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.

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

*See Tables 3 and 4 for details on the ACHD AP classification system.

†Routine TTE may be unnecessary in a year when CMR imaging is performed unless clinical indications warrant otherwise.

‡CMR may be indicated for assessment of right ventricular size and function, pulmonary valve function, pulmonary artery anatomy and left heart abnormalities. Baseline study is recommended with periodic follow-up CMR, with frequency of repeat imaging determined by anatomic and physiological findings.

§CCT may be used if CMR is not feasible and to evaluate origin and course of the coronary arteries, and cross-sectional imaging status–post-stent therapy. If cardiac CCT is used instead of CMR imaging, the frequency should be weighed against radiation exposure.

║6-minute walk test or CPET, depending on clinical indication.

ACHD indicates adult congenital heart disease; CCT, cardiac computed tomography; CMR, cardiovascular magnetic resonance imaging; CPET, cardiopulmonary exercise test; ECG, electrocardiogram; TOF, tetralogy of Fallot; and TTE, transthoracic echocardiogram.
Suivi hémodynamique

Insuffisance pulmonaire
Suivi hémodynamique

Dilatation du VD (1)
Suivi hémodynamique

Dilatation du VD (2)

Normalisation post-opératoire du VD si

\[ VTDVD < 160 \text{ ml/m}^2 \text{ et } VTSVD < 80 \text{ ml/m}^2 \]

Buechel et al. *Eur Heart J* 2005
Oosterhof et al. *Circulation* 2007
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)

Baumgartner et al. *Eur Heart J* 2010
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

Baumgartner et al. Eur Heart J 2010
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

Baumgartner et al. Eur Heart J 2010
Suivi hémodynamique
Aortopathie

Silversides et al. *Can J Card* 2010
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 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/min**

Friedli et al. JACC 1988
Hokanson et al. Am J Cardiol 2001
Suivi rythmologique
Arythmies supraventriculaires

### Tableau des arythmies supraventriculaires

<table>
<thead>
<tr>
<th>Caractéristique</th>
<th>Prévalence, %</th>
<th>CI 95%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sustained tachyarrhythmia</td>
<td>29.9</td>
<td>26.2–33.7</td>
</tr>
<tr>
<td>Atrial tachyarrhythmia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>IART</td>
<td>11.5</td>
<td>9.0–14.3</td>
</tr>
<tr>
<td>AF</td>
<td>7.4</td>
<td>5.4–9.7</td>
</tr>
<tr>
<td>Other</td>
<td>6.7</td>
<td>4.8–8.9</td>
</tr>
<tr>
<td>Ventricular tachyarrhythmia</td>
<td>14.6</td>
<td>11.8–17.7</td>
</tr>
</tbody>
</table>

### Graphique des arythmies supraventriculaires

#### Tableau des variables

<table>
<thead>
<tr>
<th>Variable</th>
<th>OR</th>
<th>CI 95%</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prior cardiac surgeries, %</td>
<td>1.4</td>
<td>1.2–1.6</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Hypertension, %</td>
<td>2.3</td>
<td>1.1–4.6</td>
<td>0.022</td>
</tr>
<tr>
<td>Right atrial enlargement, %</td>
<td>6.2</td>
<td>2.8–13.6</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>LV ejection fraction, %</td>
<td>0.93</td>
<td>0.89–0.96</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Age, y</td>
<td>1.09</td>
<td>1.05–1.12</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Prior cardiac surgeries, %</td>
<td>1.5</td>
<td>1.2–1.9</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Left atrial enlargement, %</td>
<td>3.2</td>
<td>1.5–6.8</td>
<td>0.003</td>
</tr>
</tbody>
</table>

Khairy et al. Circulation 2010
## Suivi rythmologique

**Arythmies ventriculaires/Mort subite**

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Prevalence, %</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular tachyarrhythmia</td>
<td>14.6</td>
<td>11.8–17.7</td>
</tr>
<tr>
<td>VT</td>
<td>14.2</td>
<td>11.5–17.3</td>
</tr>
<tr>
<td>VF</td>
<td>0.5</td>
<td>0.1–1.4</td>
</tr>
</tbody>
</table>

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≥180 msec (VPN 100%, Se 94.7%) (Gatzoulis et al. Circulation 1995)
  - Fragmentation des QRS? (Egbe et al. J Am Heart Assoc 2018)
  - TSV (Valente et al. Heart 2014)

- Données hémodynamiques
Facteurs prédictifs de TV inducible: âge ≥ 18 ans, palpitations, shunt palliatif antérieur, TVNS et ICT ≥ 0.6
Suivi rythmologique
Mort subite/Stratification du risque (1)

29.8% de complications
24.8% de chocs inappropriés

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

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

Khairy et al. Circulation 2008
Egbe et al. Heart 2018
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).\textsuperscript{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).\textsuperscript{215,306}

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

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Khairy et al. *Heart Rhythm* 2014

Zeppenfeld et al. *Circulation* 2007

Zeppenfeld et al. *Card Clin Electrophysiol* 2017
Mr Claude R, ddn 08/12/1952

- 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: $22 \rightarrow 3$ mm Hg

Diminution des traitements anti-HTA...