The Post-Surgical ACHD Patient

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THE POST-SURGICAL ACHD

Post-operative appearances of common repaired ACHD conditions

Review the residual anatomic and haemodynamic abnormalities that occur after surgical repair and recognise their imaging features

Review the role of echocardiography in the long-term follow-up of this patient group

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THE POST-SURGICAL ACHD – IMPORTANT PRINCIPLES

Underlying anatomy and approach to surgical repair are highly variable between patients

Key to successful echo assessment is availability of the surgical notes

Knowledge of the precise nature of the repair will aid in identification of potential long-term complications

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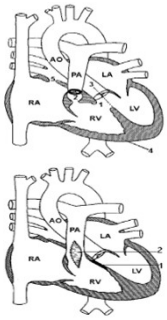
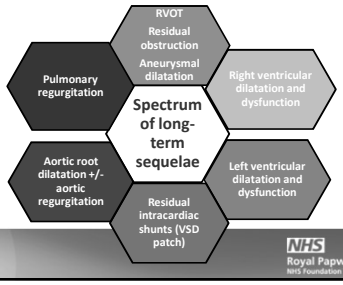
TETRALOGY OF FALLOT

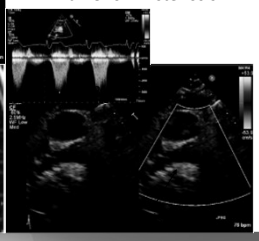


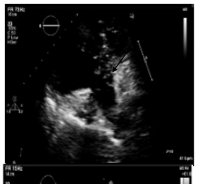
AIM OF SURGERY
Closure of the VSD
Relief of RVOT obstruction
Preserve pulmonary valve function if possible



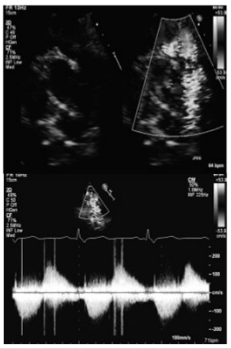
TOF – ASSESSMENT OF THE RVOT + PULMONARY VALVE

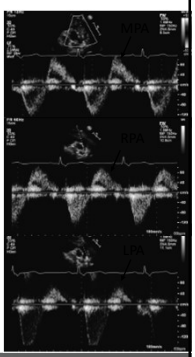
Aneurysm
Obstruction
(subvalvar/valve/supravalvar)
↑TR jet velocity -
marker of PA stenosis





TOF-ASSESSMENT OF THE PULMONARY VALVE

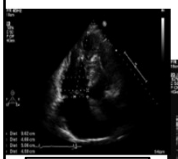


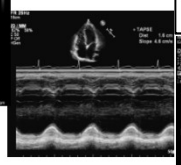


TOF – ASSESSMENT OF THE RIGHT HEART

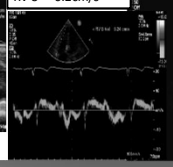
- Haemodynamic consequences of long-standing PR include RV dilatation, RVH, and dysfunction
- Indications for PVR following TOF repair include progressive RV dilatation and RV dysfunction

RVAd = 32.2cm²
RVAs = 19.9cm²
FAC = 38%
RVEDVi = 138ml/m²



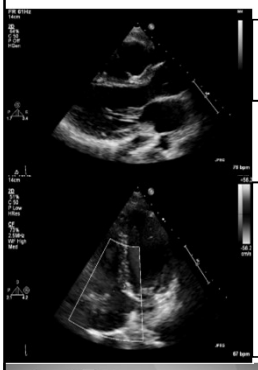


TAPSE = 1.6cm
RV S' = 6.2cm/s



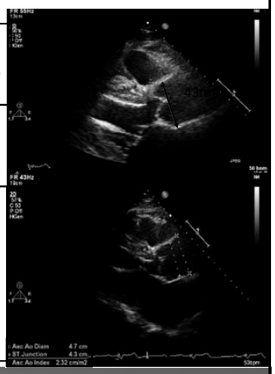
TV annulus = 4.5cm
RV base = 5.1cm
RV mid = 4.7cm
RV length = 9.6cm

OTHER POST-REPAIR FEATURES

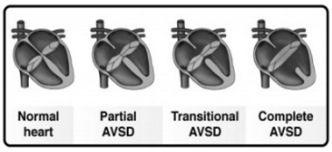


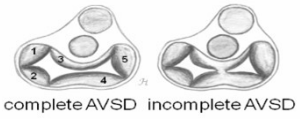
Residual VSD's are typically located at the superior portion of the patch

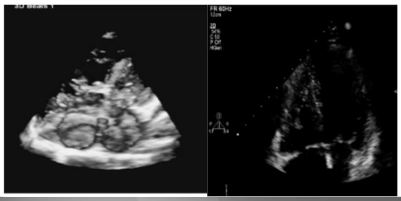
30% - AoR ≥ 40mm
AORTIC REGURGITATION
Damage to the AV during VSD closure
2° to intrinsic aortic root abnormality
Usually mild
Mod-severe AR ≈ 3.5%



ATRIO-VENTRICULAR SEPTAL DEFECT (AVSD)





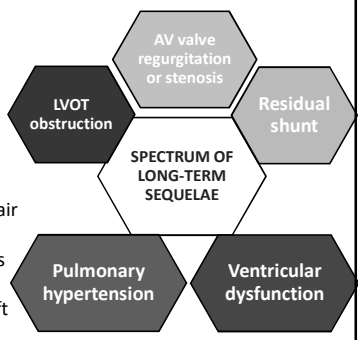


- Spectrum of lesions of deficient AV septation
- Failure of fusion of the embryonic endocardial cushions
- 5% of all congenital defects

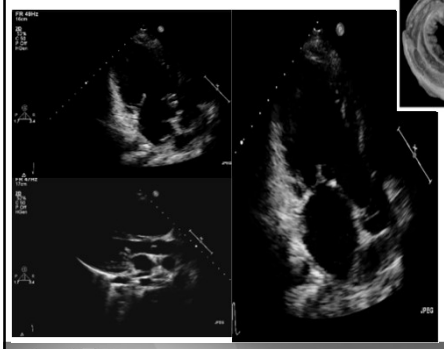
SURGICAL REPAIR OF AVSD

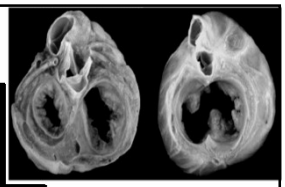
PARTIAL AVSD
Primum ASD – close interatrial communication
Cleft mitral valve – restore LAVV competency

COMPLETE AVSD
Primum ASD + Inlet VSD - early repair to avoid pulmonary vascular disease/close all intracardiac shunts
Common AV valve with single annulus – divide into competent left and right orifices



AVSD – ELONGATED LVOT

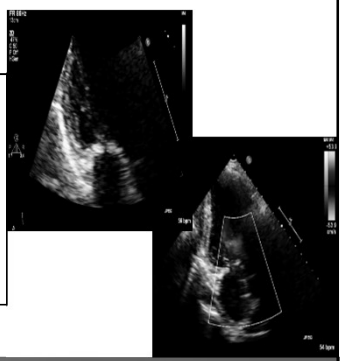




Aorta is unwedged/anteriorly displaced
→ elongated + narrowed LVOT - "goose-neck deformity"
NORMAL:
LV apex – aortic annulus = LV apex – mitral annulus
AVSD:
LV apex – aortic annulus >> LV apex – mitral annulus

REPAIRED AVSD – LEFT AVV REGURGITATION

Most common complication after repair
Most common reason for reoperation
10-15% require reoperation for LAVVR
Predictors of re-operation:
-Significant LAVVR intra-operatively
-Dysplastic LAVV
-Failure to close cleft at time of initial repair



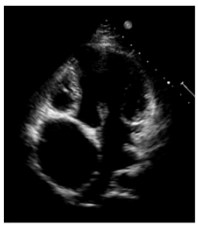
FONTAN CIRCULATION

Creation of a systemic venous to PA connection
Functionally a 'univentricular' circulation

FAILING FONTAN

- Pathway obstructions
- Ventricular dysfunction
  Diastolic dysfunction in 70%
- Thrombus formation
  (33% of RA-PA Fontan)
  (6-8% LT or EC Fontan)
- AV valve regurgitation or stenosis

FONTAN CIRCULATION

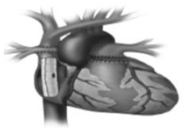


ATRIO-PULMONARY (AP)

The original palliation or RA–PA connection. RA appendage connected directly to the main PA

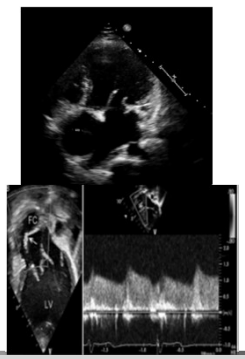
- right atrial enlargement +/- impingement of PV return +/- AV valve stenosis
- atrial arrhythmias
- stagnant flow with decreased antegrade pulmonary flow +/-thrombus
- Systemic venous hypertension leading to hepatic congestion

FONTAN CIRCULATION – TOTAL CAVOPLMONARY CONNECTION



LATERAL TUNNEL TCPC

SVC – RPA (bidirectional Glenn shunt)
IVC baffled through the lateral wall of the RA into the RPA

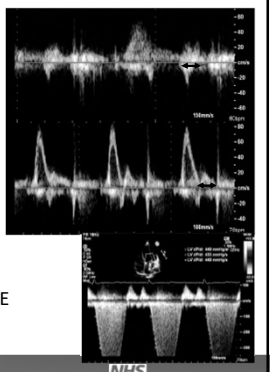


EXTRACARDIAC TCPC

SVC – RPA (bidirectional Glenn shunt)
IVC is connected via a tube graft to the RPA

Can be fenestrated – communication between FC conduit and the pulmonary venous atrium – continuous flow with mean PG of 4-8mmHg; must be distinguished from pathway leak

FONTAN CIRCULATION – IMAGING THE SINGLE VENTRICLE



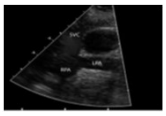
- Diastolic dysfunction common (in 70% of cases)
- Difficult to assess due to reduced preload in Fontan circulation
- E wave Decel time and PV A wave reversal duration: A wave duration >28ms indicates raised filling pressures

- Single ventricle RV – EF lower than if LV
- Morphologic LV - Biplane Simpson's EF
- Morphologic RV
- Eyeball / +dp/dt from systemic AVVR / TAPSE

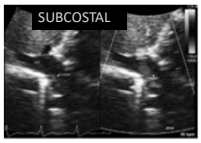
FONTAN CIRCULATION – IMAGING THE PATHWAY

SVC – RPA connection
Suprasternal notch of right supraclavicular windows
Normal appearances
- laminar flow on colour Doppler (less useful as velocities are low)
- Low velocity with phasic flow

IVC channel
Subcostal view sweeping to view IVC as it courses superiorly towards the PA
Normal appearances
- Laminar flow on colour Doppler (less useful as velocities are low)
- Low velocity with phasic flow



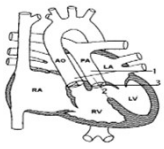
CLUES TO PATHWAY OBSTRUCTION
- dilated systemic veins upstream (hepatic and innominate veins)
- sluggish flow in the vena cavae +/- spontaneous contrast
- mean estimated gradients as low as 3 mm Hg can reflect clinically important obstruction



IMAGING THE FONTAN CIRCULATION IS CHALLENGING WITH ECHO – CMR IS COMPLIMENTARY FOR THE ASSESSMENT OF FONTAN CONNECTIONS AND THROMBUS

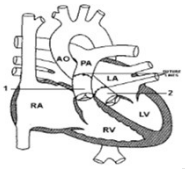
SURGICAL REPAIR OF TRANSPOSITION COMPLEXES

"Simple" transposition of the great arteries (d-TGA)

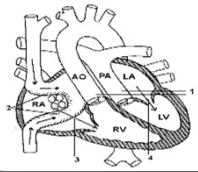


Atrial situs solitus
AV concordance
(mLA-mLV); (mRA-mRV)
VA discordance
(mLV-PA); (mRV-Ao)

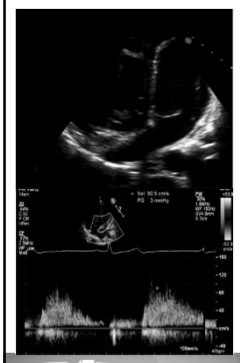
Jatene Arterial Switch



Atrial Switch
(Mustard or Senning)



ATRIAL SWITCH – MUSTARD/SENNING PROCEDURE



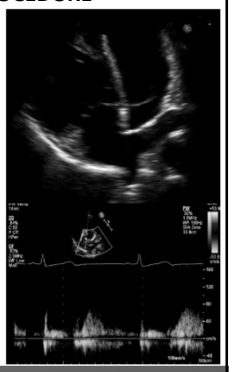
Creating of a baffle/ conduit within the atria

Systemic venous return - morphologic LV - PA

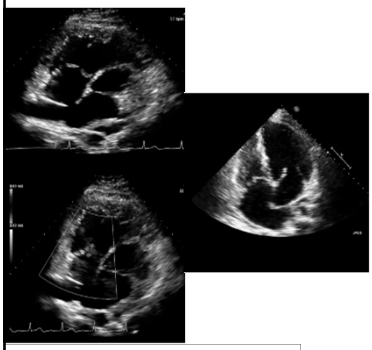
Pulmonary venous return - morphologic RV - Aorta

The morphologic RV is the systemic ventricle

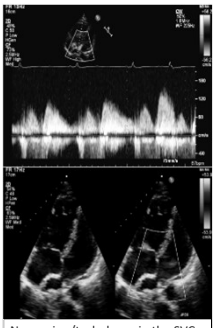
2D Imaging - Patent pathways with no anatomic narrowing
Colour Doppler - Laminar flow without evidence of leak across the baffles
PW Doppler – Vel <1.5m/s



COMPLICATIONS OF THE ATRIAL SWITCH

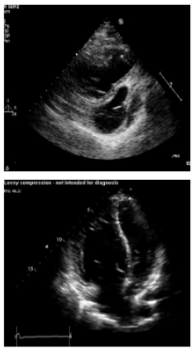


Baffle leak of IVC limb of systemic venous baffle



Narrowing/turbulence in the SVC limb of systemic venous baffle

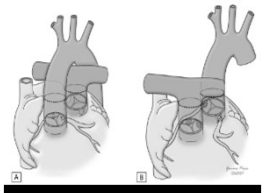
COMPLICATIONS OF THE ATRIAL SWITCH



SYSTEMIC RV FAILURE
- Hypertrophied dilated RV
- D-shaped subpulmonic LV
- Septal flattening (systole + diastole)
- Impairment of RV systolic function
- Significant RAVVR

PULMONARY HTN
- Remodelling of the subpulmonic LV
- Loss of D-shape
- Dilated PA
- Increased PASP

JATENE ARTERIAL SWITCH

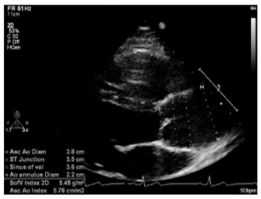


Transection of the pulmonary and aortic trunks and anastomosis of the distal ends
- neo-aortic dilatation and aortic regurgitation
- supravalvar stenosis of the PA

Translocation of the coronary arteries to the neo-aorta
- Coronary artery stenosis/occlusion

Lecompte manoeuvre – pulmonary trunk is anterior and branch PA's are draped across the aorta
- Branch PA stenosis

ARTERIAL SWITCH - COMPLICATIONS



AORTA
Dilatation of neo-aortic root

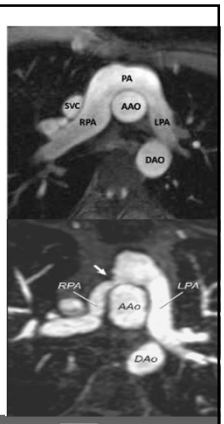
Significant AR is uncommon

STENOSIS OF THE RVOT/PA
Any level - most commonly at site of anastomosis

Imaging is challenging (high parasternal window)

Peak velocities =2 m/s across distal MPA + branch PA's within normal limits

TR jet velocity - surrogate of branch PS



CONCLUSIONS

Residual anatomic and haemodynamic abnormalities occur after surgical repair in many of the common congenital heart defects

Echocardiography plays a vital role in the long-term follow-up of this patient group

Anatomy of the surgical repair is vital to accurately identify normal versus abnormal appearances

CMR and CT are complementary imaging modalities in these patients, as some complications an anatomies are difficulty to fully characterise with echocardiography alone