

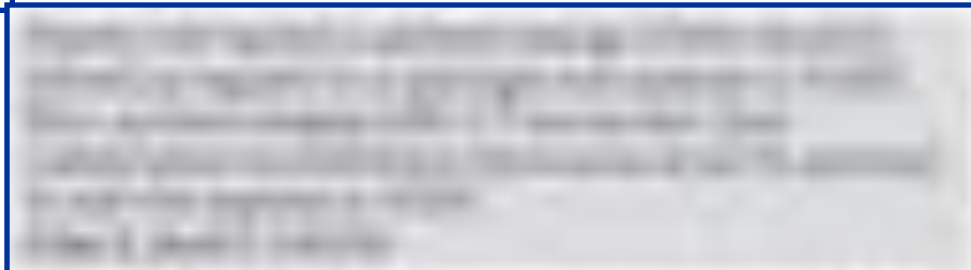
Tétralogie de Fallot



Suivi clinique

Follow-up recommendations

All patients with ToF should have periodic cardiac follow-up in a specialized GUCH centre, which in most patients should be done annually, but can be less frequent in those patients at the best end of the spectrum with minimal/stable haemodynamic disturbance. Follow-up evaluation needs to look for the complications listed above (see late clinical presentation). Echocardiography is performed as part of each visit. All patients should have CMR. The intervals for repeat studies depend on the pathology found.



Frequency of Routine Follow-Up and Testing	Physiological Stage A* (mo)	Physiological Stage B* (mo)	Physiological Stage C* (mo)	Physiological Stage D* (mo)
Outpatient ACHD cardiologist	12–24	12	6–12	3–6
ECG	24	12	12	12
TTE†	24	12–24	12	6–12
Pulse oximetry	As needed	As needed	Each visit	Each visit
Holter monitor	As needed	As needed	12–24	12–24
CMR‡/CCT§	36	24–36	12–24	12–24
Exercise test	36–60	24–60	12–24	12–24

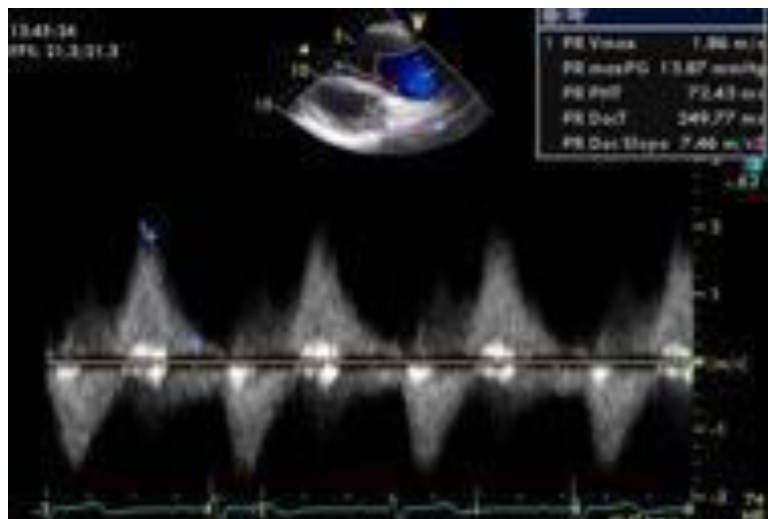
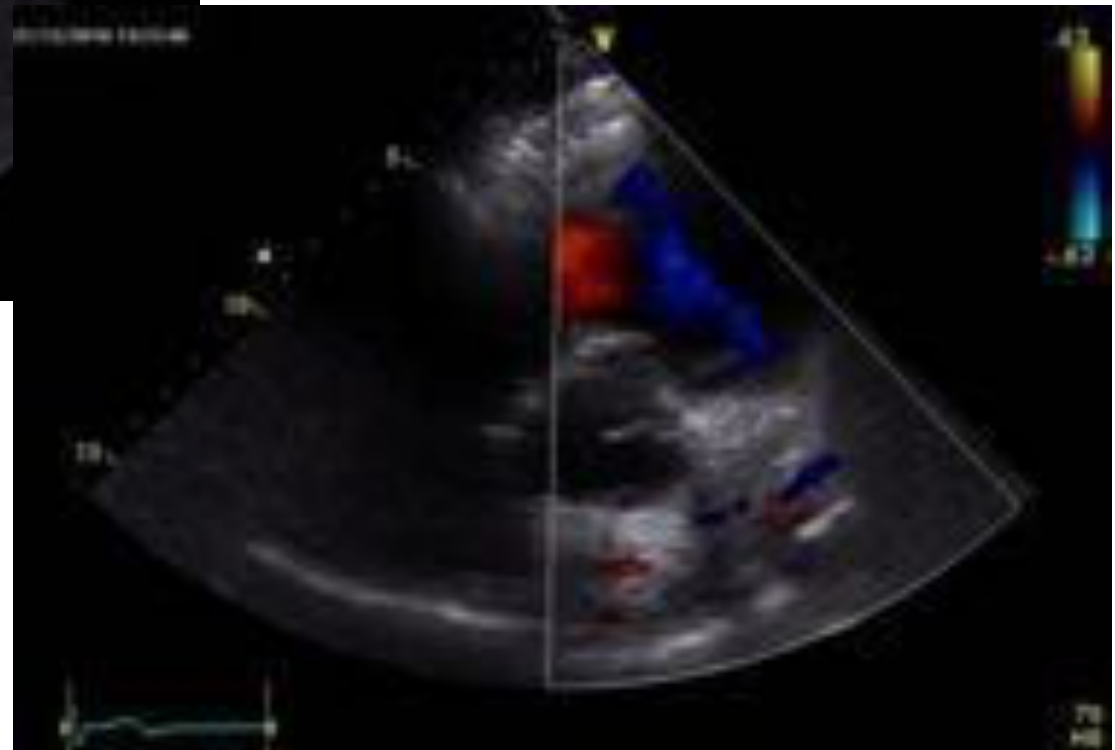
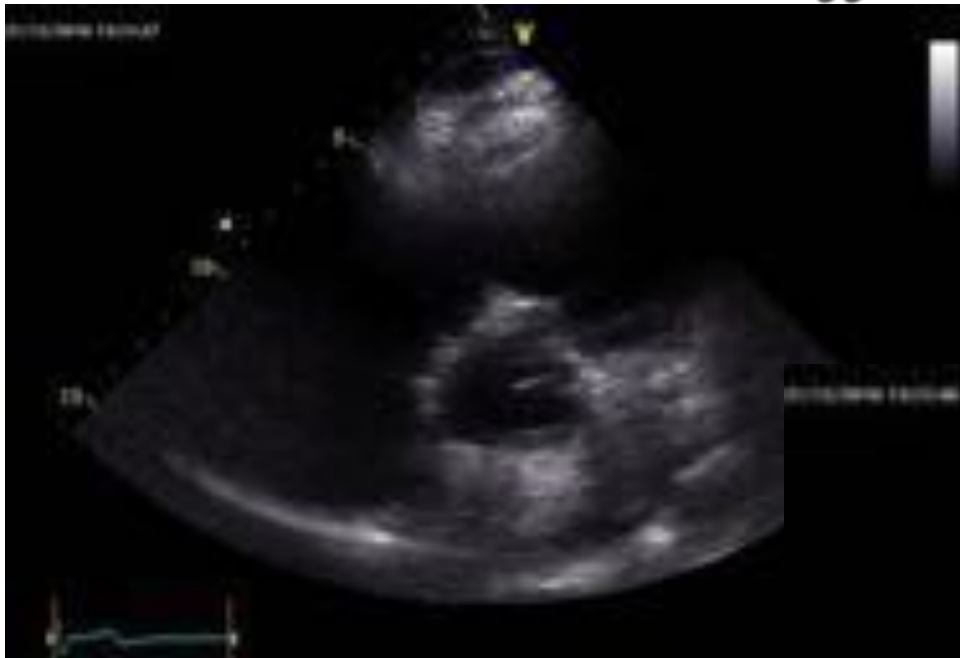
Baumgartner et al. *Eur Heart J* 2010

Warnes et al. *Circulation* 2008

Silversides et al. *Can J Card* 2010

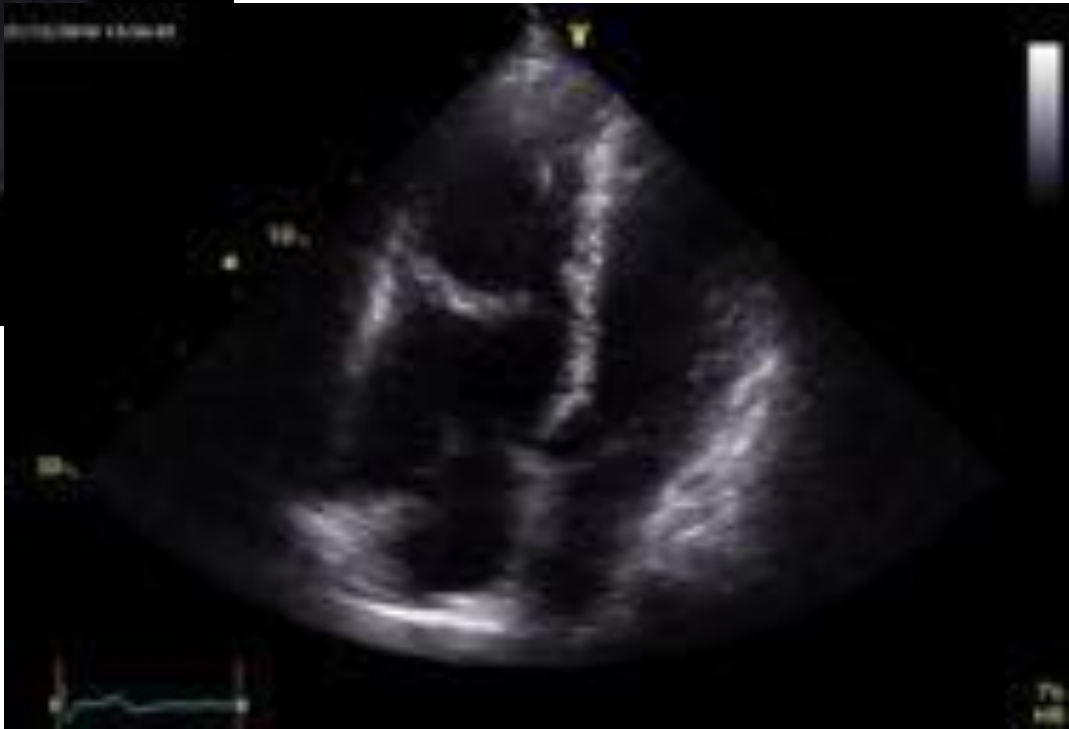
Suivi hémodynamique

Insuffisance pulmonaire



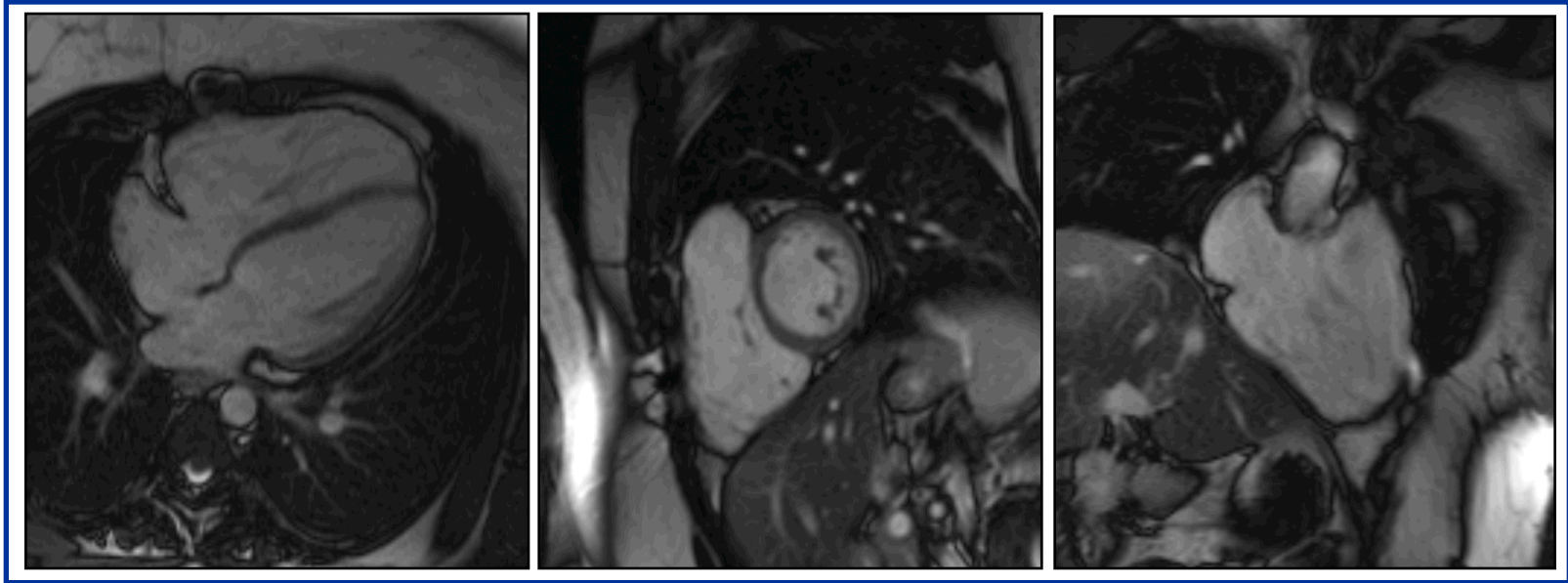
Suivi hémodynamique

Dilatation du VD (1)



Suivi hémodynamique

Dilatation du VD (2)



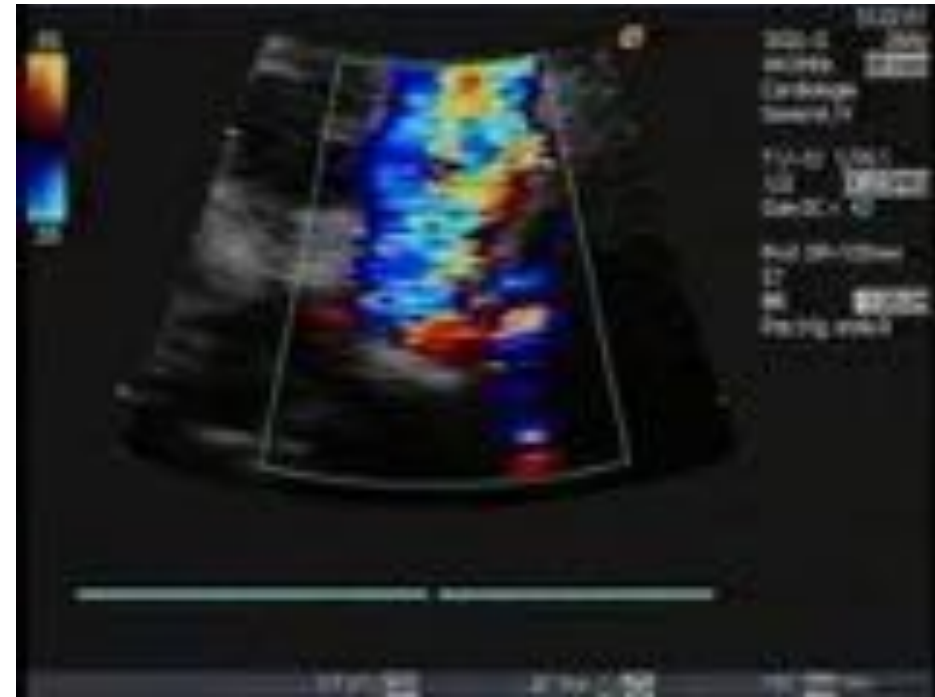
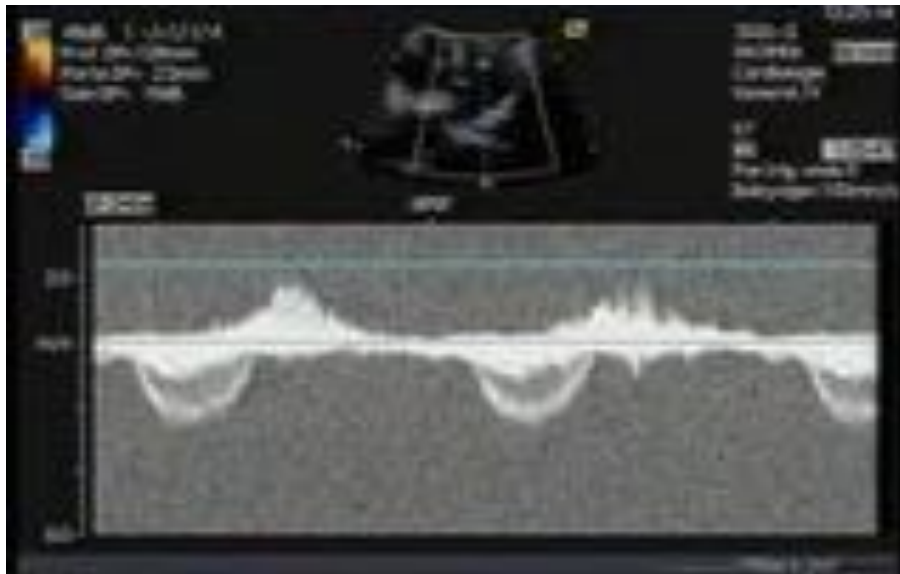
Normalisation post-opératoire du VD si

$VTDVD < 160 \text{ ml/m}^2$ et $VTSDVD < 80 \text{ ml/m}^2$



Suivi hémodynamique

Sténose des branches pulmonaires



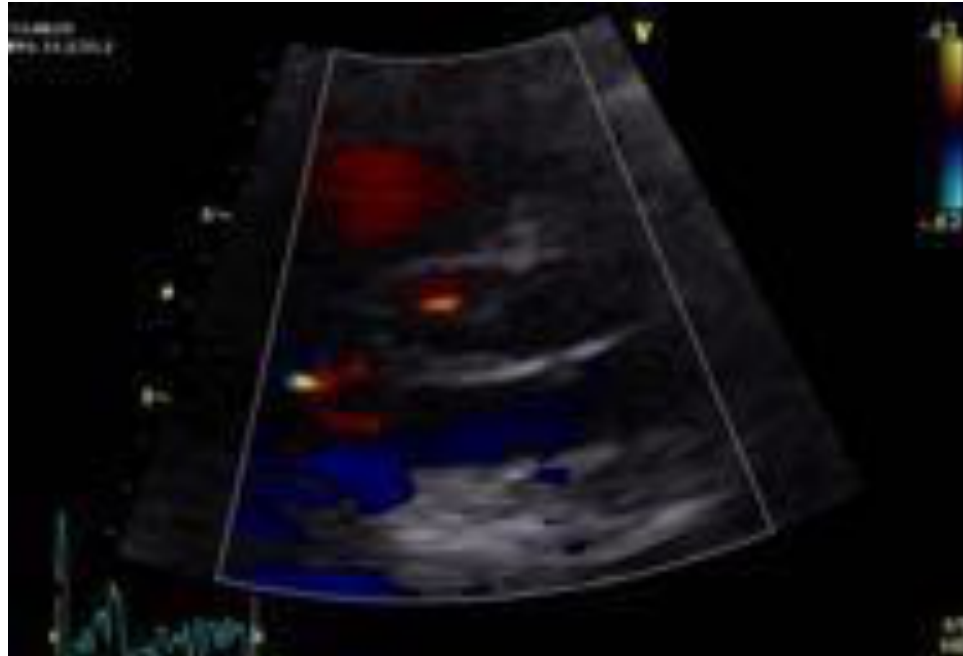
PVRep should be performed in symptomatic patients with severe PR and/or stenosis (RV systolic pressure >60 mmHg, TR velocity >3.5 m/s)

I

C

Suivi hémodynamique

CIV résiduelle



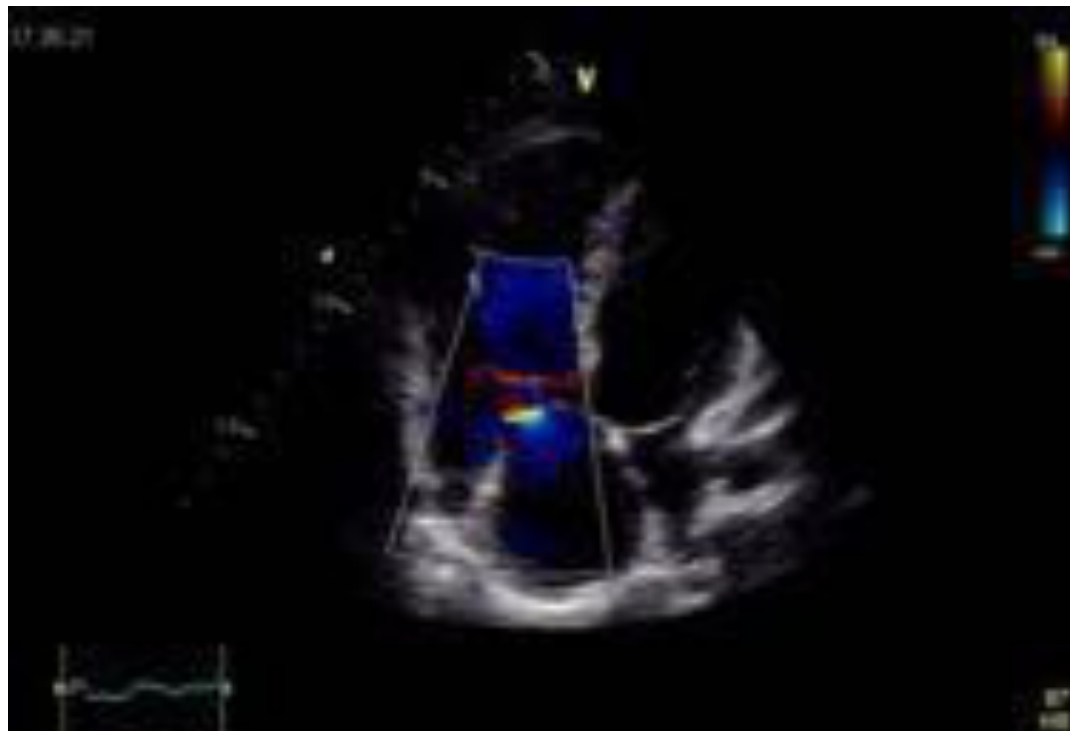
VSD closure should be considered in patients with residual VSD and significant LV volume overload or if the patient is undergoing pulmonary valve surgery

IIa

C

Suivi hémodynamique

Insuffisance tricuspide



PVRep should be considered in asymptomatic patients with severe PR and/or PS when at least one of the following criteria is present:

- Decrease in objective exercise capacity
- Progressive RV dilation
- Progressive RV systolic dysfunction
- Progressive TR (at least moderate)
- RVOTO with RV systolic pressure >80 mmHg (TR velocity >4.3 m/s)
- Sustained atrial/ventricular arrhythmias

IIa

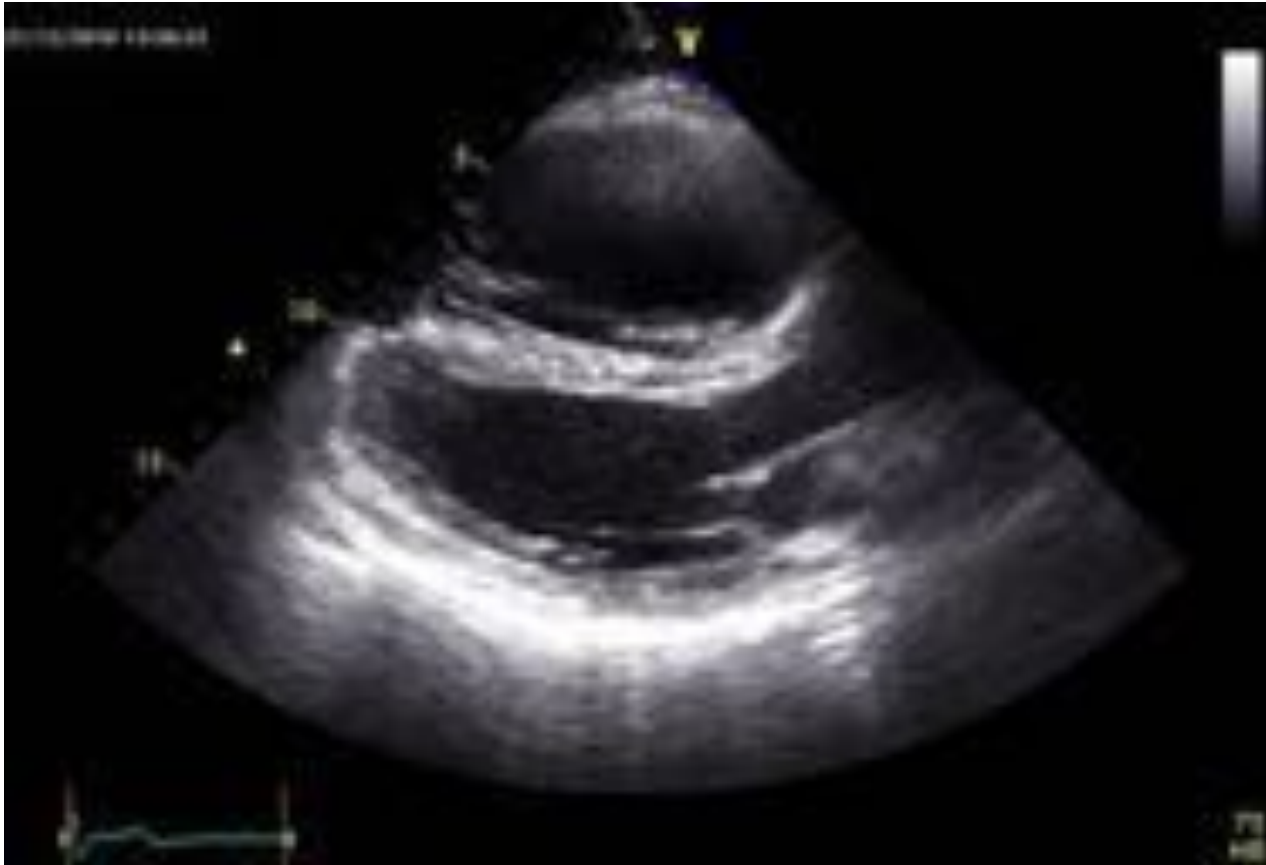
C

Suivi hémodynamique *Aortopathie*



The following statements were assessed as true or false:
 • Significant aortic regurgitation associated with aortic aneurysm progression is a contraindication to aortic dissection.
 • Aortic aneurysm progression is a contraindication to aortic dissection.
 • Aortic aneurysm progression is a contraindication to aortic dissection.

Suivi hémodynamique *FeVG*



+ strain longitudinal?

Diller et al. *Circulation* 2012

Suivi rythmologique

Troubles conductifs

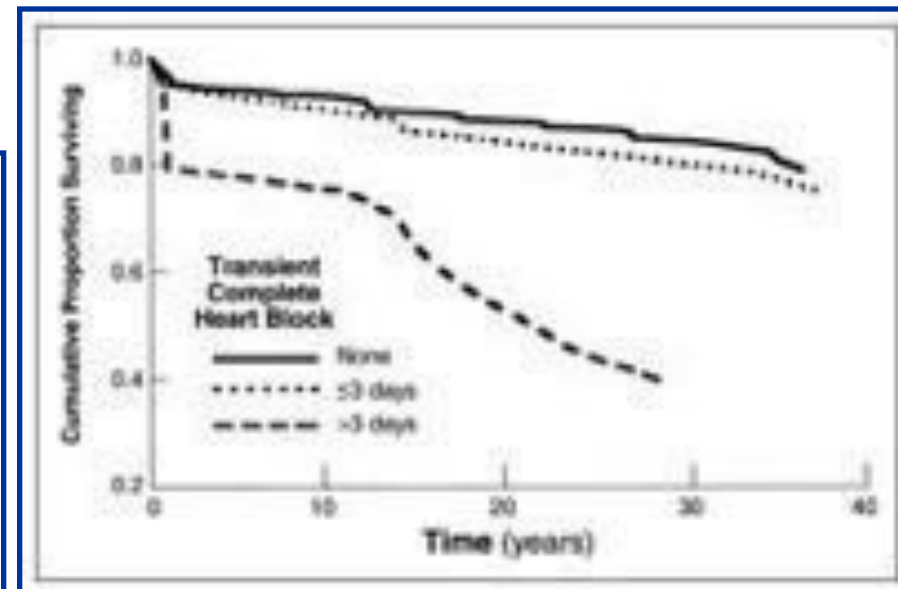
• Classiquement décrits:

- ✓ BBD 80-90% Friedli. *Arch Mal Cœur Vaiss* 1996
- ✓ BBD+HBAG 15-20% Friedli. *Arch Mal Cœur Vaiss* 1996
- ✓ BAVc transitoire ou permanent $\leq 1\%$ Friedli. *Pediatr Cardiol* 1999
- ✓ Dysfonction sinusale rare Friedli. *Pediatr Cardiol* 1999

• Quelle signification?

Facteurs prédictifs de *BAVc* tardif:

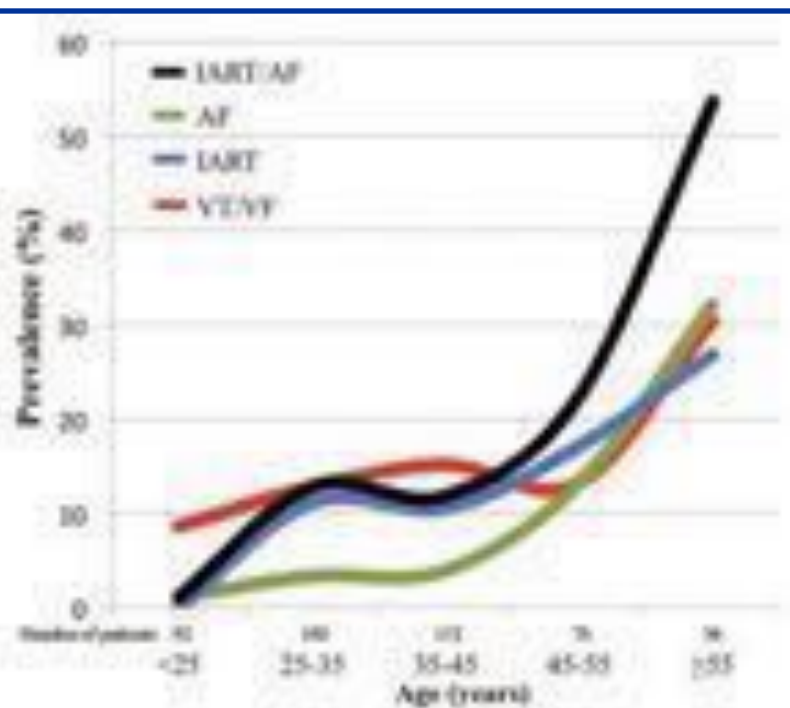
- ✓ BAVc post-opératoire transitoire
- ✓ BBD+HBAG+BAV1
- ✓ PW pour des fréquences stimulées $< 180/\text{min}$



Suivi rythmologique

Arythmies supraventriculaires

Characteristic	Prevalence, %	95% CI
Sustained tachyarrhythmia	29.9	26.2–33.7
Atrial tachyarrhythmia	20.1	17.0–23.6
IART	11.5	9.0–14.3
AF	7.4	5.4–9.7
Other	6.7	4.8–8.9
Ventricular tachyarrhythmia	14.6	11.8–17.7



Variable	OR	95% CI	P
IART			
Prior cardiac surgeries, n	1.4	1.2–1.6	<0.001
Hypertension, %	2.3	1.1–4.6	0.022
Right atrial enlargement, %	6.2	2.8–13.6	<0.001
AF			
LV ejection fraction, %	0.93	0.89–0.98	<0.001
Age, y	1.09	1.05–1.12	<0.001
Prior cardiac surgeries, n	1.5	1.2–1.9	<0.001
Left atrial enlargement, %	3.2	1.5–6.8	0.003

Suivi rythmologique

Arythmies ventriculaires/Mort subite

Characteristic	Prevalence, %	95% CI
Ventricular tachyarrhythmia	14.6	11.8–17.7
VT	14.2	11.5–17.3
VF	0.5	0.1–1.4

Khairy et al. *Circulation* 2010

Incidence annuelle de mort subite <0.2%

Intérêt de *stratifier* le risque rythmique

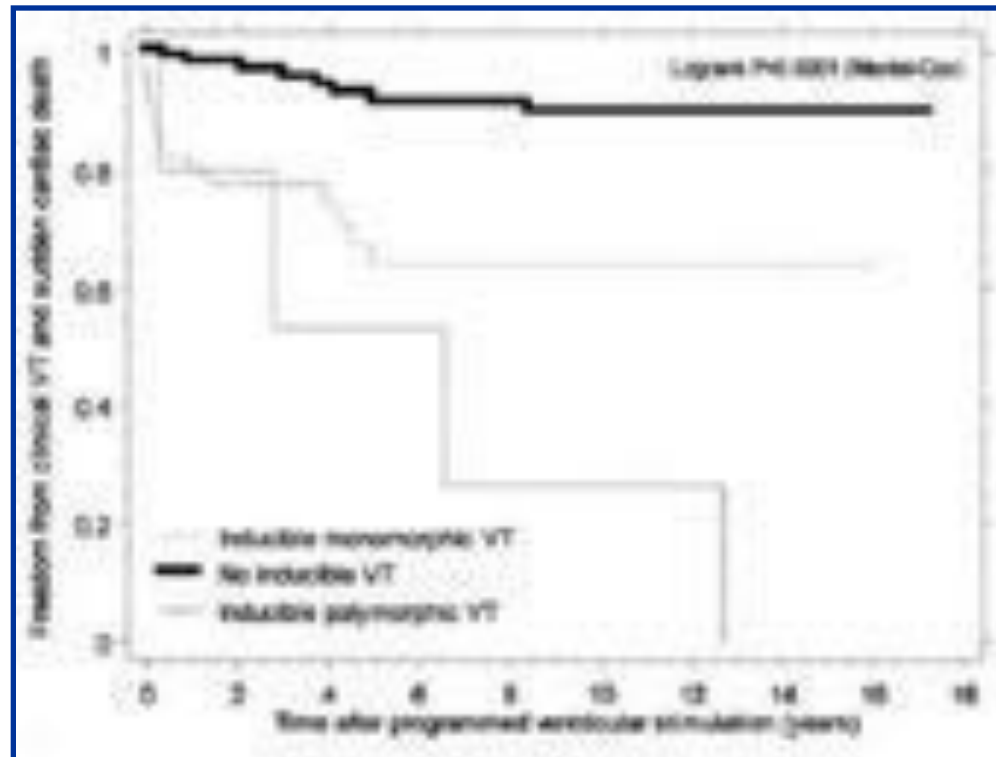
Prévention 2ndaire → DAI

Prévention 1aire → ?

FDR non invasifs

- Histoire de la cardiopathie
 - ✓ Age tardif à la cure complète, long suivi (Gatzoulis et al. *Lancet* 2000)
 - ✓ Shunt palliatif antérieur
 - ✓ Ventriculotomie/Patch transannulaire (Dietl et al. *Circulation* 1994)
- Données électrophysiologiques
 - ✓ QRS \geq 180 msec (VPN 100%, Se 94.7%) (Gatzoulis et al. *Circulation* 1995)
 - ✓ TVNS prédictive de TV clinique (Khairy et al. *Circulation* 2008) et de TV inducible (Khairy et al. *Circulation* 2004)
 - ✓ Fragmentation des QRS? (Egbe et al. *J Am Heart Assoc* 2018)
 - ✓ TSV (Valente et al. *Heart* 2014)
- Données hémodynamiques
 - ✓ Dysfonction systolique (Ghai et al. *JACC* 2002) et diastolique (Khairy et al. *Circulation* 2008) du VG, altération fonction longitudinale VG (Diller et al. *Circulation* 2012)
 - ✓ IP sévère (Gatzoulis et al. *Lancet* 2000), dilatation VD (D'Alto et al. *Heart* 1999), dysfonction VD (Knauth et al. *Heart* 2008), hypertension VD (Katz et al. *Circulation* 1982), anévrisme CCVD (Harrison et al. *JACC* 1997), réhaussement tardif (Babu-Narayan et al. *Circulation* 2006) → IRM

Facteurs prédictifs de TV inducible: âge ≥ 18 ans, palpitations, shunt palliatif antérieur, TVNS et ICT ≥ 0.6



	Sustained Monomorphic VT	Sustained Monomorphic or Polymorphic VT
Sensitivity, %	66.1 ± 6.0	77.4 ± 5.3
Specificity, %	81.6 ± 2.8	79.5 ± 2.9
Diagnostic accuracy, %	77.8 ± 2.6	79.0 ± 2.6
Positive predictive value, %	53.9 ± 5.7	55.2 ± 5.3
Negative predictive value, %	88.1 ± 2.4	91.5 ± 2.2

Suivi rythmologique

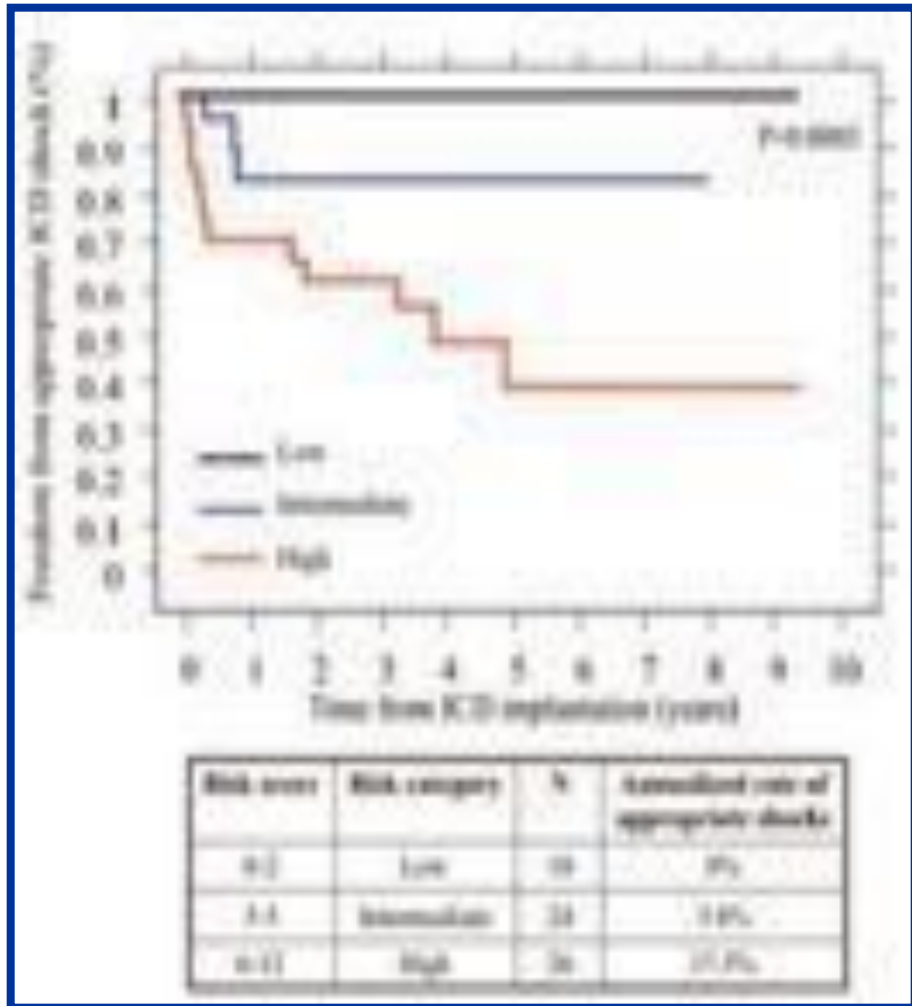
Mort subite/Stratification du risque (1)

Table 3. Risk Score for Appropriate ICD Shocks in Primary Prevention

Variable	Exp(β)	Points Attributed
Prior palliative shunt	3.2	2
Inducible sustained ventricular tachycardia	2.6	2
QRS duration ≥ 180 ms	1.4	1
Ventriculotomy incision	3.4	2
Nonsustained ventricular tachycardia	3.7	2
LVEDP ≥ 12 mm Hg	4.9	3
Total points	...	0–12

29.8% de complications

24.8% de chocs inappropriés



Khairy et al. *Circulation* 2008

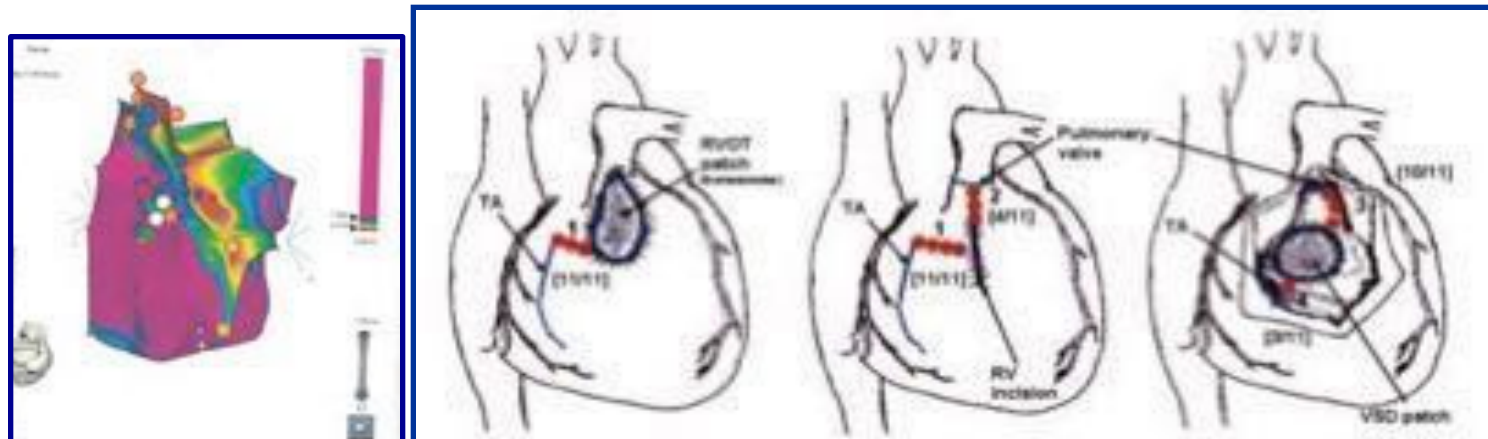
Egbe et al. *Heart* 2018



Ablation endocavitaire?

- Class I Catheter ablation is indicated as adjunctive therapy to an ICD in adults with CHD and recurrent monomorphic ventricular tachycardia, a ventricular tachycardia storm, or multiple appropriate shocks that are not manageable by device reprogramming or drug therapy (*Level of evidence: C*).^{94,320}
- Class IIa Catheter ablation can be considered for symptomatic sustained monomorphic ventricular tachycardia in adults with CHD and ICDs as an alternative to drug therapy (*Level of evidence: B*).^{215,306}

Khairy et al. *Heart Rhythm* 2014



Zeppenfeld et al. *Circulation* 2007

- Selon modalités chirurgicales
- TV monomorphe le plus souvent
- Ablation efficace

Zeppenfeld et al. *Card Clin Electrophysiol* 2017

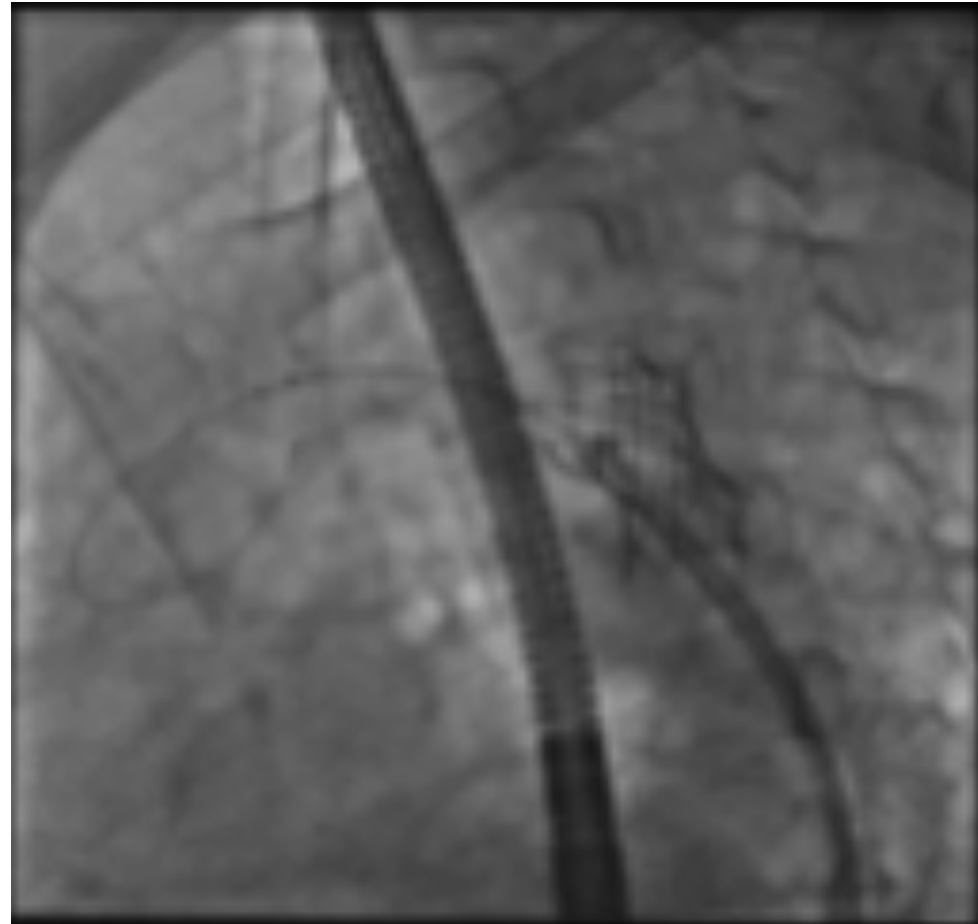
- Coarctation aortique
- Cure chirurgicale à l'âge de 11 ans
- Bicuspidie aortique saine
- HTA sous trithérapie et dyspnée d'effort progressive
- PA 152/95 mmHg, BDC réguliers, SS 2/6 RSG.

Que faites-vous?

- Reconduction de l'ordonnance de traitements anti-HTA
- Un ECG
- Je palpe ses pouls fémoraux
- Je vérifie que la PA est prise au MSG
- Explorations complémentaires (dont imagerie aortique)
- Une ETT

Angioplastie de recoarctation (stent couvert):

Gradient pic à pic: 22 → 3 mm Hg



Diminution des traitements anti-HTA...