Tetralogy of Fallot in Adults

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Introduction and Background

» TOF is the most common form of cyanotic defect, accounting for up to 10% of all CHD.

» Evolution of surgical technique, intensive care, nursing and medical management has led to a marked increase in the numbers of patients surviving into adulthood (Tutarel O, Kempney A et al, 2013)
Anatomy and Physiology

Normal heart

Tetralogy of Fallot

- Overriding aorta
- Pulmonic stenosis
- Ventricular septal defect
- Right ventricular hypertrophy
Palliation and later repair

BT shunt to increase blood flow to the lungs:

Allows for growth of PA's and child before complete repair at later date

Nursing Considerations in Adults: Original BT shunts attached subclavian artery to PA - use opposite arm for accurate BP readings
Primary complete repair

» Consists of relief of the RVOTO and closing the VSD:

» VSD closed preferably via a transatrial approach

» RVOTO resected and widened. If valve is hypoplastic, transannular patch is indicated

» Sub valvular patch repair if pulmonary annulus is adequate
Transannular Patch
Common Problems Late After TOF repair

Pulmonary regurgitation:

- All patients have PR, due to disruption of the valve integrity during the primary repair - particularly with Transannular patches.

- PR is well tolerated in young patients but as they reach adulthood the haemodynamic burden of chronic PR begins to have an impact on exercise tolerance and survival.
» Chronic free PR results in excessive volume overload of the RV

» Leading to RV dilation, dysfunction and eventually failure

» RV dilation causes broadening of the QRS & QT interval

» QRS duration greater than 180ms predisposes patients to VT & SCD
Pulmonary Valve Replacement

» One of the main goals is to preserve RV function, which involves multiple pulmonary valve replacements over the course of their lifetime

» Until 2002, open heart surgery was the only option to replace pulmonary valves

» This involved a prolonged post operative recovery. The use of CPB with it's inherent risks: thrombosis, cerebral hypoperfusion, myocardial ischemia and transfusion risks
Percutaneous Pulmonary Valve Implantation (PPVI)

The Melody Valve

The melody valve is a bovine jugular valve mounted within a platinum stent (Medtronic)
First introduced in 2002 - one of the most significant developments in CHD of the last 20 years

Benefits:

» Faster recovery - 2 night hospital stay v 1 week for sPVR

» Minimally invasive - no need for CPB

» Avoids increased morbidities associated with multiple sPVR
Complications:
» Potential compression of coronary arteries during implant
» Migration of the stent
» Stent fracture
» Endocarditis

Limitations:
» Not suitable for use in native RVOT
» Limited size 18mm - 22mm

Nursing Considerations:

Peri procedure
12 lead ECG
Telemetry
Chest pain? Consider migration of valve
Usually urgent bedside echo and may need to return to cath lab for retrieval of valve (with stand by surgical team)

Endocarditis
Patient education on symptoms and risk reduction for IE. (Prophylactic antibiotics be given before invasive dentistry in accordance with the European Society of Cardiology 2015 guidelines). Ensure your patients are aware.
Arrhythmias begin to occur approx age 25

10% of adults develop ventricular arrhythmias by the age of 25. Increasing to 30% by age 55

Atrial arrhythmias are fairly common - reentry tachycardias of RA

AF more prevalent in the older population (after age 55)

(Khairy P, Aboulhosn J et al, 2010)
Risk Factors associated with arrythmia

- VT - Ventriculotomy (transatrial/transpulmonary approach preferred)

- Extensive resection of the RVOT causes heavy scarring
Multiple surgeries increase atrial and ventricular arrhythmias

TOF patients may face at least 4 PVR's over their lifetime, hence a heavy burden of arrhythmia

(Khairy P, Aboulhosn J et al, 2010)
Arrhythmia Management

» EP testing and/or ablation is recommended for suspected or documented ventricular or atrial arrhythmias (European Society of Cardiology Guidelines, 2010)

» A considerable number of TOF patients are under the joint care of the Electrophysiologists and undergo multiple Ablations during their lifetime
VT from the RVOT scar - treated with a VT Ablation, using 3 dimensional mapping

Gatzoulis et al, 2017
Secondary prevention ICD

» Is recommended for spontaneous sustained VT (after failure to identify reversible causes) in conjunction with ablation

» For patients resuscitated from SCD

Khairy P et al 2014
Risk of sudden cardiac death

» Sudden cardiac death (SCD) is the most common cause of late mortality in TOF

» Proportionally, in comparison to other ACHD groups, TOF patients have the highest incidence of SCD - 30%

» However the overall annual incidence is low - 0.15%

Khairy et al, 2014
Risks include:

» QRS duration > 180 ms

» QRS duration increasing > 3ms per year

» Inducible VT on electrophysiology study

» VT on holter monitoring
» Severe RV dilation and/or impairment (RV ejection fraction < 45%)

» Left ventricular systolic dysfunction

» RV/LV fibrosis

» Elevated LVEDP > 12 mmHg
Primary Prevention ICD

How to reliably identify high risk TOF patients when the overall annual incidence of sudden cardiac death is low - 0.15%?
## Risk Score for ICD in Primary Prevention

<table>
<thead>
<tr>
<th>Variable</th>
<th>Points attributed</th>
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<tbody>
<tr>
<td>Prior palliative shunt</td>
<td>2</td>
</tr>
<tr>
<td>Inducible sustained VT</td>
<td>2</td>
</tr>
<tr>
<td>QRS &gt; 180 ms</td>
<td>1</td>
</tr>
<tr>
<td>Ventriculotomy incision</td>
<td>2</td>
</tr>
<tr>
<td>Nonsustained VT</td>
<td>2</td>
</tr>
<tr>
<td>LVEDP &gt; 12 mmHg</td>
<td>3</td>
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<tr>
<td><strong>Total</strong></td>
<td><strong>0-12</strong></td>
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*(Khairy, P et al 2009)*
Case Study
57 yr old male
Occupation: Lorry Driver

Diagnoses:
» Tetralogy of Fallot.
» Moderately impaired LV systolic function (Worsening LVEF of 43% on CMR)
» No evidence of obstructive CAD on CTCA

Question: ? Risk of SCD, ?For primary prevention
ICD
Interventions:
» Left BT shunt in 1964
» Complete TOF repair in 1968
» PVR (27mm Edwards Xenograft Valve and Bentall procedure (23mm St Jude AVR) 2003

Current Status:
» Aysymptomatic, denies chest pain, shortness of breath, palpitations or syncope
» Medications: Candesartan 4mg and Warfarin
» BP 138/65 HR 65 (regular)
ECG

Sinus rhythm 65, normal PR interval (183ms), RBBB, QRS duration 180ms, left axis
Chest X-ray showed:

- situs solitus,
- levocardia,
- left aortic arch,
- cardiomegaly,
- previous sternotomy,
- normal pulmonary,
- vascular markings,
- Aortic and pulmonary prosthesis
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**Khairy Score 5**

» Annual risk of SCD estimated 2-3 %

» He was referred for VT stimulation Studies - +ve

» He had insertion of ICD

*Khairy, P et al 2009*
Referred for Ablation

ICD check showed high burden of atrial flutter

He lost his job

2 months later had an inappropriate shock for fast atrial flutter HR 225

His HGV licence was suspended

He became increasingly anxious and depressed

Referred to clinical psychologist

Referred for Ablation
» Risk stratification for primary prevention remains a challenge.

» Higher risk of inappropriate shocks for sinus tachycardia or SVT's due to oversensing of T wave and QRS morphology in TOF

» ICD's can negatively impact QOL causing depression and anxiety, post shock

» Younger age group will require multiple box changes - increased risk of infection/endocarditis

Khairy et al. 2014
Pregnancy

» With good haemodynamics, usually well tolerated, similar to the general population

» Pregnancy increases fluid volume by 50% - which may tip someone with PR and severe dilation into HF

» Pre-pregnancy PVR is recommended if there is PS/PR with RV dilation

» Foetal screening is recommended for chromosomal abnormalities: Di George syndrome (22q11), Down's
Summary

» TOF patients survive well into adulthood, however PR is universal and they will face multiple PVR's

» Percutaneous PVR is a viable alternative to conventional surgery in selected patients, however further investigation is required into to the increased incidence of endocarditis.

» SCD is the leading cause of mortality - ICD's are recommended for secondary prevention. Risk stratification for primary prevention ICD's remains a challenge

» Optimal timing is required for interventions and they should have life long follow up with regular CMR's to monitor RV function
References

Holoshitz N & Hijazi Z. Transcatheter pulmonary valve replacement: valves, techniques of implantation and outcomes. Interv Cardiol 2013 5 (4)


Tutarel O, Kempny A et al. Congenital Heart Disease beyond the age of 60: emergence of a new population with high resource utilisation, high morbidity and high mortality. European Heart Journal (2014) 35, 725-732

Uebing A & Rigby M.L. The problem of infective endocarditis after transcatheter pulmonary valve implantation. Heart March 2015
Thank You

The End