Going For It: It Takes A Team

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Cardiac Center

ANCC Magnet Recognition

Cardiology 2018

Team Work in CHD Health Care

- "Interprofessional" Sport
- Everyone has a role and Everyone is important
- Everyone has the same goal:
  - **A good outcome for the patient with improved quality of life**
- Winning means "do no harm…make the patient better"
- The team needs effective, creative, and supportive management, leadership and coaching
- The patient and the family need to be part of the team
- The team needs a good "quarterback"
- The quarterback needs a good team

What is a team?

"A team is a group of people with complementary skills who are committed to a common purpose, performance goals, and approach for which they hold themselves mutually accountable."

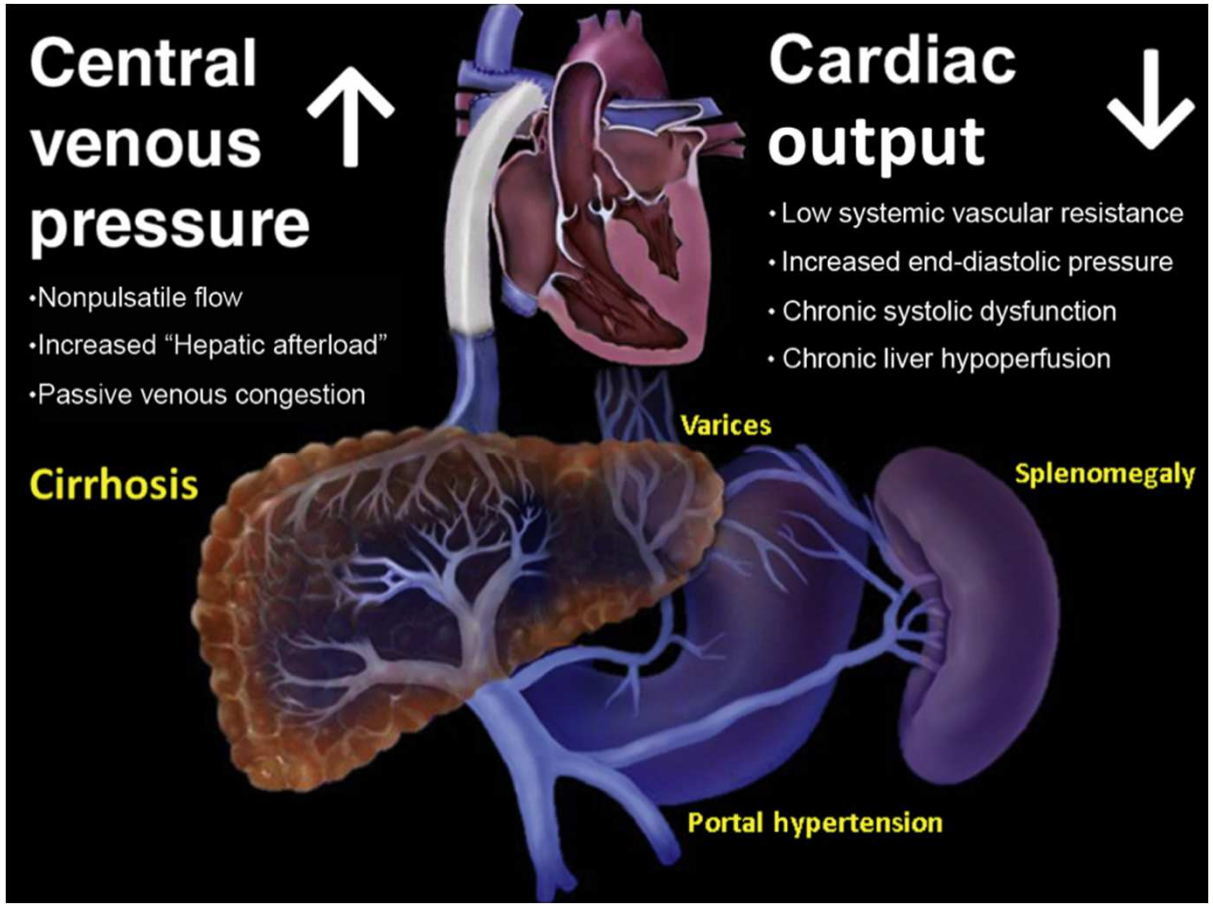
The Wisdom of Teams (Harvard Business School Press, 1993)

- Clear direction
- Open and honest communication
- Support risk taking and change
- Defined roles
- Mutually accountable
- Communicate freely
- Common goals
- Encourage differences in opinions
- Collaboration
- Team trust

Chief Complaint:

Hepatosplenomegaly, edema in a 12 year old Fontan patient

The Fontan and the Liver



HPI

- 12 year old with a balanced AVC, DORV, PS, and single ventricle CHD surgically palliated to an ECC Fontan in 2006 at another institution
- He also has a history of seizures, reactive airway disease, constipation and irritable bowel syndrome symptoms, and Factor VII deficiency
- He developed fatigue, hepatosplenomegaly, edema, and progressive AV valve insufficiency in the summer of 2016 that prompted further evaluation of his Fontan and he was started on medications
  - Hepatomegaly
  - ECC conduit with no fenestration
  - Liver imaging at outside hospital…say it is cirrhotic appearing by CT
  - Started Lasix, Digoxin…edema resolved…symptoms improved
  - Albumin normal. No casts. No lymphatic issues.
- Referred to SVSP for second opinion
  - Liver
  - Management
  - Transplant consideration

Birth History

- ✓Premature at 34 weeks
- ✓Birth weight 2.4 kg
- ✓Postnatal diagnosis
- ✓Syndromic appearing
- Cyanotic
- Prostaglandins

Past Medical History

- Prematurity
- Syndromic; concern for Noonan's
  - Microarray in 2014 showed 3p25 duplication and 7p5.3 triplication. Mom has the same variations. 3p duplication includes RAF1 gene known to be associated with Noonan syndrome. But subsequent gene sequencing for Noonan syndrome has been negative.
- Mild developmental delay
- Factor VII deficiency; not anticoagulated(Has seen hematology at another institution)
  - No bleeding issues with this at any of his surgeries. Noted to have a prolonged INR.
- Seizures(Sees neurology at a another institution)
- Migraines

Past Surgical History

- BTS as a newborn
- 10/2004: Bilateral BDG, pulmonary valve oversewn and removed leaflets, atrial septectomy
- 2006: 16 mm PTFE ECC Fontan with no fenestration
    - Junctional rhythm post op
    - Pleural effusions post op
- No non cardiac surgeries

Family History

- No known history of CHD
- Several maternal relatives have had heart murmurs including Mom and his sister but no surgery
- No history of sudden cardiac death
- No history of cardiomyopathy
- Patient and Mom have a 3p25 duplication and 7p5.3 triplication
  - 3p duplication includes RAF1 gene known to be associated with Noonan syndrome but subsequent gene sequencing for Noonan syndrome has been negative for Mom and patient

Social History

- Lives in an impoverished area
- No car
- Only dad works
- 1 brother and 1 sister. Other family in house as well.
- Medicaid…and non par with CHOP
- English is second language; primary is Spanish…pitfall… believing that everything is understood in English
- Exercises in a park near home…but only at certain times when safe to do so
- No drugs, alcohol, tobacco, vaping, piercing. No guns in the house. Feels safe in home but not in neighborhood or at school. No food concerns; breakfast and lunch at school
- 7th grade. IEP; struggles with school
- Loves basketball

Medications

Medication	Sig
albuterol HFA (VENTOLIN HFA) 108 (90 BASE) mcg/ACT Inhalation oral inhaler	2 puff(s) by Inhaled route every 4 hours as needed.
digoxin 0.125 mg Oral tablet	Take 187.5 mcg by mouth once a day. 1.5 tabs once daily
fluticasone furoate 27.5 mcg/SPRAY Nasal spray	Spray 2 spray(s) in both nostrils once a day.
fluticasone HFA (FLOVENT HFA) 110 mcg/ACT Inhalation oral inhaler	2 puff(s) by Inhaled route 2 times daily.
furosemide 20 mg Oral tablet	Take 20 mg daily.
lisinopril 5 mg Oral tablet	Take by mouth once a day.
loratadine 5 mg/5mL Oral syrup	Take 10 mL by mouth 2 times daily.
Multiple Vitamins-Minerals (VITAMINS W/ MINERALS, ADULT,) Oral tablet	Take by mouth once a day. With iron
ranitidine 150 MG Oral tablet	Take 150 mg by mouth once a day.
spironolactone 25 mg Oral tablet	Take 25 mg daily.
topiramate (TOPAMAX) 100 mg Oral tablet	Take 150 mg by mouth 2 times daily.

ROS

REVIEW OF SYSTEMS	
Eyes: Wears glasses.	**Endocrine:** Overall okay.
Gastrointestinal: Hepatomegaly. Constipation. IBS symptoms with pain/constipation. Had had endoscopy that was normal. Recent CT scan with enlarged liver and spleen, likely passive congestion.	**Genitourinary:** He is not circumcised. No issues.
Neurologic: Seizure history. On medications. Last seizure was last year. Followed by neuro. Has migraine headaches with emesis and vomiting. Topamax used to treat both.	**Psych/Development:** Dennis is in 7th grade. Has an IEP. Mild delays. Chromosomal abnormality but not Noonan's and Mom has same abnormality. .
Constitutional: No recent fevers or weight loss. No international travel. No ill contacts.	**Ears/Nose/Throat:** No hearing issues. Seasonal allergies. Snoring. Occasional epistaxis.
Hematologic: Factor VII issues. Not on aspirin or any anticoagulation.	**Integumentary/Skin:** Surgical scars. As occasional eczema but not now.
Musculoskeletal: Occasional joint pain. Has occasional edema but much better on diuretics.	**Respiratory:** RAD and seasonal allergies. Uses meds. No cough. No casts.
Immunologic: No warts. Susceptible to frequent illnesses.	**Cardiology:** ECC Fontan (only 16 mm PTFE) with no fenestration. Recent holter with mostly sinus rhythm and only one PVC. No chest pain, no syncope, no palpitations. Exercise this summer with no symptoms, normal BP, no ectopy. Baseline sat is 93-95%. CAVC and aortic valve leakage on echo and systolic and diastolic murmur on exam.

Physical Examination

- Dysmorphic facial features
- Well healed surgical scars; scar on left cheed
- No pectus
- 2/4 systolic murmur, ¼ diastolic murmur
- Hepatomegaly(4cm), firm
- Splenomegaly
- No caput medusa
- 1+ edema, very mild varicosities
- No petechiae or bruising

Testing

- ECHO
- Exercise
- US of abdomen
- Labs

Exercise Stress Testing

- Heart rate ranged from 53 bpm at rest to 160 bpm with exercise
- BP 102/50 at rest and 155/50 with exercise
- Oxygen saturation 95% at rest and 95% with exercise
- Max VO2 of 42 ml/kg/min…THIS IS OUTSTANDING!
- Sinus rhythm
- No symptoms
- No ectopy

ECHO

- 2 good sized ventricles; both with good function
- VSD
- Diagnosis is actually TOF/CCAVC
- Aorta ov
- AVC wit
- Pulmon
- Normal
- Mild ao
- ECC conduit is small with no fenestration no obstruction
- Branch PA's not well visualized

2 ventricles?

US of Abdomen

- Enlargement of the spleen measuring 18 cm in length (suggested upper limit of normal is 12 cm for this age range)
- Mild heterogeneous echotexture of the liver
- Enlarged liver; no nodules
- Normal duplex Doppler flow in the liver given history of Fontan
- No ascites
- Normal kidneys, gall bladder, pancreas

Impression

- 12 year old with TOF and balanced CCAVC s/p ECC fenestrated Fontan in 2004(fenestration now closed) with a history of seizures, Factor VII deficiency, migraines, mild developmental delay and early signs of Fontan morbidity including hepatosplenomegaly and likely hepatic fibrosis, peripheral edema
  - He has 2 ventricles, the aorta is committed to the LV, overrides VSD
  - WHY IS HE A FONTAN?
  - Is it possible to convert to 2 ventricle physiology?
  - He has 2 ventricles…but he is stable now
  - Can we takedown and convert?
  - Even if we can…should we…

Plan: Manage current symptoms and determine if conversion possible

- Maximize diuretics; increased lasix, added aldactone
- Increased the Lisinopril for AVVR and AI
- Holter
- Needs a "Fontan" cath and liver biopsy…if we stay a Fontan
- Schedule MRI; question is…can we convert?
- Schedule Cardiac Catheterization; question is…can we convert?
- Obtain hematology, genetic, and neurology records from other institution
- Consult hematology, neurology at CHOP
- Consult social work…we have issues
- Consult financial counselor
- Consider and interpreter

Do Not Forget~

- Insurance Issues
- Transportation Issues
- Factor VII issues
- Seizure issues

Need a Team Approach…a lot of moving parts

- MRI and Cath
- Liver Assessment
- Financial Counselor
- Social Work
- Hematology and Neurology
- Cardiology
- Cardiac Anesthesia
- Cardiac Surgery
- Scheduling Center
- Intake Center
- OR
- CICU
- Family and patient

Team needs a bus driver

- Clear direction
- Open and honest communication
- Support risk taking and change
- Defined roles
- Mutually accountable
- Communicate freely
- Common goals
- Encourage differences in opinions
- Collaboration
- Team trust

Need more testing

MR elastography of the Liver

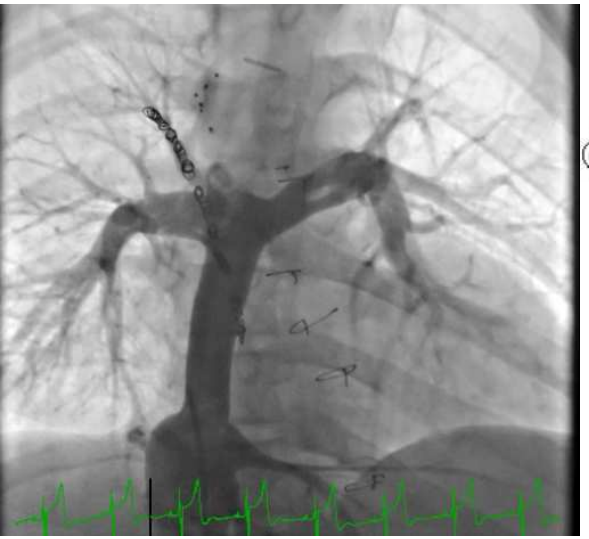
- Liver: Hepatomegaly, mild parenchymal heterogeneity, and lateral segment left hepatic lobe atrophy.
- Liver stiffness is increased Stiffness measures 4.92 kPa on EPI and 4.15 kPa on GRE sequences; normal liver stiffness in MR elastography is < 2.9kPa.
- Spleen: Splenomegaly. Otherwise, normal morphology.
- Spleen stiffness is increased Stiffness measures 9.94 kPa on EPI sequences and 5.34 kPa on GRE sequences; normal spleen stiffness in MR elastography is < 3.6 kPa.

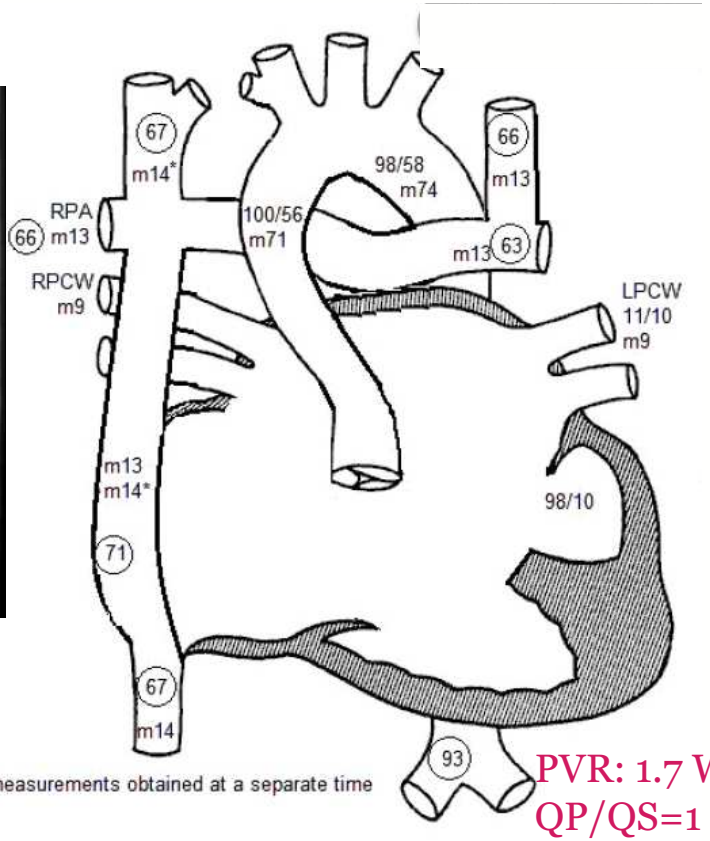
Cardiac MRI

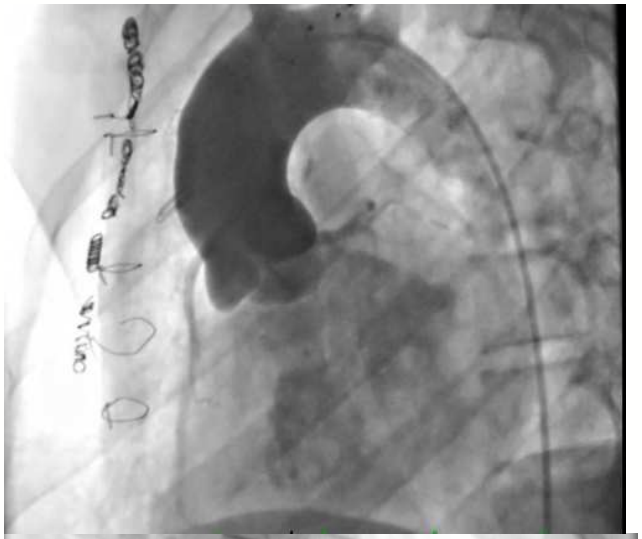
Common AV canal with normally related great vessels

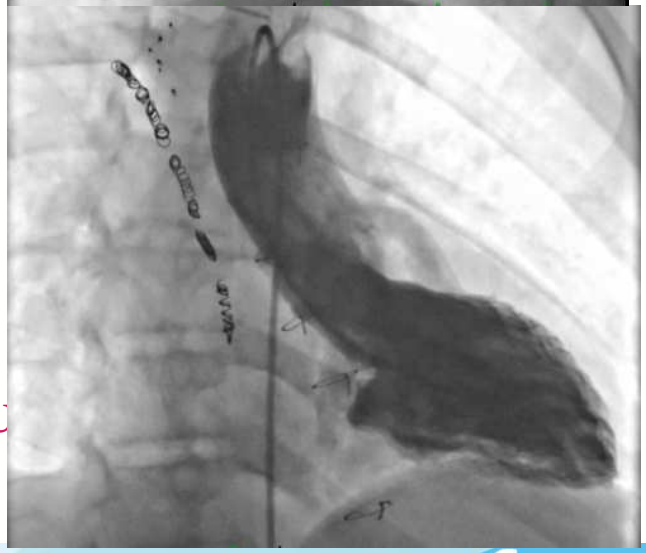
- The aorta overrides the VSD but sits mostly over the RV
- There appears to be a direct pathway from the LV to the aorta
- Low normal biventricular size and high normal biventricular ejection
- Fontan pathway and branch PA's unobstructed
- Mild AV valve regurgitation (RF 16%)
- Mild AI (RF 5%)
- Moderate collateral flow (27% of aortic flow)

Cardiac Catheterization









PVR: 1.7 WU
QP/QS=1

Factor VII Deficiency

- Mild FVII deficiency
- With a level of 35%, many patients do not have bleeding issues
- Proceed with surgery
- Low dose rVIIa (15- 25 mcg/kg) only if bleeding

	Ref. Range	10/12/2016 00:00	6/1/2017 10:09	11/20/2017 11:25
INR	Unknown		1.52	1.36
INR	Latest Ref Range: 0.8 - 1.2	1.3 (H)		
Prothrombin Time	Latest Ref Range: 11.6 - 13.8 secs	13.9(H)	18.0 (H)	16.4 (H)
Partial Thromboplastin Time	Latest Ref Range: 22.0 - 36.0 secs			29.5
APTT	Latest Ref Range: 26 - 35 sec	32		
Factor VII	Latest Ref Range: 57 - 139 %			35 (L)

Team needs to meet, discuss, and work together

- Presented at Surgical Case Conference
- Everyone reviewed/ discussed data
- Everyone discusses the options
  - Clear direction
  - Open and honest communication
  - Support risk taking and change
  - Defined roles
  - Mutually accountable
  - Communicate freely
  - Common goals
  - Encourage differences in opinions
  - Collaboration
  - Team trust

What are the options?

- Punt and stay as we are…he is fine now?
- Kick a field goal…add more medications like sildenafil?

OR

- Go for a touchdown?

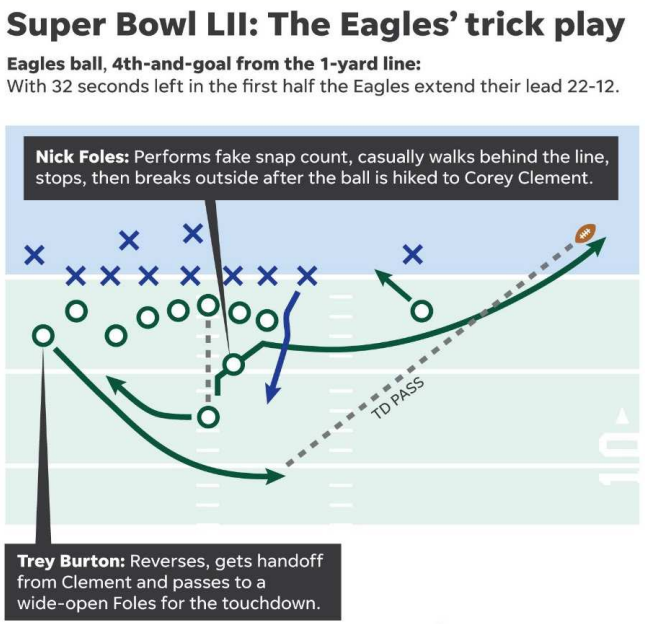
He and his family are part of the team!

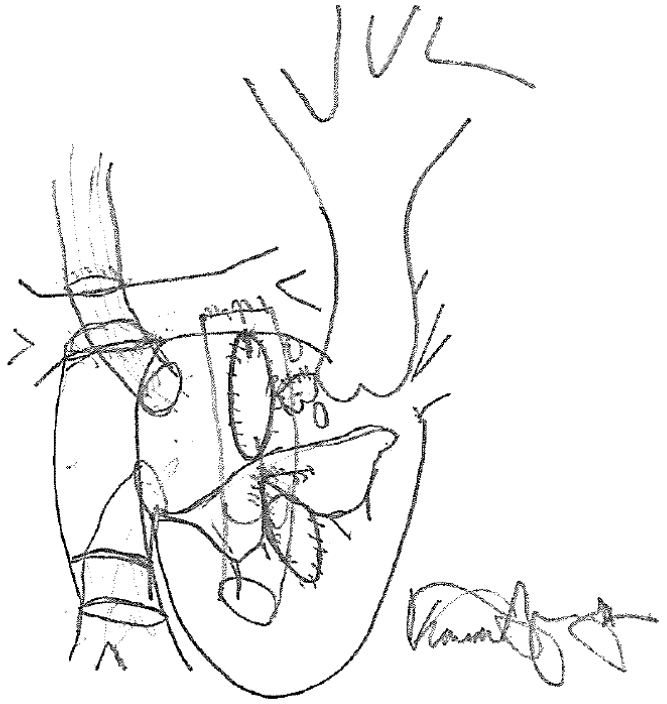
- We scheduled a family meeting
- We forgot the translator…
- We scheduled a second meeting…
- We wanted to assure Informed Consent and Assent

- Patient and family agree…let's go for it…we all get a bit tearful

Go For It!

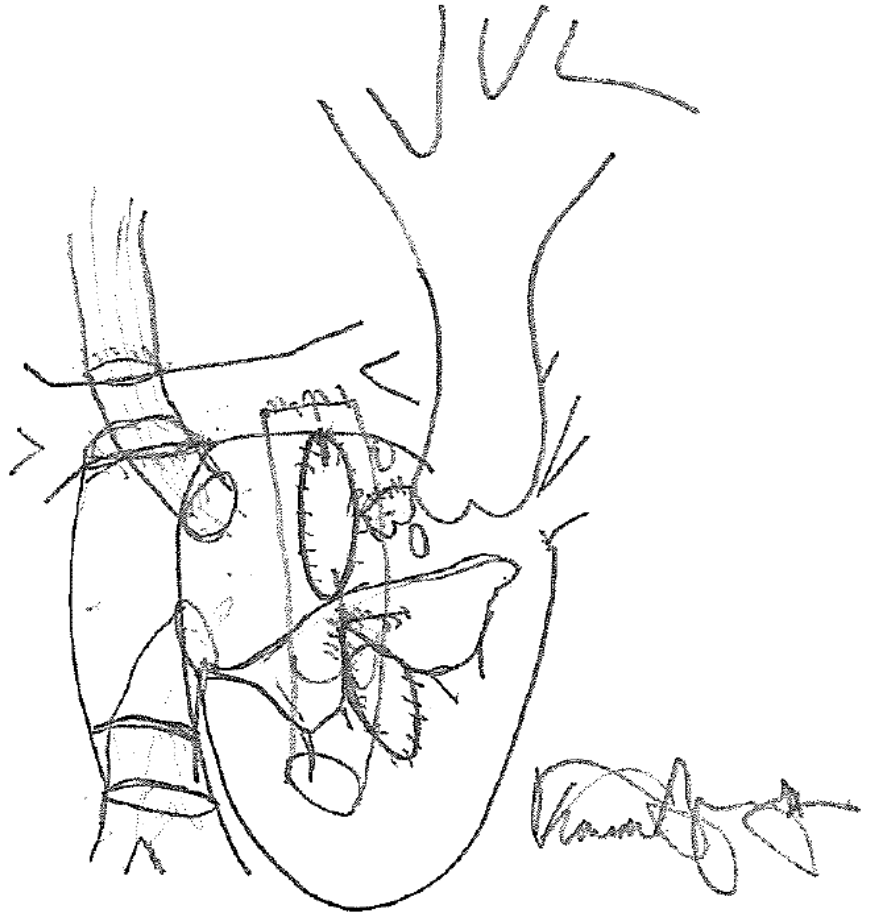
The “Philly” Special…4th and goal from the 1 yard line
The cardiac surgeon or “TLS” version





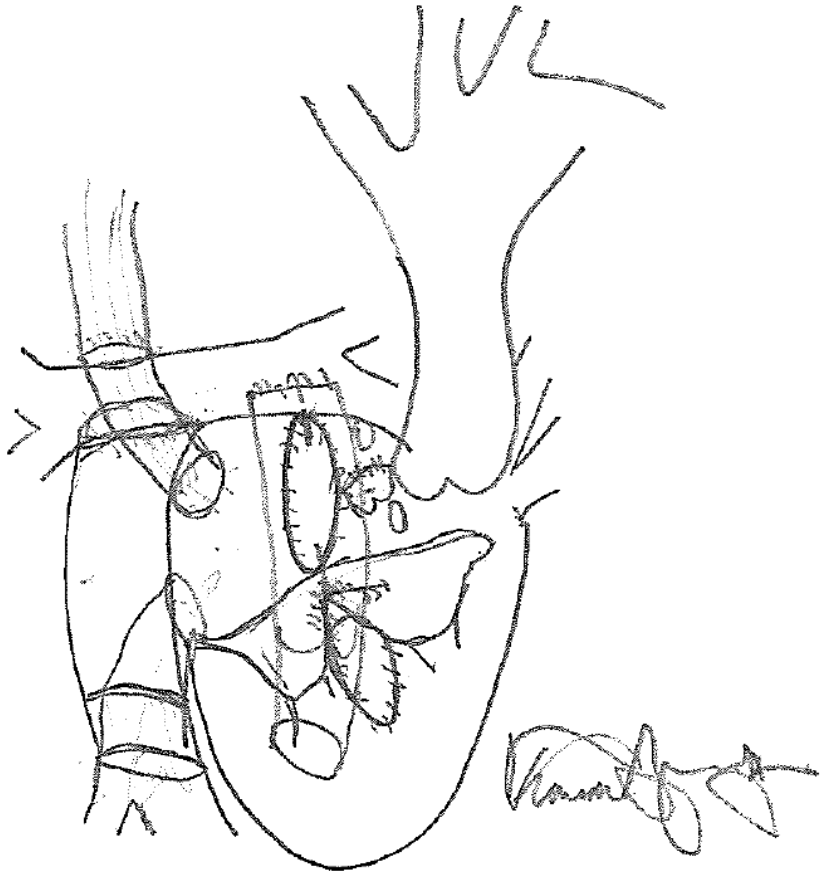
Initial steps

- Careful dissection
- Cannulation of LSVC, RSVC, Aorta, low right atrium
- CPB
- ECC conduit excised leaving a small segment on IVC for anastomosis to the RA
- RA opened



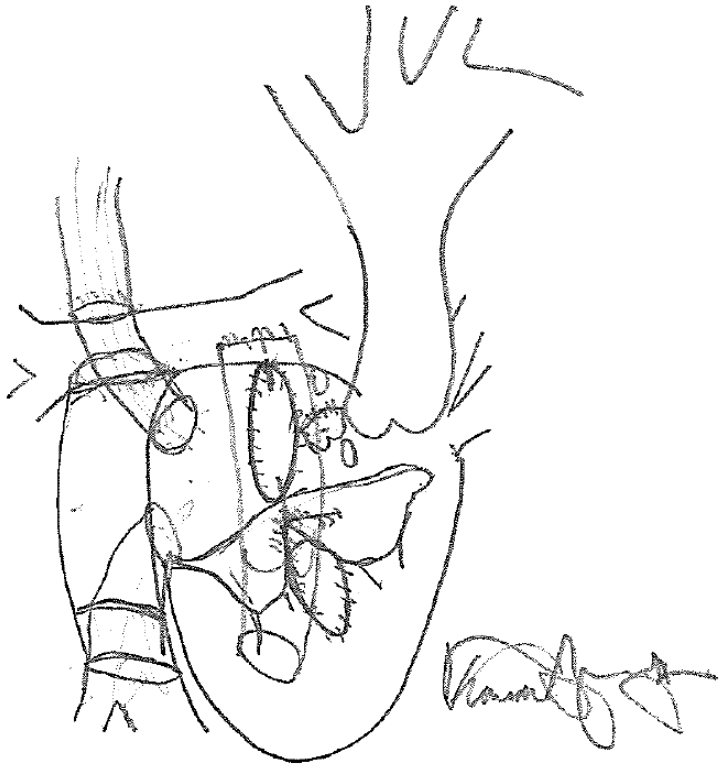
Next: Address the CCAVC

- Rastelli type C CCAVC with TOF
- Coapting aspects of the CAVV leaflets sutured together with gore-tex suture and a patch of double velour dacron cut to size with anterior extension to close the VSD component of defect
- CAVV leaflets then secured to the superior margin of the VSD patch
- Cleft of LAVV closed
- Primum ASD closed with Gore-tex patch leaving a vent for left side due to non coronary collateral return to the left atrium
- Canal repair complete



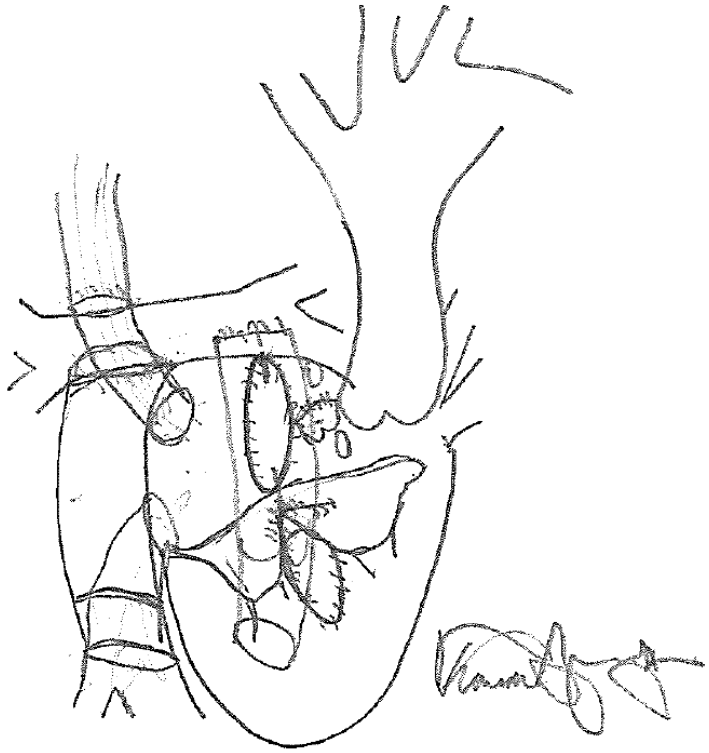
TOF: RV to PA conduit

- The original PV was oversewn and leaflets removed so…
- MPA was divided and PA opened onto the left PA
- 24 mm pulmonary homograft anastomsed to the PA bifurcation distally
- Incision in free wall of RV and muscle bundles divided and proximal anastomosis of homograft created to the RV outflow tract using a gore tex patch for hood anterior to create a gentle takeoff from the RV to the PA's
- Distal MPA proximally over sewn and the previous PV had already been oversewn at time of the Fontan



Complete the Fontan takedown and reestablish SVC and IVC connection to the right atrium

- Next the IVC was anastomosed to the inferior aspect of the right atriotomy incision
- SVC divided at entrance to the RPA and 10mm Impra graft cut and sized to connect the SVC and this was anastomosed to the right atrium
- The connection of the previous ECC conduit to the PA on the right was oversewn using a patch of homograft to close the entrance of SVC to the RPA
- Vent removed
- Left BDG left alone
- Case finished



Post op and Recovery

- Junctional rhythm initially but now sinus rhythm
- Bilateral pleural effusions, resolving with diuretics
- Discharged home after 2 week LOS
- Insurance changed, can now come to CHOP
- Socially, the family still struggles with transportation and neighborhood…social work UBER's him to appointments
- He has lost weight…without breakfast and lunch in school…oh my…
- Today?
  - Playing basketball
  - "I have never had this much energy. That is the thing I notice the most"
  - Continuing homebound instruction. Does not feel safe in school

Not a Fontan!
Dilly Dilly…or Philly Philly…or Spray Spray

Effective and Successful CHD Teams

- Need a good quarterback…a surgeon who can throw, run, catch and think on her/his feet and see things in more than 2 dimensions
- What happens in the OR is huge
- But the planning that gets you to the OR is critical
- The team management of issues post operatively is crucial
- Need to call the right plays…but be able to modify and make changes as needed…have to communicate
- Supportive, innovative, respectful, brave, and gritty team players…who perform at their best…always