

Gravidanza nella donna con cardiopatia congenita: dal counselling pre-concezionale al follow up. Il pregnancy heart team.

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Bambino Gesù
OSPEDALE PEDIATRICO

No disclosure



Bambino Gesù
OSPEDALE PEDIATRICO

Why are we speaking about pregnancy in ACHD

What does happen during pregnancy

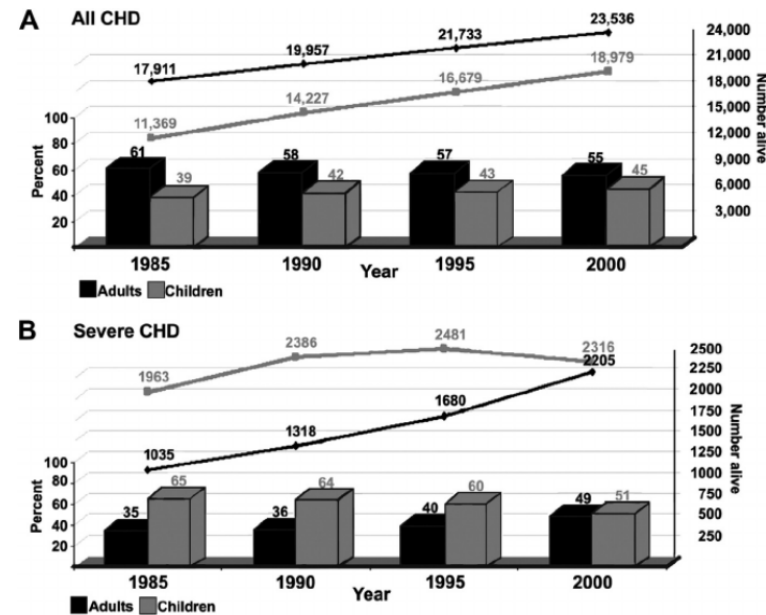
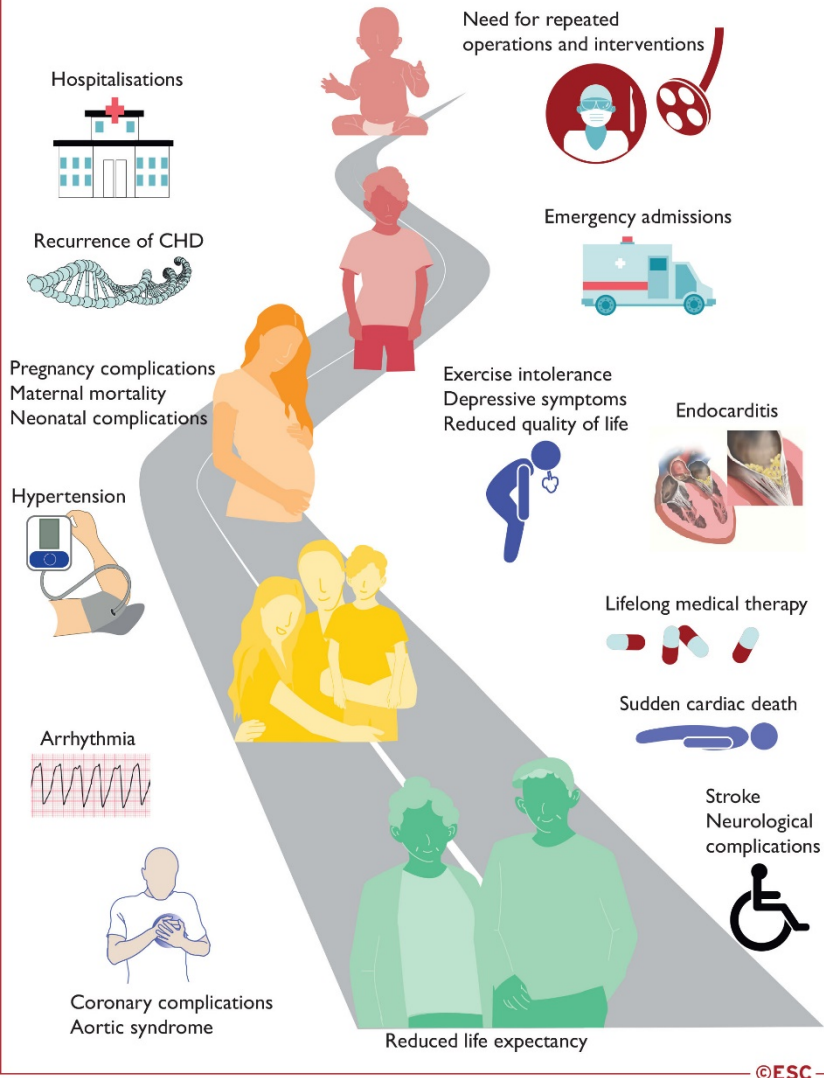
Who can or cannot get pregnant safely

When is the best time

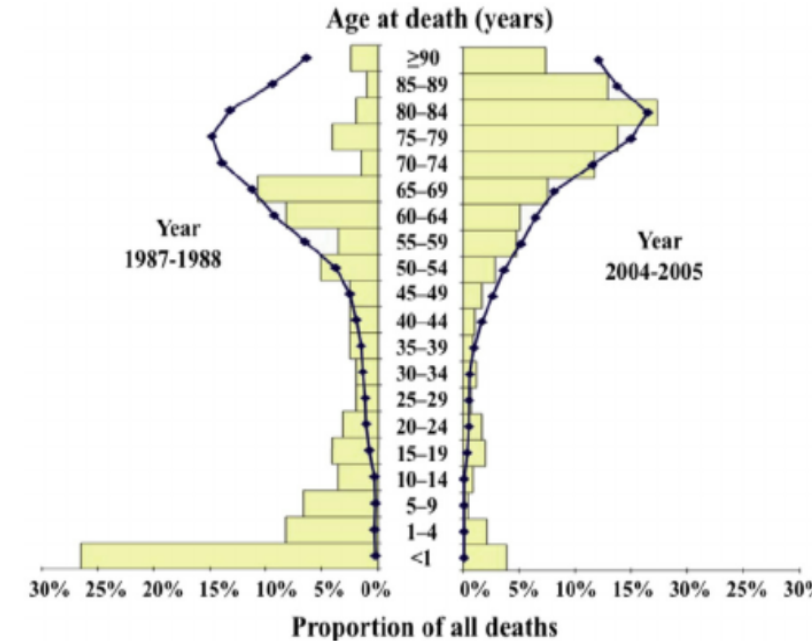
Where is the best place

Why are we speaking about pregnancy in ACOG

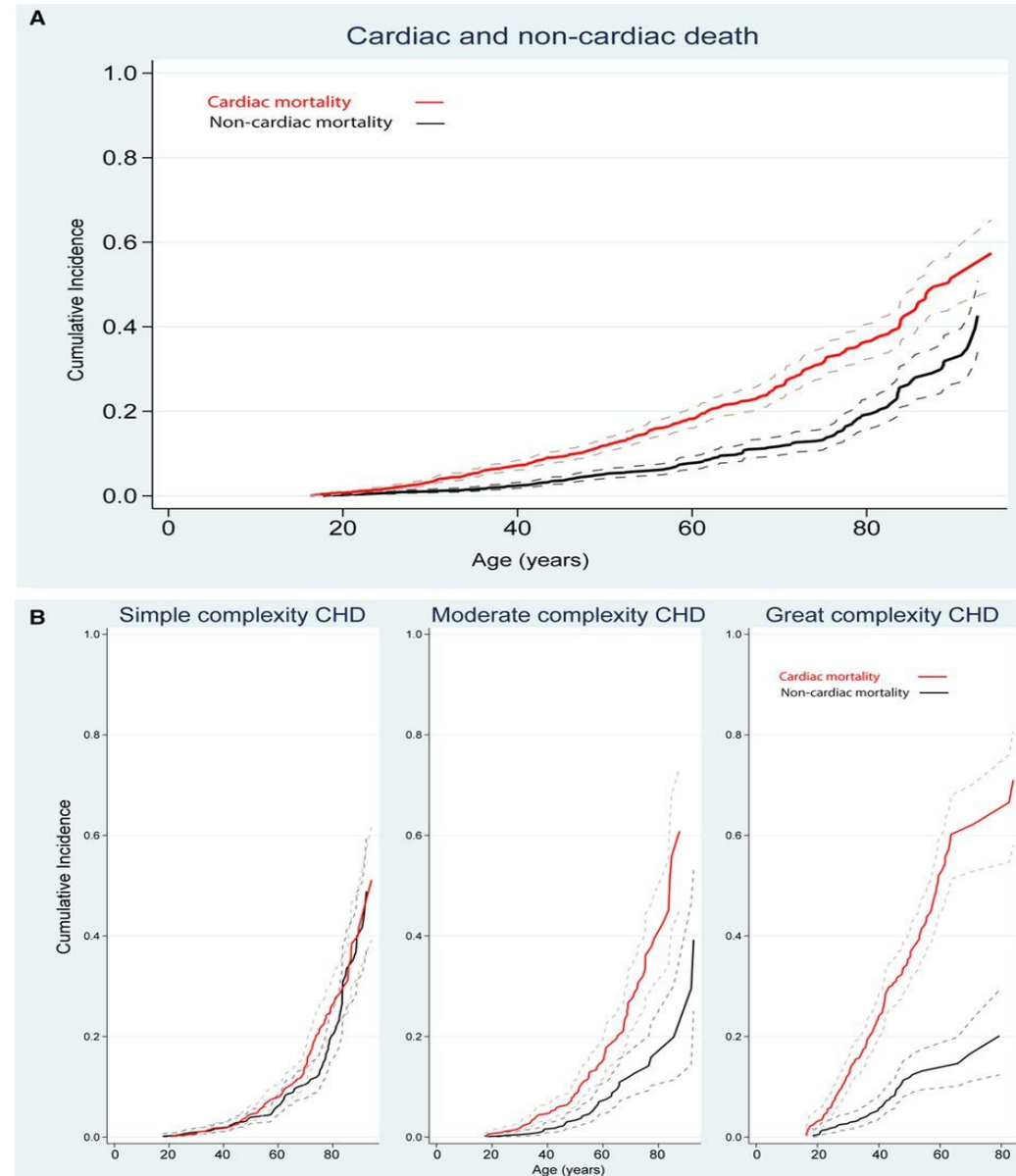
Congenital heart disease ~ A lifelong chronic condition



Number and proportion of adults and children with (A) all forms of congenital heart disease (CHD), and (B) severe CHD in 1985, 1990, 1995, and 2000. Reproduced from Marelli et al. 13

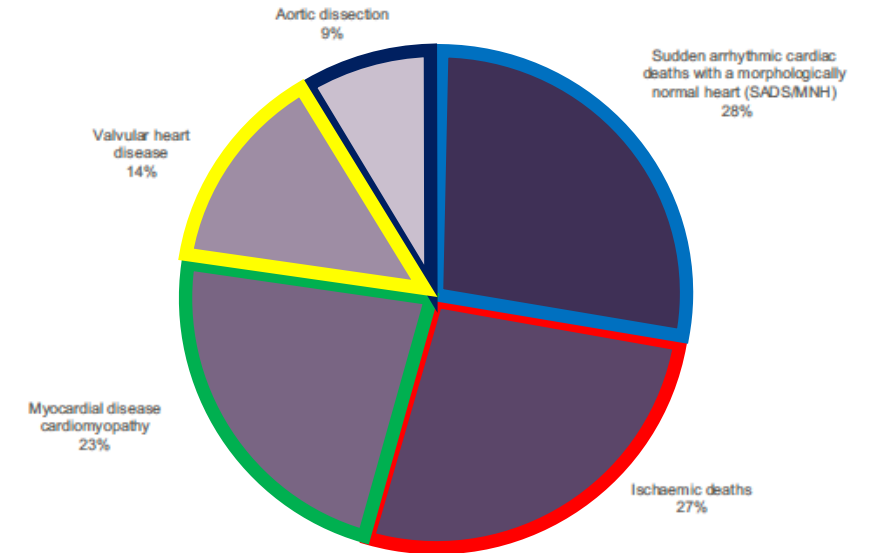
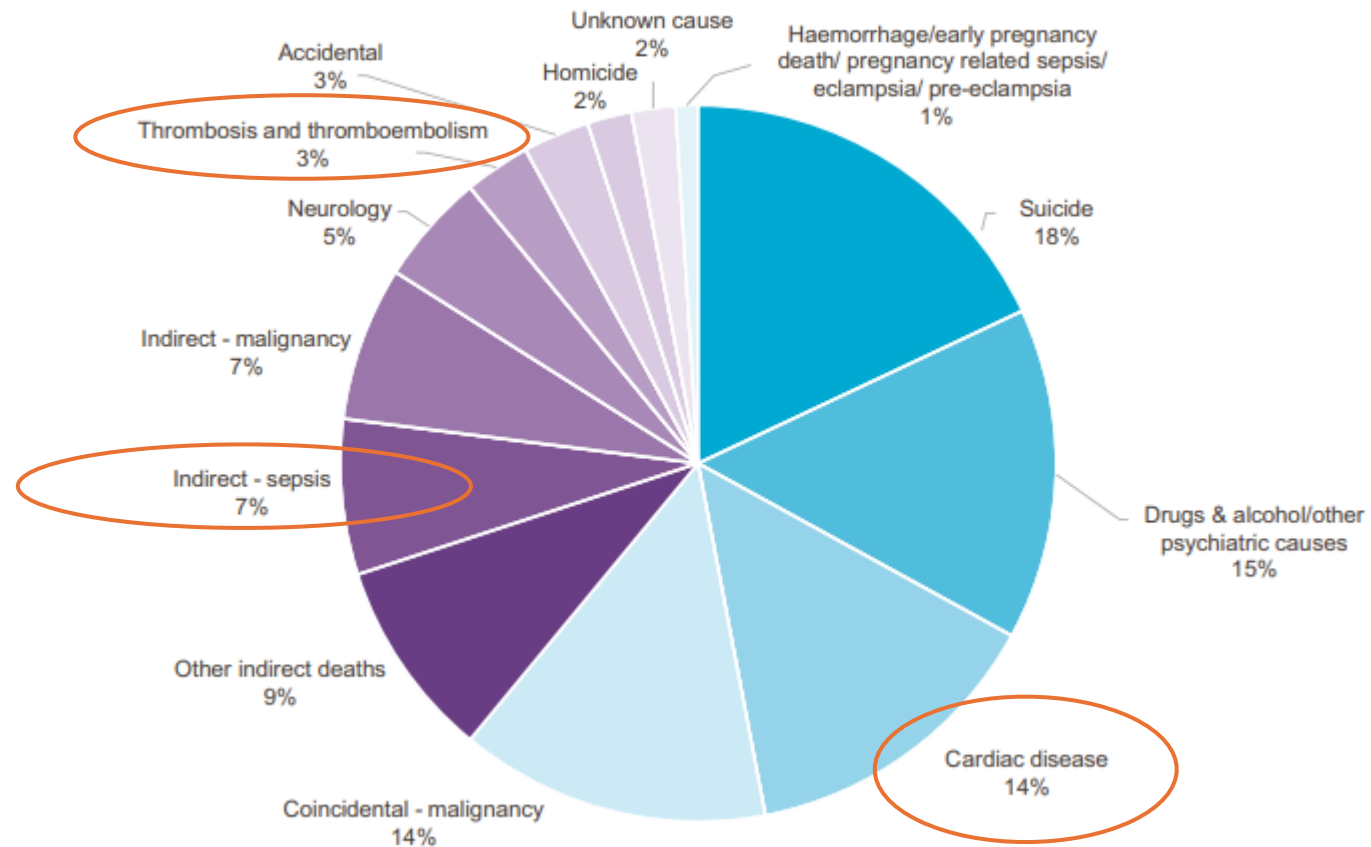


Why are we speaking about pregnancy in ACOG



Why are we speaking about pregnancy in AC

Figure 2.7: Proportion of maternal deaths by cause, amongst women who died between six weeks and one year after the end of pregnancy, UK 2021-23



What does happen during pregnancy

Significant changes occur in the maternal cardiovascular system soon after conception.

- Maintain adequate uterine perfusion
- Maintain fetal oxygenation
- Maintain fetal nutrient delivery

These changes can unmask previously unrecognised heart disease

What does happen during pregnancy

The fetoplacental unit exerts significant effects on the cardiovascular system through the production of sex hormones:

- Relaxin
- Progesterone
- Estrogen
- dehydroepiandrosterone (DHEA)
- Testosterone

Sex hormones **stimulate** endothelial synthesis of **nitric oxide** and **endogenous prostacyclin** (vasodilators) and **inhibit the activity of endothelin-1** (vasoconstrictor), resulting in vasodilation.

This, in turn, leads to incomplete filling of the vascular bed and atria, activating the **renin-angiotensin-aldosterone** system and inhibiting natriuretic hormone.

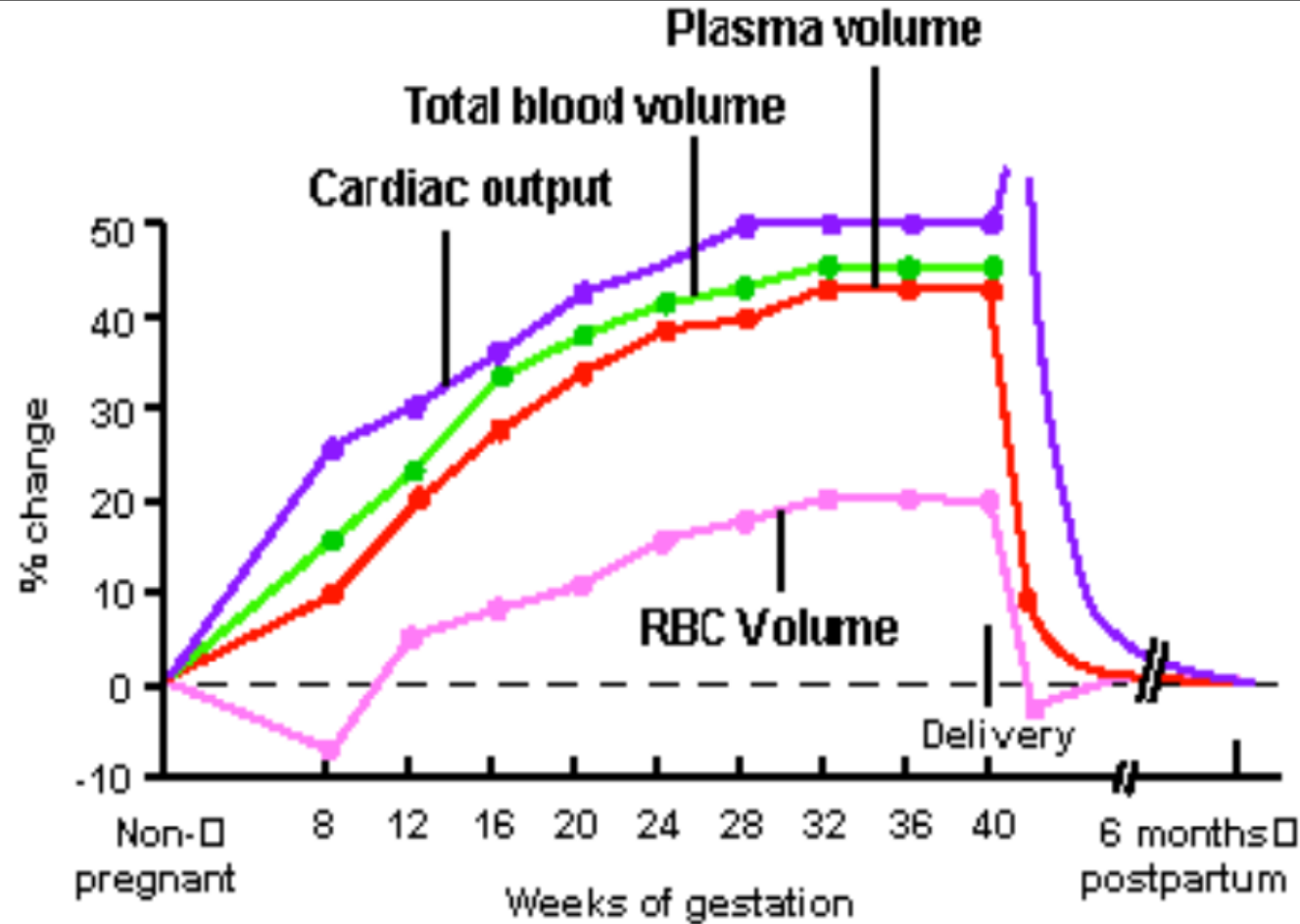
This results in significant sodium and fluid retention, dilutional anemia, and reduced blood viscosity.

What does happen during pregnancy

Cardiocirculatory changes during normal pregnancy

	Early Pregnancy	Mid-Pregnancy	Late Pregnancy
Heart rate	↑↑	↑↑↑	↑↑↑↑
Systolic Blood Pressure	↓	↔	↑↑
Diastolic Blood Pressure	↓	↓	↑↑
Stroke Volume	↑↑↑↑	↑↑↑↑↑↑	↑↑↑↑↑
Cardiac Output	↑↑↑↑↑↑	↑↑↑↑↑↑	↑↑↑↑↑↑↑
SVR	↓↓↓↓↓	↓↓↓↓↓↓	↓↓↓↓↓
Ejection Fraction	↑↑	↑↑	↑

What does happen during pregnancy



Inadequate plasma volume expansion

- IUGR (intrauterine growth restriction)
- SGA (Small for gestational age)
- Pre-eclampsia

Can lead to rapid cardiac decompensation with pre-existing or unknown CV disease

What does happen during pregnancy

Blood pressure

- Decreases as early as 7 wks
- Nadir by mid-pregnancy
- Near normal at term

SVR

- 10% decrease in 1° trimester → Preload increases & afterload decreases
- Nadir by mid-pregnancy (20-30% decrease) → Preload increases & afterload reaches a nadir
- Remains overall constant until term (small increase) → Preload reaches an acme plateau (+500cc) & afterload plateaus or increases mildly

What does happen during pregnancy

At cardiac level:

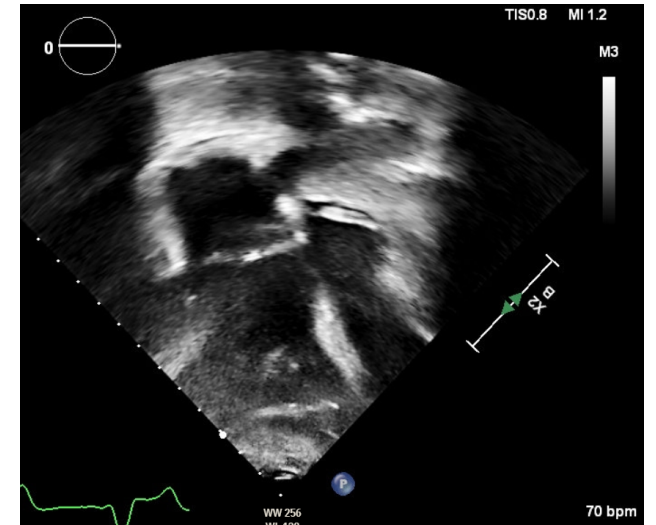
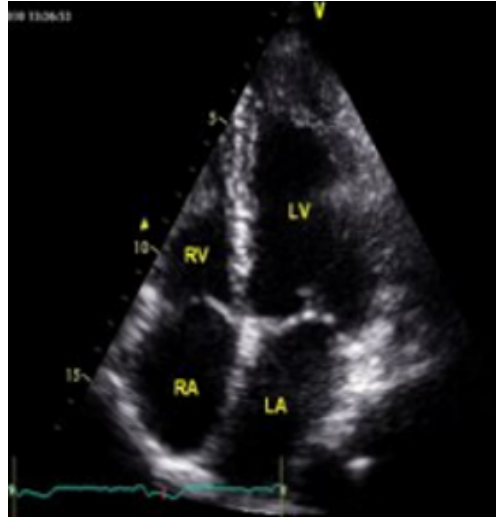
Atrial volumes increases

Ventricular volumes increases

Myocardial mass increases

LV stroke volume increase (up to 20%)

LV CO increases (early 13% ; peak 25-50% at late T2; plateau or decline by term)



What does happen during pregnancy

... in labor:

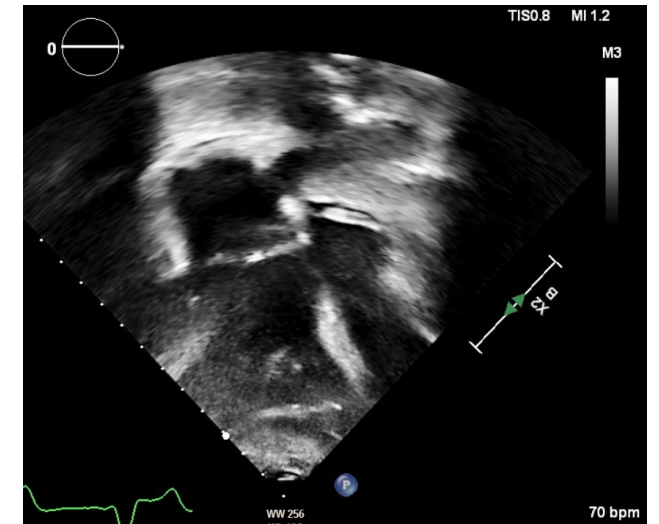
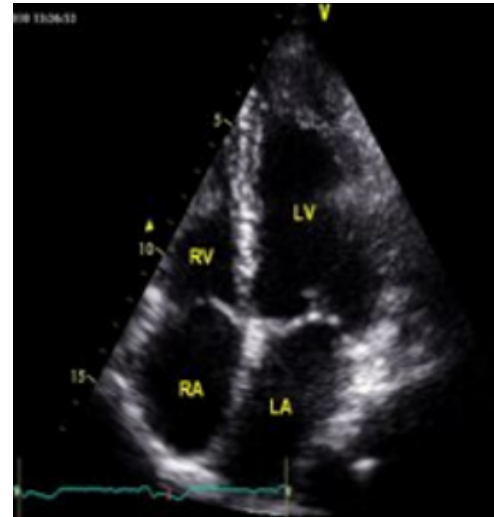
30% increase in CO during 1° stage of labor

50% increase during 2° stage

Uterine compression with contraction
increased blood volume
increased venous return
increased stroke volume

HR with variable changes

BP increases in labor



What does happen during pregnancy

Cardiac output (CO) increases as early as 5wks gestation

Increases mostly due to increased stroke volumes

Peaks around 30-35wks

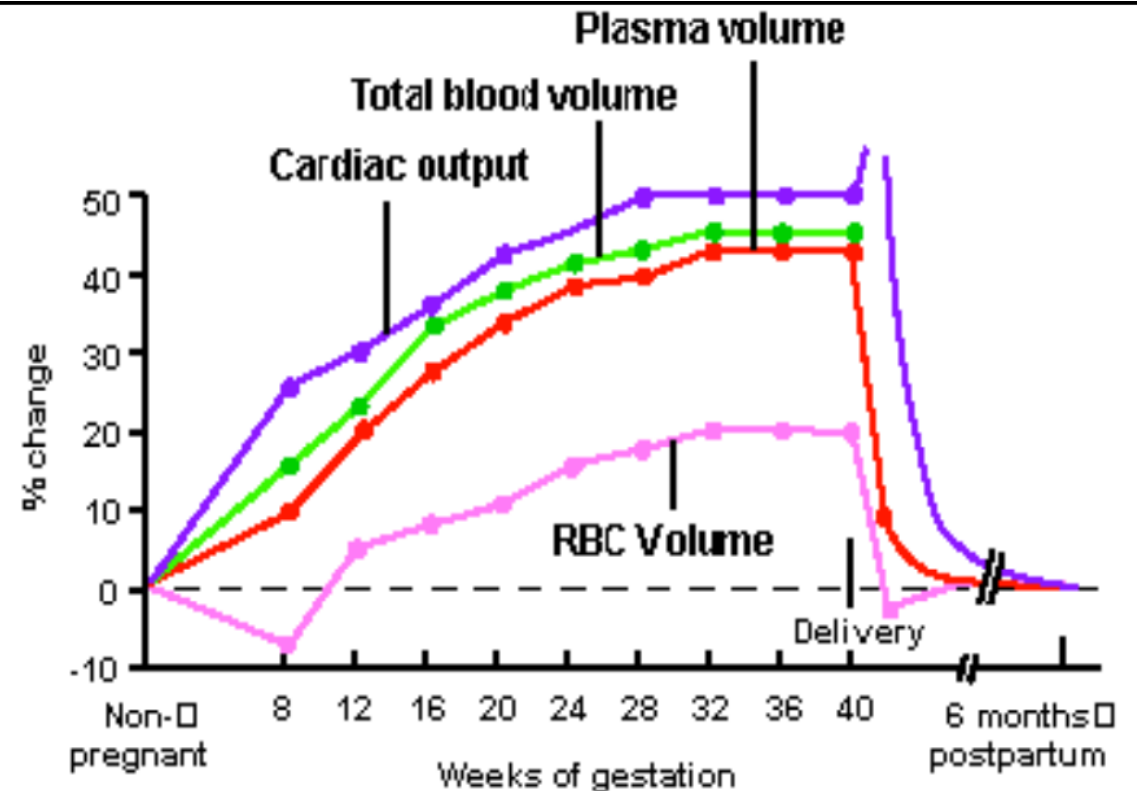
Late in pregnancy, increased CO mostly attributable to increased HR

Red blood cell mass increases up to 40%

- increased production of red cells
- Increased need for maternal iron (50% incr)
- Increased oxygen dissociation
- Increased fetal oxygen transfer

Plasma volume increase is larger

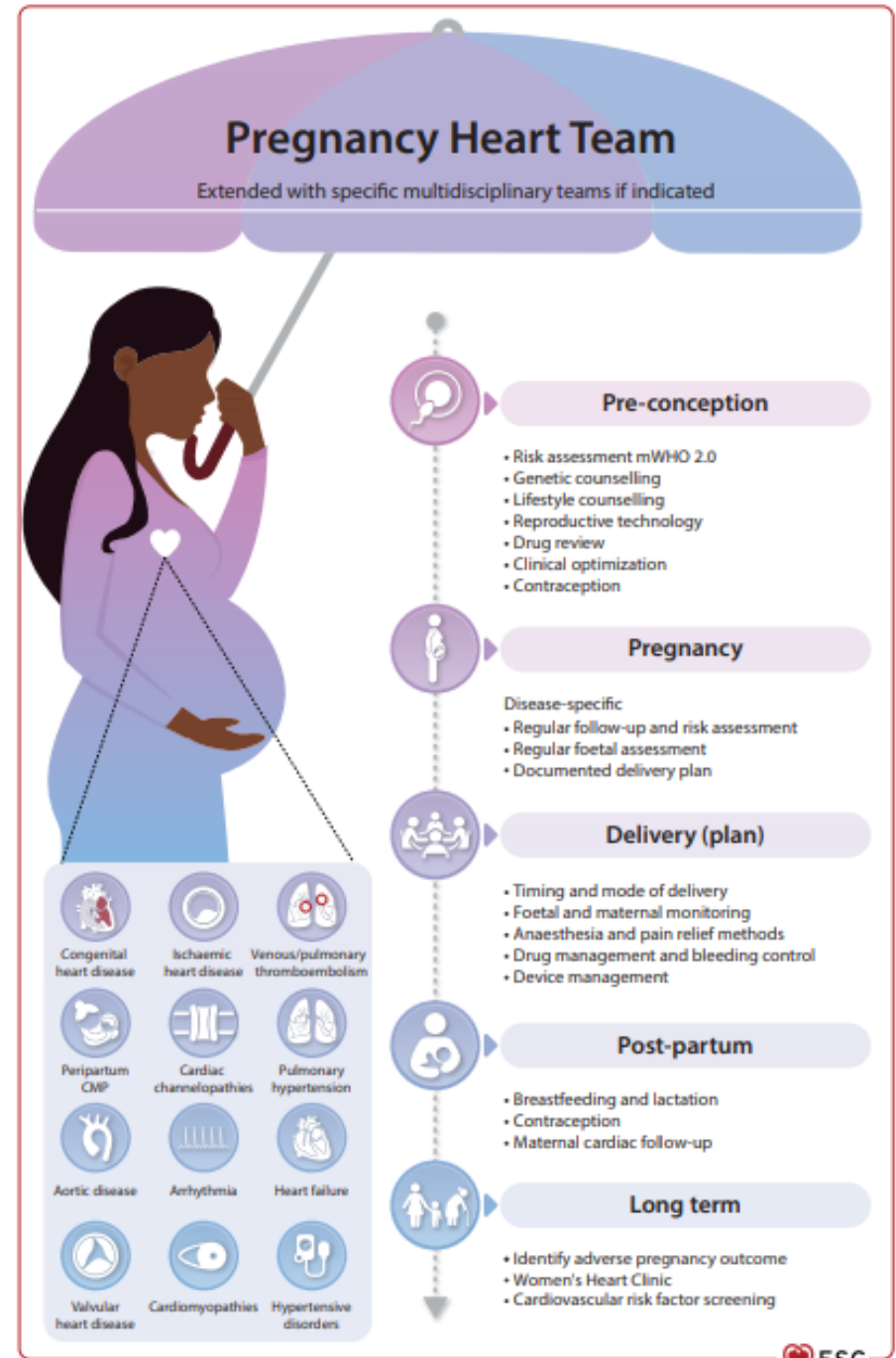
- Physiological anemia of pregnancy
- Hct 33-38%



Who can or cannot get pregnant safely

2025 ESC Guidelines for the management of cardiovascular disease and pregnancy

29 Aug, 2025



Who can or cannot get pregnant

Classificazione del rischio (mWHO 2.0)

		mWHO 2.0 I	mWHO 2.0 II	mWHO 2.0 II-III	mWHO 2.0 III	mWHO 2.0 IV
		Congenital heart disease				
		Successfully repaired simple lesions without significant residual (haemodynamic) complications (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage).	Unoperated uncomplicated atrial or ventricular septal defect. Repaired tetralogy of Fallot without significant residual haemodynamic/ arrhythmic lesions. Transposition of the great arteries with arterial switch without significant residual lesions.	Repaired atrioventricular septal defect without significant residual lesions. Uncomplicated Ebstein anomaly: mild to moderate TR, no tricuspid stenosis, no accessory pathway.	Unrepaired cyanotic heart disease (not Eisenmenger). Systemic RV with good or mildly decreased ventricular function. Uncomplicated Fontan circulation: good ventricular function, no significant valve disease or arrhythmias, good exercise tolerance, and normal arterial saturations. Ebstein anomaly with any complication.	Systemic RV with moderate or severely decreased ventricular function. Fontan with any complication. Eisenmenger syndrome.
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.
Average maternal cardiac event rates ^a	Van Hagen et al. (2016) ⁵¹	9.9%	7.7%	17.7%	28.9%	50.3%
	Silversides et al. (2018) ⁵²	3.1%	21.7%	12.8%	21.1%	35.6%
Individualize each maternal risk with the modifiers below^b (derived from CARPREG II)⁵²						
CARPREG II score: 1 point		CARPREG II score: 2 points		CARPREG II score: 3 points		
• No prior cardiac intervention indicated		• Ventricular dysfunction		• Prior cardiac event or arrhythmias		
• Late pregnancy assessment		• High-risk left-sided valve disease or outflow tract obstruction		• Baseline NYHA III/IV or cyanosis		
		• Pulmonary hypertension		• Mechanical valve		
		• Coronary artery disease				
		• High-risk aortopathy				

		mWHO 2.0 I	mWHO 2.0 II	mWHO 2.0 II-III	mWHO 2.0 III	mWHO 2.0 IV
		Valvular heart disease				
		Small or mild - pulmonary stenosis - mitral valve prolapse without significant regurgitation.		Native, homograft or tissue valve disease not considered mWHO 2.0 I or IV: mild mitral stenosis, moderate aortic stenosis. Moderate valvular regurgitation.	Uncomplicated mechanical valve with stable well controlled INRs. Moderate mitral stenosis. Severe asymptomatic aortic stenosis. Severe left-sided valvular regurgitation.	Severe mitral stenosis. Severe symptomatic aortic stenosis.
		Aortopathy				
		Non-HTAD mild aortic dilatation (<40 mm).	Turner syndrome without cardiovascular features (BAV, coarctation, AHT, aortic dilatation).	Marfan or other HTAD syndrome without aortic dilatation. Aorta <45 mm in BAV pathology. Repaired coarctation.	Moderate aortic dilatation: 40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in BAV, Turner syndrome ASI 20–25 mm/m ² , other aortic dilatation <50 mm. Marfan with previous aortic root replacement. Previous aortic dissection with stable diameter.	Severe aortic dilatation: >45 mm in Marfan syndrome or other HTAD, >50 mm in BAV, ASI >25 mm/m ² in Turner syndrome, other aortic dilatation >50 mm. Vascular Ehlers–Danlos syndrome. Severe (re)coarctation. Previous aortic dissection with increasing diameter.
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.
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Who can or cannot get pregnant safely

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					Fontan with any complication.
					Eisenmenger syndrome.
					50%

Who can or cannot get pregnant safely

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				Systemic RV with good or mildly decreased ventricular function.	
				Uncomplicated Fontan circulation: good ventricular function, no significant valve disease or arrhythmias, good exercise tolerance, and normal arterial saturations.	
				Ebstein anomaly with any complication.	

30%

Who can or cannot get pregnant safely

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/m², tetralogy of Fallot <50 mm) Ventricular d	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, >40 mm in bicuspid aortic valve, Turner syndrome ASI >25 mm/m², tetralogy of Fallot >50 mm) Vascular Ehlers–Danlos Severe (rel



European Heart Journal (2018) 39, 3165–3241
doi:10.1093/eurheartj/ehy340

2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy

ESC GUIDELINES

19-27%

40-100%

contraindicated

Clinical case: 1987

**Transposition of the great arteries with intact interventricular septum.
Infundibular-valvular pulmonary stenosis. Small apical aneurysm of the left ventricle.**

Underwent:

- Mustard procedure, Mr. Marcelletti, 1987
- Electrophysiological study negative for inducible arrhythmias, May 2003, age 16
Cardiac MRI (OPBG, October 2010): Right ventricular ejection fraction = 52%
Electrophysiological study negative for inducible arrhythmias, June 2012, age 25

Comorbidities:

- Hypothyroidism

CARDIOPULMONARY TEST 07/2020:

Exercise test stopped at **94% of maximum predicted heart rate** due to difficulty sustaining the imposed workload, **without symptoms**.

Slightly reduced exercise tolerance (duration 10 min 36 sec).

Heart rate response within normal limits (max HR 175 bpm).

Normal **blood pressure** response to exercise (max BP 140/80 mmHg).

Rare supraventricular premature beats (SVPBs) during early recovery, isolated and one couplet (RR' 260 ms) in immediate recovery.

One couplet with wide QRS (recording affected by artifacts) in phase 2 (RR' 280 ms).

SpO₂ at stopping: 98–99%.

Peak VO₂: 1440 ml/min – 26.2 ml/kg/min with peak respiratory quotient (RQ) 1.1.

24-hour Holter ECG (07/2020):

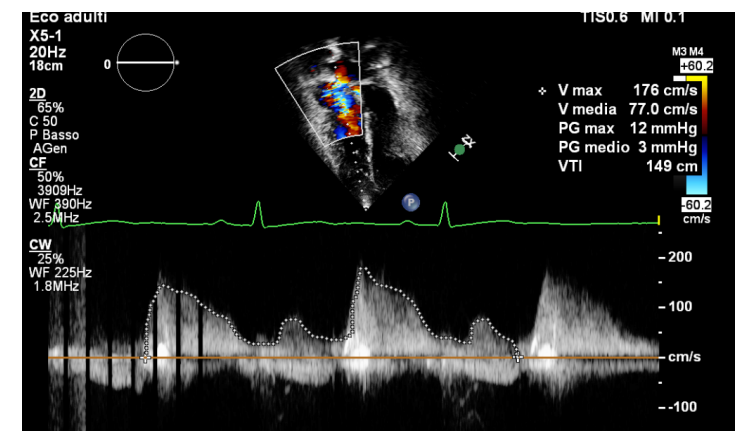
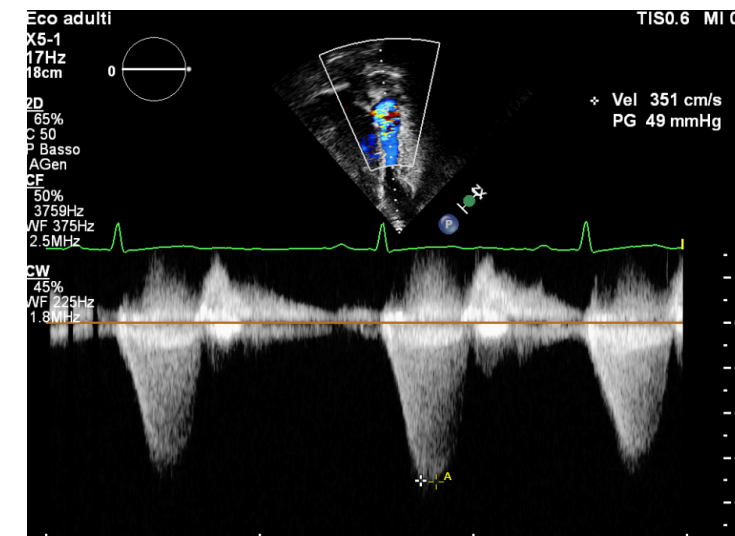
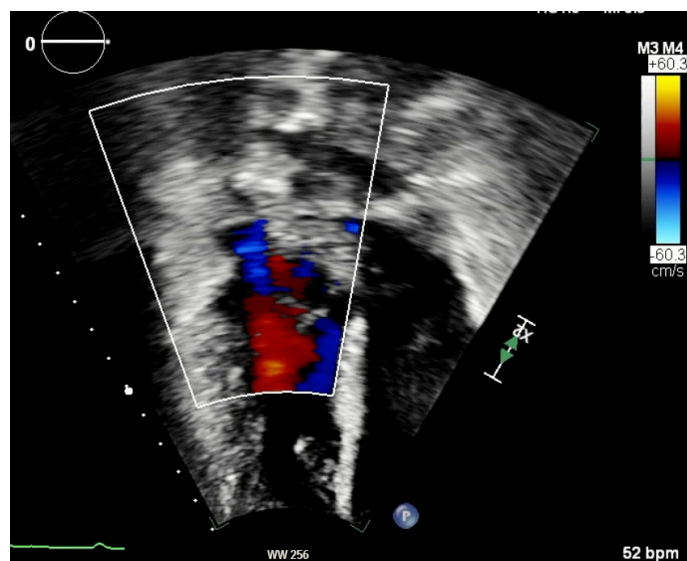
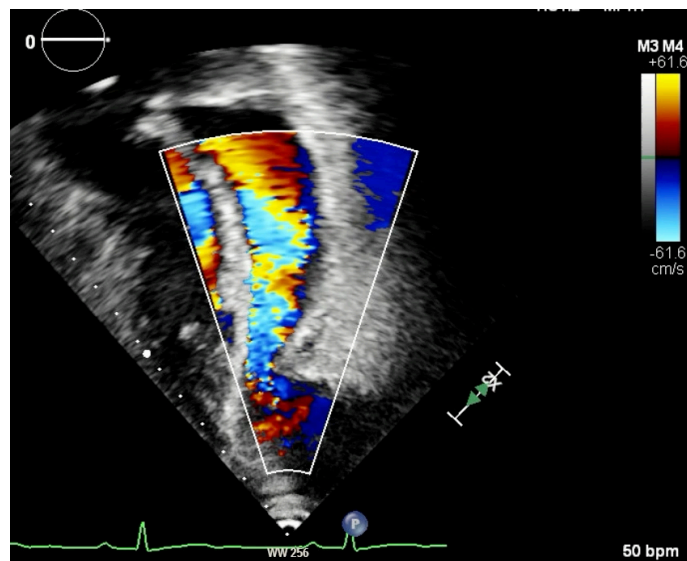
Isolated monomorphic ventricular premature beats (VPBs) (arrhythmic burden 1%), 22 couplets, one triplet.

One run of non-sustained ventricular tachycardia (7 beats at 150 bpm).

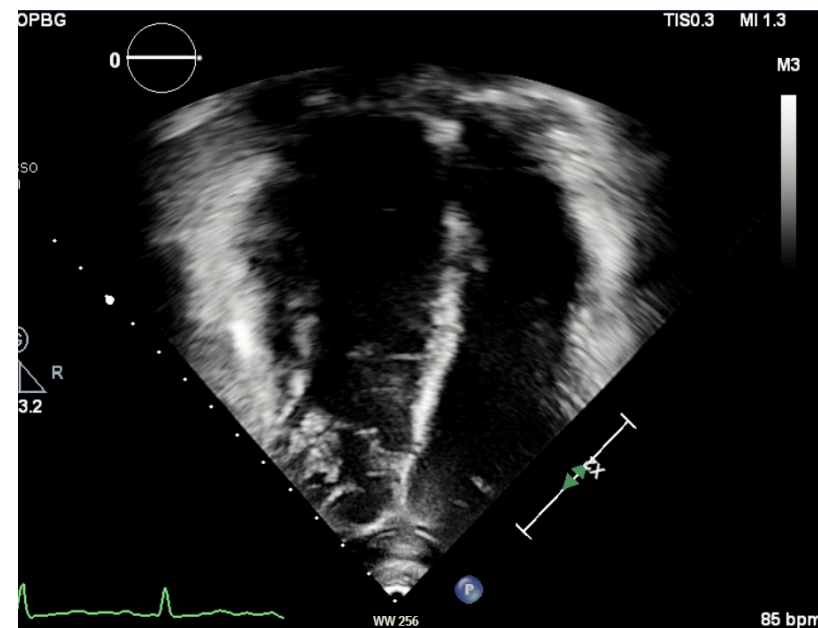
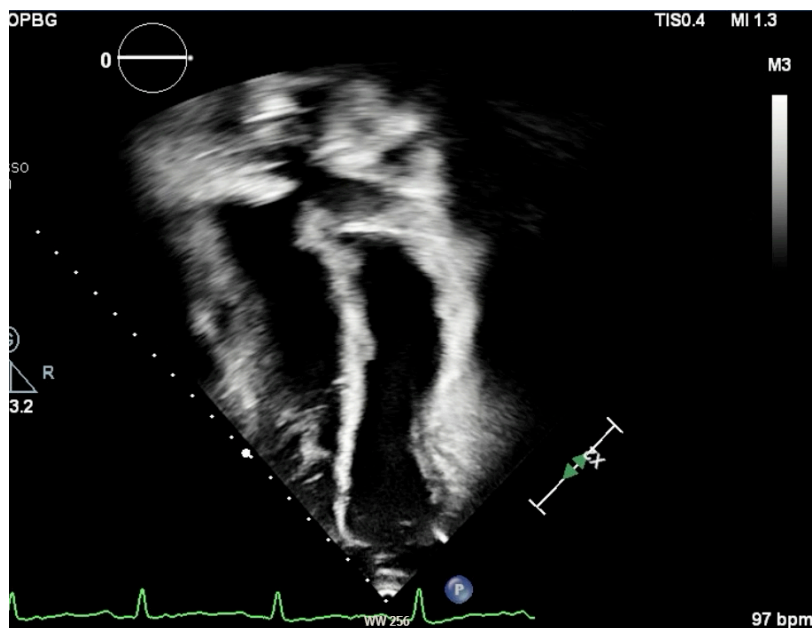
Asymptomatic

No pauses.

07/2020

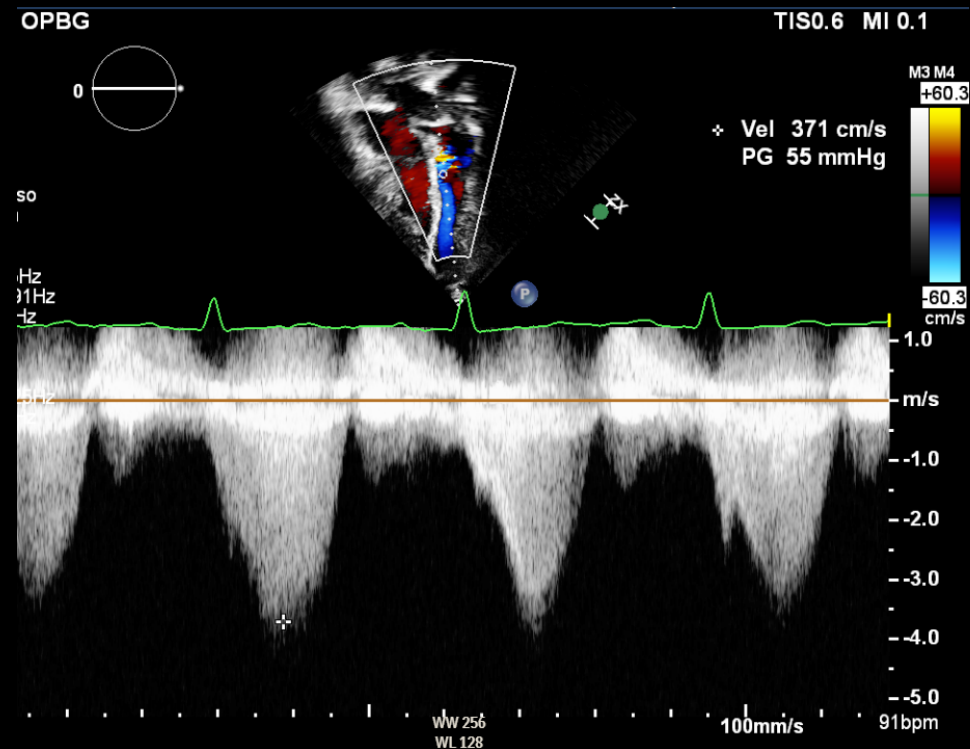
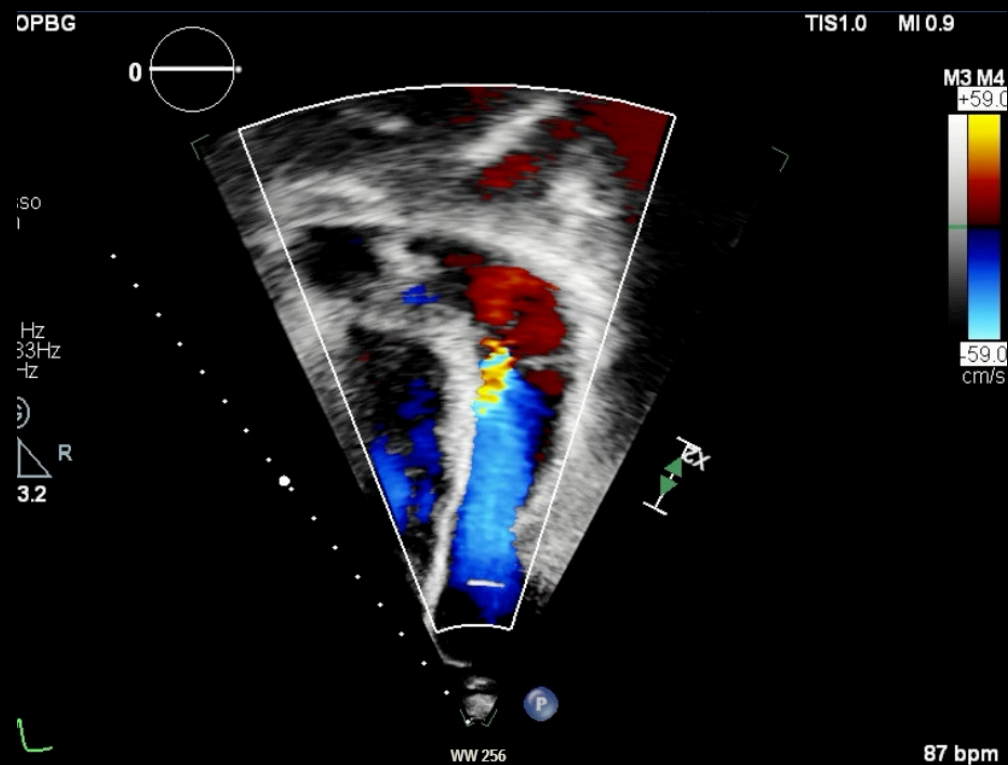


11/2020 call pregnant!

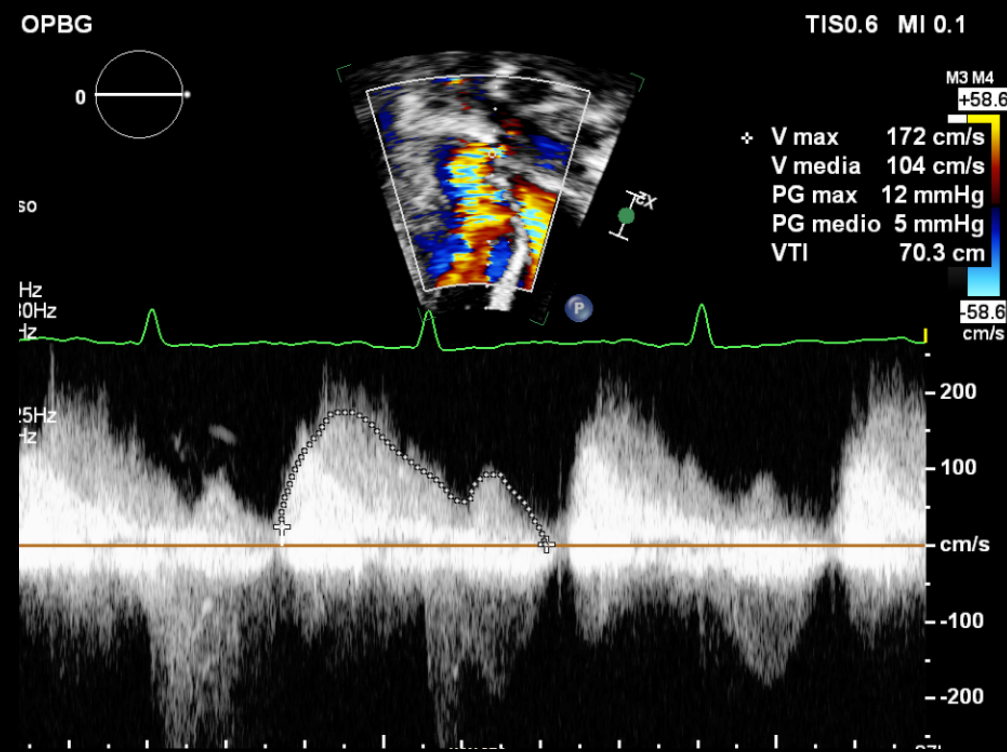
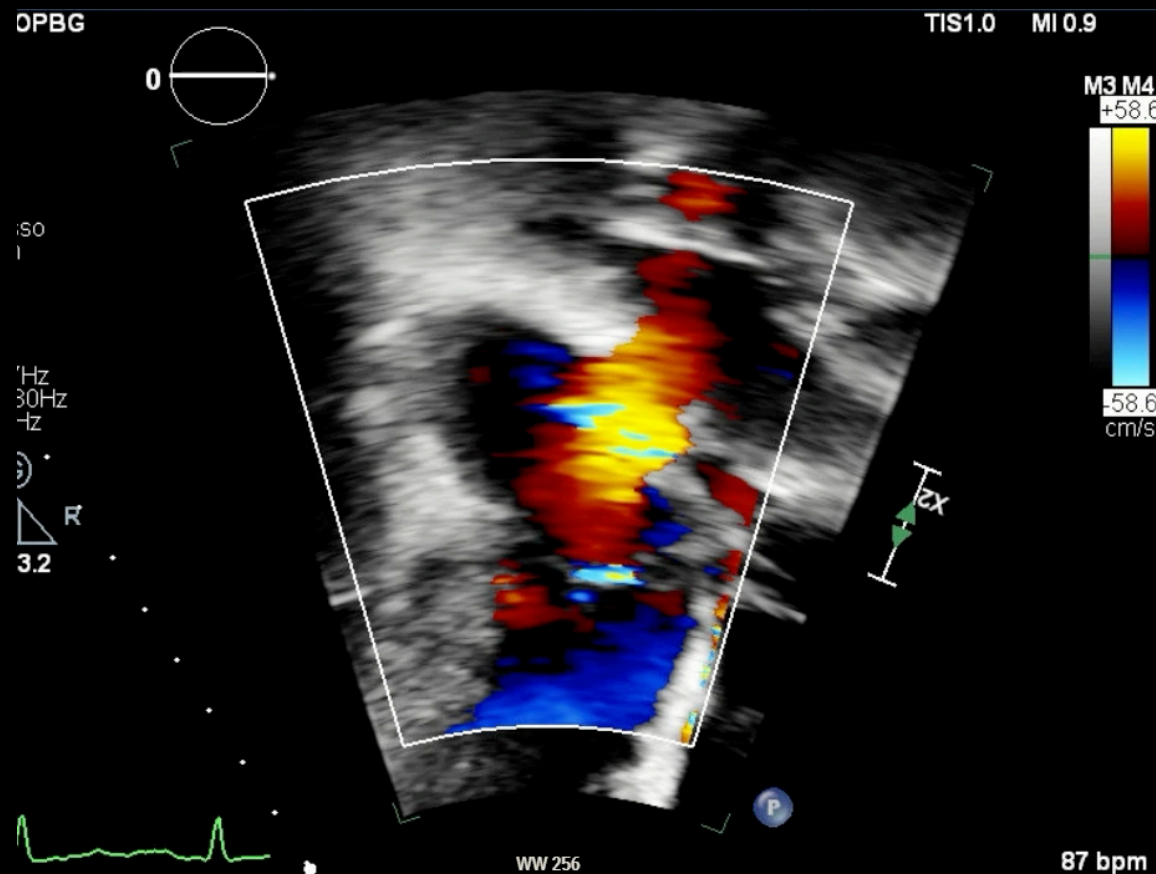


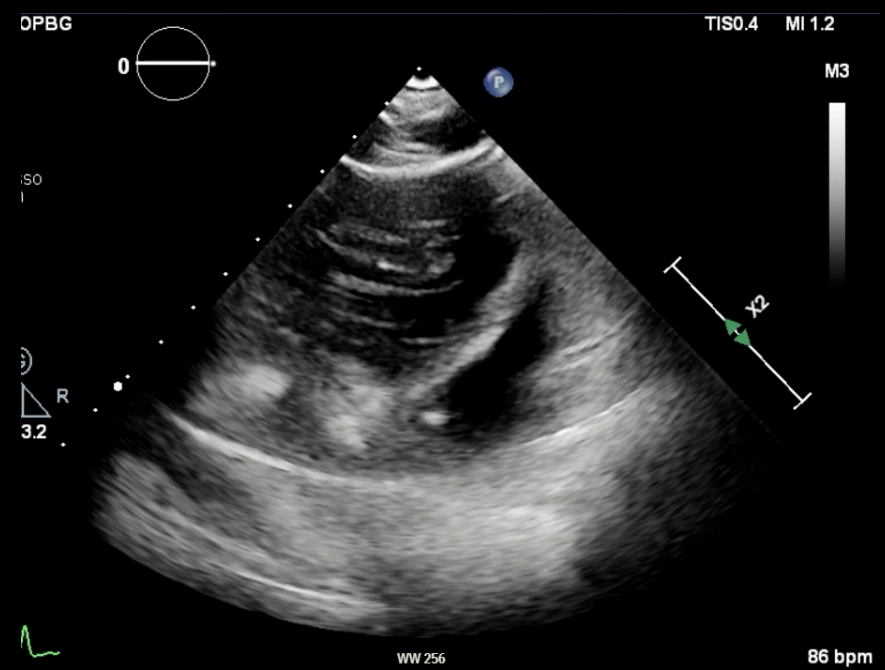
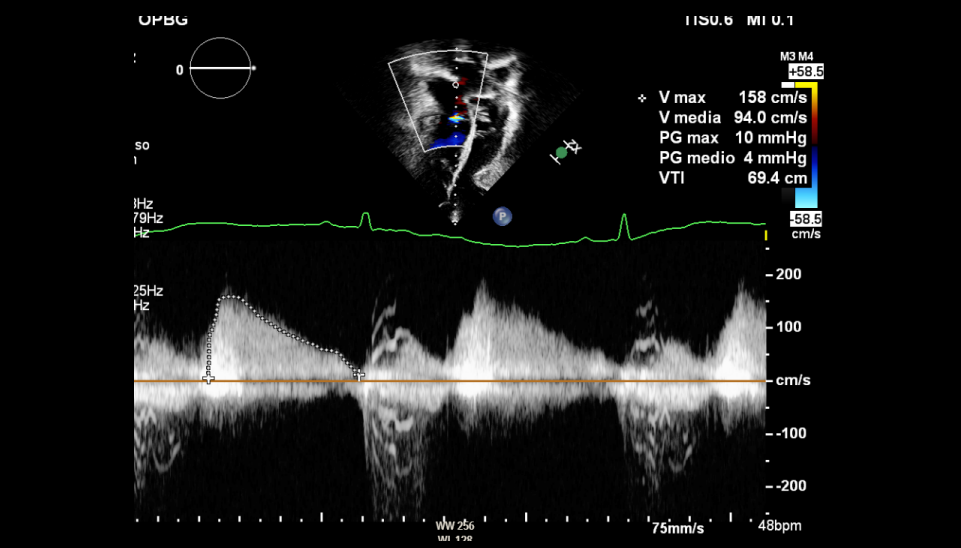
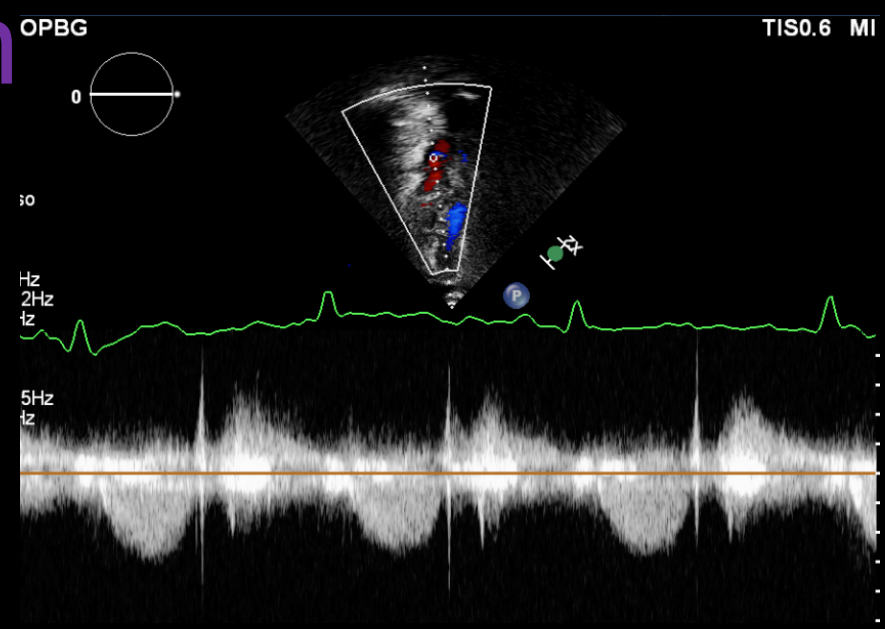
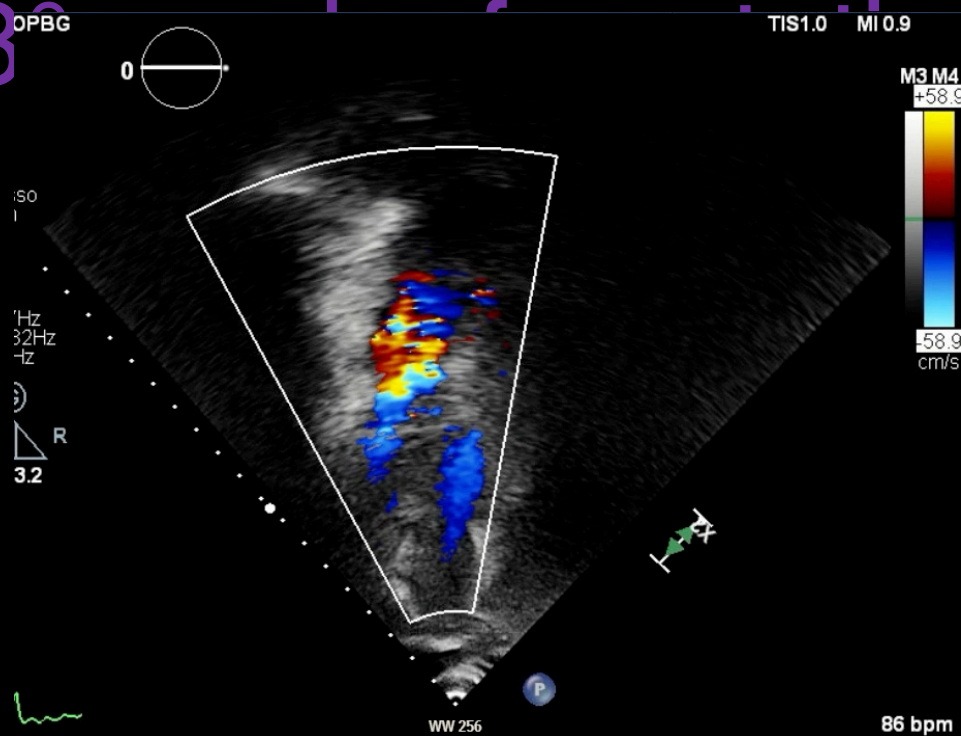
✦ Vel 329 cm/s
PG 43 mmHg

8^o week of gestation

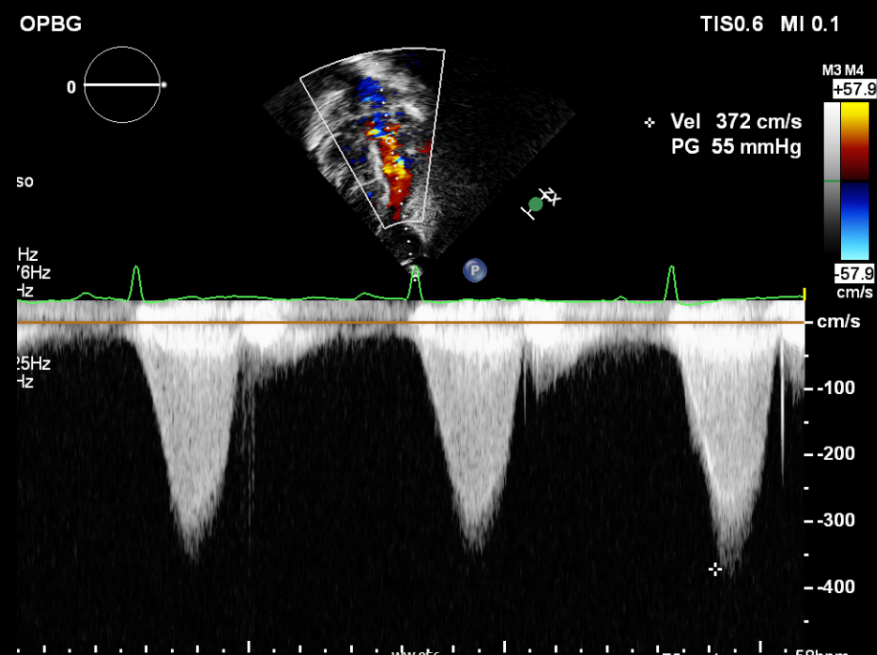
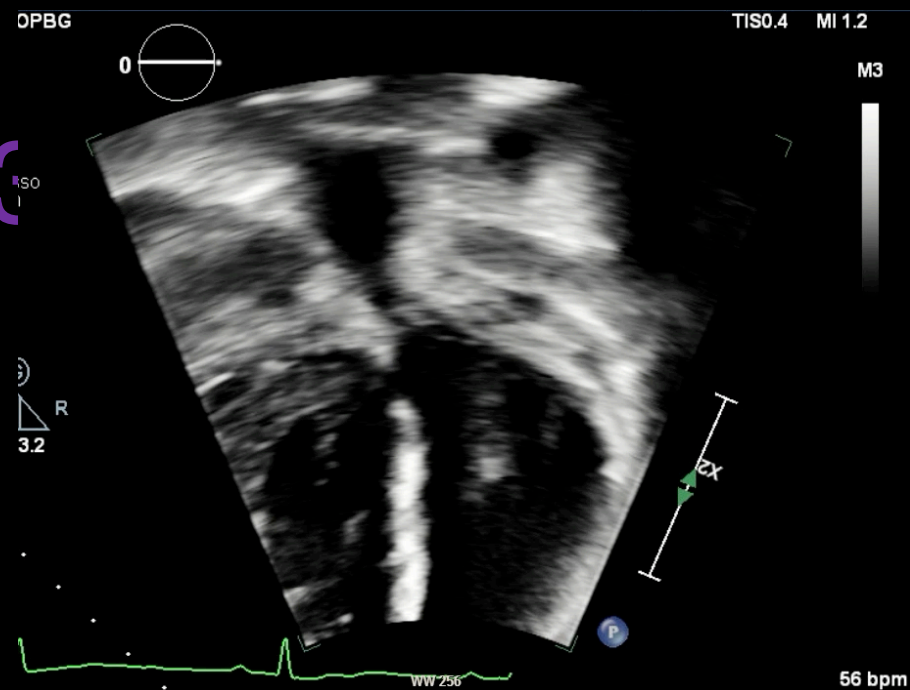
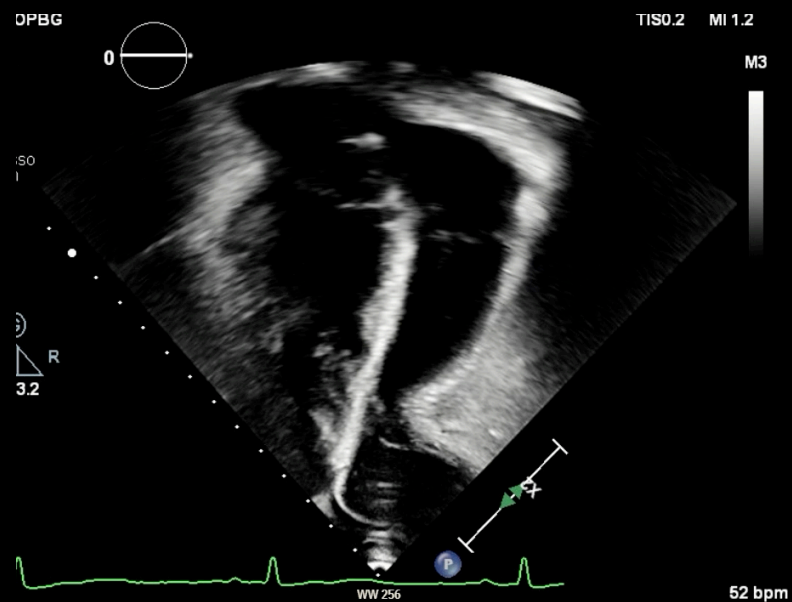


8^o week of gestation





Week of gestation 3



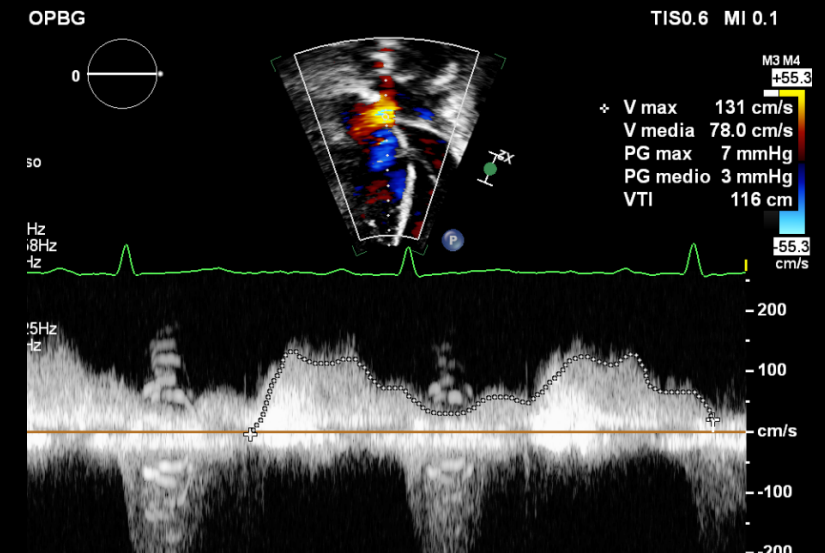
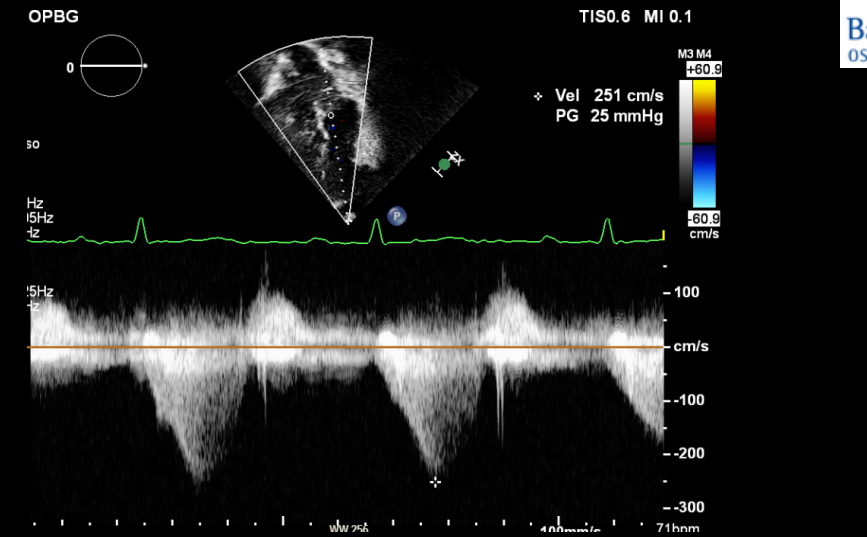
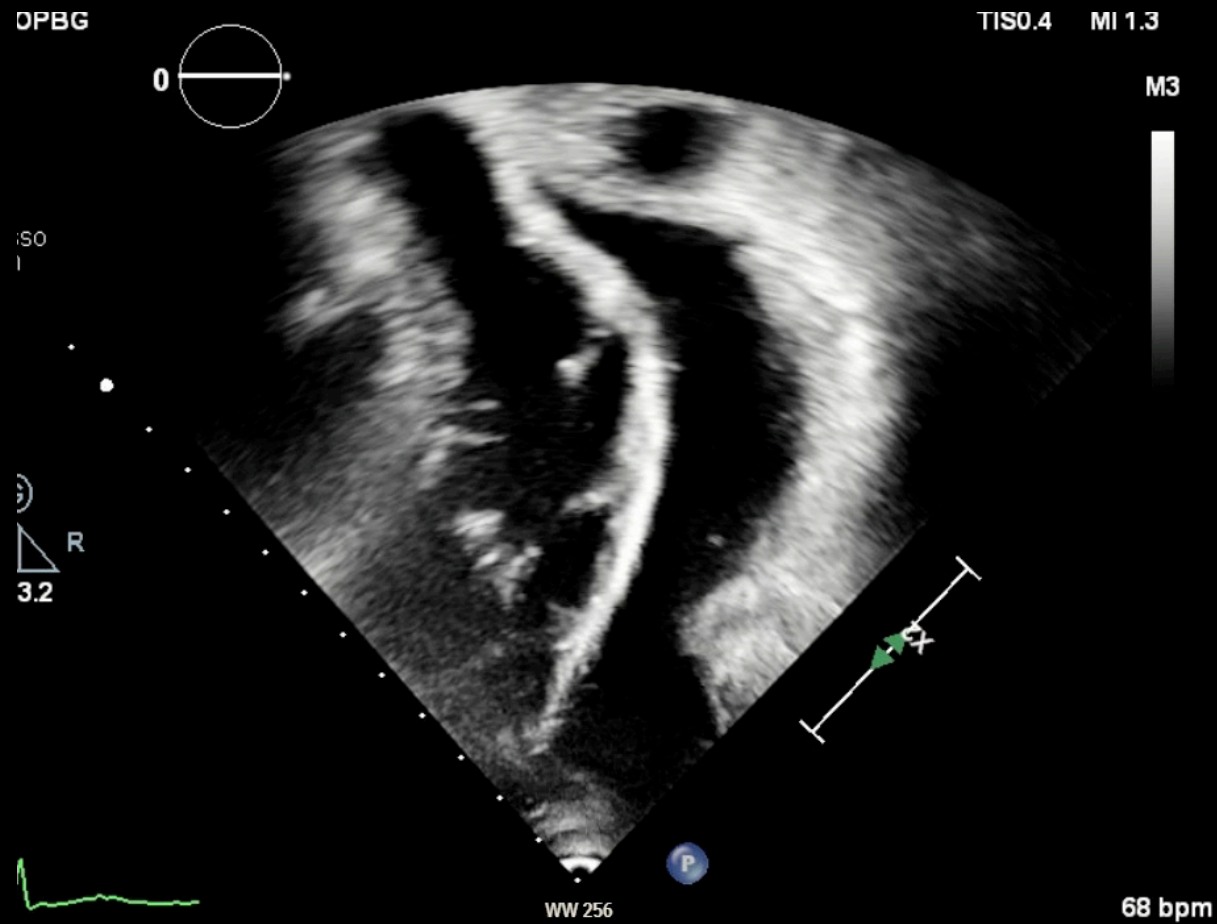
Deliver

Normal pregnancy course with cesarean delivery at 38 weeks of gestation (April 2021). Delivery without complications.

Discharged with:

- Clexane 4000 IU subcutaneously at 6:00 PM until 6 weeks postpartum, then start Cardioaspirin 100 mg on a full stomach.

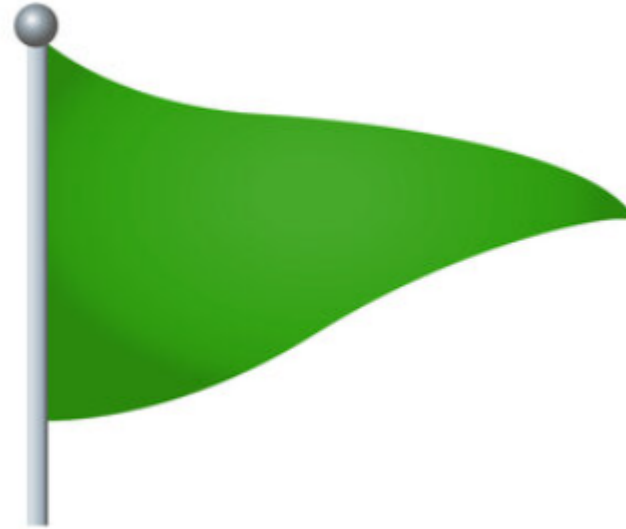
6° week post-delivery



When is the best time

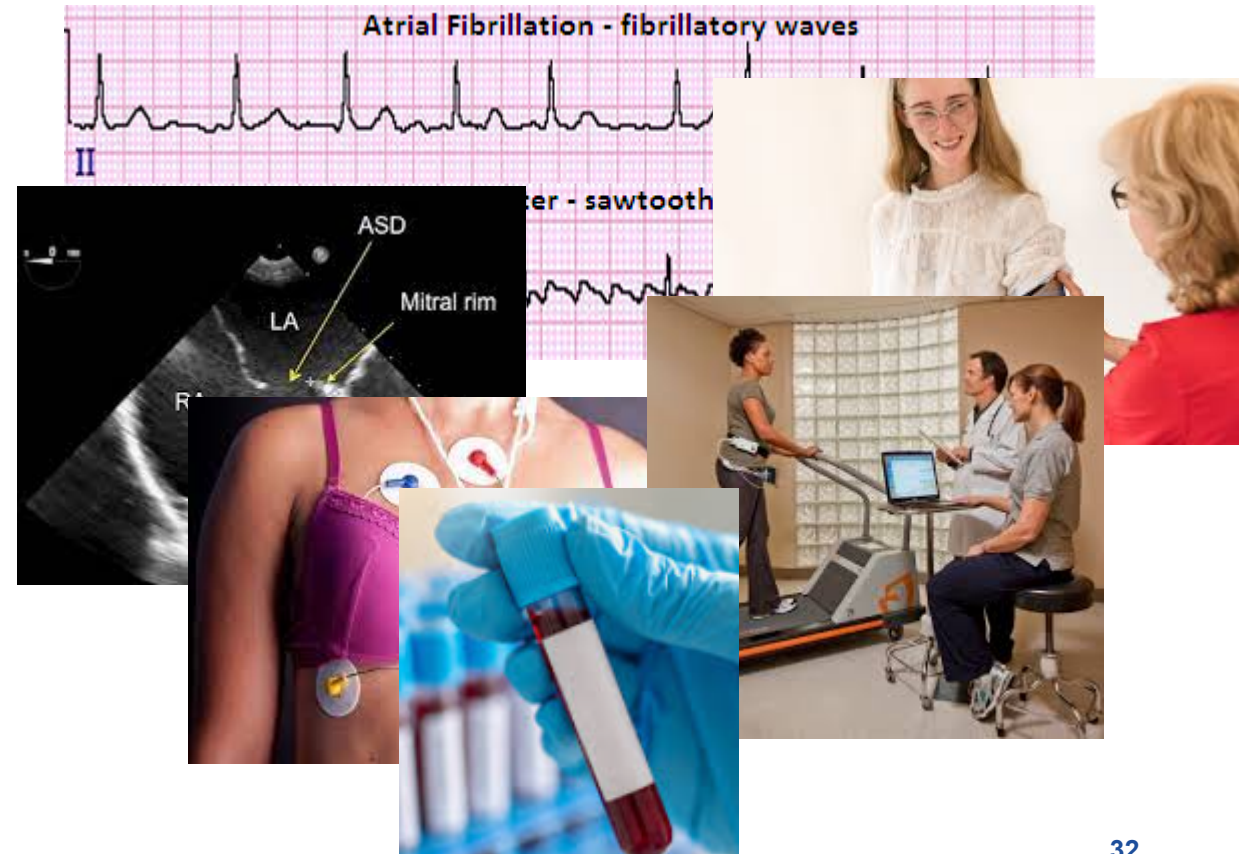
When is the best time

***THE SOONER THE
BETTER!!!***



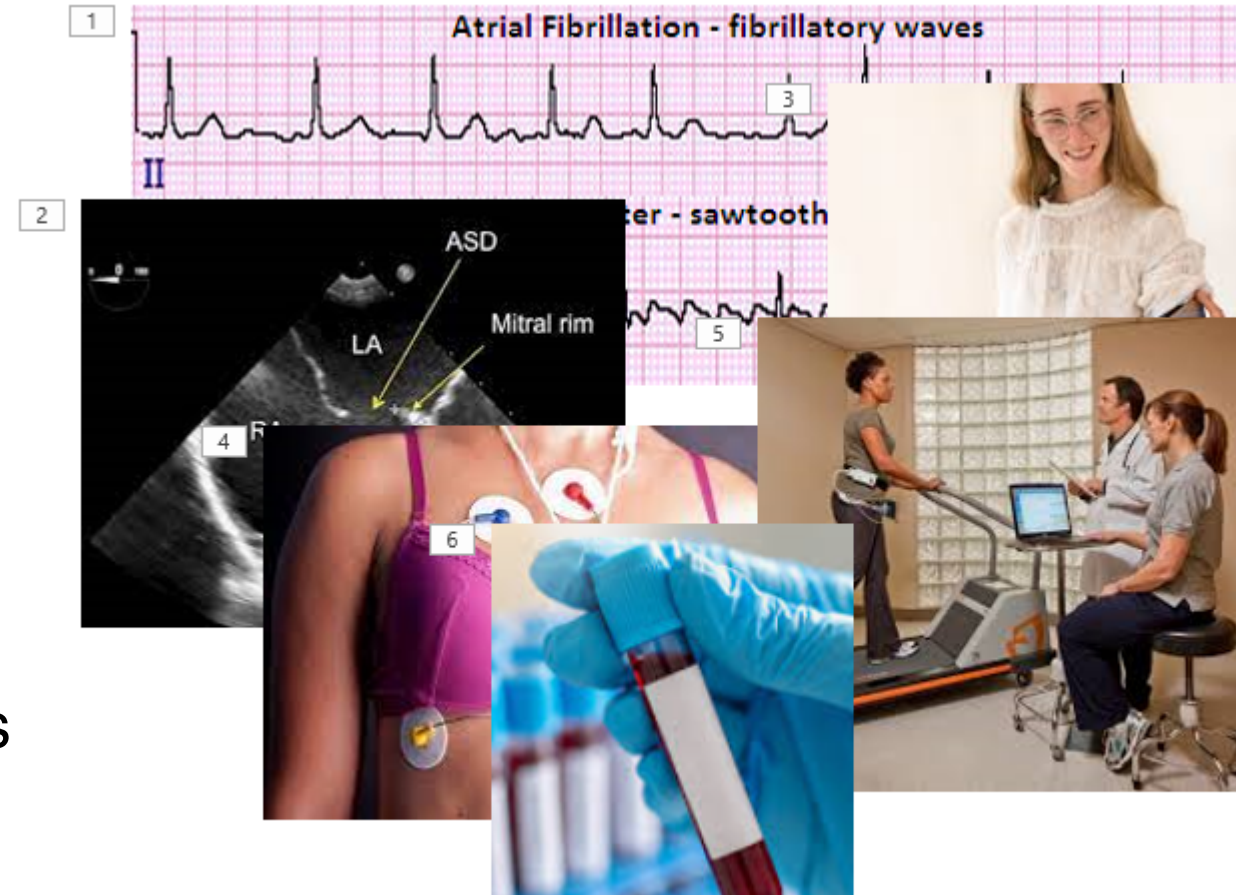
When is the best time

- Education during transition, starting at fertile age
- **Counselling** is very important to risk stratify (takes time!!!)
 - Detailed history
 - Address the risk factors
 - Investigations:
 - ECG (SR, AF, AT)
 - Echocardiogram
 - BP monitor
 - ECG monitor
 - Exercise test
 - Blood tests



When is the best time

- Education during transition, starting at fertile age
- **Counselling** is very important to risk stratify (takes time!!!)
 - Detailed history
 - Address the risk factors
 - Investigations:
 - ECG (SR, AF, AT)
 - Echocardiogram
 - BP monitor
 - ECG monitor
 - Exercise test
 - Blood tests
- **Counselling** is very important to increase patient awareness



Valve Disease

Outcomes of 613 pregnancies in women with a prosthetic valve



Registry of pregnancy and cardiac disease III

- Inclusions January 2018-April 2023
- Pregnancies from 59 centres in 27 countries
- Median follow-up 6.5 months postpartum



	Biological valve 202 pregnancies	Mechanical valve 411 pregnancies
Thromboembolic event	2% (n = 3)	9% (n = 34)
Haemorrhagic event	9% (n = 18)	20% (n = 76)
Maternal death	0%	1% (n = 4)
Foetal death	6% (n = 11)	27% (n = 109)

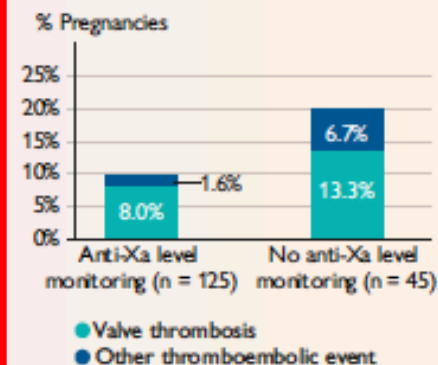
Predictor for valve thrombosis

Mitral valve prosthesis
(OR 3.3; 95% CI 1.9–8.0)

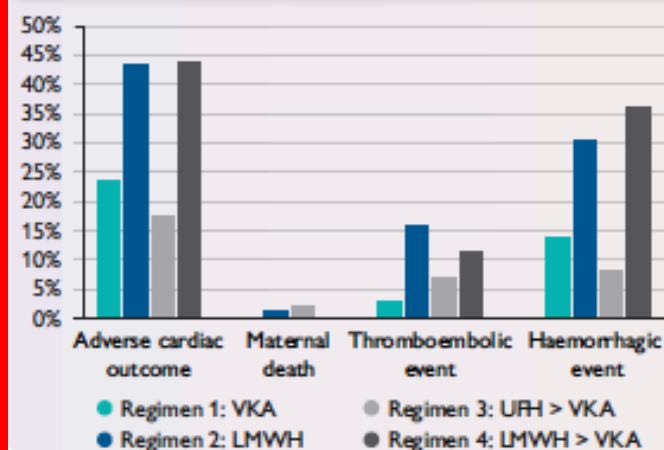
Predictor for any thromboembolic event

BMI
(OR 1.1; 95% CI 1.0–1.1)

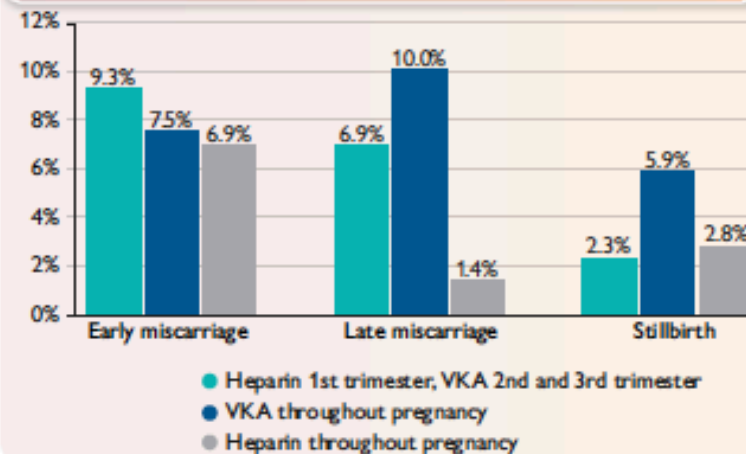
Anti-Xa level monitoring



Maternal outcomes

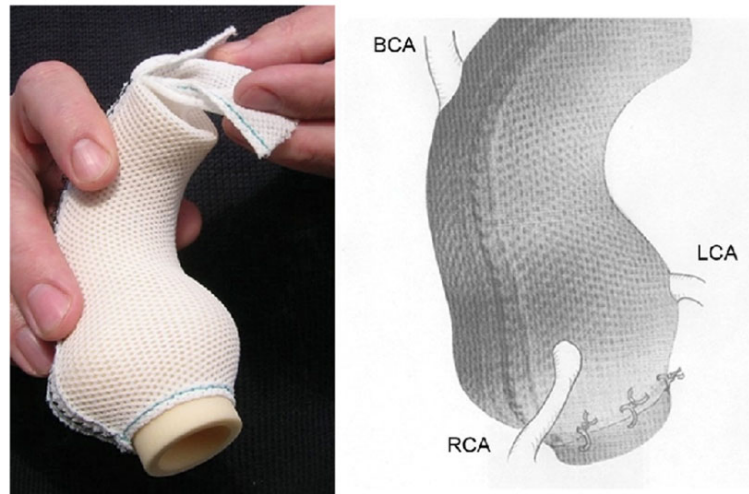


Foetal outcomes



Aortic dissection

Figure 1: On the left is the mesh sleeve on a former made by computer-aided design and rapid prototyping (3D printing). ...



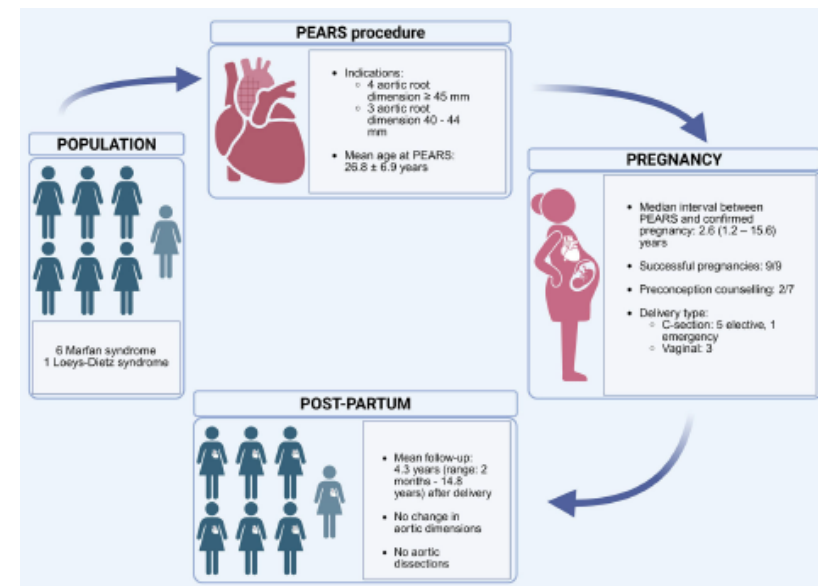
Connective tissue disorder and high risk pregnancy: a case series with personalised external aortic root support (PEARS)

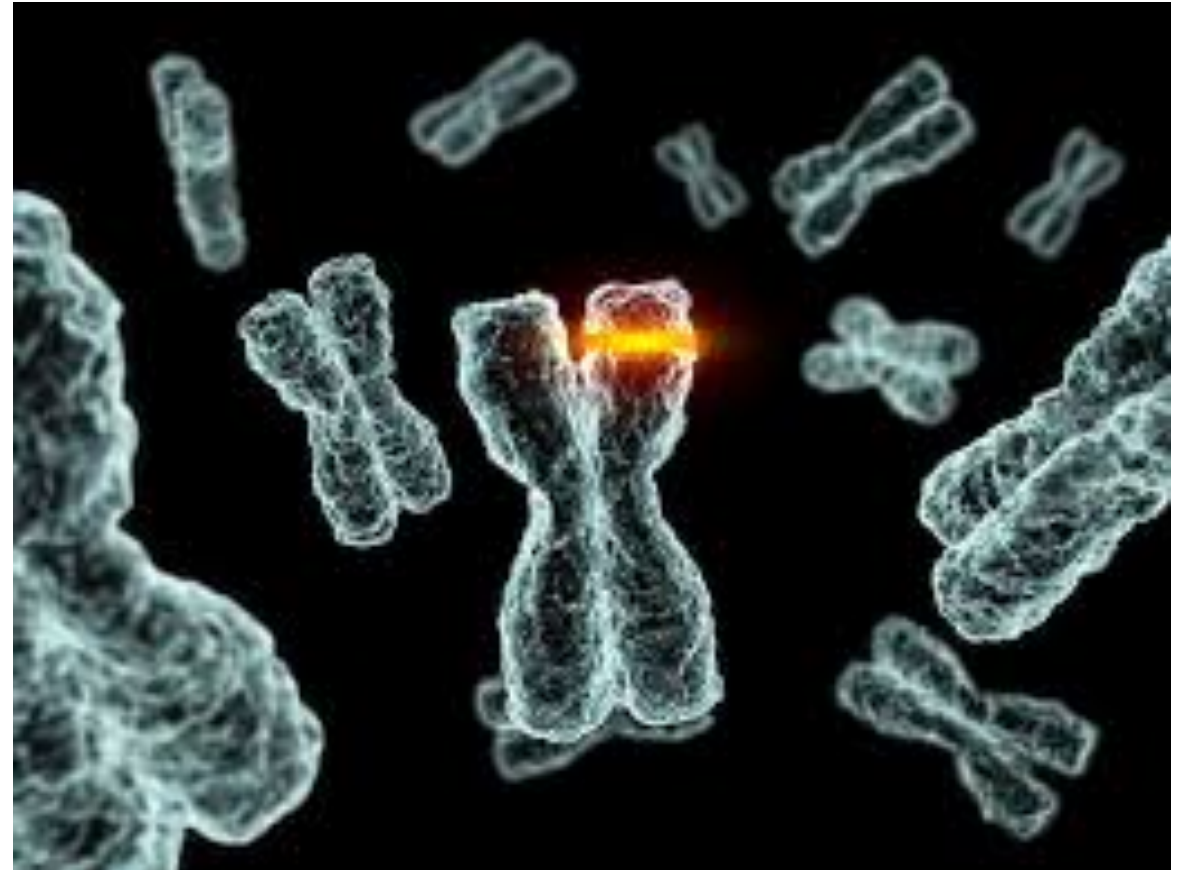
Received: 29 July 2024

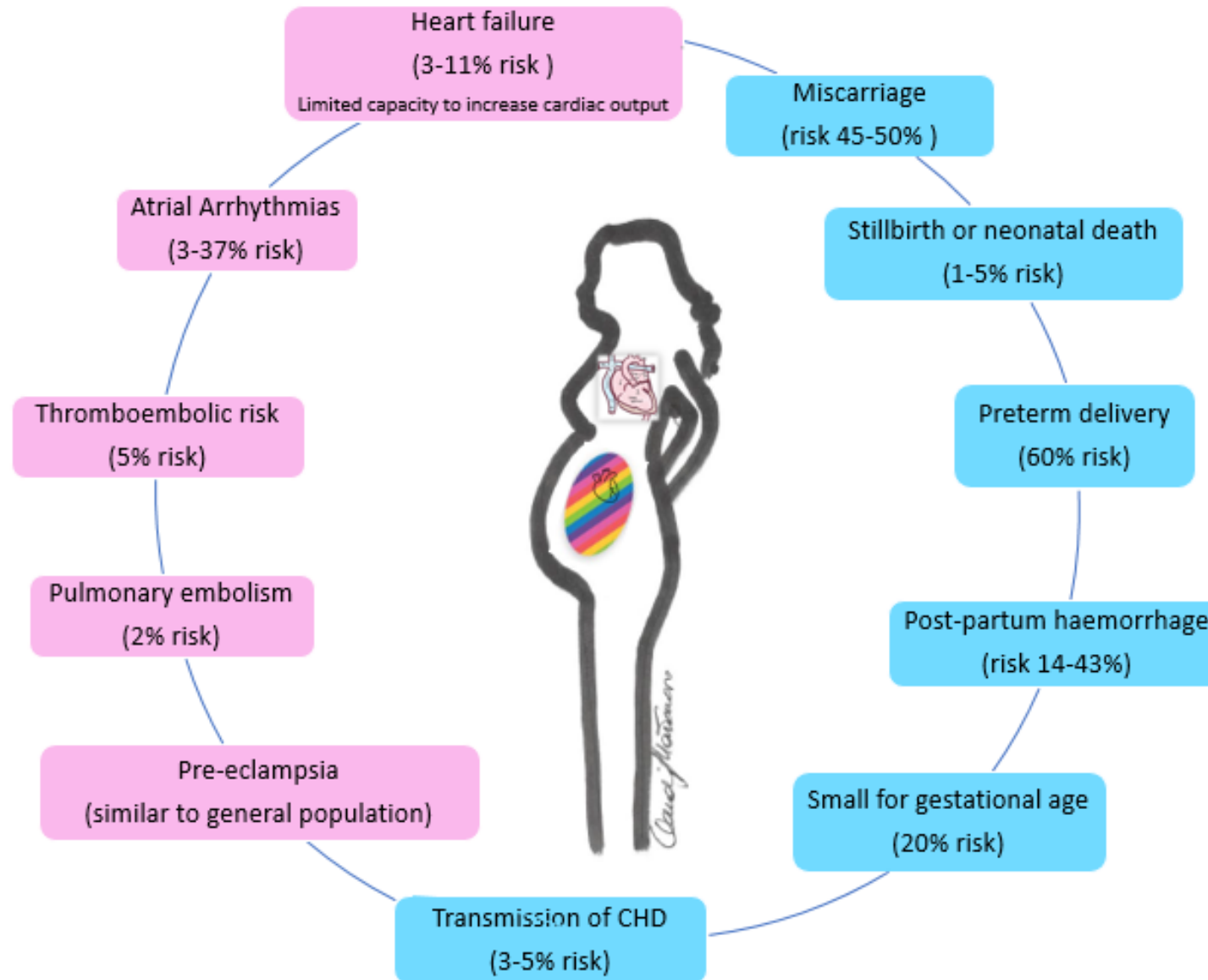
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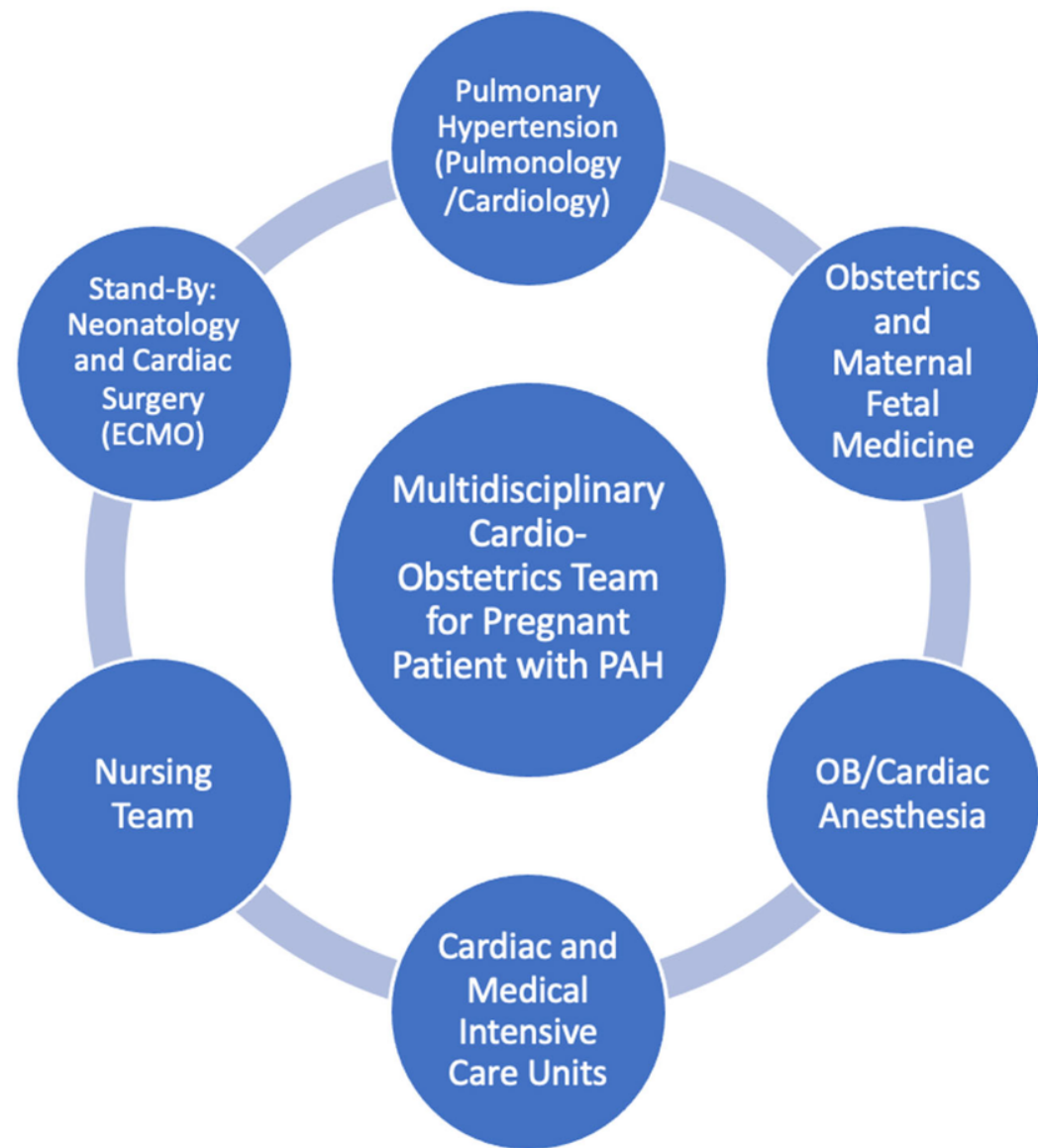
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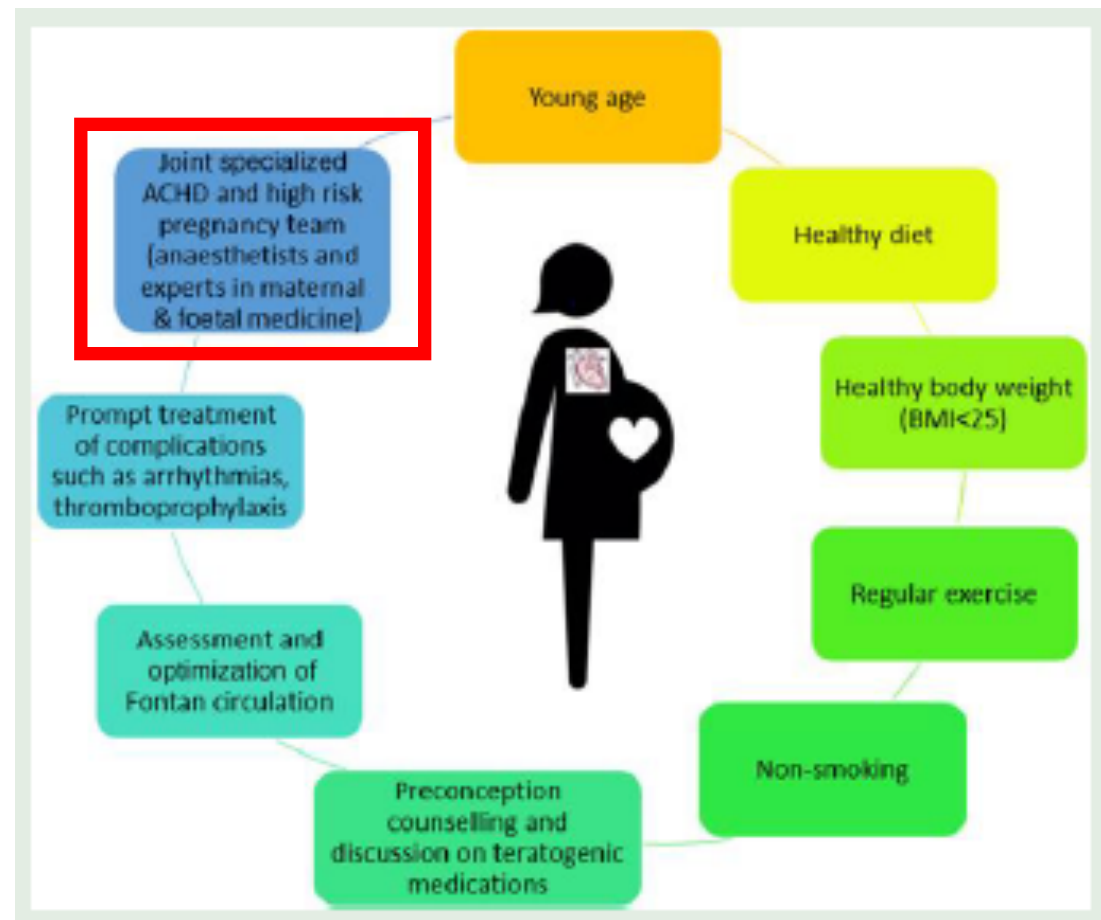






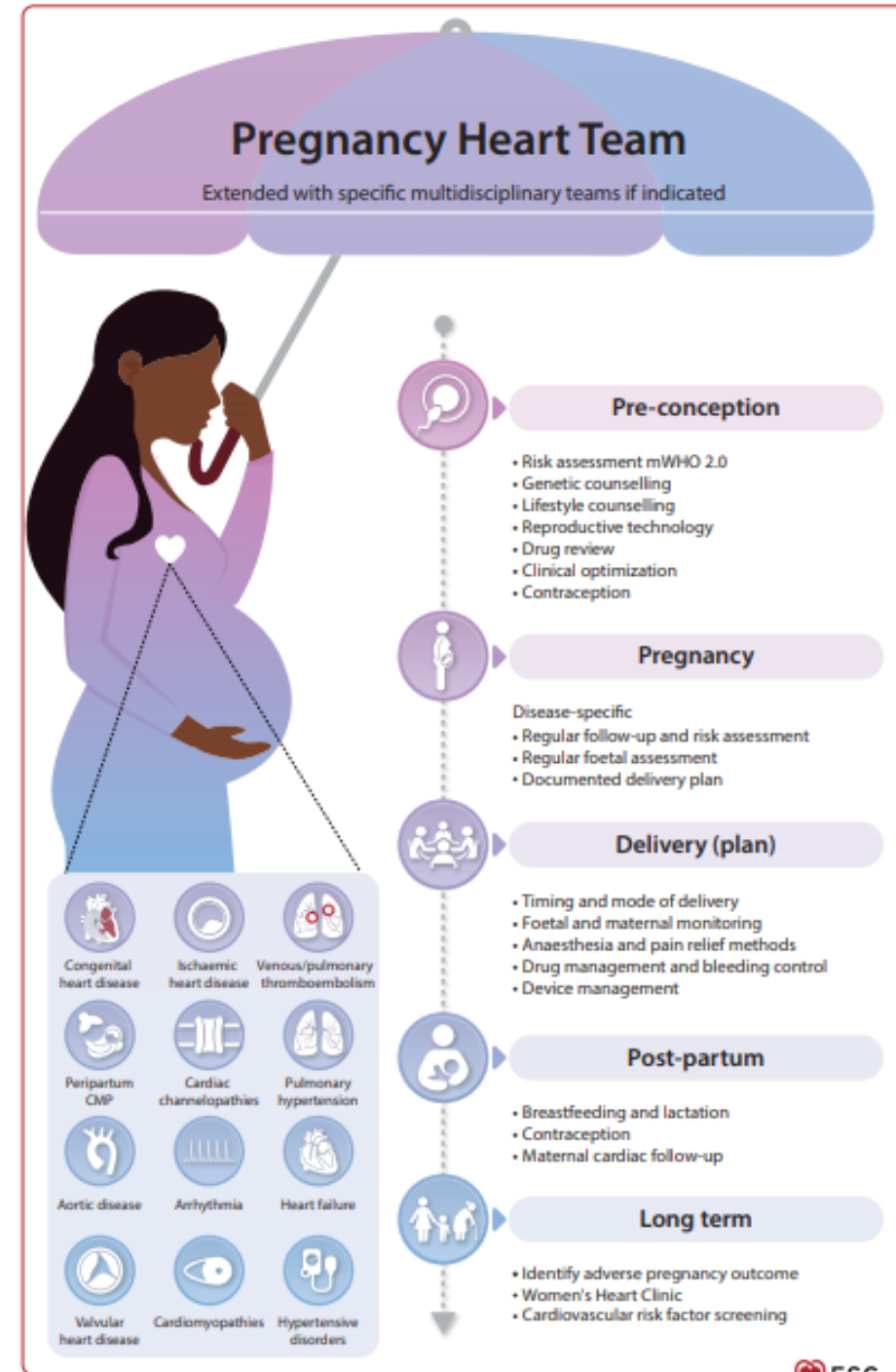
Pregnancy in patients with the Fontan operation

Claudia Montanaro ^{1,2,4†}, Siobhan Boyle ^{1,3†}, Gurleen Wander ⁴, Mark R. Johnson ^{2,4*}, Jolien W. Roos-Hesselink ⁵, Roshni Patel ^{2,4}, Isma Rafiq ¹, Candice K. Silversides ^{6‡}, and Michael A. Gatzoulis ^{1,2‡}



Where is the best place

- ACHD cardiologist
- Obstetrician with ACHD interest
- Intensivist with ACHD interest
- ACHD nurses



Where is the best place

Additional members are often involved depending on the case:

- **Cardiac surgeon** (if surgery might be needed)
- **Neonatologist** (for newborn care, especially if preterm or high-risk)
- **Electrophysiologist** (if there are arrhythmias)
- **Hematologist** (if anticoagulation or clotting issues are involved)

Neurologist

CONCLUSIONS

Why pregnancy in ACHD The ACHD is a growing population

What happens in pregnancy Hormonal changes

pre/-conception counselling

Who can deliver safely

ASAP

When is the best time

Heart Team

Where is the best place



Thank you for your attention!



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The **TRICUSPID VALVE** in **CONGENITAL** **HEART DISEASE:**

from **prenatal life**
to **adulthood**



SAVE THE DATE

October 16-17, 2026
Bambino Gesù Children's Hospital - Rome

