cardiopathies congénitales maternelles

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European Heart Journal
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ESC Guidelines on the management of cardiovascular diseases during pregnancy

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

Endorsed by the European Society of Gynecology (ESG), the Association for European Paediatric Cardiology (AEPC), and the German Society for Gender Medicine (DGesGM).

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Population of Europe of Childbearing Age

EU population 2008
499 million total*

- 105 million women in childbearing age (15-45 years).
- 5 million live births.
- 1% of pregnancies are complicated by heart disease.**

*http://epp.eurostat.ec.europa.eu/portal/page/portal/population/data
**Report on Maternal Deaths in UK RCOG

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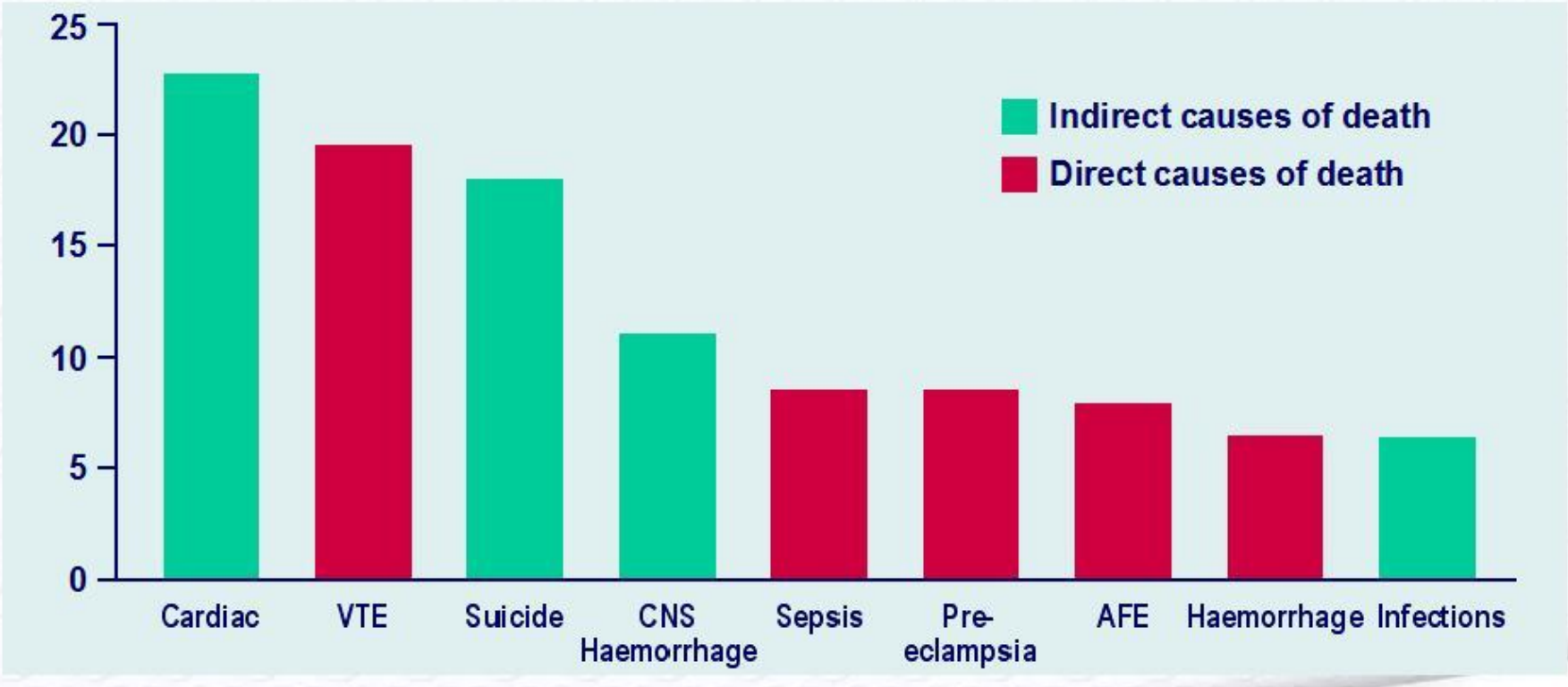
Major Causes of Maternal Death (UK 2003-2005)

Overall death rates per million maternities

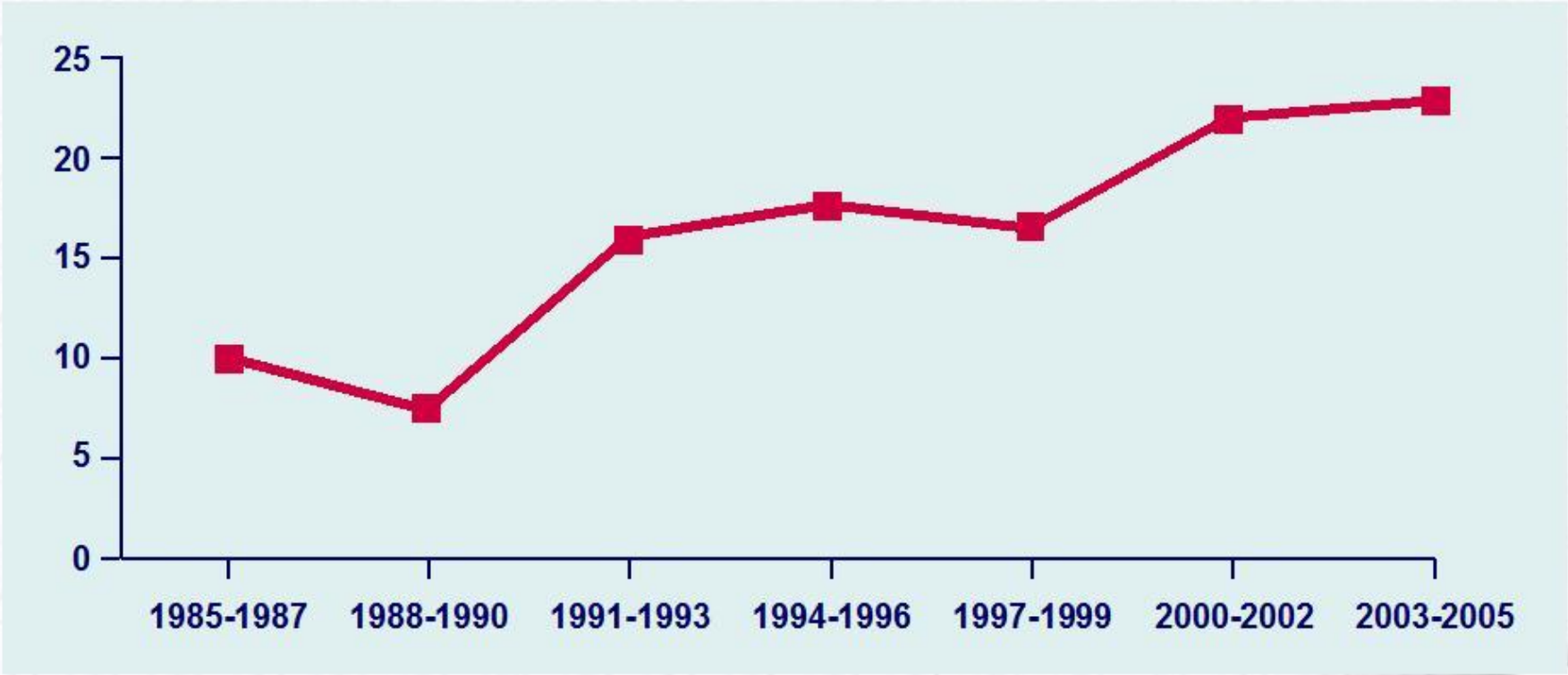


Roos-Hesselink Heart 2009;95:680-6

European Heart Journal 2011, doi: 10.1093/eurheartj/ehr218

Evolution of Maternal Mortality from Heart Disease in the UK

Cardiac



Roos-Hesselink et al. *Heart* 2009;95:680-6

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Heart Diseases during Pregnancy (I)

- Congenital heart disease:
  - most frequent cause of cardiac complications in industrialised countries (70-80%), rare in developing countries (10-20%).
- Valvular disease:
  - most frequent cause of cardiac complications in developing countries (50-90%), 15% in industrialised countries.
- Cardiomyopathies:
  - rare but severe.
- Coronary heart disease:
  - rare but increasing frequency.

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Aetiology of Cardiac Diseases in Pregnancy

Study	n	Rheumatic	Cong.	Other	Mortality	Morbidity
Siu 2001, 2002, Canada	562	Acquir. VD14 - 22%	74%	12%	1%	13%
Lesniak Sobelga 2004, Poland	259	62% Rheum 20% MVP		18% VR	0%	15%
Madazli 2010,Turkey	144	88% Rheum	12%	-	0%	6% - 66%

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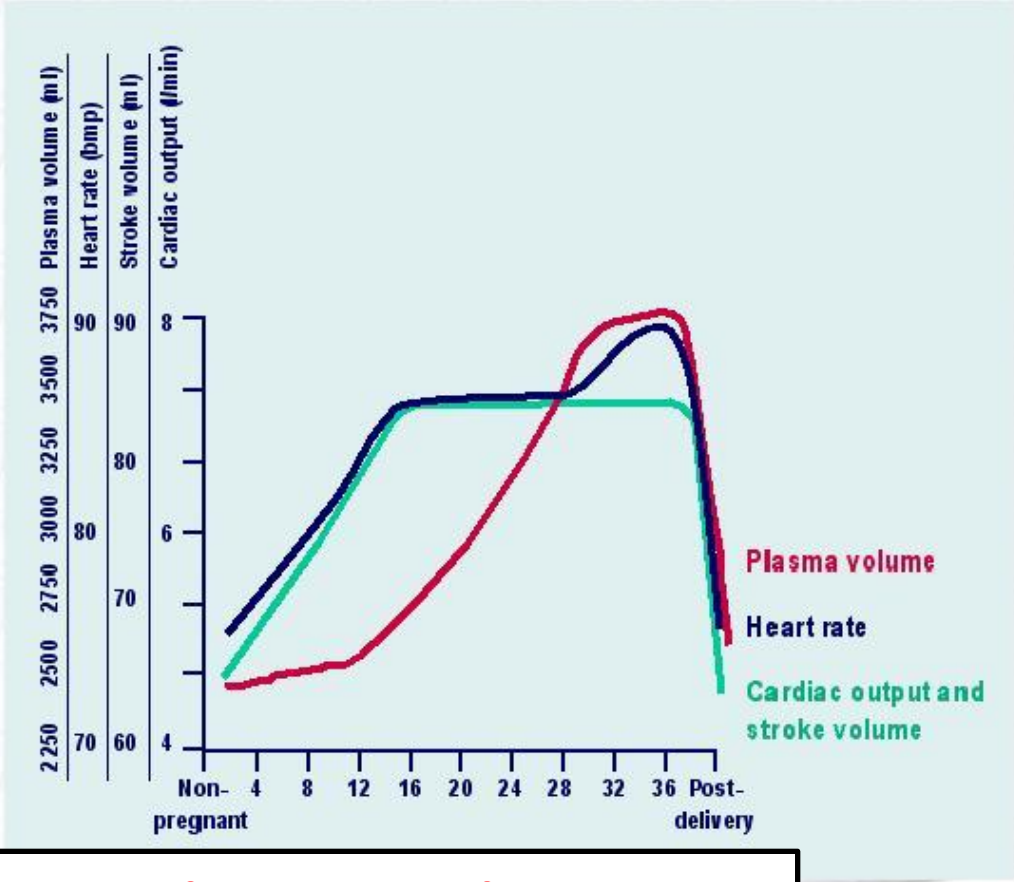
Haemodynamic Changes During Pregnancy

- ↑ blood volume ≈ 50%.
- ↑ cardiac output 30-50% maximum between, 5th and 8th months.
- ↓ systolic and diastolic blood pressure.
- ↓ systemic arterial resistance (hormones, placenta).



gradient croissant , cyanose croissante , risque majoré d' arythmies

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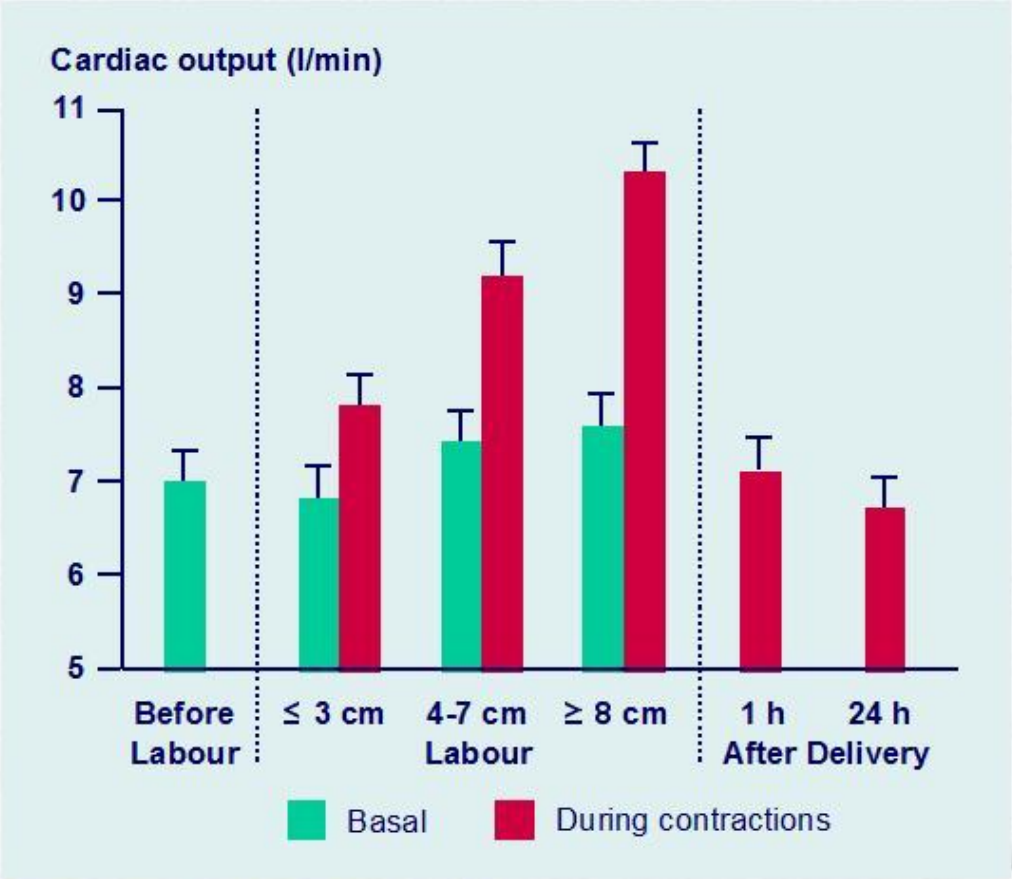
Haemodynamic Changes During Delivery

- Labour:
  - ↑ O_2 consumption,
  - ↑ baseline cardiac output,
  - ↑ cardiac output and blood pressure during uterine contractions, depending on modalities of delivery (epidural analgesia, Cesarean section)
- Post-partum:
  - ↑ blood shift from placenta,
  - ↑ preload and cardiac



Hunter et al. *Br Med J* 1992;68:540-3

Insuffisance cardiaque en post partum++++

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Other Changes during Pregnancy

- Haemostasis:
  - increased platelet adhesiveness,
  - increased concentration of coagulation factors, fibrinogen,
  - impaired fibrinolysis.

  → ***Hypercoagulability***

- Maternal glucose metabolism.

- Drug metabolism:
  - absorption, excretion, and bioavailability.

Pendant 3 à 4 semaines

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Accouchement

- Perte 500ml si accouchement par voie basse
- 1000 ml si césarienne
- Retour au débit cardiaque normal en 2 à 3 semaines

Timing and Mode of Delivery

- Favour spontaneous onset of labour and vaginal delivery in most cases of stable heart disease.
- Wide use of lumbar epidural analgesia.
- Indications for Caesarean section:
  - pre-term labour in patients on oral anticoagulants,
  - Marfan and other ascending aortic aneurysms (IIaC if > 45 mm, IIbC if 40-45 mm),
  - aortic dissection (IIaC),
  - severe aortic stenosis (IIaC),
  - Eisenmenger syndrome (IIaC).
- Multidisciplinary care for high-risk patients.

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Risk Stratification - CARPREG

Predictors of maternal cardiovascular events and risk score from the CARPREG study

Prior cardiac event (heart failure, transient ischaemic attack, stroke before pregnancy or arrhythmia).
Baseline NYHA functional class > II or cyanosis.
Left heart obstruction (mitral valve area < 2 cm^2, aortic valve area < 1.5 cm^2, peak LV outflow tract gradient > 30 mmHg by echocardiography).
Reduced systemic ventricular systolic function (ejection fraction < 40%

CARPREG risk score: for each CARPREG predictor that is present a point is assigned.
Risk estimation of cardiovascular maternal complications.
0 point 5%
1 point 27%
>1 point 75%

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Stratification

High risk states - contraindications for pregnancy

Conditions in which pregnancy risk is WHO IV (pregnancy contraindicated)
• Pulmonary arterial hypertension of any cause.
• Severe systemic ventricular dysfunction (LVEF < 30%, NYHA III-IV).
• Previous peripartum cardiomyopathy with any residual impairment of left ventricular function.
• Severe mitral stenosis, severe symptomatic aortic stenosis.
• Marfan syndrome with aorta dilated > 45 mm. • Aortic dilatation > 50 mm in aortic disease associated with bicuspid aortic valve.
• Native severe coarctation.

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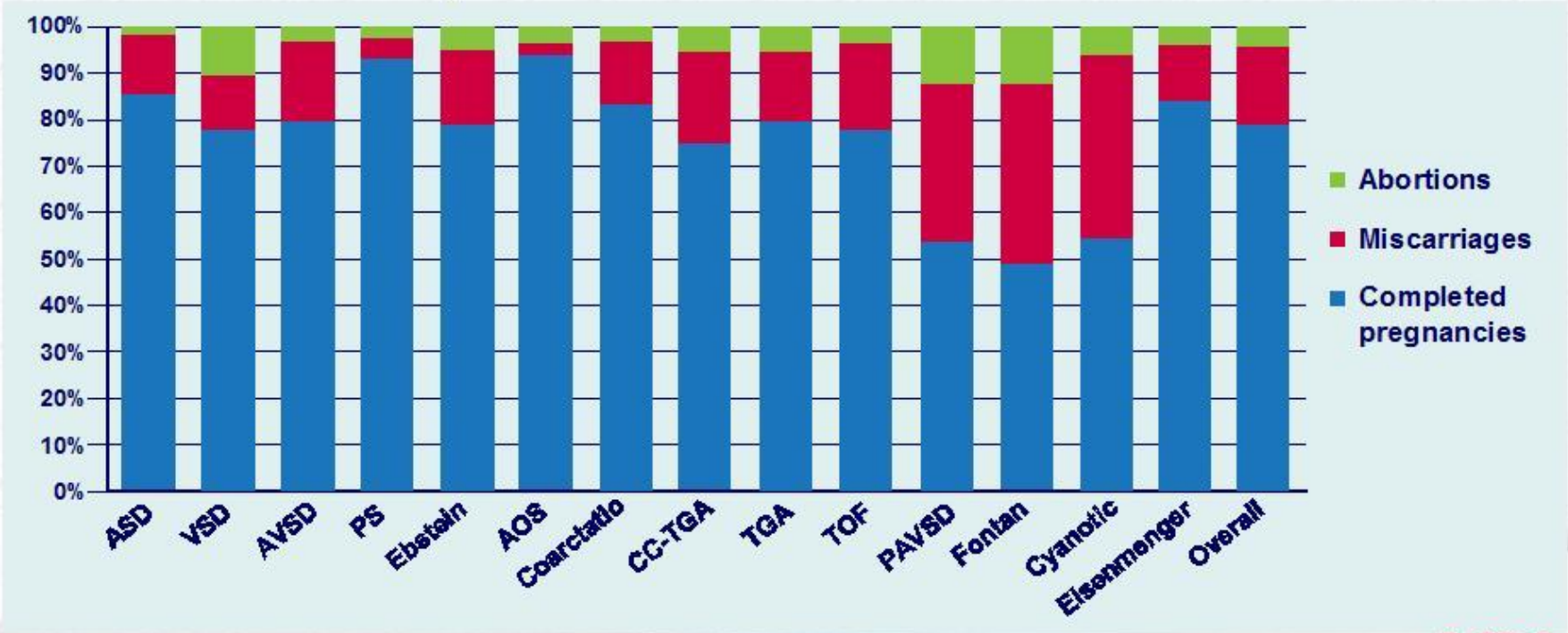
Congenital Heart Disease

- Risk for fetus depends on the underlying maternal heart disease as well as maternal ventricular and valvular function, functional class, cyanosis, use of anticoagulants.
- Overall fetal mortality 4%.



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Congenital Heart Disease

- Left to right shunts:
  - low to moderate risk.
- Right to left shunts (cyanotic heart disease):
  - moderate risk if previously repaired,
  - high fetal risk if not repaired and O_2 saturation < 85%,
  - major maternal risk (30-50% mortality) if Eisenmenger syndrome contraindication for pregnancy or early termination.
- Obstructions without shunts:
  - high risk if severe left ventricular outflow tract obstruction.

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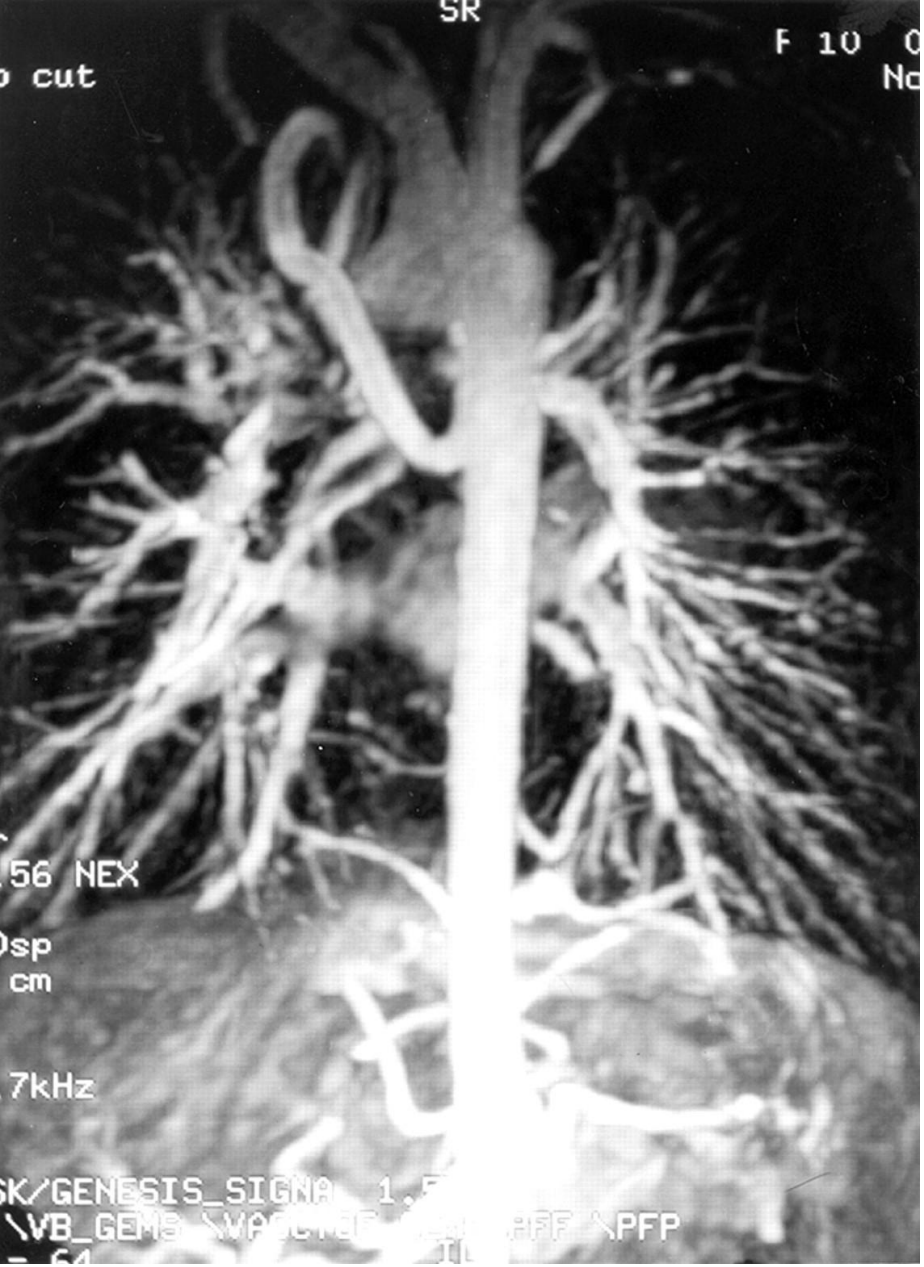
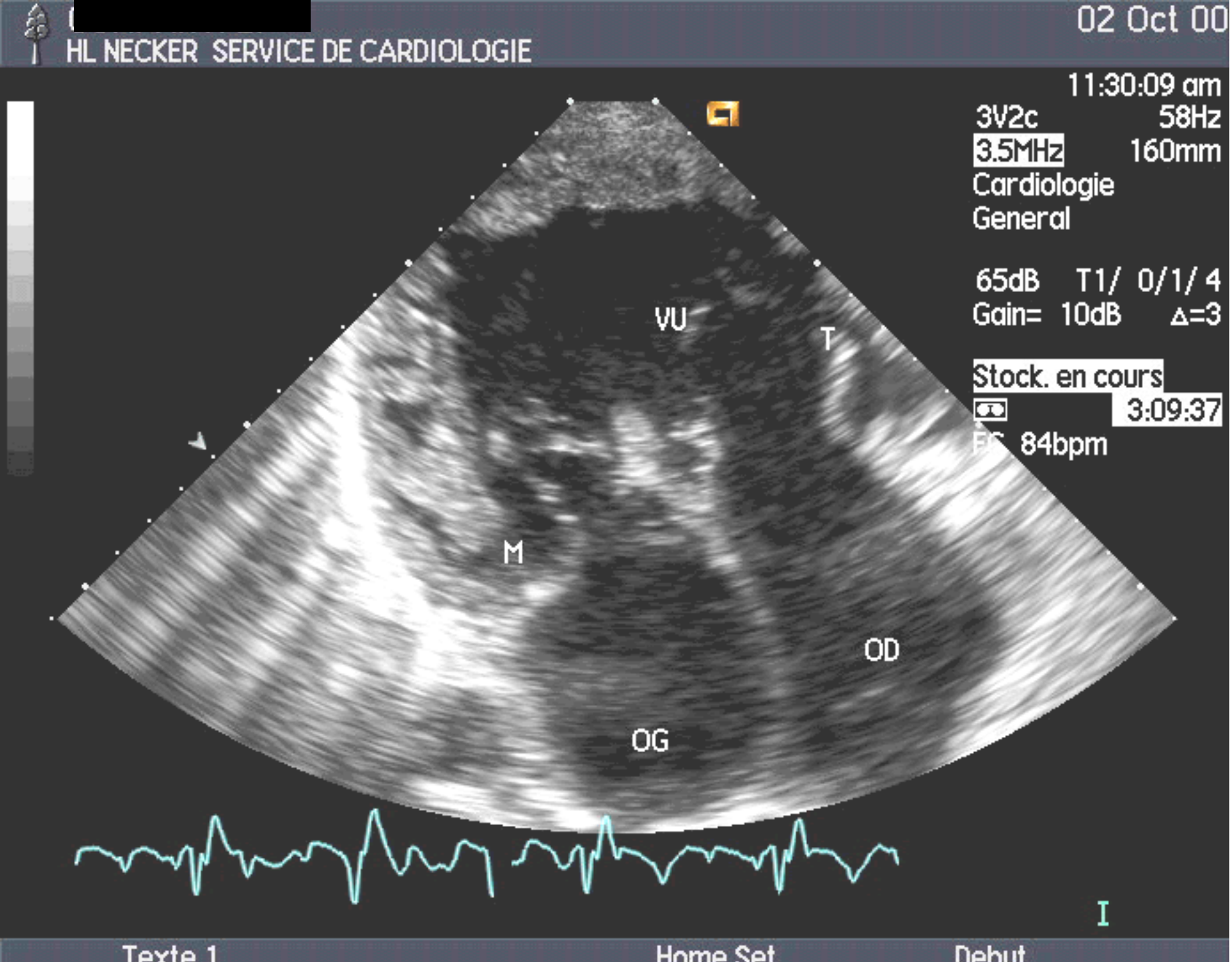


Atrésie pulmonaire avec CIV


[redacted]
02 Oct 00
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Cardiologie
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3:09:37
84bpm
VU
T
M
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OG
I
Texte 1
Home Set
Debut

Low risk patients

- Patients who have undergone previous successful surgical repair for congenital heart disease tolerate pregnancy often well if:
  - no mechanical valve is implanted,
  - the exercise tolerance is good,
  - the ventricular function is normal.

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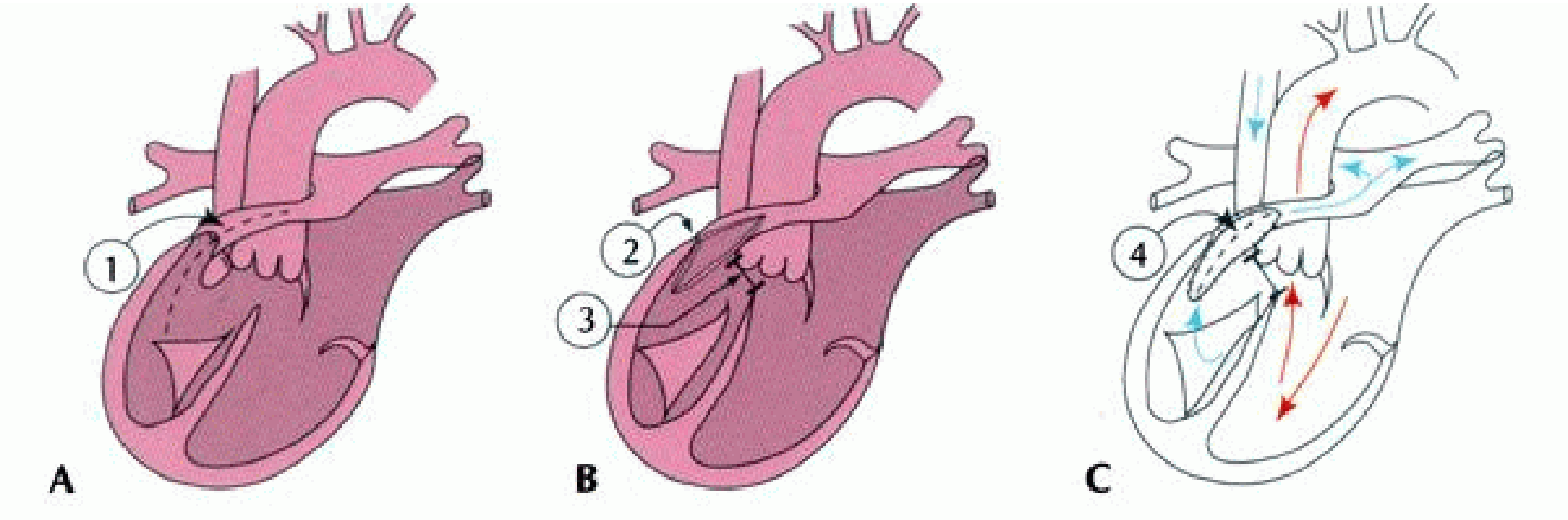
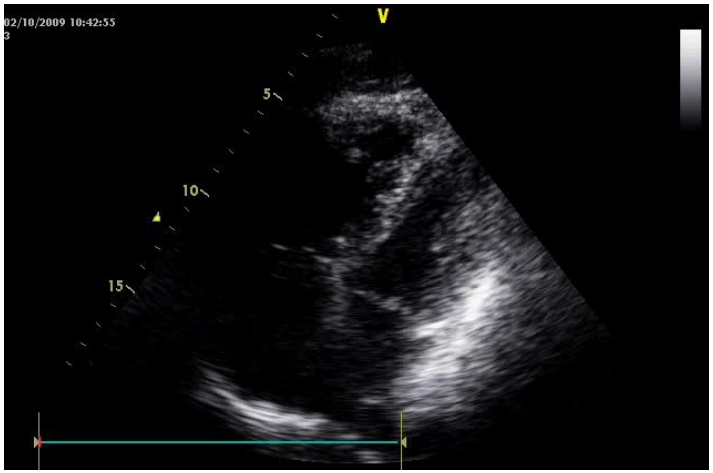
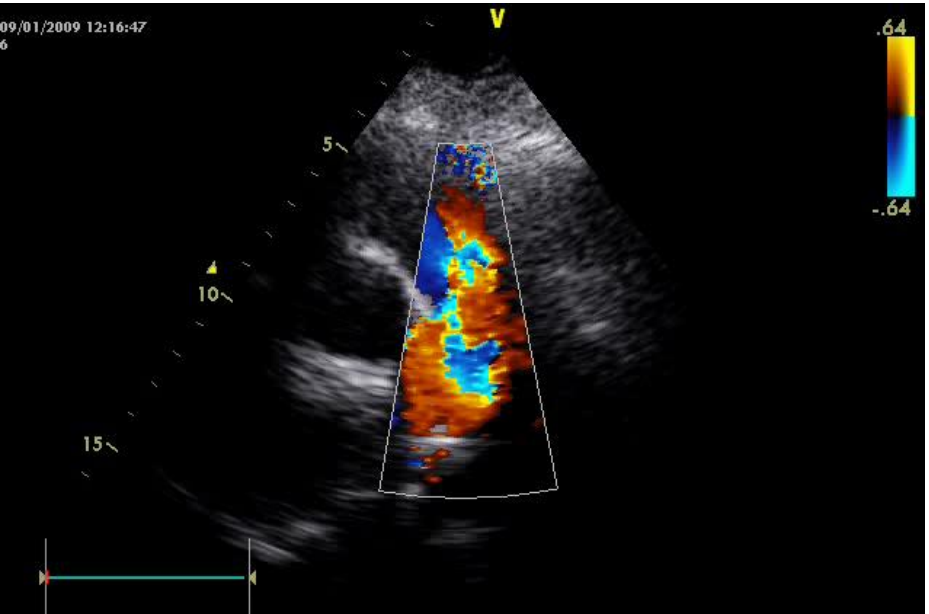
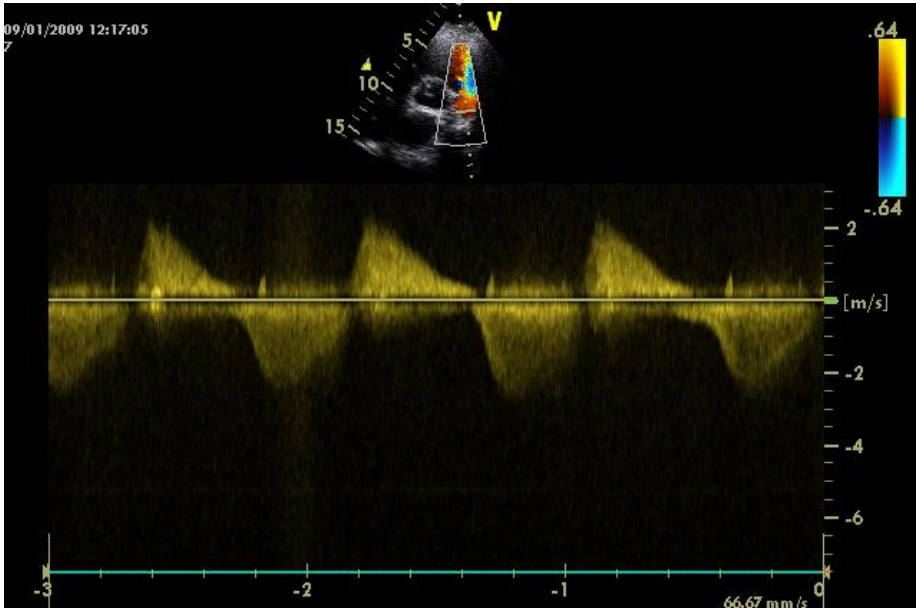
Tétralogie de Fallot



VD dilaté et IP importante







Zahara study n=123

- Evenement durant les grossesses menées a terme chez les patietnes ayant eu une tetralogie de Fallot opérées complete pregnancies in women with corrected ToF
- Evenements Cardiovasculaires 10 (8.1)

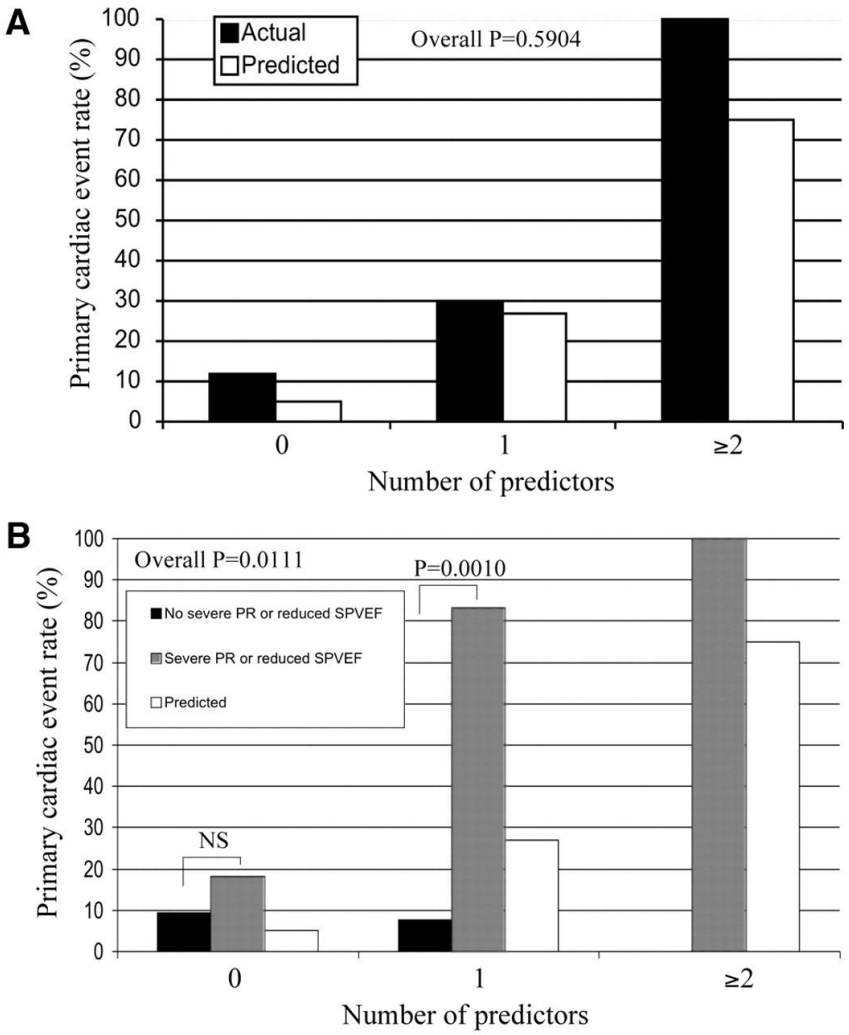
- Arythmies 8 (6.5) <1
- Defaillance cardiaque 2 (1.6) <1
- Evenements Thromboemboliques 1 (0.8) <0.3

Zahara study n=123

- Evenements Obstétricaux 73 (58.9)

- Cesarienne 25 (20.3) 6.5
- Voie basse 16 (13.0) 17
- Preeclampsie 4 (3.2) 1.4

A, Actual vs predicted primary cardiac event rates with varying numbers of predictors.



Khairy P et al. Circulation 2006;113:517-524

Congenital Heart Diseases:
Specific maternal high risk conditions WHO (III)-IV

Condition	Expected outcome
Pulmonary hypertension	Neonatal survival 87-89%. *(Bedard, EHJ 2009)*
Eisenmenger syndrome	Maternal mortality of 20-50%. Life birth 12% if O_2 saturation < 85%. *(Presbitero, Circ 1994)*
Cyanotic HD without PH	Depends on maternal oxygen saturation. Life birth 12% if O_2 saturation < 85%. *(Presbitero, Circ 1994)*
Severe LVOTO	Should be treated before pregnancy. If not, discourage pregnancy.

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Congenital Heart Diseases:
Specific defects

Specific defect	Maternal and fetal risk, management and delivery
ASD, VSD, AVSD, CoA, PST, AVST	Low to moderate risk, WHO I or II
Fallot, Ebstein's anomaly	Should be repaired before pregnancy: WHO II
Transposition of great arteries	WHO III Irreversible decline in maternal cardiac function in 10 % of pregnancies
Congenitally corrected TGA	WHO III, Fetal loss increased, Pregnancy contraindicated if EF < 40%
...	...

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Congenital HD - examples

Fontan operation

Anticoagulation treatment should be considered during pregnancy in Fontan patients.	IIa	C
Fontan patients with depressed ventricular function and/or moderate to severe atrioventricular valvular regurgitation or with cyanosis or with protein-losing enteropathy should be advised against pregnancy.	III	C

Maternal death	2%
Live births	45%
Maternal complications	20%

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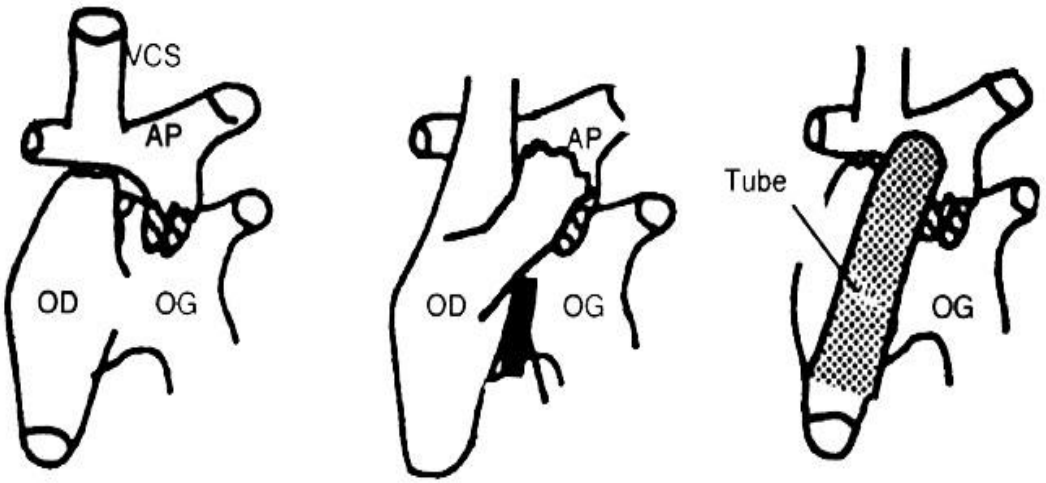
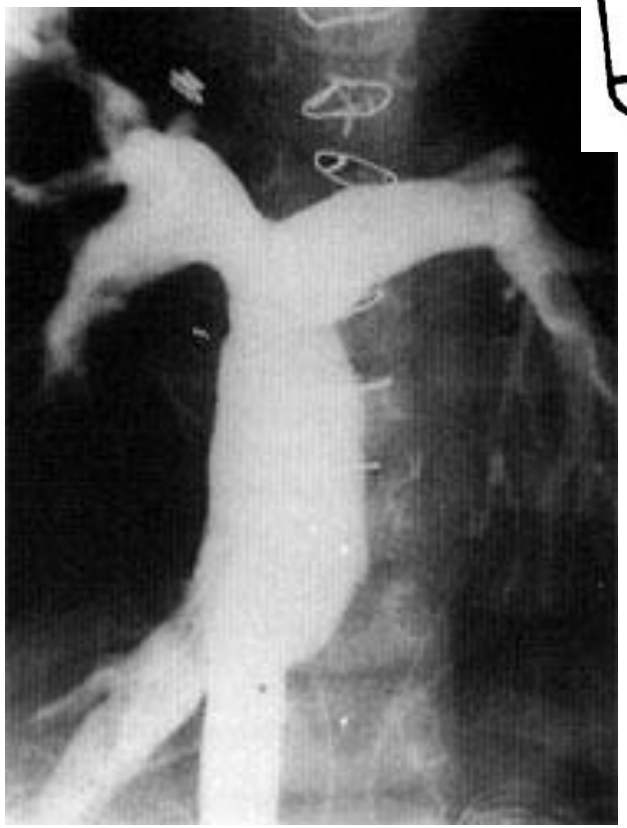
Cicatrice rythmique de Fontan





Atrial arrhythmias
Sinus failure
Intra-atrial delay
Epicardial
Elevated thresholds

Diseases of the Aorta

Increased risk of dissection during pregnancy
May lead to consider prophylactic surgery

	Risk of dissection
• Marfan syndrome	aortic Ø > 45 mm
• Bicuspid aortic valve	lower risk than Marfan
• Ehlers Danlos type IV	even if non-dilated aorta
• Turner syndrome	consider body size aortic Ø > 27 mm/m² BSA

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Aortic diseases - Delivery

Ascending aorta diameter

• < 40 mm	Vaginal delivery is favoured.
• 40-45 mm	Decision on individual basis.
• > 45 mm	Caesarean delivery should be considered.

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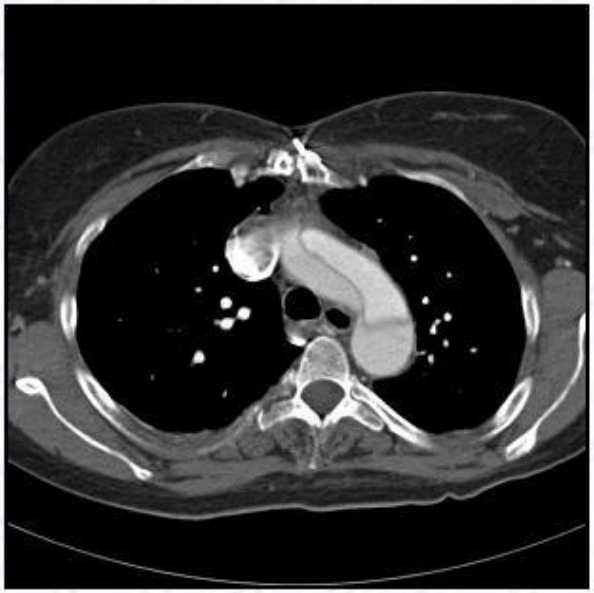
Aortic diseases



- Full assessment and counseling should be performed before pregnancy in all patients with known aortic disease.
- Patients with (or history of) type B dissection should be advised against pregnancy.
- In pregnant women with known aortic dilatation, (history of) type B dissection or genetic predisposition for dissection strict blood pressure control is recommended.
- Prophylactic surgery should be considered during pregnancy if the aortic diameter is ≥ 50 mm and increasing rapidly. When the fetus is viable, caesarean delivery followed directly by aortic surgery is recommended.

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Haut risque
Obstacles au cœur gauche

- Sténose aortique
- Sténose sous aortique
- cardiopathie obstructive
- coarctations

augmentation du gradient avec le débit

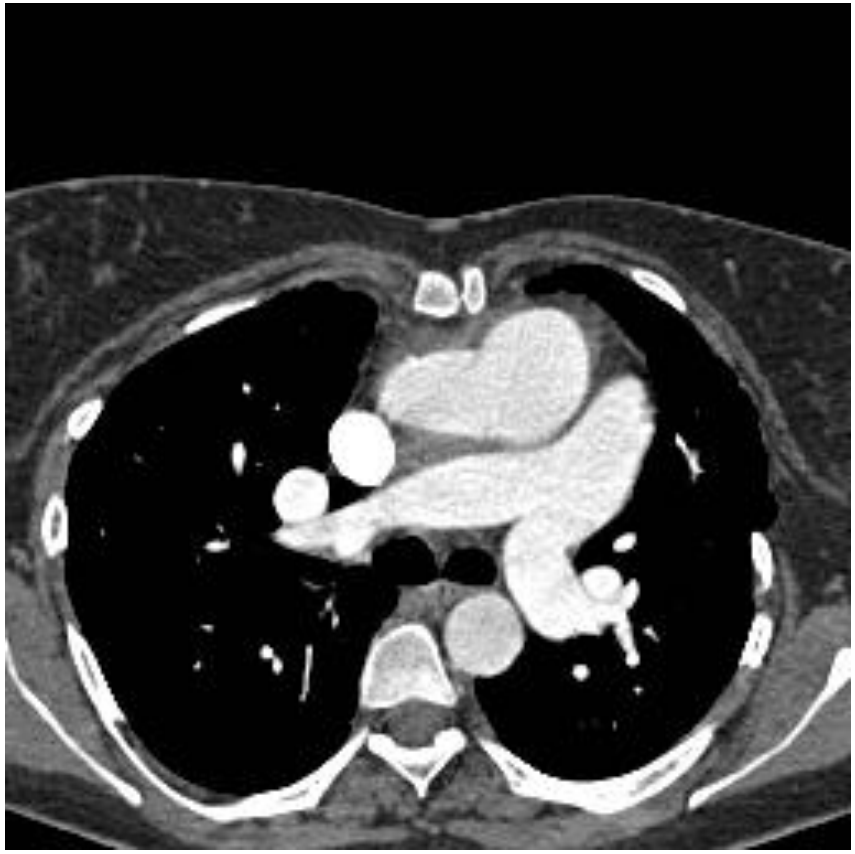
risque de syncope

efforts de poussée contre indiqués

Haut risque :Aortes fragiles

- Marfan avec aorte supérieure à 40 mm
- Dilatation aortique en dehors du Marfan (bicuspidie) risque réel de dissection
- cardiopathie cyanogène opérée tard avec dilatation aortique?

Aortic dissection



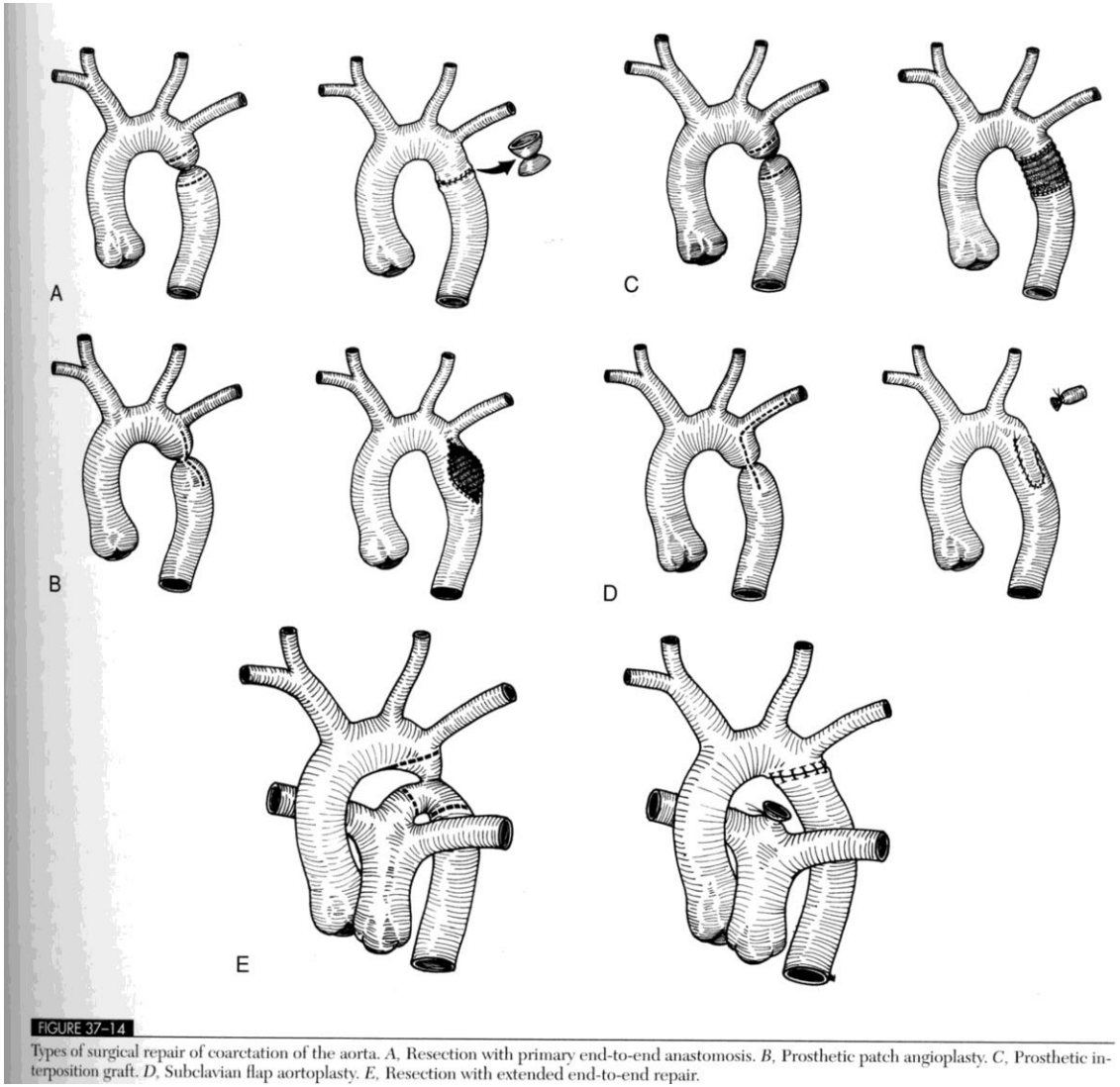


FIGURE 37–14

Types of surgical repair of coarctation of the aorta. *A*, Resection with primary end-to-end anastomosis. *B*, Prosthetic patch angioplasty. *C*, Prosthetic interposition graft. *D*, Subclavian flap aortoplasty. *E*, Resection with extended end-to-end repair.

Différents modes de réparation des coarctations

coarctation aortique : sténose le plus souvent au niveau de l'isthme aortique.

incidence de la coarctation 4 pour 10.000 naissances vivantes soit 5–8% des enfants ayant une cardiopathie congénitale.

- diagnostic peut être tardif si bien que son incidence est sous estimée.
- L'évolution naturelle: très mauvaise
- espérance de vie d'une coarctation non opérée est de 35 ans et que 90% des patients meurent avant l'âge de 50 ans .(2)
-

Outcome of pregnancy in patients after repair of aortic coarctation

- Étude rétrospective
- 126 grossesses 54 femmes
- 26 grossesses (22%) compliquées par pathologie hypertensive
  - hypertension : 21 grossesses / 14 femmes
  - pre-eclampsie : 5 grossesses / 4 femmes

- Pas d ‘évènement grave type dissection ou AVC
- FCS : 18% (X2)
- Récurrence cardiopathie gauche 4-8% selon séries.

Joris W.J. Vriend, Wim Drenthene et al, , European Heart Journal (2005) 26, 2173–2178

Arrhythmia

- Arrhythmias requiring treatment develop in up to 15% of the patients with structural and congenital heart disease.
- In haemodynamically unstable patients with tachycardias direct cardioversion should be considered.
- Atrial flutter and atrial fibrillation are rare, prefer cardioversion after anticoagulation.
- Life-threatening ventricular arrhythmias during pregnancy are rare.

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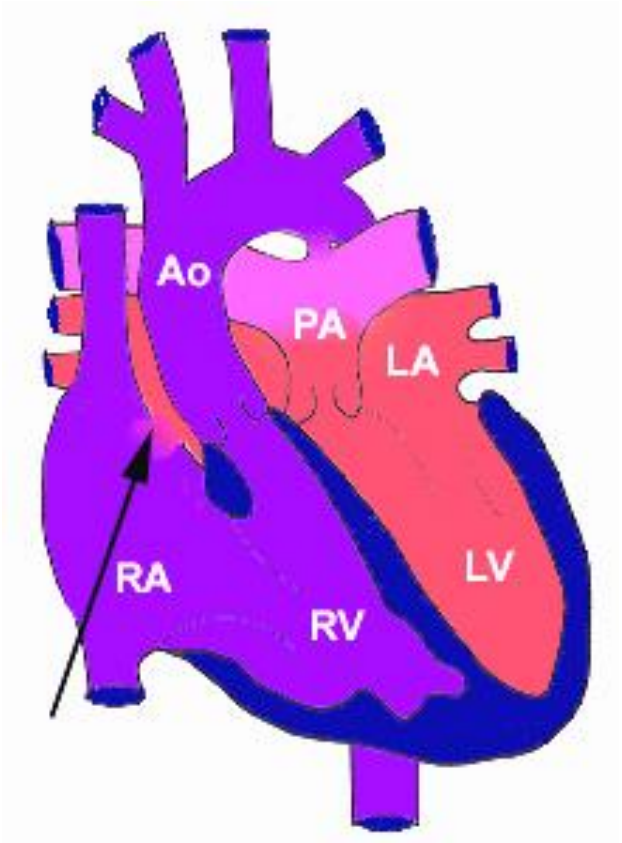
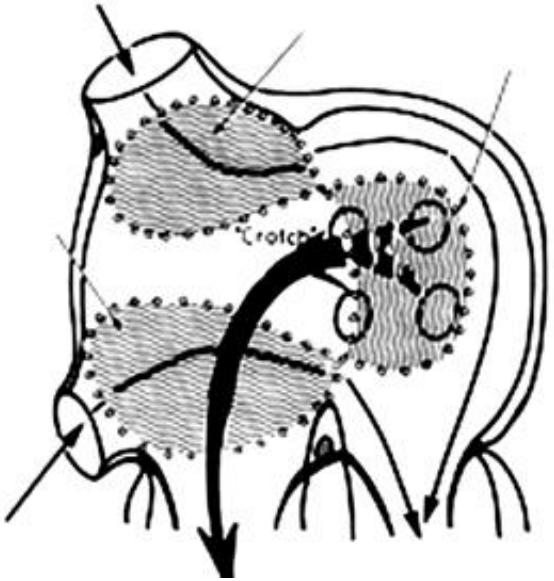
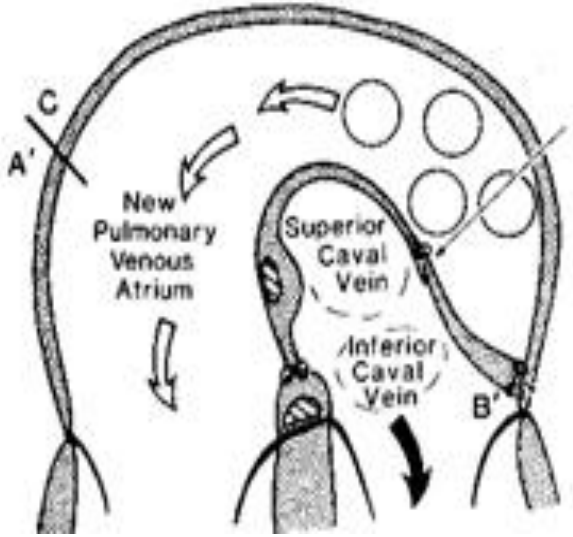


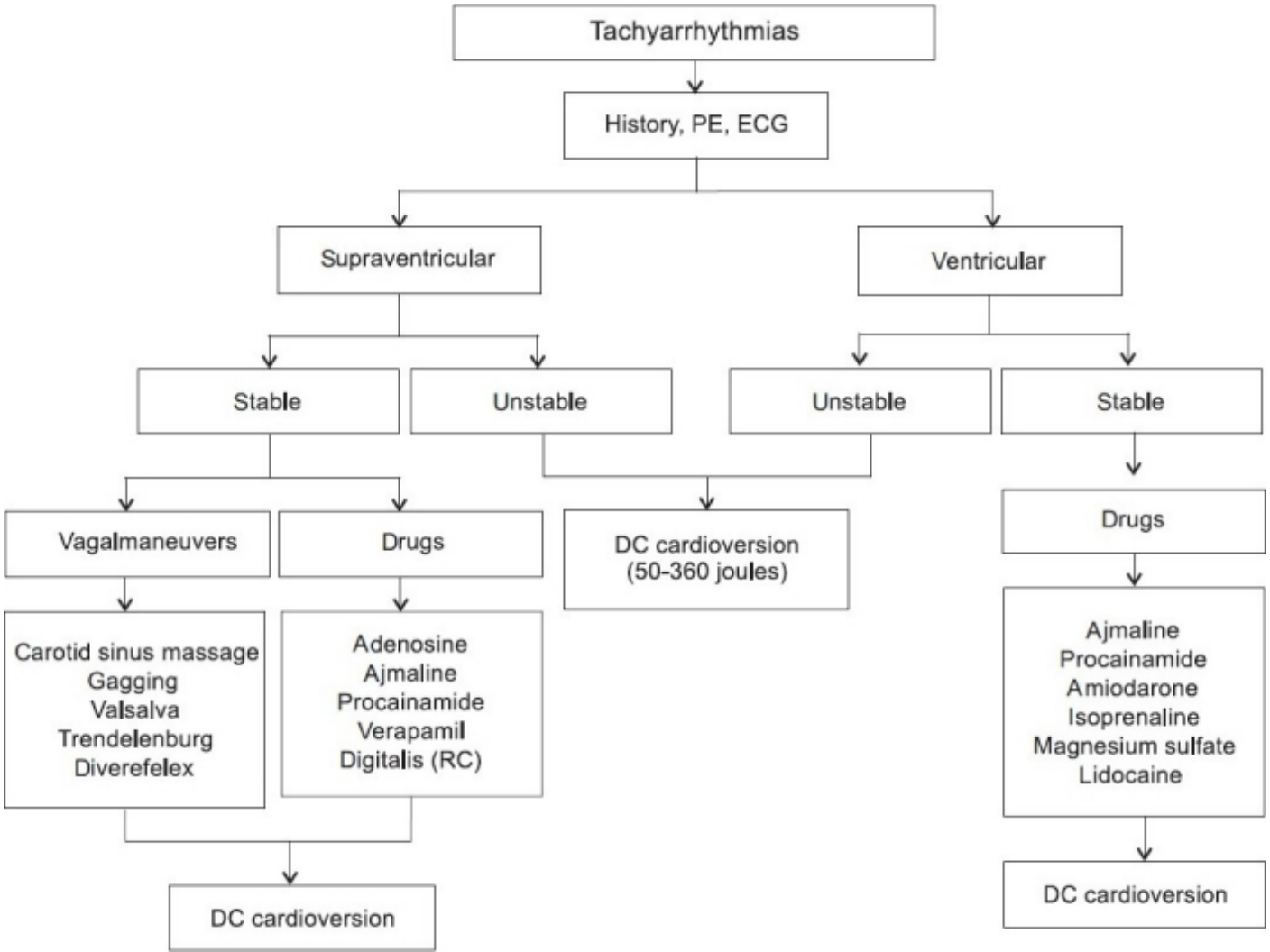
Mustard



Senning



Emergency management of atrial arrythmias



Drug therapy in pregnancy

Recommendations for drug use

Drug	Classification (Vaughan Williams for AA drugs)	FDA category	Placenta permeable	Transfer to breast milk (fetal dose)	Adverse effects
Abciximab	Monoclonal antibody with antithrombotic effects	C	Unknown	Unknown	Inadequate human studies; should be given only if the potential benefit outweighs the potential risk to the fetus.
Acenocoumarol	Vitamin K antagonist	D	Yes	Yes (no adverse effects reported)	Embryopathy (mainly first trimester), bleeding (see further discussion in Section 5 for use during pregnancy).
Acetylsalicylic acid (low dose)	Antiplatelet drug	B	Yes	Well-tolerated	No teratogenic effects known (large datasets).
Adenosine	Antiarrhythmic	C	No	No	No fetal adverse effects reported (limited human data).
Aliskiren	Renin inhibitor	D	Unknown	Unknown	Unknown (limited experience).
Amiodarone	Antiarrhythmic (Class III)	D	Yes	Yes	Thyroid insufficiency (9%), hyperthyroidism, goitre, bradycardia, growth retardation, premature birth.
Ampicillin, amoxicillin, cephalosporins, erythromycin, mezlocillin, penicillin	Antibiotics	B	Yes	Yes	No fetal adverse effects reported.

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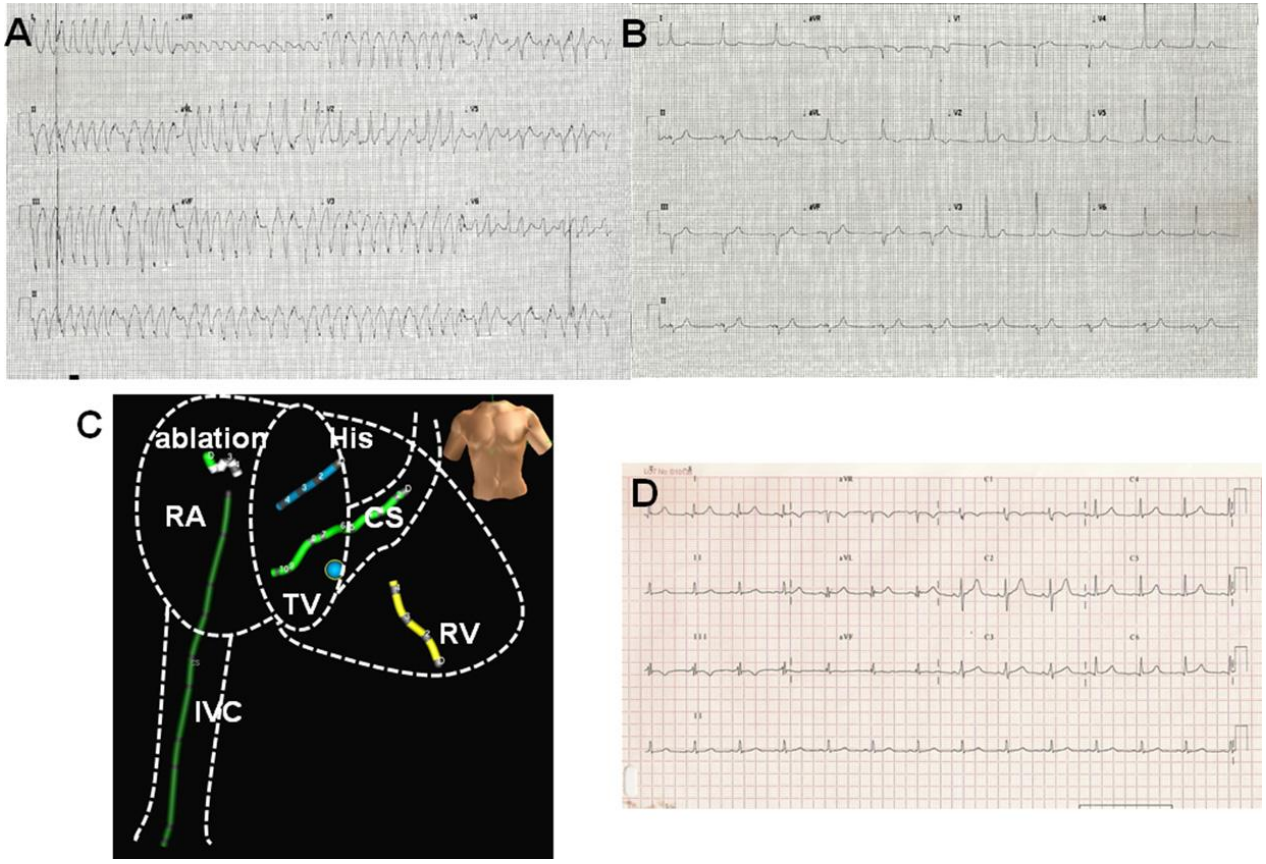
WPW ablation non-fluoroscopic 3D catheter navigation systems



European Journal of Obstetrics & Gynecology and Reproductive Biology, 2011 ZR Manjaly

Congenital Heart Diseases (CHD): Essential messages

- Women with CHD may tolerate pregnancy well.
  The risk depends on the underlying specific constellation.
- All patients with CHD should be seen by the end of the first trimester and an individualized follow up plan should be established.
- Vaginal delivery can be planned in most patients.
- Discuss high risk conditions, contraindications and indications for Caesarean delivery on an individual basis.

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Grossesse et endocardite

Incidence: jusqu ’à 7% des patientes

Bactériémies:

- 3,6%* à 7%** des accouchements

Situations obstétricales à haut risque:

- Rupture prématurés de la poche des eaux **
- Pyélonéphrite (traitement des bactériuries asymptomatiques)

->Prophylaxie pendant le travail si

- valve
- Atcd endocardite
- Cardiopathie cyanogène

° *Presbitero, Circul. 1994*

**Sugrue , Br Heart J. 1980*

***Tiossi, Arq Bras Cardiol 1994*

Risque de récurrence

- Descendance des cardiopathies majeurs : TGV, Fallot, CAV
- 727 individus, 393 nouveaux nés.
- Risque global de 4% (16 nn)

Lancet J Burn 1998

Pregnancy contraindications in cong. HD

- Women with pulmonary hypertension.
- Women with an oxygen saturation below 85% at rest.
- Patients with transposition of the great arteries and a systemic right ventricle with > moderate impairment of RV function and/or severe TR.
- Fontan patients with depressed ventricular function and/or moderate to severe atrioventricular valvular regurgitation or with cyanosis or with protein losing enteropathy.

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Congenital Heart Diseases (CHD): Essential messages

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complications Obstetriques et cardiaques : interactions fortes

- Hemorragie de pre partum
- pre-eclampsie
- Hemorragie du postpartum
- Mauvaise tolerance possible , même si atteinte obstetricale mineure (pas de réserve “myocardique”, pas de tolérance à une hypoxémie supplementaire)