Preconception Planning and Peripartum Medical Management in Genetic Aortopathies

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Vascular Genetics Symposium

Outline

- Background and pregnancy physiology
- Risk assessment and preconception counseling
- Aortopathy management during pregnancy
- Delivery recommendations

I have no disclosures

DISCLAIMER: I am not an obstetrician (just a cardiologist who has learned a lot from them)

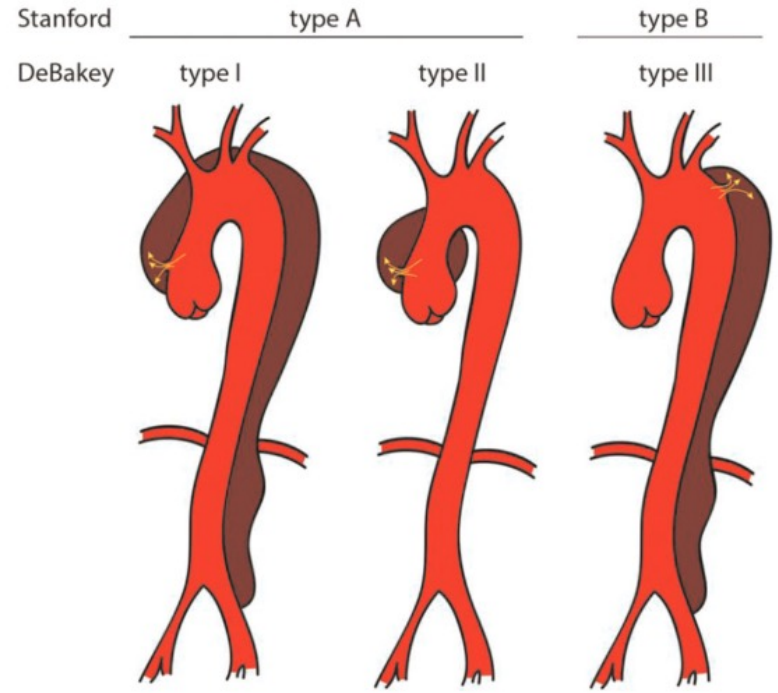
Dissection risk

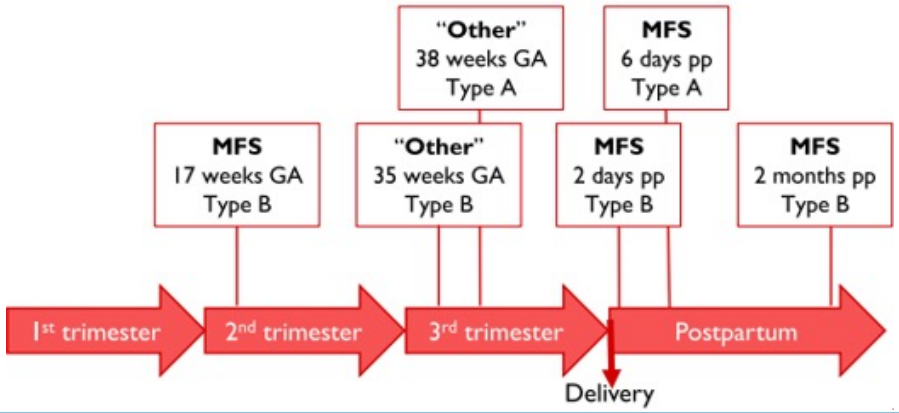
Pregnancy is a risk factor for aortic dissection in women with aortopathy due to hemodynamic and hormonal changes

ROPAC III: Pregnancy-related aortic dissection rate in women with known aortopathy diagnosis prior to pregnancy, under surveillance in specialized clinics, was ~3.5%

Dissection may occur throughout pregnancy or up to 12 weeks postpartum; most commonly in the 3rd trimester or early post-partum

Dissection risk is associated with type of mutation, size of aorta, HTN, rapid aortic growth

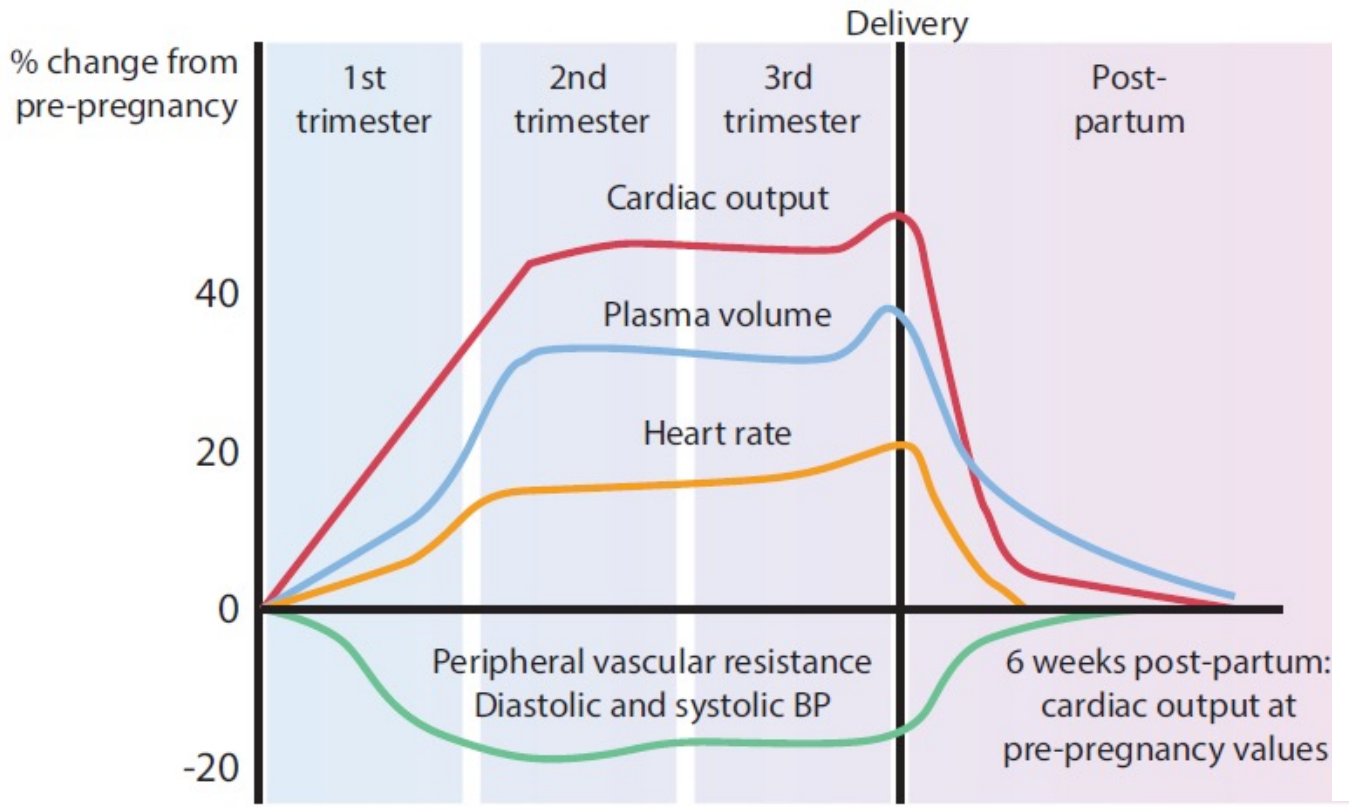




Peters et al, on behalf of ROPAC. Pregnancy outcomes in women with heritable thoracic aortic disease. *EHJ-QCCO*,2025

Physiologic changes in pregnancy

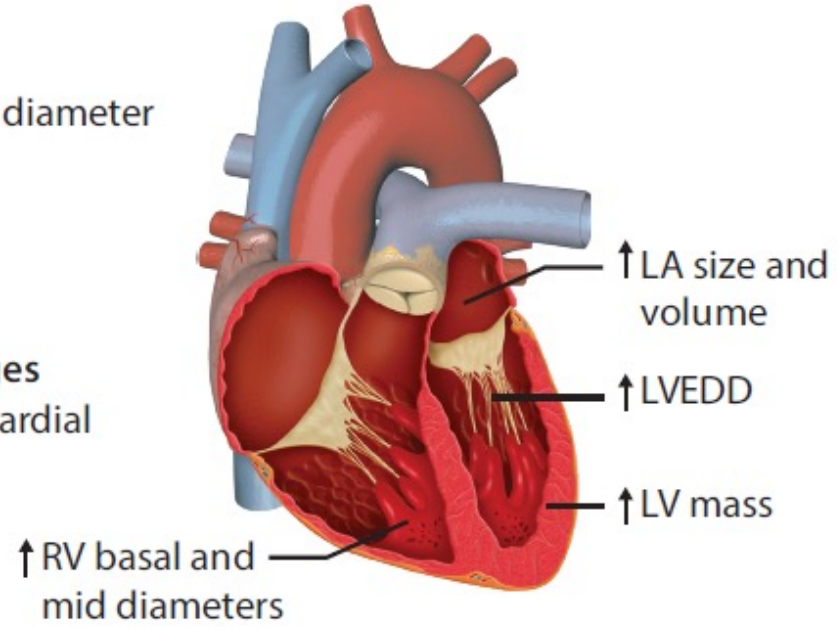
Haemodynamic changes



Echocardiographic changes

Unchanged
- Aortic root diameter
- LVEF
- RVEF
- SPAP

Small changes
- Small pericardial effusion

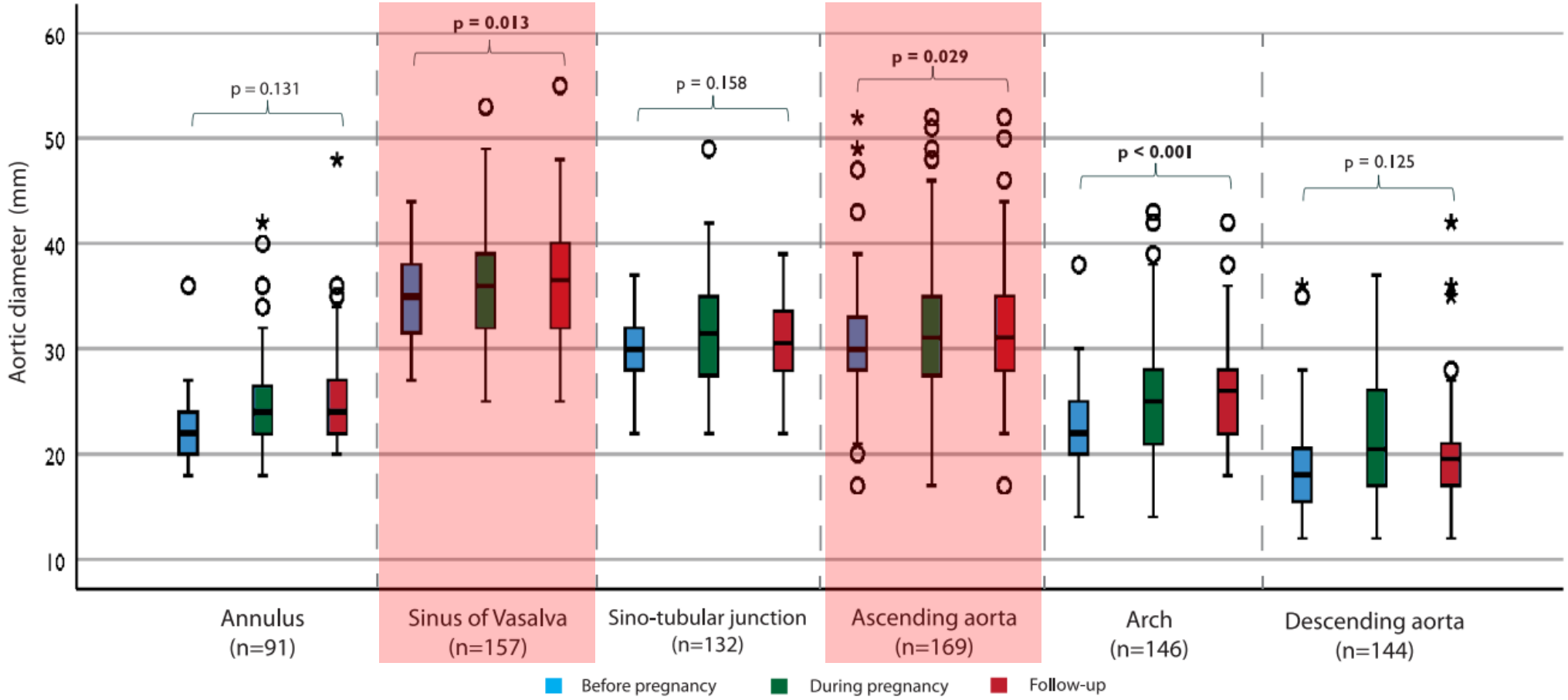


De Backer et al. 2025 ESC guidelines for the management of cardiovascular disease and pregnancy. EHJ, 2025.

Hemodynamic changes in pregnancy

↓ Systemic vascular resistance (~30%)	↓ Blood pressure (~10 mmHg)	↓ Pulmonary vascular resistance (~24%) and ↑ pulmonary blood flow	↑ Cardiac output (30-50%); plateaus at ~24 weeks gestation
↑ HR (10-20 bpm)	↑ Risk of arrhythmia	↑ Tidal volume and minute ventilation	↑ Risk of blood clots
	↑ GFR	Remodeling of the vascular tree	

Changes in aortic diameter during pregnancy in HTAD



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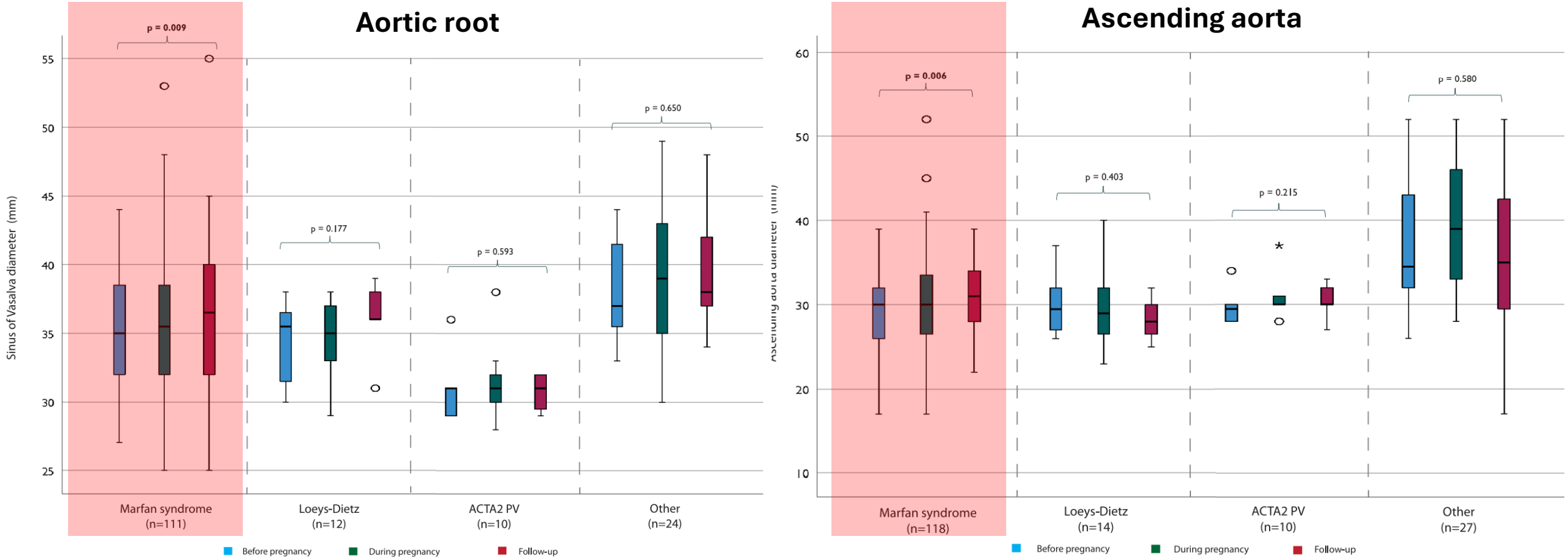
Aortic size based on diagnosis group



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Multidisciplinary cardio-obstetric care from preconception through postpartum decreases adverse cardiac events

Davis et al. JACC 2021; 77(14):1763-77

Preconception counseling for all women of childbearing age with a known CV diagnosis

Maternal and fetal risk stratification

Counselling on the risk of dissection and the recurrence risk

Imaging of the entire aorta before pregnancy

Optimizing cardiac lesions

Discussing pre-pregnancy interventions

Reviewing safety of medications in pregnancy; safe titration off ARB (losartan, etc)

Preconception genetic counseling

- Autosomal dominant inheritance
- Variable expressivity
- Preimplantation genetic diagnosis
- Prenatal testing

Making a detailed plan for the pregnancy, labor, and delivery

Risk prediction tools help assess the risk of maternal cardiac events during pregnancy

mWHO

- Provides a first impression about the potential risk of pregnancy, based on expert opinion
- Stratification is based on the underlying diagnoses and may also give direction as to who should be referred immediately, and who may be evaluated in non-tertiary centres

CARPREG II

- Derived from, and validated in, a large Canadian cohort of women with heart disease in general, of whom 64% had congenital heart disease

Risk stratification tools should always be supplemented by lesion-specific data and clinical judgement

		mWHO 2.0 I	mWHO 2.0 II	mWHO 2.0 II–III	mWHO 2.0 III	mWHO 2.0 IV
		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.
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 • No prior cardiac intervention indicated • Late pregnancy assessment			**CARPREG II score: 2 points** • Ventricular dysfunction • High-risk left-sided valve disease or outflow tract obstruction • Pulmonary hypertension • Coronary artery disease • High-risk aortopathy		**CARPREG II score: 3 points** • Prior cardiac event or arrhythmias • Baseline NYHA III/IV or cyanosis • Mechanical valve	

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Pregnancy considerations by diagnosis

	Marfan	Loeys-Dietz	Vascular Ehlers Danlos	BAV	Turner
Dissection risk	~1% if aorta <4 cm ~10% if aorta >4 cm	High, depending on specific type ~1-19%	High Maternal mortality rate 4-25%	Low, <1%	High, 1-10%
Other risks	Mitral regurgitation Heart failure Arrhythmias Dural abnormalities	Mitral regurgitation Heart failure Arrhythmias Bleeding Uterine rupture	Uterine rupture PROM Bleeding Dural abnormalities Dissections of other arteries	Aortic stenosis or regurgitation	HTN Diabetes
Caution with pregnancy	Aorta >4.5 cm	Aorta >4.5 cm (or >4 cm if FH of dissection)	All patients	Aorta>5 cm	Aortic size index >2.5 cm/m2
Prophylactic surgery	Aorta >4.5 cm Moderate-severe AI Rapid dilation (>2-3 mm/yr)	Aorta ≥4.5 cm (class I recommendation for TGFBR1 and TGFBR2; IIa for SMAD3, TGFB2, TGFB3, SMAD2)	No well-established guidelines, >4.0 cm reasonable	Aorta>5.0 cm	Aortic size index >2.5 cm/m2

Management during pregnancy

Managed by a multidisciplinary cardio-OB team

Strict BP control during pregnancy

- **Goal ~120/80**

Echocardiogram every 4–12 weeks for moderate to high risk disease

- At least once each trimester; more frequent depending on aortic diameter, aortic growth rate, and underlying condition

Beta blocker therapy throughout pregnancy and PP

Monitoring for red flags

- Acute chest or back pain; new neurologic event
- Progressive aortic dilation
- Hypertensive disorders of pregnancy

Delivery recommendations

Vaginal delivery

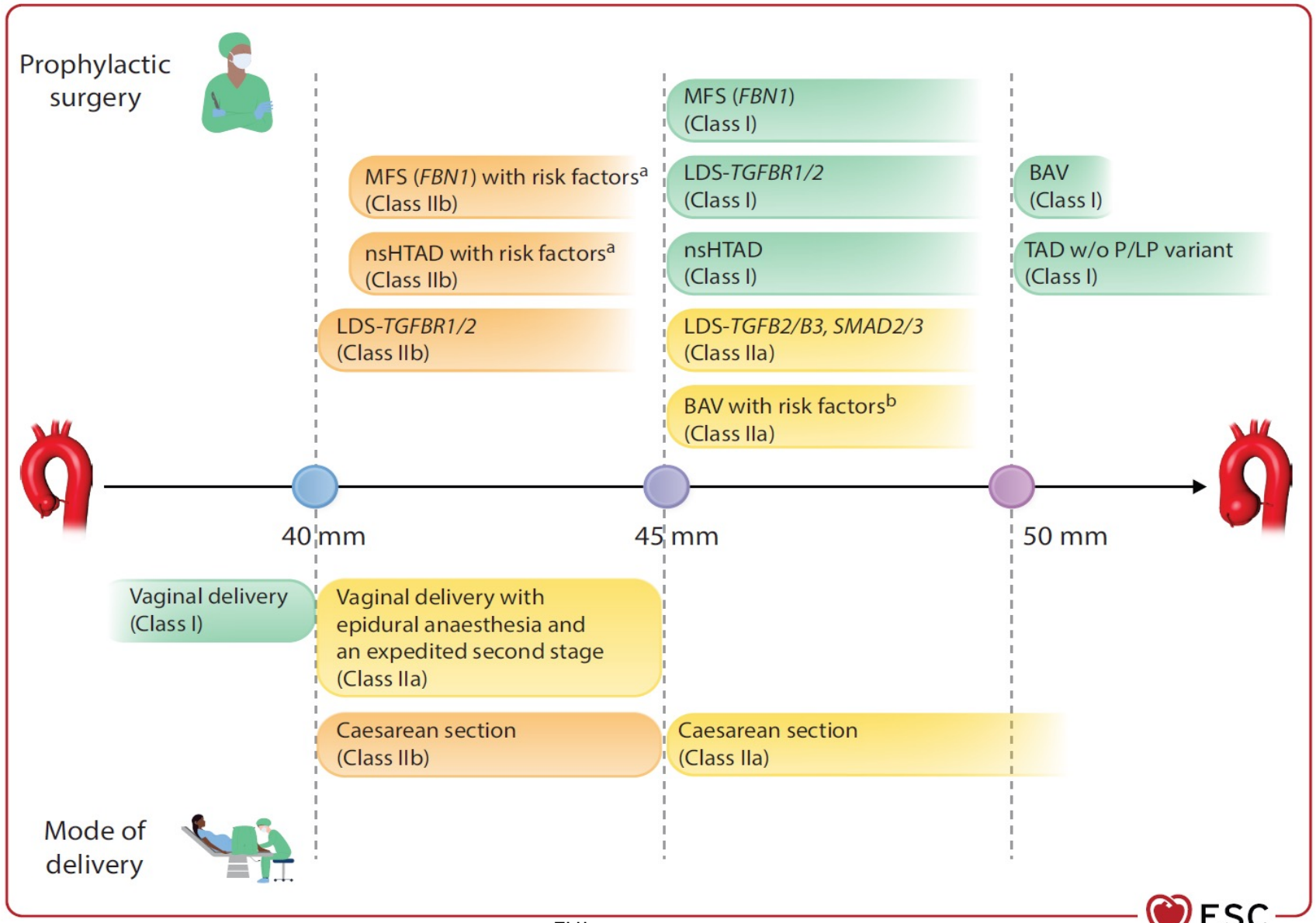
- Recommended if aorta **<4.0 cm**

Vaginal delivery with epidural anesthesia and an expedited/assisted second stage

- Reasonable if aorta **4.0-4.5 cm**

Caesarian delivery

- **Prior aortic dissection**
- **VEDS, at 37 weeks**
- Reasonable if ascending aorta **>4.5 cm**



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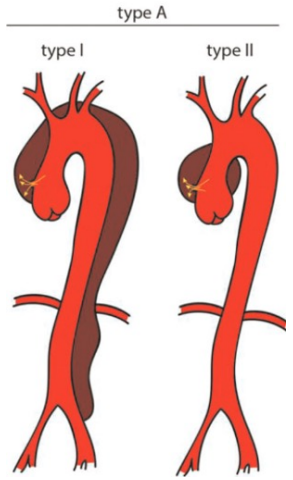
Aortic dissection during pregnancy



Acute type A dissection

Prior to viability:
Repair with pregnancy in situ

>28 weeks GA:
C-section delivery followed by urgent repair

Viability-28 weeks:
Shared decision making, individualized management

Acute type B dissection

Medical management preferred

Endovascular or surgical intervention if acute complications (malperfusion)

Timing of Cardiac Surgery during Pregnancy

A patient-level meta-analysis

Pregnant patients who underwent cardiac surgery	1st trimester*	2nd trimester*	3rd trimester*	C-section prior to surgery
386	58	182	146	120
Maternal mortality 7.3%	12.1%	4.6%	8.8% P= 0.089	8.3% OR=1.40 (0.57 - 3.43)
Foetal mortality 26.5% *Excl. CS patients	44.8%	33.9%	29.4% P= 0.233	6.7% OR=0.11 (0.05 - 0.25)

- C-section prior to surgery may be considered
- If not feasible or safe, trimester stage should not delay surgery

Vascular Genetics Symposium

Eur Heart J, Volume 43, Issue 29, 1 August 2022, Pages 2801–2811, https://doi.org/10.1093/eurheartj/ehac234

Summary

Pregnancy is a risk factor for aortic dissection

Risk of type A dissection is related to aortic size; type B dissection can occur at normal aortic dimensions

Preconception risk assessment and avoiding an unplanned pregnancy are essential

Management in multidisciplinary cardio-obstetric program recommended

Regular aortic imaging throughout pregnancy is recommended

Delivery recommendations depend on aortic size and genetic mutation

THANK YOU