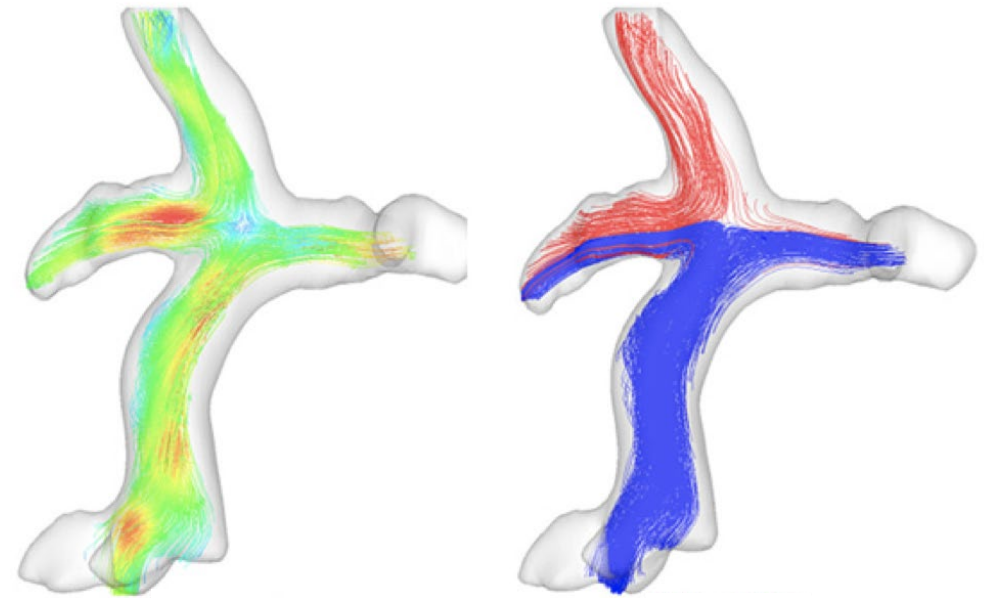
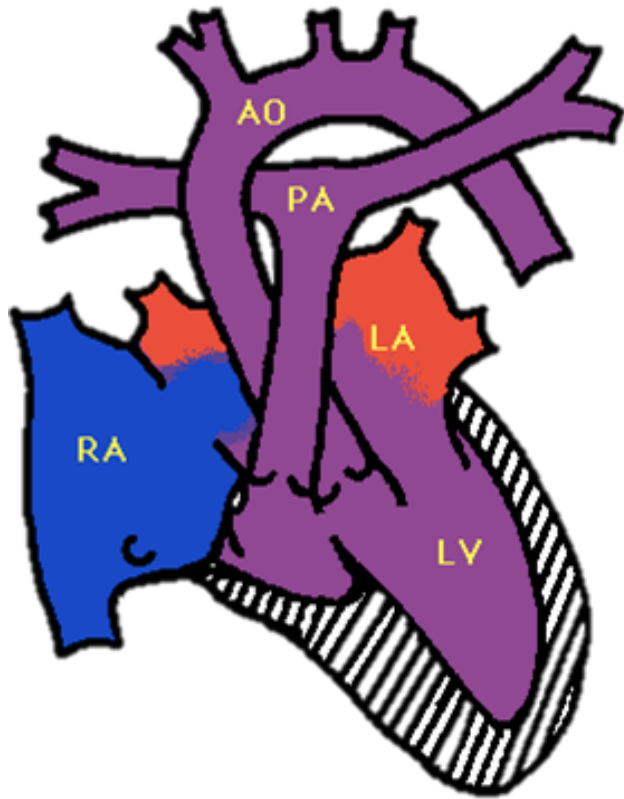


Fontan circulation - late health considerations

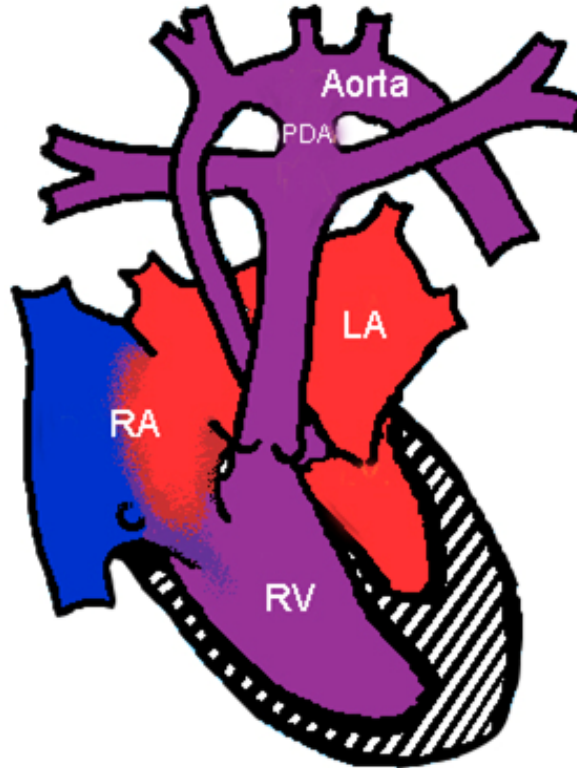
Rafael Alonso Gonzalez
Director, Toronto ACHD Program
Assistant professor of Medicine
Peter Munk Cardiac Centre
Toronto - Canada



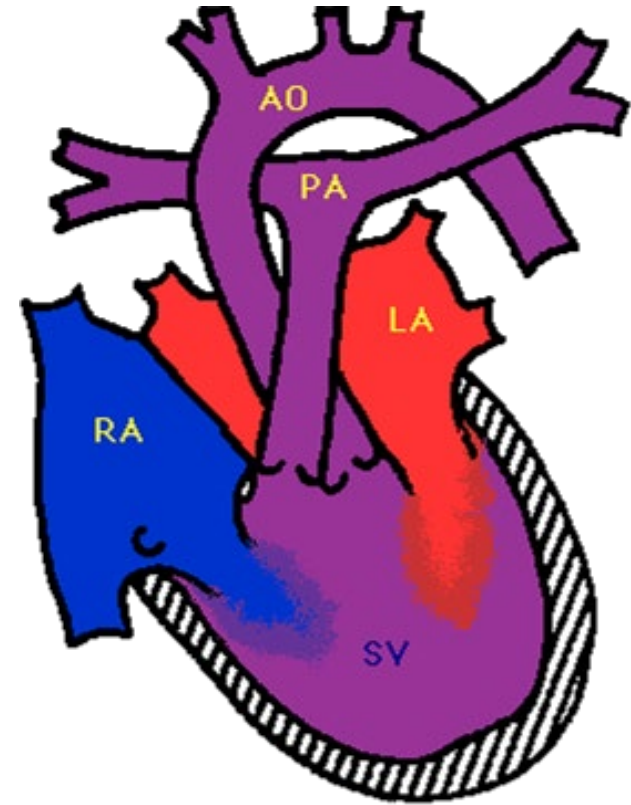
Diverse morphology



Tricuspid atresia
Dominant left ventricle

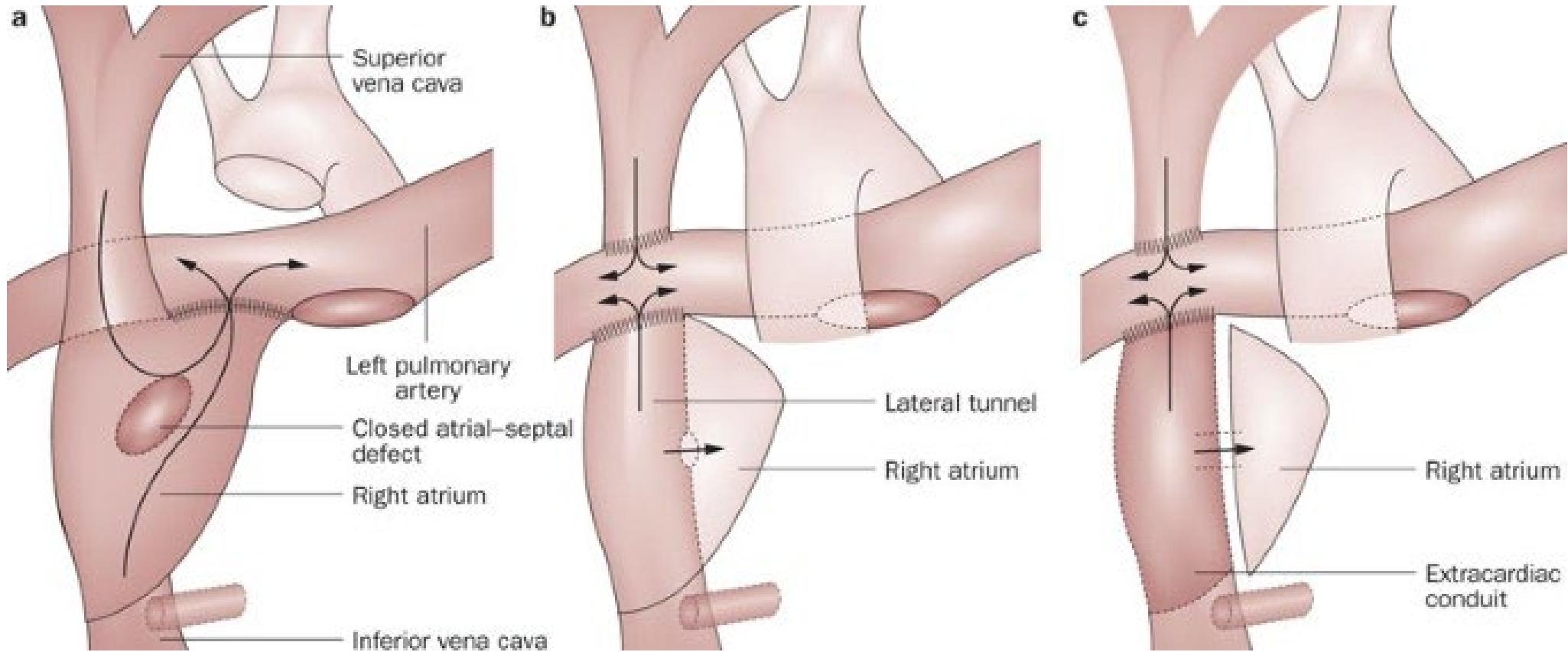


Hypoplastic LH syndrome
Dominant right ventricle



Univentricular heart
Single ventricle

Evolution of Fontan surgery



**Atrio-pulmonary
Fontan**

Lateral tunnel

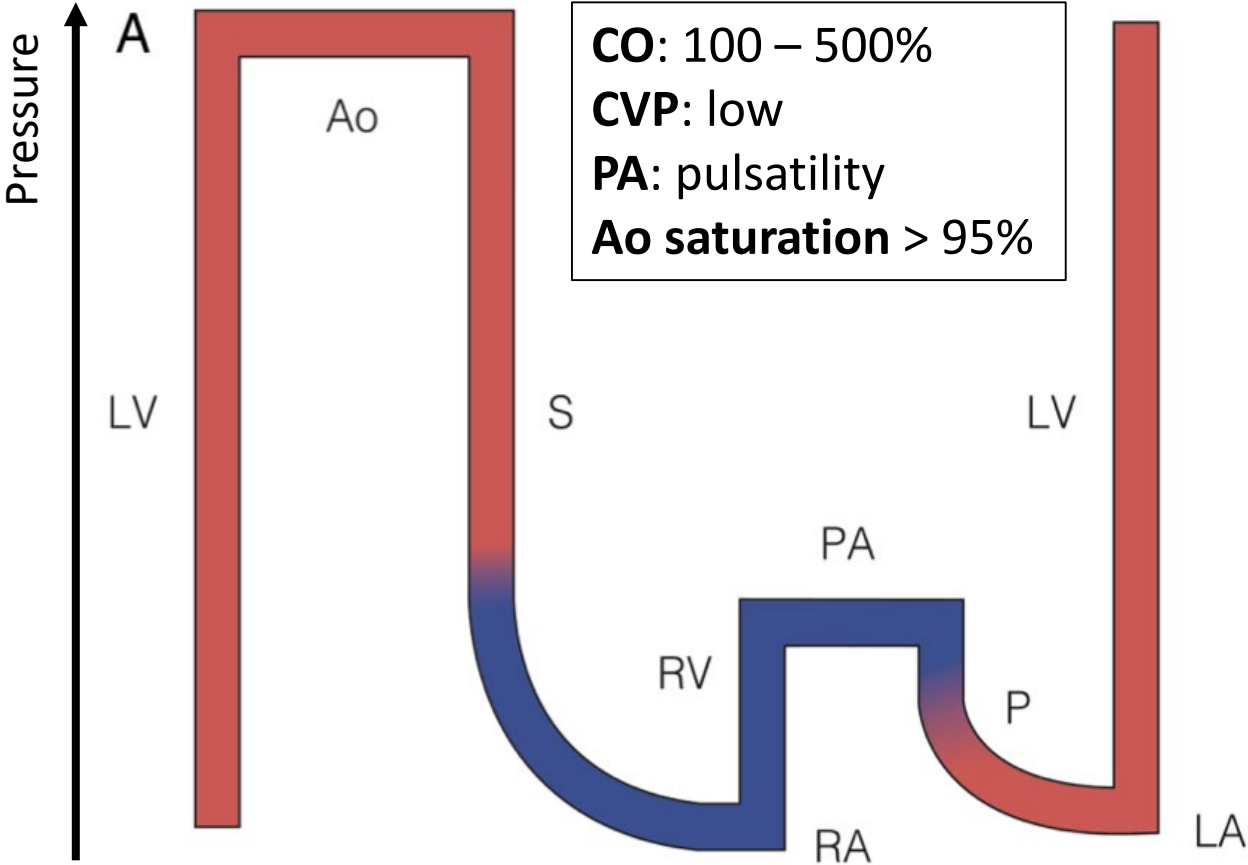
Extra-cardiac conduit

Total cavo-pulmonary connection

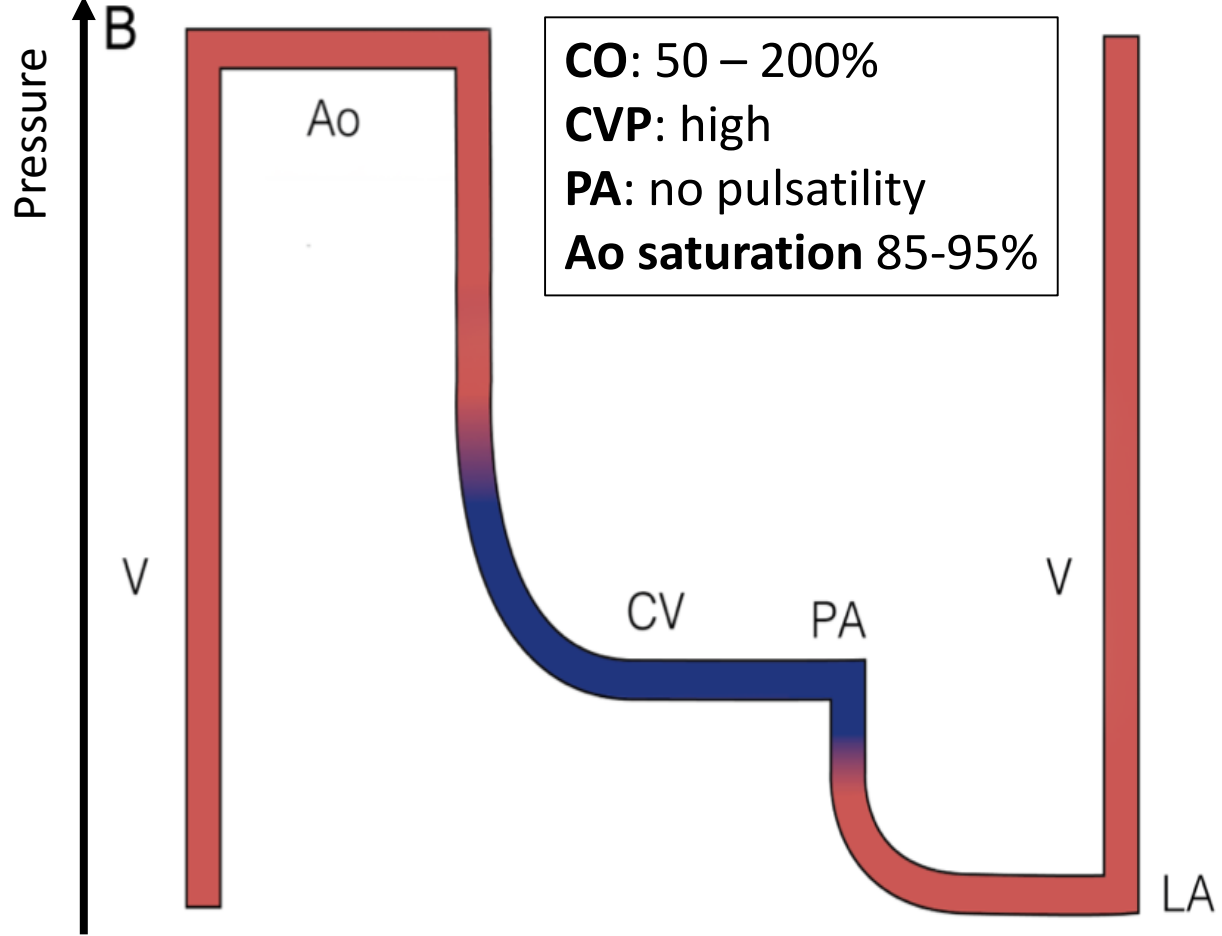
Normal circulation vs Fontan

circulation

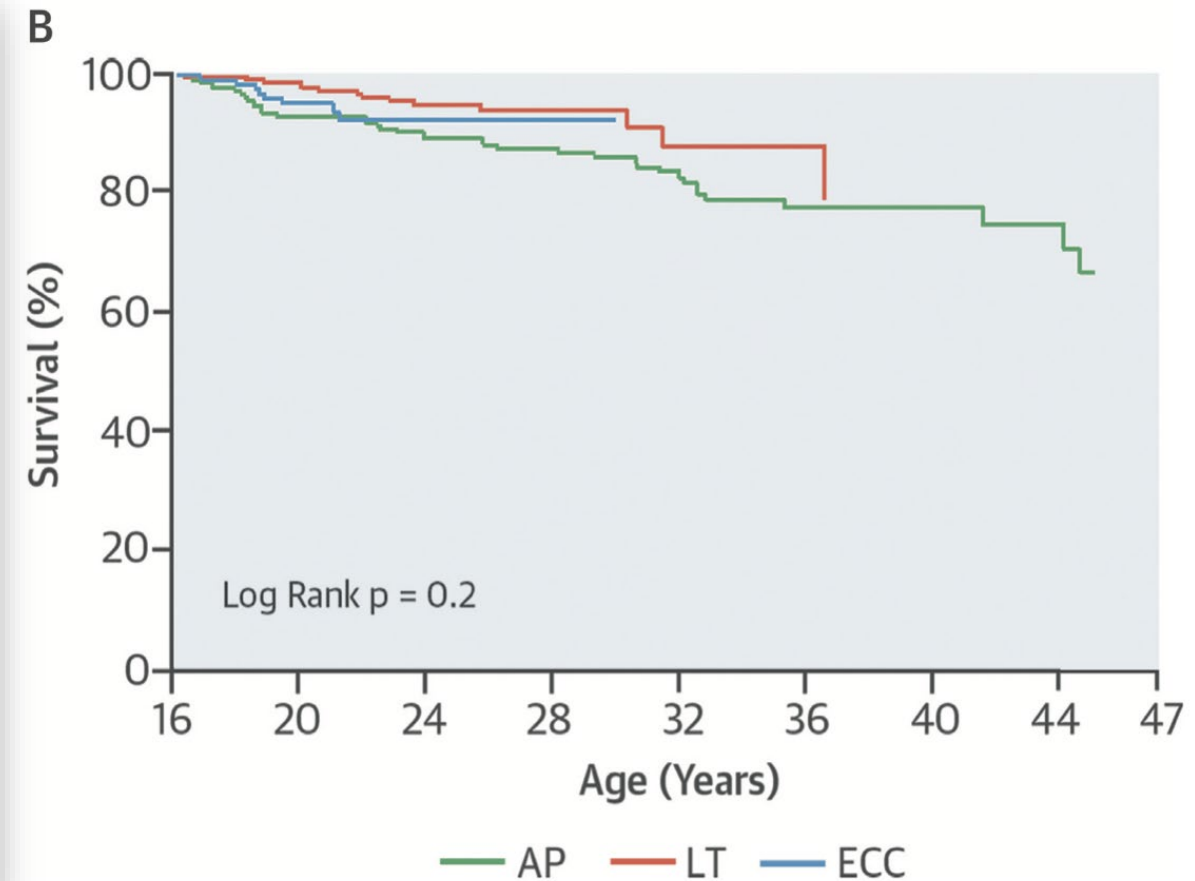
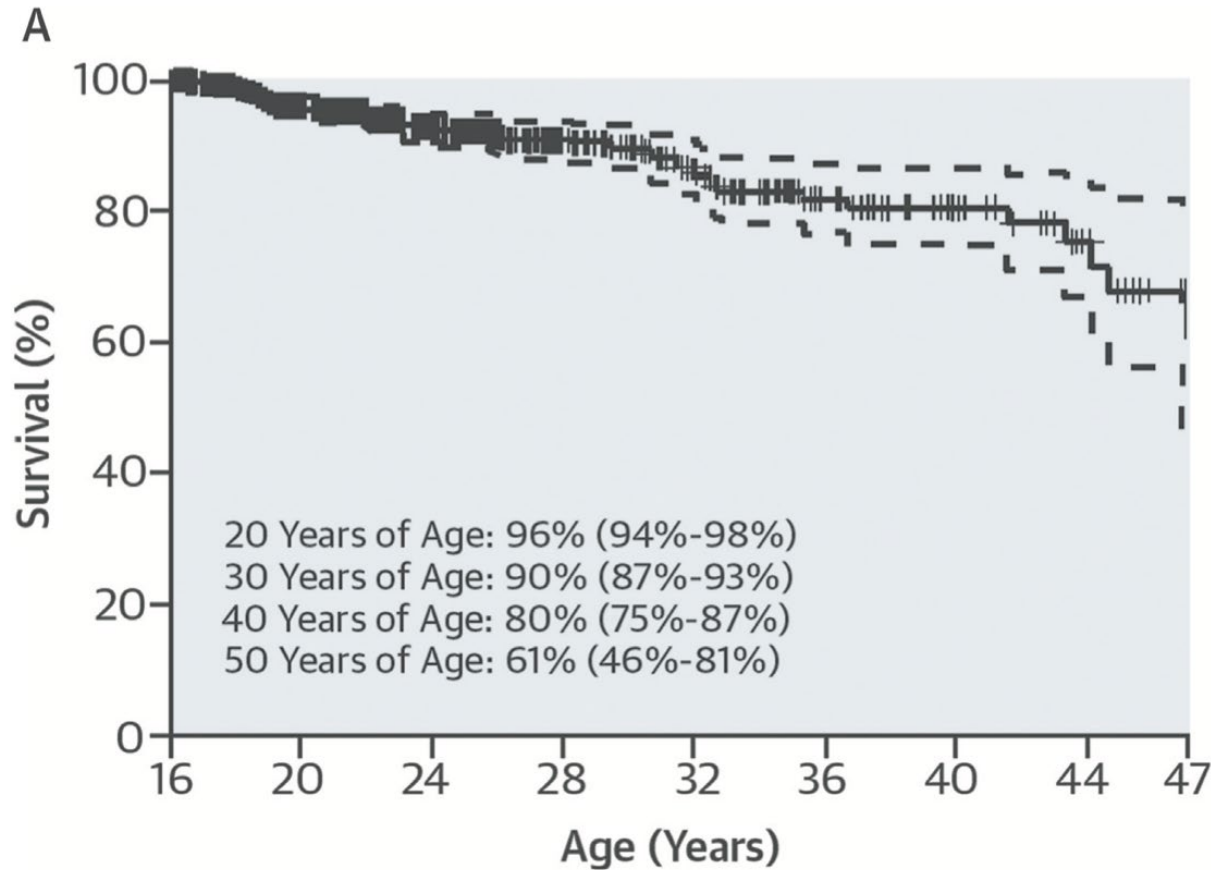
Normal circulation



Fontan circulation

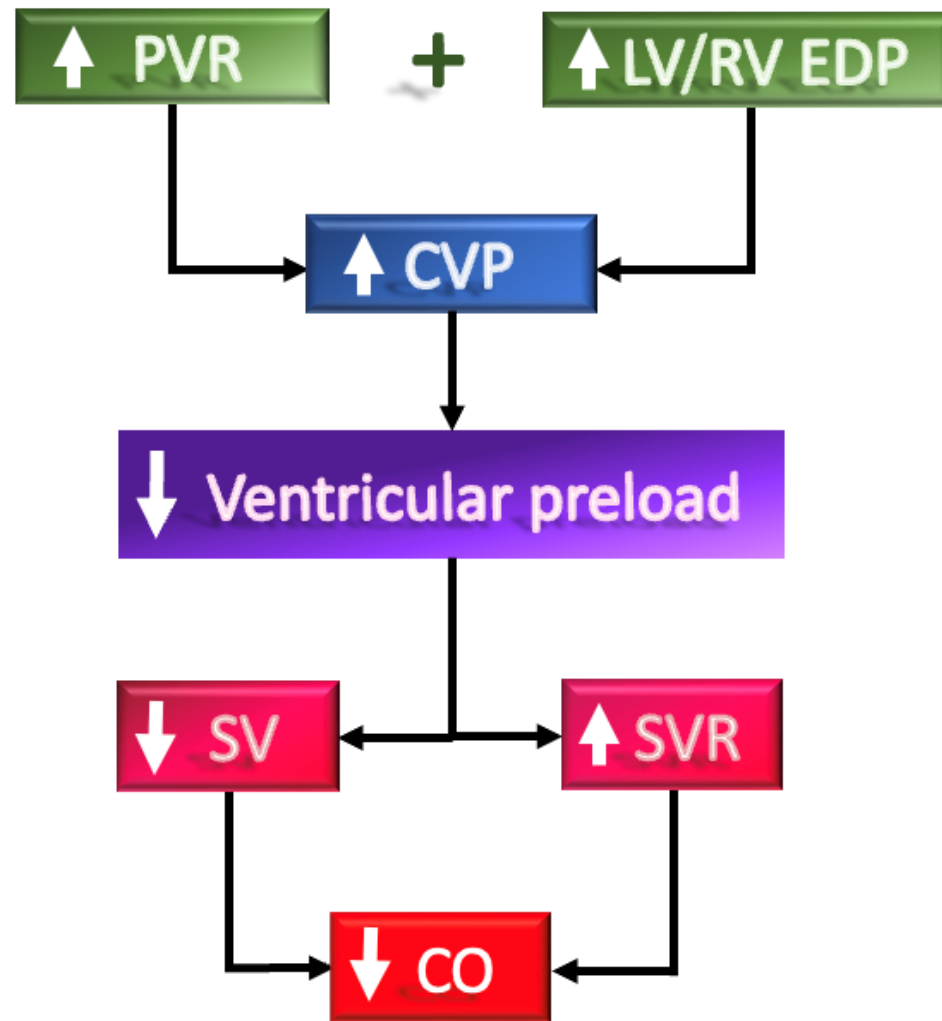
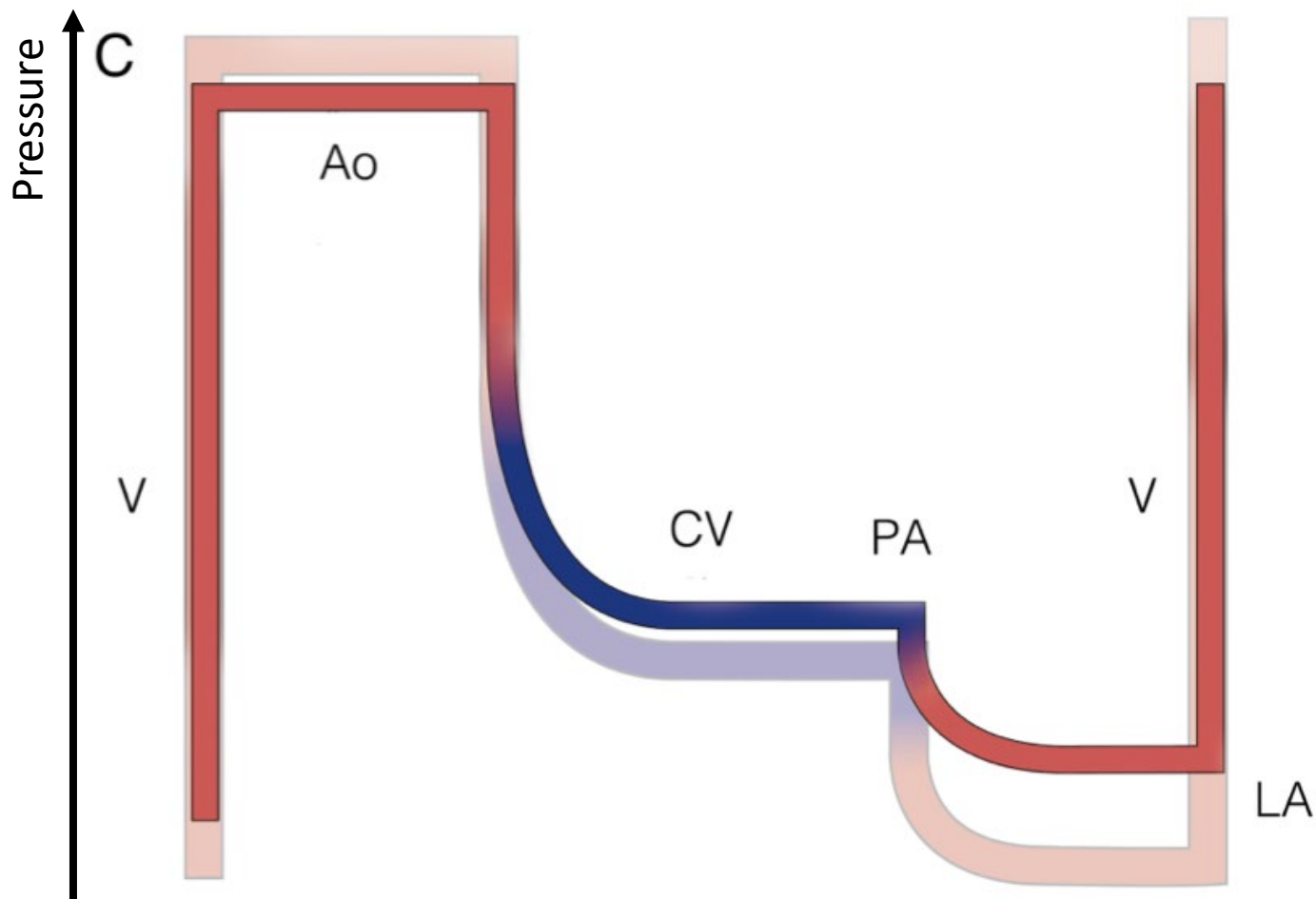


Survival after Fontan surgery

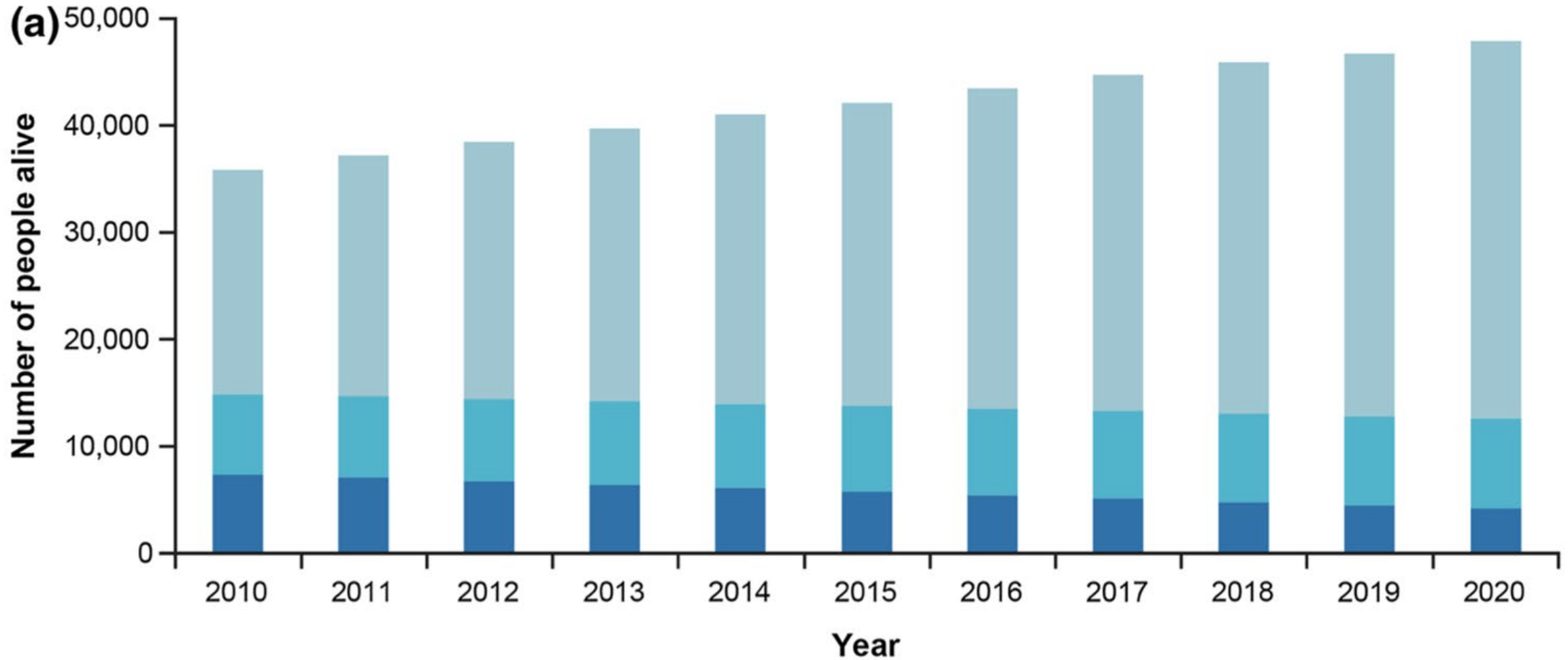


Natural history of Fontan circulation

Late Fontan circulation

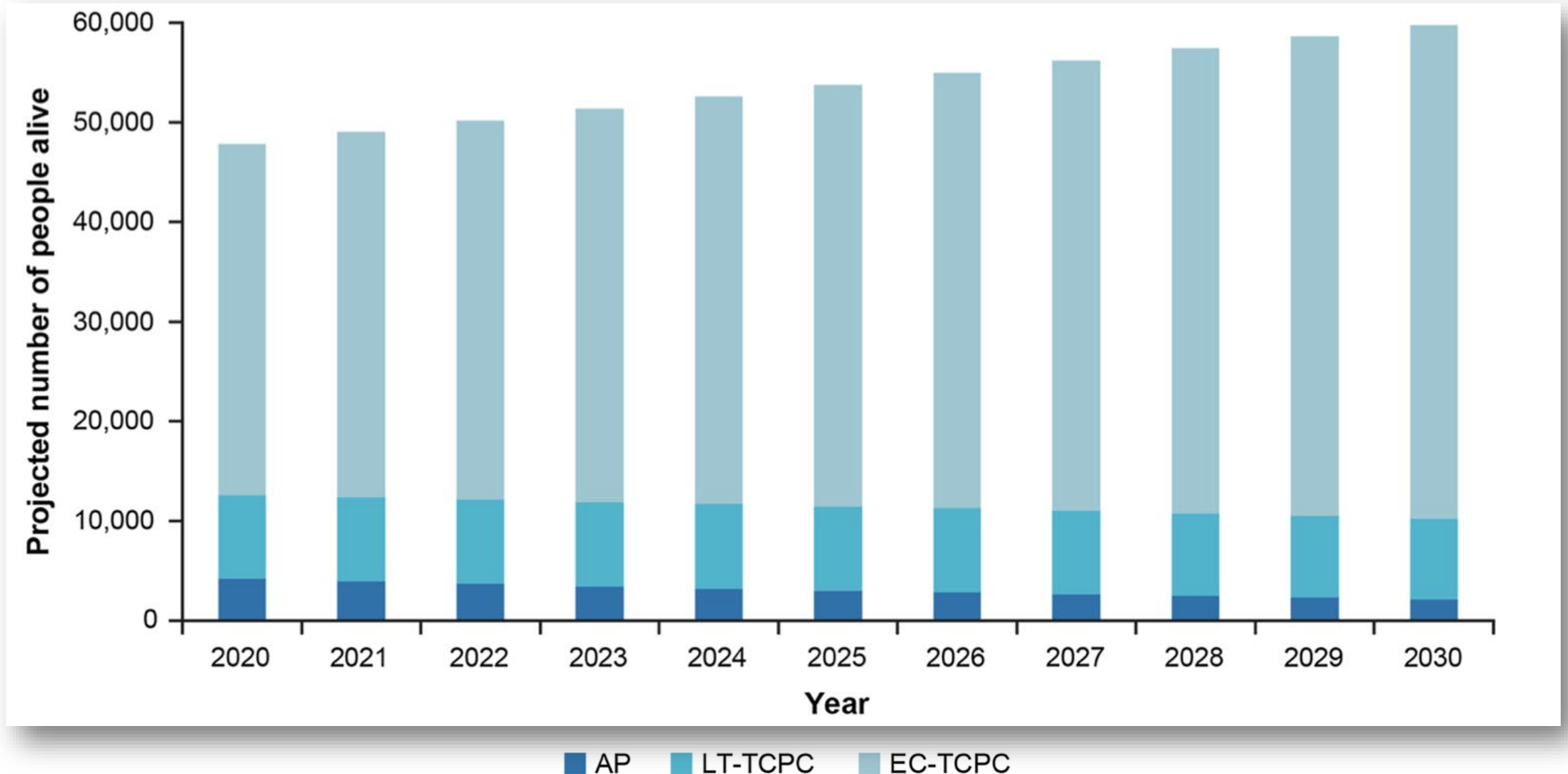


Persons living with Fontan

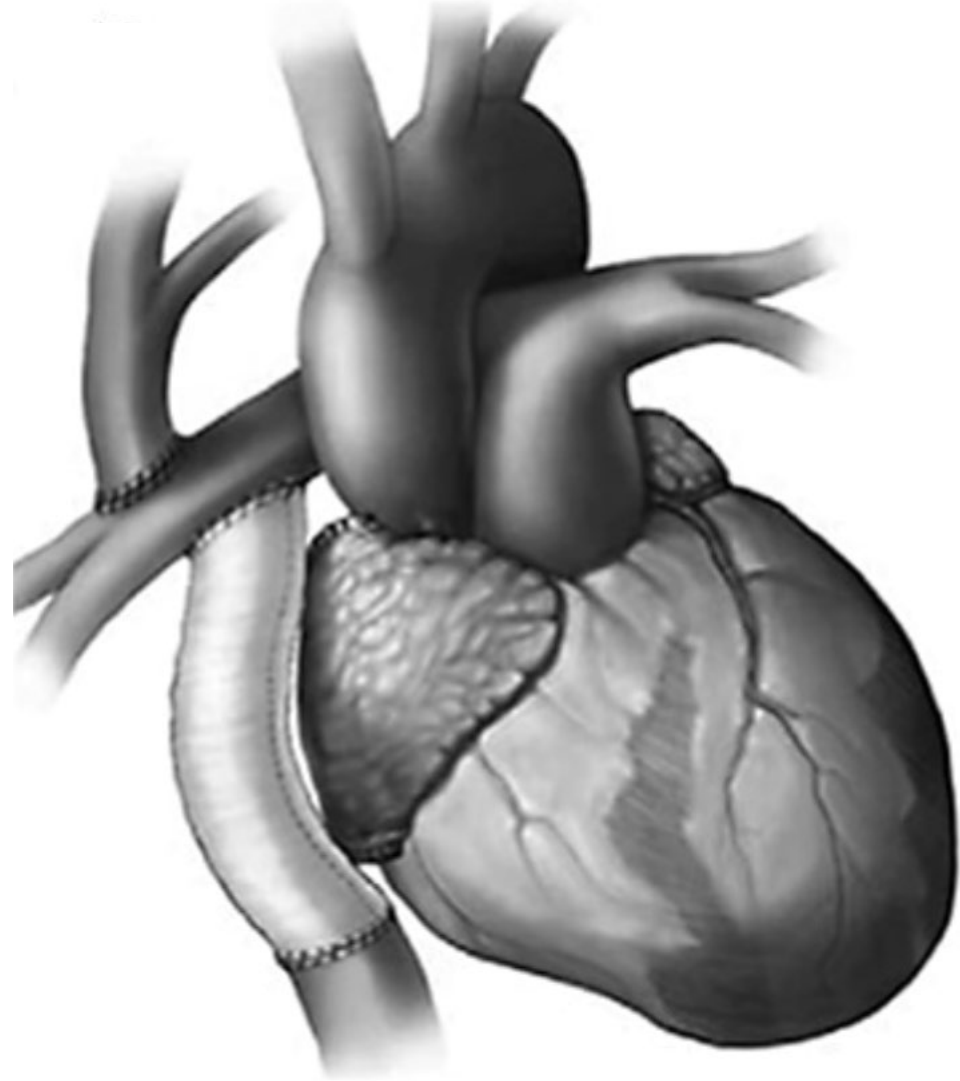


■ AP ■ LT-TCPC ■ EC-TCPC

Persons living with Fontan



Heart failure



Systolic dysfunction

Diastolic dysfunction

AV valve regurgitation

Fontan pressures/Clot

Arrhythmias

ECG (yearly)

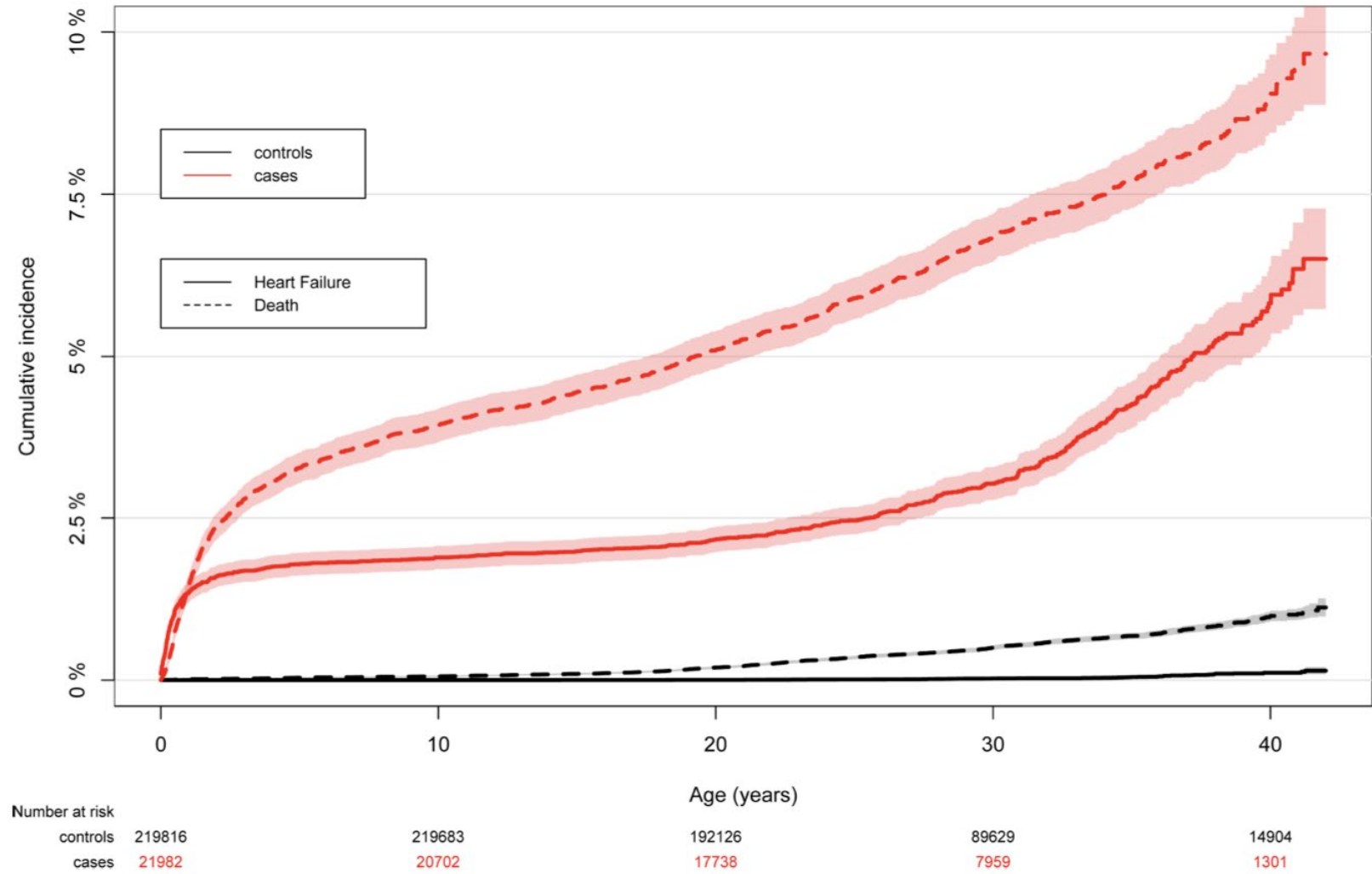
**Echocardiogram
(yearly)**

**Holter monitor
(2-3 years)**

**CMR/CT
(baseline/2-3 years)**

**Right heart
catheterization
(baseline /5-10 years)**

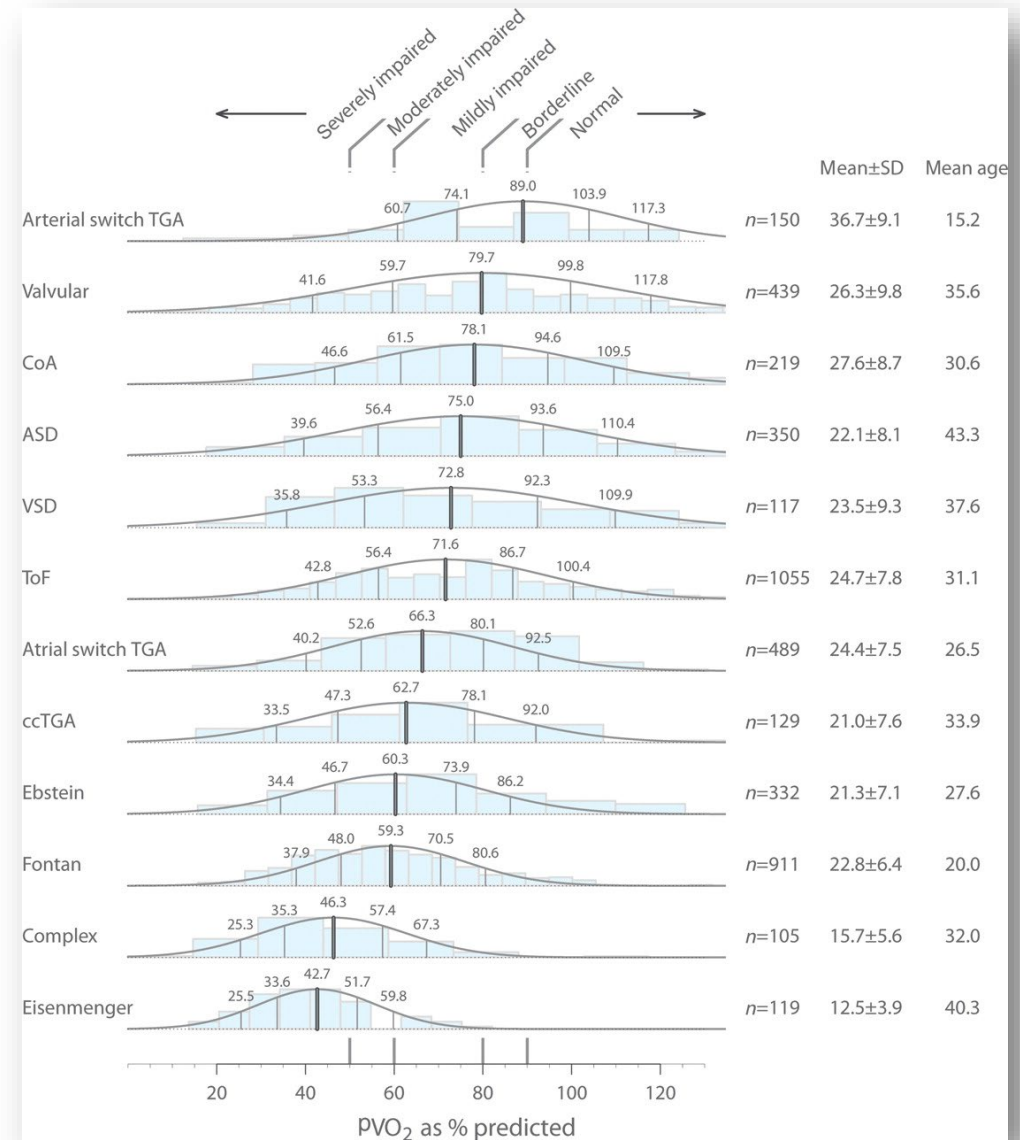
Heart failure



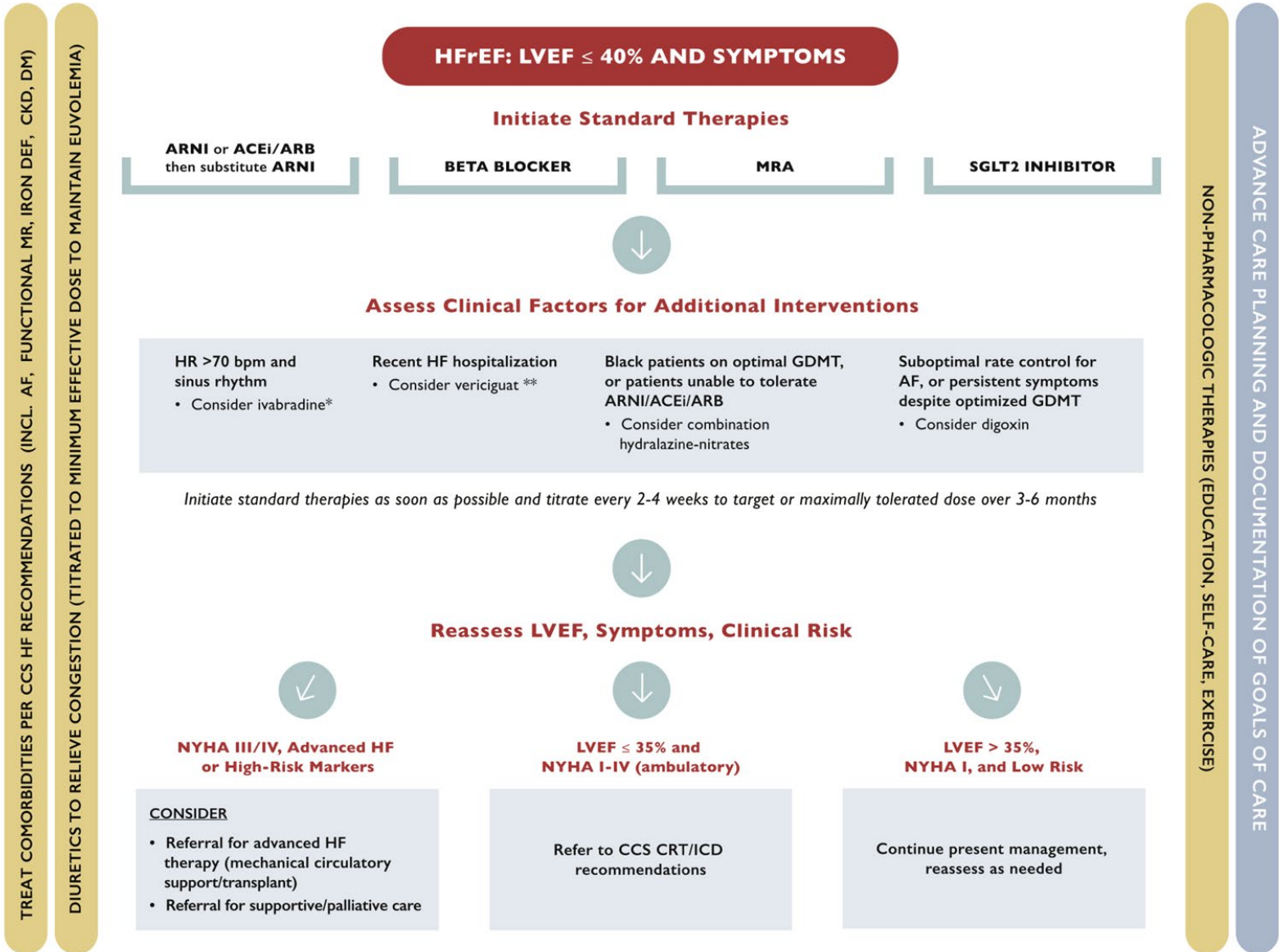
Failing Fontan circulation

Clinical features

- Progressive venous congestion
- Peripheral oedema
- Ascites
- Protein-losing enteropathy
- Plastic bronchitis
- Cyanosis
- Exercise intolerance



HF medication in acquired heart



A close-up of Darth Vader's helmet, centered in the frame. The helmet is dark and metallic, with its characteristic breathing apparatus. The background is a blurred city skyline at night, with numerous yellow and white lights from buildings and streetlights. The overall tone is dark and ominous.

**WELCOME TO THE DARK SIDE OF
SCIENCE**

DATA SCIENCE

HF medication in Fontan circulation



**OPINIONS ONLY
EXIST BECAUSE
OF LACK OF
DATA**

2 Pulmonary vascular disease



PVR elevated > 2 indexed Wood units
Endothelin receptor antagonists and PDE-5 inhibitors may be considered with elevated PVR in the absence of a high ventricular end-diastolic pressure (ESC Class IIb, Level C)

3 Systolic dysfunction

Single ventricle ejection fraction <50%
Beta-blockers, RAAS blockers extrapolated from biventricular heart failure



4 Diastolic dysfunction

End-diastolic pressure or pulmonary capillary wedge pressure ≥ 12 mmHg
Diuretics (careful monitoring to avoid decreased preload)

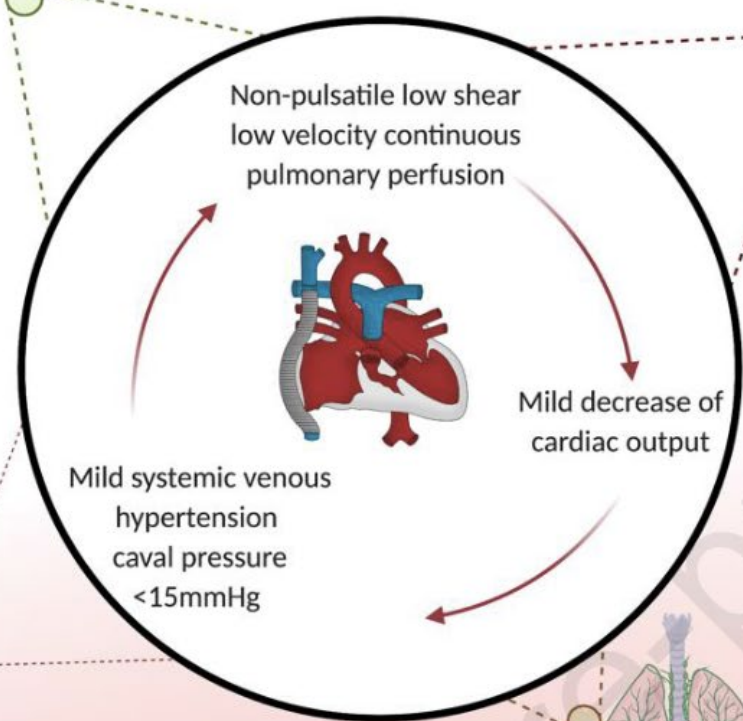
5 Atrial arrhythmia

Maintenance of sinus rhythm: rhythm control strategy
Beta-blockers
Caution with amiodarone, dofetilide and sotalol

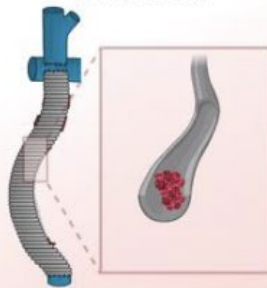
6 Atrio-ventricular valve regurgitation

Diuretics, RAAS blockers
Outflow tract obstruction
Beta-blockers in case of dynamic obstruction

7 Extra-cardiac organ lymphatic dysfunction and Fontan associated liver disease



1 Fontan pathway thrombosis



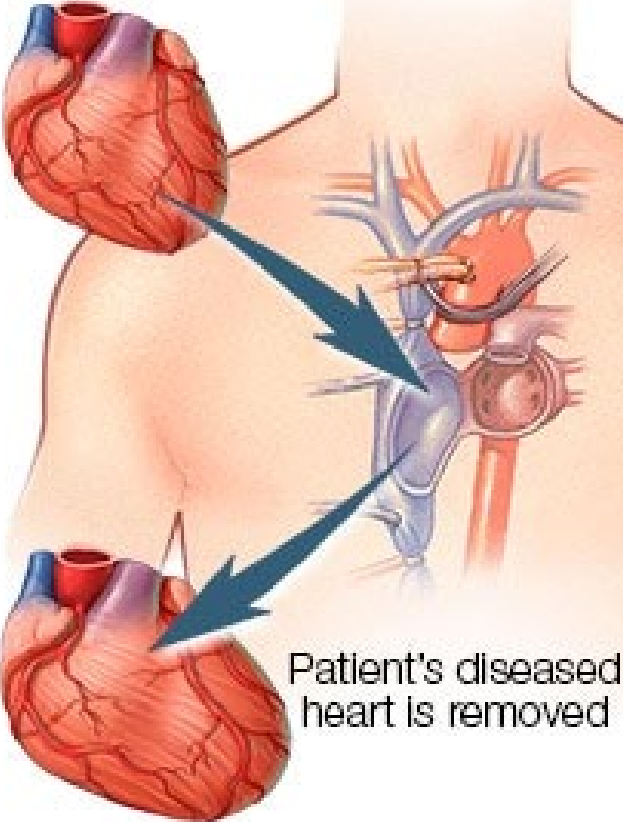
Anticoagulation with a vitamin K antagonist in presence or history of atrial thrombus, past thromboembolic event or atrial arrhythmia (AHA, ESC Class I Level C)
DOAC reporting safety and long term efficacy data pending

What's
next

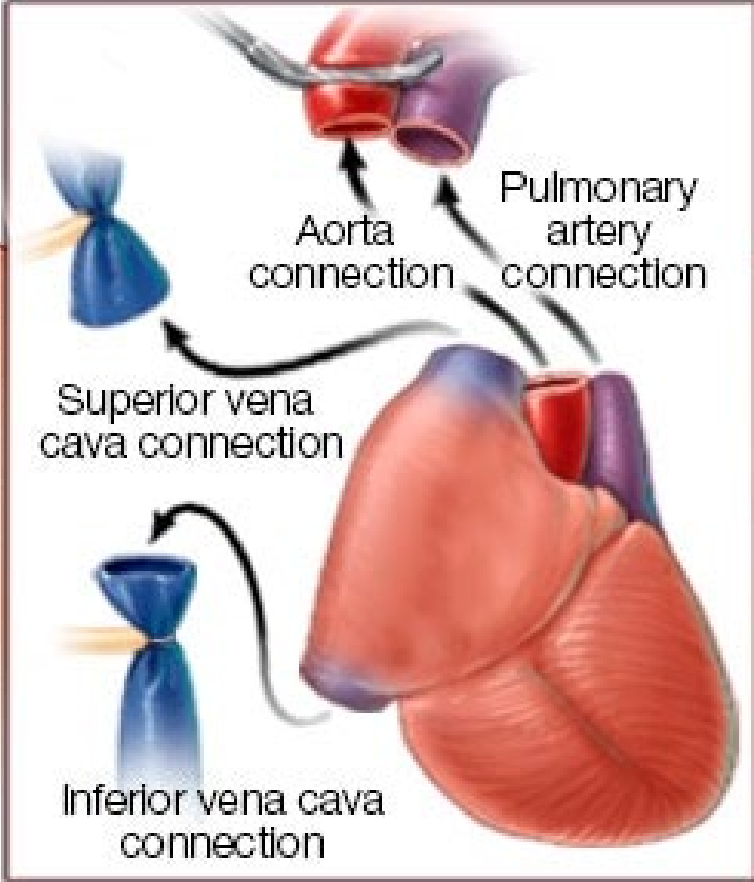


Heart transplant procedure

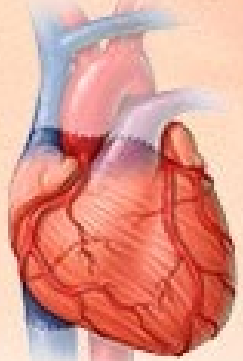
Donor heart



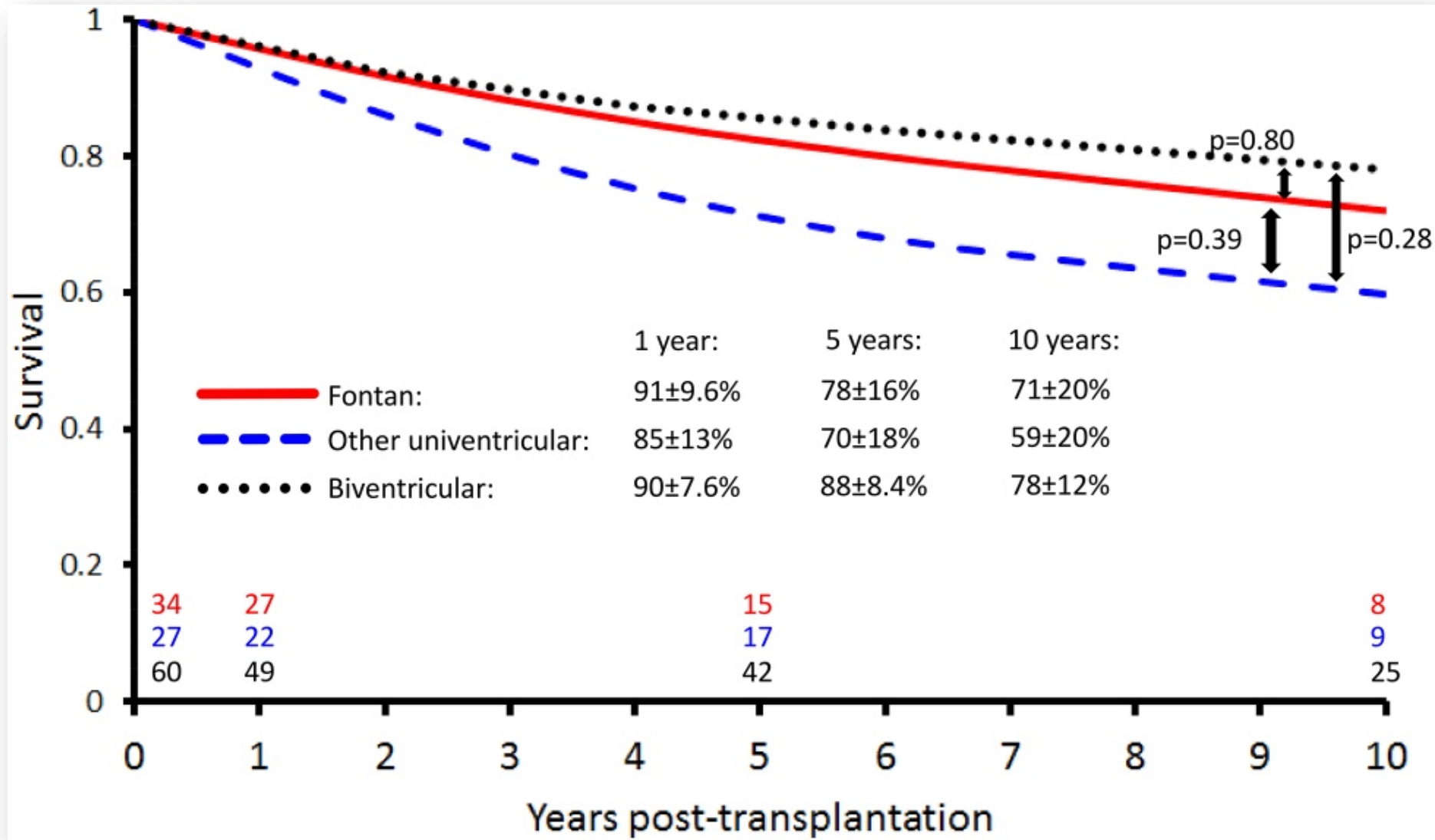
Patient's diseased heart is removed



Donor heart in place

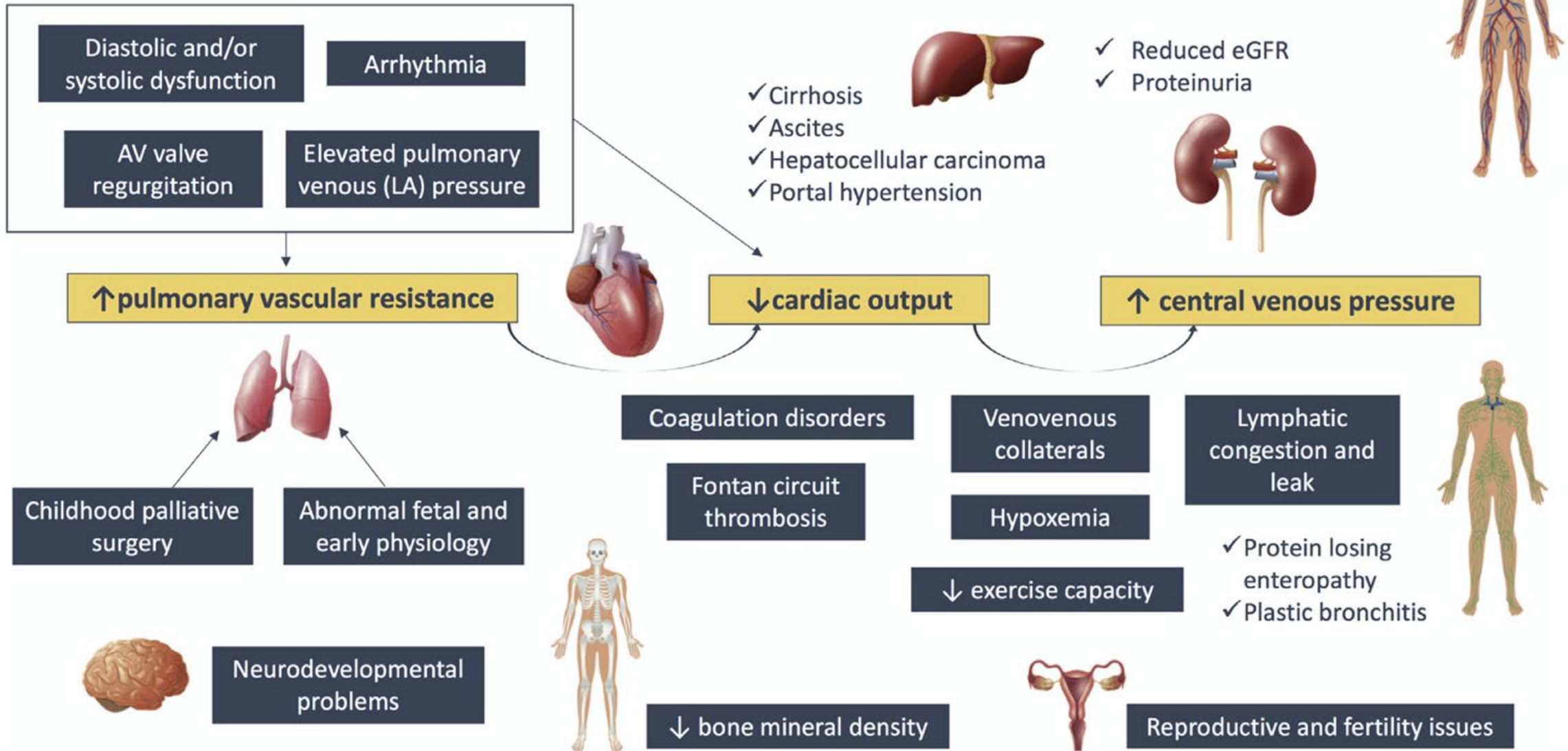


Transplant survival

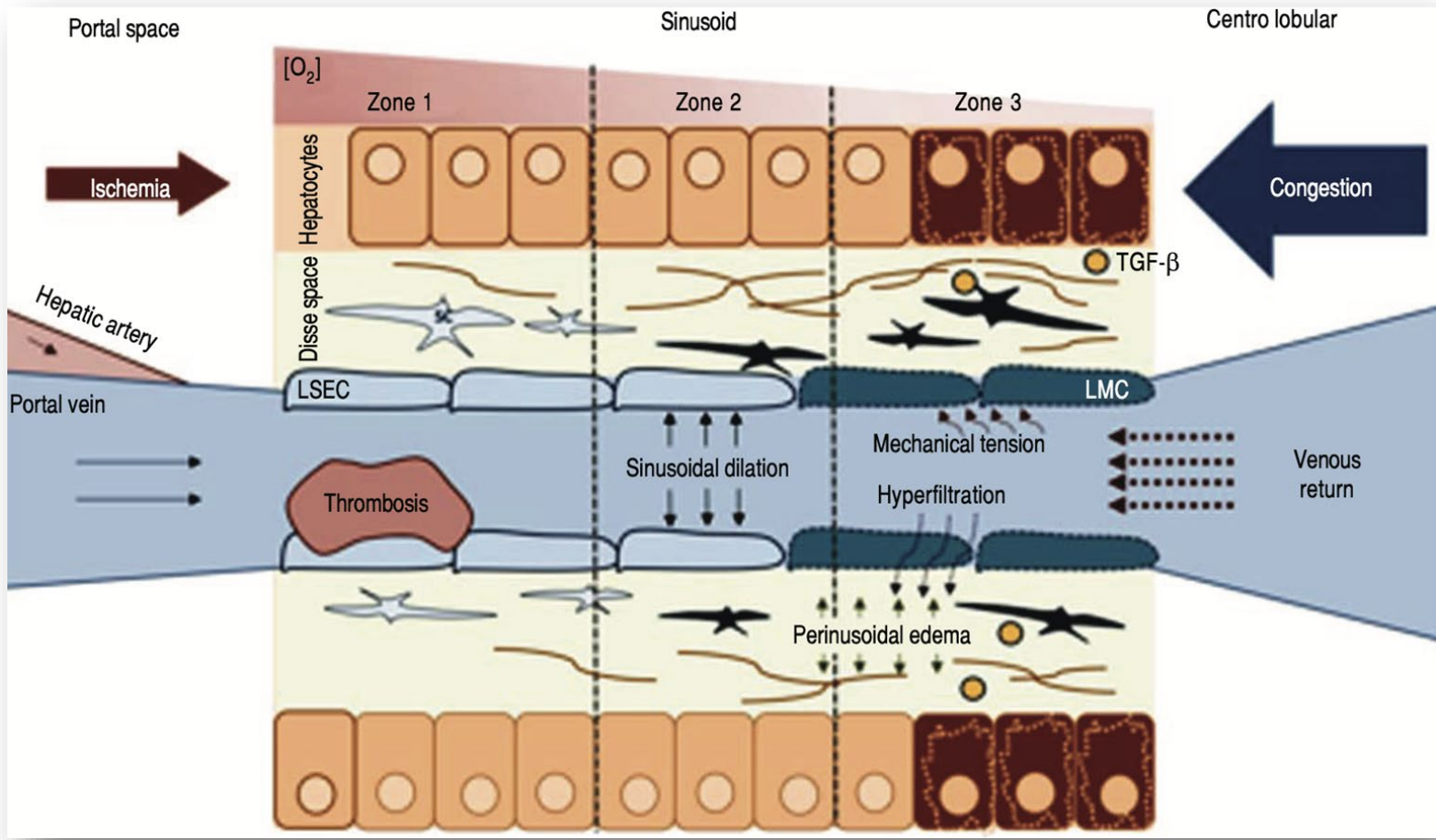
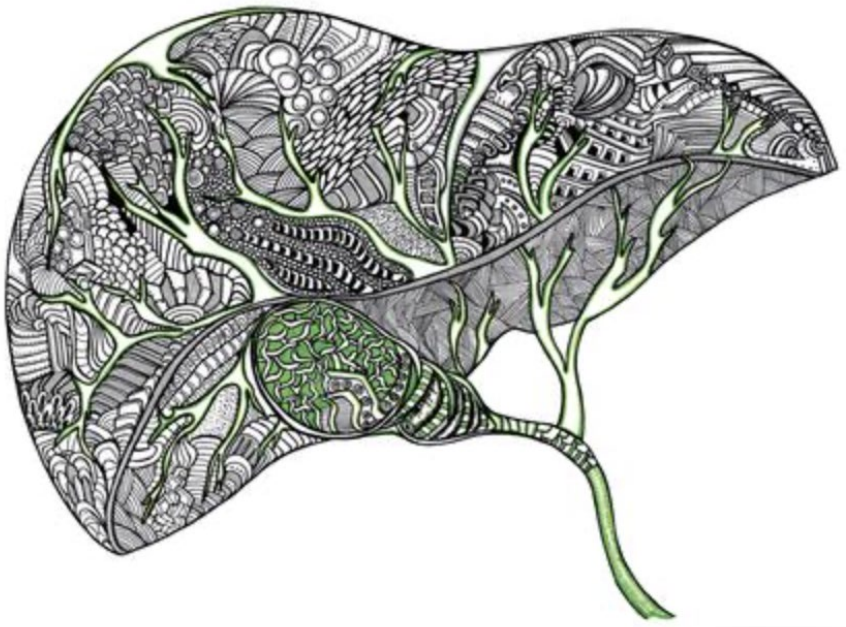




The interlinked and multisystem disturbances of a Fontan circulation



The liver



Hepatic dysfunction

Hemodynamic changes

Elevated CV pressure	Elevated hydrostatic pressure
Impaired lymphatic drainage	Perisinusoidal edema
Hypoxia	Increased matrix stiffness
Sinusoidal dilatation and mechanical stress	Congestion

Fontan surgery

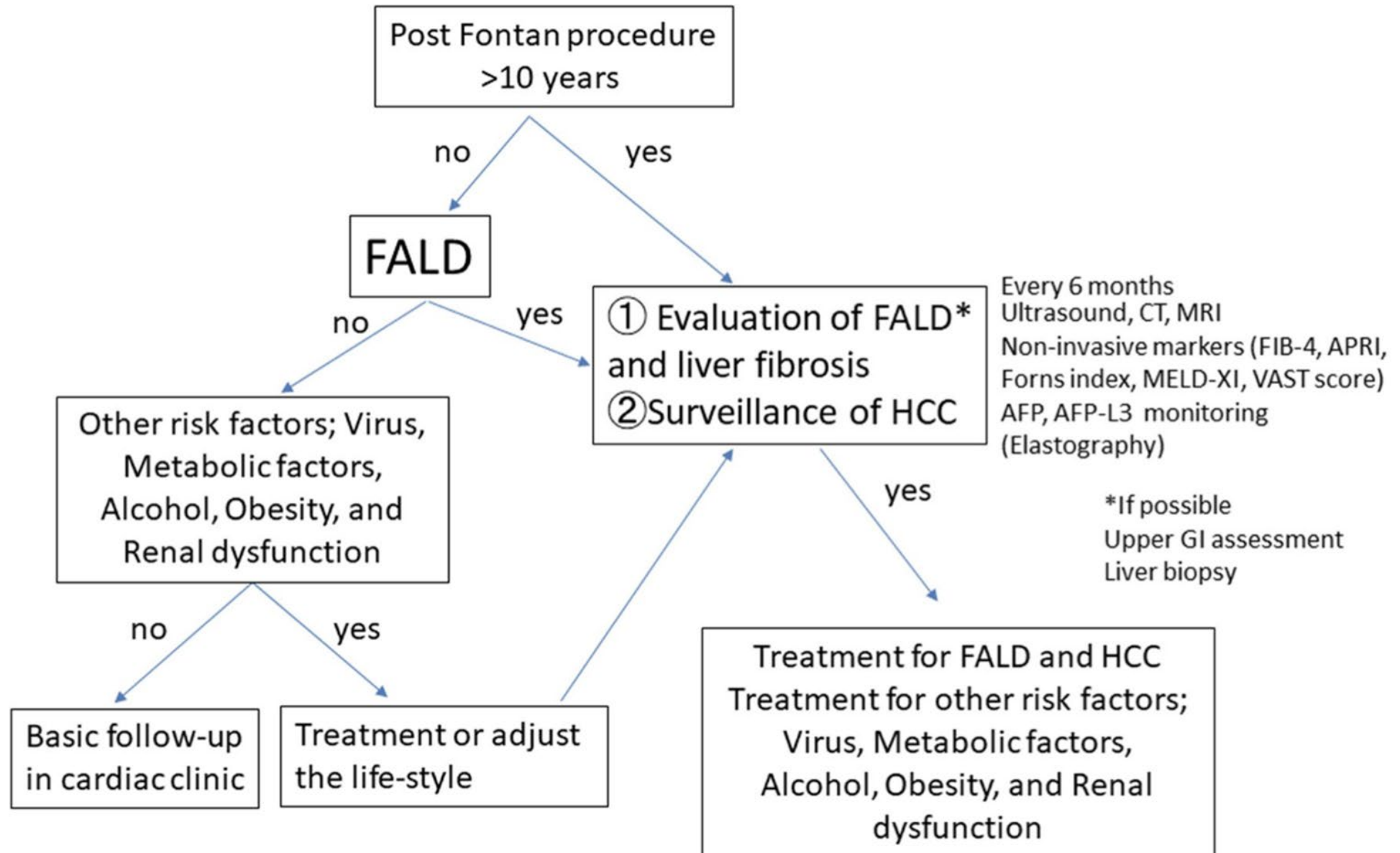


Complications of Fontan

Mental disorders (anxiety and behavioral disorders) and mental developmental delay
 Arrhythmia, thromboembolism, protein enteropathy, heart failure
 Pulmonary arteriovenous fistulae

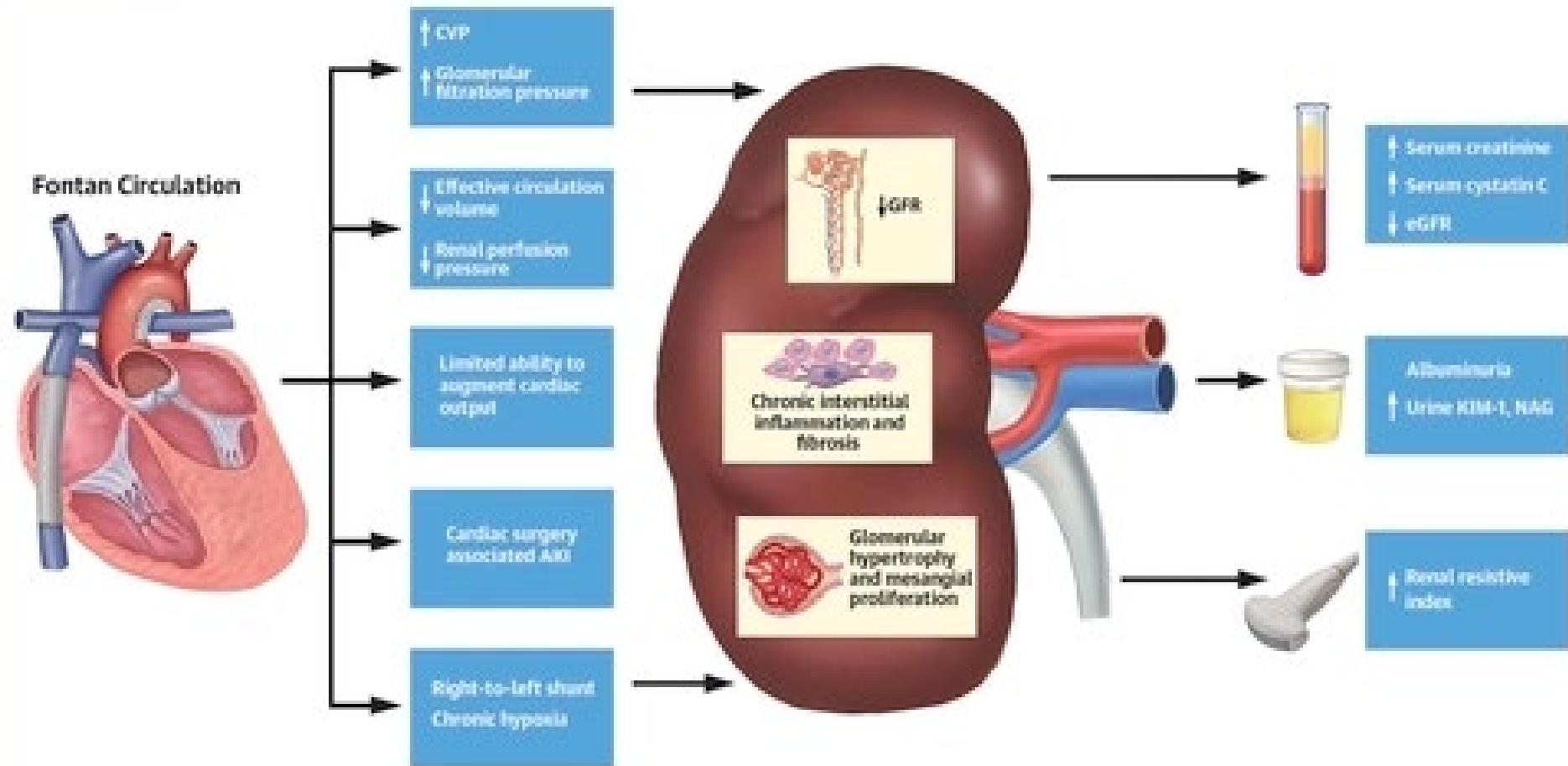
Virus	Metabolic factors
Alcohol	Obesity Renal dysfunction

FALD-HCC surveillance





CENTRAL ILLUSTRATION: Pathophysiology of Fontan-Associated Renal Disease



Zafar, F. et al. J Am Coll Cardiol. 2020;76(3):334-41.

The kidneys

LABORATORY TESTING

Direct parameters

Urea

Creatinine

Creatinine clearance (24h
urine)

Albumin

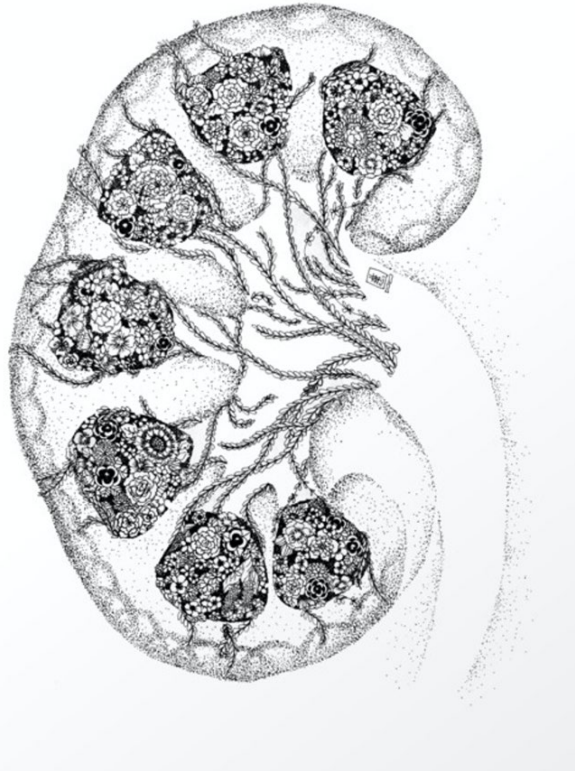
Cystatin C

Calculated parameters

eGFR

Urine albumin/Cr ratio (uACR)

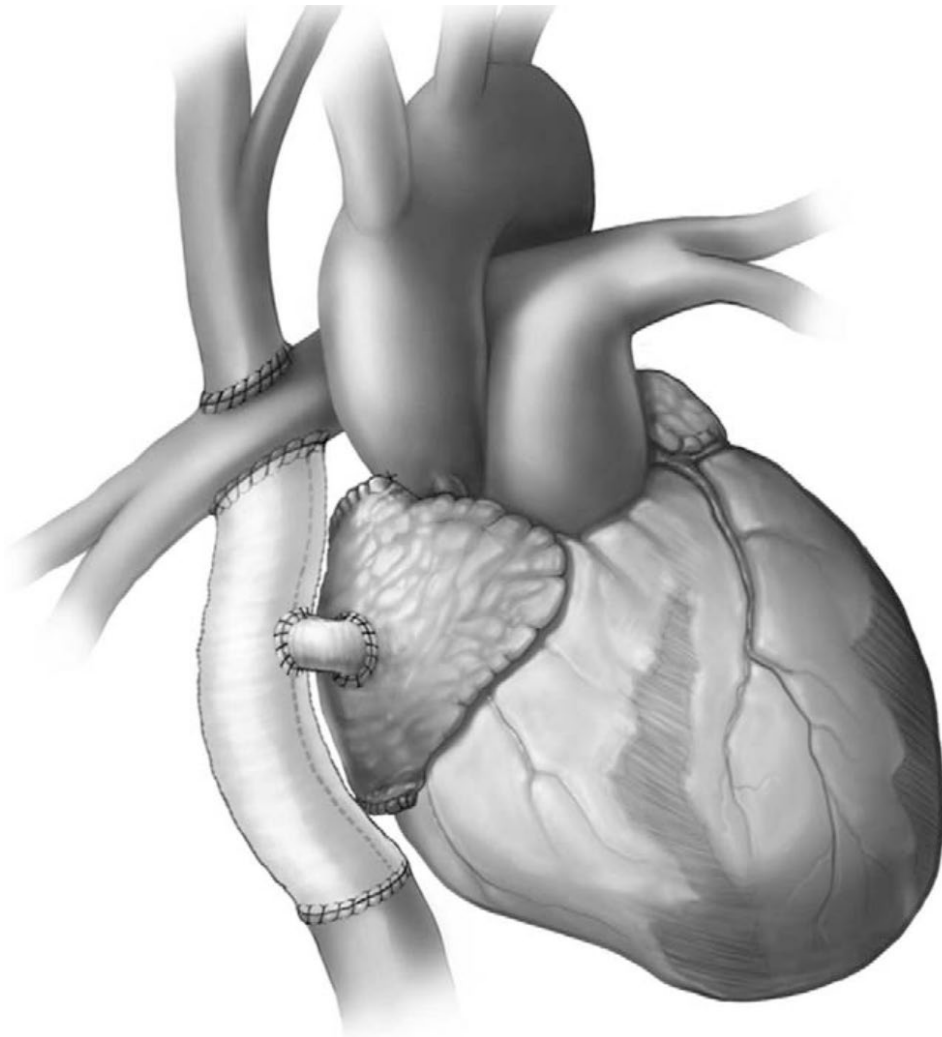
BUN



Impaired eGFR - 10-15% Fontan patients

Albuminuria - 30% Fontan patients

Fontan – multisystemic disease



LUNG

Restrictive lung disease
Pulmonary hypertension
Pulmonary hemorrhage
Plastic bronchitis

LIVER

Congestive hepatopathy
Cardiac cirrhosis
Fontan associated liver disease

IMMUNOLOGY/ INFECTIOUS DISEASE

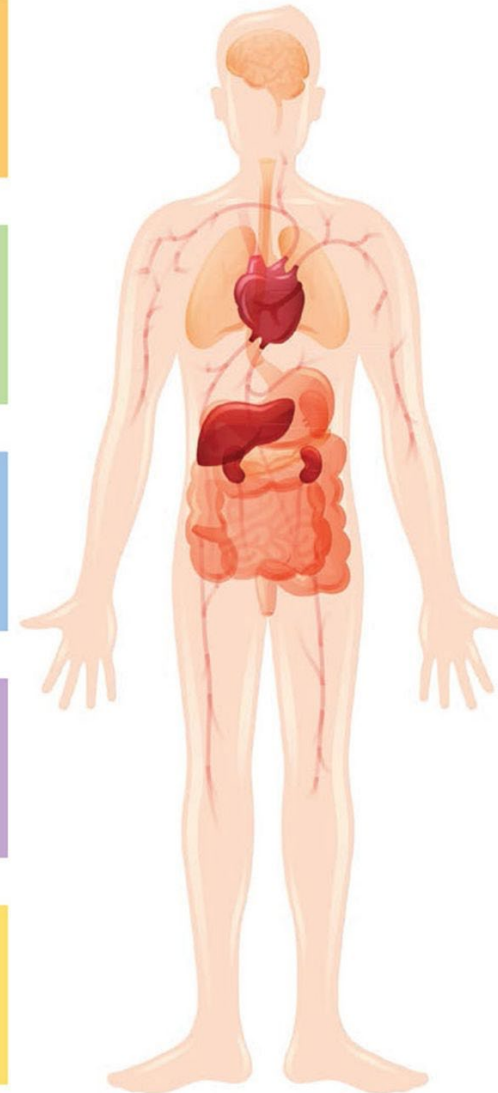
Protein-losing enteropathy
Infective endocarditis
Pneumonia
Brain abscess

HEMATOLOGY

Secondary erythrocytosis/Iron
deficiency/Hyperuricemia
(Cyanotic CHD)
Thromboembolism
Anemia

ONCOLOGY

Low-dose ionizing radiation
and malignancy
Hepatocellular carcinoma
Age-appropriate cancer
screening



PSYCHOSOCIAL

Depression
Anxiety
Neurodevelopment deficits

ENDOCRINE

Thyroid
Calcium hemostasis/Bone health
Obesity/Metabolic syndrome
Diabetes
Dyslipidemia

RENAL

Chronic kidney disease
Cardiorenal syndrome

VASCULAR

Cerebrovascular disease
Peripheral venous/arterial disease
Aortopathy
Endothelial dysfunction
Hypertension

15

Doctor's
Appointment

16

22

23

