

Morphology of tetralogy of Fallot: the range in spectrum from absent pulmonary valve to pulmonary atresia

S. Yen Ho FRCPath, FESC, FHEA



Brompton Cardiac Morphology

Webpage: www.rbht.nhs.uk/morphology

E-mail: Morphology@rbht.nhs.uk LinkedIn: Brompton Cardiac Morphology

www.rbht.nhs.uk/Morphology

Tetralogy of Fallot

Malformation first described by Niels Stensen in 1671

Etienne-Louis Arthur Fallot
1888

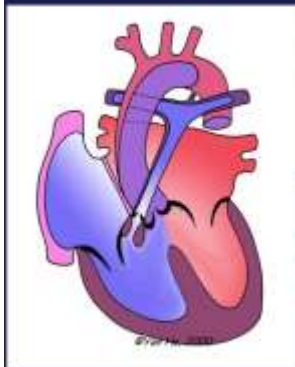
Contribution à l'anatomie pathologique de la maladie bleue (cyanose cardiaque)



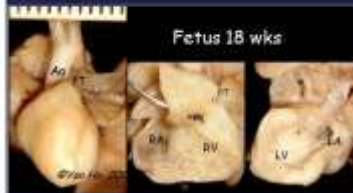
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Tetralogy of Fallot



- Ventricular septal defect
- Overriding aorta
- Subpulmonary stenosis
- RV hypertrophy



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Tetralogy of Fallot – anatomy and variants

~ 5% of congenital heart disease

With pulmonary stenosis

With pulmonary atresia (PA with VSD)

First surgical repair by Lillehei et al in 1954

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Tetralogy of Fallot – anatomy and variants

The RV muscle bundles

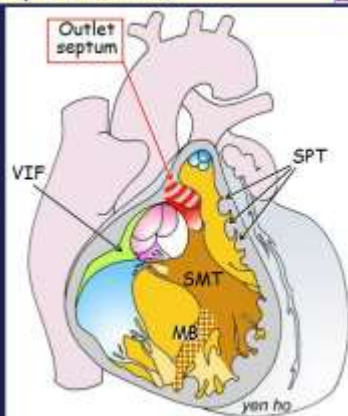
Septomarginal trabeculation / trabecula septomarginalis (SMT)

Ventriculo-infundibular fold (VIF)

Septo-parietal trabeculations (SPT)

Moderator band (MB)

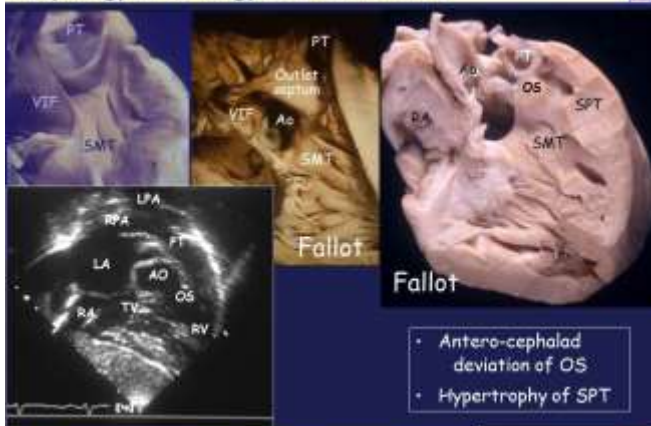
Outlet septum



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Morphology: Tetralogy of Fallot with PS



- Antero-cephalad deviation of OS
- Hypertrophy of SPT

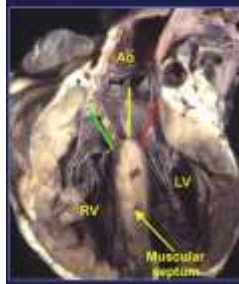
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Basic malformation +
Variable morphologic features

- Extent of aortic override
- Right ventricular margins of VSD
- Nature of sub-pulmonary obstruction
- Pulmonary valve malformation
- Associated malformations

Overriding aorta



Variability in Aortic Override

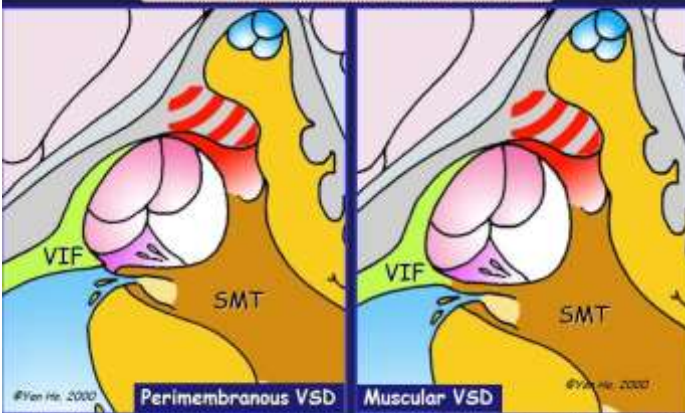
Variability in Ventriculo-arterial connections

- Aorta predominantly supported by LV (effectively concordant connections)
- Aorta predominantly supported by RV (effectively double outlet connection)

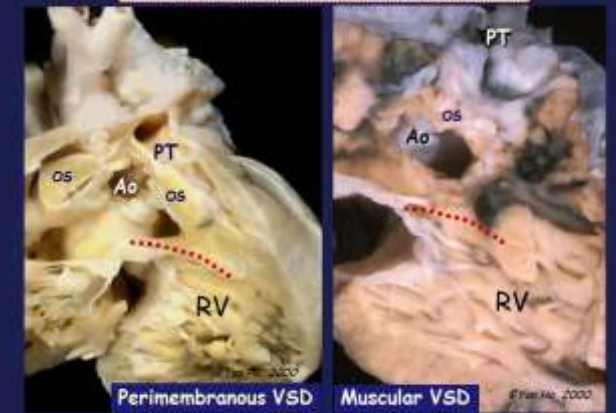
- at mouth of infundibulum } stenotic to atretic
- pulmonary valve
- within body of right ventricle
- distally within pulmonary arteries



VSD and AV conduction bundle

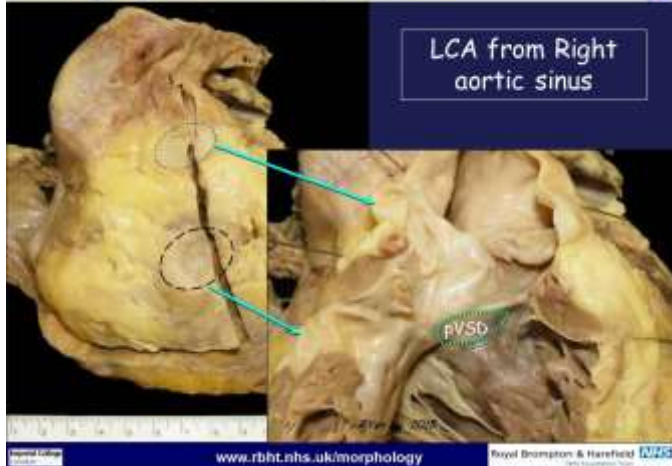
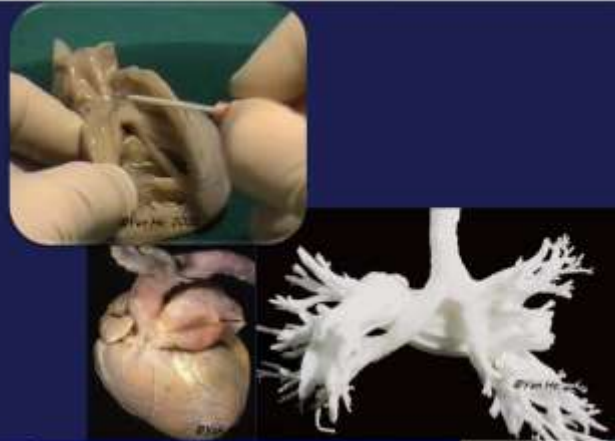


VSD and AV conduction bundle



Associated malformations

- Pulmonary atresia ⇔ Later
- "Absent" leaflets of pulmonary valve
- Straddling tricuspid valve
- Atrioventricular septal defect
- Coronary arterial anomalies
- Right aortic arch
- Muscular inlet VSD
- Arterial wall



VSD patch, muscular resection
± RVOT patch/conduit



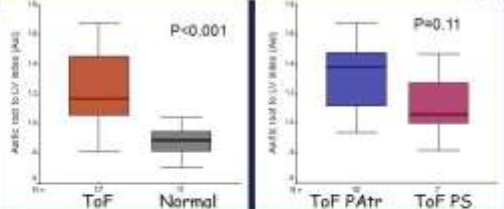
Pulm valve calcification
TV regurgitation
Aortic root dilatation



Tetralogy of Fallot: Aortic root

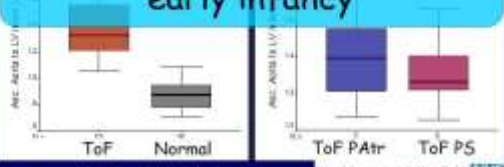
Tan et al, Circulation 2005

Aortic root



Aortic dilation present from early infancy

Asc. Aorta

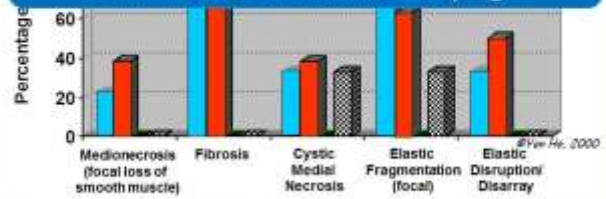


Tetralogy of Fallot: Aortic root

Tan JL et al, Circulation 2005

TOF infants/children TOF adults
Normal infants/children Normal adults

Histologic abnormalities present in aortic media from an early age

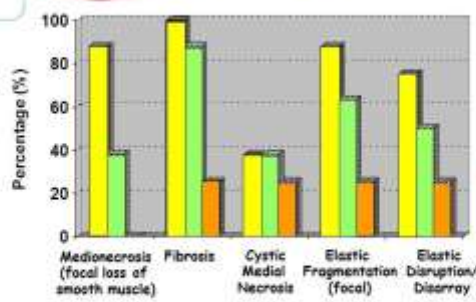


Percentage of abnormal histological features of Grade 2 or Grade 3 in TOF versus normal groups. Grading 1,2,3 according to Schlatmann & Becker, Am J Card 1977

Tetralogy of Fallot: Aortic root

Tan JL et al, Circulation 2005

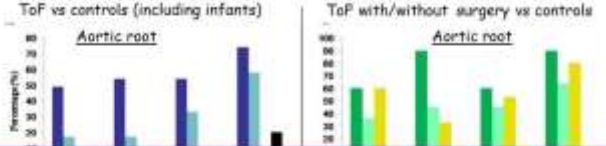
Ao Root Asc Aorta Desc Thoracic aorta



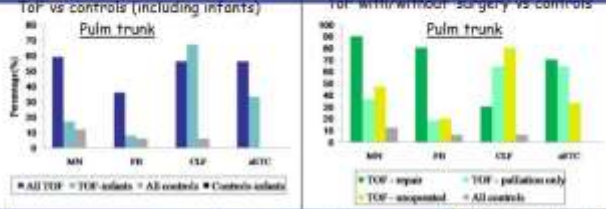
Aortic distribution of the various abnormal histological features of Grade 2 or 3 in the adult TOF group (n=8)

Tetralogy of Fallot: Pulmonary trunk

Histology changes grade ≥2



Marked intrinsic histological abnormalities in the PT of Tetralogy compared to controls. Bedard et al, JACC 2009



Tetralogy of Fallot: anatomy and variants

- Pulmonary atresia ←
- "Absent" leaflets of pulmonary valve
- Straddling tricuspid valve
- Atrioventricular septal defect
- Coronary arterial anomalies
- Right aortic arch
- Muscular inlet VSD
- Arterial wall



Yen Ho
Morphology@rbht.nhs.uk

Tetralogy of Fallot: with pulmonary atresia

- RV outlet to aorta via VSD
- Nature of atretic outlet (muscular/imperforate membrane)
- Type of VSD (perimembranous/muscular/multiple)
- Other intracardiac defects
- What supplies the lungs ?

Aortic override



Concordant ↔ DORV

Nature of atretic outlet

- Well formed outlet septum **Deviated**
- Imperforate pulmonary valve
- Muscular atresia

Example of muscular atresia

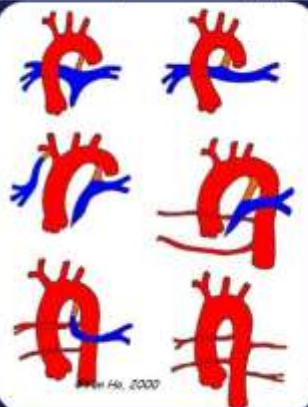


- RV outlet to aorta via VSD
- Nature of atretic outlet
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Source of pulmonary blood supply



pulmonary blood supply ?



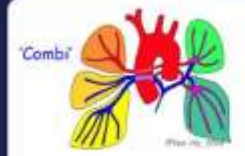
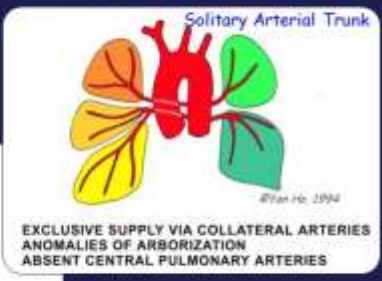
Variations

- Duct: unilateral or bilateral
- Systemic-to-pulmonary collateral arteries
- Others, e.g. :
Aorto-pulmonary window
Fifth aortic arch
Coronary-pulmonary fistula

Tetralogy of Fallot: with pulmonary atresia

pulmonary blood supply ?

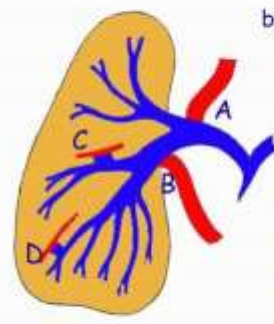
Variations



RIGHT UPPER LOBE VIA COLLATERAL ARTERY
OTHER LOBES VIA CENTRAL PULMONARY ARTERIES
- FED VIA THREE COLLATERAL ARTERIES

Tetralogy of Fallot: with pulmonary atresia

Anastomoses between SPCA and central PAs
8/9 cases



	Right	Left
A. Extrapulmonary	2	1
B. Hilar	0	1
C. Lobar	5	7
D. Segmental	6	1
	13	10

Ho et al, 1992

Tetralogy of Fallot: with pulmonary atresia

SPCA

PA

Anastomosis between SPCA and central PAs

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Tetralogy of Fallot: with pulmonary atresia

SWIGART ET AL: ESOPHAGEAL ARTERIES

Bronchial arteries (300 specimens)

Fig. 4. Types of bronchial arterial anastomosis in perspective. Schematically shown, as if from the dorsal aspect. The classification is based upon origin, number, and course of the vessels.

Cauldwell et al 1948, revised by Swigart

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Tetralogy of Fallot: with pulmonary atresia



Imperforate membrane
Muscular atresia
Absence of central PAs

pulmonary blood supply ?

Associated malformations

Yen Ho
E-mail: Morphology@rbht.nhs.uk
LinkedIn: Brompton Cardiac Morphology