Grown-up Congenital Heart Disease

La Diagnostica non invasiva

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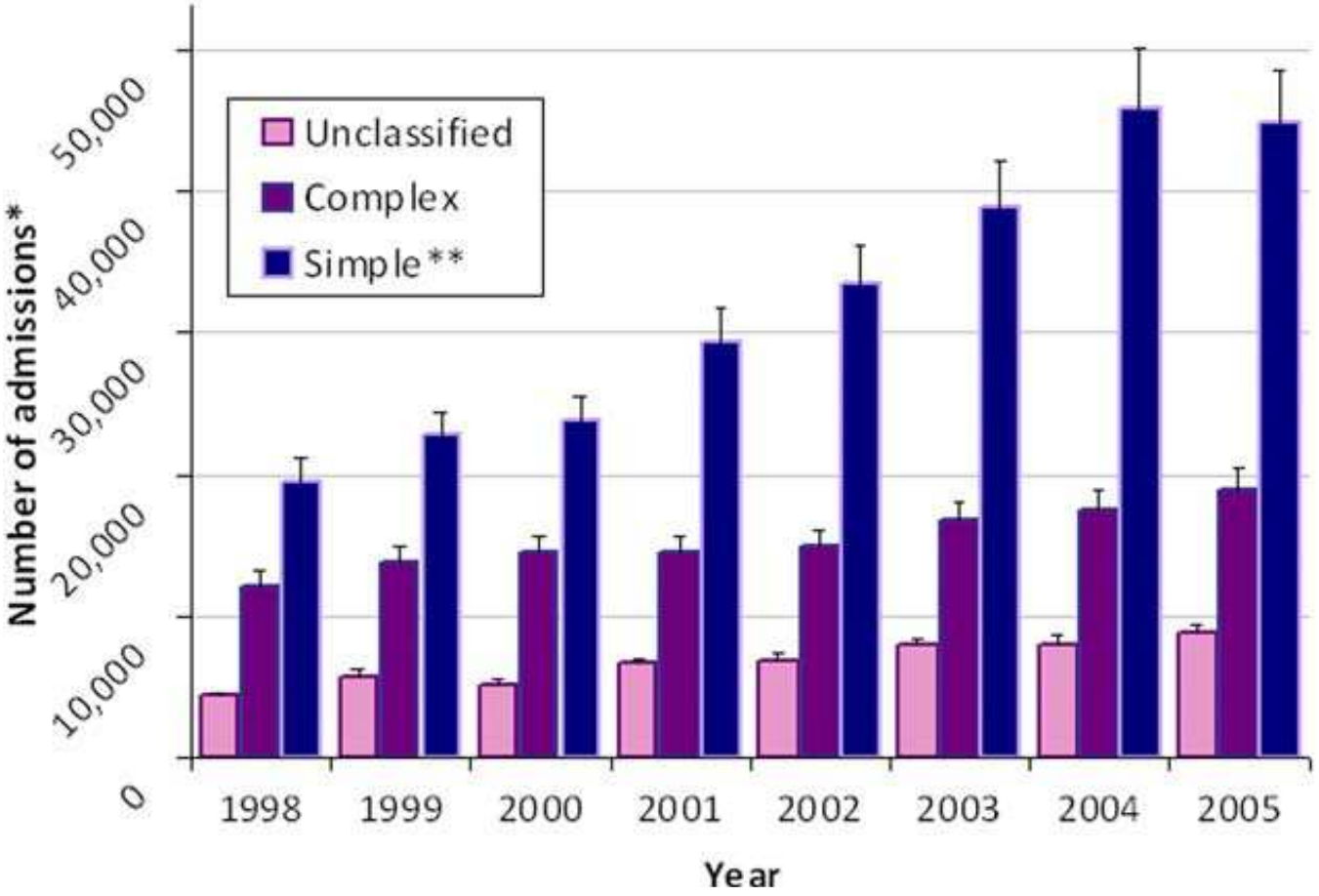
XL° Congresso Nazionale della Società Italiana
di Cardiologia Pediatrica

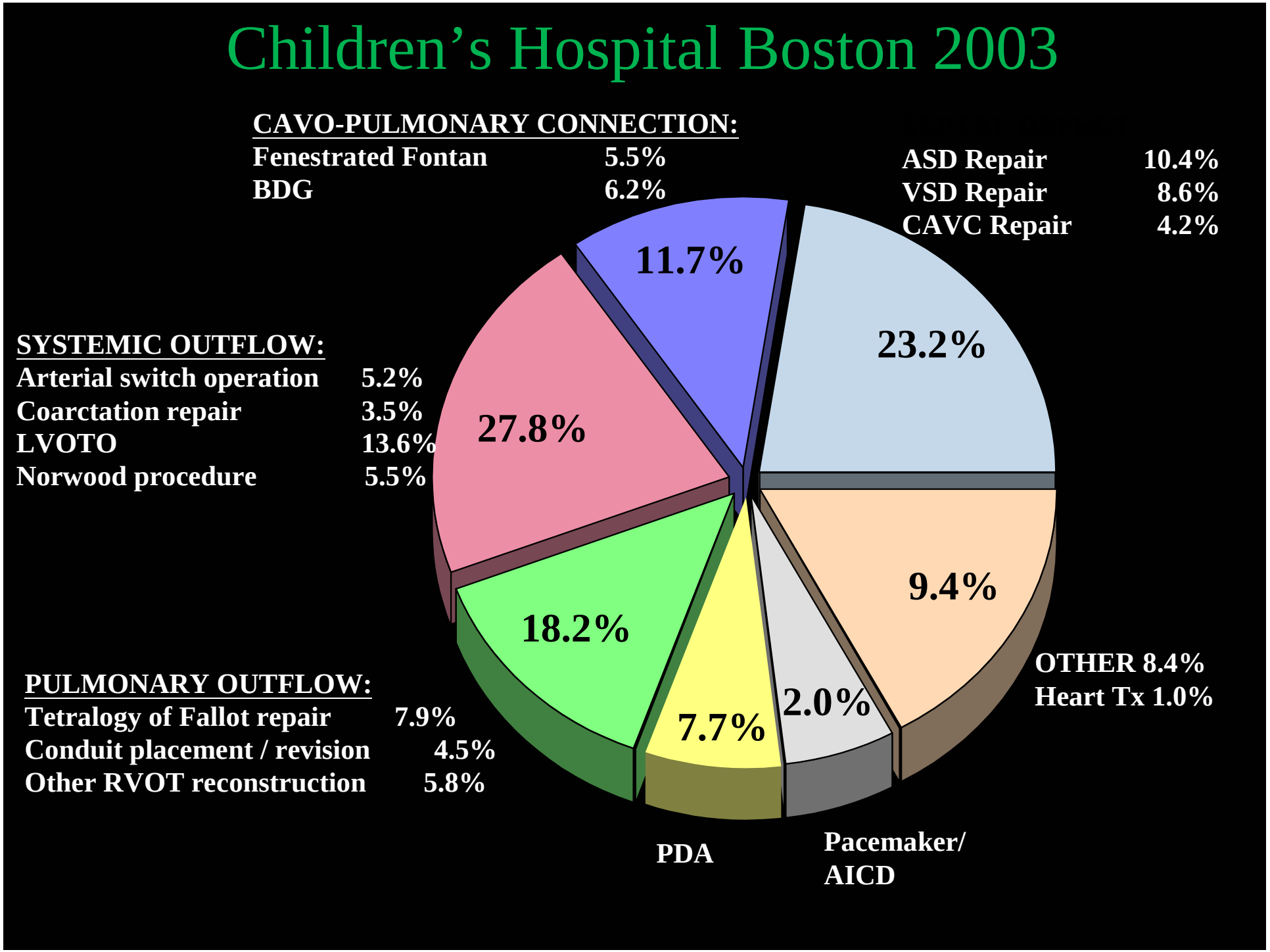
Milano, 24 settembre 2010

Annual Number of GUCH Admissions in the US Categorized by Level of Defect Complexity



Opotowsky AR, et al JACC 2009;54: 460–7

Children's Hospital Boston 2003



Complications After TOF Repair

Mechanical problems

- Significant Pulmonary regurgitation
- RV Dilatation
- Restrictive RV
- Residual RVOTO
- Aneurysmal dilatation of the RVOT
- Residual VSD
- AR with or without root dilatation
- LV dysfunction
- Infective endocarditis

Mechanical Problems After D-Transposition Corrections

- Atrial Switch (Mustard/Senning)
  - → Moderate systolic dysfunction of the RV {(50%) (only few present with CHF)}
  - → Severe systemic TR (1/3rd)
  - → Baffle leak or obstruction
- Arterial Switch
  - → Data about long term consequences still unavailable.
  - → Concerns include:
    - supra neopulmonary artery stenosis
    - ostial coronary artery disease
    - progressive neoaortic valve regurgitation

Long-term Complications After Atrial/Cavo-Pulmonary Connections

- Right atrial thrombus formation
- Obstruction of the Fontan circuit
- Ventricular dysfunction
- Protein losing enteropathy
- Excessive aortopulmonary collaterals
- Fenestration of atrial baffle
- Cyanosis/Venous Hypertension
- Pulmonary Arteriovenous Fistulas (a consequence of the bidirectional Glenn procedure)

Trend in Cardiac Catheterization for GUCH in a 20 Years Period

Open Cardiovasc Med J. 2009; 3: 124–127.

NonInvasive Imaging in GUCH

- There has been a shift form cardiac catheterization to non-invasive imaging modalities for patients with GUCH, which is similar to that which has occurred in pediatric cardiology over the last two decades
- These include echocardiography (transthoracic and transoesophageal), MRI, which are essential in specialist GUCH centres and, recently, CT

Echocardiography

- Echocardiography remains the first-line investigation and continues to evolve, with improved functional assessment using threedimensional echocardiography, Doppler, and contrast echocardiography
- Transoesophageal echocardiography, with superior image quality in adults, is advantageous in certain indications, but is required in a minority of examinations.

Strengths of Echocardiography

- Basic cardiac anatomy including orientation and position of the heart, venous return, connection of the atria and ventricles, and origin of the great arteries.
- Evaluation of the morphology of cardiac chambers, ventricular function, and detection and evaluation of shunt lesions, as well as the morphology and function of heart valves.
- Assessment of ventricular volume and pressure overload
- Doppler echocardiographic information, which includes haemodynamic data such as gradients across obstructions and right ventricle pressure/pulmonary artery pressure [obtained from tricuspid regurgitation velocity], but also flow calculations

Limitations of Echocardiography

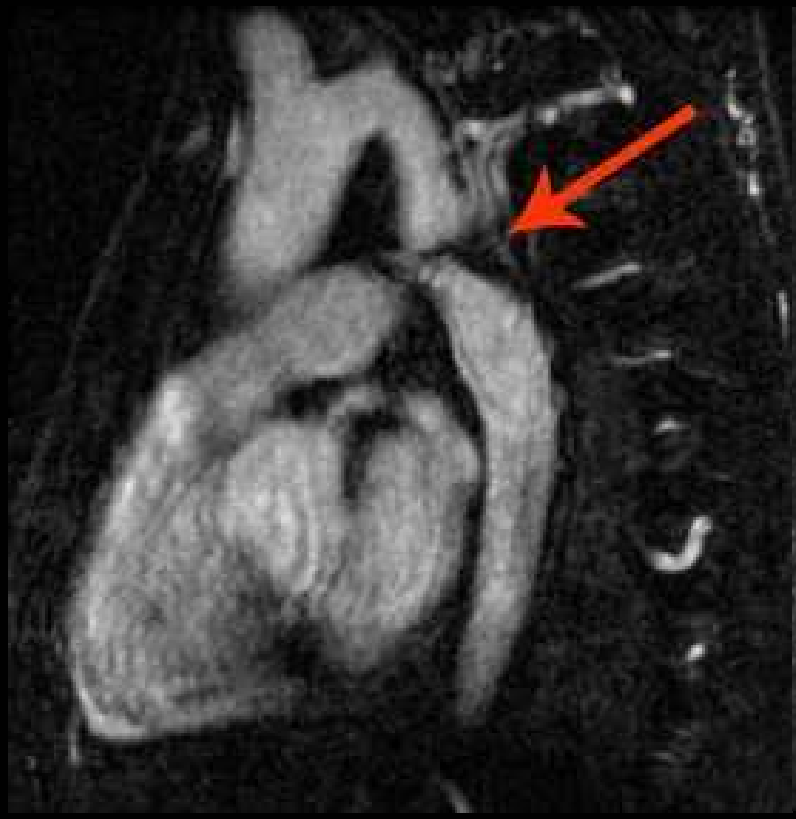
- It is highly user dependent, requiring special expertise in GUCH patients
- Assessment of ventricular volumes and function may be complicated by geometry and regional incoordination, particularly in systemic and non-systemic RVs or univentricular hearts
- Doppler gradients may sometimes be misleading,particularly in right ventricular outflow tract obstruction, CoA, and stenoses in series
- Venous return and great arteries may be difficult to image

Cardiovascular Magnetic Resonance

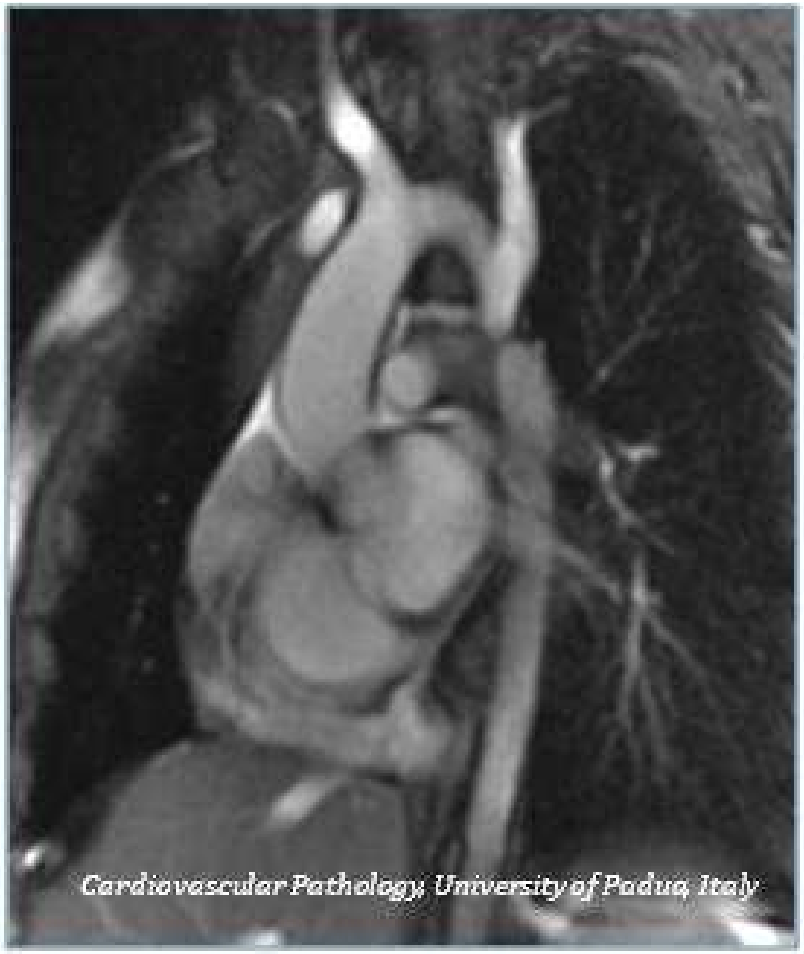
Strengths of CMR in GUCH

- Unrestricted access to cardiovascular anatomy and function, including the systemic and pulmonary venous connections, the right ventricle and pulmonary arteries, and the whole aorta, without ionizing radiation
- Well suited for repeated, life-long follow-up investigation, if needed
- Versatility, including measurements of biventricular size and function regardless of chamber geometry, measurements of flow volumes, characterization of tissues, and assessment of myocardial function, viability, and perfusion, when required
- Applicable in women with CHD during pregnancy, but without gadolinium contrast agent, unless essential.

Coarctation



Adult-type Aortic Arch Coarctation



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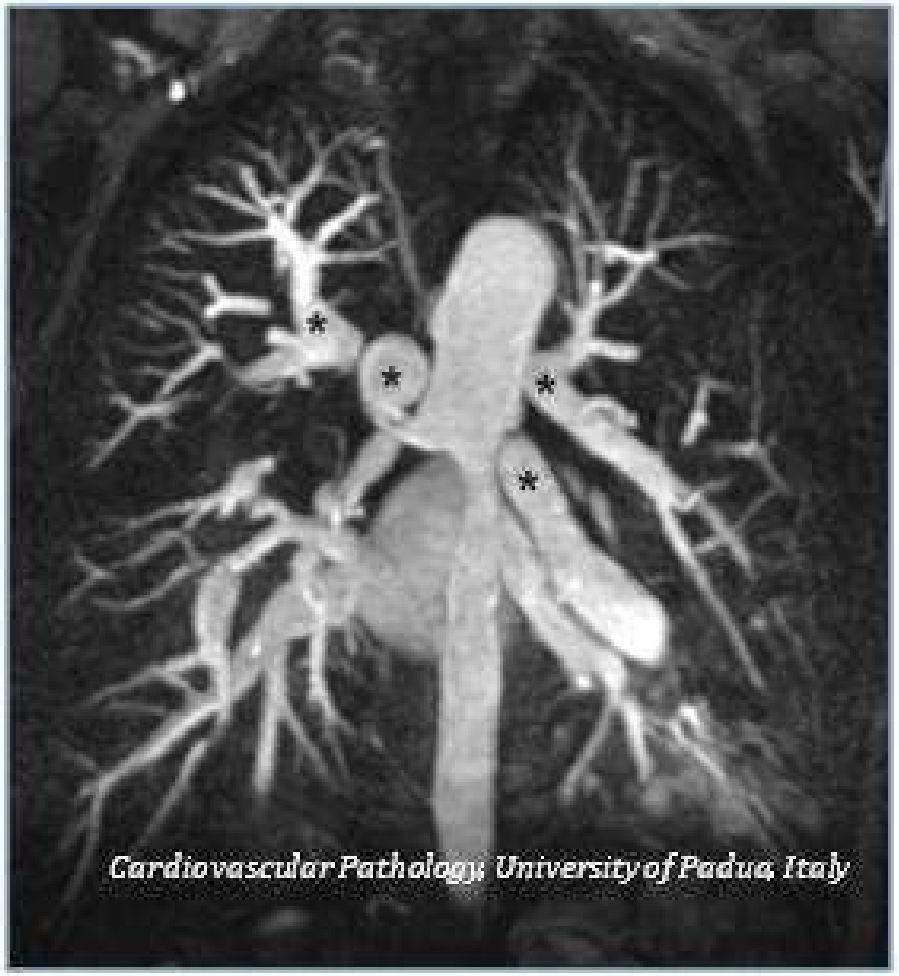
Pulmonary Atresia With VSD



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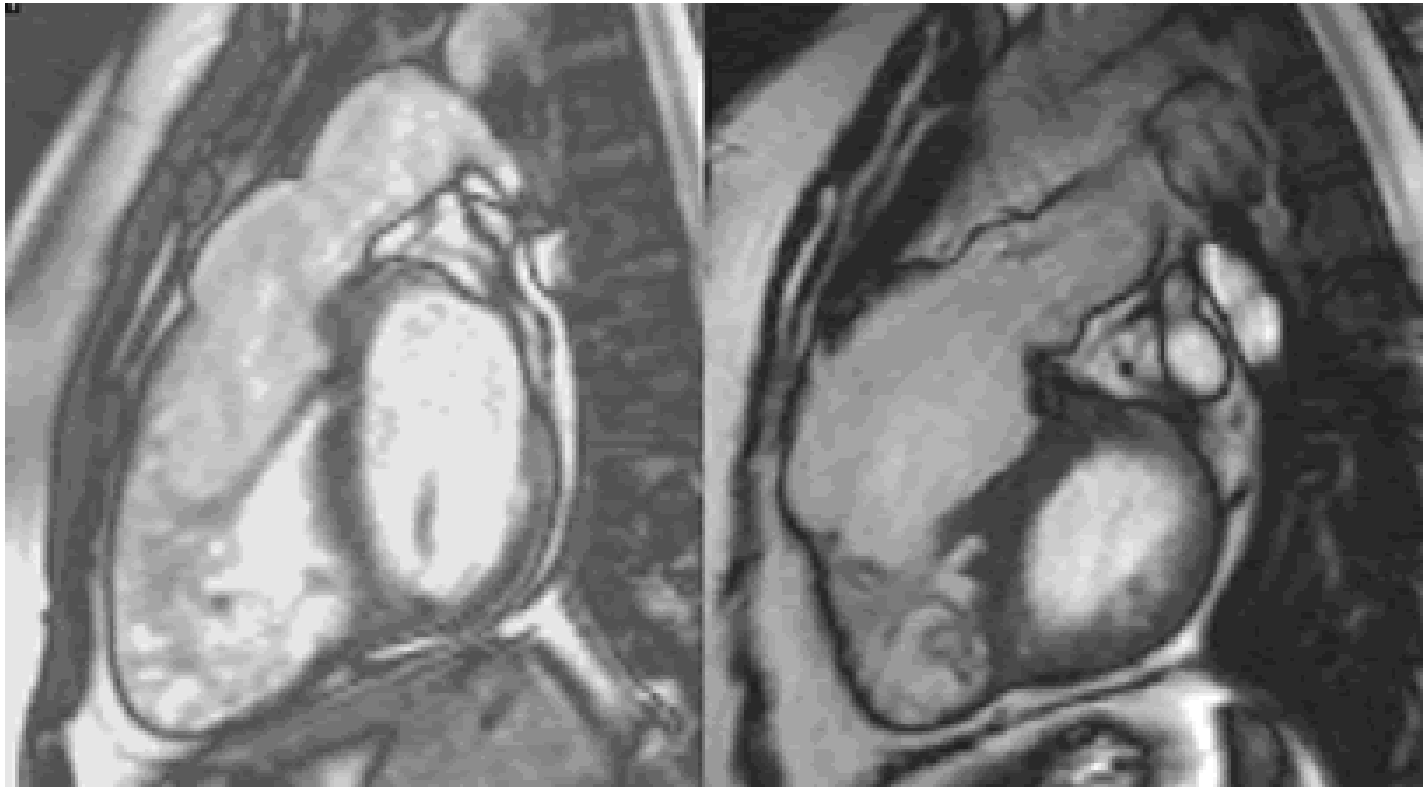
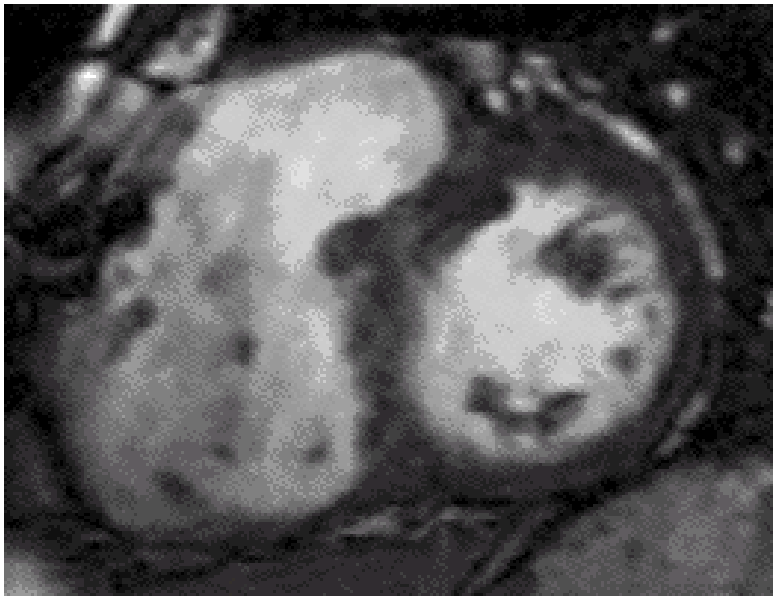
Repaired Tetralogy of Fallot

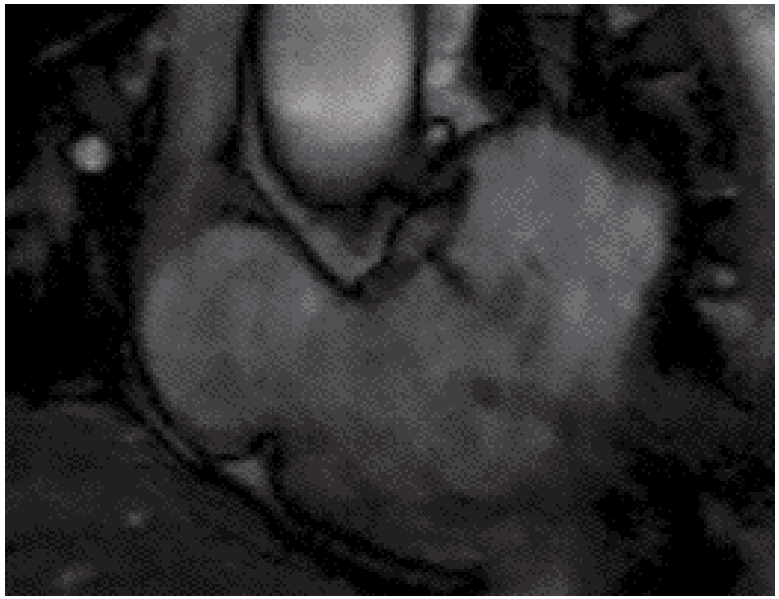
RVOT Akinesia/Aneurysm



Babu-Narayan SV: CMR of repaired tetralogy of Fallot

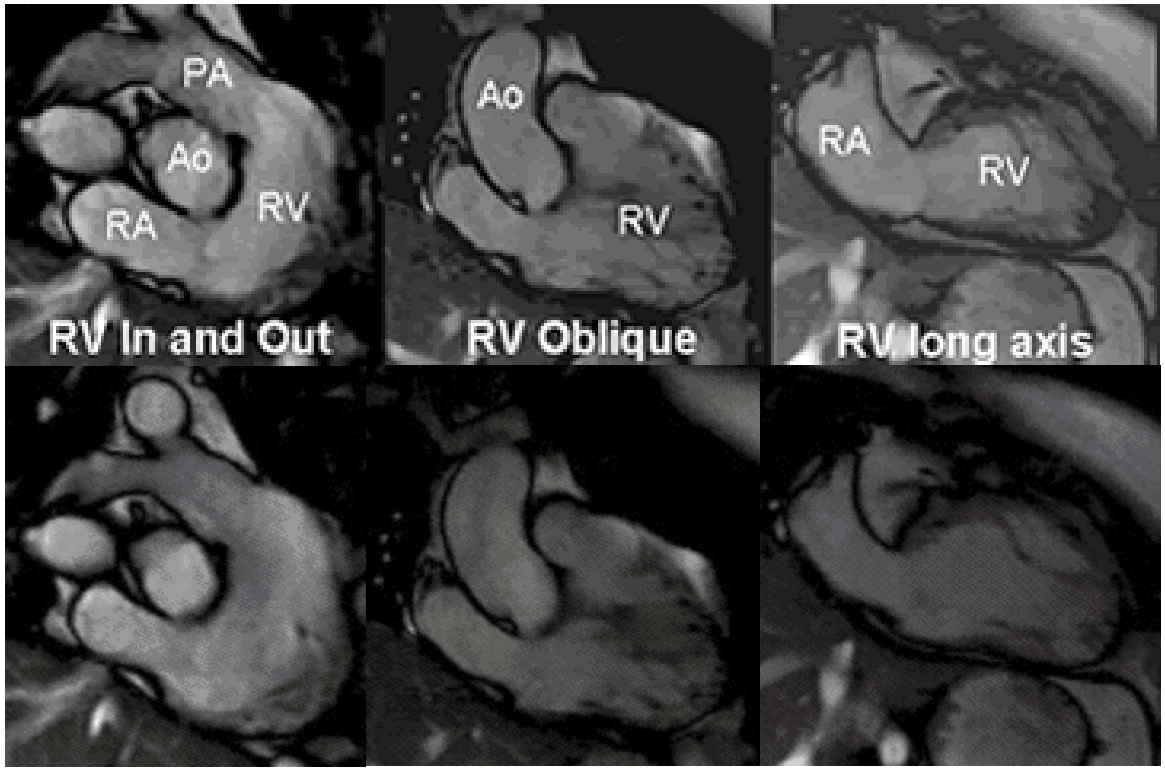
Residual VSD / Patch Leak





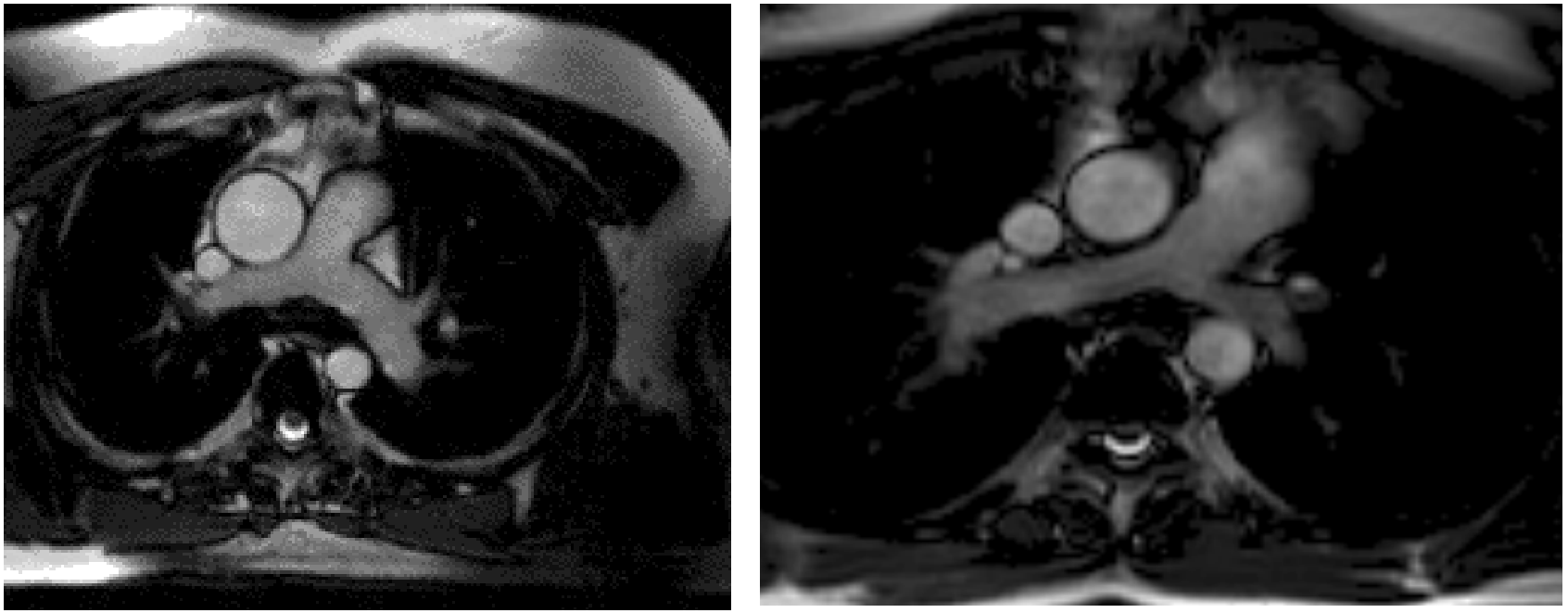
Babu-Narayan SV: CMR of repaired tetralogy of Fallot

Assessment of Regional and Global RV Function



Babu-Narayan SV: CMR of repaired tetralogy of Fallot

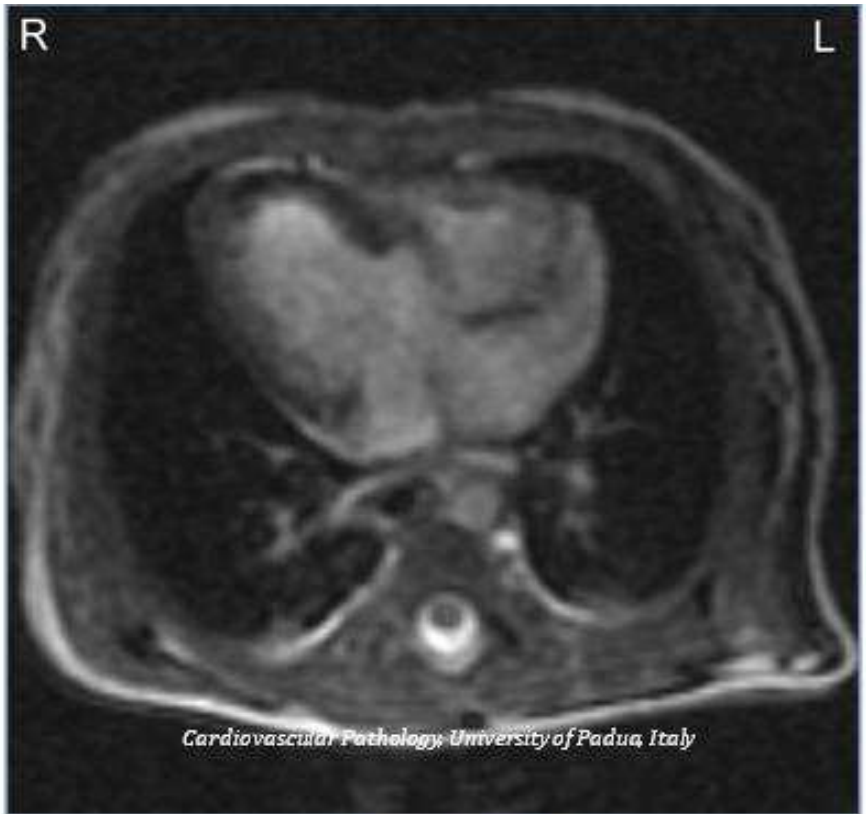
Assessment of LPA Stenosis



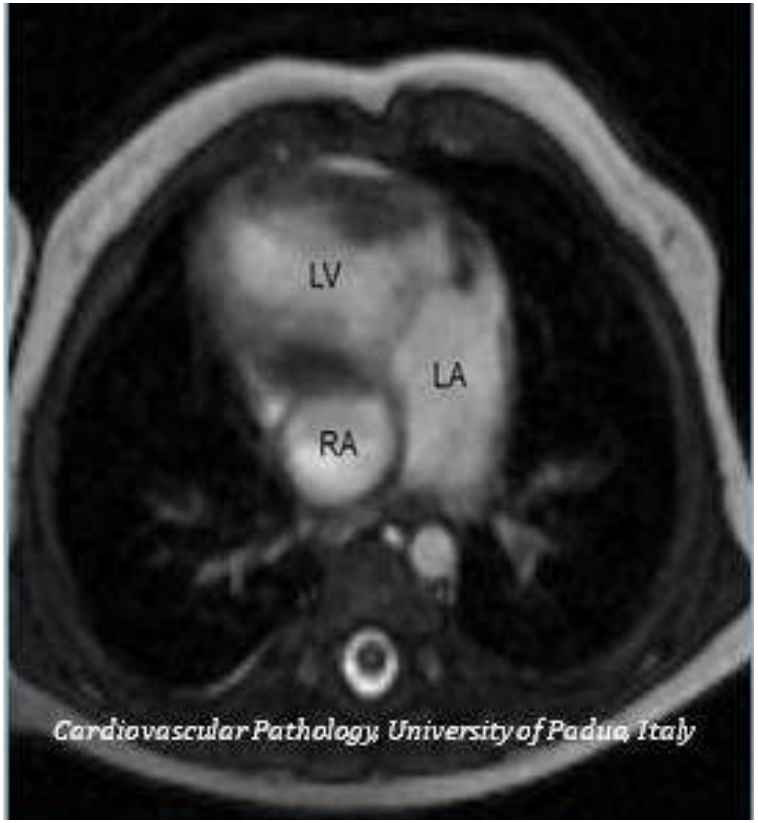
Babu-Narayan SV: CMR of repaired tetralogy of Fallot

Univentricular Heart

Double Inlet LV



Tricuspid Atresia



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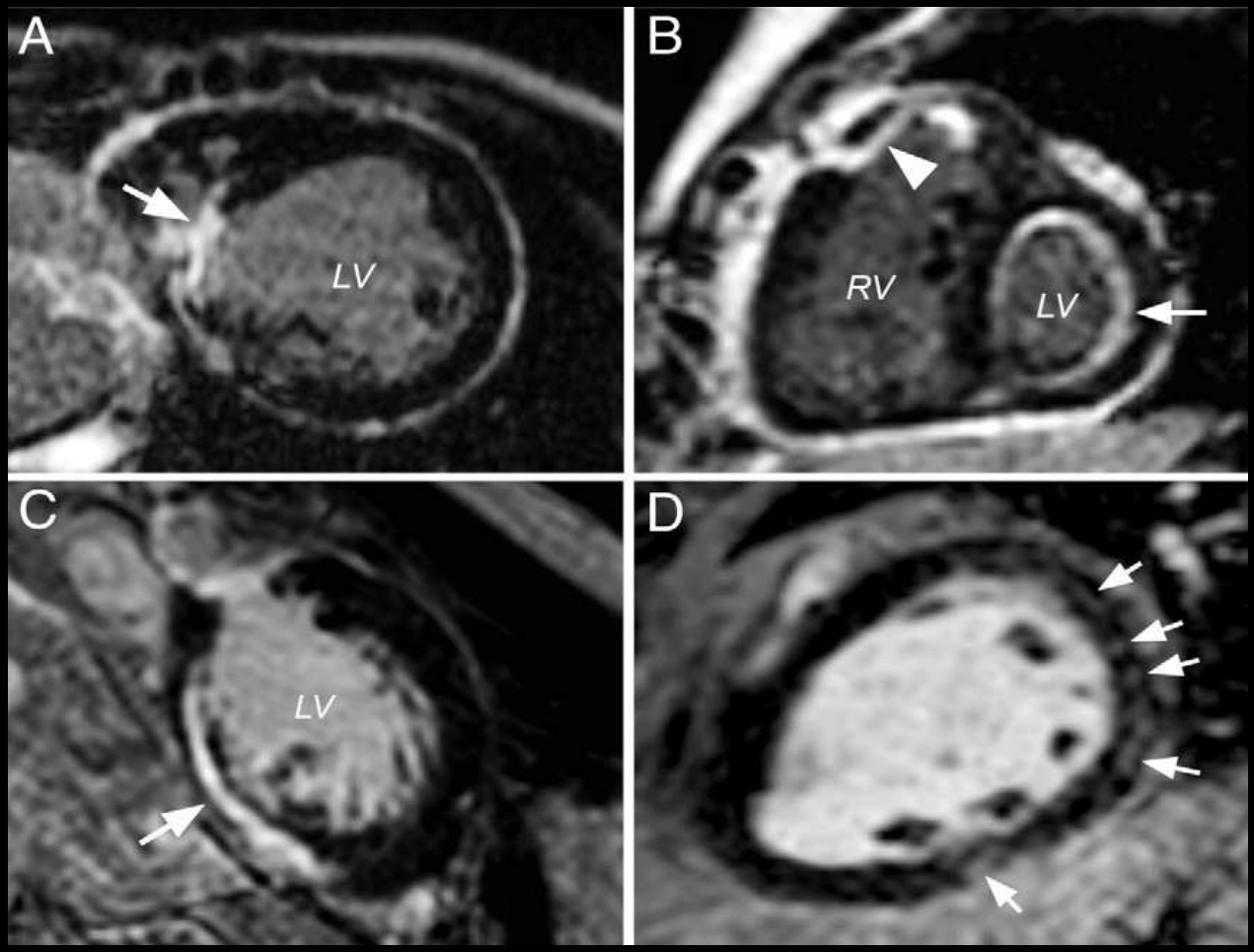
Cardiac Magnetic Resonance Versus Routine Cardiac Catheterization

- In patients with single-ventricle physiology considered for a bidirectional Glenn operation, routine cardiac catheterization is associated with higher rates of minor adverse events, longer hospital stay, and higher hospital charges than CMR.

Brown DW, et al.*Circulation. 2007;116:2718-2725*

Detection and quantification of myocardial fibrosis/scar

Locations and Patterns of LGE Late After Fontan Operation



Rathod RH, et al. JACC 2010;55:1721–8

Myocardial Fibrosis Identified by CMR Late GE Is Associated With Adverse Ventricular Mechanics and Ventricular Tachycardia Late After Fontan Operation

	All Patients (n = 90)	LGE Absent (n = 65)	LGE Present (n = 25)	p Value
EDV_i ($ml/BSA^{1.3}$)	87 [66–108]	82 [63–98]	100 [79–158]	0.004†
EDV_i (ml/BSA)	100 [76–127]	95 [73–115]	123 [92–171]	0.003†
ESV_i ($ml/BSA^{1.3}$)	36 [27–53]	34 [26–44]	63 [35–87]	<0.001†
ESV_i (ml/BSA)	41 [31–65]	39 [29–52]	66 [40–102]	<0.001†
SV_i (ml/BSA)	55 ± 18	54 ± 17	58 ± 19	0.36*
EF (%)	53 ± 12	56 ± 10	45 ± 14	<0.001*
$Mass_i$ ($g/BSA^{1.3}$)	50 [41–69]	45 [38–59]	63 [49–89]	<0.001†
$Mass_i$ (g/BSA)	57 [46–76]	52 [42–72]	73 [56–98]	0.001†
Mass/volume ratio (g/ml)	0.6 [0.5–0.7]	0.6 [0.5–0.8]	0.6 [0.5–0.7]	0.72†
RWMA	31 (34%)	18 (28%)	13 (52%)	0.05‡
Any ventricular arrhythmia	25 (28%)	13 (20%)	12 (48%)	0.02‡
Ventricular ectopy	19 (21%)	9 (14%)	10 (40%)	0.01‡
NSVT	17 (19%)	7 (11%)	10 (40%)	0.005‡
Sustained ventricular tachycardia	6 (7%)	3 (5%)	3 (12%)	0.3‡
Arrhythmia-related cardiac arrest	3 (3%)	1 (2%)	2 (8%)	0.2‡
Pacemaker	12 (13%)	10 (15%)	2 (8%)	0.5‡
Defibrillator	2 (2%)	2 (3%)	0 (0%)	1‡

Rathod RH, et al. JACC 2010;55:1721–8

Bidirectional Glenn

Aorto-Pulmonary Colleterals in a Patients with
Complete Obliteration of the Left-sided Pulmonary Veins

Indications where CMR should be regularly used (superior to echo) when the information is essential for patient management

- Quantification of RV volumes and right ventricular ejection fraction [tetralogy of Fallot, systemic RV]
- Evaluation of the RVOTO and RV–pulmonary artery conduits
- Quantification of pulmonary regurgitation
- Evaluation of pulmonary arteries (stenoses, aneurysms) and the aorta (aneurysm, dissection, coarctation), of systemic and pulmonary veins (anomalous connection, obstruction, etc.)
- Collaterals and arteriovenous malformations (CT is superior)
- Coronary anomalies and coronary artery disease (CT is superior)
- Evaluation of intra- and extracardiac masses (CT is superior)
- Quantification of myocardial mass (LV and RV)
- Detection and quantification of myocardial fibrosis/scar (gadolinium LE)
- Tissue characterization (fibrosis, fat, iron, etc.)

Kilner PJ, et al: *ESC Guidelines for CMR in GUCH*. EHJ 2010

Limitations of CMR in GUCH

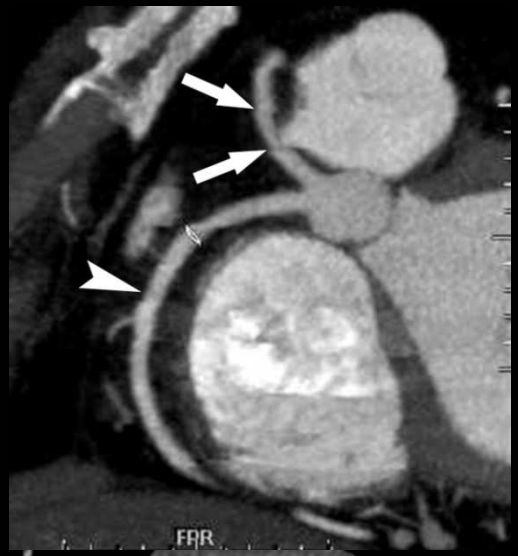
- ECG-gated cine images and flow maps are typically acquired over a breath-hold and not in real time. Because of the acquisition period and the typical dimensions of the voxels, thin mobile structures may not be well seen
- Experience is needed for appropriate velocity acquisition and interpretation
- Cardiovascular magnetic resonance lacks the portability of echo and is not available during open heart surgery
- Implanted pacemakers and defibrillators generally preclude CMR

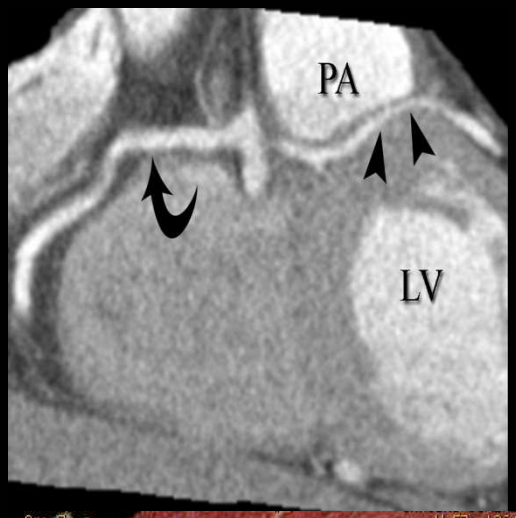
Computed Tomography

- CT plays an increasing role in imaging of GUCH patients, providing excellent spatial resolution and rapid acquisition time.
- It is particularly good for imaging epicardial coronary arteries and collateral arteries, and for parenchymal lung disease.
- Ventricular size and function can be assessed, with inferior temporal resolution compared with CMR

Multi Detection Computed Tomography

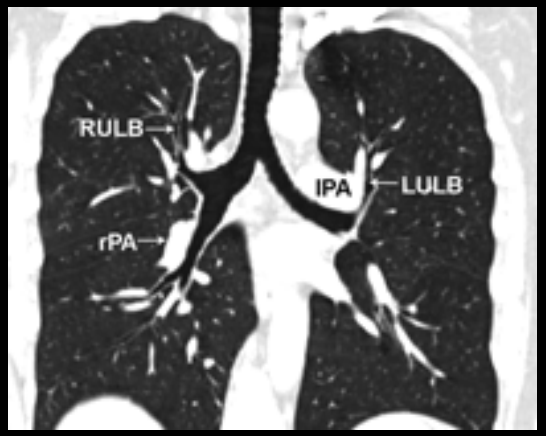
Anomalous Coronary Arteries

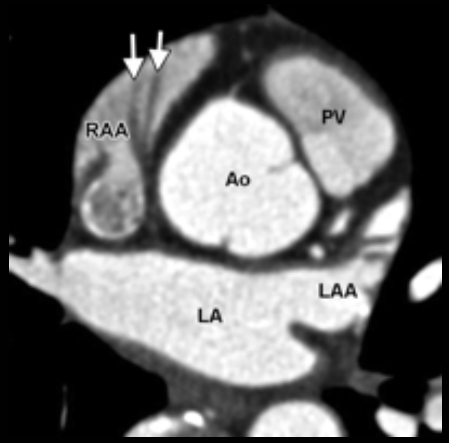


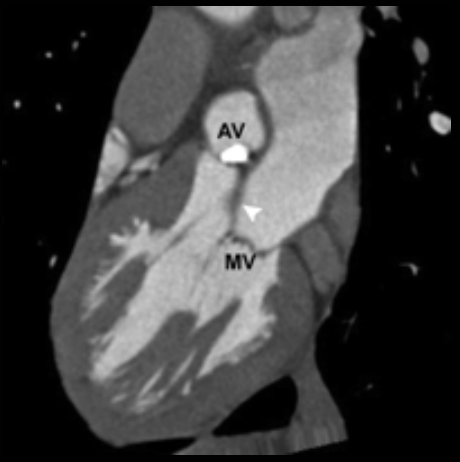


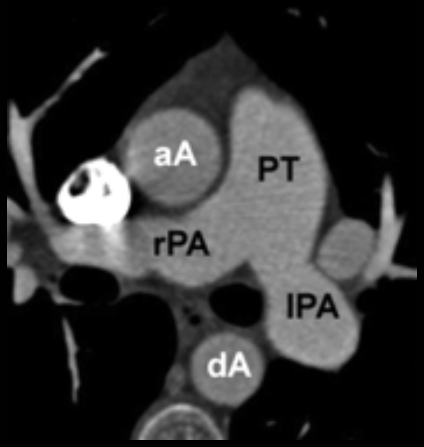
Multi Detection Computed Tomography

Sequential-Segmentary Approach

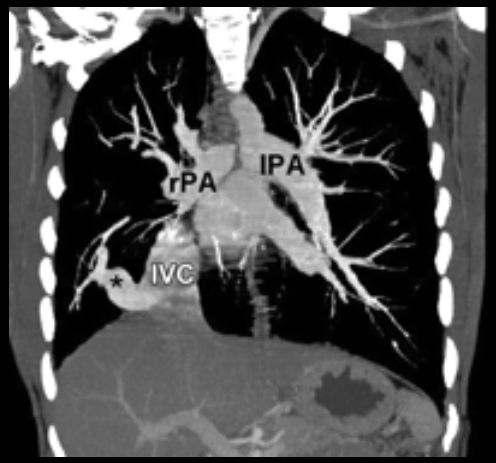


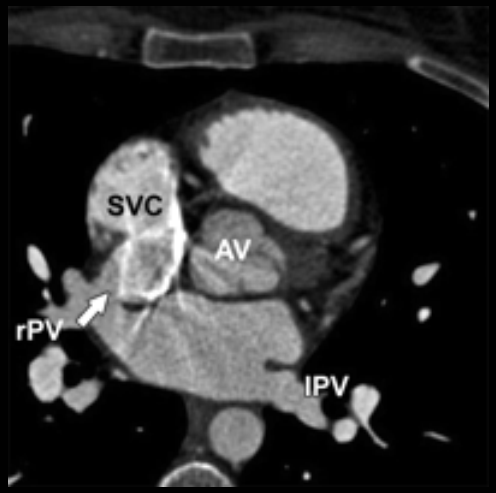


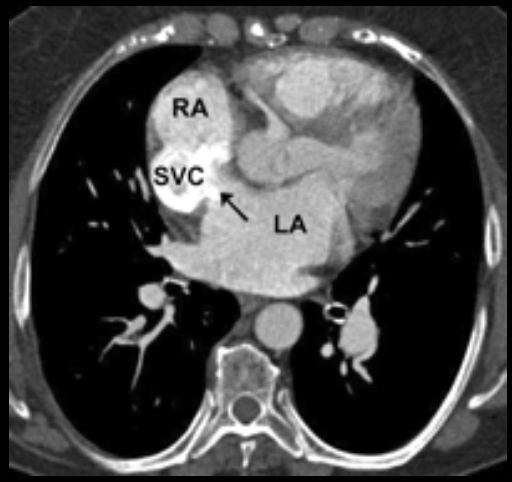




MDCT- Cardiac and Extracardiac Congenital Anomalies







Computed Tomography

- The major drawback of most current CT systems is its high dose of ionizing radiation, making serial use unattractive.
- CT is currently more widely available than CMR and thus plays a role in acute situations.
- Moreover, recent developments, such as ECG triggered acquisition and newer rotational techniques, reduce the amount of radiation substantially, which may possibly make CT a more attractive alternative to CMR in the coming years

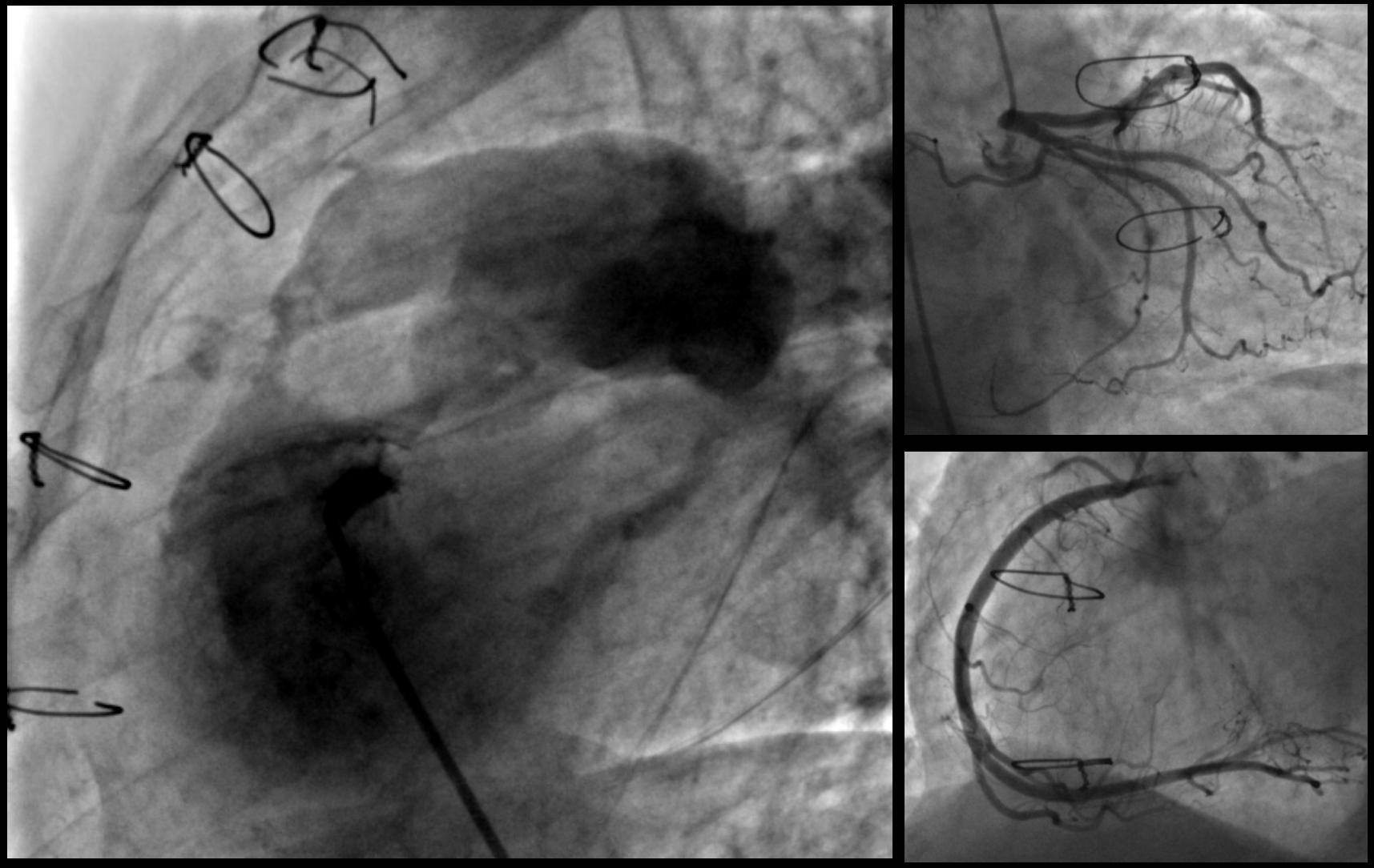
Cardiac Catheterization

Cardiac catheterization is now reserved for resolution of specific anatomical and physiological questions, or for intervention

- Continuing indications include assessment of PVR, LV and RV diastolic function, pressure gradients, and shunt quantification when non-invasive evaluation leaves uncertainty, coronary angiography, and the evaluation of extracardiac vessels such as aortic pulmonary collateral arteries.
- In shunt lesions with Doppler echocardiographically documented pulmonary hypertension (PAP >50% of systemic pressure), catheterization remains essential for therapeutic decision making [if PAH is severe, testing of vasoreactivity may be required for the decision to intervene (shunt closure)]
- Before surgery, coronary angiography should be performed in men ≥40 years of age, postmenopausal women, and patients with signs of or risk factors for CAD

RV–Pulmonary Artery Conduits Obstruction

76 years old



Conclusions

- Strategies for investigation of anatomy and physiology of CHD are changing rapidly, with a shift from invasive studies to noninvasive protocols involving not only echocardiography but, more recently, CMR and cardiac CT.
- Echocardiography remains the first-line investigation and continues to evolve
- CMR has become increasingly important in GUCH patients (particularly useful for volumetric measurements, assessment of vessels, and detection of myocardial fibrosis)
- CT plays an increasing role in imaging of GUCH patients, providing excellent spatial resolution and rapid acquisition time.