

2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy

Dr Nadjib HAMMOUDI

Institut de cardiologie
Hôpital de la Pitié-Salpêtrière. Paris

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The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC).

Endorsed by: the International Society of Gender medicine (IGM), the German Institute of Gender in Medicine (DGesGM), the European Society of Anaesthesiology (ESA), and the European Society of Gynecology (ESG).

Authors/Task Force Members: Vera Regitz-Zagrosek (Chairperson) (Germany), Jolien W. Roos-Hesselink (Co-Chairperson) (The Netherlands), Johann Bauersachs (Germany), Carina Blomström-Lundqvist (Sweden), Renata Cífková (Czech Republic), Michele De Bonis (Italy), Bernard Jung (France), Mark R. Johnson (UK), Ulrich Kintscher (Germany), Peter Kranke (Germany), Irene Marthe Lang (Austria), Joao Morais (Portugal), Petronella G. Pieper (The Netherlands), Patrizia Presbitero (Italy), Susanna Price (UK), Giuseppe M. C. Rosano (UK/Italy), Ute Seeland (Germany), Tommaso Simoncini (Italy), Lorna Swan (UK), Carole A. Warnes (USA).

Table 1 Classes of recommendations

Classes of recommendations	Definition	Suggested wording to use
Class I	Evidence and/or general agreement that a given treatment or procedure is beneficial, useful, effective.	Is recommended/ is indicated.
Class II	Conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of the given treatment or procedure.	
<i>Class IIa</i>	<i>Weight of evidence/opinion is in favour of usefulness/efficacy.</i>	Should be considered.
<i>Class IIb</i>	<i>Usefulness/efficacy is less well established by evidence/opinion.</i>	May be considered.
Class III	Evidence or general agreement that the given treatment or procedure is not useful/effective, and in some cases may be harmful.	Is not recommended.

Table 2 Levels of evidence

Level of evidence A	Data derived from multiple randomized clinical trials or meta-analyses.
Level of evidence B	Data derived from a single randomized clinical trial or large non-randomized studies.
Level of evidence C	Consensus of opinion of the experts and/or small studies, retrospective studies, registries.

Introduction

- Pays à haut revenus
 - maladies CV = cause principale de mortalité maternelle
 - augmentation de l'âge maternel,
 - meilleure prise en charge des cardiopathies (notamment congénitales)
- Autres pays
 - valvulopathies rhumatismales ++

Adaptation CV pendant la grossesse

- **Augmentation du débit cardiaque** (+50% par rapport à l'état basal)

- Augmentation du VES (élévation de la pré-charge, baisse de la post-charge, remodelage VG et OG)
- Augmentation de la FC

>> Déstabilisation d'une maladie cardiaque latente

- **Etat procoagulant**

>> Elévation du risque thrombo-embolique

- **Modification de la pharmacocinétique des médicaments (HBPM...)**

- **Contre indication de certains médicaments** (Insuffisance cardiaque..)

- **Accouchement** (travail, anesthésie, risque hémorragique)

Information de la patiente bilan avant la conception

- **Evaluer le risque maternel et fœtal** d'une grossesse
 - ECG / échocardiographie
 - **Epreuve d'effort**
 - imagerie de coupe de l'ensemble de l'aorte** en cas de pathologie de l'aorte.
- **Informar la patiente** du risque et/ou de la contre indication à la grossesse
- **Proposer un conseil génétique**
 - maladie autosomiques dominante: QT long, Marfan....

Evaluation du risque maternel

- Cardiopathie:
 - sévérité et type de la valvulopathie (Sténose >> régurgitation)
 - fonction VG,
 - niveau de pression pulmonaire,
 - arythmies,
 - taille de l'aorte
 - Cardiopathies congénitales (HTAP, cyanose, dysfonction du ventricule systémique)
- Comorbidités
- Statut fonctionnel / capacité à l'exercice
- Antécédents personnels et familiaux de syndrome aortique

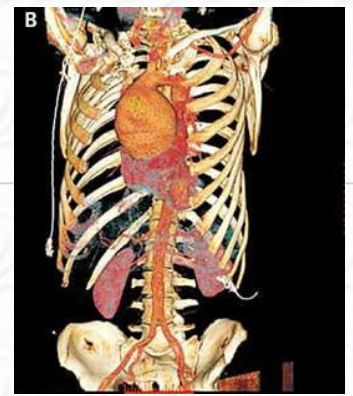
Table 3 Modified World Health Organization classification of maternal cardiovascular risk

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Counselling	Yes	Yes	Yes	Yes: expert counselling required	Yes: pregnancy contraindicated: if pregnancy occurs, termination should be discussed
Care during pregnancy	Local hospital	Local hospital	Referral hospital	Expert centre for pregnancy and cardiac disease	Expert centre for pregnancy and cardiac disease
Minimal follow-up visits during pregnancy	Once or twice	Once per trimester	Bimonthly	Monthly or bimonthly	Monthly
Location of delivery	Local hospital	Local hospital	Referral hospital	Expert centre for pregnancy and cardiac disease	Expert centre for pregnancy and cardiac disease
				valve, Turner syndrome AS 20–25 mm/m ² , tetralogy of Fallot <50 mm) Ventricular tachycardia	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
Maternal cardiac event rate	2.5–5%	5.7–10.5%	10–19%	19–27%	40–100%

3.3.3 Pregnancy heart team

In women with a moderate or high-risk of complications during pregnancy (mWHO II–III, III, and IV), pre-pregnancy counselling and management during pregnancy and around delivery should be conducted in an expert centre by a multidisciplinary team: the pregnancy heart team. The minimum team requirements are a cardiologist, obstetrician, and anaesthetist, all with expertise in the management of high-risk pregnancies in women with heart disease. Additional experts

Pathologie aortique



- Origine génétique: Marfan, Loeys–Dietz, Ehlers–Danlos
- Non génétique

>>Risque de dissection

>>Troisième trimestre et post-partum(suivi à 6mois)

Evaluation du risque (AVANT LA GROSSESSE)

- **Dans quel syndrome?**

- Marfan (CI si Aorte >45mm)

- Ehlers–Danlos de type IV (très haut risque quelque soit la taille de l'aorte)

- **Quelle est la taille de l'aorte (indexation surface corporelle, >25 mm/m²)**

- >> Imagerie totale de l'aorte

- **Taux de progression de la dilatation**

- **Histoire personnelle ou familiale de Sd aortique**

Table 8 Aortic diseases (2)

	Marfan	Bicuspid aortic valve	LoeysDietz	Turner	Vascular Ehlers-Danlos
Advise not to become pregnant	Ascending aorta >45 mm (or >40 mm in family history of dissection or sudden death)	Ascending aorta >50 mm	Ascending aorta >45 mm (or >40mm in family history of dissection or sudden death)	ASI >25 mm/m ²	All patients

Table 9 Management of aortic disease (1)

Recommendations	Class	Level
All aortic diseases		
It is recommended that women with aortic disease have counselling about the risk of aortic dissection.	I	C
Imaging of the entire aorta (CT/MRI) is recommended before pregnancy in patients with a genetically proven aortic syndrome or known aortic disease.	I	C
In bicuspid aortic valve patients, imaging of the ascending aorta is recommended before pregnancy.	I	C
When a woman with known aortic dilatation (history of) dissection or genetic predisposition for dissection becomes pregnant, strict blood pressure control is recommended.	I	C

Table 9 Management of aortic disease (2)

Recommendations	Class	Level
Repeated echocardiographic imaging every 4–12 weeks (depending on diagnosis and severity of dilatation) is recommended during pregnancy and 6 months post-partum in patients with ascending aorta dilatation.	I	C
For imaging of pregnant women with dilatation of the distal ascending aorta, aortic arch, or descending aorta, MRI (without gadolinium) is recommended.	I	C
It is recommended to deliver all women with aortic dilatation or (history of) aortic dissection in an experienced centre with a pregnancy heart team, where cardiothoracic surgery is available.	I	C
In patients with an ascending aorta <40 mm , vaginal delivery is recommended.	I	C

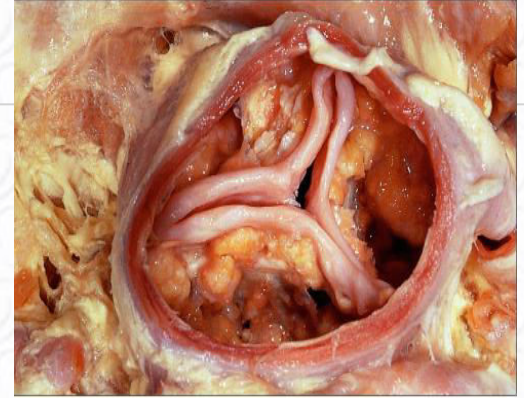
Table 9 Management of aortic disease (3)

Recommendations	Class	Level
In patients with an ascending aorta >45 mm, caesarean delivery should be considered.	Ila	C
In patients with (history of) aortic dissection, caesarean delivery should be considered.	Ila	C
Prophylactic surgery should be considered during pregnancy if the aorta diameter is >45 mm and increasing rapidly.	Ila	C
When the foetus is viable, delivery before necessary surgery should be considered.	Ila	C
In patients with an aorta 40–45 mm, vaginal delivery with epidural anaesthesia and an expedited second stage should be considered.	Ila	C

Table 9 Management of aortic disease (4)

Recommendations	Class	Level
Specific syndromes		
In patients with vascular Ehlers–Danlos syndrome, celiprolol is recommended.	I	C
Beta-blocker therapy throughout pregnancy should be considered in women with Marfan syndrome and other heritable thoracic aortic diseases.	IIa	C
Pregnancy is not recommended in patients with severe dilatation of the aorta (heritable thoracic aortic disease such as Marfan syndrome >45 mm, bicuspid aortic valve >50 mm or >27 mm/m ² BSA, or Turner syndrome ASI >25 mm/m ² BSA).	III	C
Pregnancy is not recommended in patients with vascular Ehlers–Danlos syndrome.	III	C

Valvulopathies



Evaluation du risque (AVANT LA GROSSESSE)

● Quelle valvulopathie / quelle sévérité?

- Sténoses >> régurgitations
- RM > RA

● Fonction VG / pression pulmonaire

● Retentissement fonctionnel (NYHA / test à l'effort)

- Anatomie favorable à un geste percutané (Sténose mitrale++)

Table 10 Management of native valvular heart disease (1)

Recommendations	Class	Level
Pre-pregnancy evaluation, including echocardiography, and counselling is recommended for any woman with known or suspected valvular disease.	I	C
Mitral stenosis		
In patients with symptoms or pulmonary hypertension, restricted activities and beta-1-selective blockers are recommended.	I	B
Diuretics are recommended when congestive symptoms persist despite beta-blockers.	I	B
Intervention is recommended <u>before pregnancy</u> in patients with MS and valve area <u><1.0 cm²</u> .	I	C
Therapeutic anticoagulation using heparins or VKA is recommended in case of atrial fibrillation, left atrial thrombosis, or prior embolism.	I	C

Table 10 Management of native valvular heart disease (2)

Recommendations	Class	Level
Intervention should be considered <u>before pregnancy</u> in patients with MS and valve area <1.5 cm² .	Ila	C
Percutaneous mitral commissurotomy should be considered <u>in pregnant</u> patients with severe symptoms or systolic pulmonary artery pressure >50 mmHg despite medical therapy .	Ila	C
Aortic stenosis		
Intervention is recommended before pregnancy in patients with severe aortic stenosis if:		
• they are symptomatic	I	B
• OR LV dysfunction (LVEF <50%) is present	I	C
• OR when they develop symptoms during exercise testing	I	C

Table 10 Management of native valvular heart disease (3)

Recommendations	Class	Level
Intervention should be considered before pregnancy in asymptomatic patients with severe AS when a fall in blood pressure below baseline during exercise testing occurs.	Ila	C
Balloon aortic valvuloplasty should be considered during pregnancy in patients with severe aortic stenosis and severe symptoms.	Ila	C
Chronic regurgitant lesions		
Surgical treatment is recommended before pregnancy in patients with severe aortic or mitral regurgitation <u>with symptoms of impaired ventricular function</u> or ventricular dilatation.	I	C
Medical therapy is recommended in pregnant women with regurgitant lesions when symptoms occur.	I	C

Gestion du traitement anticoagulant

Valve mécanique

- Risque de thrombose de valve

-AVK 0 à 4%

-HBPM 4 à 9%

- Risque d'embryopathie

-AVK 0.6 à 10% (mais <2% si faible dose)

-HBPM aucun

Figure 3 Flowchart on anticoagulation in mechanical valves and low-dose VKA

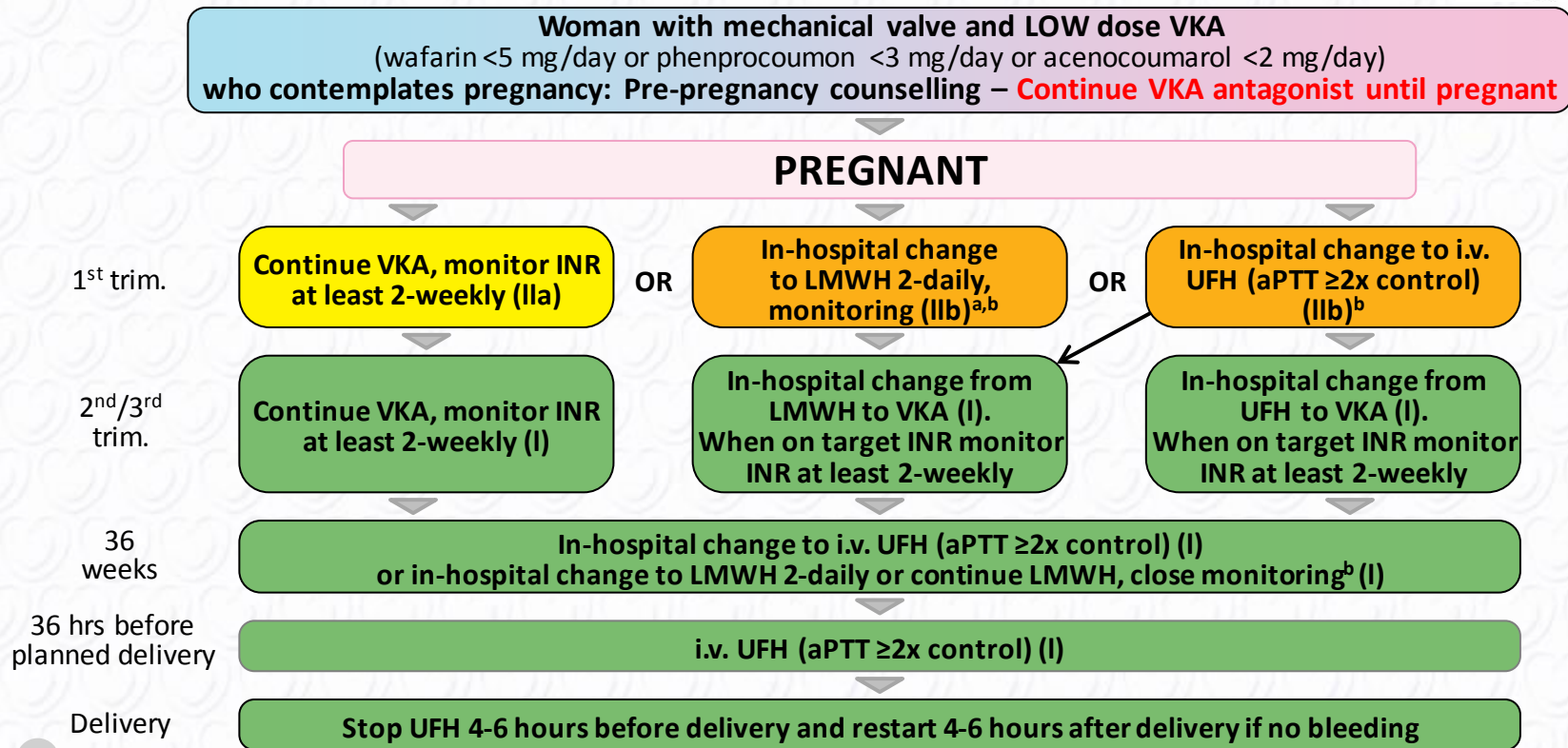


Figure 2 Flowchart on anticoagulation in mechanical valves and high-dose VKA

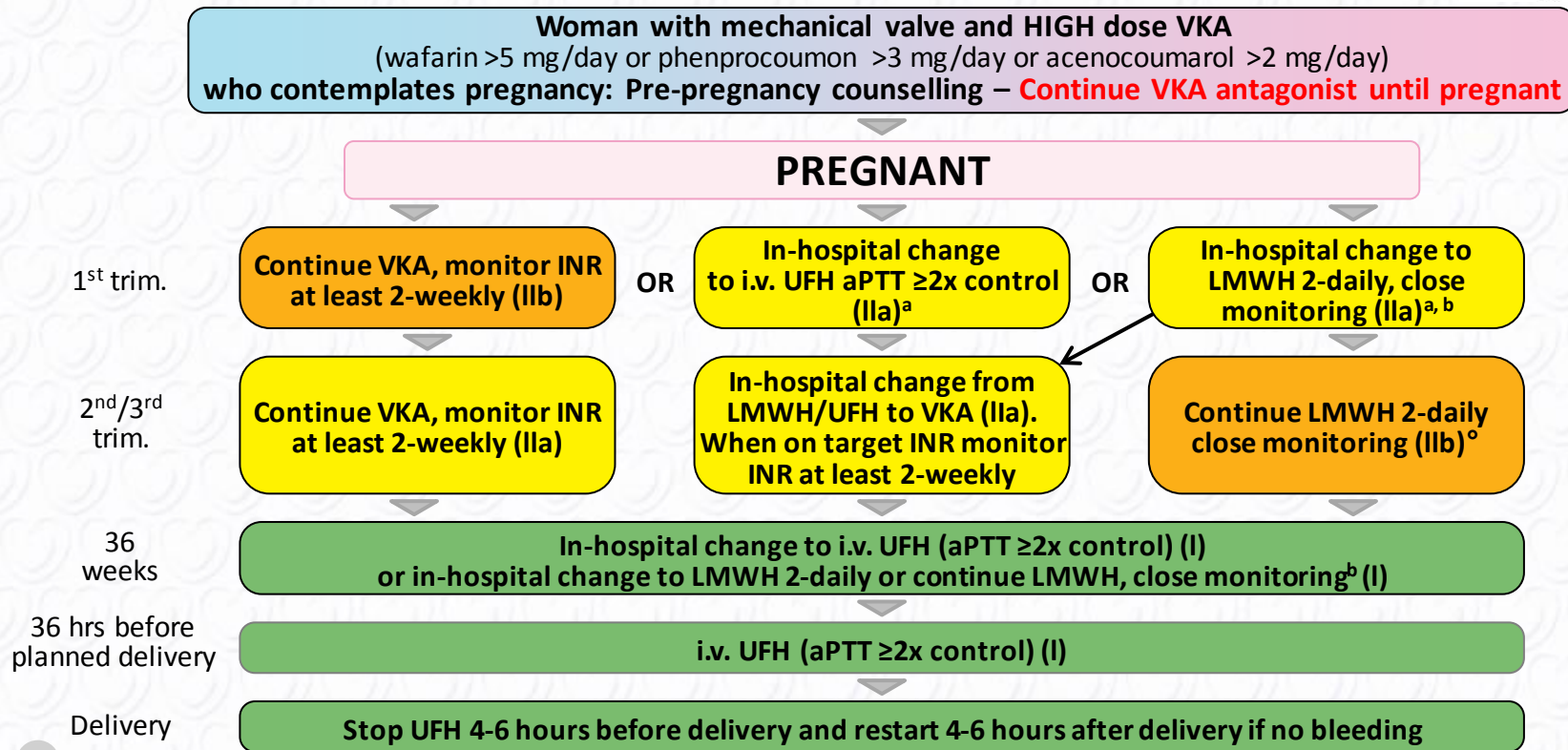


Table 11 Management of prosthetic heart valves (2)

Recommendations	Class	Level
In pregnant women on a VKA, it is recommended to perform INR monitoring weekly or every 2 weeks .	I	C
In pregnant women with LMWH , it is recommended to target anti-Xa levels 4–6 h post-dose at 0.8–1.2 U/l (aortic valve prosthesis) or 1.0–1.2 IU/mL (mitral and right-sided valve prostheses) .	I	C
It is recommended to replace LMWH with intravenous UFH (aPTT $\geq 2\times$ control) at least 36 h before planned delivery . UFH should be continued until 4–6 h before planned delivery and restarted 4–6 h after delivery if there are no bleeding complications.	I	C
It is recommended to anticipate the timing of delivery to ensure safe and effective peripartum anticoagulation.	I	C
Immediate echocardiography is recommended in women with mechanical valves presenting with dyspnoea and/or an embolic event.	I	C

Table 11 Management of prosthetic heart valves (4)

Recommendations	Class	Level
Discontinuation of VKAs between weeks 6 and 12 , and replacement with adjusted-dose intravenous UFH (aPTT $\geq 2\times$ control) or adjusted-dose LMWH ^c twice daily (see separate recommendations), should be considered in patients with a warfarin dose >5 mg/day (or phenprocoumon >3 mg/day or acenocoumarol >2 mg/day).	IIa	C
During the second and third trimesters, LMWH ^c with anti-Xa level monitoring and dose adjustment (see separate recommendations) may be considered in women who need a high dose of VKA ^e after patient information and consent.	IIb	C
In pregnant women with LMWH, in addition to monitoring peak anti-Xa levels, monitoring pre-dose levels targeted at ≥ 0.6 IU/mL may be considered.	IIb	C
LMWH is not recommended when weekly anti-Xa level monitoring and dose-adjustment is not available.	III	C

Insuffisance cardiaque

Dilated Cardiomyopathy



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● CMD

- évaluation attentive avant la grossesse
- information de la patiente
- FEVG / statut fonctionnel
- Attention nécessité d'arreter: IEC/ AA2/Entresto...

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Diagnosis (if otherwise well and uncomplicated)	<p>Small or mild</p> <ul style="list-style-type: none"> – pulmonary stenosis – patent ductus arteriosus – mitral valve prolapse <p>Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage)</p> <p>Atrial or ventricular ectopic beats, isolated</p>	<p>Unoperated atrial or ventricular septal defect</p> <p>Repaired tetralogy of Fallot</p> <p>Most arrhythmias (supraventricular arrhythmias)</p> <p>Turner syndrome without aortic dilatation</p>	<p>Mild left ventricular impairment (EF >45%)</p> <p>Hypertrophic cardiomyopathy</p> <p>Native or tissue valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis)</p> <p>Marfan or other HTAD syndrome without aortic dilatation</p> <p>Aorta <45 mm in bicuspid aortic valve pathology</p> <p>Repaired coarctation</p> <p>Atrioventricular septal defect</p>	<p>Moderate left ventricular impairment (EF 30–45%)</p> <p>Previous peripartum cardiomyopathy without any residual left ventricular impairment</p> <p>Mechanical valve</p> <p>Systemic right ventricle with good or mildly decreased ventricular function</p> <p>Fontan circulation. If otherwise the patient is well and the cardiac condition uncomplicated</p> <p>Unrepaired cyanotic heart disease</p> <p>Other complex heart disease</p> <p>Moderate mitral stenosis</p> <p>Severe asymptomatic aortic stenosis</p> <p>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)</p> <p>Ventricular tachycardia</p>	<p>Pulmonary arterial hypertension</p> <p>Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV)</p> <p>Previous peripartum cardiomyopathy with any residual left ventricular impairment</p> <p>Severe mitral stenosis</p> <p>Severe symptomatic aortic stenosis</p> <p>Systemic right ventricle with moderate or severely decreased ventricular function</p> <p>Severe aortic dilatation (>45 mm in Marfan syndrome or other HTAD, >50 mm in bicuspid aortic valve, Turner syndrome ASI >25 mm/m², tetralogy of Fallot >50 mm)</p> <p>Vascular Ehlers–Danlos</p> <p>Severe (re)coarctation</p> <p>Fontan with any complication</p>
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
Maternal cardiac event rate	2.5–5%	5.7–10.5%	10–19%	19–27%	40–100%

Cardiopathie du péri-partum

- Rare mais grave
- **Décision au cas/cas de l'attitude par rapport au fœtus**
 - état maternel?
 - viabilité du fœtus ?
 - avis des parents?
- **Traitement médical habituel + Bromocriptine (prolactine)**
- Discuter life-vest si FEVG <35%
- **Récupération dans près de 50% des cas: temporiser+++ avant DAI et greffe**

Table 13 Management of cardiomyopathies and heart failure (2)

Recommendations	Class	Level
As rapid diagnosis and decision-making is crucial for all pregnant women with acute HF, a pre-specified management algorithm and an interdisciplinary team should be established.	Ila	C
Patients in cardiogenic shock/dependent on inotropes should be transferred early to a facility where mechanical circulatory support is available.	Ila	C
Bromocriptine treatment should be accompanied by prophylactic (or therapeutic) anticoagulation.	Ila	C
Due to the high metabolic demands of lactation and breastfeeding, preventing lactation may be considered in patients with severe HF.	IIb	B
In patients with PPCM, bromocriptine treatment may be considered to stop lactation and enhance recovery (LV function).	IIb	B
In women with PPCM and DCM, subsequent pregnancy is not recommended if LVEF does not normalize.	III	C

Table 13 Management of cardiomyopathies and heart failure (3)

Recommendations	Class	Level
HCM		
In patients with HCM, the same risk stratifications as for non-pregnant women are recommended.	I	C
In patients with HCM, it is recommended that beta-blockers are continued in women who used them before pregnancy.	I	C
In patients with HCM, beta-blockers should be started in women who develop symptoms due to outflow tract obstruction or arrhythmia during pregnancy.	IIa	C
In HCM, cardioversion should be considered for persistent atrial fibrillation.	IIa	C

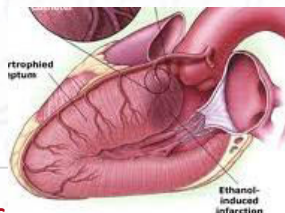


Table 16 Management of hypertension (1)

Recommendations	Class	Level
Low-dose aspirin (100–150 mg daily) is recommended in women at high or moderate risk of pre-eclampsia from week 12 to weeks 36-37.	I	A
In women with gestational hypertension or pre-existing hypertension superimposed by gestational hypertension, or with hypertension and sub-clinical organ damage or symptoms, initiation of drug treatment is recommended at SBP >140 mmHg or DBP >90 mmHg. In all other cases, initiation of drug treatment is recommended if SBP ≥150 mmHg or DBP ≥95 mmHg.	I	C
SBP ≥170 mmHg or DBP ≥110 mmHg in a pregnant woman is an emergency, and hospitalization is recommended.	I	C
Methyldopa, labetalol, and calcium antagonists are recommended for the treatment of hypertension in pregnancy.	I	B

Table 17 Prevention and treatment of venous thrombo-embolism (1)

Recommendations	Class	Level
LMWH is recommended for the prevention and treatment of VTE in pregnant patients.	I	B
For high-risk women, it is recommended to give a weight-related prophylactic dose of LMWH (e.g. enoxaparin 0.5 mg/kg once daily)	I	B
A documented assessment of risk factors for VTE before pregnancy or in early pregnancy is recommended in all women.	I	C
It is recommended that the therapeutic dose of LMWH is based on body weight.	I	C
Thrombolytics to manage patients with pulmonary embolism is only recommended in patients with severe hypotension or shock.	I	C
In high-risk women, it is recommended to convert LMWH to UFH at least 36 h prior to delivery and stop the UFH infusion 4–6 h prior to anticipated delivery. aPTT should be normal before regional anaesthesia.	I	C

Table 18 Drug use in pregnancy

Recommendations	Class	Level
Before pharmacological treatment in pregnancy is started, it is recommended to check Table 19 for clinical safety data.	I	C
In the absence of clinical safety data, it is recommended to check the electronic drug table (www.safefetus.com) for pre-clinical safety data.	I	C
In the absence of adequate human safety data, decision-making should be based on individual drug efficacy and safety profiles, and the available animal data, and the decision must be made together with the patient.	IIa	C
Decision-making based on former FDA categories alone is no longer recommended.	III	C

Table 19 Drugs and safety data

Drugs	Classification (Vaughan Williams for antiarrhythmic drugs)	Former FDA category	Placenta permeable	Transfer to breast milk (foetal dose)	Pre-clinical/ clinical safety data
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The table on drugs and safety data can be found in the Full text of the Guidelines and it is available at: www.escardio.org/guidelines and on the European Heart Journal web site (<https://academic.oup.com/eurheartj/articlelookup/doi/10.1093/eurheartj/ehy340>).

Accouchement

Les **modalités doivent être discutées à l'avance et expliquées aux parents**

- **≥ 40 semaines** pour toutes les mères « cardiaques »
- **Préférer la voie basse**
- **Césarienne si:**
 - pathologie aortique
 - patientes sous AVK (risque d'hémorragie fœtale intra crânienne)
 - IC sévère / HTAP sévères
 - complications obstétricales

MERCI

'What to do' and 'what not to do' messages from the Guidelines (1)

Recommendations	Class	Level
General recommendations		
Pre-pregnancy risk assessment and counselling is indicated in all women with known or suspected congenital or acquired cardiovascular and aortic disease.	I	C
It is recommended to treat high risk patients in specialized centres by a multidisciplinary team: the pregnancy heart team.	I	C
Echocardiography is recommended in any pregnant patient with unexplained or new cardiovascular signs or symptoms.	I	C
Vaginal delivery is recommended as first choice in most patients; for most important exceptions see below.	I	C
Prophylactic antibiotic therapy to prevent endocarditis during delivery is not recommended.	III	C

'What to do' and 'what not to do' messages from the Guidelines (2)

Recommendations	Class	Level
Pregnancy and pulmonary hypertension or congenital heart disease		
Right heart catheterization is recommended to confirm the diagnosis of PAH (group 1). This can be performed during pregnancy but with very strict indications, optimal timing, and shielding of the fetus.	I	C
Treatment dose LMWH is recommended in pregnant patients with chronic thrombo-embolic pulmonary hypertension.	IIa	C
Pregnancy is not recommended in patients with PAH.	III	B
Pregnancy is not recommended in patients with a systemic right ventricle and moderate or severely decreased ventricular function.		
Pregnancy is not recommended in patients after Fontan operation and any associated complication.	III	C

'What to do' and 'what not to do' messages from the Guidelines (3)

Recommendations	Class	Level
Management of aortic disease		
All aortic diseases		
Imaging of the entire aorta (CT/MRI) is recommended before pregnancy in patients with a genetically proven aortic syndrome or known aortic disease.	I	C
When a woman with known aortic dilatation, (history of) dissection, or genetic predisposition for dissection becomes pregnant, strict blood pressure control is recommended.	I	C
Repeated echocardiographic imaging every 4–12 weeks (depending on the diagnosis and severity of dilatation) is recommended during pregnancy and 6 months post-partum in patients with ascending aorta dilatation.	I	C
It is recommended to deliver all women with aortic dilatation or (history of) aortic dissection in an experienced centre with a pregnancy heart team, where cardiothoracic surgery is available.	I	C
In patients with an ascending aorta <40 mm, vaginal delivery is recommended.	I	C

'What to do' and 'what not to do' messages from the Guidelines (4)

Recommendations	Class	Level
In patients with an ascending aorta <40 mm, vaginal delivery is recommended.	I	C
Specific syndromes		
Pregnancy is not recommended in patients with vascular Ehlers–Danlos syndrome.	III	C
Management of native valvular heart disease		
Mitral stenosis		
In patients with symptoms or pulmonary hypertension, restricted activities and beta-1-selective blockers are recommended.	I	B
Diuretics are recommended when congestive symptoms persist despite beta-blockers.	I	B
Intervention is recommended before pregnancy in patients with MS and valve area <1.0 cm ² .	I	C
Therapeutic anticoagulation using heparins or VKA is recommended in case of AF, left atrial thrombosis, or prior embolism.	I	C

'What to do' and 'what not to do' messages from the Guidelines (5)

Recommendations	Class	Level
Chronic regurgitant lesions		
Surgical treatment is recommended before pregnancy in patients with severe aortic or mitral regurgitation and symptoms, impaired ventricular function, or ventricular dilatation.	I	C
Medical therapy is recommended in pregnant women with regurgitant lesions when symptoms occur.	I	C
Management of prosthetic heart valves		
It is recommended to choose the valve prosthesis in women contemplating pregnancy in consultation with a pregnancy heart team.	I	C
It is recommended to manage pregnancy in women with mechanical valves in a centre with a pregnancy heart team.	I	C

'What to do' and 'what not to do' messages from the Guidelines (6)

Recommendations	Class	Level
If delivery starts while on VKA or in less than 2 weeks after discontinuation of a VKA, caesarean section is indicated.	I	C
It is recommended to discontinue VKA and start adjusted-dose intravenous UFH (aPTT ≥ 2 control) or adjusted-dose LMWH (see separate recommendations) at the 36th week of gestation.	I	C
It is recommended to anticipate the timing of delivery to ensure safe and effective peripartum anticoagulation.	I	C
Immediate echocardiography is indicated in women with mechanical valves presenting with dyspnoea and/or an embolic event.	I	C
During the second and third trimesters until the 36th week, VKAs are recommended in women needing a low dose. ^a	I	C

'What to do' and 'what not to do' messages from the Guidelines (7)

Recommendations	Class	Level
Management of coronary artery disease		
ECG and measurement of troponin levels is recommended when a pregnant woman has chest pain.	I	C
Primary coronary angioplasty is recommended as the preferred reperfusion therapy for STEMI during pregnancy.	I	C
Breastfeeding is not recommended in mothers who take antiplatelet agents other than low-dose aspirin due to a lack of data.	III	C
Management of cardiomyopathies and heart failure		
Anticoagulation is recommended in patients with intracardiac thrombus detected by imaging or with evidence of systemic embolism.	I	A

'What to do' and 'what not to do' messages from the Guidelines (8)

Recommendations	Class	Level
It is recommended to treat women with heart failure during pregnancy according to current guidelines for non-pregnant patients, respecting contraindications for some drugs in pregnancy.	I	B
It is recommended to inform women with HFrEF about the risk of deterioration of the condition during gestation and peripartum.	I	C
Therapeutic anticoagulation with LMWH or VKAs according to stage of pregnancy is recommended for patients with AF.	I	C
In HFrEF, it is recommended that beta-blockers are continued in women who used them before pregnancy, or that they are installed with caution if symptoms persist.	I	C
In patients with PPCM and DCM, counselling for recurrence risk during subsequent pregnancy is recommended in all cases, even after recovery of LV function.	I	C

'What to do' and 'what not to do' messages from the Guidelines (9)

Recommendations	Class	Level
HCM		
In patients with HCM, it is recommended that beta-blockers are continued in women who used them before pregnancy.	I	C
Management of arrhythmias		
Acute management (intravenous administration of drugs) of SVT and AF		
Immediate electrical cardioversion is recommended for any tachycardia with haemodynamic instability and for pre-excited AF.	I	C
Long-term management (oral administration of drugs) of SVT and AF		
Beta-1-selective blockers or verapamil ^b are recommended for the prevention of SVT in patients without pre-excitation on resting ECG.	I	C

'What to do' and 'what not to do' messages from the Guidelines (10)

Recommendations	Class	Level
Flecainide ^c or propafenone ^c are recommended for the prevention of SVT in patients with WPW syndrome.	I	C
Beta-1-selective blockers are recommended for rate control of AT or AF.	I	C
Acute management (intravenous administration of drugs) of ventricular tachyarrhythmias		
Immediate electrical cardioversion is recommended for both sustained unstable and stable VT.	I	C
Long-term management (oral administration of drugs) of ventricular tachyarrhythmias		
Beta-blocking agents are recommended during pregnancy and post-partum in patients with long QT syndrome or catecholaminergic polymorphic ventricular tachycardia.	I	C

'What to do' and 'what not to do' messages from the Guidelines (11)

Recommendations	Class	Level
Management of hypertension		
Low-dose aspirin (100–150 mg daily) is recommended in women at high or moderate risk of pre-eclampsia from week 12 to week 36 -37.	I	A
In women with gestational hypertension or pre-existing hypertension superimposed by gestational hypertension, or with hypertension and subclinical organ damage or symptoms, initiation of drug treatment is recommended at SBP >140 mmHg or DBP >90 mmHg. In all other cases, initiation of drug treatment is recommended at SBP ≥150 mmHg or DBP ≥95 mmHg.	I	C
SBP ≥170 mmHg or DBP ≥110 mmHg in a pregnant woman is an emergency, and hospitalization is recommended.	I	C

'What to do' and 'what not to do' messages from the Guidelines (12)

Recommendations	Class	Level
Methyldopa, labetalol, and calcium antagonists are the drugs of choice for the treatment of hypertension in pregnancy.	I	C
It is recommended to expedite delivery in pre-eclampsia, and with adverse conditions such as visual disturbances or haemostatic disorders.	I	C
In severe hypertension, drug treatment with intravenous labetalol, oral methyldopa, or nifedipine is recommended.	I	C
Management of venous thrombo-embolism		
LMWH is recommended for the prevention and treatment of VTE in pregnant patients.	I	B
For high-risk women, it is recommended to give a weight-related prophylactic dose of LMWH (e.g. enoxaparin 0.5 mg/kg once daily).	I	B

'What to do' and 'what not to do' messages from the Guidelines (13)

Recommendations	Class	Level
It is recommended that the therapeutic dose of LMWH is based on body weight.	I	C
Thrombolytics to manage patients with pulmonary embolism are only recommended in patients with severe hypotension or shock.	I	C
In high-risk women, it is recommended to convert LMWH to UFH at least 36 h prior to delivery and to stop the UFH infusion 4-6 h prior to anticipated delivery. aPTT should be normal before regional anaesthesia.	I	C
Drug use in pregnancy		
Before pharmacological treatment in pregnancy is started, it is recommended to check drugs and safety data.	I	C
In the absence of clinical safety data, it is recommended to check the supplementary data and www.safefetus.com for pre-clinical safety data.	I	C
Decision making based on former FDA categories alone is no longer recommended.	III	C

Essential messages (1)

- Risk estimation should be individualized depending on the underlying cardiac diagnosis, ventricular and valvular function, functional class, presence of cyanosis, PAPs, and other factors.
- Indications for intervention (surgical or catheter) in the majority of patients do not differ in women who consider pregnancy compared with other patients. There are a. **few exceptions, such as some degree of aortic dilatation and severe asymptomatic MS**
- In women with a moderate or high-risk of complications during pregnancy (mWHO II–III, III, and IV), pre-pregnancy counselling and management during pregnancy and around delivery should be performed in an expert centre by a multidisciplinary team: the pregnancy heart team.
- All women with congenital or other possibly genetic heart disease should be offered foetal echocardiography in weeks 19-22 of pregnancy.
- A delivery plan should be made between 20-30 weeks of pregnancy detailing induction, management of labour, delivery, and post-partum surveillance.

Essential messages (2)

- Induction of labour should be considered at 40 weeks of gestation in all women with cardiac disease.
- Vaginal delivery is the first choice for the majority of patients.
- Indications for caesarean section are:
 - pre-term labour in patients on OACs,
 - aggressive aortic pathology,
 - acute intractable HF,
 - severe forms of PH (including Eisenmenger's syndrome).
- Pregnancy termination should be discussed if there is a high-risk of maternal morbidity or mortality, and/or of foetal abnormality.
- Pregnancy, and consequently fertility treatment, is contraindicated in women with mWHO class IV.

Essential messages (3)

- All patients with known cardiac or aortic disease need investigations and counselling about the risks of pregnancy pre-pregnancy or before assisted reproductive therapy.
- The **following patients should be counselled against pregnancy**:
 - with a Fontan operation and additional comorbidities (ventricular dysfunction, arrhythmias, or valve regurgitation),
 - with PAH,
 - severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV),
 - severe (re-)coarctation,
 - systemic right ventricle with moderate or severely decreased ventricular function,
 - with vascular Ehlers-Danlos,
 - with severe aortic dilatation or (history of) aortic dissection,

Essential messages (4)

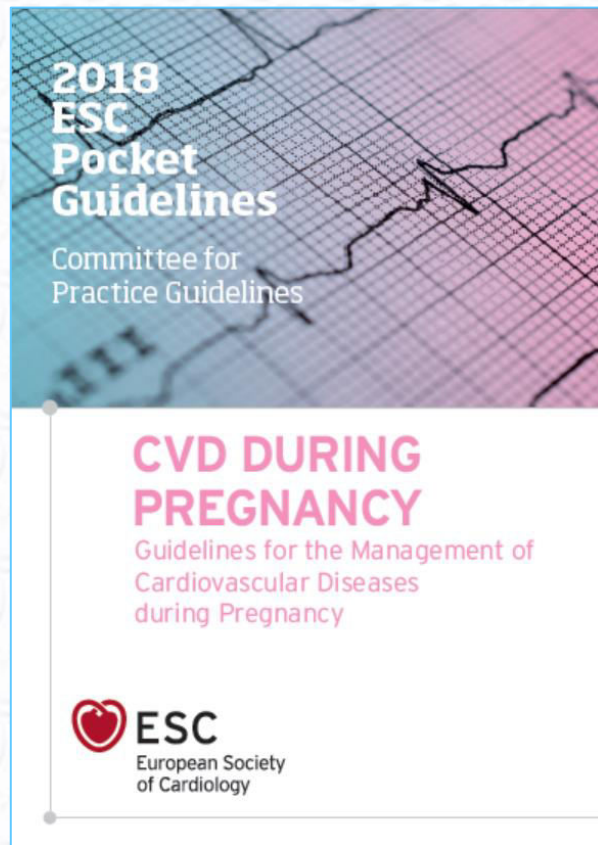
- with severe MS (even when asymptomatic),
- patients with severe AS who are symptomatic, or asymptomatic patients with impaired LV function or a pathological exercise test,
- if LVEF does not normalize in women with previous PPCM.
- Women with a mechanical valve prosthesis are at high-risk of maternal morbidity (especially valve thrombosis and bleeding) and even mortality, and should be managed by a pregnancy heart team in expert centres.
- LMWH should **only be used when weekly monitoring of anti-Xa levels with dose adjustment is available.**
- Women with HF during pregnancy should be treated according to current guidelines for non-pregnant patients, respecting contraindications for some drugs in pregnancy (see table ‘Recommendations for drug use in pregnancy’). When inotropes or more advanced treatment is necessary, transport to an expert centre is recommended.

Essential messages (5)

- It is recommended to inform women with DCM and HFrEF about the risk of deterioration of the condition during gestation and peripartum.
- In women with PPCM and DCM, subsequent pregnancy is not recommended if LVEF does not normalize.
- Patients with congenital LQTS and catecholaminergic polymorphic VT are recommended beta-blockers during pregnancy and post-partum.
- Initiation of antihypertensive drug treatment is recommended in all women with persistent elevation of BP $\geq 150/95$ mmHg and at values $>140/90$ mmHg in women with:
 - gestational hypertension (with or without proteinuria),
 - pre-existing hypertension with the superimposition of gestational hypertension,
 - hypertension with subclinical organ damage or symptoms at any time during pregnancy.

Essential messages (6)

- Women at high or moderate risk of pre-eclampsia should be advised to take 100-150 mg of acetylsalicylic acid daily from week 12 to week 36-37 in addition to their hypertension treatment.
- Methyldopa, labetalol, and calcium antagonists are recommended for the treatment of hypertension in pregnancy.
- LMWH is the agent of choice for VTE prophylaxis and treatment.
- Thrombolytics to treat thrombo-embolism should only be used in patients with severe hypotension or shock.
- In the case of an emergency, drugs that are not recommended by the pharmaceutical industry during pregnancy and breastfeeding should not be withheld from the mother. The potential risk of a drug and the possible benefit of the therapy must be weighed against each other.



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Table 14 Surveillance levels at time of delivery in women with arrhythmias (1)

Risk for arrhythmia with Haemodynamic compromise at delivery		Level of Surveillance ^a	Class ^b	Level ^c
Low risk	PSVT, AF, idiopathic VT, low-risk LQTS, WPW syndrome	1	I	C
Medium risk	Unstable SVT, VT, ICD carriers, VT and structural heart disease, Brugada syndrome; moderate risk: LQTS, catecholaminergic polymorphic VT	2	I	C
High risk for life threatening arrhythmia	Unstable VT in structural heart disease/congenital heart disease, unstable VT/TdP in high-risk LQTS patients, short QT syndrome, high-risk catecholaminergic polymorphic VT	3	I	C

Table 14 Surveillance levels at time of delivery in women with arrhythmias (2)

Descriptions of actions to be planned	Surveillance level		
	Low 1	Medium 2	High 3
Consult cardiologist	x		
Consultation with multidisciplinary team including arrhythmologists at specialized centre		x	x
Mode and location of delivery as advised by obstetricians	x	x	
Caesarean delivery recommended			x
Monitor cardiac rhythm (telemetry, external rhythm monitor)		(x)	x
Intravenous line		x	x
Arterial line			x

Table 14 Surveillance levels at time of delivery in women with arrhythmias (3)

Descriptions of actions to be planned	Surveillance level		
	Low 1	Medium 2	High 3
Prepare for intravenous administration of adenosine		x	
Prepare for intravenous administration of a beta-blocker		x	x
Prepare for intravenous administration of selected antiarrhythmic drugs			x
External cardioverter defibrillator at site		x	x
Delivery at thoracic operating theatre			x
Prepare for transfer to cardiac intensive care unit post-partum if needed			x

Table 15 Management of arrhythmias (1)

Recommendations	Class	Level
Acute management (intravenous administration of drugs) of SVT and AF		
Vagal manoeuvres, followed by adenosine if these fail, are recommended for acute conversion of PSVT.	I	C
Immediate electrical cardioversion is recommended for any tachycardia with haemodynamic instability and for pre-excited AF.	I	C
Beta-1-selective blockers should be considered for acute conversion of PSVT.	IIa	C
Ibutilide or flecainide may be considered for termination of atrial flutter and AF in stable patients with structurally normal hearts. ^c	IIb	C
Long-term management (oral administration of drugs) of SVT and AF		
Beta-1-selective blockers or verapamil is recommended for the prevention of SVT in patients without pre-excitation on resting ECG.	I	C
Flecainide ^e or propafenone ^e are recommended for the prevention of SVT in patients with WPW syndrome.	I	C

Table 15 Management of arrhythmias (2)

Recommendations	Class	Level
Long-term management (oral administration of drugs) of SVT and AF (cont'd)		
Beta-selective blockers are recommended for rate control of AT or AF.	I	C
Flecainide ^e , propafenone, ^e or sotalol ^f should be considered to prevent SVT, AT, and AF if AV nodal blocking agents fail.	IIa	C
Digoxin and verapamil should be considered for rate control of AT or AF if beta-blockers fail.	IIa	C
Catheter ablation with electroanatomical systems should be considered in experienced centres in cases of drug-refractory and poorly tolerated SVT.	IIa	C
Acute management (intravenous administration of drugs) of ventricular tachyarrhythmias		
Immediate electrical cardioversion is recommended for sustained both unstable and stable VT.	I	C
For acute conversion of sustained, haemodynamically stable, monomorphic VT (e.g. idiopathic VT), a beta-blocker, sotalol, ^f flecainide, ^e procainamide, or overdrive ventricular pacing should be considered.	IIa	C

Table 15 Management of arrhythmias (3)

Recommendations	Class	Level
Long-term management (oral administration of drugs) of Ventricular tachyarrhythmias		
ICD (preferably one chamber) is recommended prior to pregnancy if clinically indicated but also during pregnancy, preferably using echocardiographic guidance or mapping, especially if the foetus is beyond 8 weeks of gestation, if indication emerges.	I	C
Beta-blocking agents are recommended during pregnancy and post-partum in patients with long QT syndrome or catecholaminergic polymorphic VT.	I	C
Beta-blocking agents or verapamil ^{d,e} are recommended for the prevention of idiopathic sustained VT if associated with severe symptoms or haemodynamic compromise.	I	C
In idiopathic sustained VT, sotalol ^f or flecainide ^e should be considered for prevention if other drugs fail.	IIa	C
Catheter ablation with electroanatomical mapping systems may be considered in experienced centres in sustained drug-refractory and poorly tolerated VT if there are no other alternatives.	IIb	C