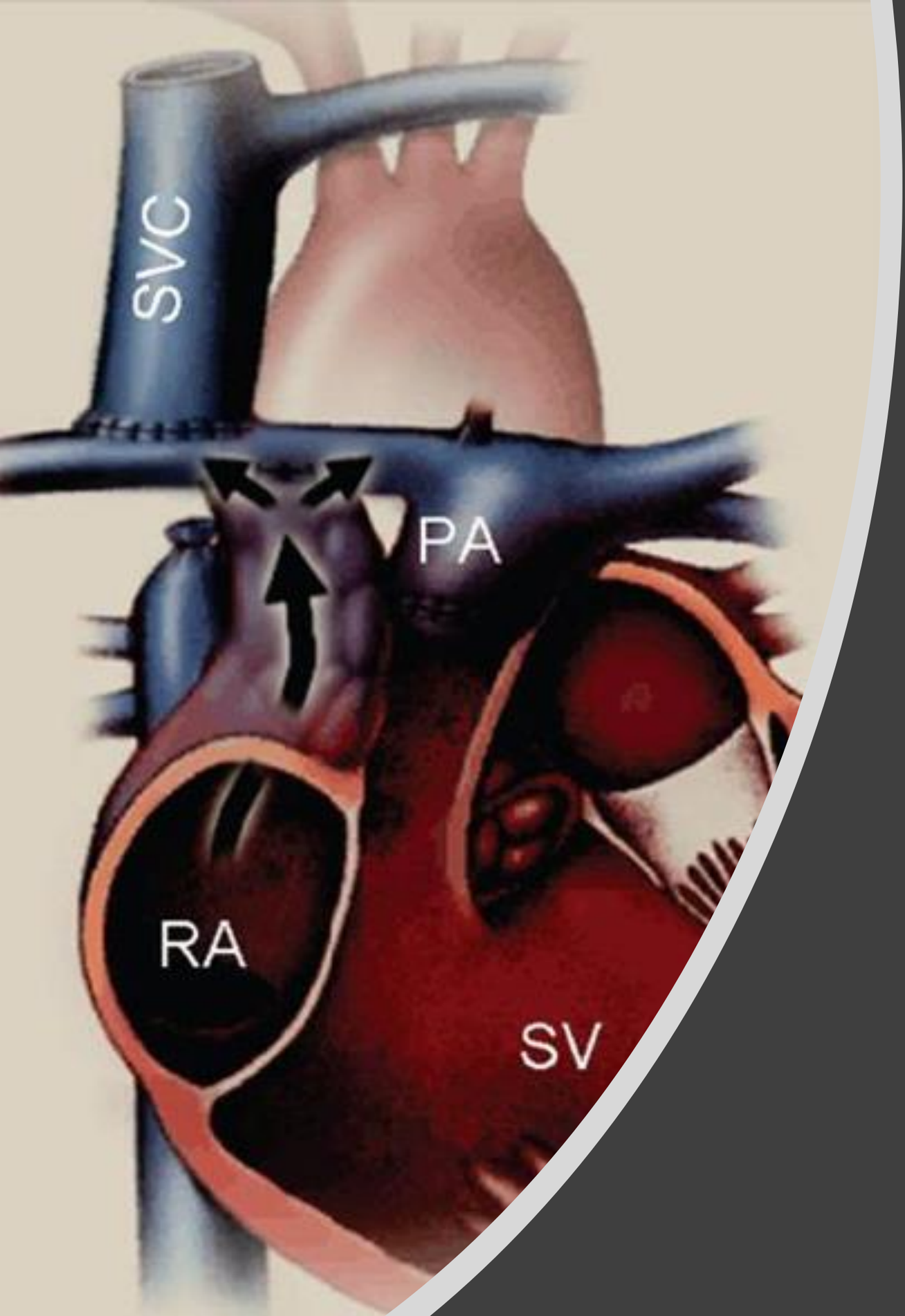


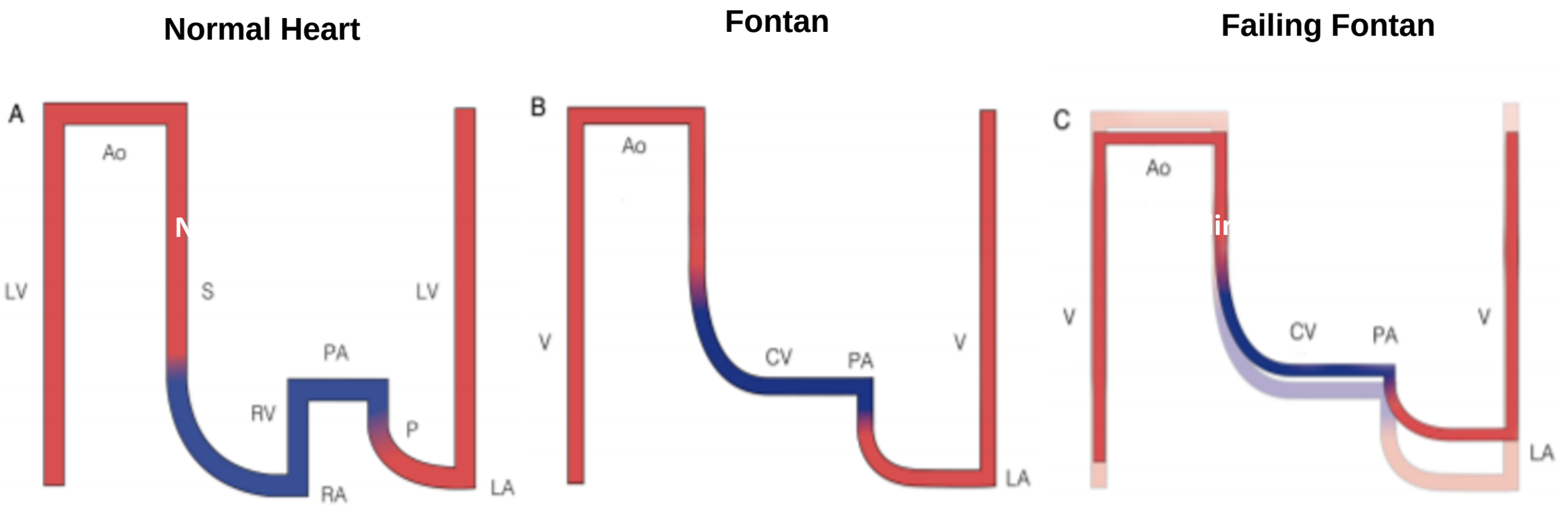
Failing Fontan

Dr Louise Coats

Adult Congenital Cardiologist

Freeman Hospital and Newcastle University

Newcastle upon Tyne, UK

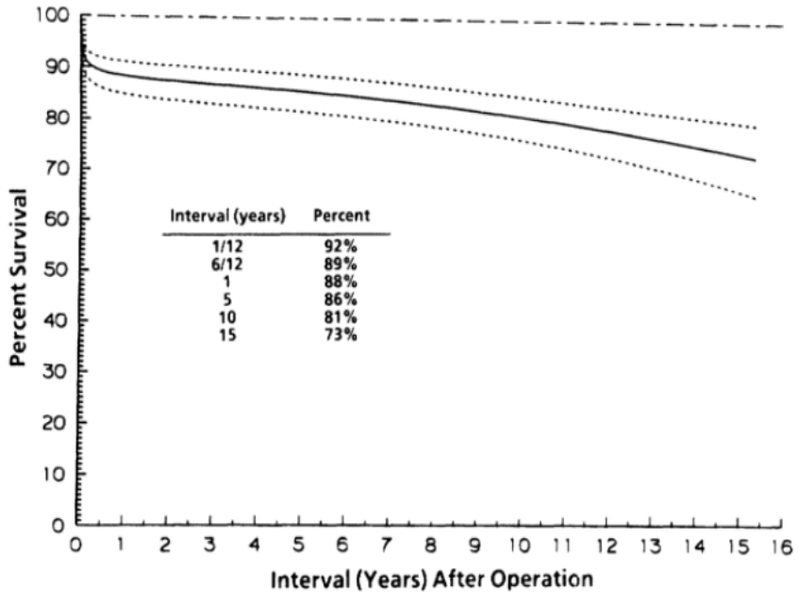


Why does the Fontan Fail?

- Non-pulsatile pulmonary blood flow
- → systemic venous hypertension
- → ventricular preload deficiency → low cardiac output
- → increased afterload

Gewillig et al. Heart 2016

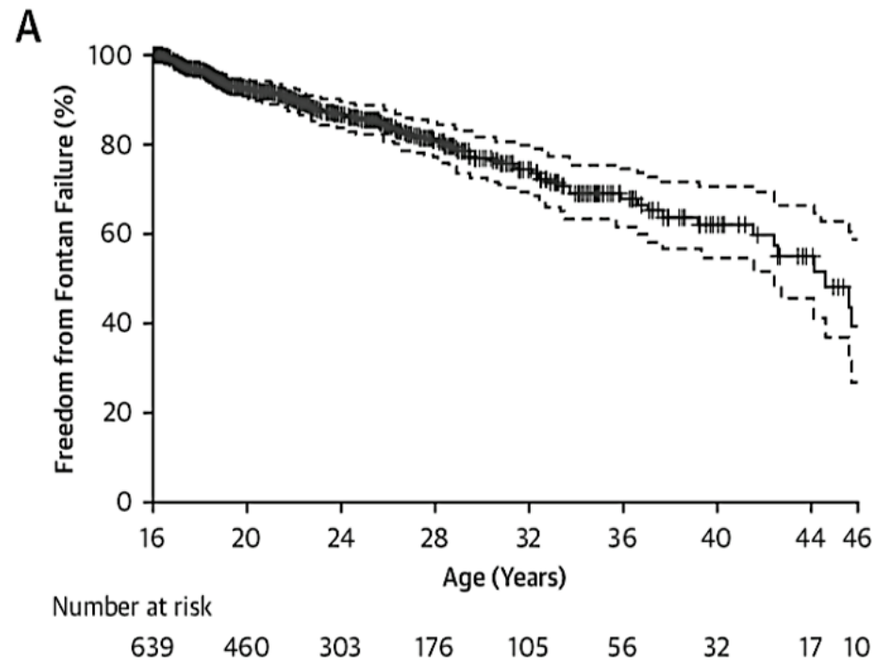
Every Fontan Circulation Eventually Fails



Years Post Fontan: 1 y → 10 y → 15 y

Mortality: 12% → 19% → 27%

Fontan F et al. Circ. 1990



Patient Age: 30 y → 40 y → 50 y

Fontan Failure: 23% → 38% → 70%

Dennis et al. JACC 2018

	Patient's age (years)								
	20	**25**	**30**	**35**	**40**	**45**	**50**	**55**	**60**
ASD	25	26	32	38	42	47	52	57	61
Valvar disease	29	31	36	40	45	49	54	59	63
VSD	28	30	36	40	44	49	53	59	63
Aortic Coarctation	32	33	38	43	47	52	56	62	66
AVSD	33	34	39	44	48	52	57	62	66
Marfan syndrome	37	38	42	46	50	54	59	64	68
Tetralogy of Fallot	37	38	42	47	50	54	60	65	69
Ebstein anomaly	42	43	47	51	54	59	63	68	72
Systemic RV	46	48	51	55	59	63	67	72	76
Eisenmenger syndrome	57	58	62	65	69	73	77	81	84
Complex CHD	58	59	63	67	70	74	78	82	85
Fontan	64	65	68	72	75	78	82	86	91

Age differenc

- >40
- 30-40
- 20-30
- 10-20
- 5-10
- 2-5
- <2

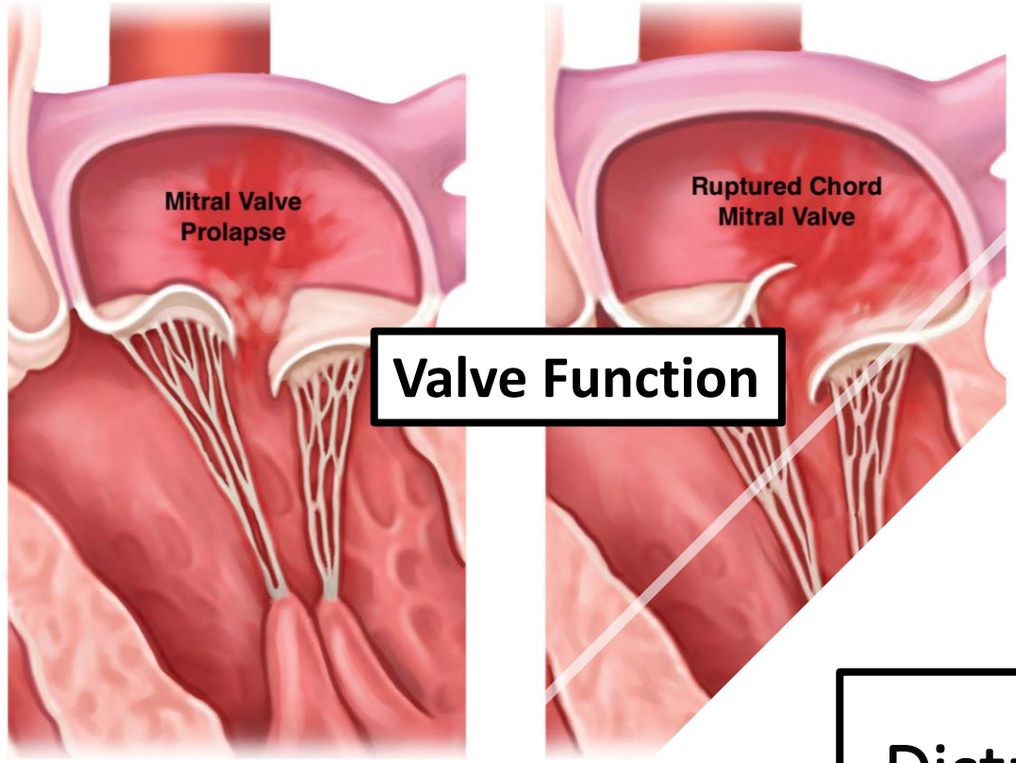
Diller GP et al. Circulation 2015

When does the Fontan Fail?

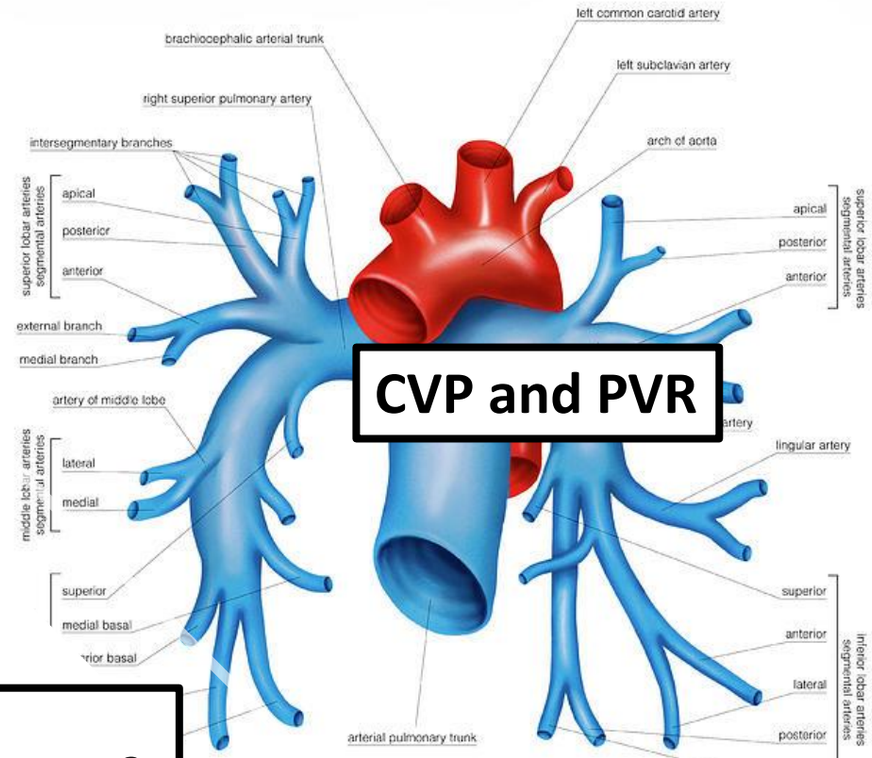
Fontan patients requiring hospitalisation due to heart failure have a high mortality

24% at 1 year
35% at 3 years

(CONCOR) Zomer et al. Int J Cardiol 2013

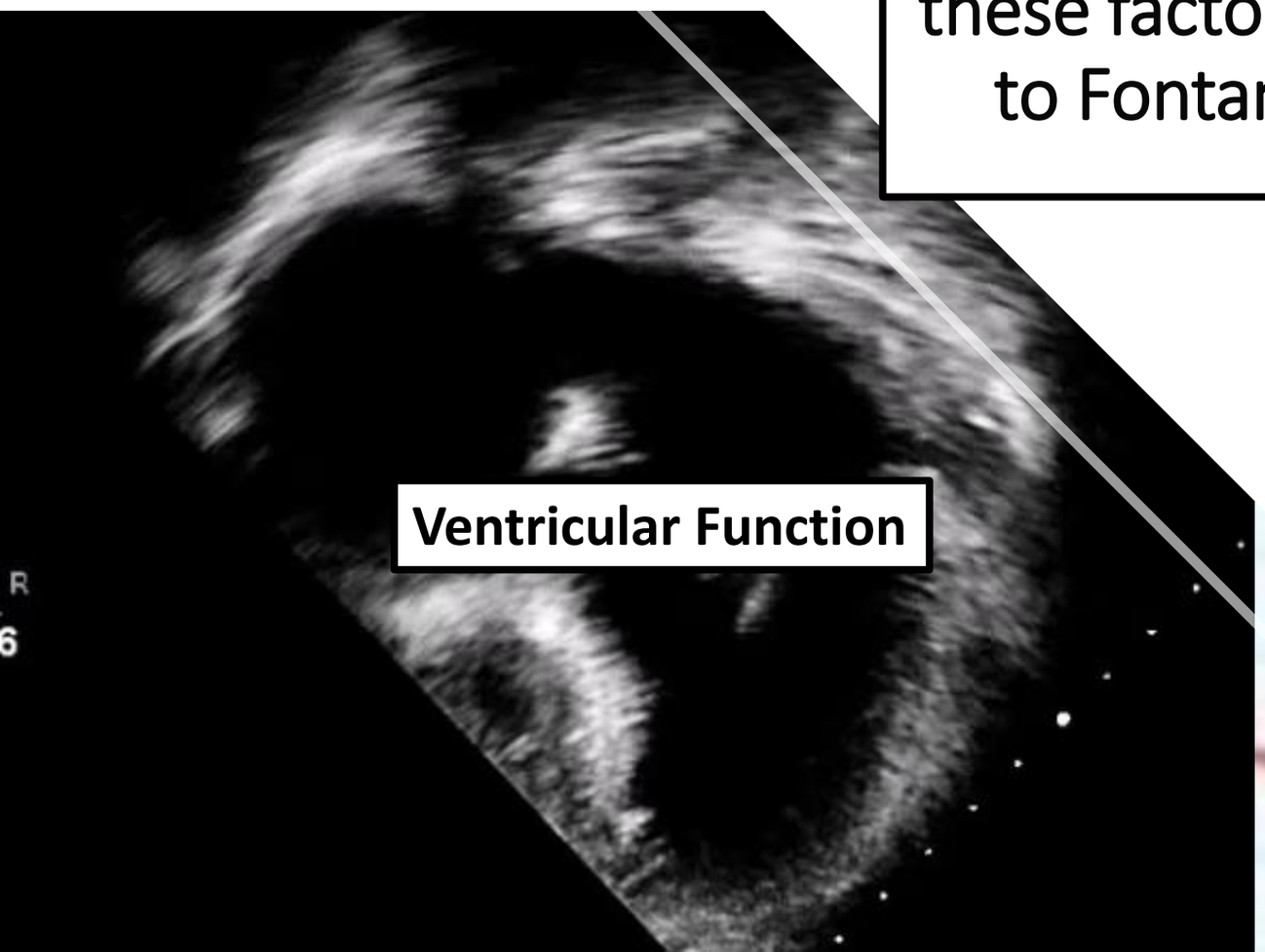


Valve Function

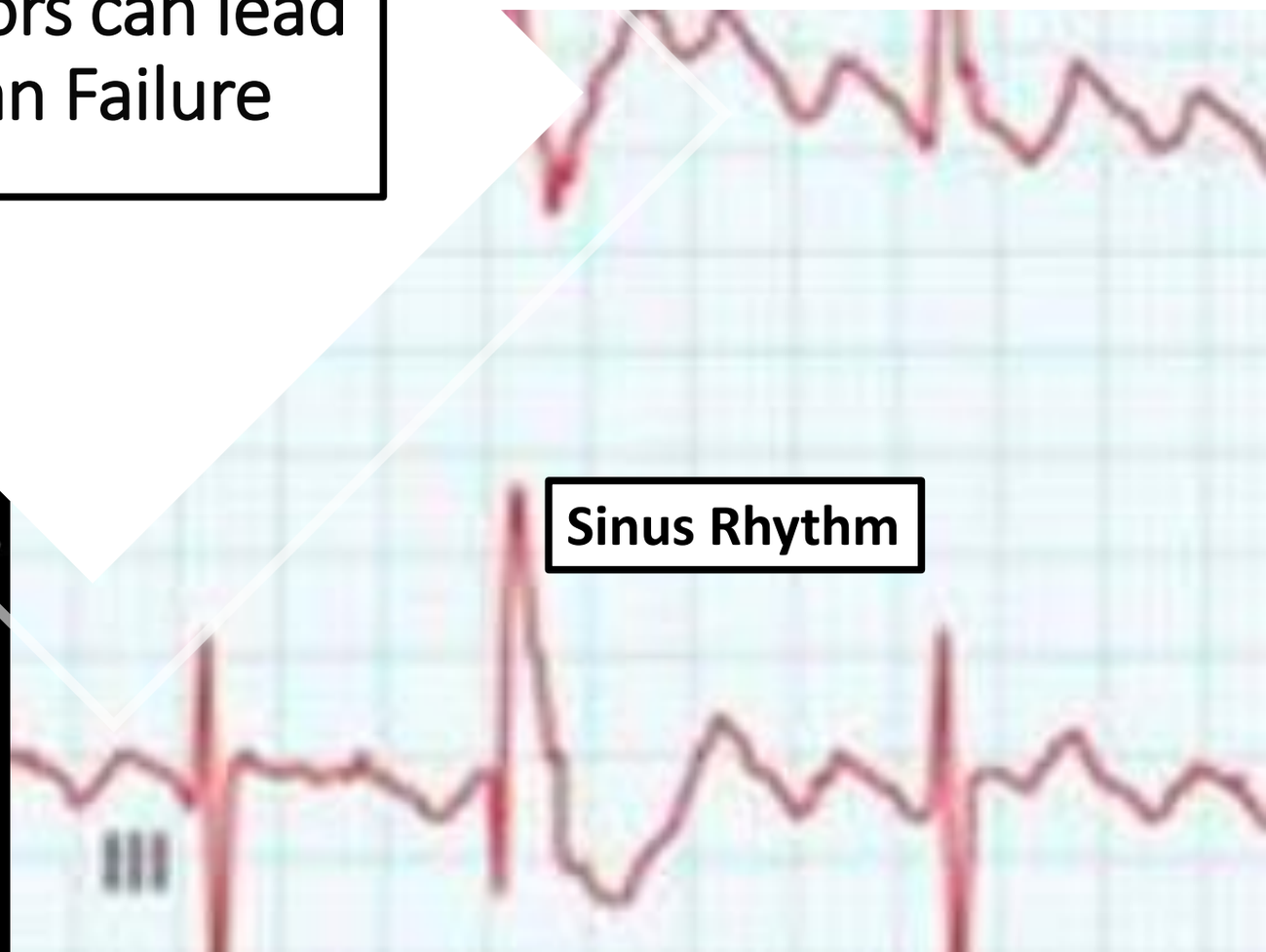


CVP and PVR

Disturbance of any of these factors can lead to Fontan Failure



Ventricular Function



Sinus Rhythm

Fontan failure with **preserved** ventricular function

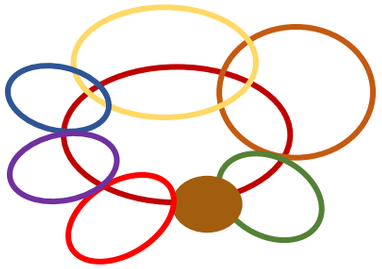
Fontan failure with **impaired** ventricular function

Two Types of Fontan Failure

Fontan Failure with Preserved SV Function

- ✓Sinus Rhythm
- ✗CVP and PVR
- ✓Ventricular Function
- ✓Valves

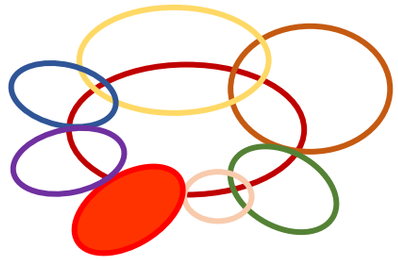
Osteoporosis



- Treatment of PLE
- Diet
- Vitamin D + Ca
- Exercise

Coagulation disorders due to:

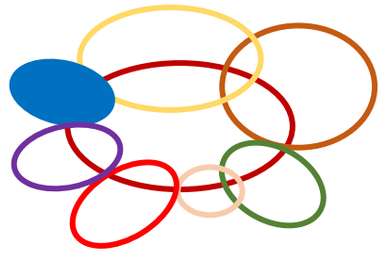
- Pro-inflammatory condition
- Low flow
- Liver dysfunction + portal hypertension
- PLE



Management:

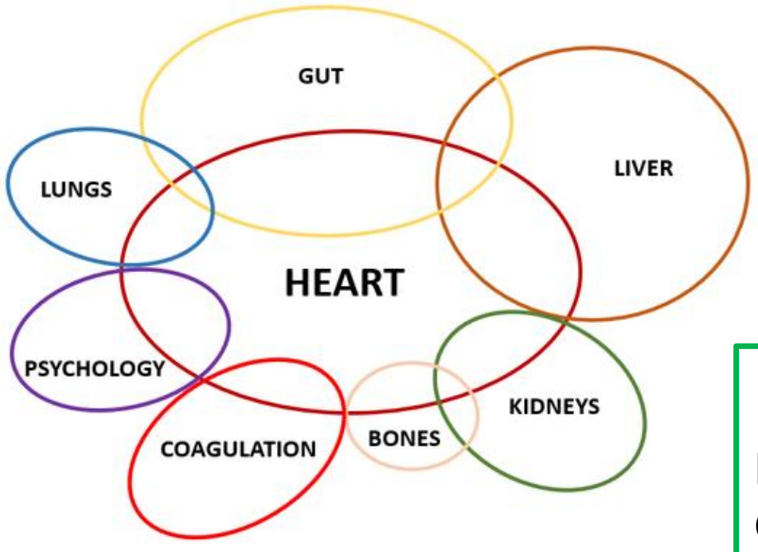
- VKA, ASA, DOAC

Plastic Bronchitis



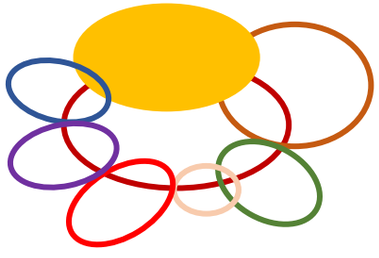
Management:

- Diuretics + MRA
- Bronchodilators
- Physiotherapy
- Diet
- Catheter intervention on lymphobronchial communications



Protein Losing Enteropathy

- ↑ Venous Pressure
- Lymphatic congestion
- Mesenteric vasc inflammation



Management:

- diet, medication, cardiac causes, anaemia, sleep apnoea

Psychology

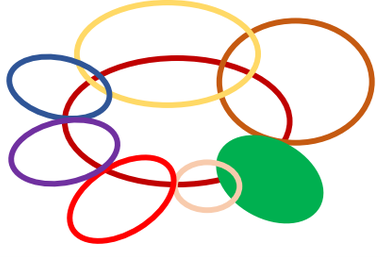
- Poorer QOL
- Negative Illness perception

Management

- ?Physical Activity ?Transition

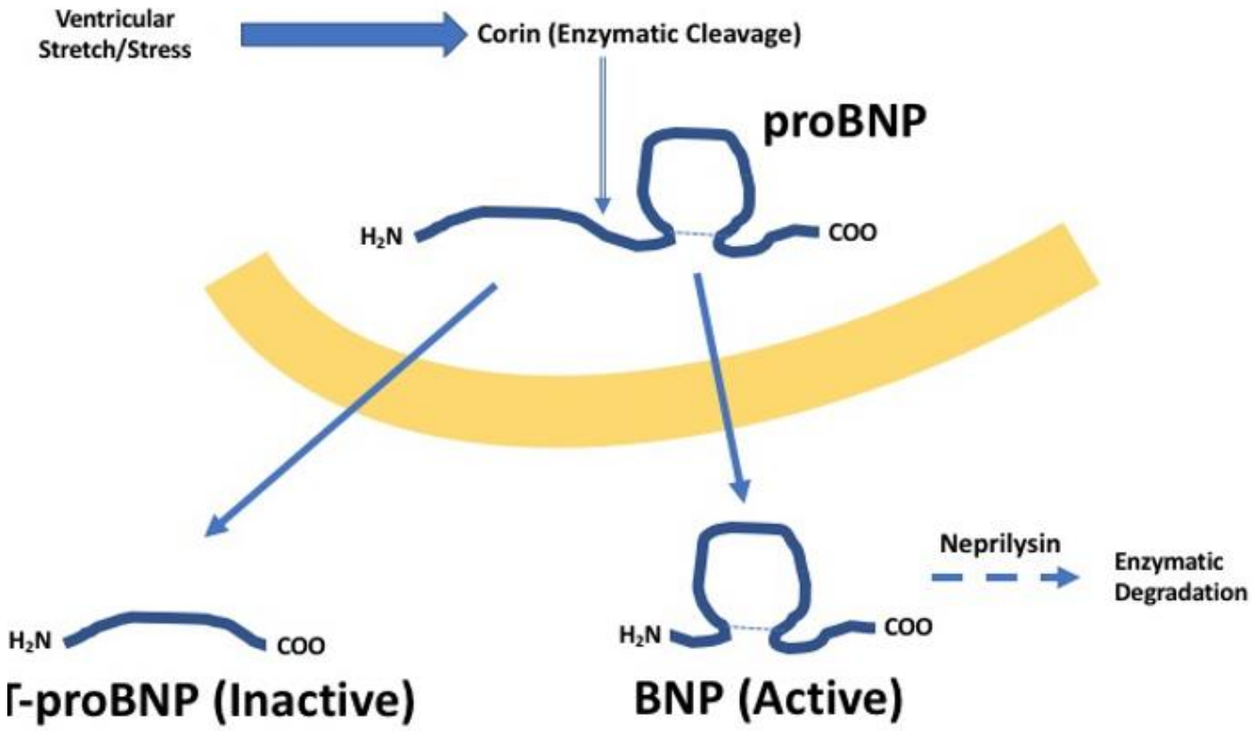
Renal Dysfunction

Reduced blood flow, venous congestion, cyanosis
Autonomic dysfunction
Extracellular fluid retention



When does Fontan heart failure management start?

The Role of BNP in Fontan?



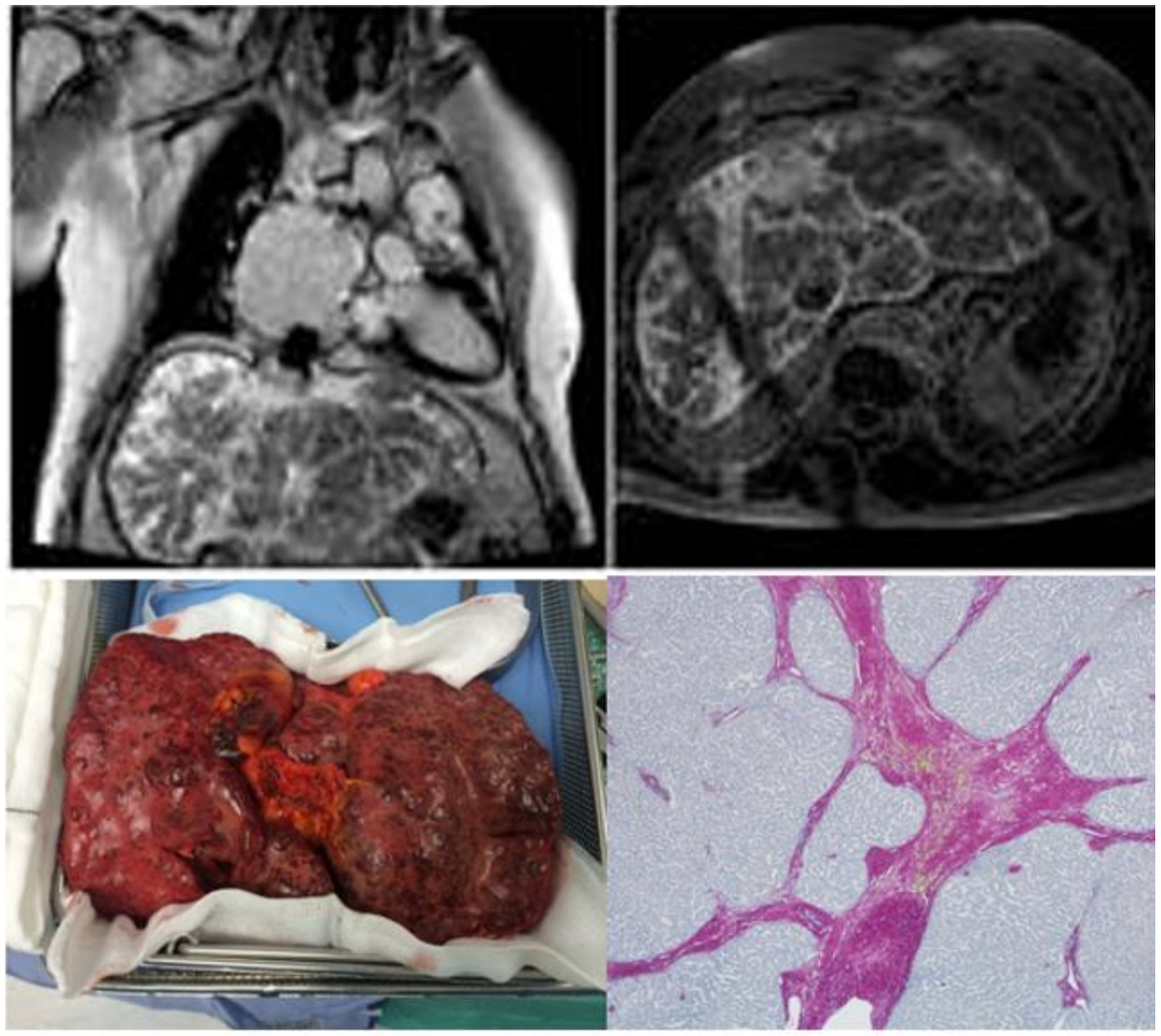
- Higher in AP Fontan
- Normal in majority up to 15 years after TCPC
- Predictor of late morbidity and mortality
- But not **sensitive**

Cardiopulmonary Exercise Testing

- **Strong predictors of death and transplantation:**
  - Declining peak VO2
  - Peak VO2 < 16.6 mL/kg/min
  - Peak heart rate < 122 bpm,
  - Heart rate reserve < 72 bpm
  - Exercise Oscillatory Ventilation

Udholm et al. OpenHeart 2018

Single Measures less Helpful…..Surveillance and Longitudinal Measurement is Key



Liver Disease is Universal and often progresses Silently

- Monitor progression
- Detect hepatocellular carcinoma early

Osteoporosis

- Treatment of PLE
- Diet
- Vitamin D + Ca
- Exercise

Coagulation disorders due to:

- Pro-inflammatory condition
- Low flow
- Liver dysfunction + portal hypertension
- PLE

Management:

- VKA, ASA, DOAC

Plastic Bronchitis

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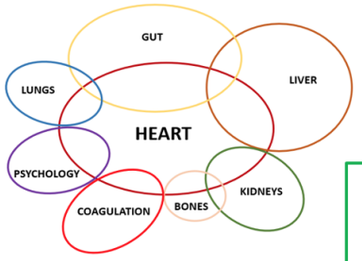


Psychology

- Poorer QOL
- Negative Illness perception

Management

- ?Physical Activity ?Transition

Renal Dysfunction

Reduced blood flow, venous congestion, cyanosis
Autonomic dysfunction
Extracellular fluid retention

CHUF
Children's Heart Unit Fund

Fontan Failure with Impaired SV Function

Medical Therapy

Betablockers

- Carvedilol: **Ishibashi et al.,** *Circ J 2012*

ACE inhibitors

- Enalapril: **Hsu et al.,** *Circulation 2010*

MRA

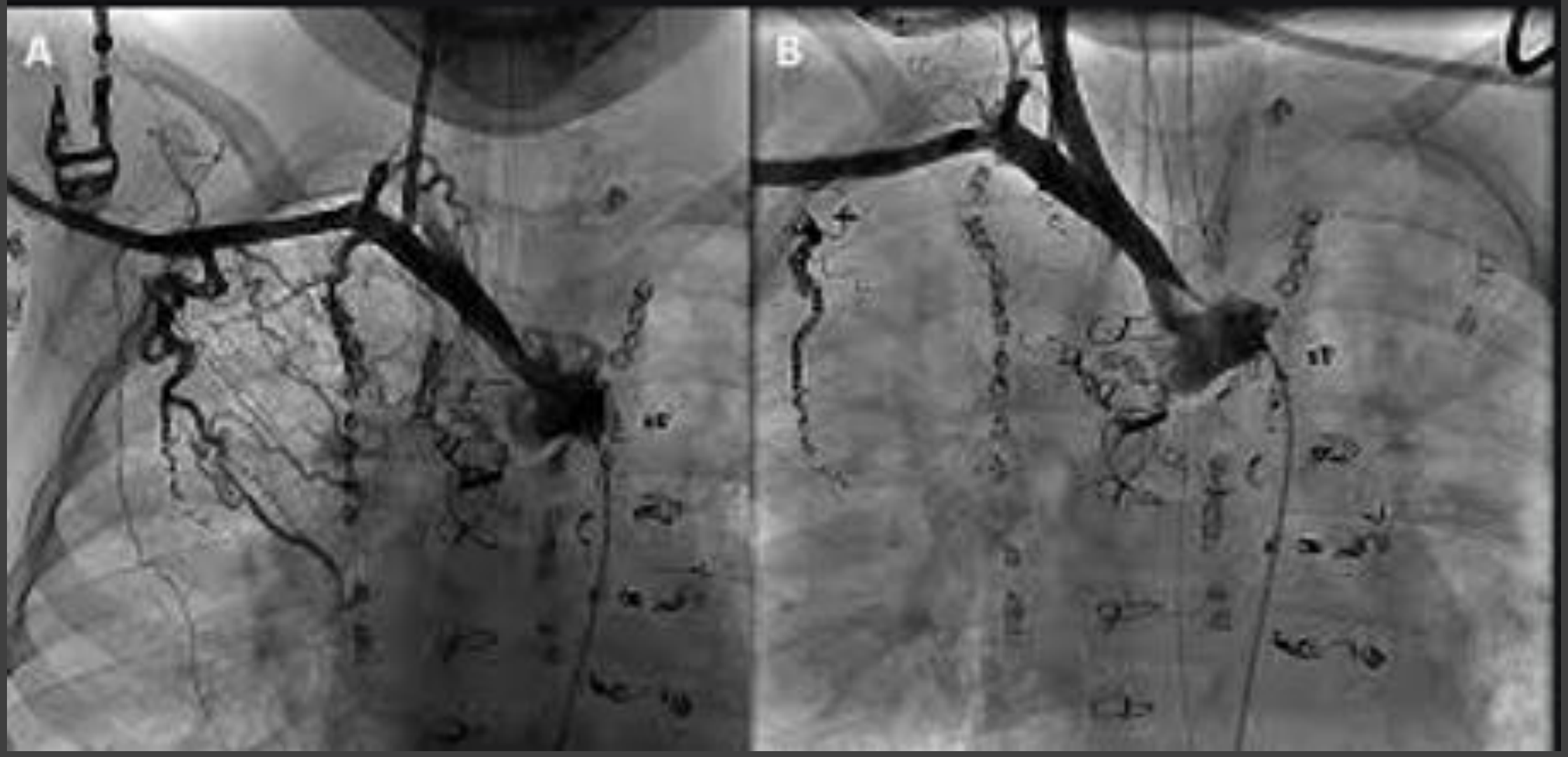
- Spironolactone: **Mahle et al.,** *Congenit Heart Dis 2009*

Diuretics

- Risk of reduced preload with cardio-renal syndrome

PDE-5

- Sildenafil: **Giardini et al.,** *Eur Heart J 2008*



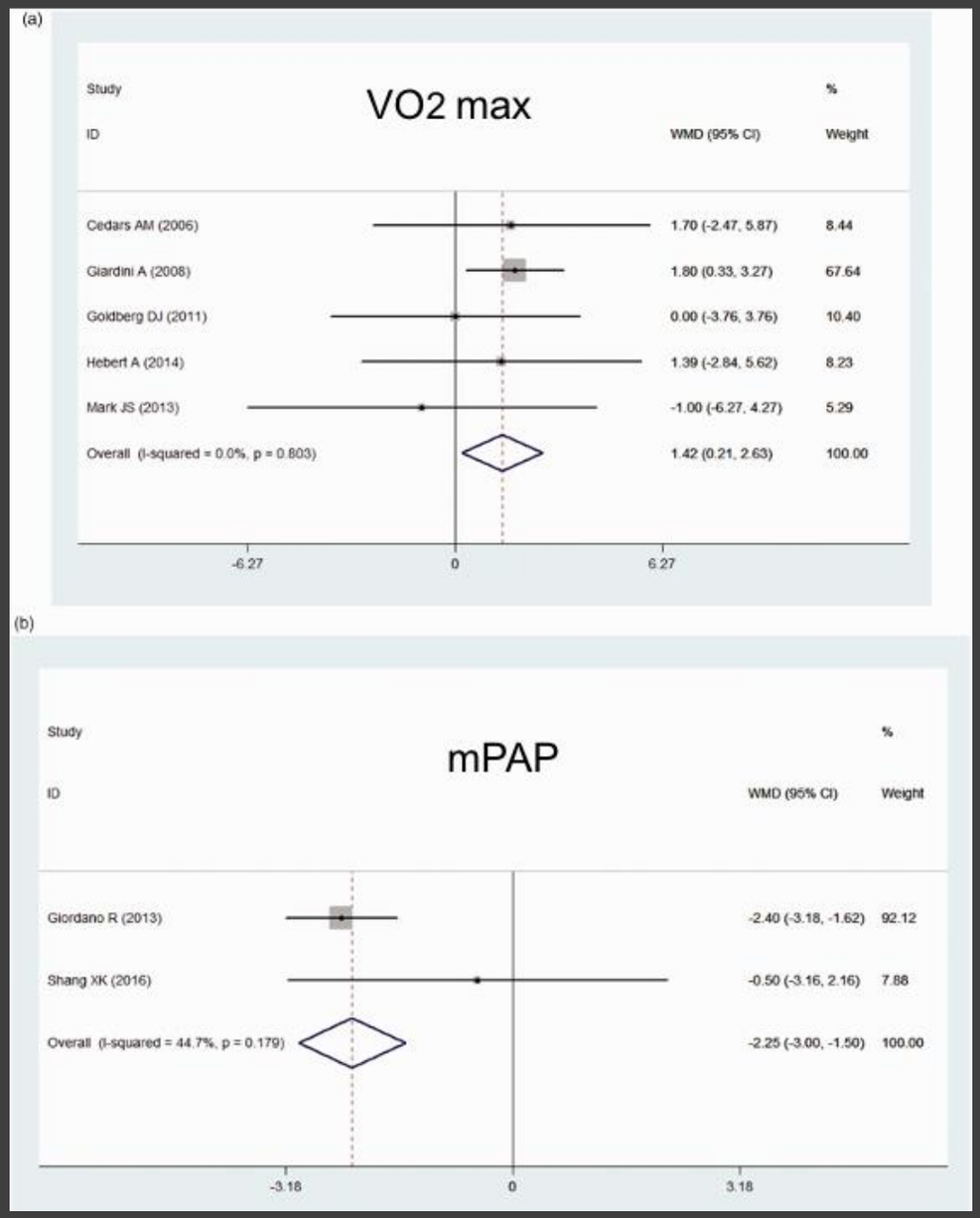
Fontan Failure with Impaired SV Function

Consider coil embolization of AP collaterals

Fontan Failure with Preserved SV Function

Creation of Fontan fenestration

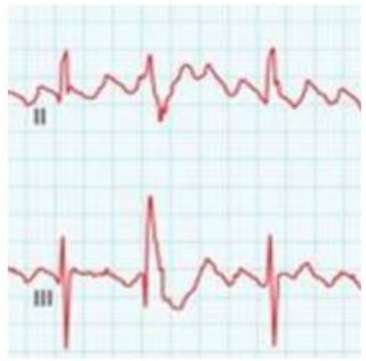
Pulmonary vasodilator therapy



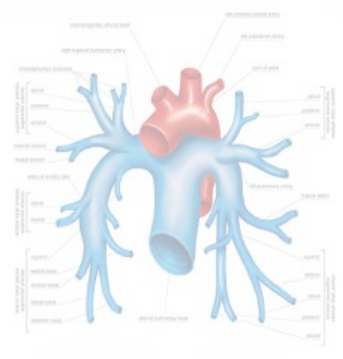
Wuang W et al. Pulm Circ 2019;9(1):2045894018790450.

Disturbance of any of the Following Factors can lead to Fontan Failure

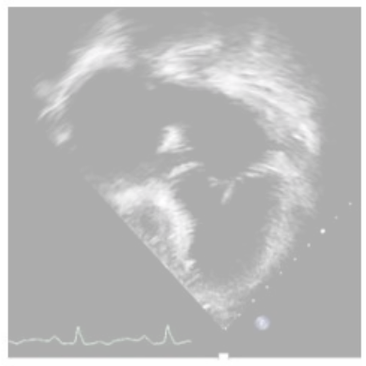
Medication
DCC
Ablation
Conversion



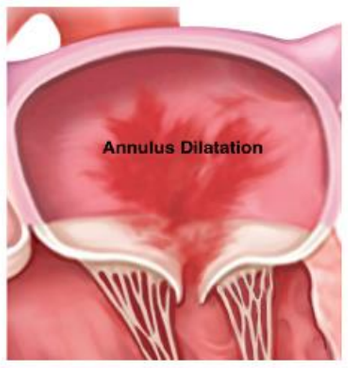
Sinus Rhythm



CVP and PVR



Ventricular Function



Valves

Conventional
Intervention
or
Surgery

Fontan patients requiring hospitalisation due to heart failure have a high mortality

---

24% at 1 year

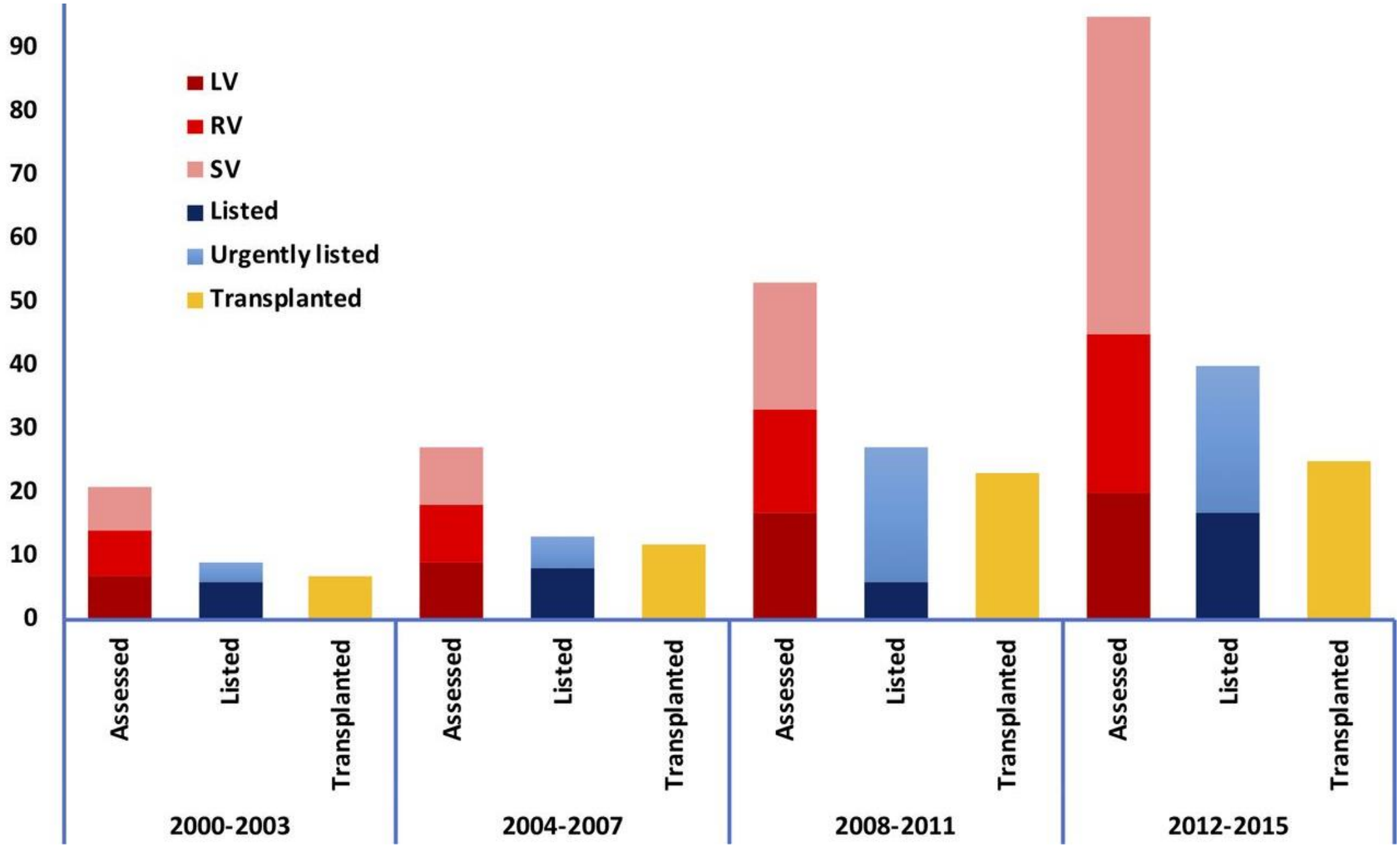
35% at 3 years

(CONCOR) Zomer et al. Int J Cardiol 2013

Advanced Fontan Failure Therapies

Transplant

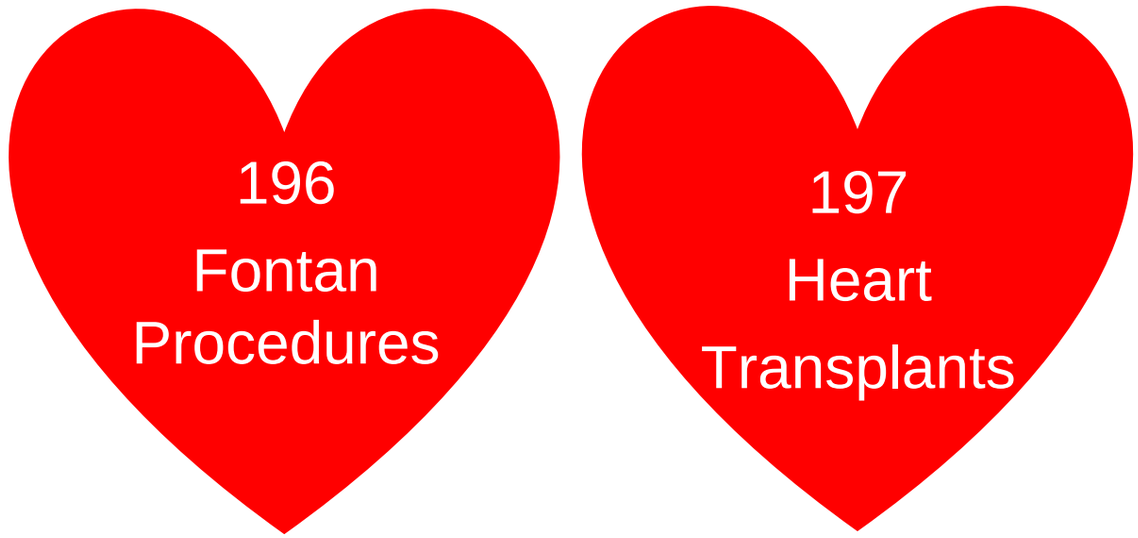
Mechanical Circulatory Support



Trends in Assessments for OHT Freeman Hospital, Newcastle upon Tyne 2000-2015

Crossland DS et al. Heart 2019

Challenges



- Complex Anatomy
- Venous Access
- Previous Sternotomies
- Collaterals
- FALD
- Cardiac Cachexia
- Psychology/Support

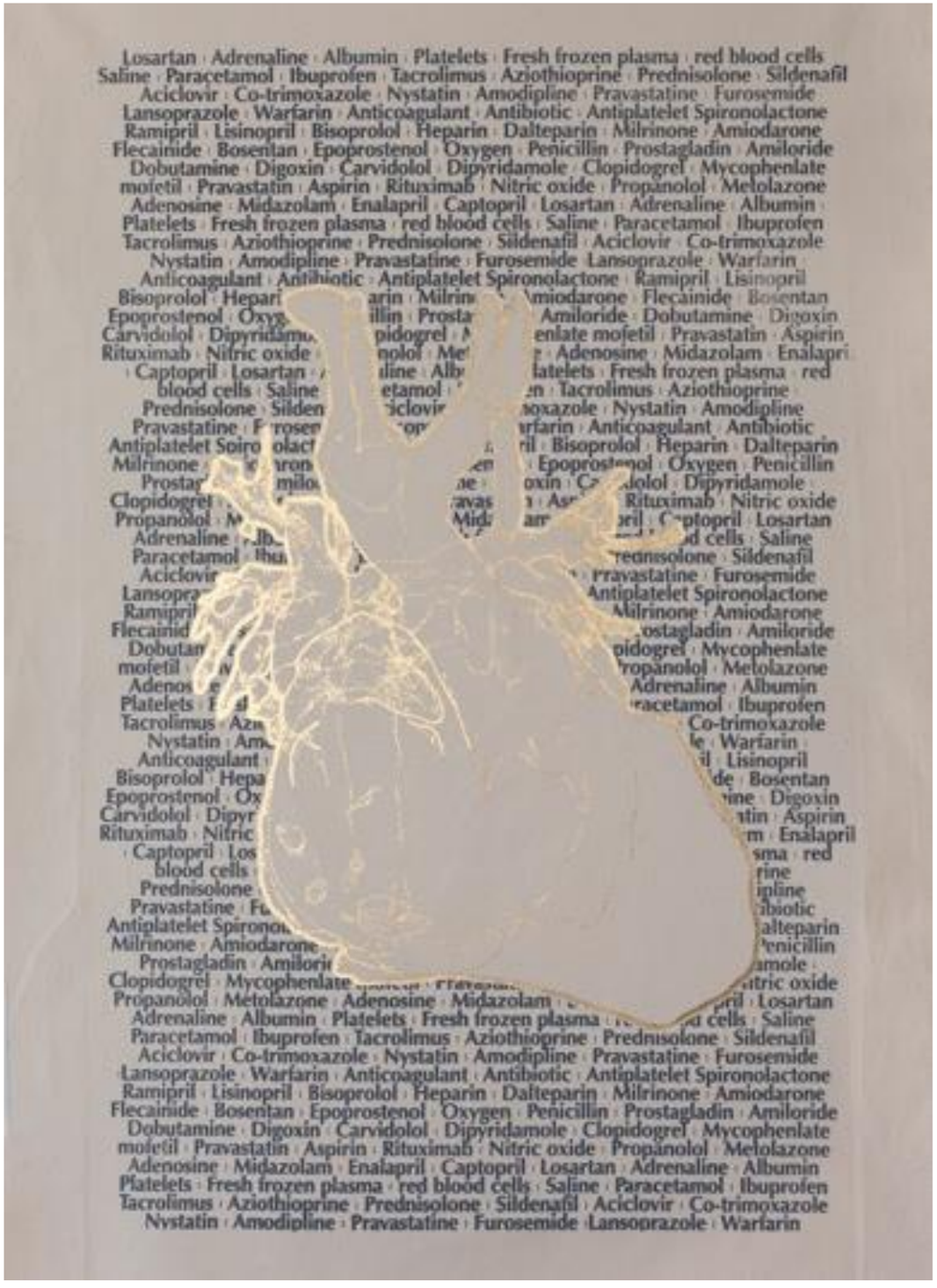
Early Referral is Key

- Is there a perception gap between what we think is important and what patients think is important?
- Most of our evidence is focused on mortality – is it the right outcome?
- What do patients really think/feel/want?

What Matters to Fontan Patients?

The Future for the Patient

Regular follow-up
Healthy lifestyle
Participation in Research



Medication by Sophie Layton

www.insidetheheart.org

Thank You

NENC-CHDN

North East and North Cumbria Congenital Heart Disease Network

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