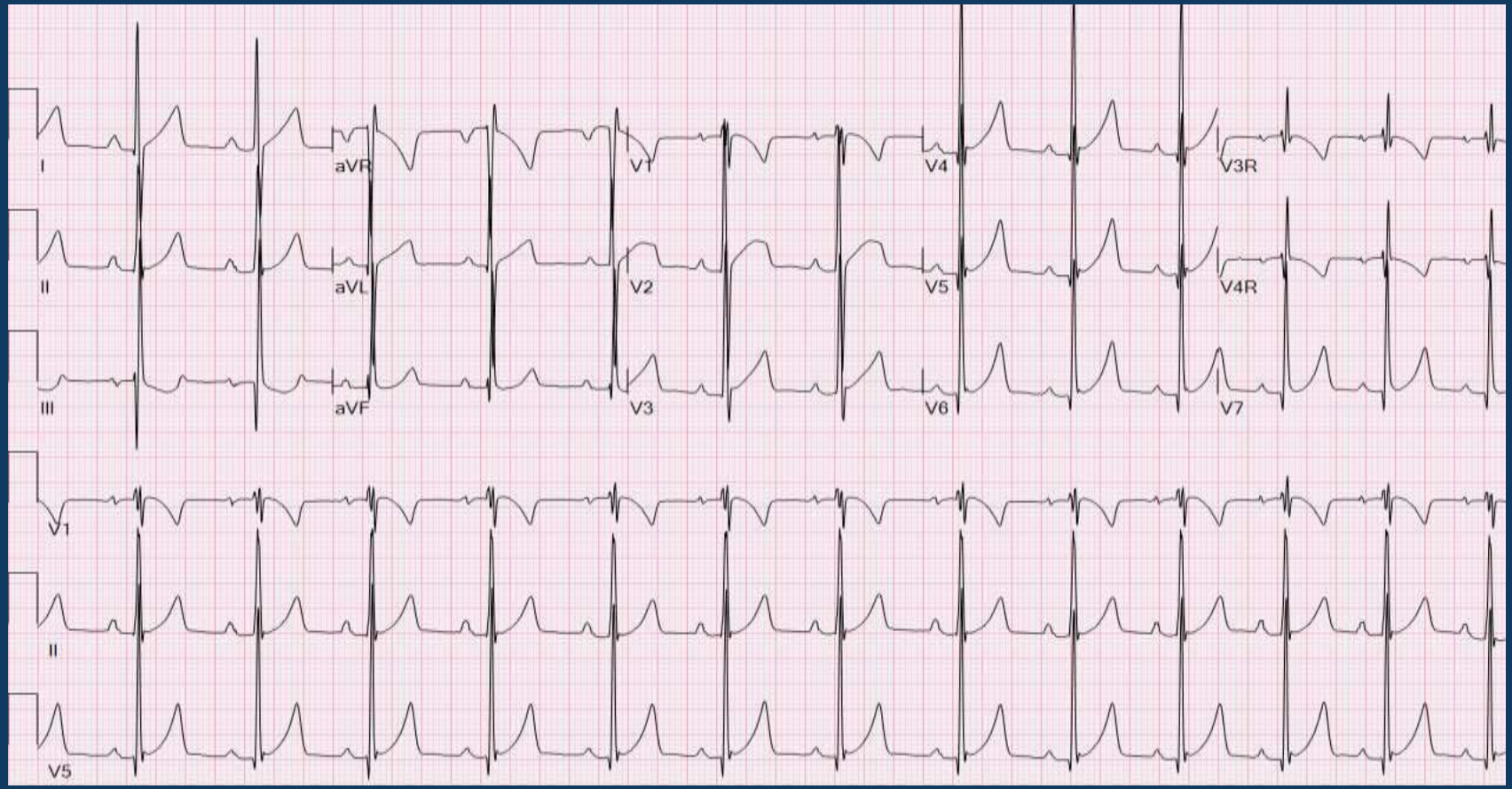
LVNC Case Presentation



LVNC Case Presentation

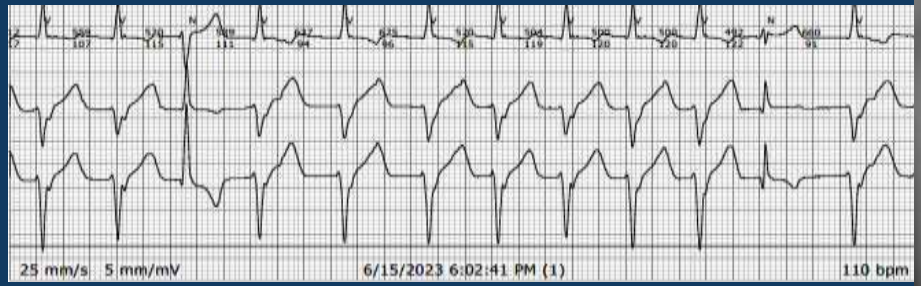


LVNC Case Presentation

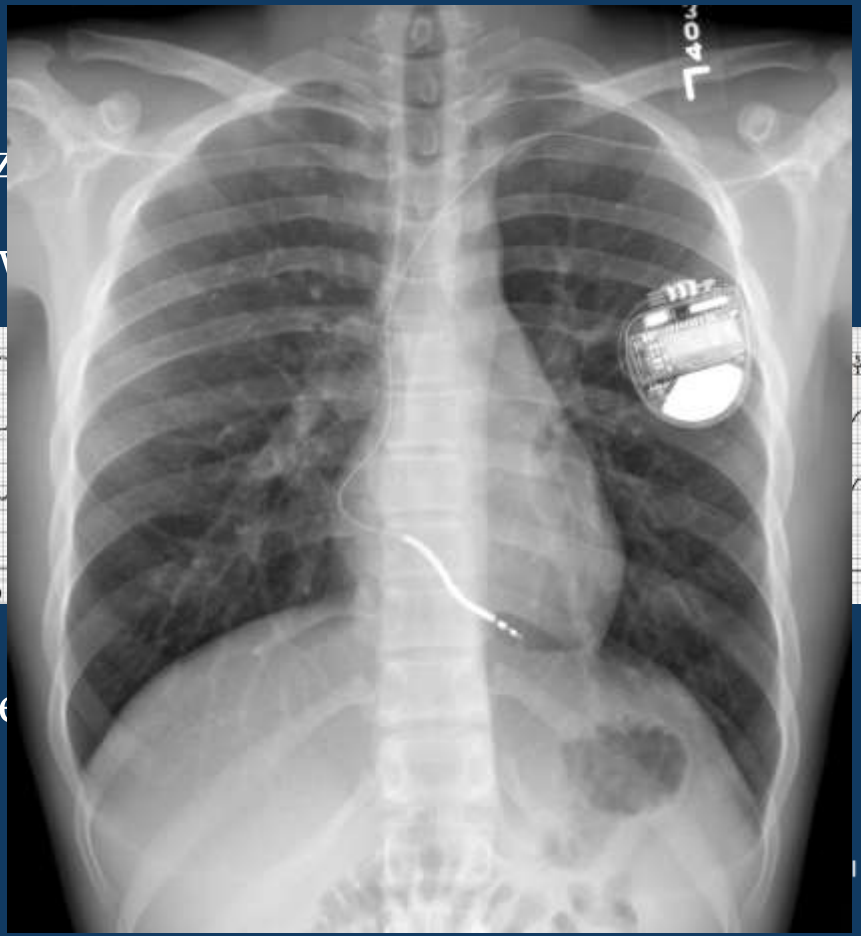


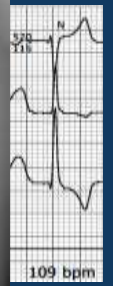
LVNC Case Presentation

- Systolic cardiac function normaliz
- Presented at the age of 17 with nev



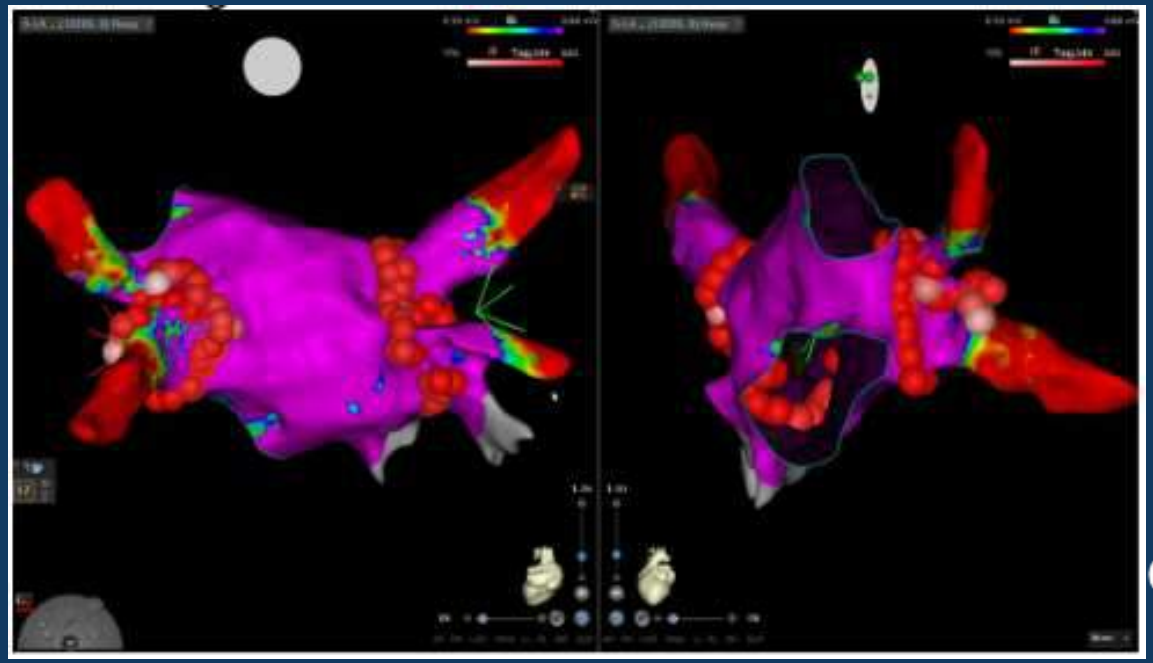
- Echo showed moderately depresse





LVNC Case Presentation

- Goal directed medical therapy → systolic function stabilized
- Inappropriate shock at age 21 and noted to have diastolic dysfunction



LVNC Case Presentation

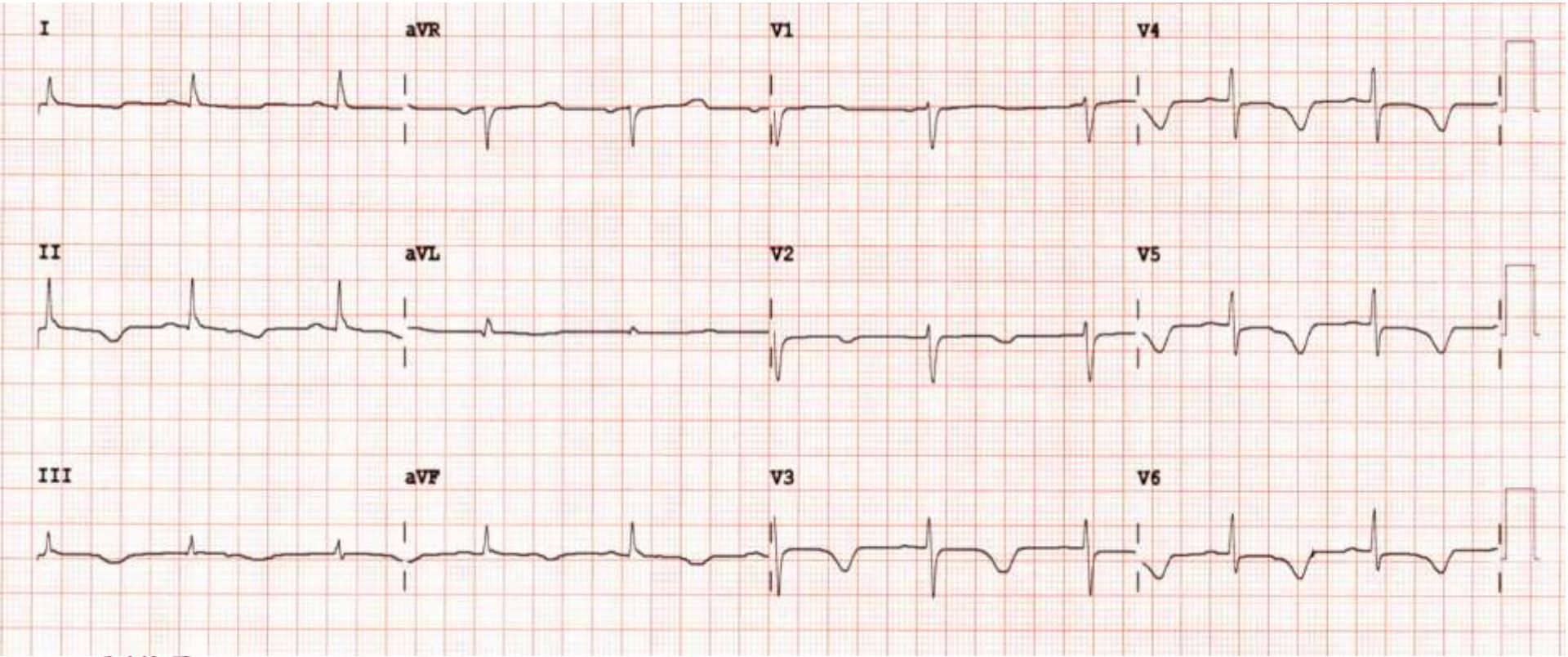
- Currently managed as outpatient with goal directed medical therapy by our adult congenital heart failure service (Toprol XL, Lisinopril, Eliquis)
- Genetic testing showed VUS in TTN and HCN4
- Being monitored for progressive restrictive changes
- Doing clinically well – working at Zara

Contemporary Approach to LQT

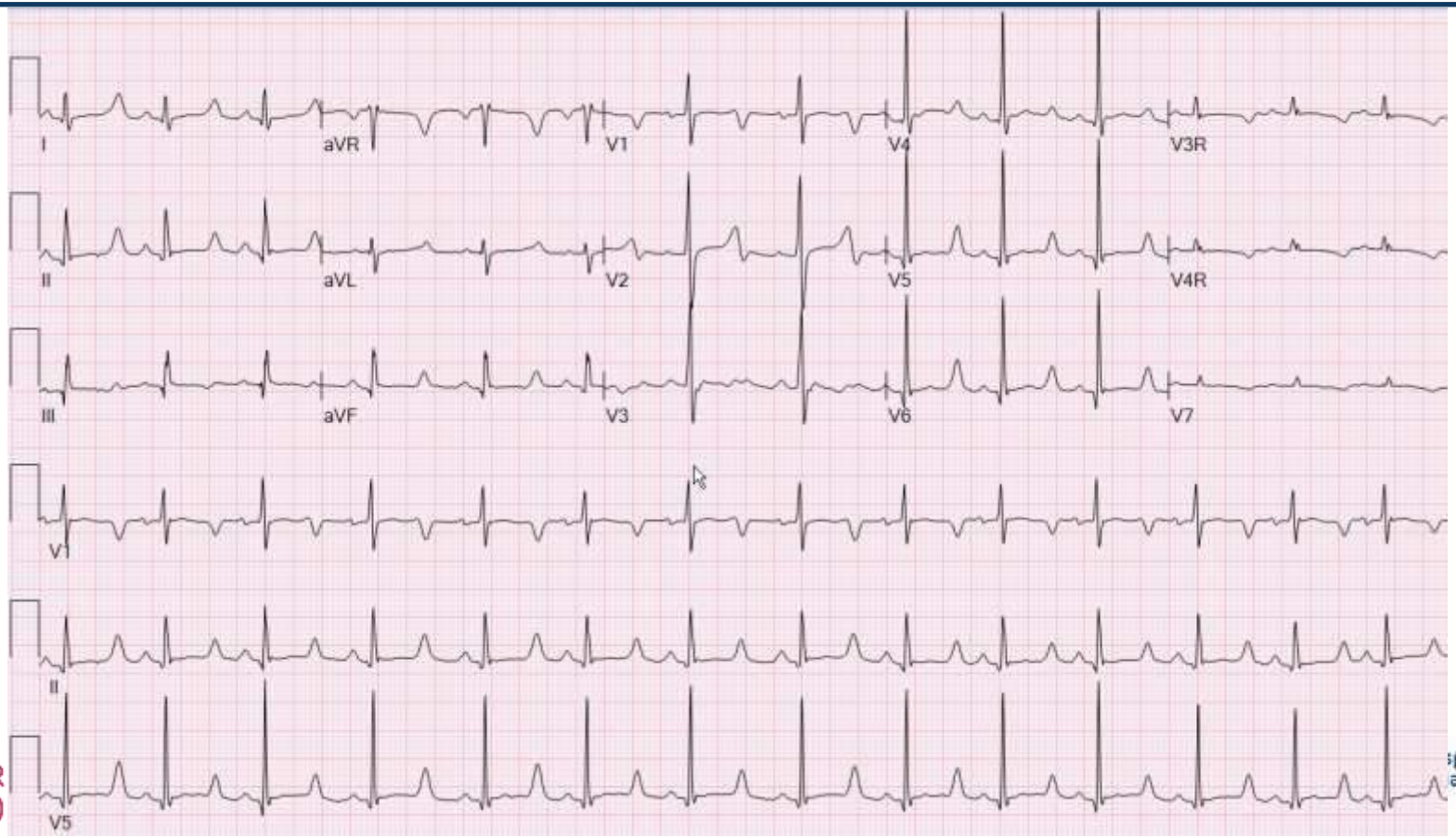
Case #3: An 11-year-old boy ("Johnny") presents with a family history of LQTS; the proband (mother) presented with an aborted cardiac arrest in the middle of the night as an adolescent. Mom has an ICD. Her QTc is 560 msec. She has an ICD, but it has never gone "off".
Johnny has a QTc of 520 msec and is asymptomatic. Gene test confirms LQT3 (same mutation).

What do you want to do now?

MOM's EKG

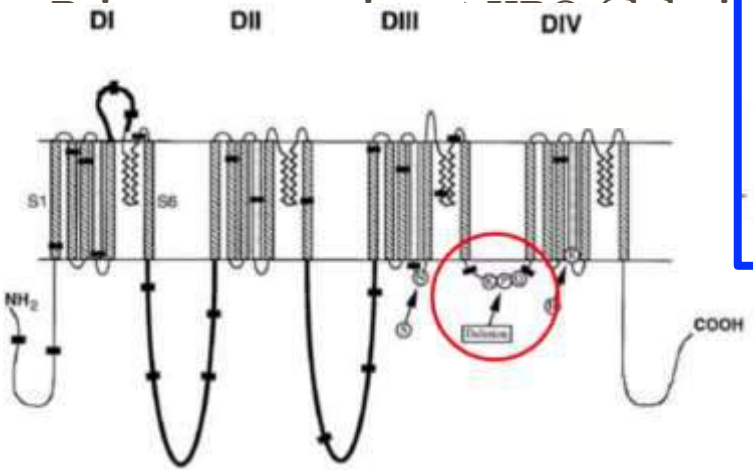


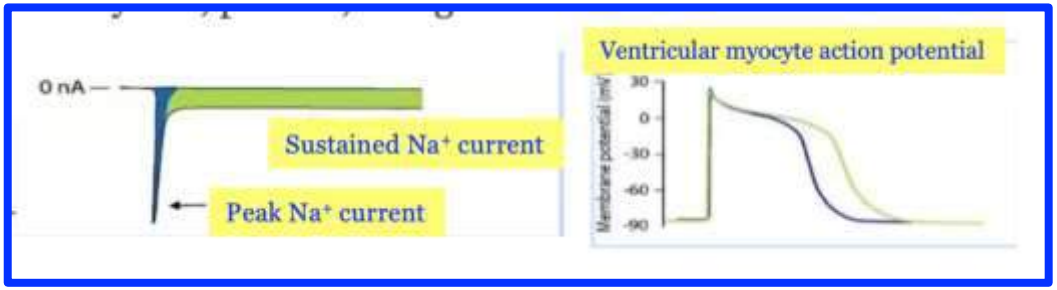
Johnny's EKG



LQT III

- LQT III autosomal dominant disease
- Mutation affects the inactivation of the sodium channel and causes a gain of function





- Cardiac events occur rest or inactivity
- ≅ 10% of cases of SIDS
- First cardiac event tends to be more lethal than LQT1 and LQT2 in children

Circulation. 2016;134:872–882.
J Am Coll Cardiol. 2003;42:103–109.

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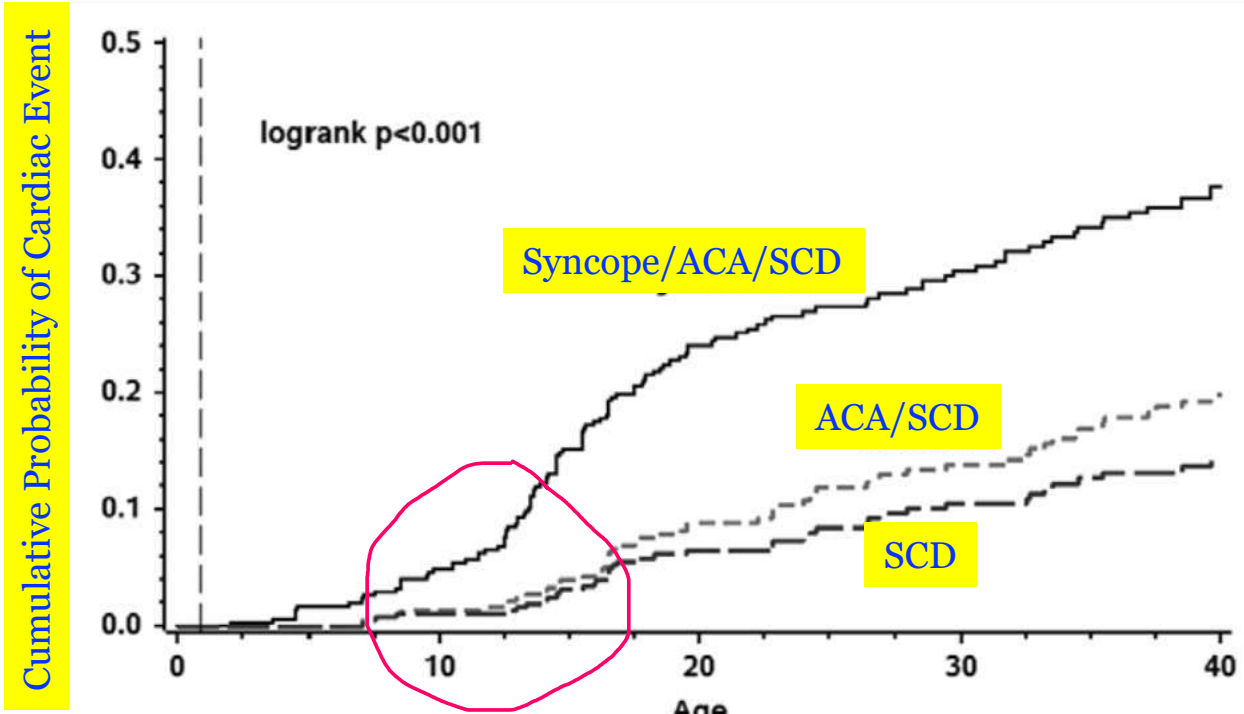
LQT III



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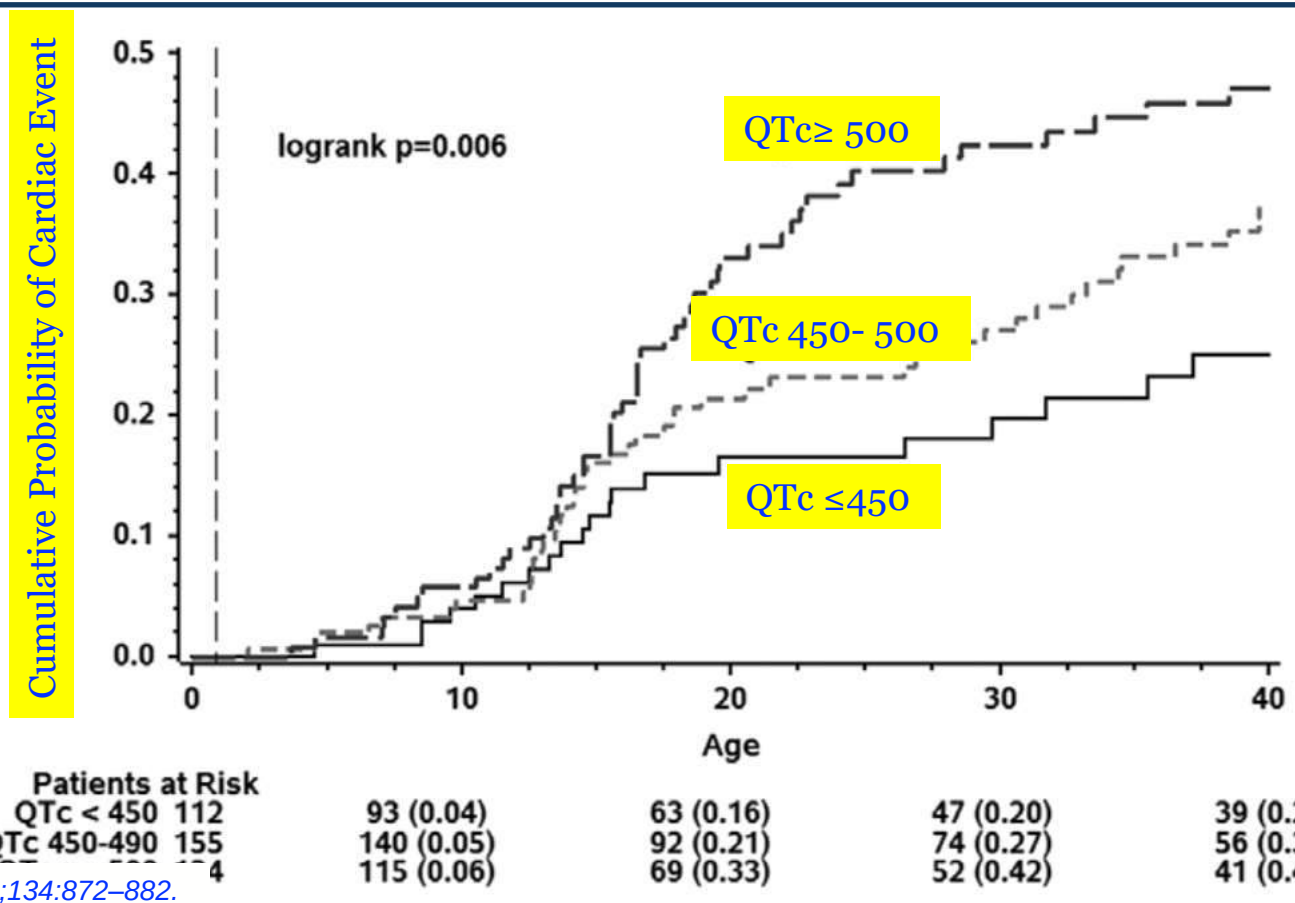
LQT III



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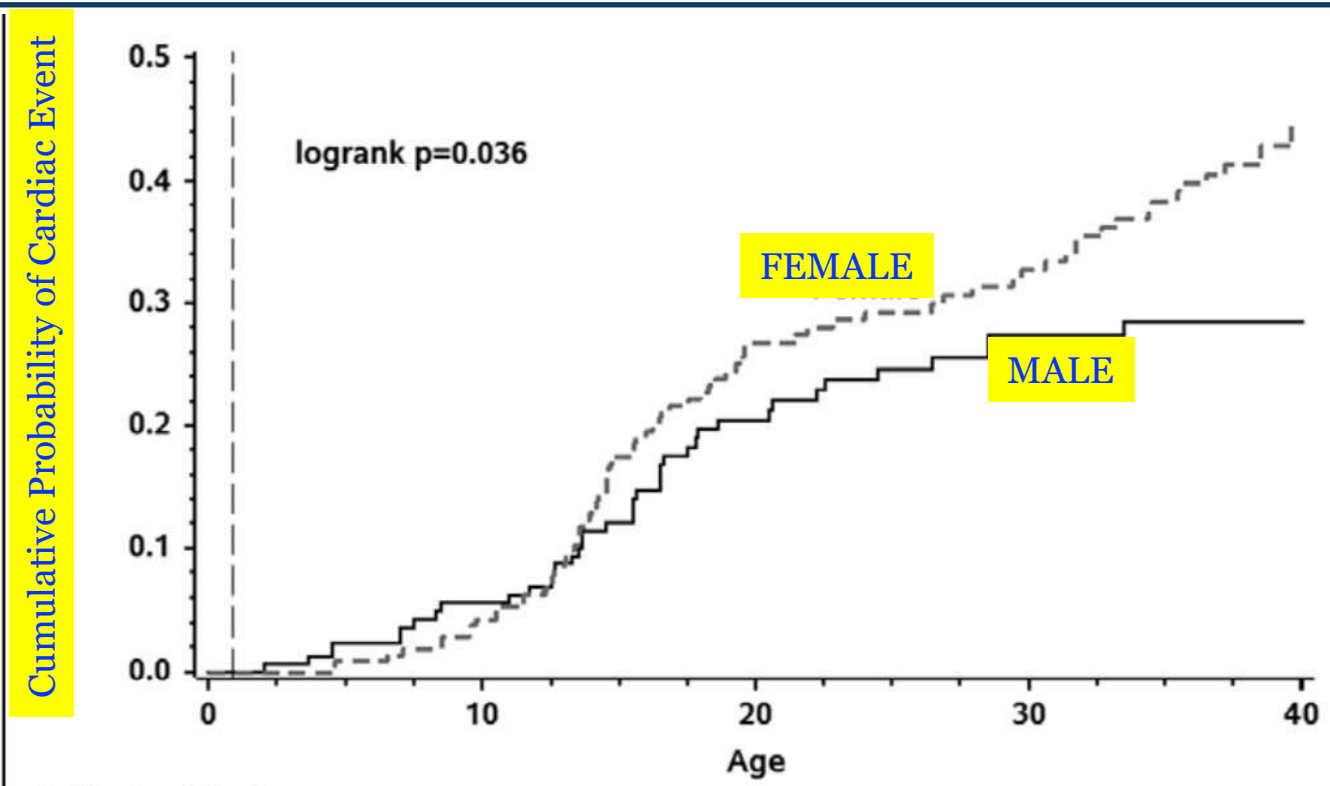
LQT III



Circulation. 2016;134:872–882.

Multivariate cox Model analyses for risk of cardiac events: First Acute Cardiac Event or LQT3 Related Sudden Cardiac Death

Parameter	*P* Value	Hazard Ratio	95% Confidence Interval	
			LCL	UCL
Syncope	0.023	2.03	1.10	3.72
β-Blockers among females*	0.032	0.20	0.05	0.87
β-Blockers among males*	0.308	0.51	0.14	1.88
E1784K mutation	0.001	0.09	0.02	0.37
D1790G mutation	0.049	0.30	0.09	0.99
QTc per 10 ms (up to 500 ms)	<0.001	1.33	1.19	1.48
Year of birth (after 1955)	<0.001	1.06	1.03	1.09

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Circulation. 2016;134:872–882.

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Gene-Specific Therapy With Mexiletine Reduces Arrhythmic Events in Patients With Long QT Syndrome Type 3

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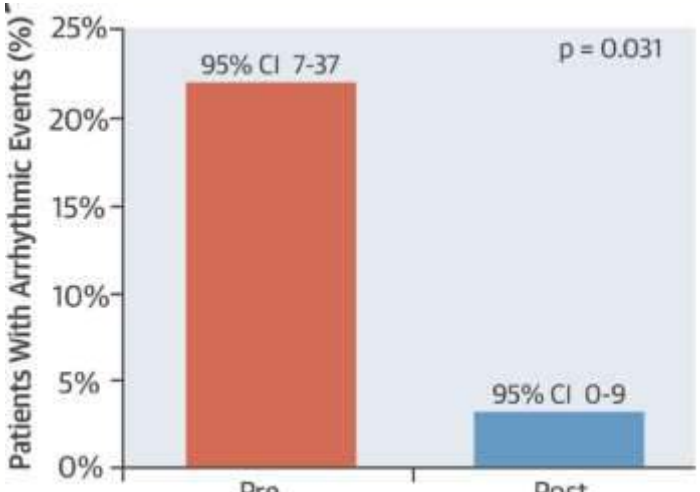

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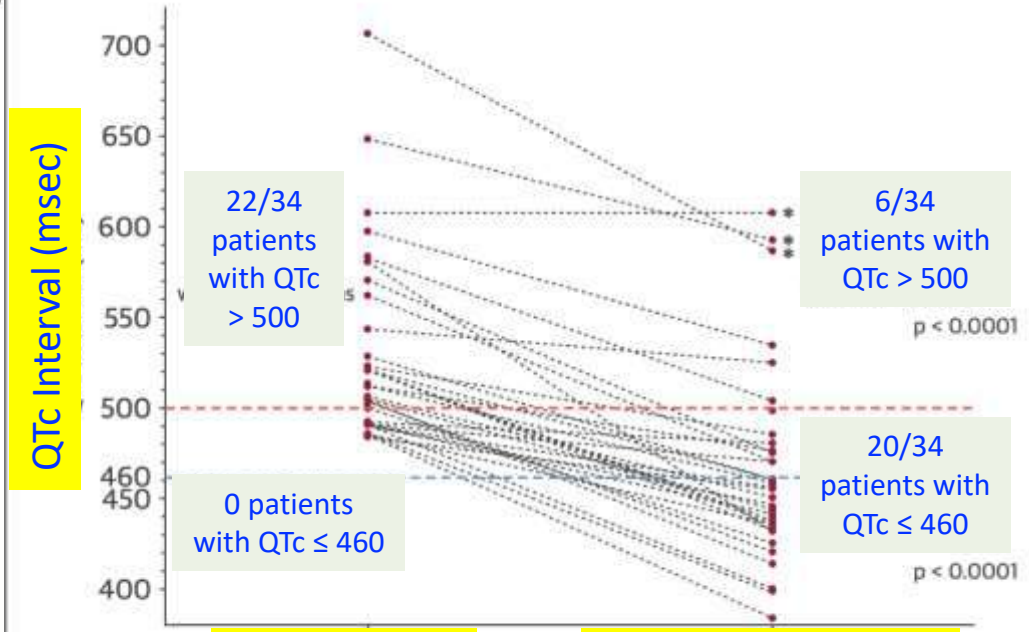
Effect of Mexiletine on QTc Interval Values

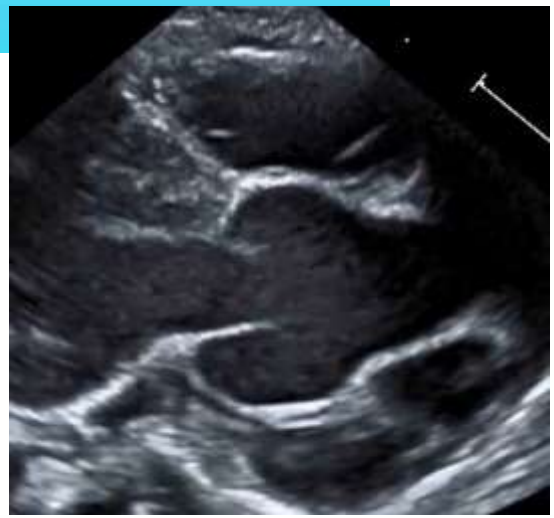


LQT3

- **Events tend to happen during rest/sleep/inactivity**
- Beta-blockers (still a role females>males)
- May be a role for atrial pacing to prevent bradycardia, limit pauses at night without an ICD.
  - Holters to look for bradycardia at night is important
- Consider mexiletine or flecainide (must prove it)
  - If flecainide watch for BrS pattern
- If syncope on beta-blockers likely ICD
- No exercise restrictions (still be smart)
- Avoid QT prolonging drugs
-

Case Presentation: Aortopathy

- Prenatal-
  - Club foot and thumb deformity
- Post natal-
  - Aortic root dilation on screening echo
- Genetics evaluation (10 y.o.)
  - Pectus carinatum
  - Hypertelorism
  - Bifid uvula
  - Micrognathia
  - Hypermobility



Loeys Dietz Syndrome

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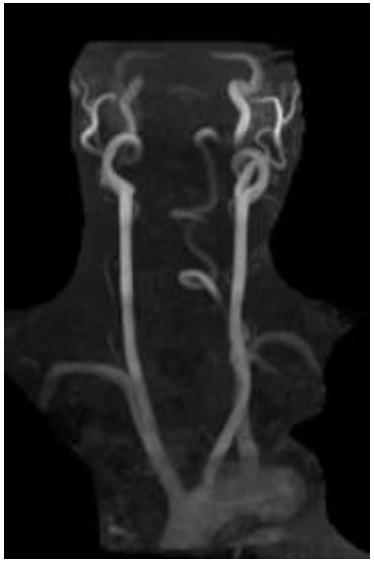
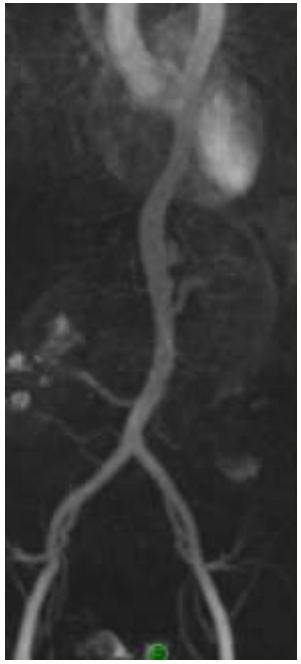
Risk Factors

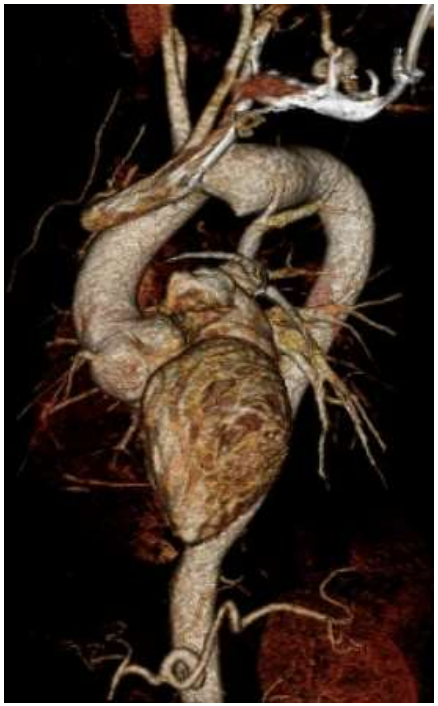
Gene involved- TGFBR2

MRA





CTA



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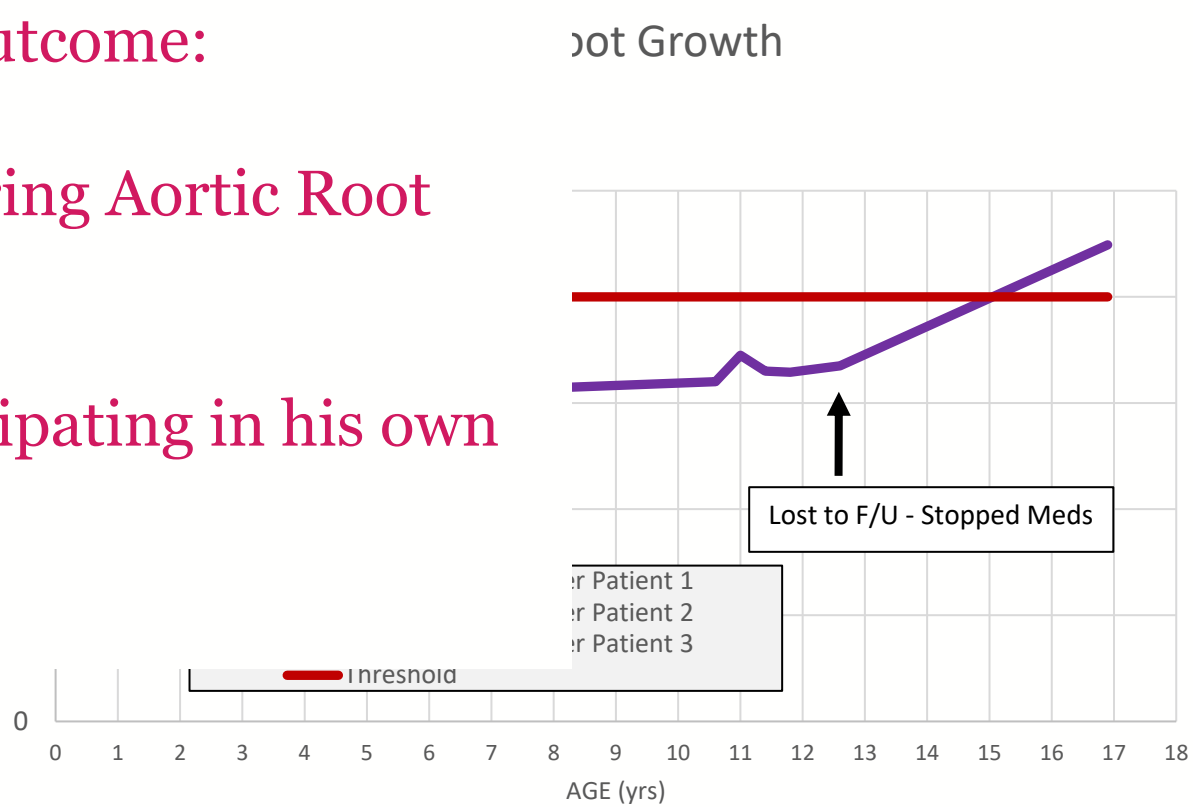
Aortic Diameter

Outcome:

s/p Valve Sparing Aortic Root Replacement

Actively participating in his own medical care



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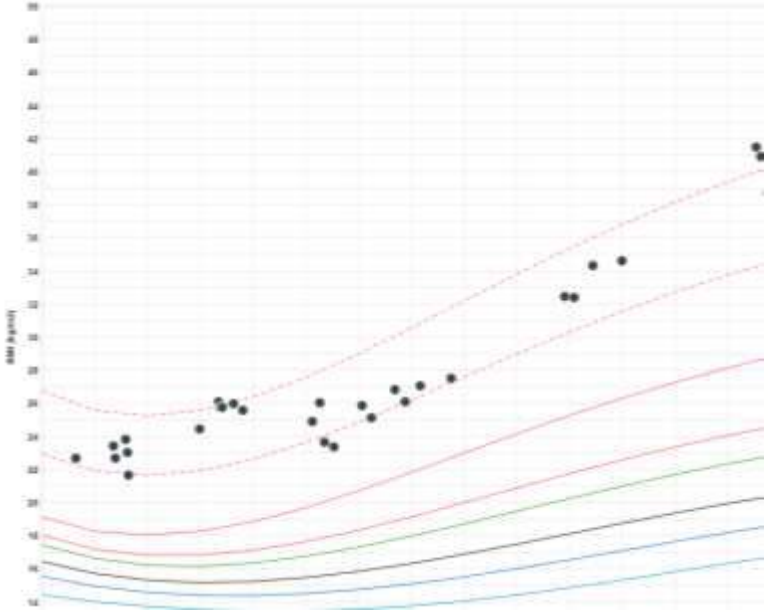
CASE: BENEFITS OF OBESITY MANAGEMENT

17 year old referred for dyslipidemia, obesity and elevated BP



SCREENING CASE #1

- Birth history (GA <37 weeks (premature birth), pregnancy related complications (pre-eclampsia, gestational hypertension, gestational diabetes, birth weight)
- Early childhood history: breastfeeding and duration, introduction of solids, periods of rapid weight gain
- Health behaviors: Diet, exercise, social structure (meals/who cooks/composition), sleep, smoking
- Other conditions (e.g. poorly controlled asthma, PCOS, h/o cancer, transplant, diabetes, inflammatory conditions)
- Medications (e.g., steroid, ADHD)
- Family history (HTN, HC, kidney disease, diabetes, premature cardiovascular/coronary artery disease: <55 years in males, <65 years in females)
- Vitals (BMI and BMI trend, BP (manual +/- ABPM))
- Labs (Lipid panel, CMP, A1c, TSH, free T4)
- Additional testing: echocardiogram

- Birth history: 37 weeks, **gestational diabetes and maternal obesity**
- Early childhood history: bottle fed; introduction of solids at 6 months; **rapid weight gain ~5 years of age** (more sugar sweetened beverage intake)

Health Behaviors:

- Diet: **skips breakfast**, family starting to prepare more meals at home; limited V/F
- Exercise: **limited exercise**
- No smoking or vaping
- Sleep: stays up late playing video games (**<6hours per night +habitual snoring**); sleep study pending
- Other conditions: none
- Medications: previously took Vyvanse
- Family history: mother with HC, HTN, obesity, preDM; **+FHx PCAD**

Health Factors:

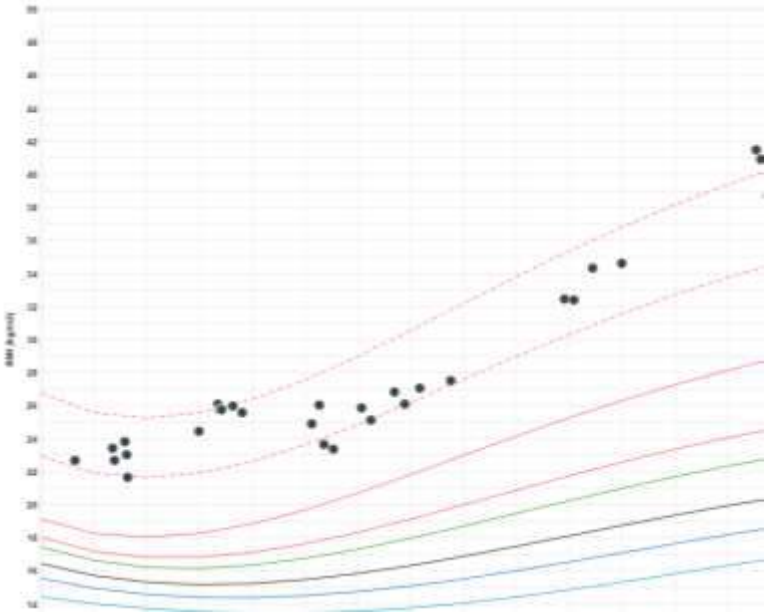
Vitals: BMI 42, **BP 126/60** (no antihypertensive meds); ABPM not available due to arm circumference

- **Labs: TC-C 184, TG 340, HDL-C 27, LDL-C 140; non-HDL 157**; **HgbA1c 5.8%**; **AST 43; ALT 40**

Imaging: concentric remodeling (relative thickness: 2*LVPWd/LVEDd >0.42), LVMI 40g/m^2.7. Normal function

COUNSELING AND TREATMENT CASE #1

17 year old referred for dyslipidemia, obesity and elevated BP



- Severe obesity
- Elevated blood pressure (possible masked hypertension)
- Combined dyslipidemia
- Possible sleep disordered breathing/obstructive sleep apnea
- Pre-diabetes
- Concentric remodeling

CVH score: **poor** 32 (Diet 0, PA 0, Sleep 20, Smoke 100; BMI 0, BG 60, BP 75, Lipids 20)

Risk Stratification: **high risk**

CKM: Stage2

INTERVENTION

Diet and exercise counseling (nutrition and weight management); Portion/ decrease carbs (1/4), omega-3 enriched diet

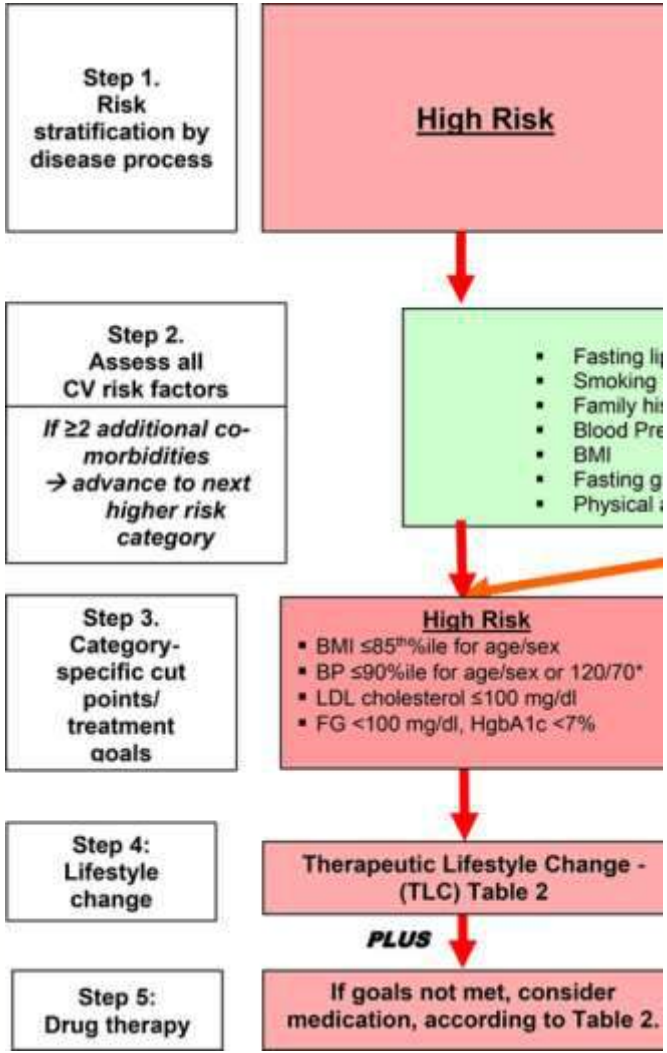
Psychology counseling

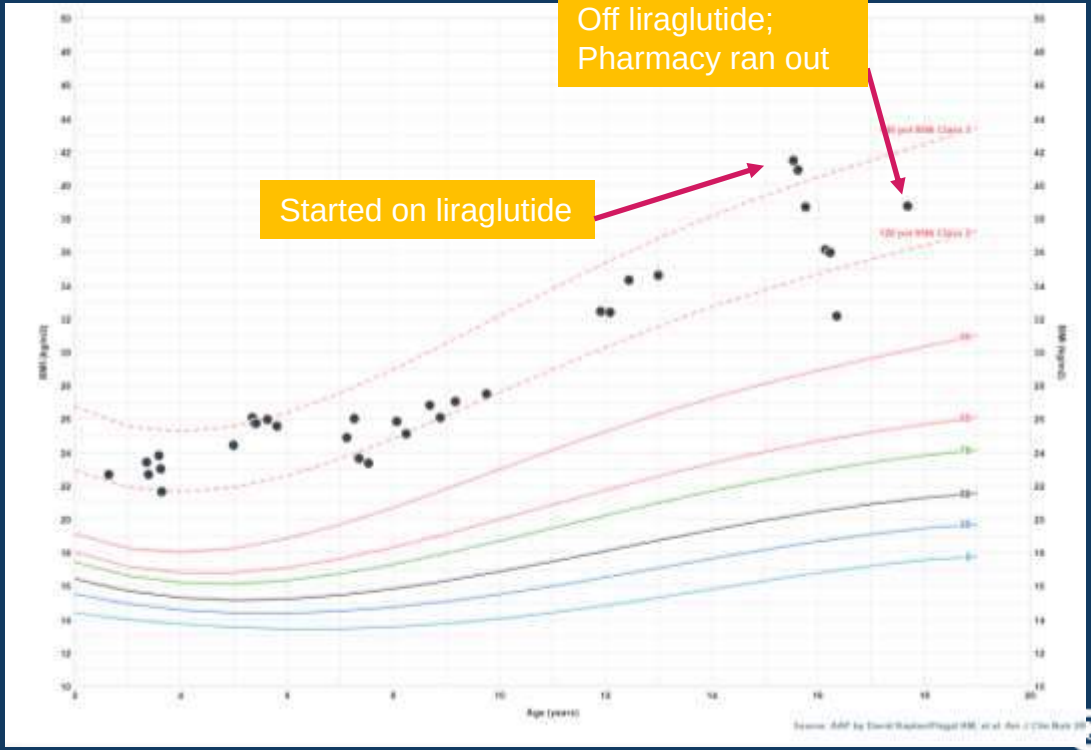
Exercise: 5hrs dynamic +3 core/weight lifting (motivation)

Sleep study +/-CPAP

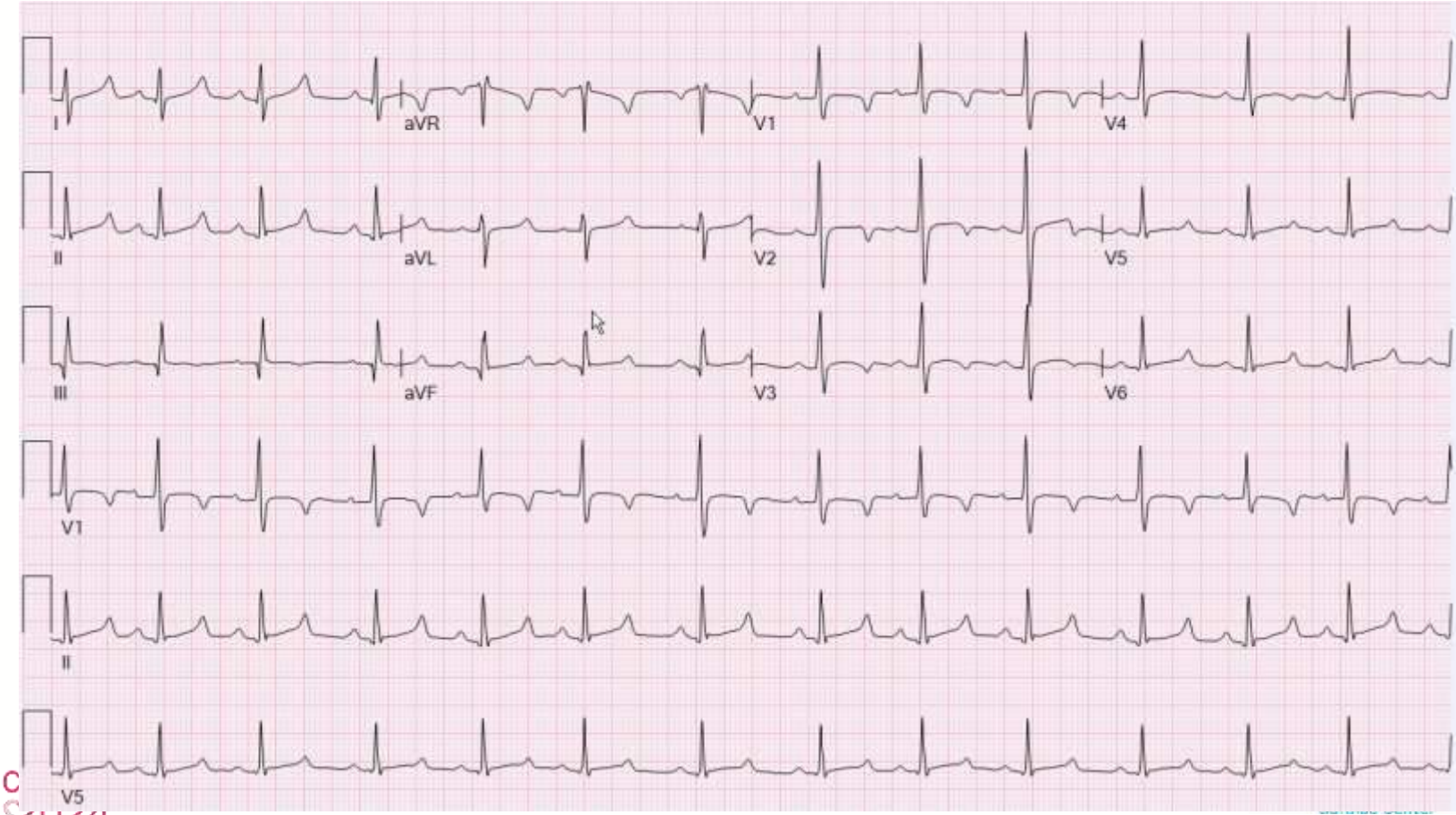
Repeat labs (typically give 3 months)

- if persistent t/c statin
  - TG >400mg/dL, icosapent ethyl (2-4g per day)
- if ABPM abnormal t/c antihypertensive agent



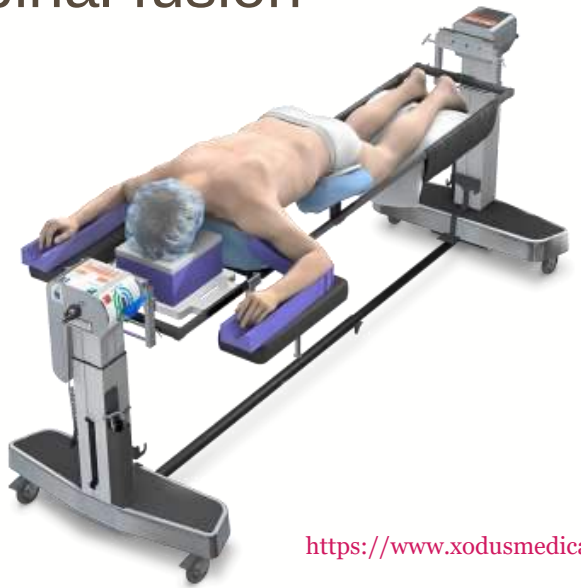

Off liraglutide;
Pharmacy ran out
Started on liraglutide

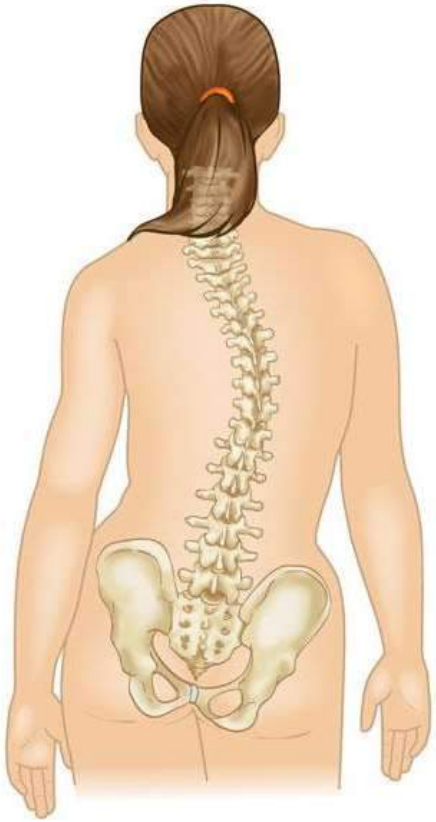
After Flecainide 50 mg PO BID –QTc shortened by 40 msec



CASE DESCRIPTION

14-year-old female with history of **Trisomy 21**, history of idiopathic scoliosis scheduled for posterior spinal fusion

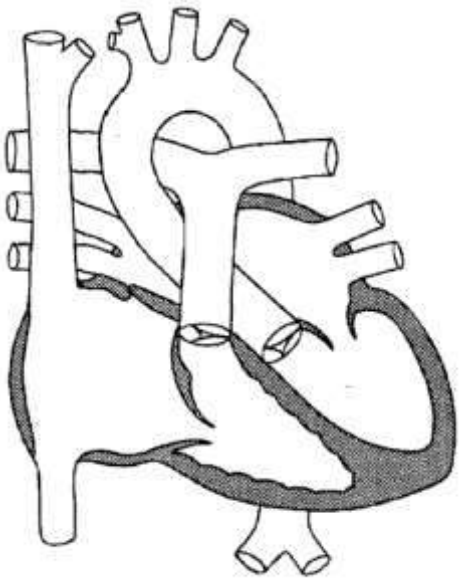




https://www.xodusmedical.com/BackPacks

TRISOMY 21

CARDIAC	RESP	GI/RENAL	NEURO	ENDO	SKELETAL	OTHER
• Had a TTE at birth, reported to be normal	• No hx of atlantoaxial instability • No prior airway evaluations or intubations • Mild intermittent asthma with occasional flare ups with URI	• Unremarkable	• Doing well in school	• No hypothyroidism	• No issues	• None



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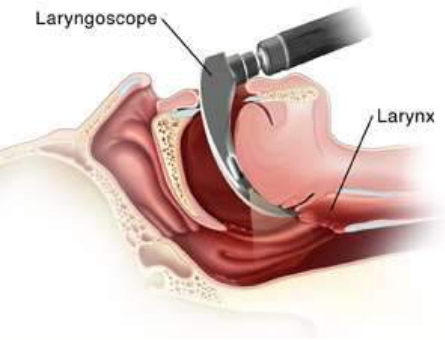
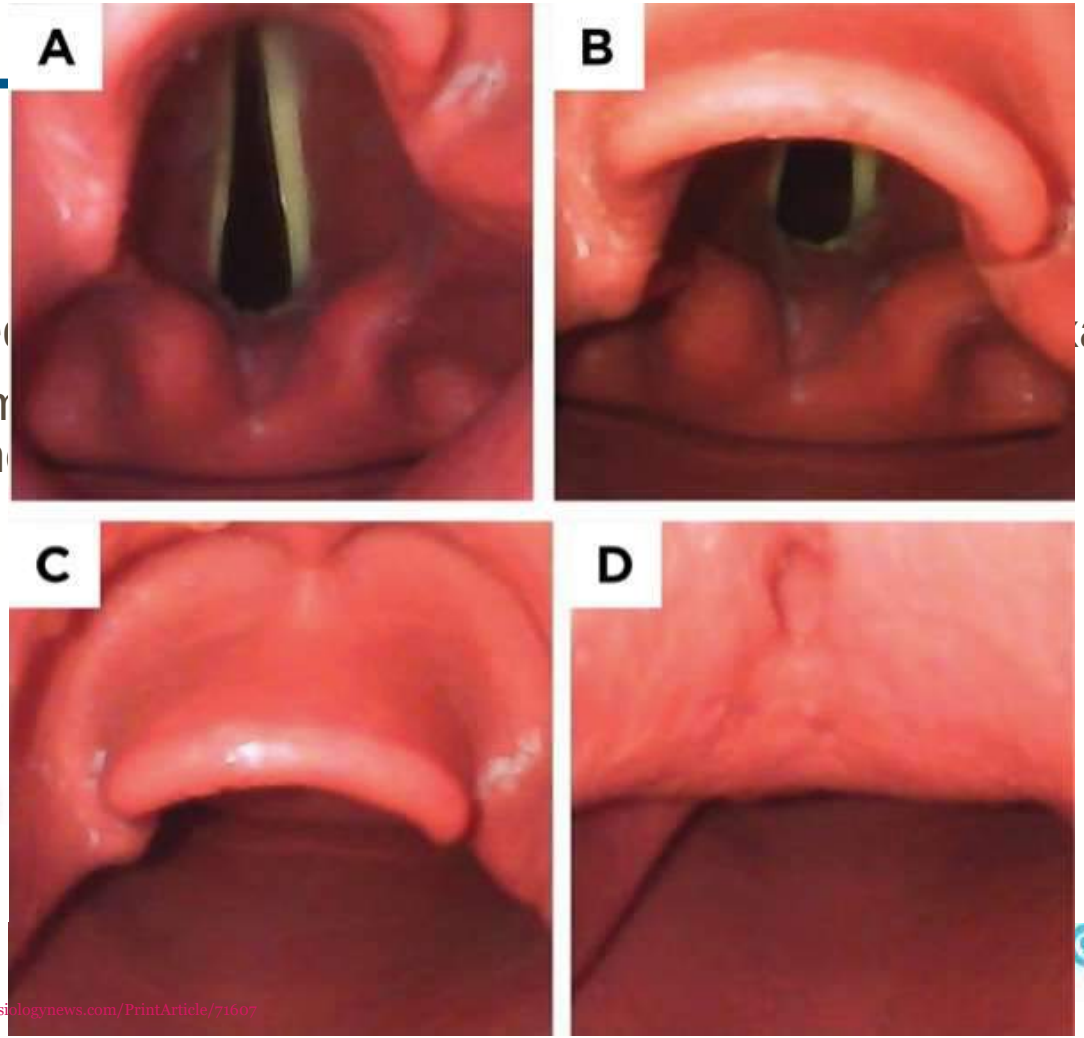
https://www.youtube.com/watch?v=oHPcuuKW984&t=90s

ANESTHE

- PIV placed in
- Uncomplicate ant
- Airway placem blade with 6.0 endotrach





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https://www.anesthesiologynews.com/PrintArticle/71607

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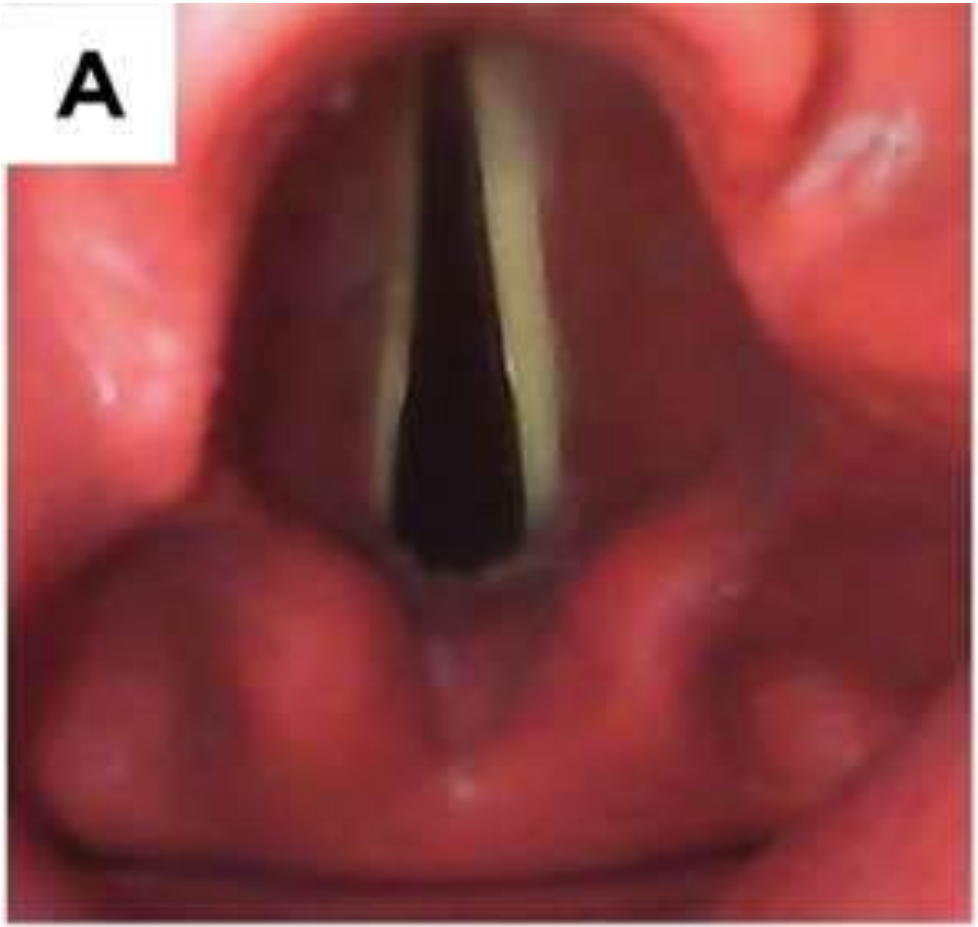
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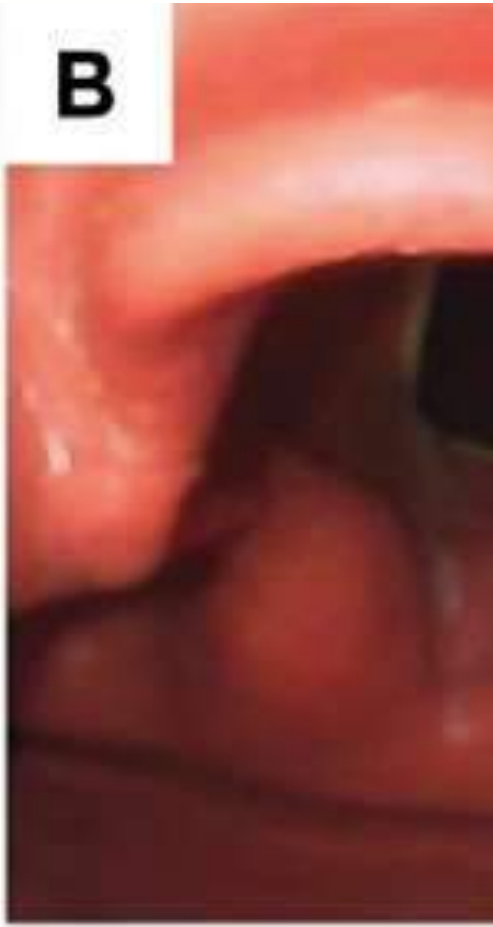
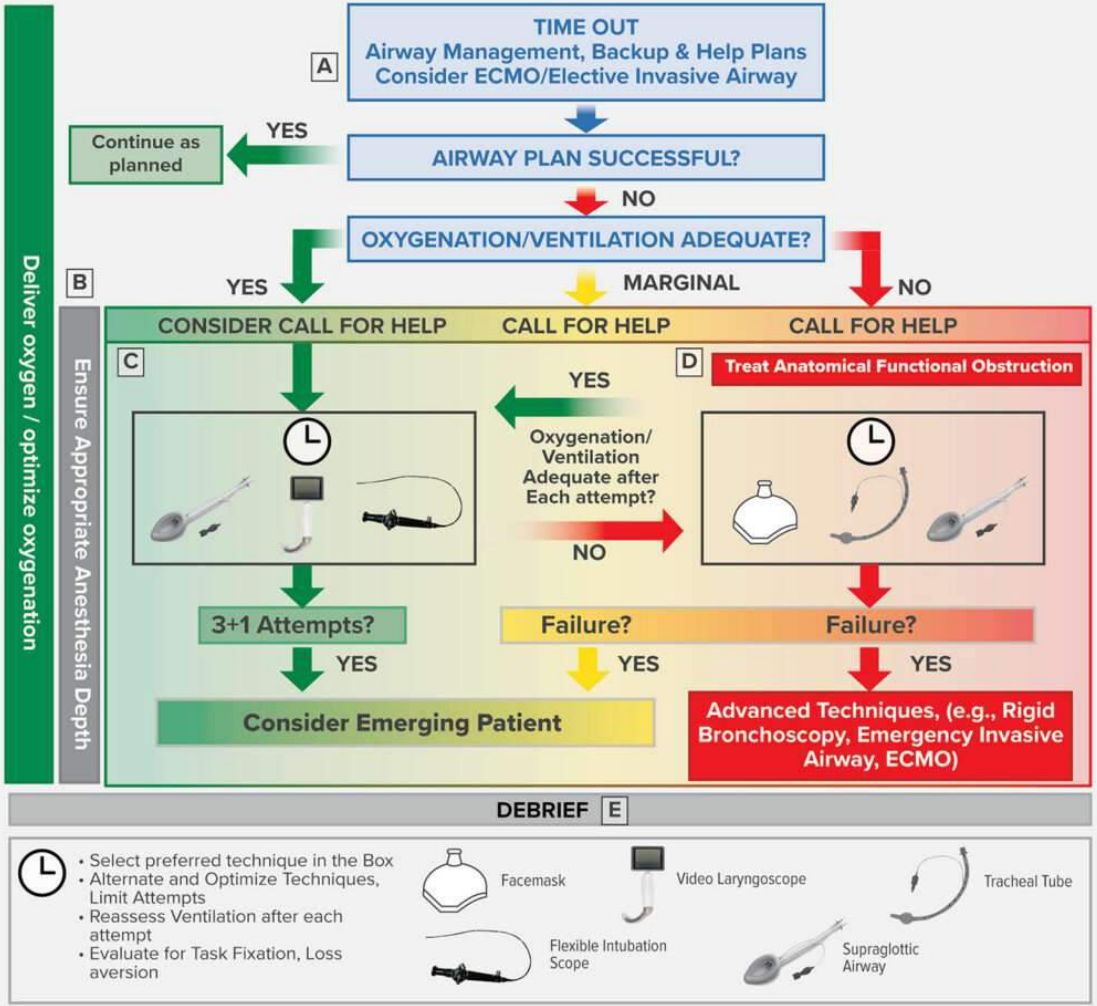

B

ANESTHETIC PLAN

- PIV placed in preoperative area
- Uncomplicated induction with Propofol, Midazolam, Rocuronium
- Airway placement attempt 1 with direct laryngoscopy with Mac 3 blade with 6.0 endotracheal tube, Grade 1 view with direct laryngoscopy
- 6.0 ETT (age appropriate for this patient) **would not pass beyond sub-glottic area**
- Second attempt with 5.0 ETT, **would not pass beyond sub-glottic area (with direct laryngoscopy)**
- Third attempt with hyperangulated Videolaryngosco

Difficult Airway Infographic: *Pediatric Patients*



https://www.apsf.org/article/anesthesia-patient-safety-foundation-update-2022-american-society-of-anesthesiologists-practice-guidelines-for-management-of-the-difficult-airway/

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Deliver oxygen / optimize oxygenation

B

YES

MARGINAL

Ensure Appropriate Anesthesia Depth

CONSIDER CALL FOR HELP

CALL FOR HELP

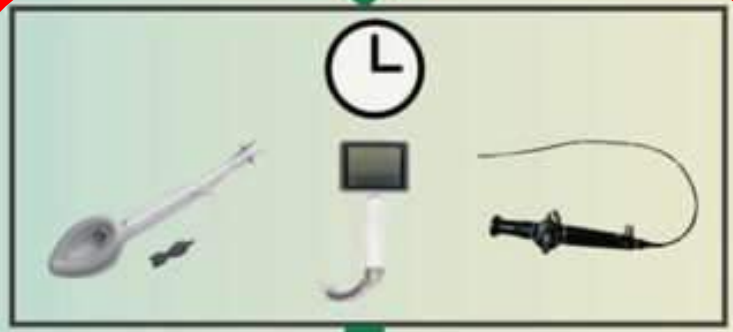
CALL FOR

C

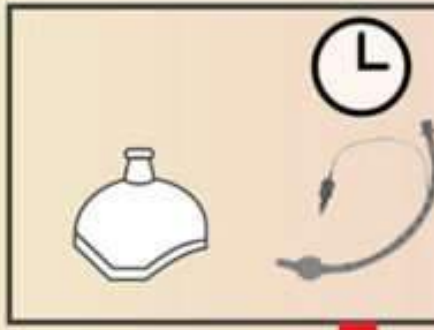
D

Treat Anatomical Fun

YES



Oxygenation/
Ventilation
Adequate after
Each attempt?



NO

3+1 Attempts?

Failure?

Failure

YES

YES

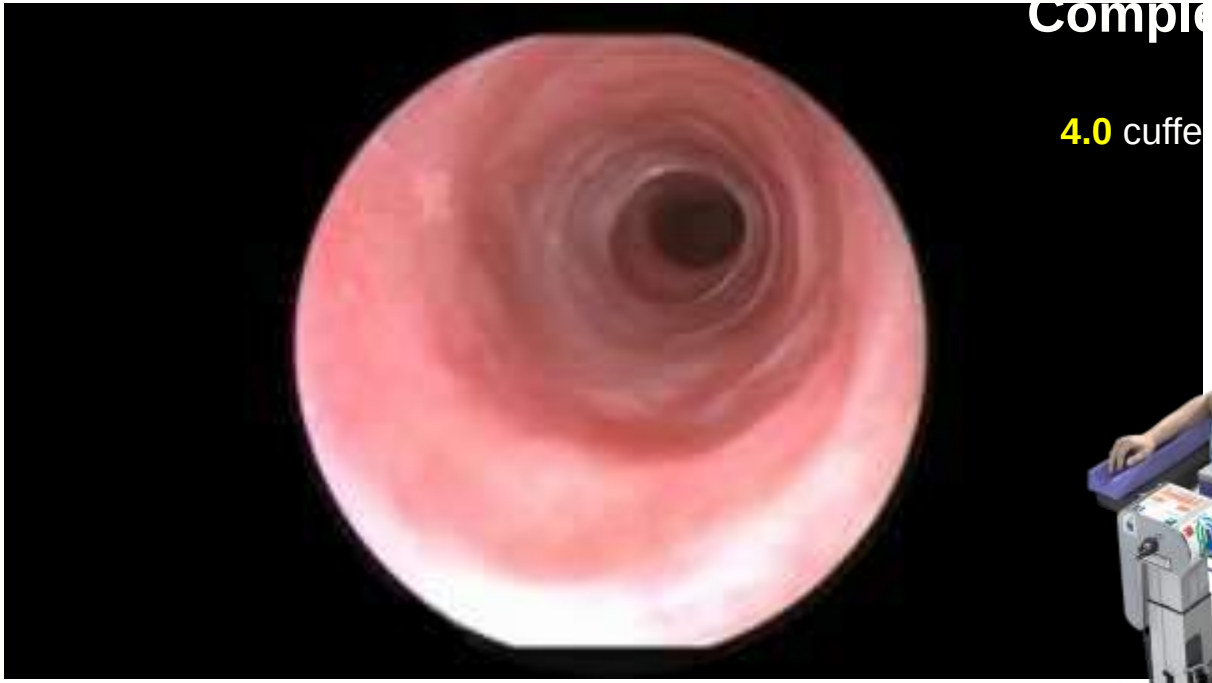
Consider Emerging Patient

Advanced Techniqu

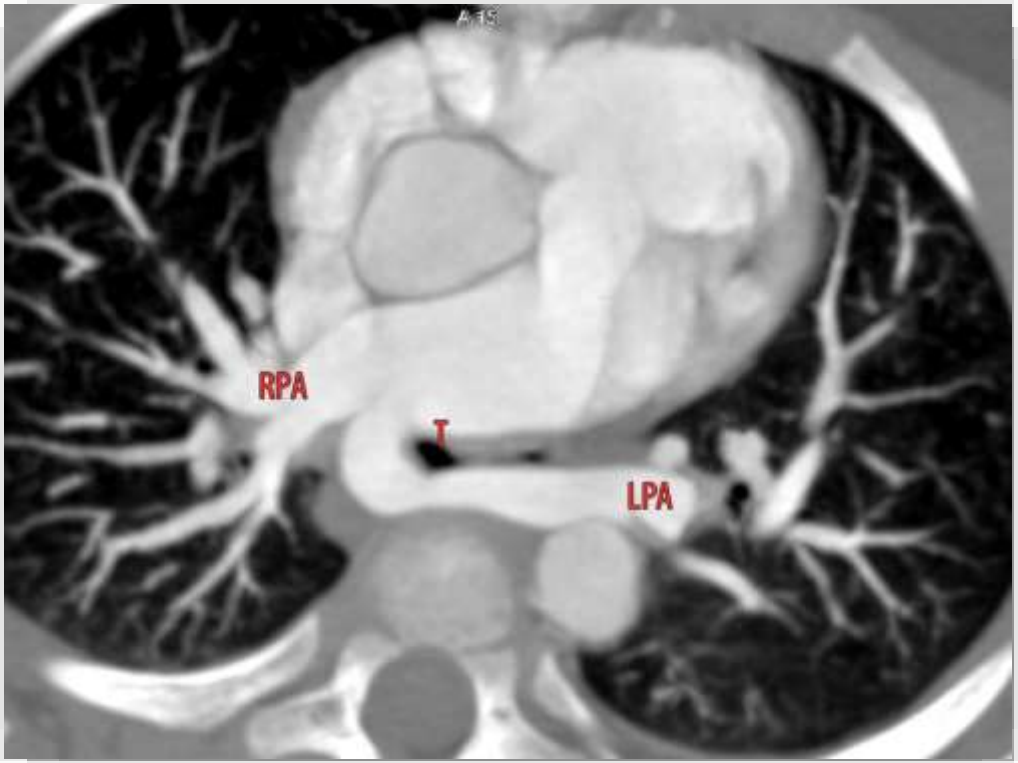
Bronchoscopy, Emer

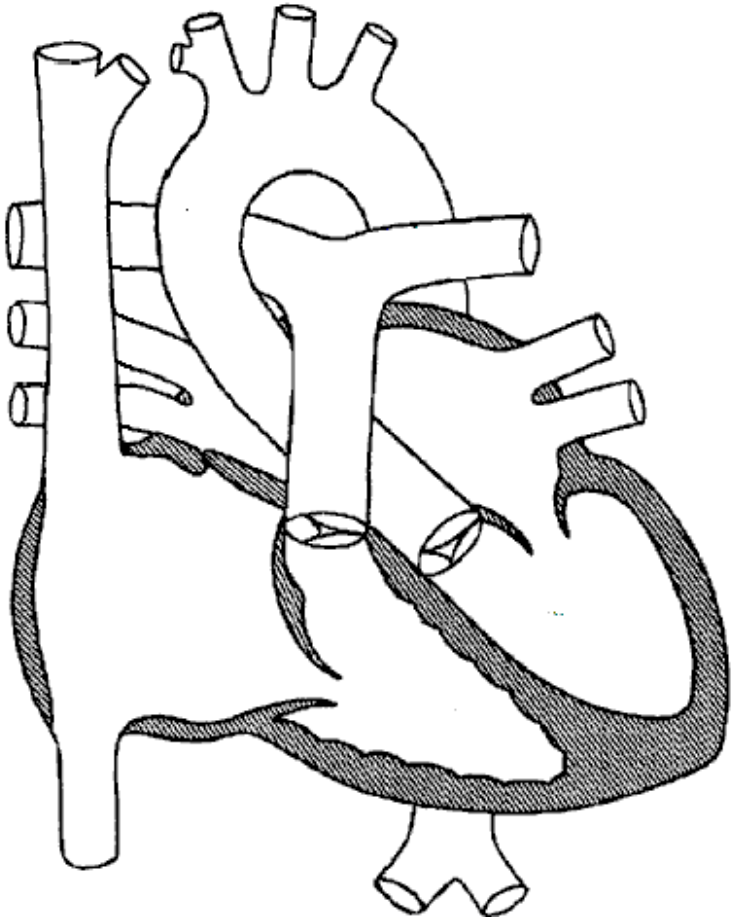
Airway, EC

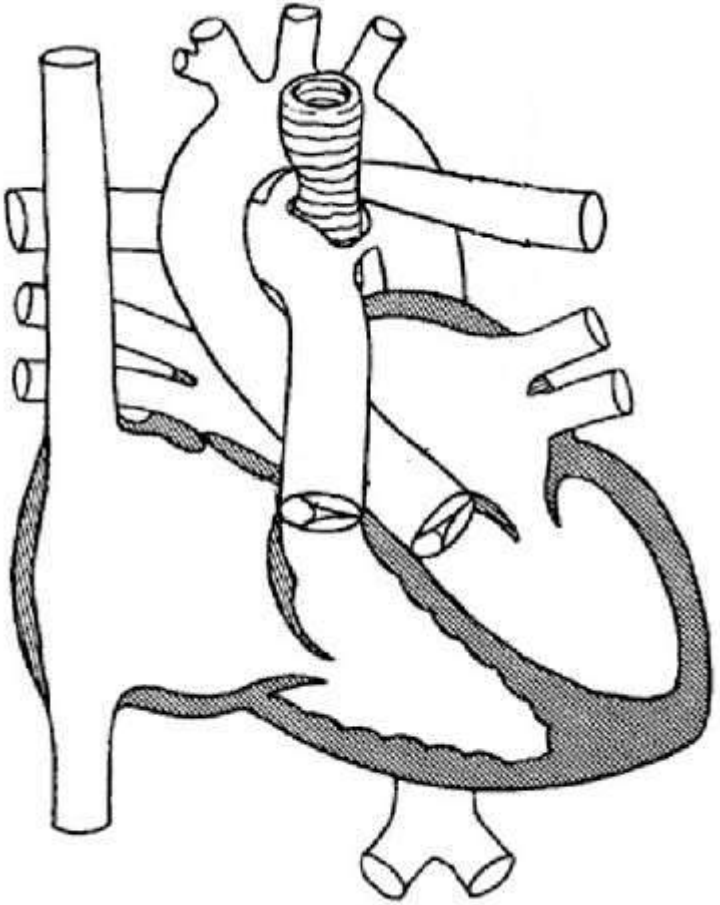
ENT EVALUATION IN THE OR



LPA SLING WITH TRACHEAL STENOSIS







CARDIOLOGY
2024

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LESSONS LEARNED

- Importance of thorough preoperative evaluation
- “Asthma” history, was likely from tracheal compression
- Early recognition and calling for help early before it is too late