Tetralogy of Fallot

Andrea Frederick RN, NP
Nurse Practitioner, Adult Congenital Cardiology
Stanford Hospital and Clinics
Palo Alto, California

I. Embryology

A. The heart starts as a tube. Two sections of the tube, the truncus arteriosus and the bulbus cordis, grow towards each other. The truncus arteriosus twists 180° as it grows down towards the bulbus cordis. This twisting separates the aorta and the pulmonary artery. Anterior deviation of the twisting causes tetralogy of Fallot (TOF). Anatomic features include:

1. Smaller right ventricular outflow tract (RVOT) and pulmonary valve (PV)
2. Impeded flow from right ventricle (RV)
3. Anterior malaligned ventricular septal defect (VSD).
4. Enlarged aortic root which overrides the VSD
5. Right ventricular hypertrophy (RVH) develops as a result of the RV pumping against the small RVOT and PV

II. Anatomy

A. Right sided obstruction may occur at three levels.

1. Obstruction along the RVOT
2. Hypoplasia/stenosis of the pulmonary valve (PS)
3. Stenosis of the pulmonary arteries

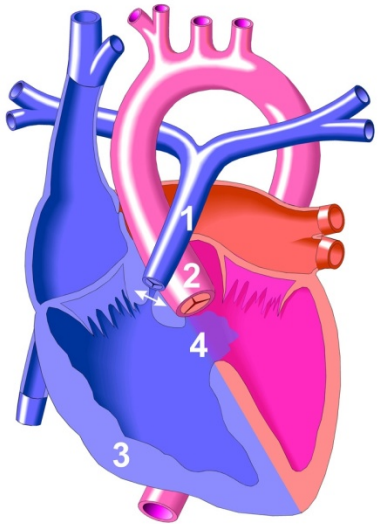
B. Aorta (Number 2 in illustration)

1. Sits over the VSD, called an ‘overriding’ aorta.
2. Aortic root dilation
   a. Occurs with significant sub-pulmonary stenosis and right to left shunting across the VSD
   b. Results from increased blood flow across the aortic valve

C. Right ventricular hypertrophy (Number 3 in illustration)

1. occurs if TOF repair is not done early in life.

D. Ventricular septal defect (Number 4 in illustration)



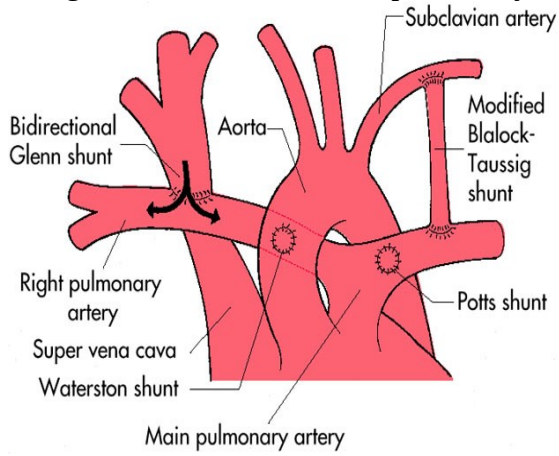
E. Variants of TOF
1. TOF with pulmonary atresia
2. TOF with pulmonary atresia with multiple aorta-pulmonary collaterals (MAPCA's)
3. TOF with absent pulmonary valve
4. TOF with double outlet RV
5. TOF with atrial septal defect (ASD)

III. Physiology/clinical findings
A. Neonate (Unrepaired)
1. Cyanosis secondary to right to left shunting across the VSD
2. Loud, harsh systolic ejection murmur (SEM) from right sided obstruction
3. Severe sub-PS results in very little forward flow through the RVOT
a. Results in increased cyanosis
b. May hear decreased systolic ejection murmur (SEM) from decreased flow
4. Clubbing not developed with early repair

B. Repaired Adult with TOF
1. Pathology related to right ventricular outflow track obstruction and pulmonary regurgitation.
a. Right ventricular dilation and dysfunction from increased volume load due to pulmonary valve regurgitation
b. Murmurs
(1) To and fro murmur heard best at the left upper sternal border (LUSB) from pulmonary stenosis and regurgitation
(2) Systolic ejection murmur on upper back from pulmonary artery stenosis
(3) Systolic murmurs on lateral lung fields from branch pulmonary artery stenosis
2. Pathology related to tricuspid regurgitation
a. Chordal tissue of the septal leaflet of the tricuspid valve attach to the intraventricular septum
b. Leaflets separate as right ventricle dilates resulting in tricuspid valve regurgitation
3. Pathology related to aortic valve
a. Regurgitation may be present
b. Dilation of aortic root
c. Diastolic murmur from aortic valve regurgitation
4. Pathology of conduction system
a. Right bundle branch block (RBBB)
(1) Almost always present and results from:
(a) Ventriculotomy
(b) Infundibular resection

(c)RV dilation
(2) S2 may be widely split
(3) Risk of ventricular tachycardia and sudden death from RBBB > 180 miliseconds

IV. Types of Repairs
A. Palliative Shunts
1. Classic Blalock-Taussig Shunt (BT shunt)
a. Subclavian artery anastamosed to pulmonary artery
(1) First preformed in 1944.
(2) Palliative procedure done in infancy with complete 'repair' later in life
(3) Thoracotomy scar on the side where the shunt was placed
(4) Diminished pulse on affected arm due to use of subclavian artery to supply blood to the pulmonary arteries
b. Arm on operated side should not be used for blood pressure assessment or to draw blood
2. Modified BT shunt
a. Connection with Gore-Tex tube graft between subclavian artery and pulmonary arteries
b. May complete assessment of pulse and BP in both arms
3. Waterston shunt
a. Ascending aorta connected to main or right pulmonary artery (RPA)
b. May lead to pulmonary hypertension
4. Potts Shunt
a. Descending Aorta connected to left pulmonary artery (LPA)



B. Primary TOF Repair
1. Closure of VSD
2. Opening of the RVOT
a. Surgery depends upon size of pulmonary valve and arteries
b. RV to PA conduit
c. Trans-annular patch
d. Augmentation of pulmonary arteries

C. Repeated intervention in the adult patient may include:

1. Pulmonary Valve replacement
2. Relief of RVOT
3. RV-PA conduit change
4. PA augmentation
5. Cavo-tricupsid isthmus ablation (MAZE procedure for intractable arrhythmias)
6. Tricuspid valve repair or annuloplasty
7. Aortic root replacement
8. Aortic valve replacement
9. Closure of residual VSD's or ASD's

V. Long Term Complications (See Problem List included with these documents)

A. Pulmonary regurgitation
B. Right ventricular dilation
C. Right ventricular dysfunction
D. Right sided heart failure
E. Tricuspid regurgitation
F. Residual RVOTO
G. Branch PA stenosis
    1. May develop at the site of the original BT shunt
    2. May not have been present since birth but never adequately repaired
H. Endocarditis
I. Aortic regurgitation +/- aortic root dilation
    1. May result from damage to valve from VSD closure
    2. May be intrinsic aortic root abnormality
J. LV dysfunction
K. Atrial arrhythmia
    1. May be a result of a scar line
    2. May be a result of multiple areas of fibrosis
L. Ventricular arrhythmia
M. Sudden death

References:

Bailliard F, Andersen H, et al: Tetalogy of Fallot, *Orphanet Journal of Rare Diseases* 1750-1172, 2009.

Gatzoulis M, Webb G, Daubeney P: Diagnosis and Management of Adult Congenital Heart Disease. *Elselvier Unlimited*, 2003.

Hickey E, Veldtman G, Bradley T, et al: Late Risk of outcomes for adults with repaired tetralogy of Fallot from inception and cohort spanning four decodes, *Journal of Cardiothoracic Surgery*, 35:156-164, 2009.

Huehnergarth K, Gurvitz M, stout K, et al: Repaired Tetralogy of Fallot in the adult: monitoring and management, *Heart*: 1663-1669, 2008.