Single ventricle pathophysiology: beyond childhood

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Overview

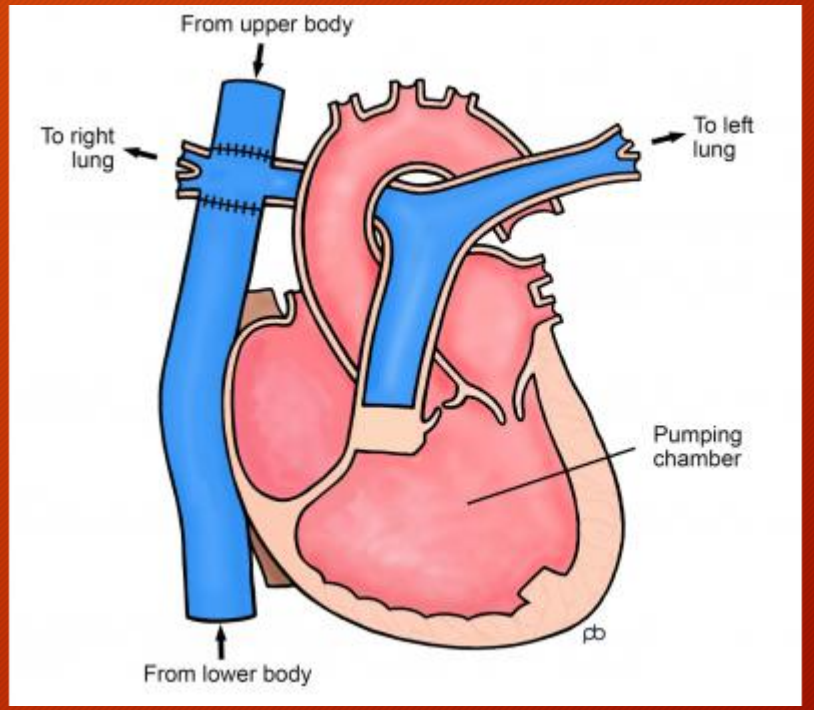
- Intro: what is a single ventricle? What is a Fontan circulation?
- Cardiovascular changes in Fontan circulation
- Complications by system:
  - Heart: valves, pump, rhythm
  - Lungs: perfusion, pressures
  - Liver: FALD
  - Renal
  - GI
  - GU
  - Brain

Introduction: single ventricle

- Definition/ anatomy:
- 1 ventricle due to ventricular hypoplasia or irreparable communication between the 2 ventricles
- 3.1-4.9 per 10 000 live births
- Not all “Fontans”
- Some patients have no intervention or other palliation e.g. PDA stent, PA band
- Some stop at Norwood stage 1 or stage 2

Introduction- Fontan

- No sub-pulmonic ventricle
- Passive flow to the lungs
- All cardiac output generated by one ventricle
- Aims: secure pulmonary perfusion, separate oxygenated blood, improve saturations and ET

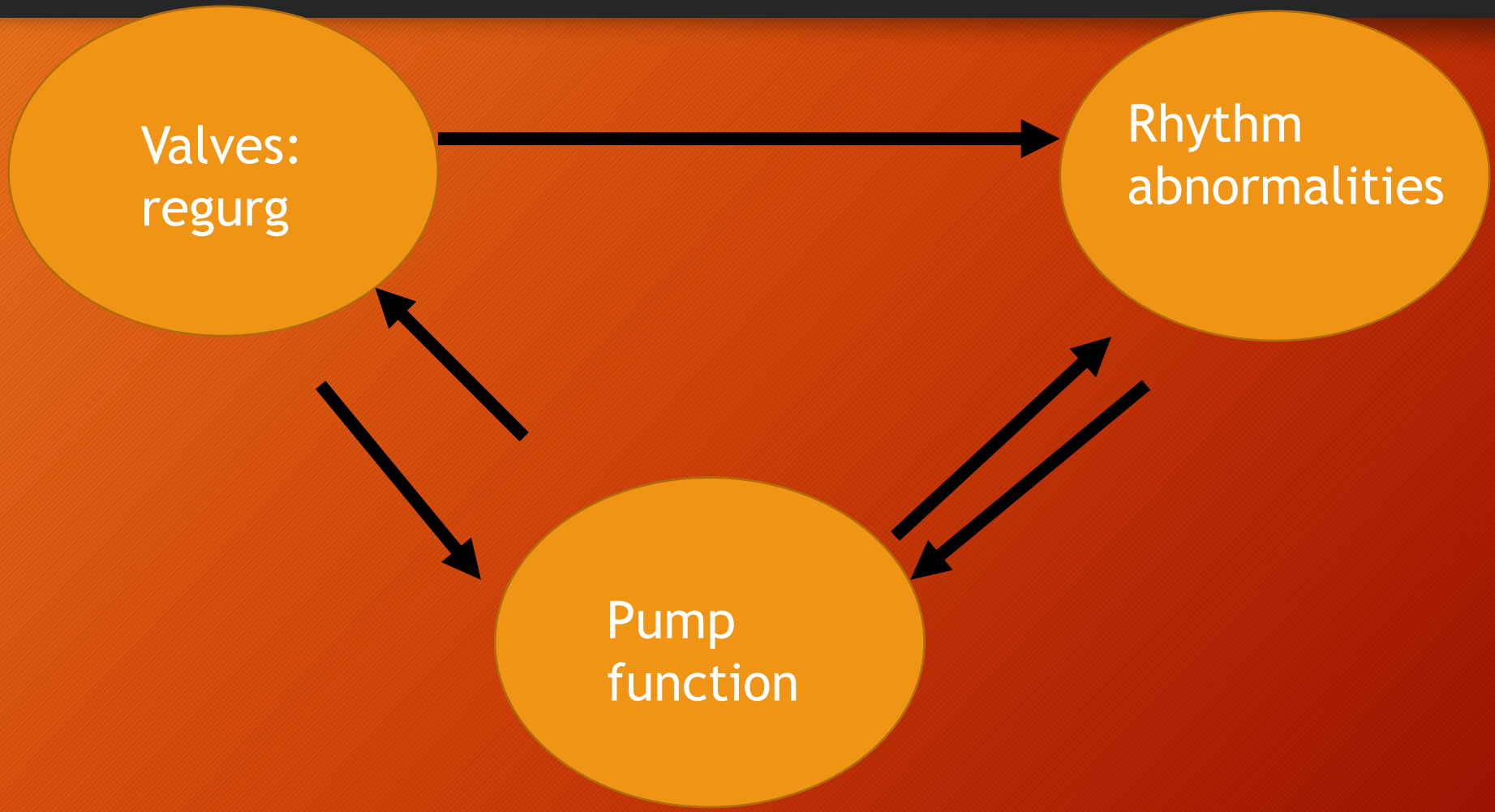


Fontan-pathophysiology

- Increase in systemic venous pressure
- Increase in afterload
- Increased sympathetic drive
- Increased vascular resistance

- Pulmonary and systemic vascular congestion

Cardiac complications

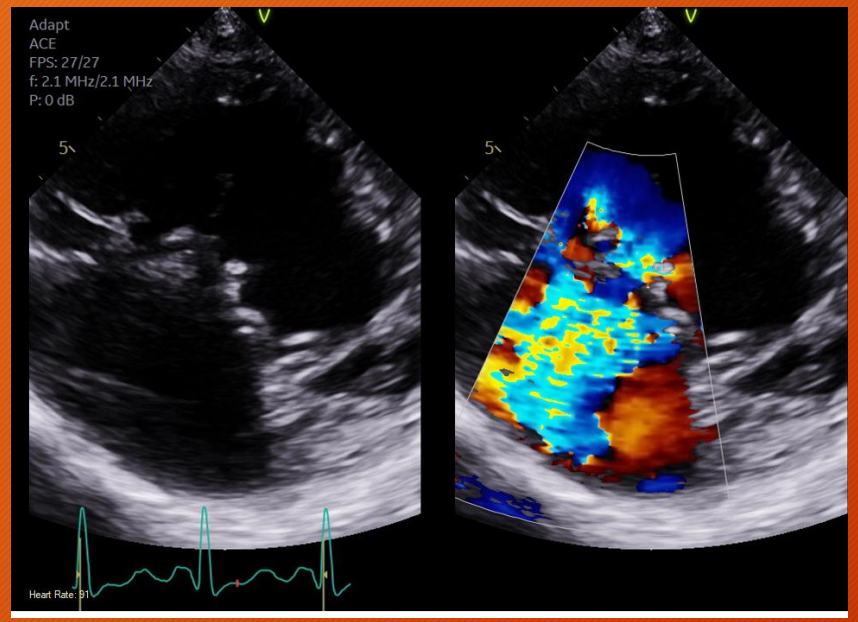


AV valve regurgitation

- 75% have some degree following Fontan completion
- Most common in anatomical tricuspid valves or common AV valve
- Valvar- cleft, prolapse
- Subvalvar- abnormal papillary muscles, abnormal chordal insertion leading to tethering
- “Functional”- failure of coaption due to atrial and ventricular dilatation
- Self-propagating

AV valve regurg

Echo image



AVSD- unrepaired

- Post bi-directional Glenn
- PAs to small for Fontan completion

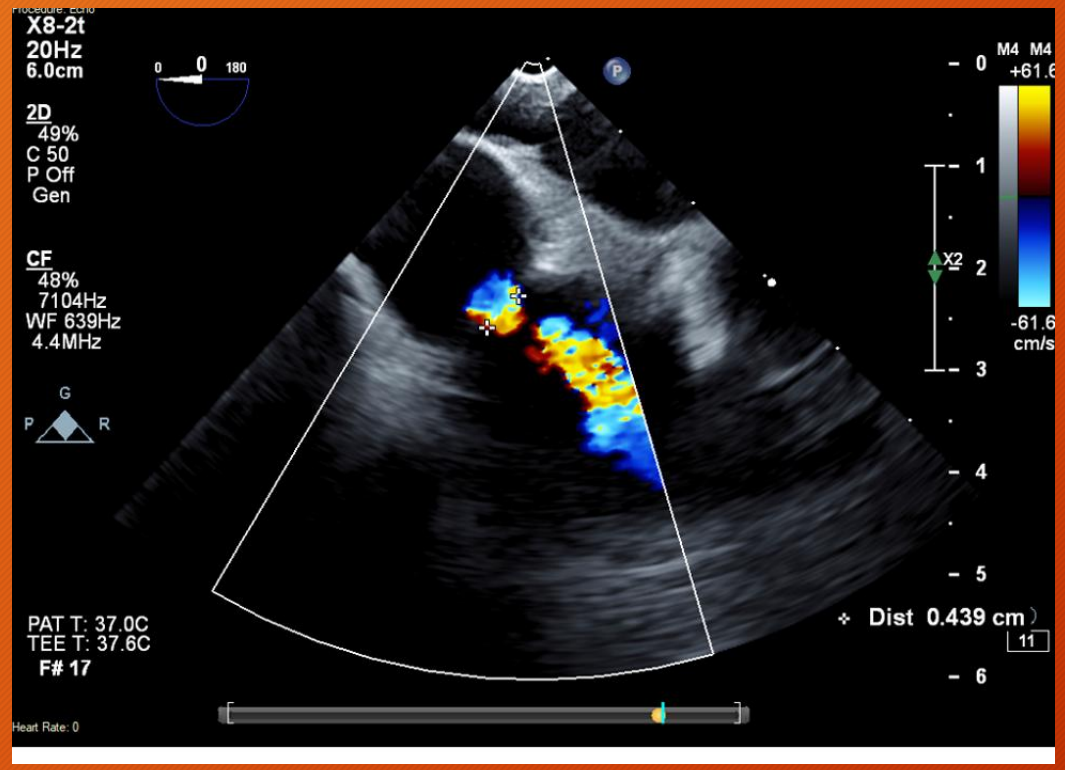
Reduced pump function

- Caused by: AV valve regurgitation, one ventricle doing the work of two, single RV, chronically raised sympathetic drive
- Management:
- Medical-
  - Four pillars of HF therapy
  - Associated side effects- often hypotension related, renal
- Intervention- collateral occlusion
- Surgical- valve repair/ replacement

Rhythm

- Atrial arrhythmia (~20% of adults) causes:
  - AV valve regurgitation causing atrial dilatation
  - Raised ventricular pressures
- Management:
  - Medication- usually anticoagulated, anti-arrhythmics
  - Ablation- very tricky due to access
- Ventricular arrhythmia-meds, ablation, subcut ICD
- Bradycardia requiring pacing ~6-30% epicardial

Fenestration



TCPC
Right to left shunt
Underwent percutaneous closure

Fenestration

- Reduces central venous pressure, improves systemic preload, avoids excess pulmonary flow and increased PVR
- Associated with desaturation, route for systemic thromboemboli

- Often closed in pre-transition or early post transition period
  - May improve saturations if closed
  - ?risks if increased PVR in higher risk groups, ?may not improve sats, veno-venous collaterals

Transplant?

- Data based on relatively small samples
- 1 year and 10 year survival based on current data suggest reduced relative to congenital heart disease with double pump
- Possible reasons:
  - Increased incidence of end organ damage prior
  - Number of previous sternotomies?
  - More complex plumbing?

Multi-system involvement

In addition:

- Liver
- GU
- Brain

Multi-system

- Lungs: ventilation: perfusion mis-match, PH, plastic bronchiolitis
- Liver: hepatic congestion, fibrosis, cirrhosis, HCC
- Renal: hypoperfusion, medication
- Gut: hypoperfusion, gut oedema and malabsorption, PLE
- GU: implications for pregnancy, menorrhagia, medication side effects including teratogenicity
- Brain: stroke, psychological aspects

- Haematology: thrombocytopenia, clotting factor derangement

Monitoring

Non-exhaustive checklist

Will vary between patients

Fontan Screening Protocol

Revised March 2022

	Baseline	Annual	3-5 yrly	IN ACCORDANCE WITH SYMPTOMS / INDICATED BY MRI OR LIVER FONTAN MDT
FBC	√	√		
U&E	√	√		
LFT	√	√		
AST	√	√		
ELF SCORE	√		√	
HEP B AND C SCREENING	√			
CLOTTING SCREEN	√	√		
12 LEAD ECG	√	√		
ECHOCARDIOGRAM	√	√		
CXR	√			√
CARDIAC MRI (IF MRI COMPATIBLE)	√		√	
LIVER ULTRASOUND	√	√		
CARDIOPULMONARY EXERCISE TEST	√		√	
24 HR TAPE				√
CARDIAC CATHETERISATION				√
UPPER GI ENDOSCOPY				√
LIVER MRI				√
LIVER MDT DISCUSSION		√		
PRE-PREGNANCY COUNSELLING	√	√		
LIFESTYLE ADVICE	√	√		
FONTAN NURSE APPOINTMENT	TRANSITION	12/12LY TEL (ALTERNATING WITH DR APPT)		
MEDICAL OP REVIEW	√	12/12 LY FtF (ALTERNATING WITH NURSE APPT)		

Conclusions

- Multiple cardiac complications in single ventricle pathophysiology
- Precipitate further cardiac complications
- In turn, precipitate non-cardiac complications
- Non cardiac complications (and medications for) can exacerbate other cardiac and non-cardiac complications

- Counselling, vigilance, timely action
- Increasing body of evidence to guide us

Questions?

Bibliography

- Incidence: Garcia AM, Beatty JT, Nakano SJ. Heart failure in single right ventricle congenital heart disease: physiological and molecular considerations. Am J Physiol Heart Circ Physiol. 2020 Apr 1;318(4):H947-H965. doi: 10.1152/ajpheart.00518.2019. Epub 2020 Feb 28. PMID: 32108525; PMCID: PMC7191494.
- Tseng SY, Siddiqui S, Di Maria MV, Hill GD, Lubert AM, Kutty S, Opotowsky AR, Possner M, Morales DLS, Quintessenza JA, Alsaied T. Atrioventricular Valve Regurgitation in Single Ventricle Heart Disease: A Common Problem Associated With Progressive Deterioration and Mortality. J Am Heart Assoc. 2020 Jun 2;9(11):e015737. doi: 10.1161/JAHA.119.015737. Epub 2020 May 16. PMID: 32419552; PMCID: PMC7429008.
- Bakhtiyar SS, Sakowitz S, Ali K, Chervu NL, Verma A, Si MS, D'Alessandro D, Benharash P. Survival After Cardiac Transplantation in Adults With Single-Ventricle Congenital Heart Disease. J Am Coll Cardiol. 2023 Sep 19;82(12):1226-1241. doi: 10.1016/j.jacc.2023.06.037. PMID: 37704313.
- Zhu A, Meza JM, Prabhu NK, McCrary AW, Allareddy V, Turek JW, Andersen ND. Survival After Intervention for Single-Ventricle Heart Disease Over 15 Years at a Single Institution. Ann Thorac Surg. 2022 Dec;114(6):2303-2312. doi: 10.1016/j.athoracsur.2022.03.060. Epub 2022 Apr 14. PMID: 35430225.
- Hassan A, Chegondi M, Porayette P. Five decades of Fontan palliation: What have we learned? What should we expect? J Int Med Res. 2023 Oct;51(10):3000605231209156. doi: 10.1177/03000605231209156. PMID: 37910851; PMCID: PMC10621298.
- Saiki H, Kuwata S, Iwamoto Y, Ishido H, Taketazu M, Masutani S, Nishida T, Senzaki H. Fenestration in the Fontan circulation as a strategy for chronic cardioprotection. Heart. 2019 Aug;105(16):1266-1272. doi: 10.1136/heartjnl-2018-314183. Epub 2019 Mar 2. PMID: 30826770
- Ozawa H, Hoashi T, Ohuchi H, Kurosaki K, Ichikawa H. Long-Term Outcomes After Fenestration Closure in High-Risk Fontan Candidates. Pediatr Cardiol. 2021 Aug;42(6):1356-1364. doi: 10.1007/s00246-021-02619-9. Epub 2021 Apr 21. PMID: 33881601.
- Corno AF, Koerner TS, Salazar JD. The pendulum of Fontan fenestration. Transl Pediatr. 2023 Jan 31;12(1):104-107. doi: 10.21037/tp-22-562. Epub 2023 Jan 16. PMID: 36798929; PMCID: PMC9926132.