Arrhythmias after Fallot Repair

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Pôle de cardiologie pédiatrique et congénitale
Arrhythmias factors and Fallot patients

1) Factor associated with hemodynamic status before correction
   - hypoxemia
   - high output
   - pressure overload

2) Factor associated with surgery:
   - suture lines
   - patches
   - direct trauma of conducting system
   - residual lesions

3) Cardiovascular risk factors during life:
   - HBP, diabetes mellitus, tobacco use, dyslipidemia, SAS, aging

Substrate for arrhythmias:
   - Reentry +++

Genetic factors?
   - TBX5
   - NKX 2.5
   - Long QT genes

Goldmuntz E et al. Circulation 2001
Chiu SN et al. Int J Cardiol 2017:249:156

Hypertrophy
Dilatation
Ischemia
Electrical dyssynchrony

Fibrosis
Arrhythmias up to 30% of Fallot patients with long follow up
Sustained ventricular arrhythmias up to 10%
Sudden cardiac death incidence 0.2%/year

Sudden cardiac death related to:
1) Ventricular tachycardias
2) Rapid conducted supraventricular tachycardias
3) (acute AV block)

AV block essentially in the early post operative period (0.6% in the modern area)

Increasing rate of supraventricular tachycardia with age changing from organized tachycardia to atrial fibrillation

Interventionnal arrhythmia therapy in 20.4%
In adults, frequent association of multiple arrhythmias

Association with mortality and heart failure

Mouws EM et al. *Heart Rhythm* 2018;15:503
High incidence of atrial fibrillation, compared to control population

Mandalenakis Z et al. *Circulation* 2018;137:928
How to change this arrhythmia profile?

Changing surgical management?

- Early complete correction. Ideal timing?
- Increase cardiac protection, decrease aortic cross-clamp time?
- Limit ventricular incisions?
- Limit patches (indundibular)?
- Pulmonary valve sparing techniques to decrease redo procedures?
- Incisions orientations guided by potentials tachycardia isthmuses in infants?
- Arrhythmia prophylactic surgery in adults?
- Earlier redo surgery for residual lesions/pulmonary regurgitation?

Aggressive management of lifestyle and cardiovascular risk factors?
Stimulation in Fallot

In cases of:

- Complete AV block: permanent/paroxysmal
- (Symptomatic sinus node dysfunction)
- Cardiac resynchronisation:
  . NYHA ≥ II, QRS > 150 ms (or > 120 ms with LBBB pattern), LV EF ≤ 35%
  . Indication for permanent ventricular stimulation and LV EF < 40%

How to stimulate?

Khairy P et al. Heart Rhythm 2014;11:e102
Epicardial stimulation for:

- Children to spare venous access
- Patients with residual shunts to avoid cardiac emboli

Mode of stimulation:

- Single left ventricular lead in infants with good haemodynamics
  - Atrial and ventricular leads in other cases

Contemporary problems:

- 30% lead failure at 10 years
- High threshold stimulation
- Difficult epicardial stimulation in patients with redo surgeries
  - Devices related infections (endocardial ++)

Leadless pacemakers?
Long life batteries?

MacLeod KA. *Heart* 2010;96:1502
Khairy P et al. *Circulation* 2006;113:2391
Kelvin CL et al. *Heart rhythm* 2015;12:566
Cardiac resynchronisation in Fallot patients?

Improvement similar to cardiomyopathies with low LV EF with large QRS
But short series and follow-up

Koyak Z et al. Europace 2018

Merchant FM et al. Pacing Clin Electrophysiol 2014
Thambo JB et al. Int J Cardiol 2013
Thambo JB et al. Heart Rhythm 2010
RV resynchronisation?

Possible haemodynamic improvement with stimulation of the most delayed RV wall in fusion with normal ventricular activation

In selected patients
A need for multicentric and long term follow-up

Janousek J et al. Circ Arrhythm Electrophysiol 2019
Early postoperative period

Junctional ectopic tachycardia (JET) the most frequent arrhythmia but rare between 5 and 6% of patients

Association with younger age, longer aortic cross-clamping/by-pass time
Tendency to increase mortality
No evolution to complete AV block

AV conduction tissue inflammation/trauma

Dodge K et al. *Thorac Cardiovasc Surg* 2002;123:624
Supraventricular arrhythmias

Late postoperative period

Arrhythmias from right atrium with a majority of macro-reentries
(++) cavo-tricuspid isthmus reentry ie common flutter
Most of the time first arrhythmia after adolescence

Supra ventricular arrhythmias most of the time around scars/patches suture lines

Clear goal for ablation
Good success rate of percutaneous ablation with contemporary tools (>90% acute success, 15% of recurrences)

Contemporary issues:
- Early detection to decrease related heart failure and mortality
- How to decrease incidence?

Large and early indication with avoidance of medical treatment
Question of thromboembolic prevention in cases of supraventricular arrhythmias

Starting age? NOAC? VKA? Other drug? Duration after ablation?

Khairy P et al. Heart Rhythm 2014;11:e102
Atrial fibrillation: the last frontier

No sufficient data in CHD patients
Pranata R et al. Indian Pacing Electrophysiol J 2019;epub

Clear substrate? Right sided? Left sided with pulmonary veins?

Haemodynamic and common risk factors optimization +++
Poor efficacy and tolerance of anti-arrhythmic drugs

Percutaneous ablation with pulmonary veins isolation?
Simple
Low complication rate
Efficacy < 50%

Surgical treatment with Cox-Maze III?
Variable efficacy between 30 and 80%
High complication rate
Need for cardiac stimulation > 10%

Hybrid ablation?

Deal BJ, Mavroudis C.
Card Electrophysiol Clin 2017;9:329
### Ventricular arrhythmias

Risk factors for ventricular tachycardia/sudden cardiac death

<table>
<thead>
<tr>
<th>Standard clinical variables</th>
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</thead>
<tbody>
<tr>
<td>Older age at time of repair</td>
</tr>
<tr>
<td>Prior large palliative shunts</td>
</tr>
<tr>
<td>Older chronologic age</td>
</tr>
<tr>
<td>Recurrent syncope</td>
</tr>
<tr>
<td>Pulmonary regurgitation</td>
</tr>
<tr>
<td>Residual pulmonary stenosis</td>
</tr>
<tr>
<td>Severe RV enlargement</td>
</tr>
<tr>
<td>Depressed RV function</td>
</tr>
<tr>
<td>Depressed LV function</td>
</tr>
<tr>
<td>High-grade ventricular ectopy on Holter or exercise test</td>
</tr>
<tr>
<td>Prolonged QRS duration on electrocardiogram (&gt;180 ms)</td>
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<th>Advanced testing</th>
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<tbody>
<tr>
<td>Positive ventricular stimulation at electrophysiology study</td>
</tr>
<tr>
<td>Large RV size on CMR</td>
</tr>
<tr>
<td>Large pulmonary regurgitant fraction on CMR</td>
</tr>
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**CMR** = cardiac magnetic resonance; **LV** = left ventricular; **RV** = right ventricular.


strong predictive negative accuracy but only fair predictive positive accuracy
No clear consensus for ventricular arrhythmias risk stratification in these patients

<table>
<thead>
<tr>
<th>Variable</th>
<th>Exp(j)</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>


Utility of invasive electrophysiological study for intermediate risk?

With MRI

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<th>Variable</th>
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<td>Prior palliative shunt</td>
<td>2</td>
</tr>
<tr>
<td>QRS duration ≥180 ms</td>
<td>1</td>
</tr>
<tr>
<td>Ventriculotomy incision</td>
<td>2</td>
</tr>
<tr>
<td>Previous VT</td>
<td>2</td>
</tr>
<tr>
<td>LV EF &lt;45%</td>
<td>2</td>
</tr>
<tr>
<td>RV EF &lt;30%</td>
<td>3</td>
</tr>
<tr>
<td>Total points</td>
<td>0-12</td>
</tr>
</tbody>
</table>

Bokma JP et al. *JAMA Cardiol* 2017;2:618

Cohort analysis: low arrhythmias rate in primary prevention with this approach

Probst J et al. *Int J Cardiol* 2018;15:269
ICD in Fallot patients

Secondary prevention: aborded SCD, VT with haemodynamic compromise

Primary prevention???

Problem of complication rate +++
- Technical problem (++ lead revision): up to 30% after 4 years
- Inappropriate therapies: up to 6% after several years

DAIT4F French registry in process
Decrease complication rate of ICD with subcutaneous ICD?

Actually no stimulation for bradycardia / anti-tachycardia pacing
Caution with low weights < 25 kgs
Risk of inappropriate shocks in young patients

Moore JP et al. Circ Arrhythm electrophysiol 2016;9
Mecanisms of ventricular arrhythmias in Fallot Patients

VT isthmuses are now well known
Most frequent : isthmus 3

Zappenfeld K et al. Circulation 2007;116:2241-2252
Kapel GF et al. Circ Arrhythm Electrophysiol 2015;8:102-109
Utility of electrophysiologic study in the management of ventricular arrhythmias

Precise substrate location

Scar / patches location
Slow conducting channel location
Activation map during VT

Acute success rate 90%
between 10 and 20% of recurrences

Zeppenfeld K et al. Card Electrophysiol Clin 2018

VT ablation and block lines validation

Zeppenfeld K et al. Circulation 2007;116:2241-52
Kriebel T et al. Am Coll Cardiol 2007;50:2162-68
Kappel GF et al. Circ Arrhythm Electrophysiol 2015;8:102-9
Van Zyl M et al. Heart Rhythm 2016;13:1449
Causes of percutaneous ablation failure

Valvular prosthesis / homografts

Conduction tissue proximity

Local Hypertrophy, calcifications / epicardial source

Kapel GF et al. Circ Arrhythm electrophysiol 2014;7:889
Causes of percutaneous ablation failure

Caution with percutaneous pulmonary valves with possible arrhythmia « protected isthmus »

Discussion of arrhythmia evaluation before surgery in adults ++

Combes N. et al. Eur Heart J 2019;epub
Ventricular arrhythmias surgical treatment

With redo surgery with primary haemodynamic indication

Ideally:

- guided by electrophysiological study before isthmuses location
- With lesion control for achievement of complete block of conduction

Indication:

- Secondary prevention
- Primary prevention in selected patients?
  - Systematically?

When perform EP study in patients referred for pulmonary valve intervention?

Rotes AS et al. Circ Arrhythm Electrophysiol 2015;8:110-16
Conclusion

A life-long burden of arrhythmias in Fallot patients

An increasing knowledge of arrhythmias mechanisms

An increasing efficacy of interventional treatment with ablation therapies

A lot of improvement needed

- Early detection of arrhythmias
  - Complications of PMK / ICD
  - A better assessment of SCD risk

Multidisciplinary staffs with congenital cardiologists, cardiac surgeons, electrophysiologists to increase arrhythmia prognosis in CHD patients