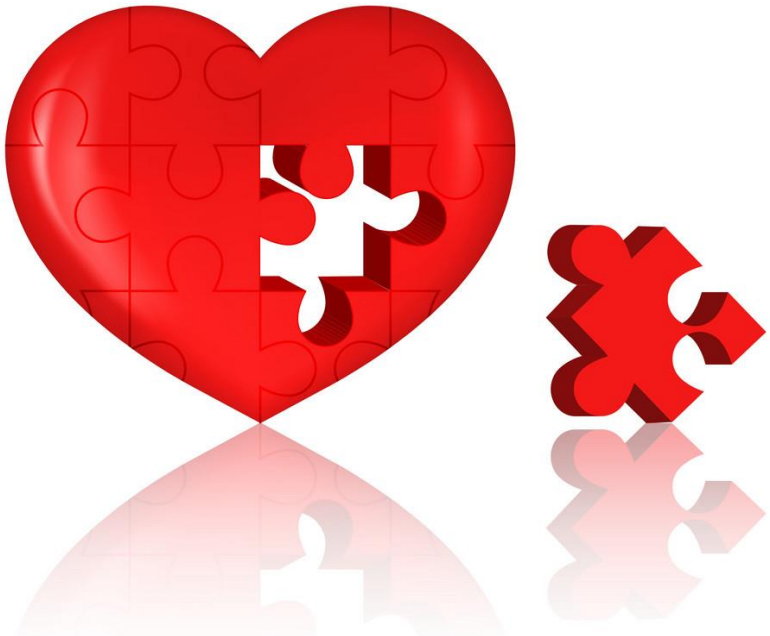
The Fontan in Pregnancy

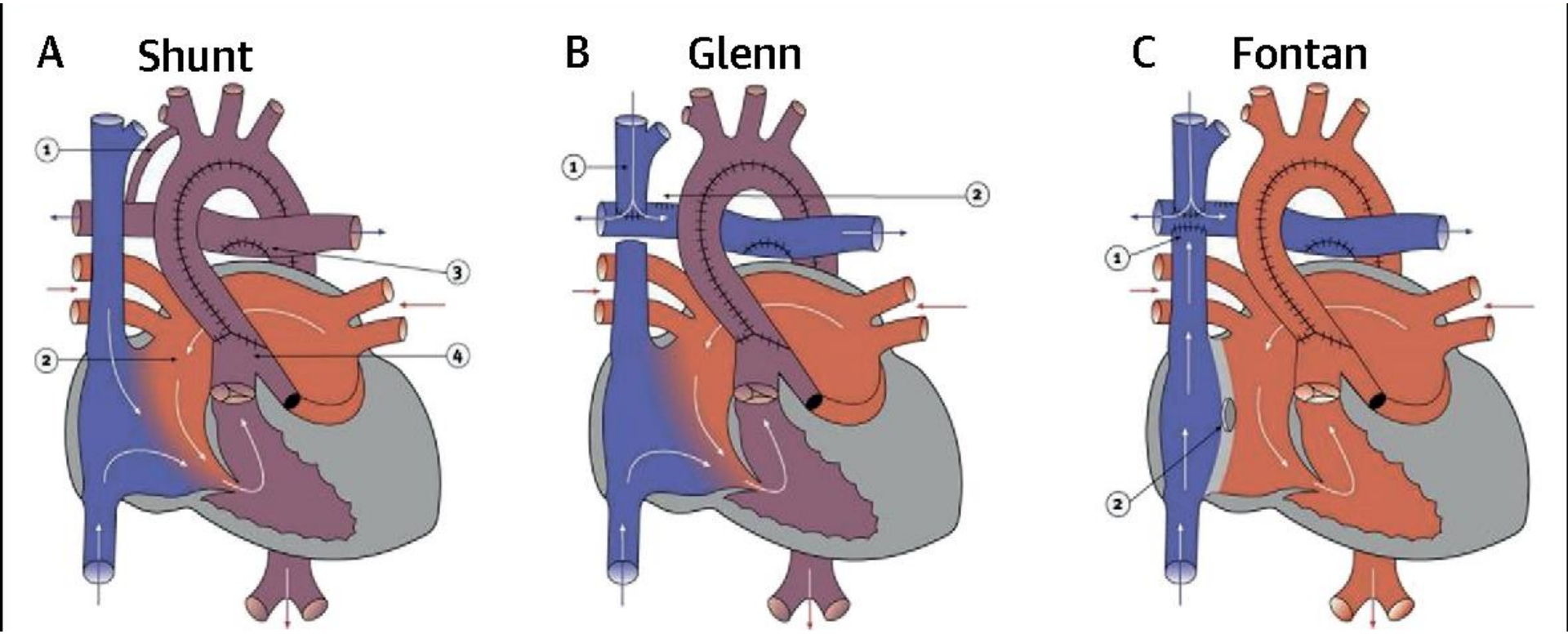
Dr Stephanie Curtis



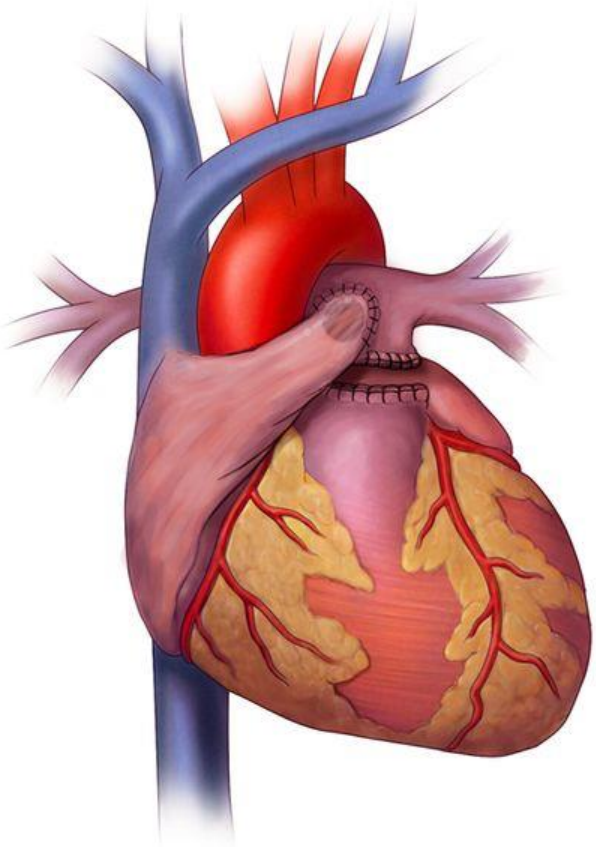
University Hospitals Bristol NHS
NHS Foundation Trust

ACHD Nurse Study Day October 2019

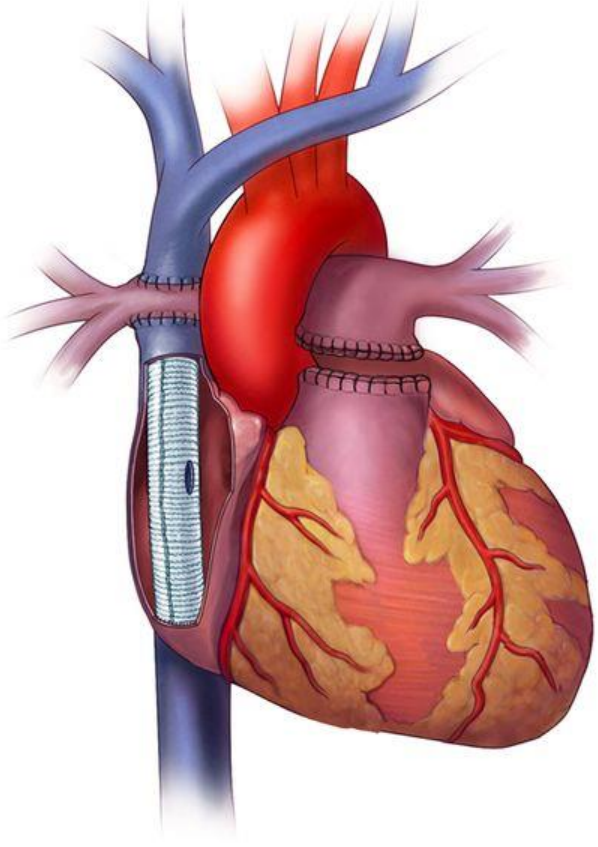
What is the Fontan?



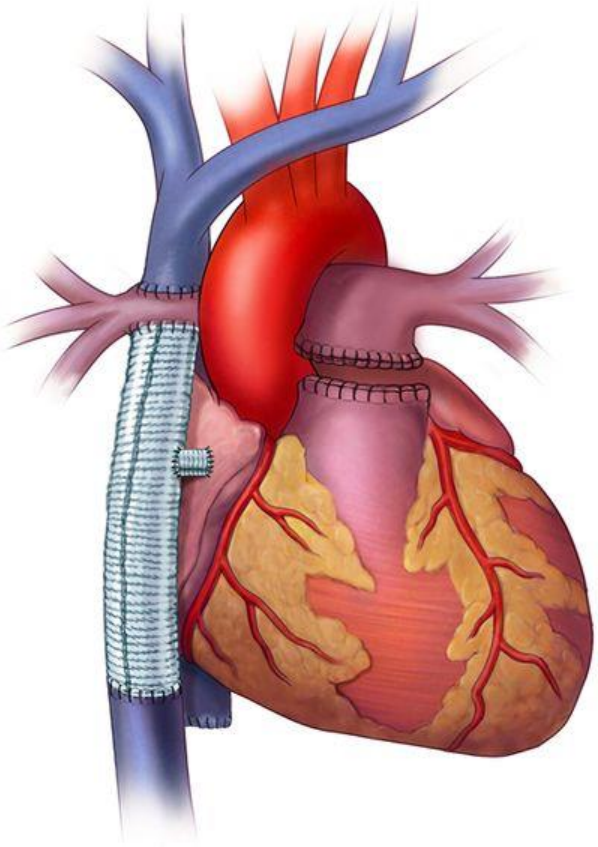
AP Fontan

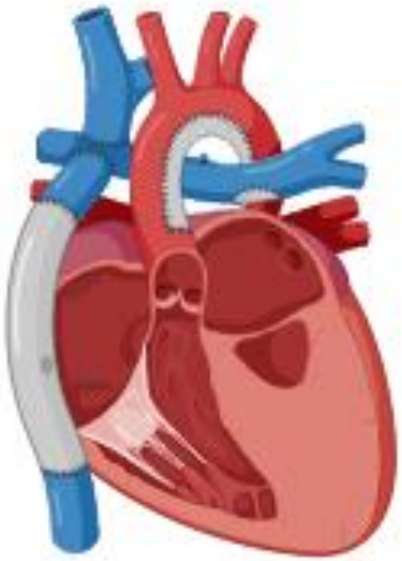


Lateral tunnel

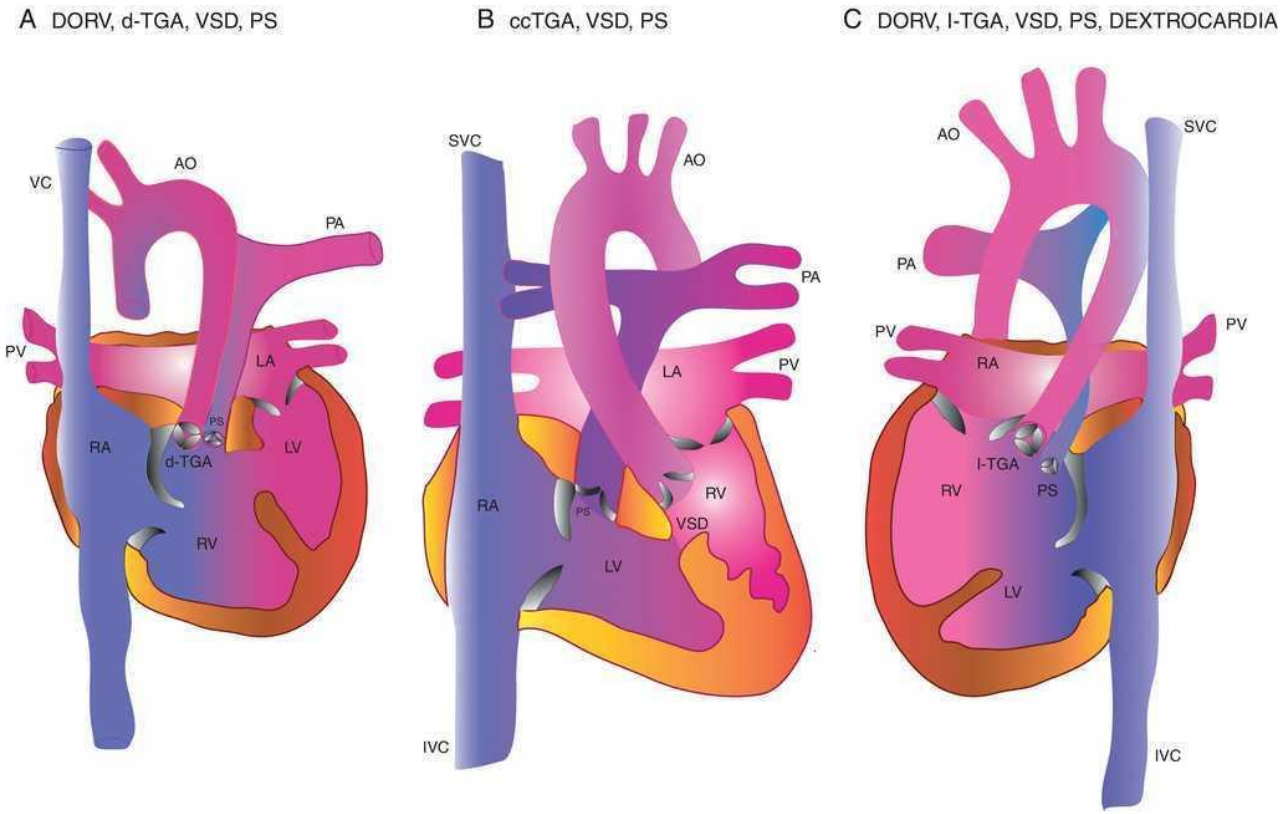


TCPC



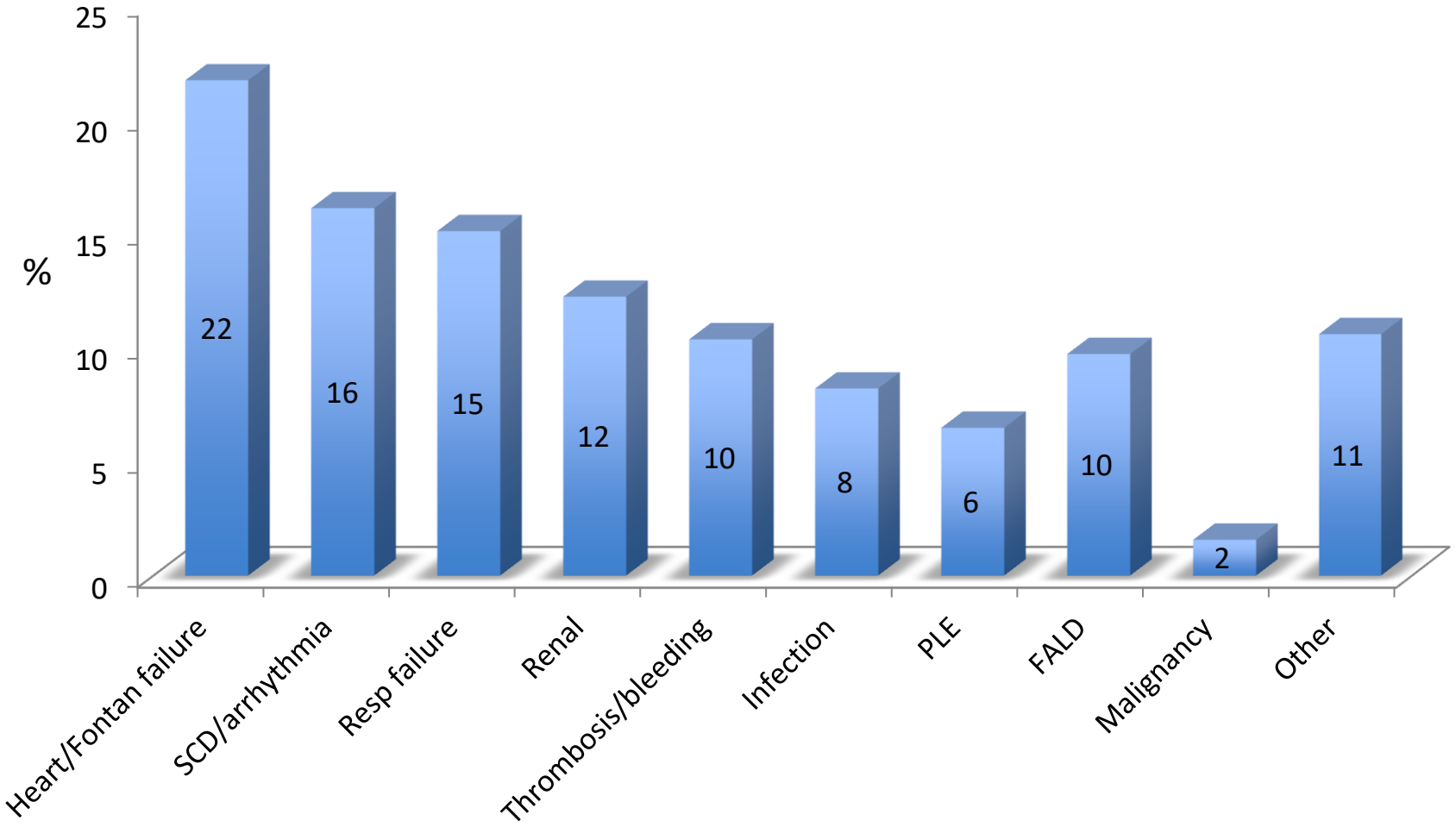


All Fontans are the same…ish

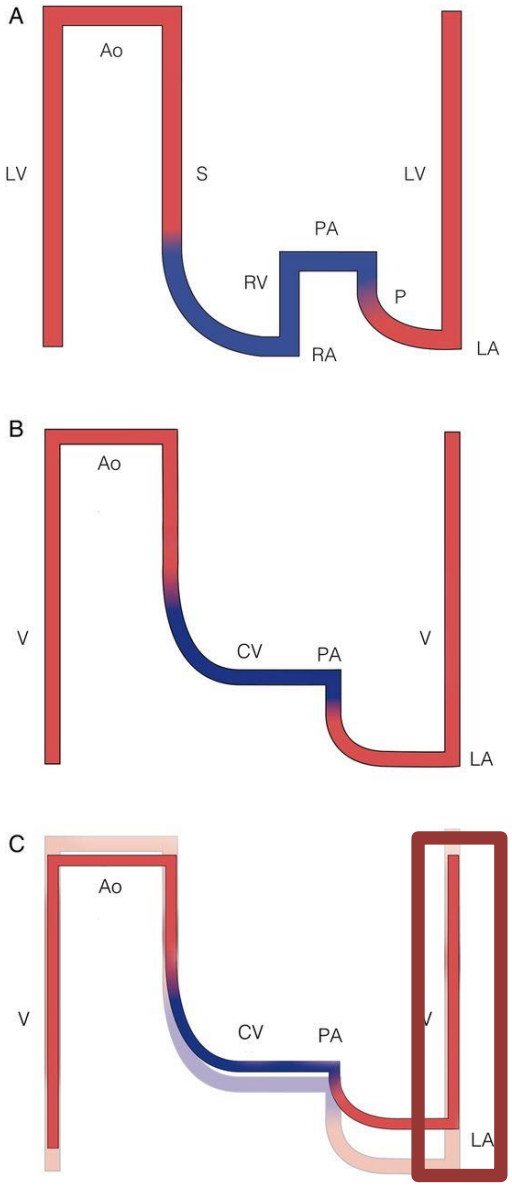


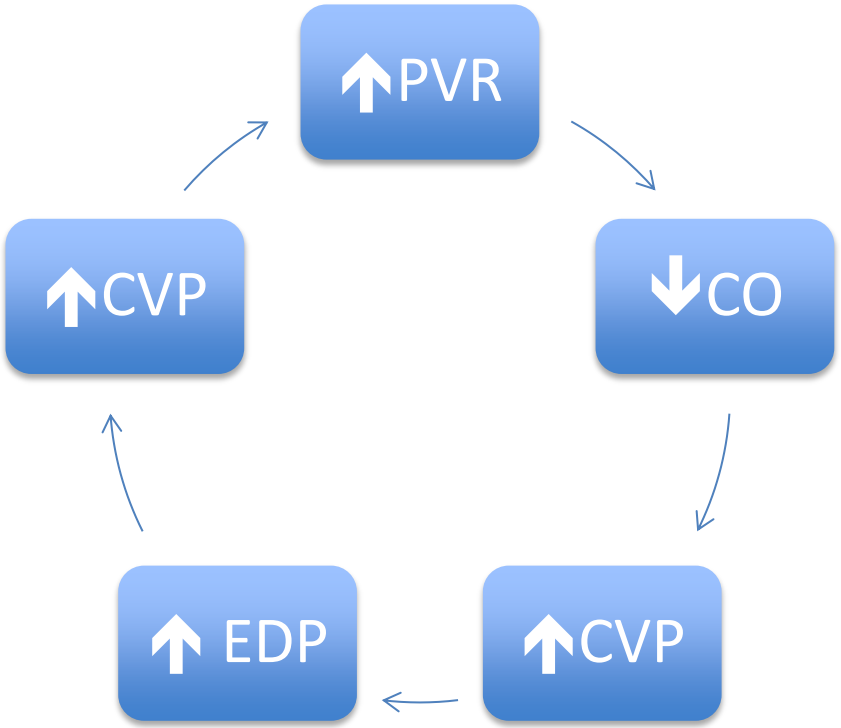
Rev Argent Anestesiol 2017;75:33-7

Fontan failure is the commonest cause of death



Alsaied …Veldtman et al…Heart 2017





CVP **has** to be high

Heart

Marc Gewillig, and Stephen C Brown Heart 2016;102:1081-1086

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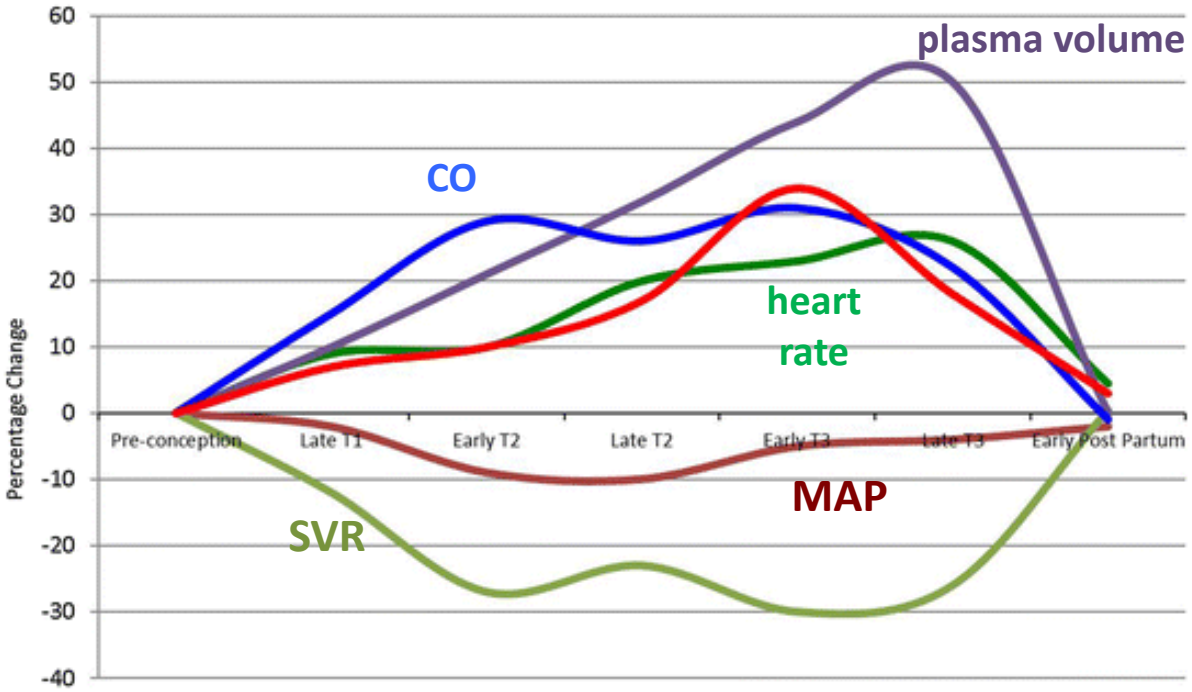
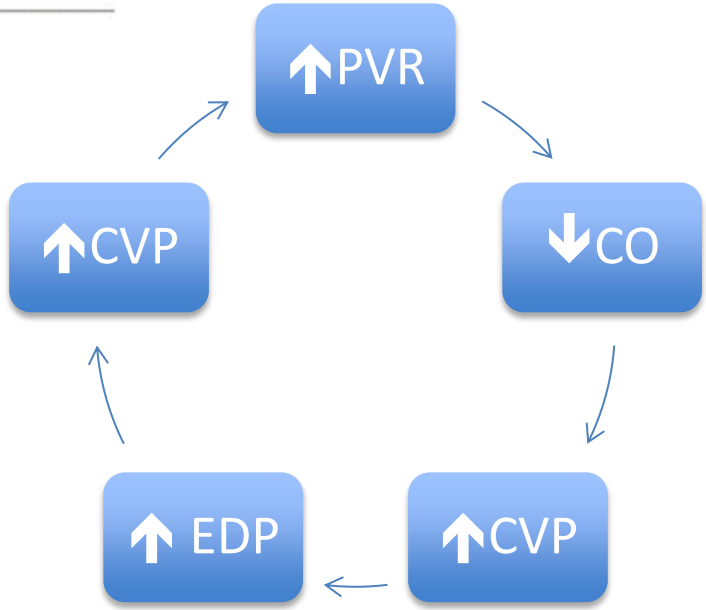
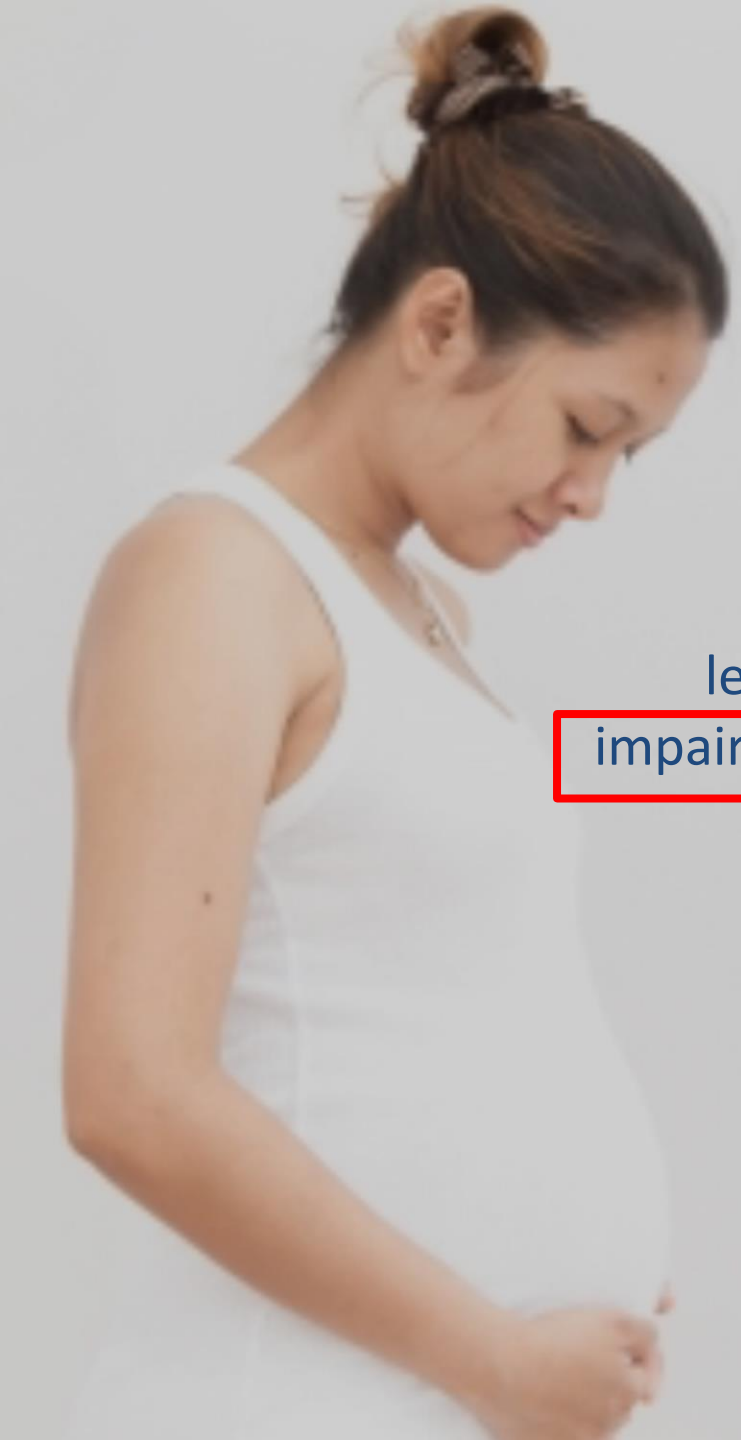




Ashrafi & Curtis. Cardiology and Therapy 2017

pregnancy is a
hypercoagulable
state



predictors of maternal events in pregnancy

NYHA Class> II
cyanosis
previous cardiac event

left heart obstruction (mitral/aortic stenosis)
impaired systemic ventricular function (EF < 40%)

severe pulmonary regurgitation and poor RV
mechanical valve
dilated aorta

Drenthen W et al, ZAHARA. Eur Heart J. 2010 Sep;31(17):2124-32
Siu SC, Circulation. 2001 Jul 31;104(5):515-21
Khairy P et al. Circ 2006;113(4):517-24

Definitions 1/3:
Predictors of cardiovascular events

CARPREG	Points
• Prior arrhythmias or cardiac event	1
• NYHA functional class > II or cyanosis	1
• Left heart obstruction	1
• Systemic ventricular dysfunction (EF < 40%)	1

Siu 2001

Total points	Risk %
0	5
1	27
>1	75

ZAHARA I	Points
• Prior arrhythmias	1.5
• NYHA functional class > II	0.75
• Left heart obstruction	2.5
• Cardiac medication before pregnancy	1.5
• Systemic AV valve regurgitation	0.75
• Pulmonary AV valve regurgitation	0.75
• Mechanical valve prosthesis	4.5
• Cyanotic heart disease	1.0

Drenthen 2010

Total points	Risk %
0	2.9
0.5 - 1.5	7.5
1.51 - 2.50	17.5
2.51 - 3.50	43.1
> 3.51	70.0

Khairy et al.

- Severe pulmonary regurgitation or subpulmonary ventricular dysfunction
- Smoking history

Khairy 2006

predictors of fetal events in pregnancy

NYHA Class> II
cyanosis
left heart obstruction (mitral/aortic stenosis)
smoking
anticoagulants

severe pulmonary regurgitation and poor RV
mechanical valve
dilated aorta

Drenthen W et al, ZAHARA Investigators. Eur Heart J. 2010 Sep;31(17):2124-32.
Siu SC, Circulation. 2001 Jul 31;104(5):515-21
Khairy P et al. Circ 2006;113(4):517-24

Table 3 **Modified World Health Organization classification of maternal cardiovascular risk**

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Diagnosis (if otherwise well and uncomplicated)	Small or mild – pulmonary stenosis – patent ductus arteriosus – mitral valve prolapse Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage) Atrial or ventricular ectopic beats, isolated	Unoperated atrial or ventricular septal defect Repaired tetralogy of Fallot Most arrhythmias (supraventricular arrhythmias) Turner syndrome without aortic dilatation	Mild left ventricular impairment (EF >45%) Hypertrophic cardiomyopathy Native or tissue valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis) Marfan or other HTAD syndrome without aortic dilatation Aorta <45 mm in bicuspid aortic valve pathology Repaired coarctation Atrioventricular septal defect	Moderate left ventricular impairment (EF 30–45%) Previous peripartum cardiomyopathy without any residual left ventricular impairment Mechanical valve Systemic right ventricle with good or mildly decreased ventricular function Fontan circulation. If otherwise the patient is well and the cardiac condition uncomplicated Unrepaired cyanotic heart disease Other complex heart disease Moderate mitral stenosis Severe asymptomatic aortic stenosis Moderate aortic dilatation (40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in bicuspid aortic valve, Turner syndrome ASI 20–25 mm/m^2, tetralogy of Fallot <50 mm) Ventricular tachycardia	Pulmonary arterial hypertension Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV) Previous peripartum cardiomyopathy with any residual left ventricular impairment Severe mitral stenosis Severe symptomatic aortic stenosis Systemic right ventricle with moderate or severely decreased ventricular function Severe aortic dilatation (>45 mm in Marfan syndrome or other HTAD, >50 mm in bicuspid aortic valve, Turner syndrome ASI >25 mm/m^2, tetralogy of Fallot >50 mm) Vascular Ehlers–Danlos Severe (re)coarctation Fontan with any complication
Risk	No detectable increased risk of maternal mortality and no/mild increased risk in morbidity	Small increased risk of maternal mortality or moderate increase in morbidity	Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity	Significantly increased risk of maternal mortality or severe morbidity	Extremely high risk of maternal mortality or severe morbidity

what might happen?

low CO

poor fetal outcome

IUGR, prematurity, death, miscarriage, SGA

excessive cardiac demand

heart failure

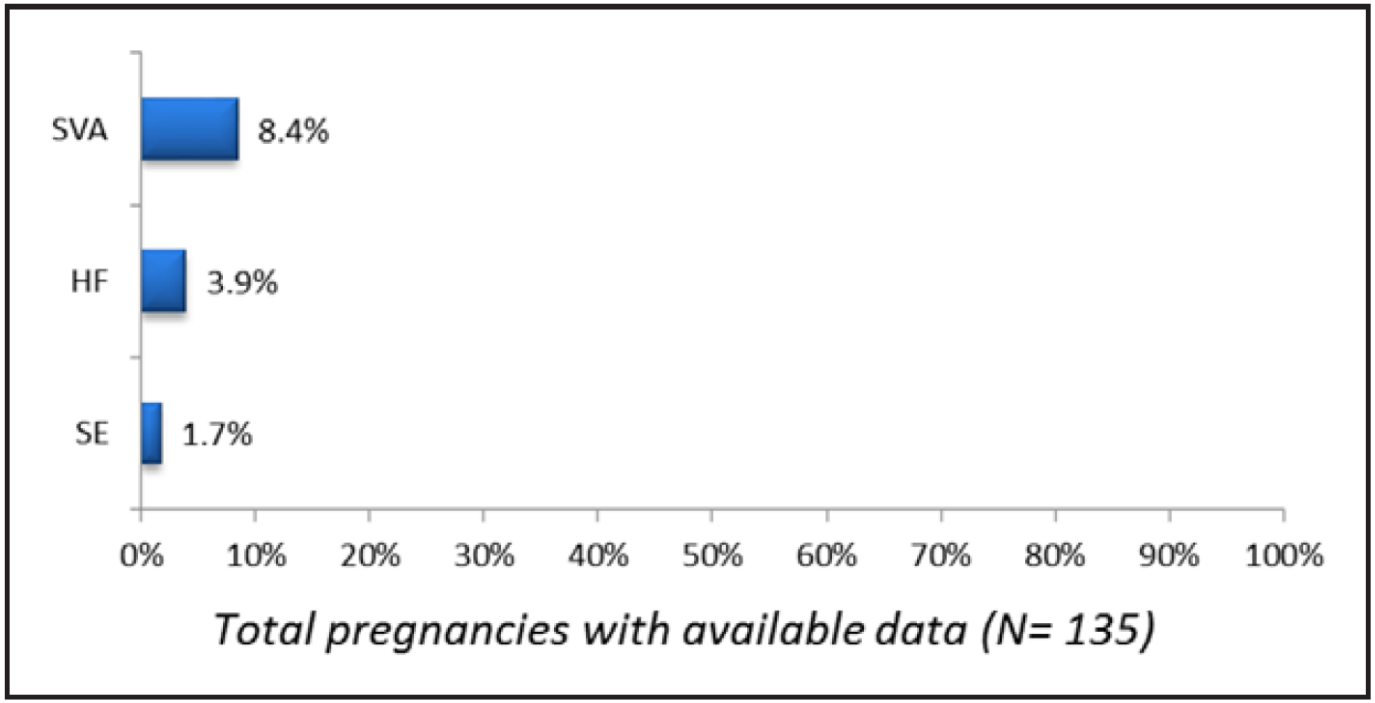
electrical irritability

arrhythmia

hypercoagulability

clotting/bleeding

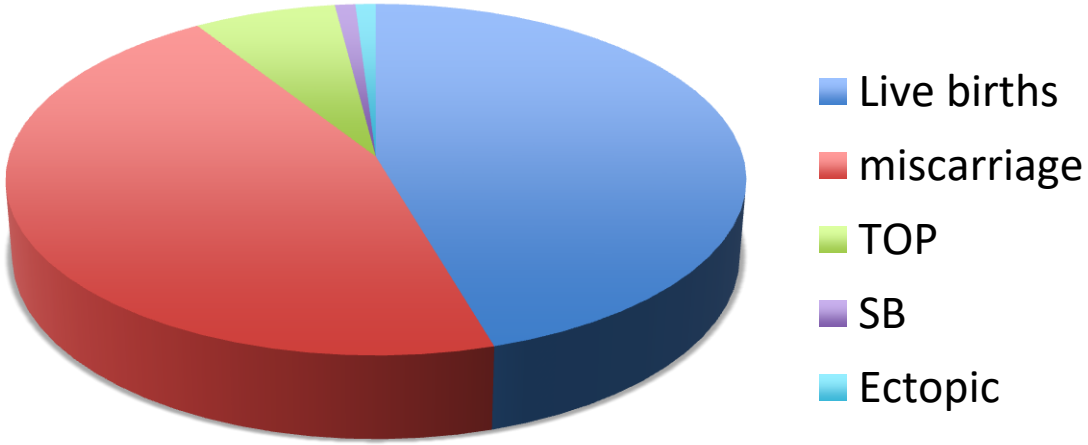
serious cardiac complications are uncommon…



255 pregnancies, 133 women, 1986-2015

Garcia Ropero 2018 Circulation: Cardiovascular Quality and Outcomes

…but babies do badly



55% fetal loss

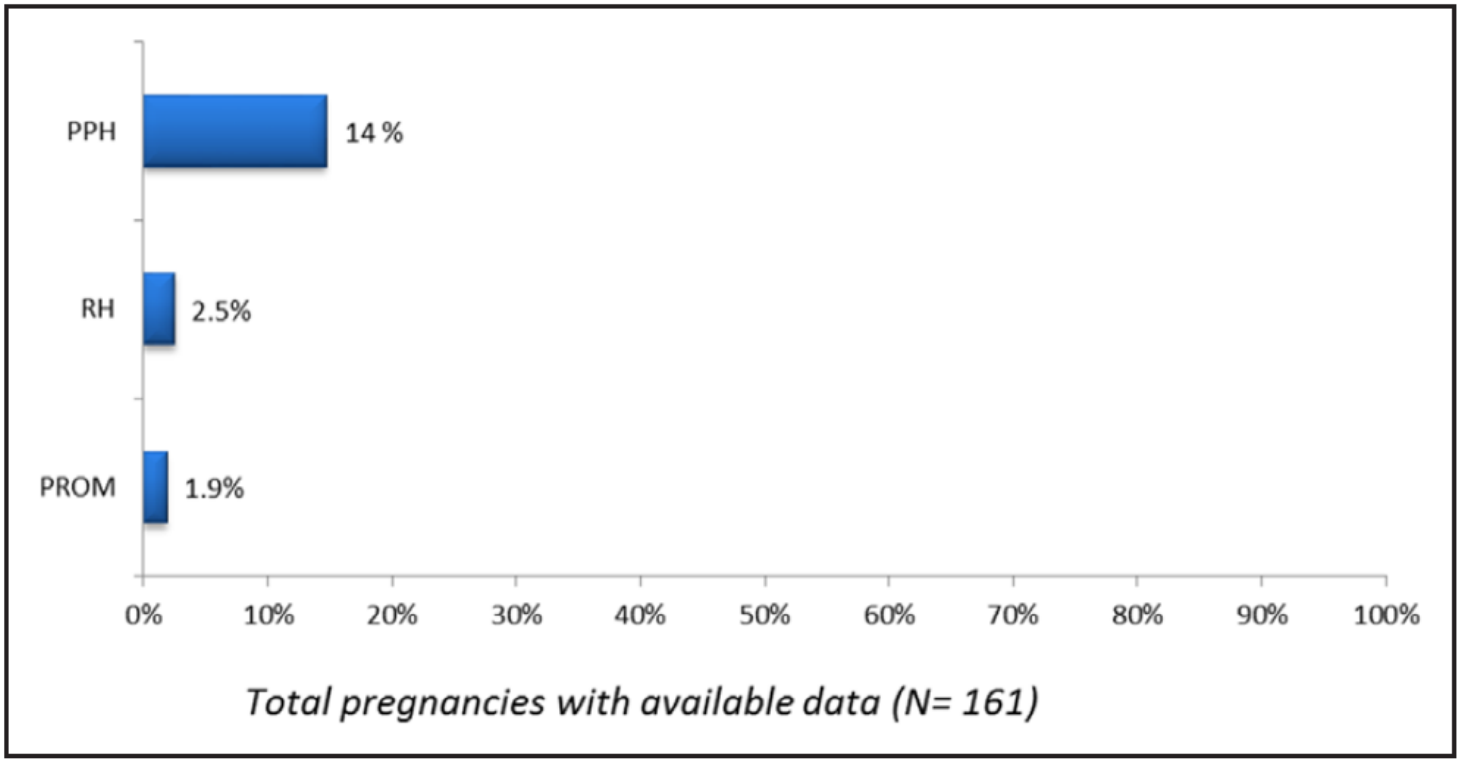
5% NND

59% premature

20% SGA

Garcia Ropero 2018 Circulation: Cardiovascular Quality and Outcomes

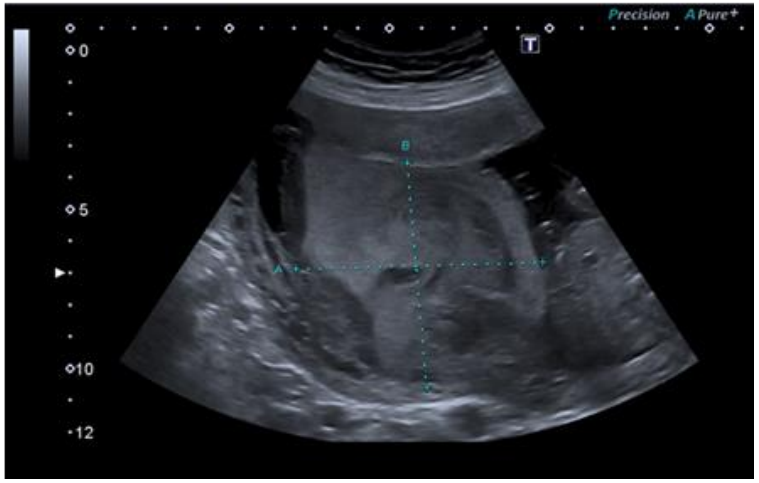
bleeding happens

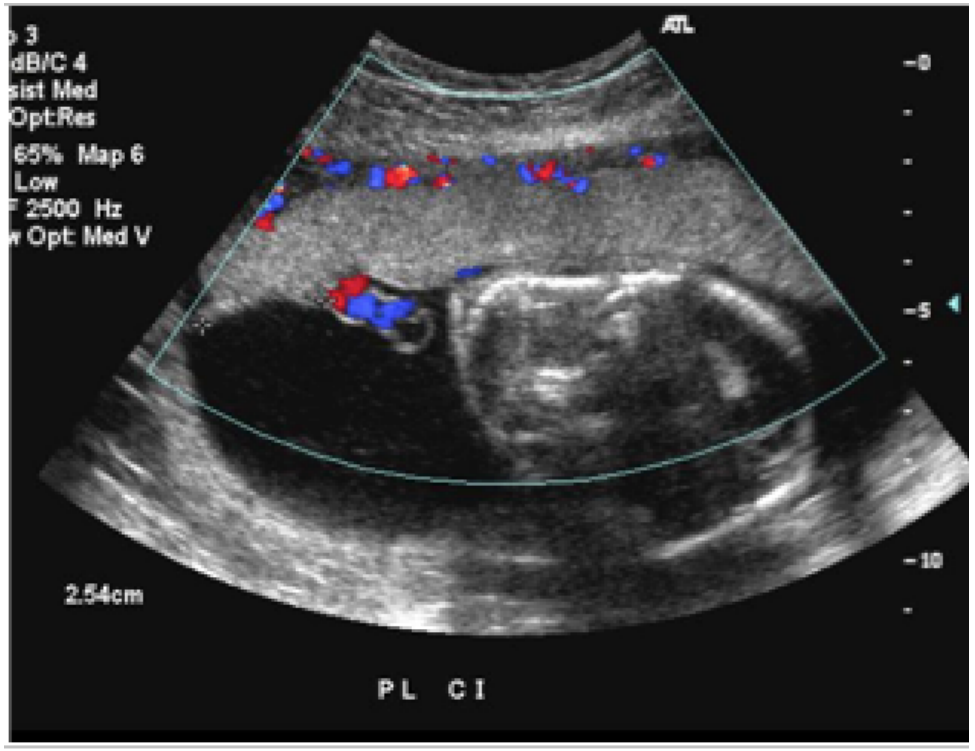


89% antiplatelet or anticoagulation

Garcia Ropero 2018 Circulation: Cardiovascular Quality and Outcomes

the placenta is abnormal





subchorionic fibrin deposition
infarcts
haematoma

Phillips AL et al. IJC 2019.

symptoms?

medication?

echo

CPET

MRI

any intervention pre-pregnancy?

maternal and fetal risks?

what can we do to reduce the risk?

reduce cyanosis

echoes

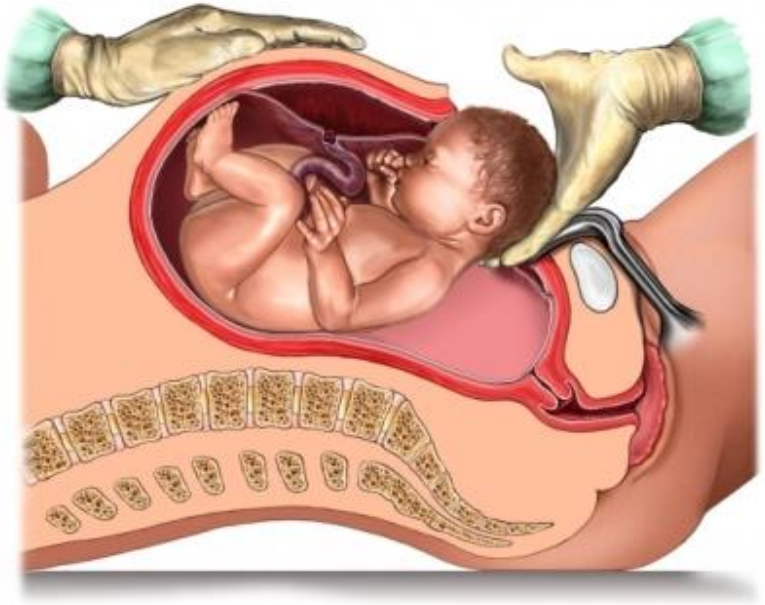
LFTs

clexane

frequent fetal scans

deliver early

SENIOR TEAM!



CS in 60%

epidural anaesthesia

shorter active 2nd stage

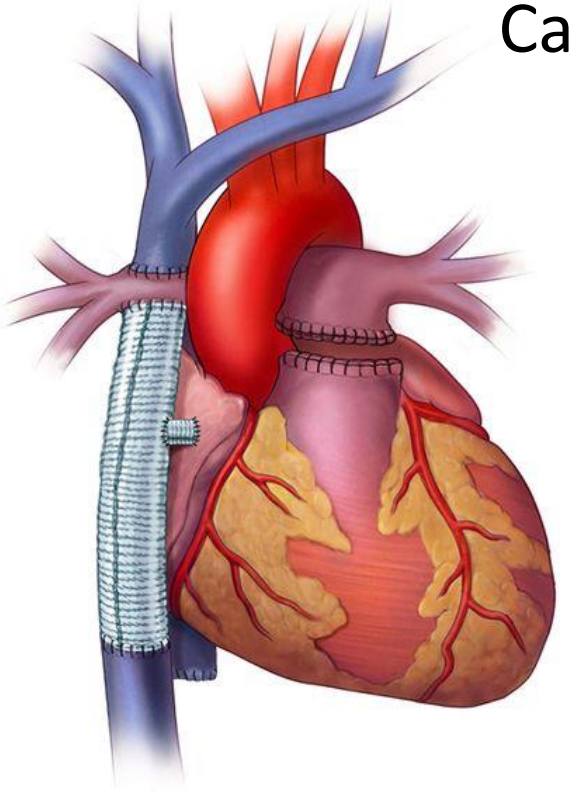
E Gelson et al. BJOG 2008. 115 (3):398–402
Balci A et al. Am Heart J. 2011 Feb;161(2):307-13

Laura
18 years old
Fontan

2005	A	1st T misc	
2006	A	1st T misc	
2006	B	1st T misc	
2008	C	SVD term	
2008	D	SVD 35 weeks	
2009	D	1st T misc	
2009	B	em CS term	
2009-13	B	>4 x 1st T misc	
2016	E	29 weeks el CS	Short gut due to NEC
2017	F	2nd T misc severe IUGR 17 weeks	
2018	G	1st T misc	
2019	H	PROM 25 weeks	NND 6 days
2019	D	1st T misc	

8 women, 2005-2019
all good function
all sats>90%

High risk WHO II-III
Low rate of cardiac complications
High rate of fetal loss
Careful monitoring by cardiac obstetric team



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