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I Master di Risonanza Magnetica Cardiaca

Cardiopatie congenite – Parte 1

Dott.ssa Veronica Spadotto

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CARDIOPATIE CONGENITE

- **ANATOMIA**

- **VALUTAZIONE FUNZIONALE**
  - **Funzione ventricolare**
  - **Funzione valvolare**
  - **Shunts/direzioni dei flussi**

- **APPLICAZIONI ULTERIORI**
  - **Caratterizzazione tissutale**

CARDIOPATIE CONGENITE

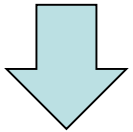
ANATOMIA delle cardiopatie congenite (non corrette vs corrette)

Conoscenza clinica del tipo di malformazione, degli interventi correttivi e delle possibili conseguenze a lungo termine

CARDIOPATIE CONGENITE

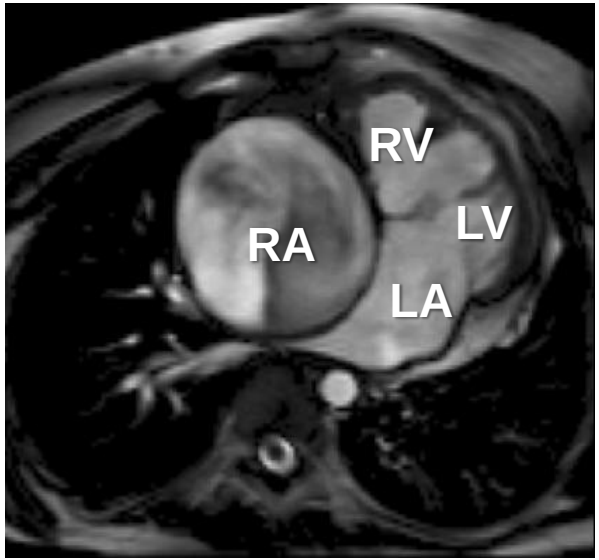
ANATOMIA delle cardiopatie congenite (non corrette vs corrette)

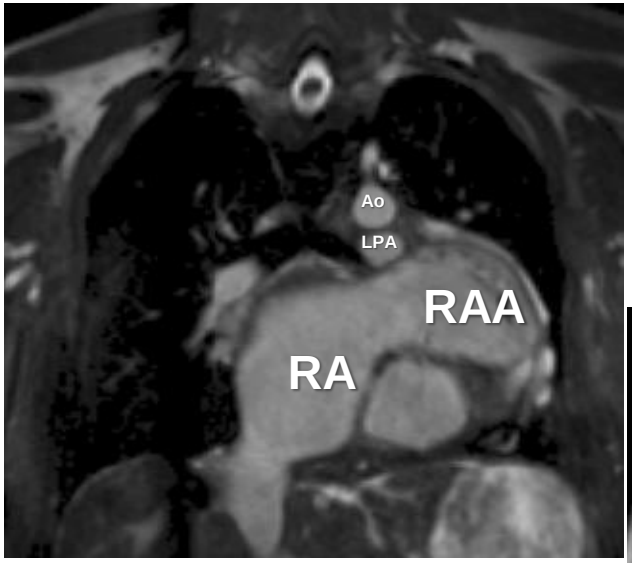
Conoscenza clinica del tipo di malformazione, degli interventi correttivi e delle possibili conseguenze a lungo termine

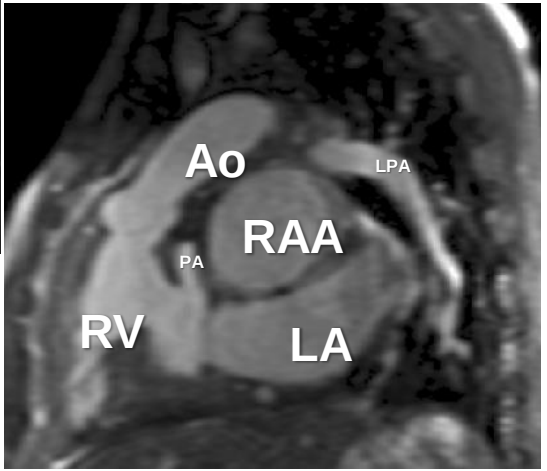


SCELTA del protocollo MRI

CARDIOPATIE CONGENITE







CARDIOPATIE CONGENITE

Sequenze maggiormente utilizzate in MRI per la valutazione delle CHD

CINE imaging
Phase velocity Mapping
4D flow sensitive velocity mapping
Contrast Enhanced Magnetic Resonance angiography (CE-MRA)
3D steady-state free processing non contrast imaging (3D SSFP)

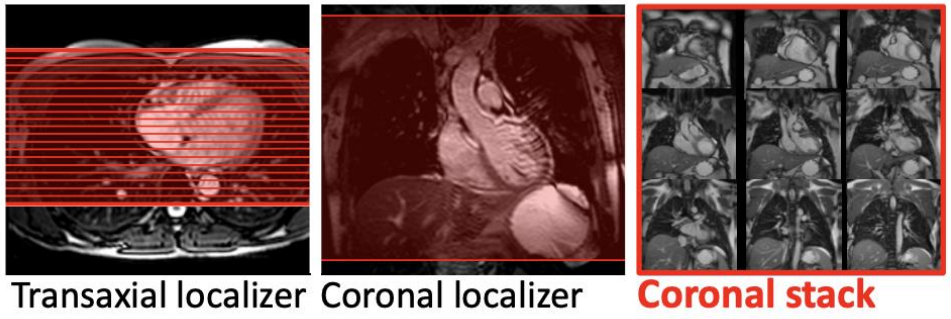
CARDIOPATIE CONGENITE

MULTISLICE IMAGING

Per iniziare: stacks multislices trasversi, coronali e sagittali, es SSFP bright-blood > 20 slices almeno ad ogni sospensione del respiro

Costituiscono la base per le successive acquisizioni cine

CARDIOPATIE CONGENITE



«stacks» anatomici

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CINE imaging

Permette la visualizzazione dei flussi e dei movimenti del cuore e delle pareti vascolari (con gating ECG; a respiro trattenuto o libero)

- Acquisire stacks contigui di immagini transassiali o coronali per coprire l'intero volume cardiaco e il mediastino
- Spessore slices 5-7 mm
- Attenzione! Strutture sottili (valvole, zone periferiche dei jets) si vedono bene solo se perpendicolari alla slice

SSFP cine imaging: buon contrasto sangue-tessuto > buona misurazione di volumi/masse e valutazione delle valvole.

CARDIOPATIE CONGENITE

SSFP cine imaging: buon contrasto sangue-tessuto > buona misurazione di volumi/masse e valutazione delle valvole.
Acquisizioni ripetibili e precise grazie al trattenimento del respiro

A. «Cross cutting» dei jet valvolari o delle valvole (posizionamento di una slice ortogonale per visualizzarne orientamento e direzione)
B. Stack obliquo di immagini cine con slices di circa 5 mm, senza gaps

NB maggiori artefatti in caso di disomogenità di B0 (devices)

CARDIOPATIE CONGENITE

CARDIOPATIE CONGENITE

Table 2 Cine steady-state free precession

	Infant/small child		Large child/adult
In-plane resolution (mm)	1.2-2.0		1.5-2.5
Slice thickness (mm)	4-6		5-8
Inter slice gap (mm)	0-2		0-4
Respiratory compensation	Free-breathing	Breath-holding	Breath-holding
Number of signal averages	3	1	1
Reconstructed phases per R-R interval	20-30		
ECG gating	Retrospective		

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MODULO «VENTRICOLOGRAFIA»

Cine CMR: 2 CH e 3 CH del ventricolo sinistro, 3 CH ventricolo destro, 4 camere
Cine CMR: stack di slices continue a coprire entrambi i ventricoli (asse corto e/o assiale)

Key reporting elements: LVEDV, LVESV, RVEDV, RVESV, LVSV, RVSV, LVEF, RVEF, massa LV (e RV), cinetica regionale

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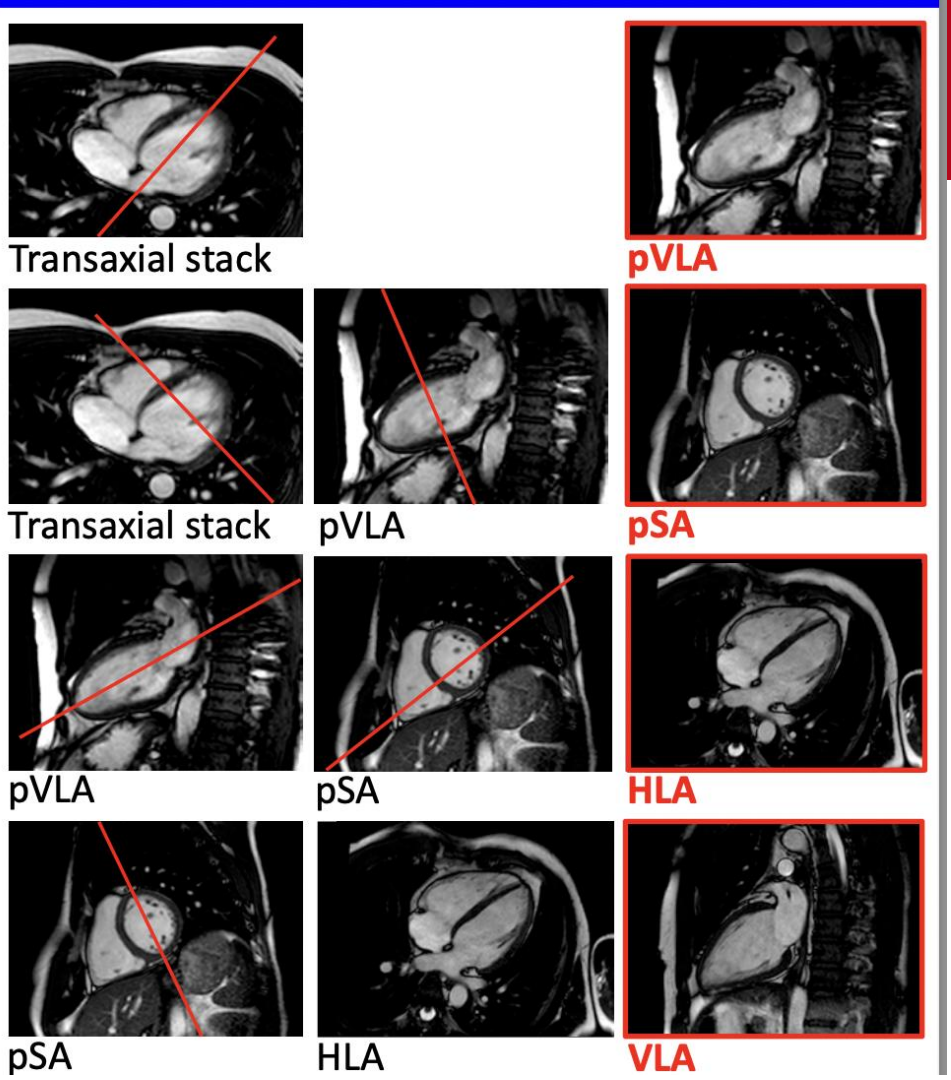

Planning del modulo «ventricolografia»

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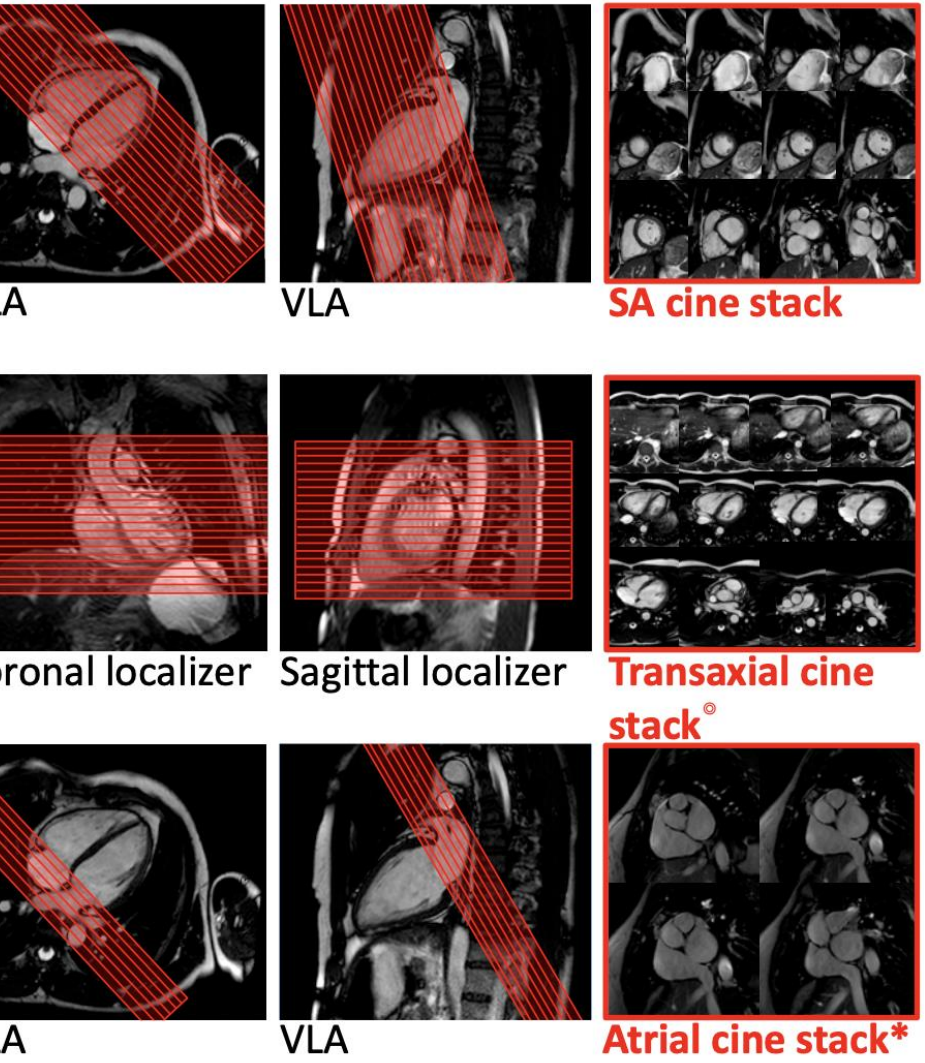

Planning del modulo «ventricolografia»

Adattato da: ESC/EACVI CHD/CMR guide, 2014

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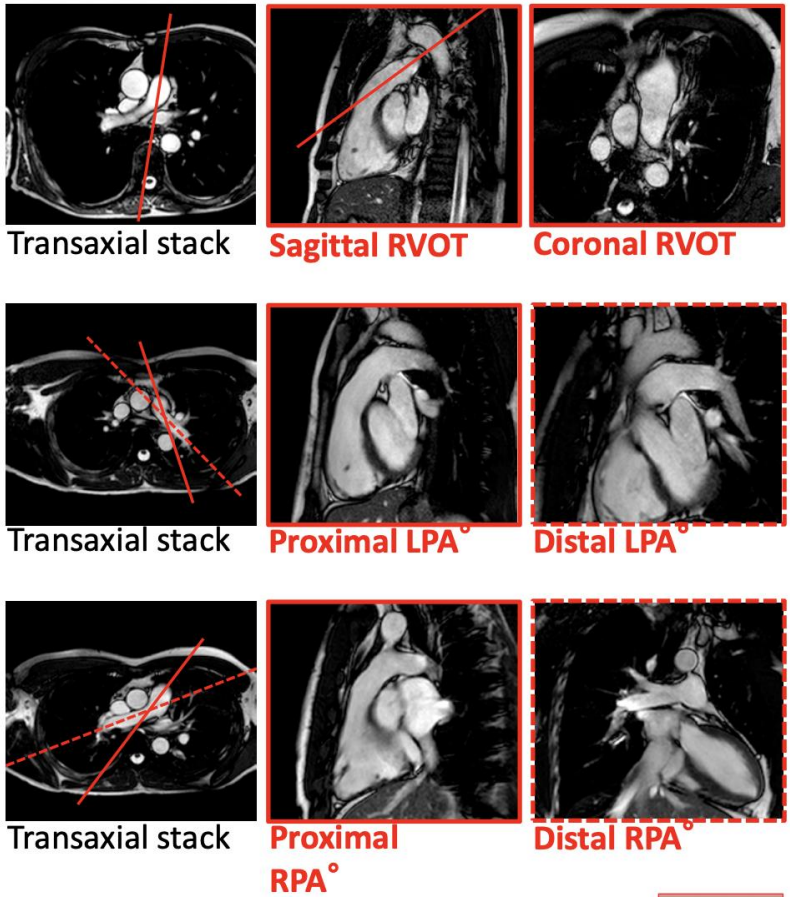

Piani utili nelle CHD

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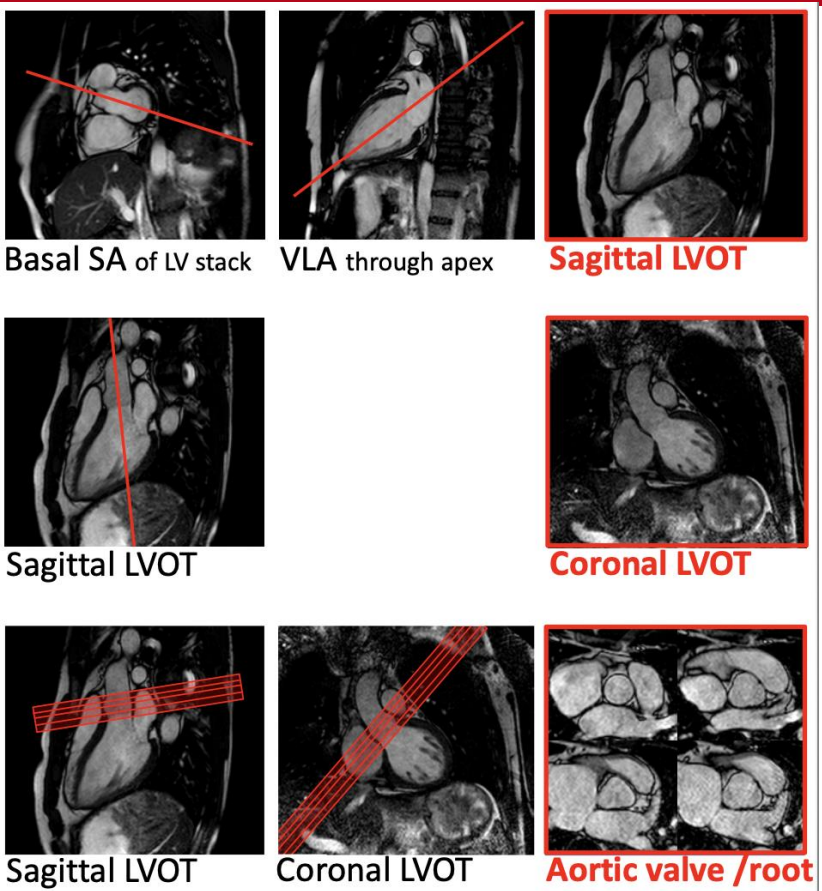

Piani utili nelle CHD

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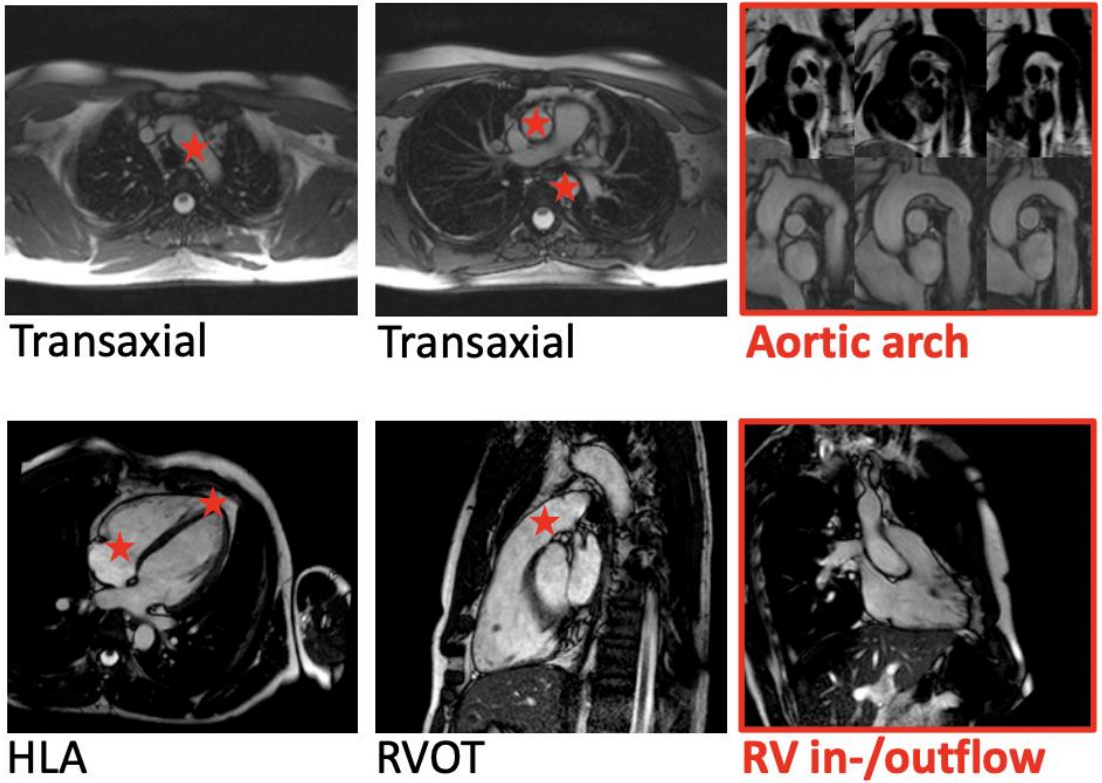

Piani utili nelle CHD

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CARDIOPATIE CONGENITE

«Black blood»imaging (fast spin echo – FSE, ECG gated):
Utile per visualizzazione statica delle strutture vascolari (il sangue ad alto flusso risulta nero)
Acquisizione in un solo battito cardiaco (single shot) o multipli (fast spin echo) > risoluzione spaziale migliore
Meno suscettibili agli artefatti da flusso turbolento/devices
Possibili slices anche di 2 mm
Possibile pesatura in T1 e T2 per caratterizzazione tissutale

CARDIOPATIE CONGENITE

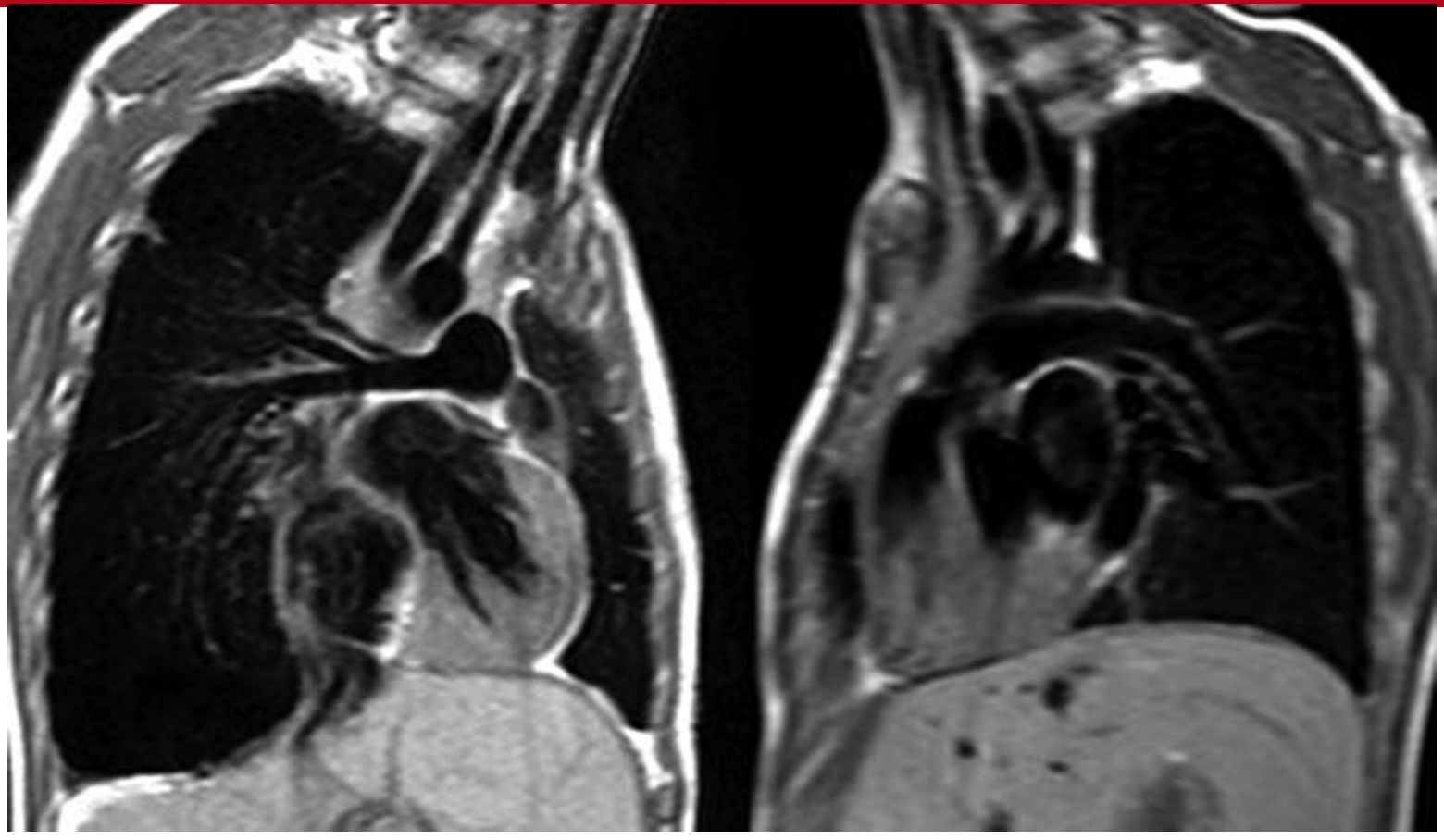

Table 1 Fast (turbo) spin echo

	Infant/small child		Large child/adult
In-plane resolution (mm)	1.0-2.0		1.5-2.5
Slice thickness (mm)	2-3		4-6
Echo train length	12-24		16-32
Image acquisition timing	3-4 R-R		1-2 R-R
Respiratory compensation	Free-breathing	Breath-holding	Breath-holding
Number of signal averages	3	1-2	1
Trigger delay	Diastole or systole		Diastole

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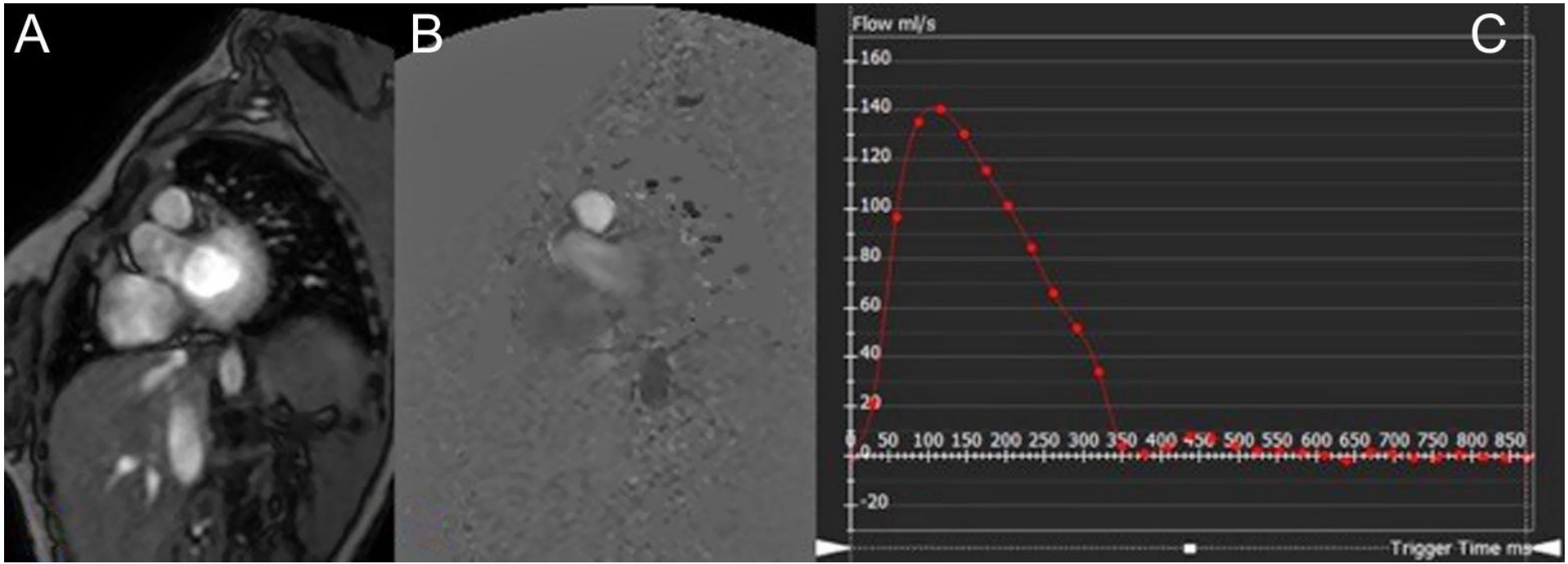
PHASE VELOCITY MAPPING

Forniscono misure adeguate di velocità e volumi di flusso (es. gittata cardiaca, quantificazione shunts, quantificazione di flusso attraverso collaterali, volumi di rigurgito, velocità del jet attraverso le stenosi)

Velocity encoding direction: parallela o perpendicolare al piano

Perpendicolare (nella direzione del selection gradient) per misurare il volume del flusso

Misura area della sezione del lume e della velocità media del flusso attraverso l'area > curva di flusso


A
B
C
Flow ml/s
160
140
120
100
80
60
40
20
0
-20
0 50 100 150 200 250 300 350 400 450 500 550 600 650 700 750 800 850
Trigger Time ms

CARDIOPATIE CONGENITE

Table 4 Velocity-encoded phase-contrast cine for quantitative flow measurements

	Infant/small child	Large child/adult
In-plane resolution (mm)	1.0-1.3	1.3-2.0
Slice thickness (mm)	5	6-8
Number of signal averages	3	
Reconstructed phases per R-R interval	25-30	
Velocity encoding (cm/s)	Artery 200, vein 100, atrioventricular-valve inflow 150	
Cardiac/respiratory motion	Retrospective ECG-gating with free-breathing	

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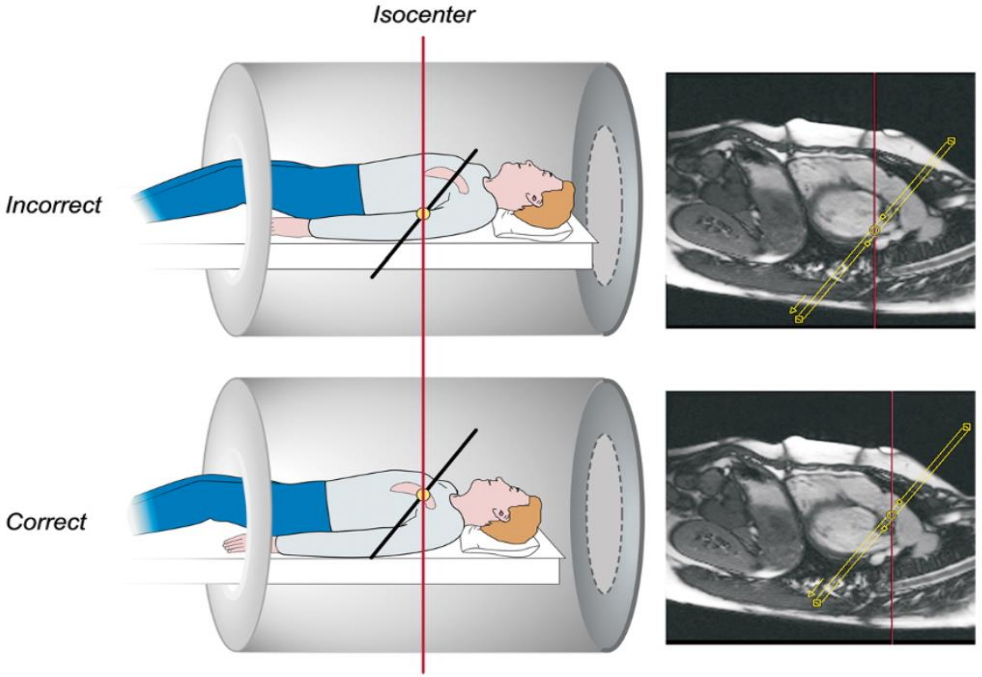

Figure 11 Proper positioning of the imaging plane for main pulmonary artery blood flow measurement. PC CMR velocity and flow measurements are most accurate when the location of interest is at the isocenter of the scanner during the acquisition. Most MR scanners will slide the patient table so that the center of the imaging plane (yellow circle) is at the scanner's isocenter (vertical red line). Prescribing the imaging plane so that the center of the image is at the same level in the superoinferior dimension as the location of interest is therefore recommended.

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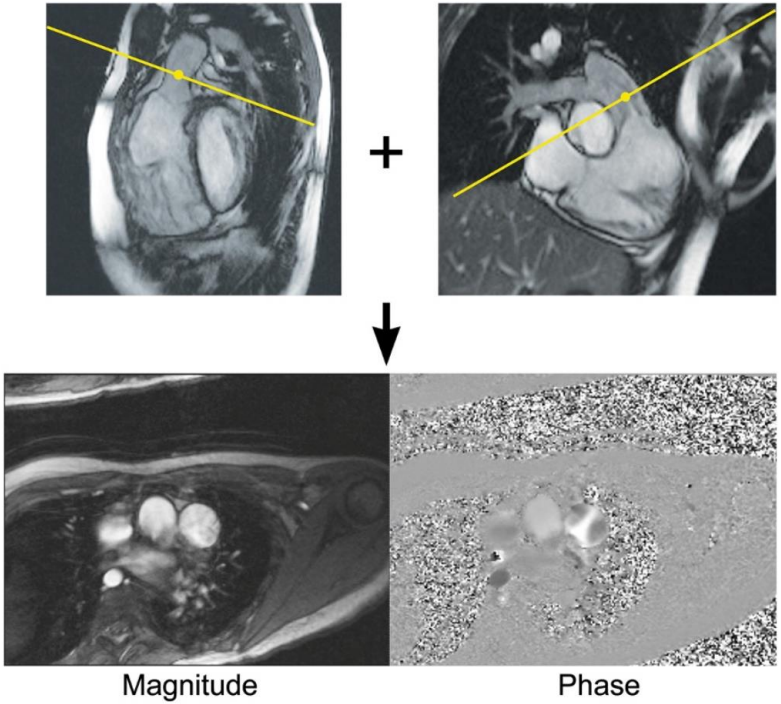

Figure 10 Planning a CMR phase-contrast acquisition to measure flow in the main pulmonary artery. The PC CMR imaging plane is simultaneously viewed and adjusted on orthogonal views of the main pulmonary artery (top row) to ensure that it is oriented perpendicular to the blood vessel. The resulting magnitude and phase images are shown (bottom row).

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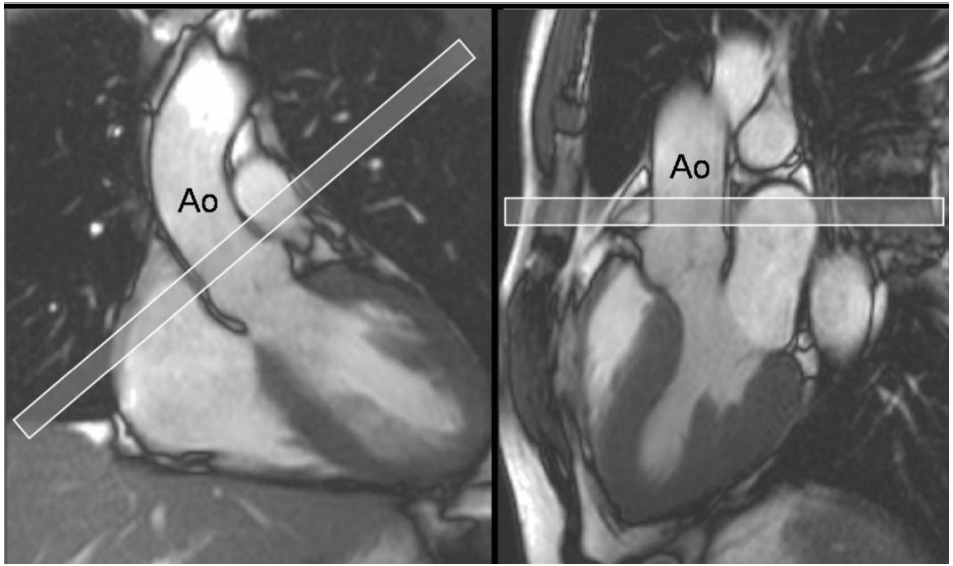

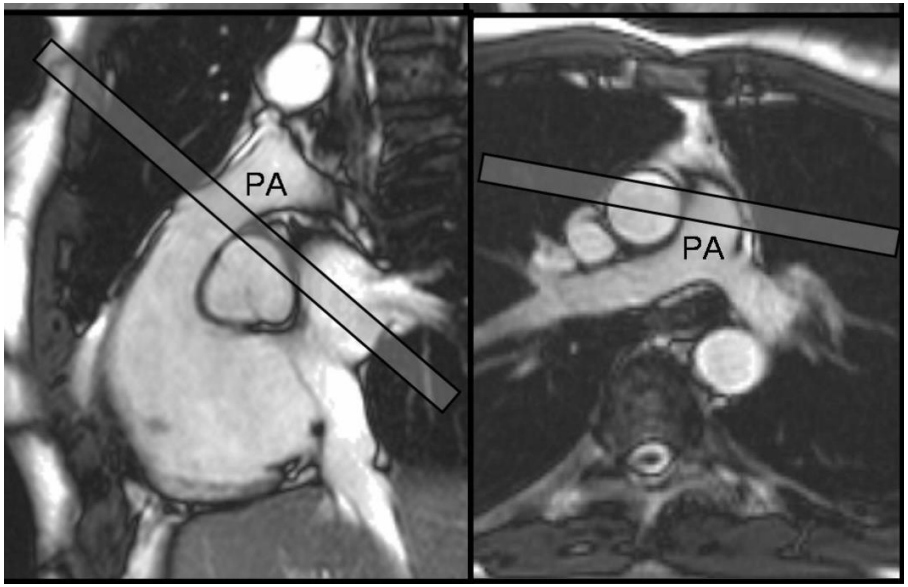

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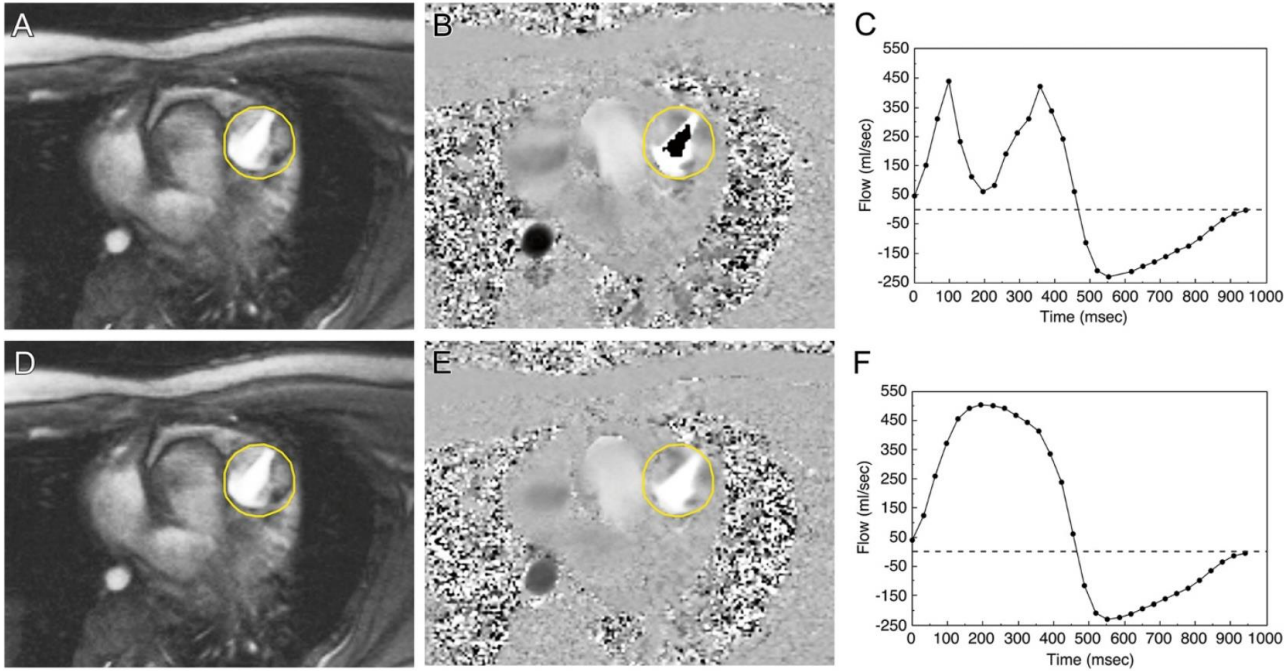

Figure 9 Effect of aliasing on phase-contrast cine CMR (PC CMR) flow measurements. Sixteen-year-old patient with surgically repaired tetralogy of Fallot and mild pulmonary valve stenosis. PC CMR was performed in the main pulmonary artery with the velocity range (v_{enc}) set incorrectly at 200 cm/sec (top row) and then with the v_{enc} set correctly at 300 cm/sec (bottom row). Magnitude (**A**, **D**) and phase images (**B**, **E**) in systole, and the resulting flow curves (**C**, **F**) generated from analyzing the region of interest (yellow contour) are shown. Because the peak velocity is 260 cm/sec, aliasing (**B**) and flow underestimation (**C**) are seen with a v_{enc} of 200 cm/sec but not with a v_{enc} of 300 cm/sec (**E** and **F**).

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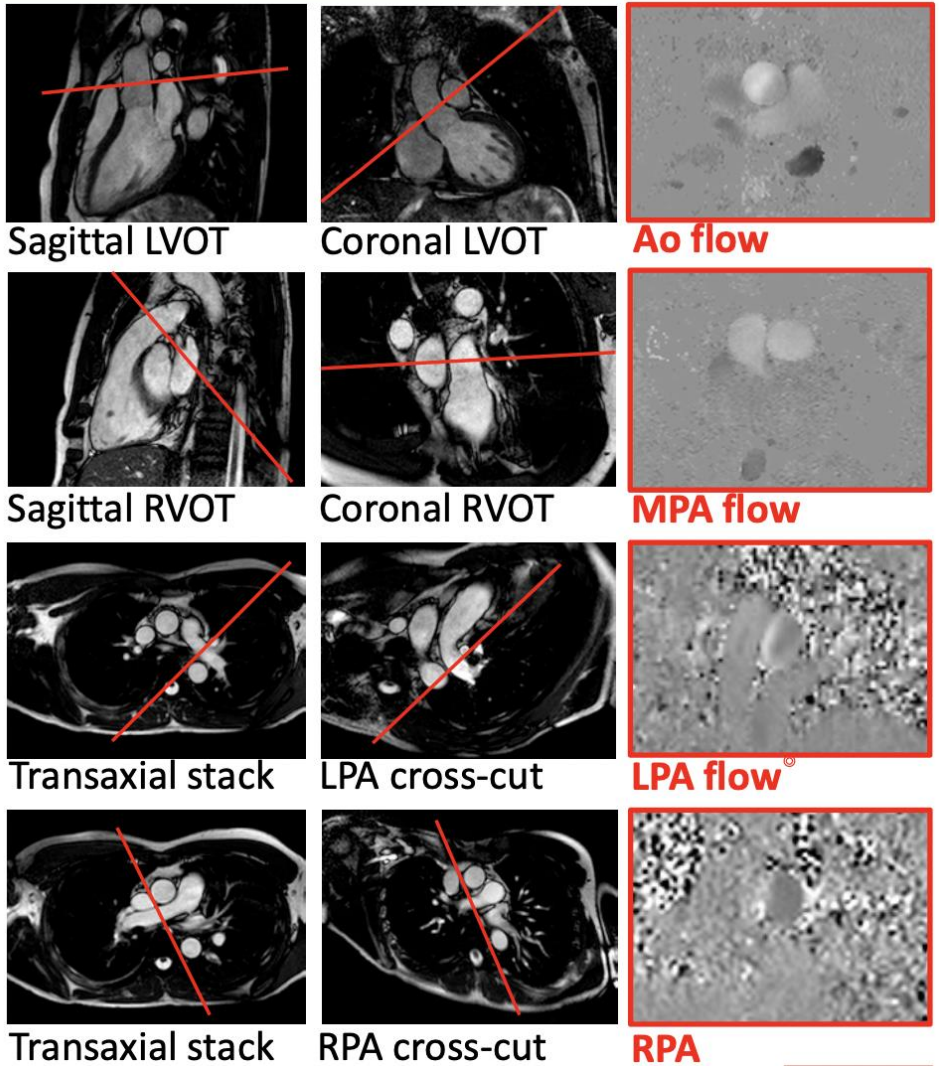

PC- Piani utili nelle CHD

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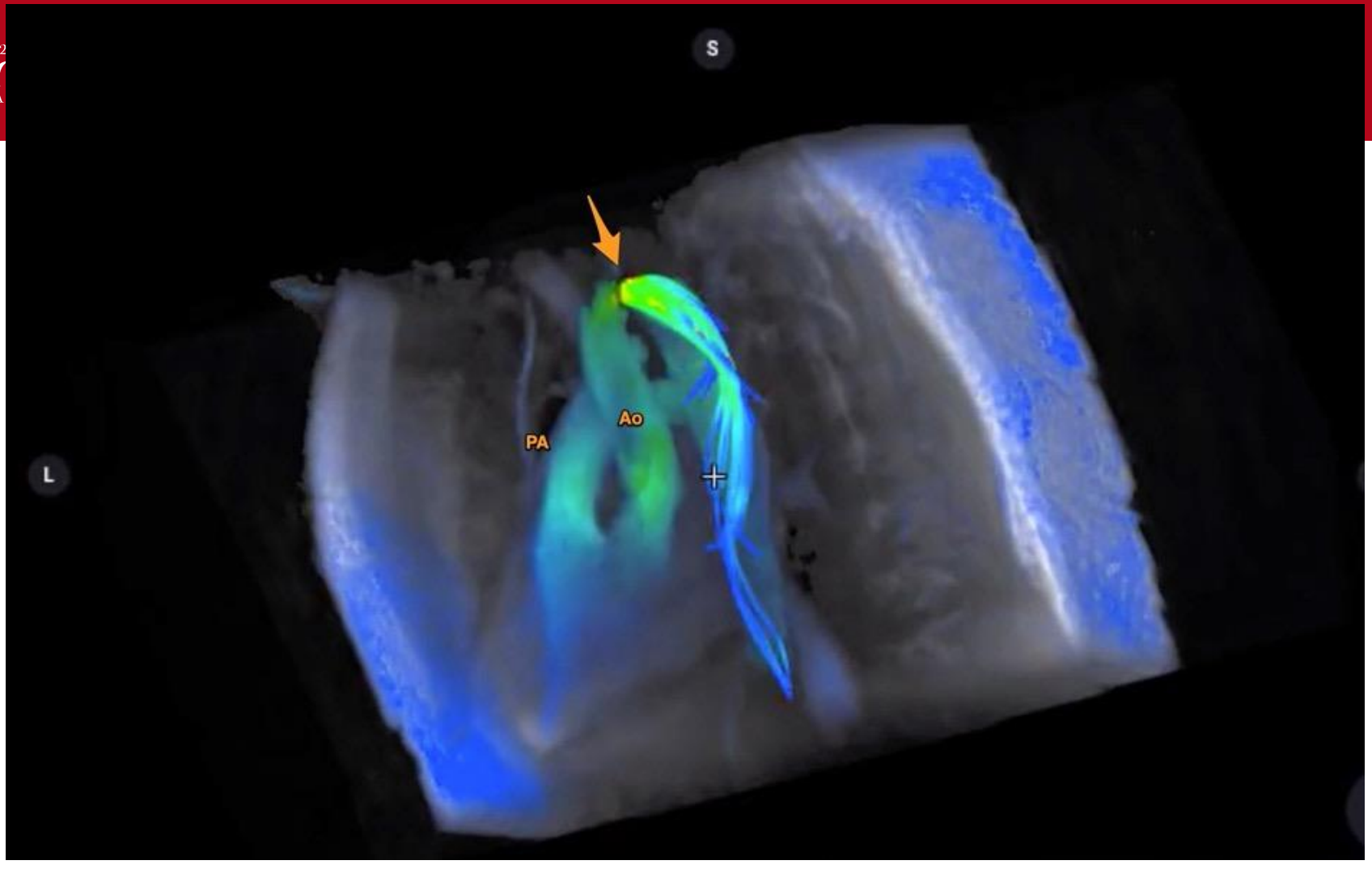
CARDIOPATIE CONGENITE

4D flow sensitive velocity mapping

- Misurazione di tutte le tre componenti spaziali dei flussi E della componente temporale durante il ciclo cardiaco > quantificazione e visualizzazione di jet anche complessi
- Possibilità di acquisizione di grossi volumi con una acquisizione singola
- Possibilità di analisi dell'immagine anche a posteriori in qualunque piano contenuto nel volume di acquisizione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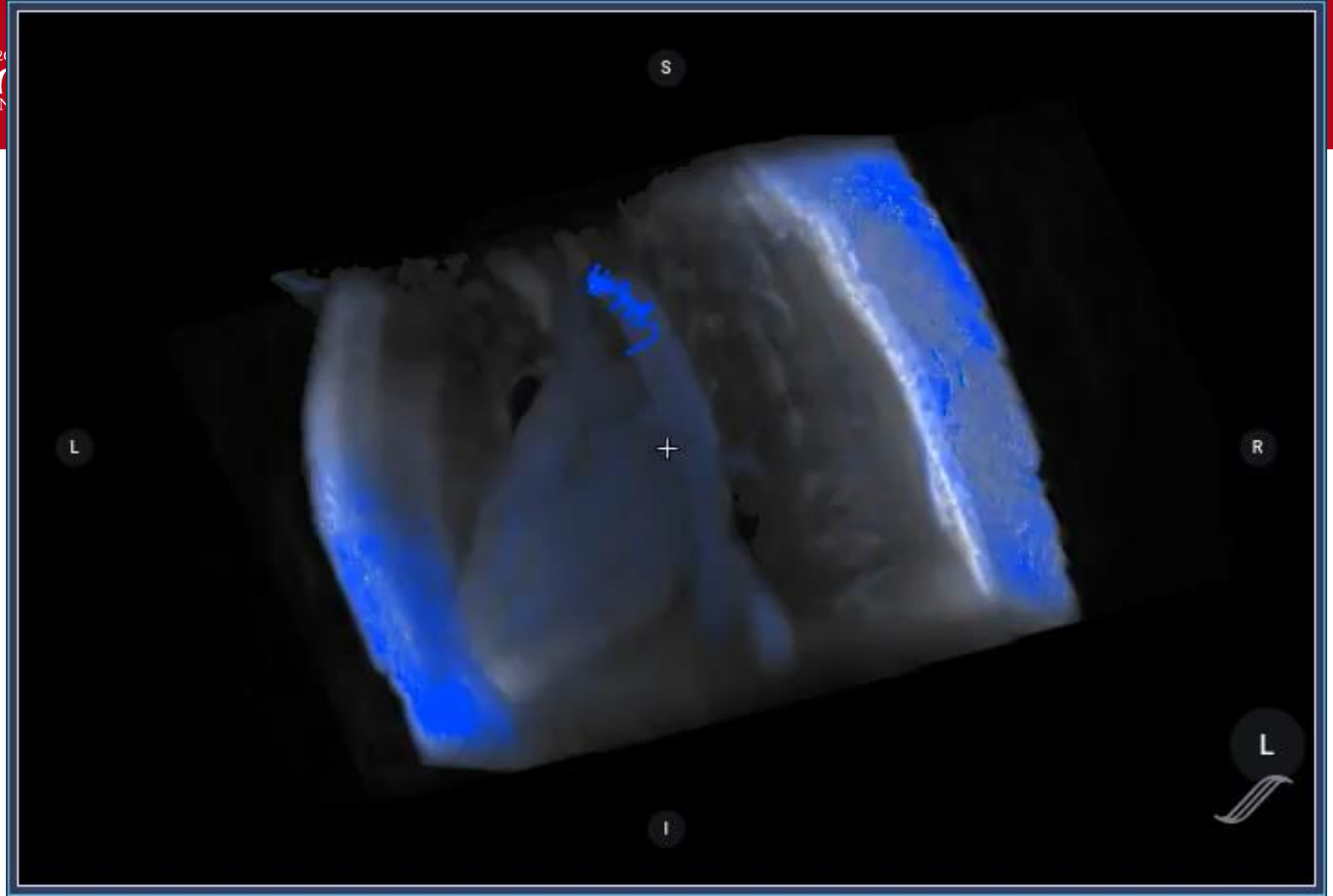
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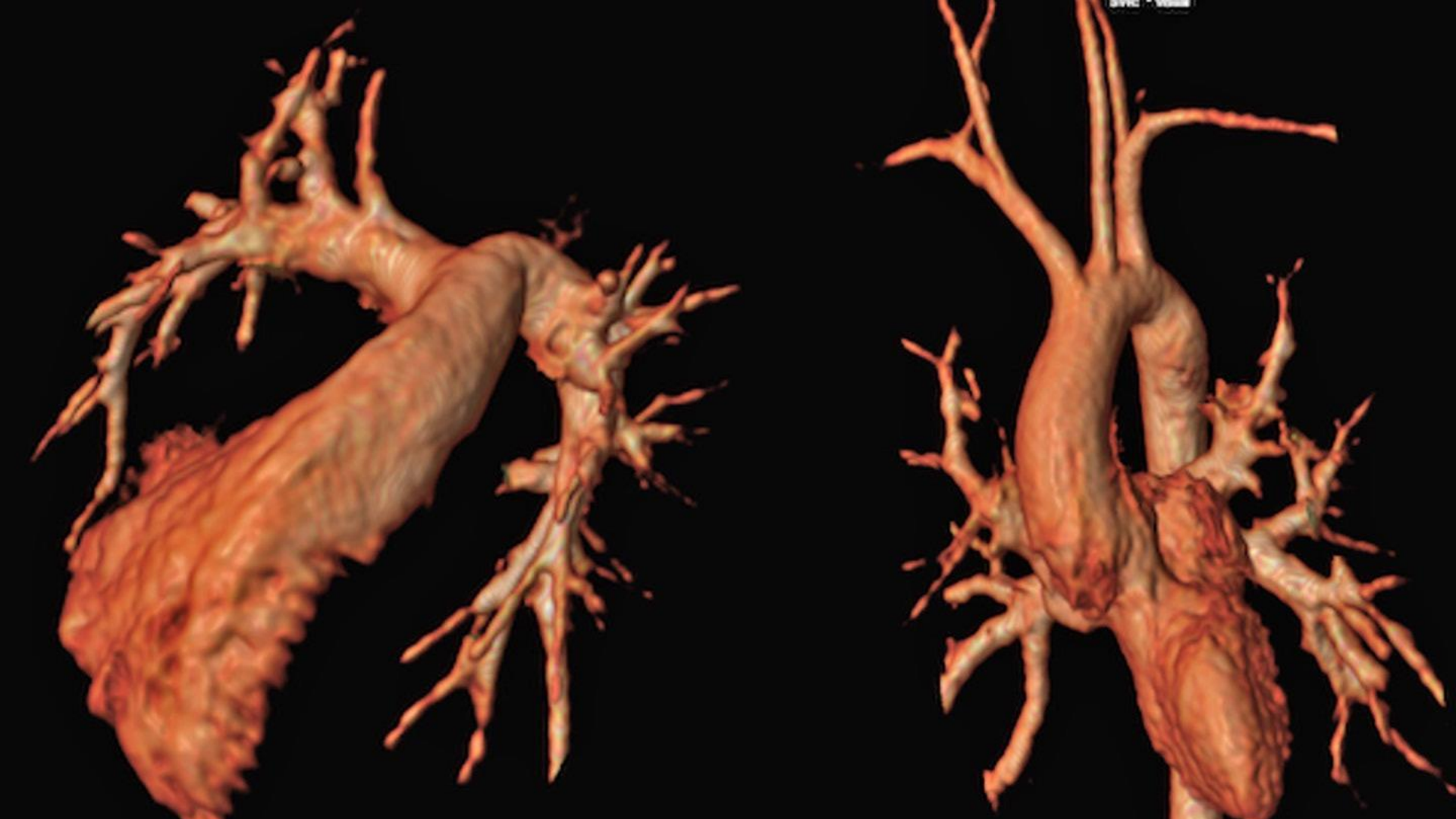
CARDIOPATIE CONGENITE

Contrast Enhanced Magnetic Resonance Angiography (CE-MRA)

- Acquisizioni 3D angiografiche dopo iniezione di mdc paramagnetico
- Acquisizione veloce (anche singolo respiro) con buona risoluzione spaziale
- Importanza del timing corretto (concentrazione del contrasto nella zona anatomica di interesse)
- Time-resolved: più semplici, peggiora la risoluzione spaziale

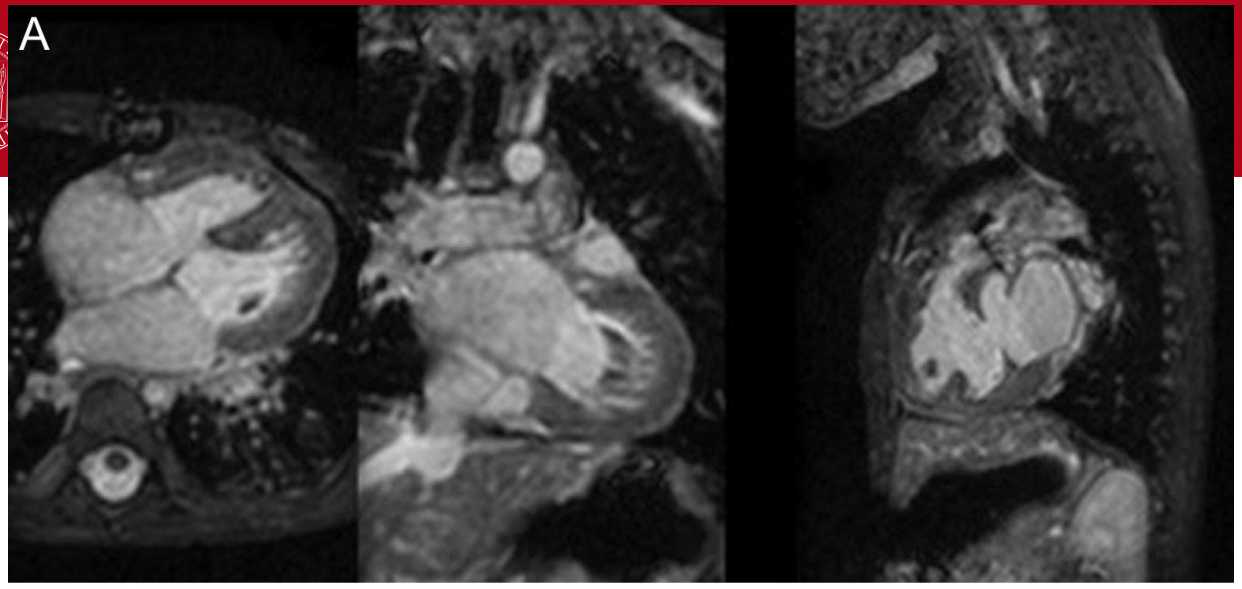


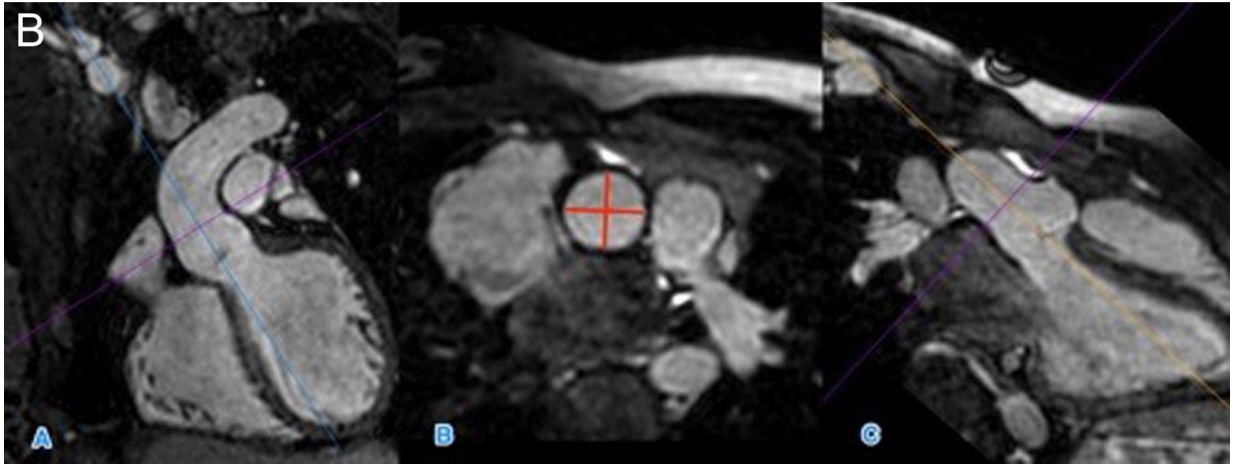
CARDIOPATIE CONGENITE

L’orientamento dei difetti/lesioni non è necessariamente nei piani classici dello spazio!
Utilità MPR (multi planar reconstruction)/3D

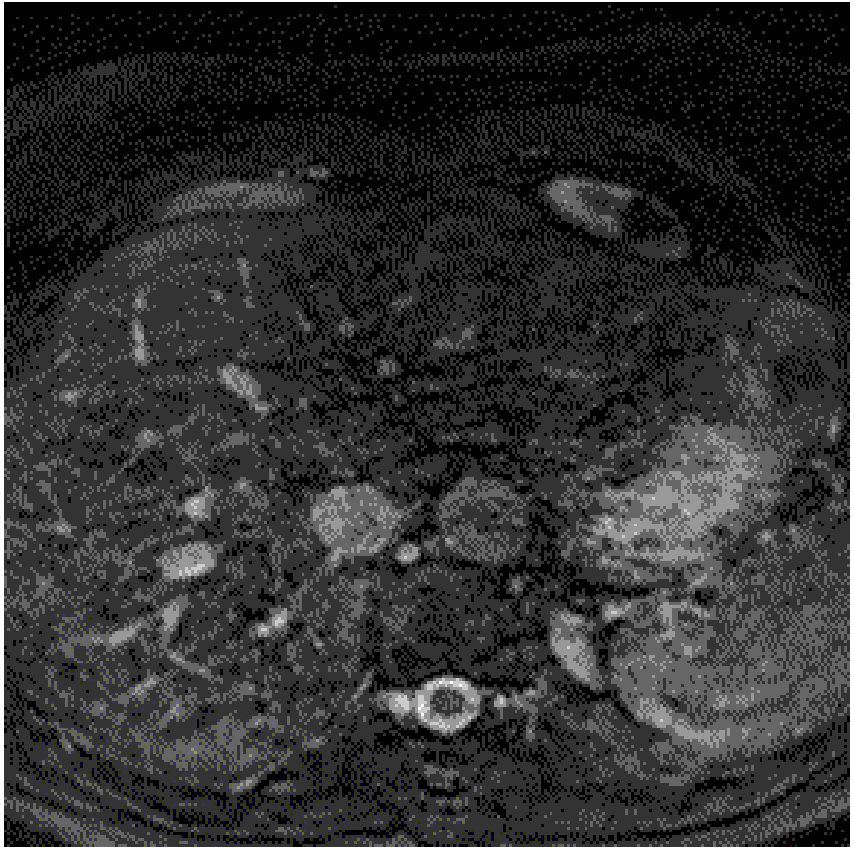
CARDIOPATIE CONGENITE

3D steady-state free processing non contrast imaging (3D SSFP)
Immagini bright blood
PROs: Permettono imaging 3D, sincronizzate con l'ECG, SENZA necessità di mdc (Fontan > diluizione contrasto per via della mancata opacizzazione del flusso cavale); anche a respiro singolo o con gating respiratorio grazie al navigatore diaframmatico > angioMR coronarica; ricostruzione immagini post acquisizione
CONs: acquisizione lunga in cui è necessaria immobilità; molto sensibile ad artefatti da devices; non utilizzabile per valutare il movimento delle strutture; molto sensibili ad aritmia


A


B
A
B
C

CARDIOPATIE CONGENITE



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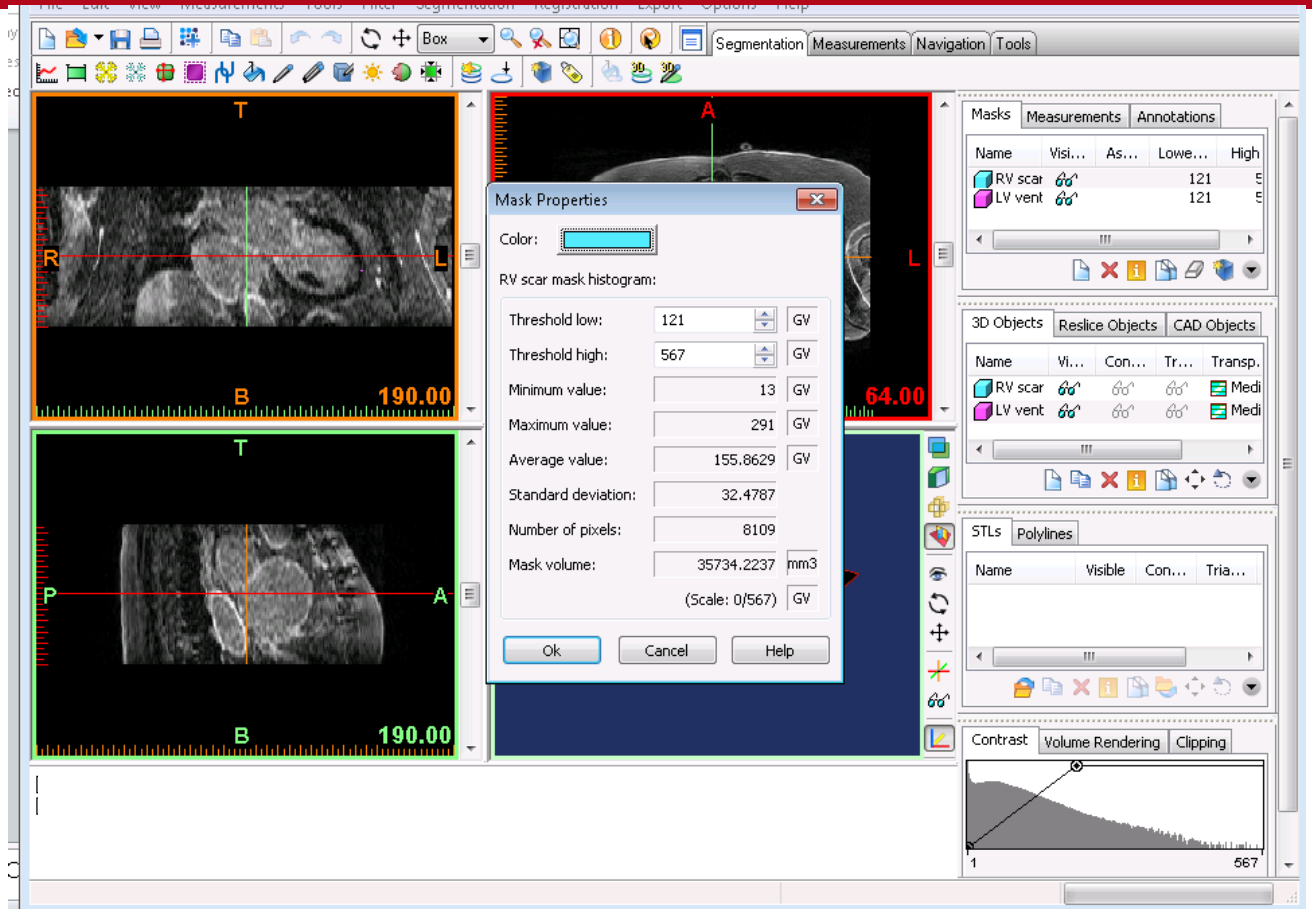

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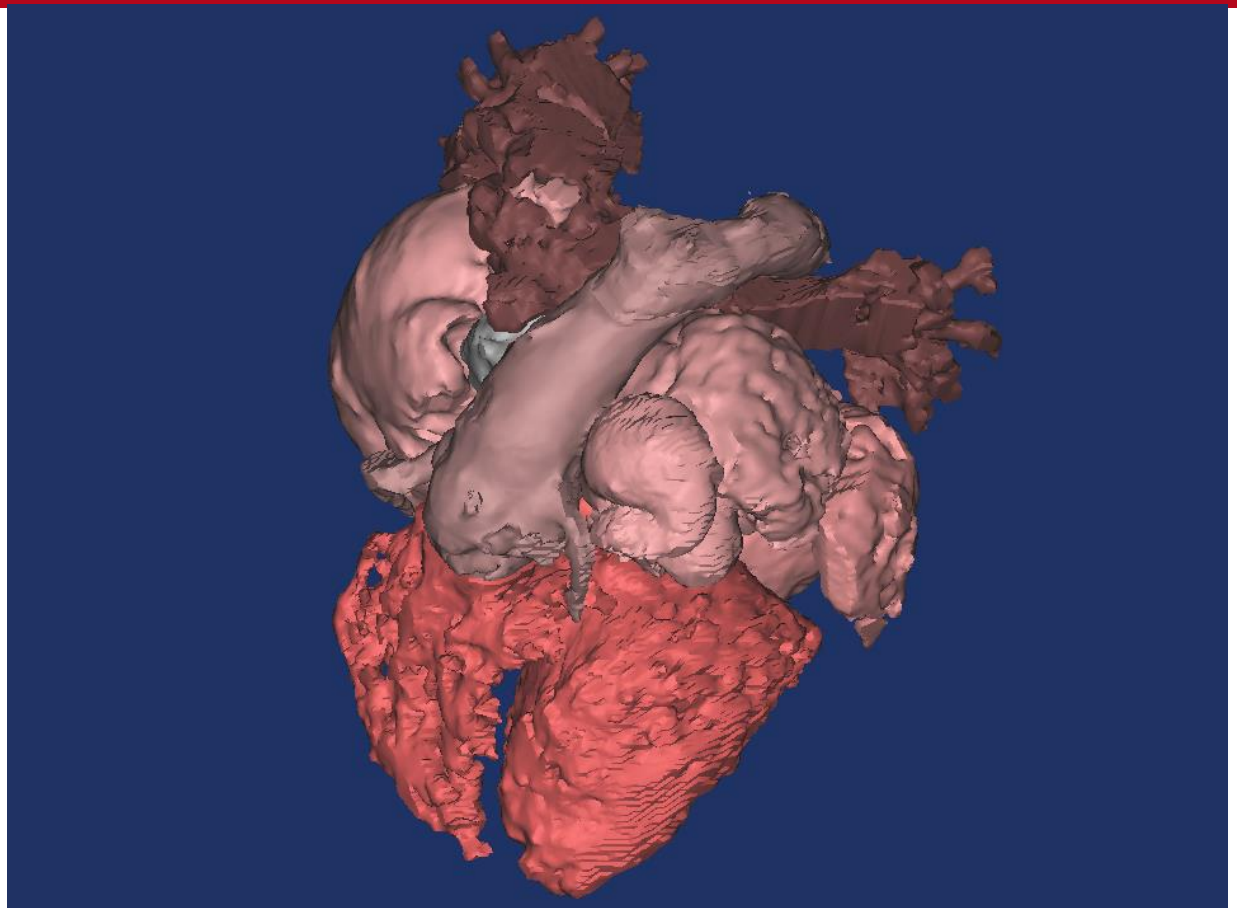

CARDIOPATIE CONGENITE

Table 3 ECG and respiratory navigator-gated 3D steady-state free precession

	Infant/small child	Large child/adult
Isotropic resolution (mm^3)	1.2-1.5	1.3-2.0
Navigator window (mm)	3	5
Image acquisition duration (ms)	40-60	80-150
Trigger delay	End-systole or mid-diastole	Mid-diastole

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CARDIOPATIE CONGENITE

Altre tecniche utilizzate in MRI per la valutazione delle CHD

Perfusion imaging
LGE IR (scar, trombosi)
T1 mapping
MR-augmented cardiac cath

CARDIOPATIE CONGENITE

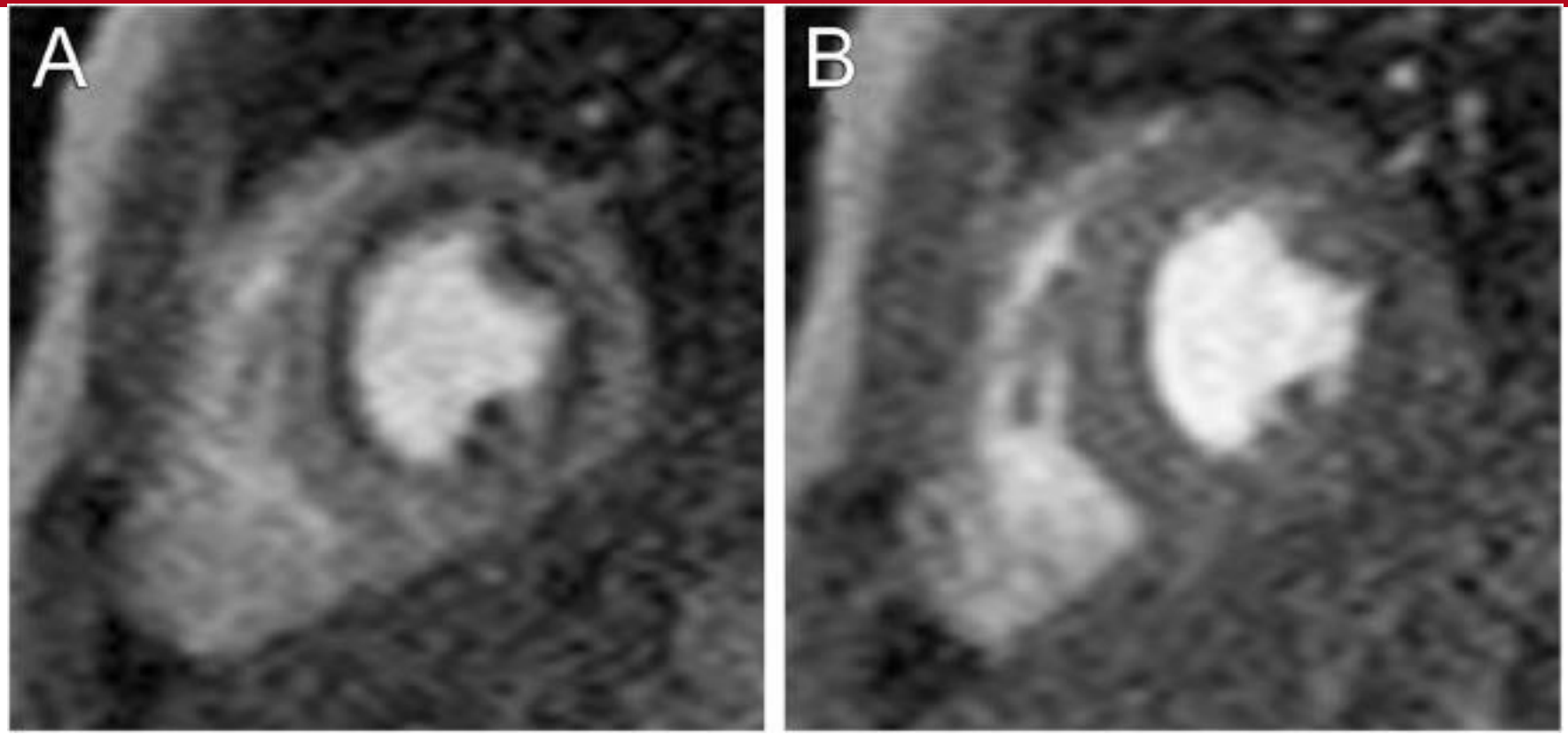

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Table 5 T1-weighted saturation-recovery gradient echo for first pass myocardial perfusion

	Infant/small child	Large child/adult
In-plane resolution (mm)	1.5-2.0	2.0-2.5
Slice thickness (mm)	5-8	8-10
Image acquisition timing	1 R-R or 2 R-R	1 R-R

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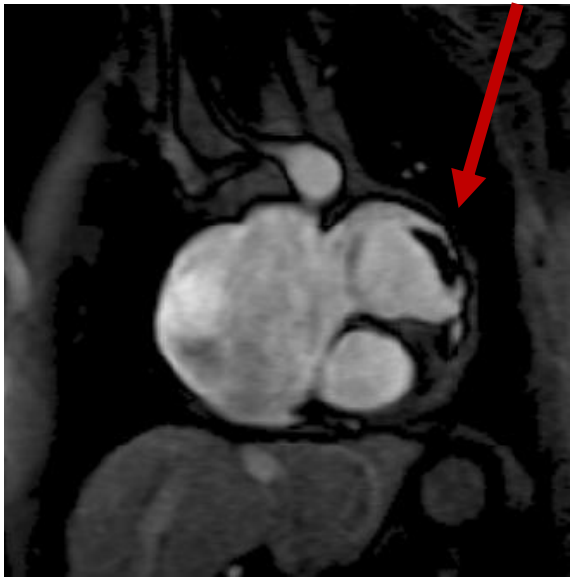

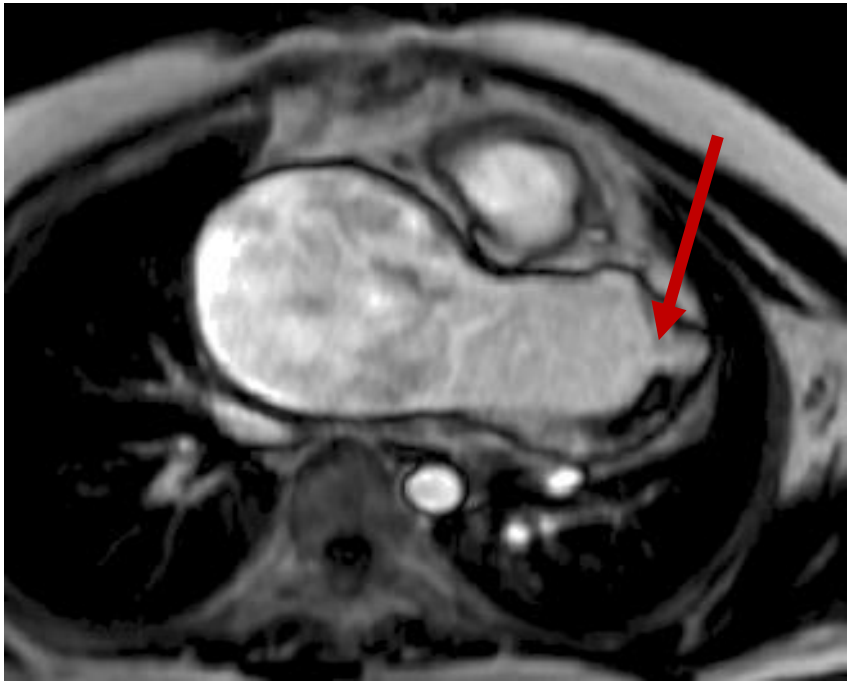

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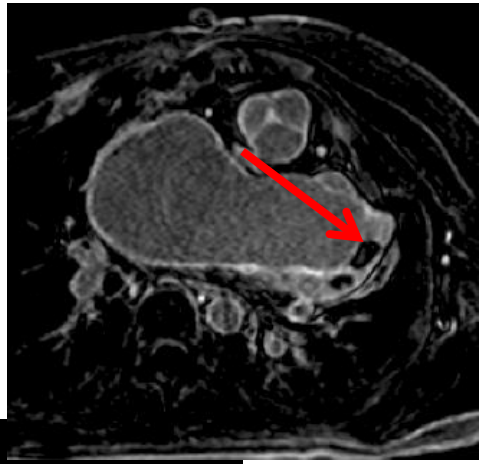

CARDIOPATIE CONGENITE

Table 6 T1-weighted inversion-recovery gradient echo for late gadolinium enhancement

	Infant/small child	Large child/adult
In-plane resolution (mm)	1.0-1.5	1.2-2.0
Slice thickness (mm)	5	6-8
Views per segment	8-16	16-28
Number of signal averages	1-2	1
Image acquisition timing	3 R-R or 4 R-R	2 R-R
Trigger delay	Diastole or systole	Diastole

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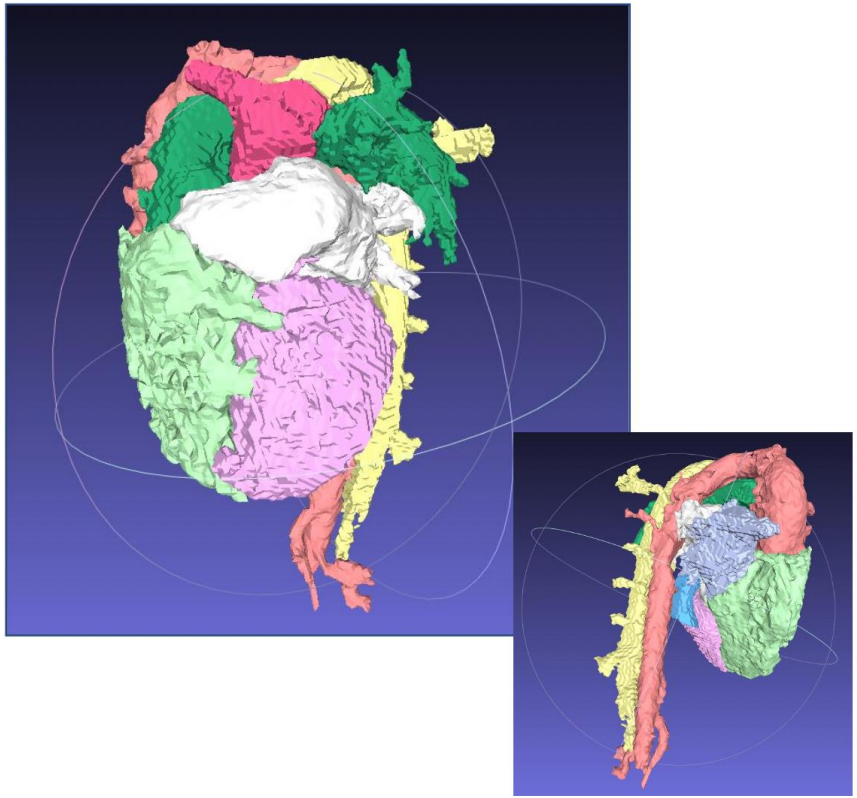
MODULO «LGE»

- LGE imaging in left ventricular 2-chamber and 3- chamber views, right ventricular 3-chamber view, 4-chamber view
- LGE imaging with a stack of contiguous slices to completely encompass both ventricles planned in a ventricular short-axis or axial orientation
- LGE imaging in an orthogonal plane to confirm LGE if present

Key reporting elements: location, extent and thickness of LGE

CARDIOPATIE CONGENITE



IVC
LV
RV
AO
PA
Baffle
Hemi
RA
LA

CARDIOPATIE CONGENITE

ALTRI ASPETTI TECNICI

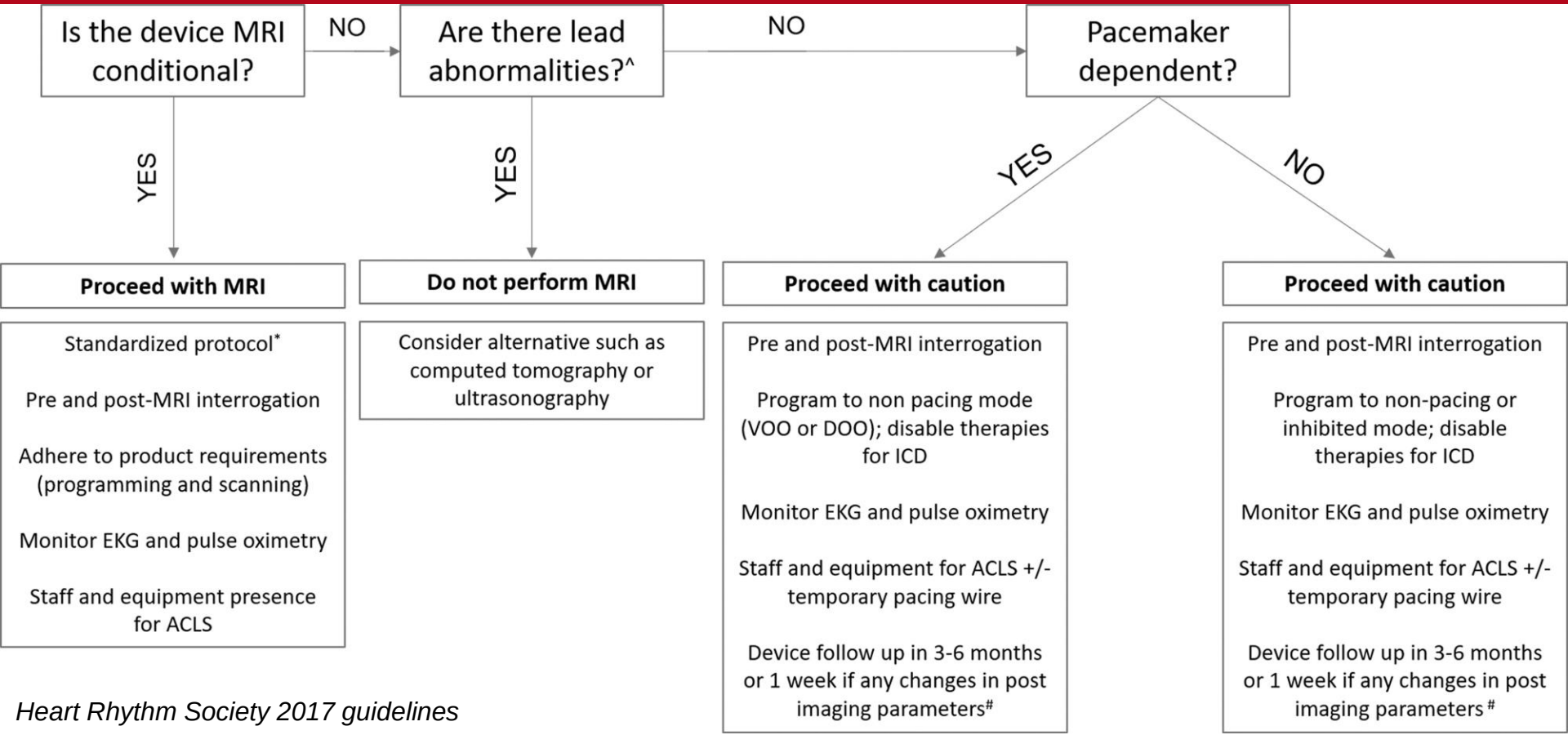
- **Dispersione calore più veloce nel bambino > coprire il paziente**
- **Compliance del paziente - Sedazione necessaria nei più piccoli**
- **Devices intracardiaci**
- **Aritmie**

Acceleration technique	Comment
• Reduce **number of slices** acquired per breath-hold	• Increases overall scan time
• Reduce **number of phases** for each breath-hold: - by reducing **acquisition matrix** (scan or phase percentage) - by reducing **FOV**	• Reduces SNR • Increases spatial resolution
• Increase **voxel size**	• Decreases spatial resolution
• Use **parallel imaging**	• Prone to artefacts
• Use **respiratory navigator**	• Increases overall scan time
• Acquire images in **inspiration**	• Varying slice position with each breath-hold
• Use **real time imaging**	• Reduces image quality
• Consider **general anaesthesia**	
• Ensure **correct understanding** of breath-hold technique	• If patient has no respiratory problems

Scarsa compliance respiratoria

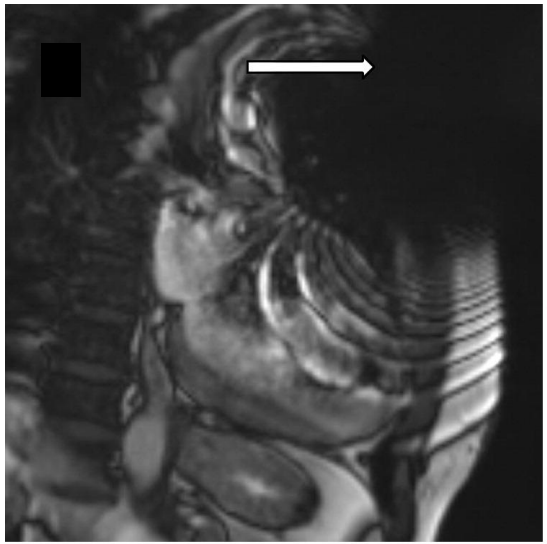
Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE

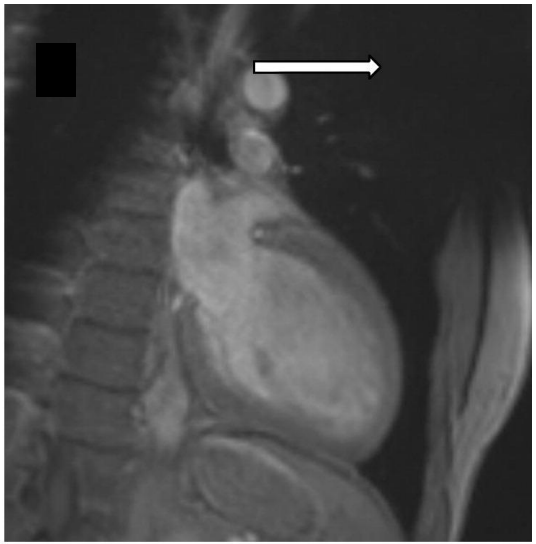


Heart Rhythm Society 2017 guidelines

CARDIOPATIE CONGENITE



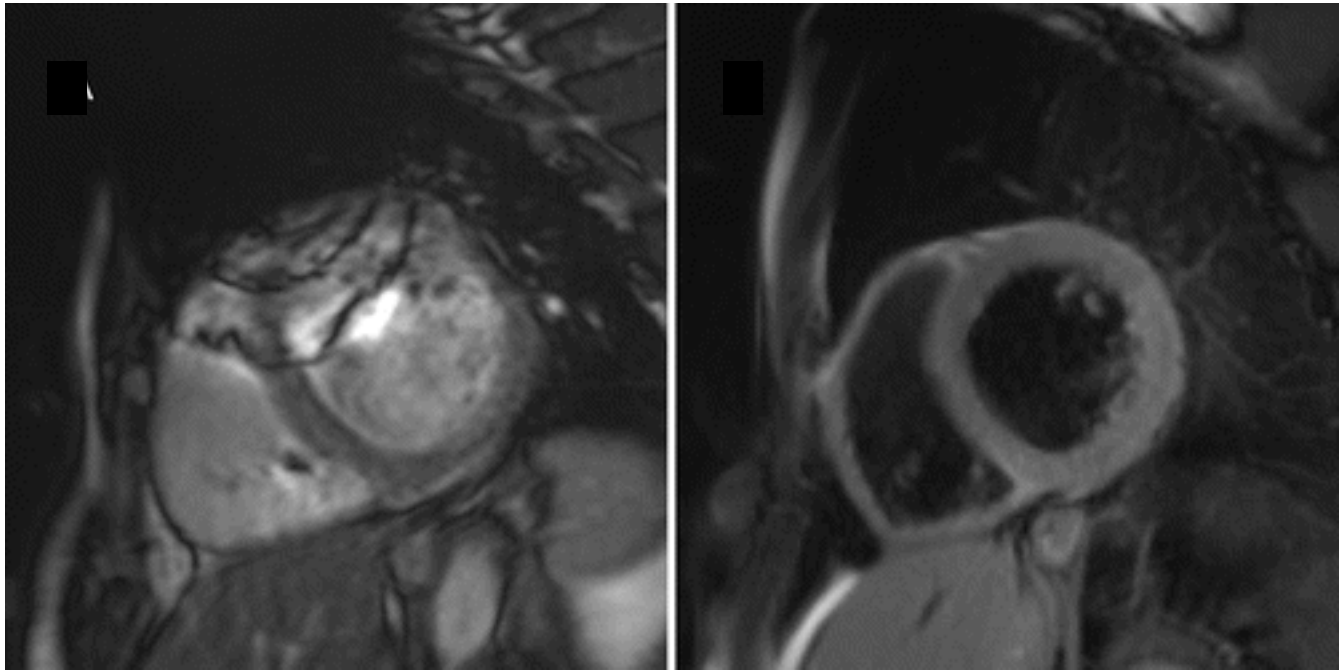
SSFP



Gradient echo

Devices

CARDIOPATIE CONGENITE



SSFP

Spin echo

Devices

Technique	Comment
• **Ensure correct lead position**	
• **Try again**	• You might be lucky!
• **Heart rate** and/or **rhythm control** before scanning	• Use beta-blockers or other antiarrhythmic medication
• Use **arrhythmia rejection**	• Increases breath-hold time
• Use **prospective triggering**	• Cardiac diastole is not entirely visualised
• Use **real-time imaging**	• Reduces temporal and spatial resolution as well as SNR
• Scan **in inspiration**	• If heart signal capture is suboptimal • Reduces vasovagal arrhythmias
• **Increase NSA**	• Signal averaging can be useful for e.g. delayed contrast imaging
• **Alternative sequence**	• E.g. turbo field echo rather than SSFP, white blood imaging rather than black blood sequences

ARITMIE

Adattato da: ESC/EACVI CHD/CMR guide, 2014

1222 · 2022
800 ANNI

UNIVERSITÀ DEGLI STUDI DI PADOVA

I Master di Risonanza Magnetica Cardiaca

Cardiopatie congenite – Parte 2

Dott.ssa Veronica Spadotto

CARDIOPATIE CONGENITE

Cardiac Situs	**Situs solitus** **Situs inversus** **Situs ambiguous** Based on atrial morphology Bronchial situs is a surrogate
Cardiac Position	**Levocardia** **Dextrocardia** **Mesocardia** Based on position in thorax
Cardiac Segments	**Atrial Segment** **Ventricular Segment** **Arterial Segment**
Connections	**Veno-Atrial Connection** **Atrio-Ventricular Connection** **Ventricular-Arterial Connection**

Sequential segmental analysis

Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE

Cardiac Situs

Situs Solitus

- Anterior RA (on right)
- Posterior LA (on left)

Situs Inversus

- Posterior RA (on left)
- Anterior LA (on right)

Situs Ambiguous

Right **Left**

- RA or LA isomerism

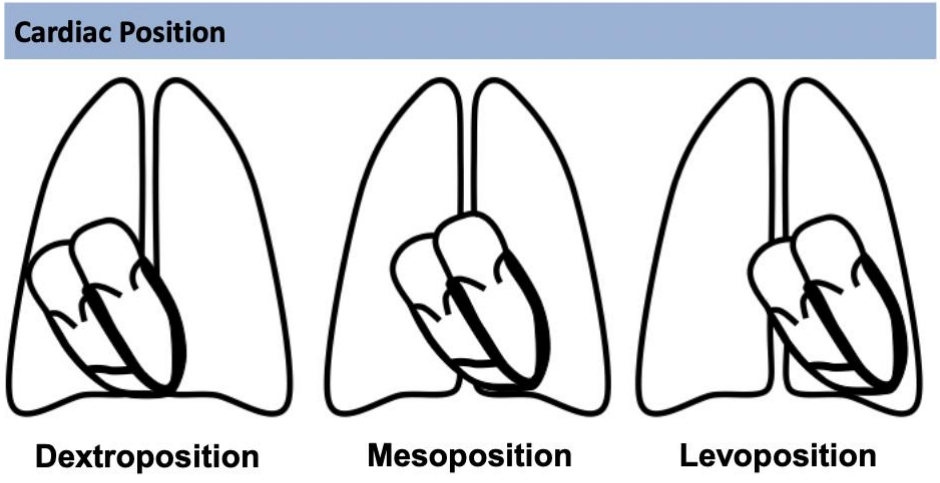
Abdominal Situs

Situs Solitus **Situs Inversus** **Heterotaxia**

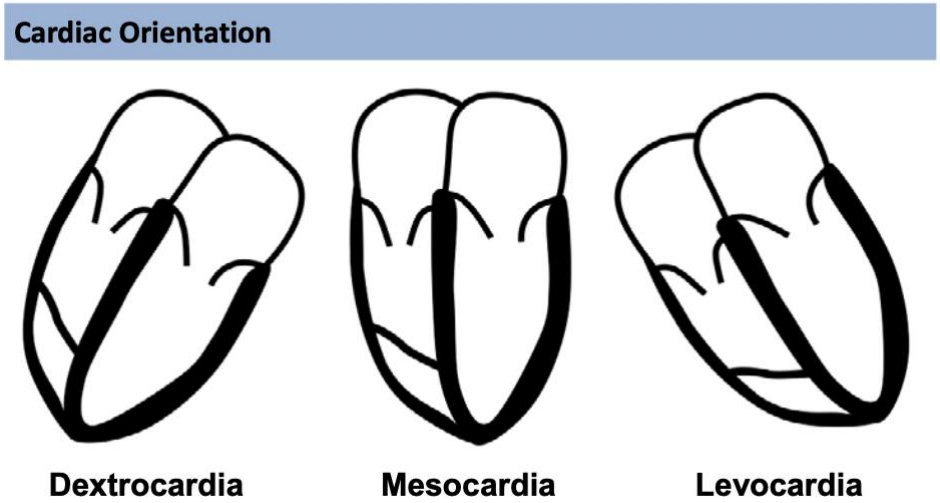
Sequential segmental Analysis: situs

Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE



Sequential segmental Analysis: posizione e orientamento del cuore



Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE

Sequential segmental Analysis: segmenti cardiaci

Atrial Segment

Right atrium

- Broad, based, triangular appendage
- Short and vertical bronchus arrangement

Left atrium

- Narrow, tubular appendage
- Long and horizontal bronchus arrangement

Ventricular Segment

Right ventricle

- Trabeculated
- TV associated, TV attachments to the septal moderator band
- Muscular infundibulum between inlet and outlet

Left ventricle

- Smooth walled
- MV associated, MV attachments to papillary muscles

Arterial Segment

Pulmonary Trunk

- Bifurcation to RPA and LPA

Aorta

- Left- or right-sided
- Coronary arteries
- Regular branches

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CARDIOPATIE CONGENITE

Sequential segmental Analysis: connessioni

Veno-Atrial Connection

- IVC and SVC connections
- Presence of left SVC (90% left SVCs drain to RA via coronary sinus)
- Normal, partial or total anomalous pulmonary venous drainage

Atrio-Ventricular Connection and Inlets

AV concordance

- RA is connected to RV, LA to LV

AV discordance

- RA is connected to LV, LA to RV

The valves go with the ventricles

Double inlet, mitral atresia, tricuspid atresia, AVSD

Ventricular-Arterial Connection

VA concordance

- LV is connected to Ao
- RV to MPA

VA discordance

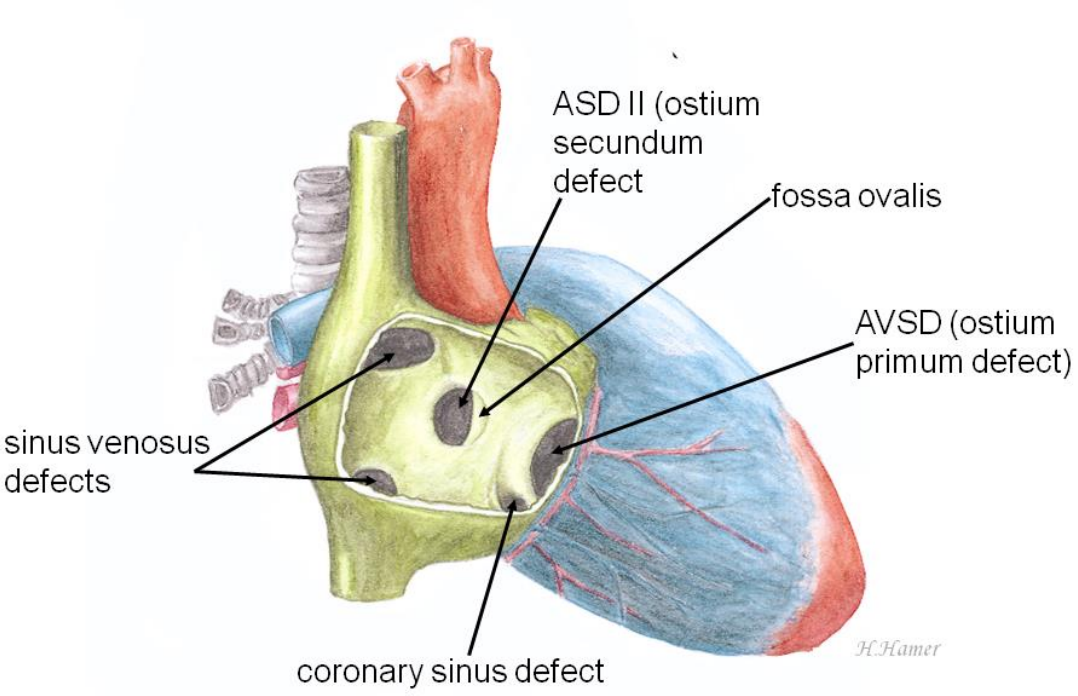
- LV is connected to MPA
- RV to Ao

Double outlet, single outlet

(e.g. pulmonary atresia or truncus arteriosus)

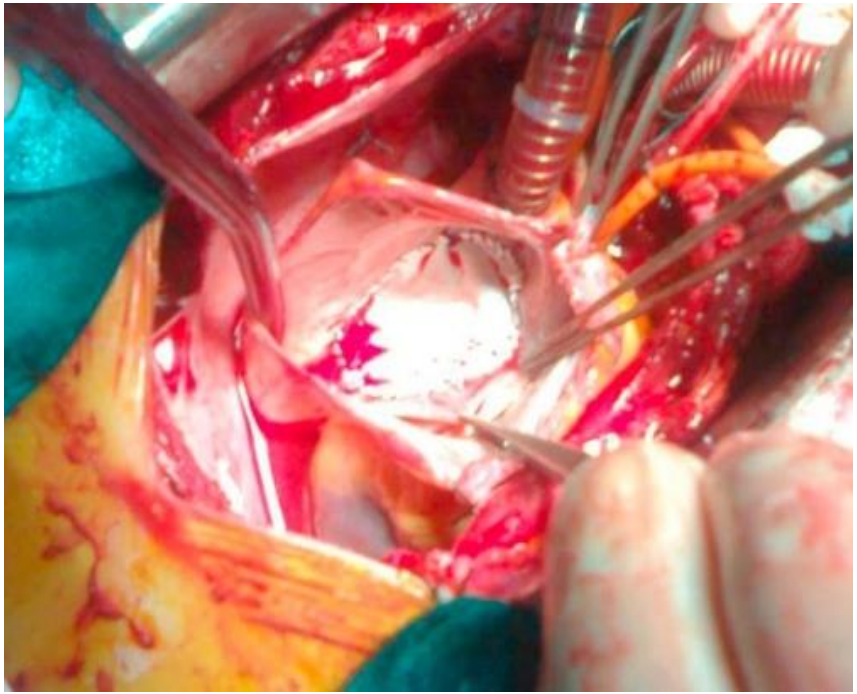
Adattato da: ESC/EACVI CHD/CMR guide, 2014

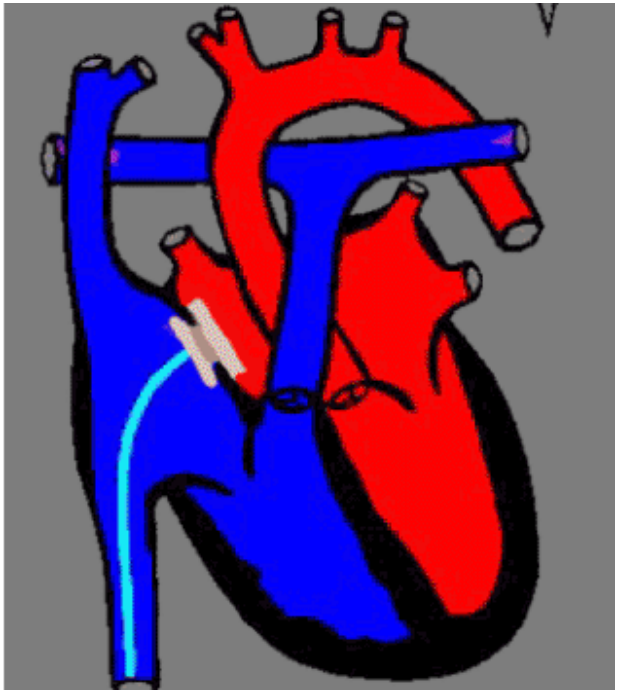
CARDIOPATIE CONGENITE: difetti interatriali



Types of atrial septal defects with their locations

CARDIOPATIE CONGENITE: difetti interatriali





Correzione chirurgica (tutti i tipi) o percutanea (ostium secundum)
Sequele a lungo termine: shunt residuo

CARDIOPATIE CONGENITE: difetti interatriali

PROTOCOLLO DIA TIPO OSTIUM SECUNDUM

Standard imaging

1.Cine CMR: stack di slices sottili in piano 4CH che includa interamente il setto interatriale

2.Cine CMR: stack di slices sottili in piano sagittale obliquo perpendicolare al SIA che lo includa interamente

3.Modulo ventricolografia

4. PC CMR: AAo, PA

Eventualmente anche:

1. PC CMR: 1–3 slices parallele al piano del SIA, in modo da ottenere una «en face view» of the defect con through-plane velocity encoding

2. PC CMR: stack di slices sottili in 4 CH e/o in piano obliquo sagittale perpendicolare al setto interatriale che lo includa interamente, con in-plane velocity encoding nella direzione del flusso del DIA

3. CE-MRA o 3D SSFP per l'albero vascolare toracico

Key reporting elements: numero e posizione dei difetti, misurazione dei margini dei difetti, parametri ventricolari, Qp/Qs

CARDIOPATIE CONGENITE: difetti interatriali

Fratz et al. Journal of Cardiovascular Magnetic Resonance 2013, 15:51

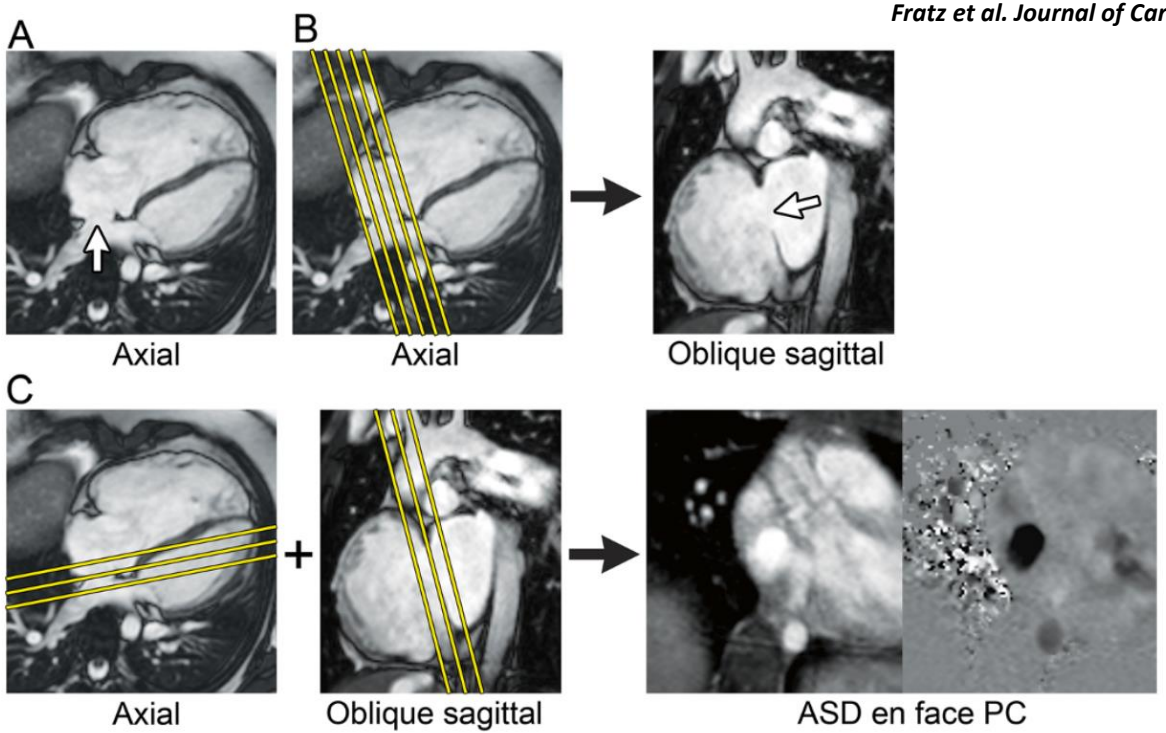


Figure 17 Secundum atrial septal defect (ASD) imaging protocol. A. Axial steady-state free precession (SSFP) image in a 10-year-old with a large secundum ASD (white arrow). **B.** The axial SSFP image is used to plan a stack of oblique sagittal SSFP images to visualize the ASD and the superior and inferior defect margins. **C.** The axial and oblique sagittal images are used together to plan a stack of phase-contrast (PC) cine images to visualize the ASD flow *en face*. This provides insight into the oval shape of the defect and may demonstrate additional ASDs.

CARDIOPATIE CONGENITE: difetti interatriali

CARDIOPATIE CONGENITE: difetti interatriali

PROTOCOLLO DIA TIPO SENO VENOSO

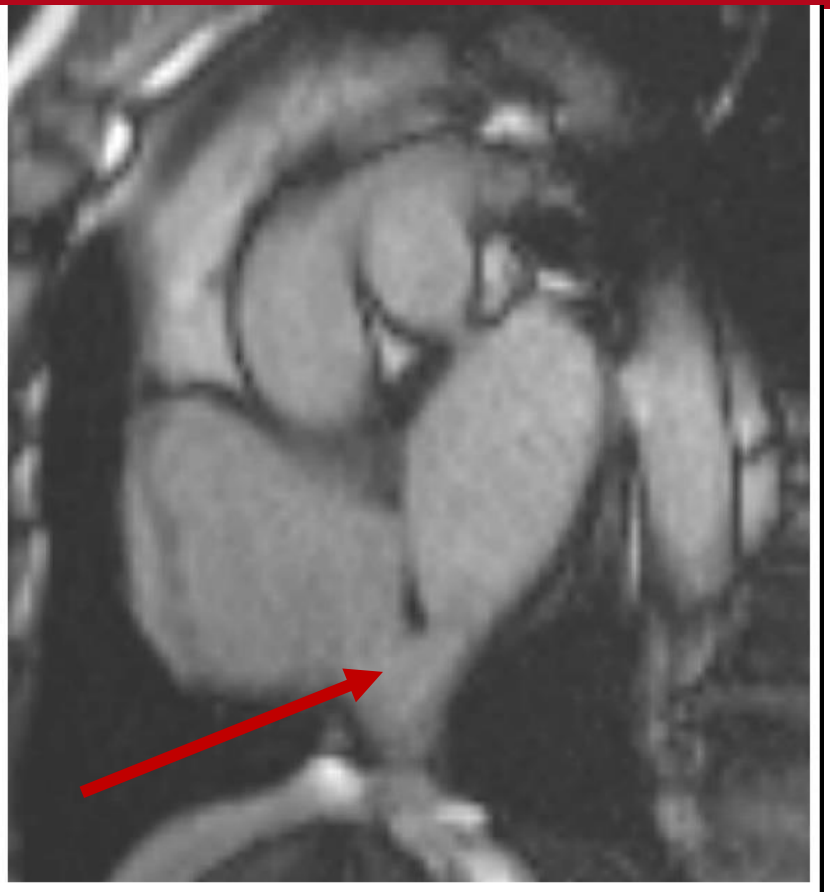
Standard imaging

1. Cine CMR: stack assiale da metà fegato fino alla sommità dell'arco aortico
2. Cine CMR: piano sagittale obliquo perpendicolare al piano del difetto
3. Modulo ventricolografia
4. CE-MRA or3D SSFP per l'albero vascolare toracico
5. PC CMR: AAo, AP

Key reporting elements: posizione e dimensioni del difetto, ritorni venosi polmonari, parametri ventricolari, Qp/Qs

CARDIOPATIE CONGENITE: difetti interatriali

CARDIOPATIE CONGENITE: difetti interatriali



CARDIOPATIE CONGENITE: difetti interatriali

Fratz et al. Journal of Cardiovascular Magnetic Resonance 2013, 15:51

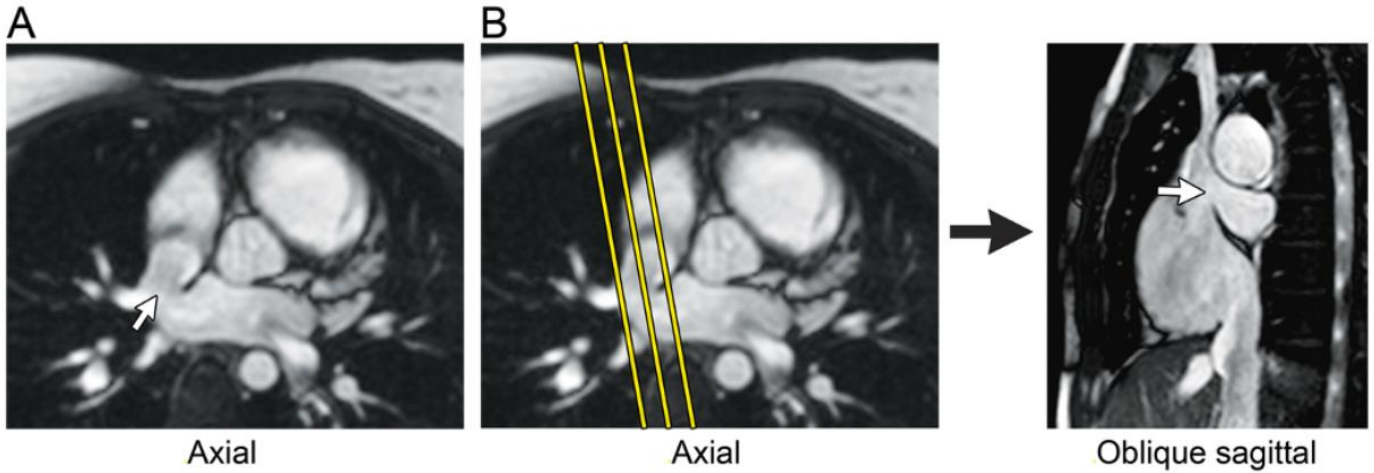
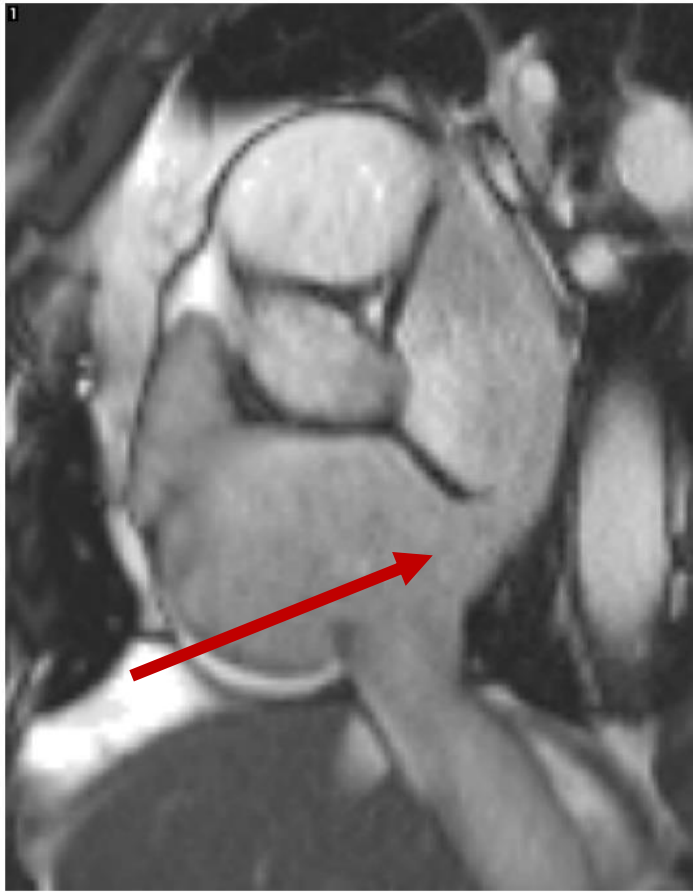


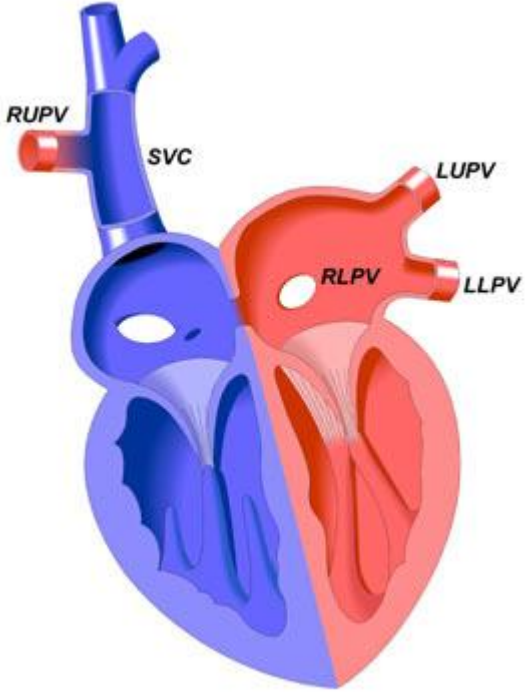
Figure 18 Sinus venosus septal defect imaging protocol. A. Axial steady-state free precession (SSFP) cine image in a 22-year-old with a large sinus venosus septal defect (white arrow). **B.** The axial SSFP image is used to plan a stack of oblique sagittal SSFP cine images to visualize the defect in an orthogonal view and assess its superoinferior dimension.

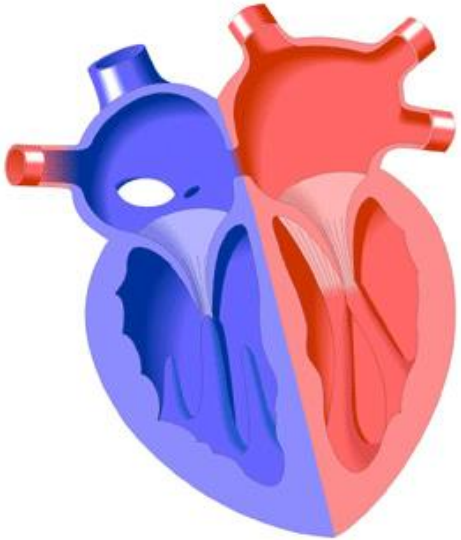
CARDIOPATIE CONGENITE: difetti interatriali



CARDIOPATIE CONGENITE: ritorni venosi polmonari anomali

Una o più vene polmonari arrivano alle sezioni destre (direttamente, tramite il flusso cavale superiore o inferiore, o con un collettore)- PARZIALI o TOTALI





CARDIOPATIE CONGENITE: ritorni venosi polmonari anomali

- **Il PAPVD delle vene di destra si associa quasi sempre a DIA tipo seno venoso.**
- **Il PAPVD delle vene di sinistra non si associa a DIA seno venoso, e spesso arriva in atrio destro tramite un collettore venoso.**

CARDIOPATIE CONGENITE: ritorni venosi polmonari anomali

Standard imaging

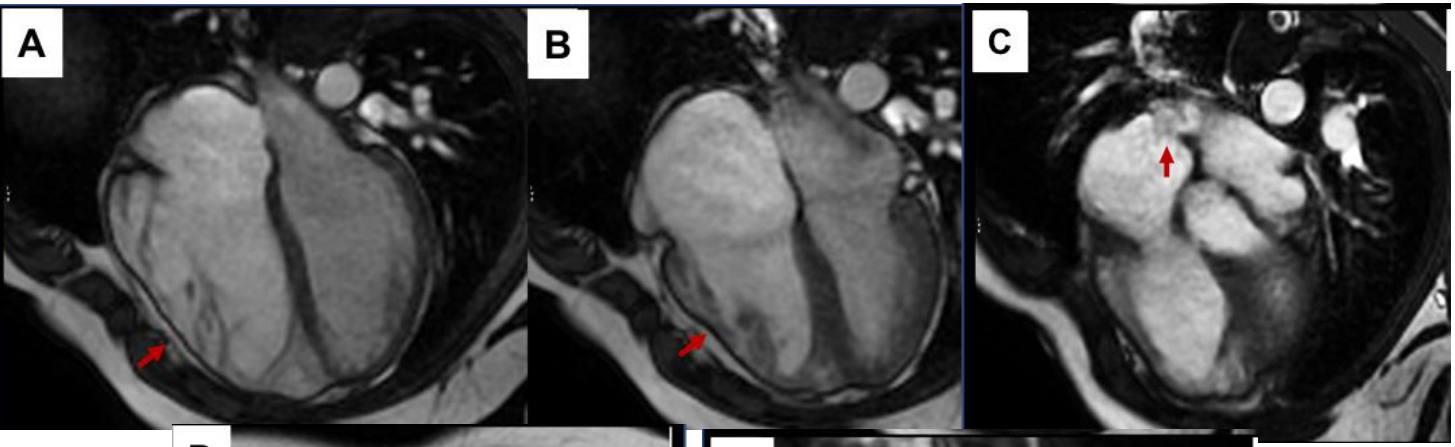
1. Cine CMR: stack assiale da metà addome alla sommità dell'arco aortico
2. Cine CMR: piano obliquo per valutare la/e vena/e anomala/e in asse lungo
3. Modulo ventricolografia
4. CE-MRA o 3D SSFP per la l'albero vascolare toracico
5. PC CMR: AAo, AP, APDx, APsn

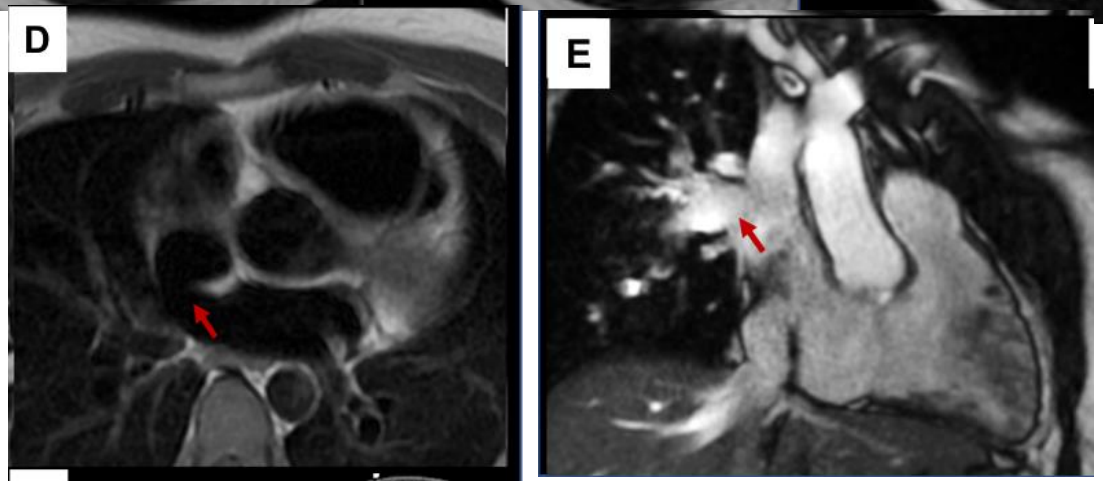
Eventualmente anche:

PC CMR: vena anomala

Key reporting elements: numero, sede, drenaggio delle vene polmonari; parametri ventricolari; Qp/Qs, distribuzione del flusso nelle arterie polmonari

CARDIOPATIE CONGENITE:
Ritorni venosi polmonari anomali





DIA tipo Seno venoso superiore + RVPAP vene pp dx

CARDIOPATIE CONGENITE: shunt pretricuspidalici

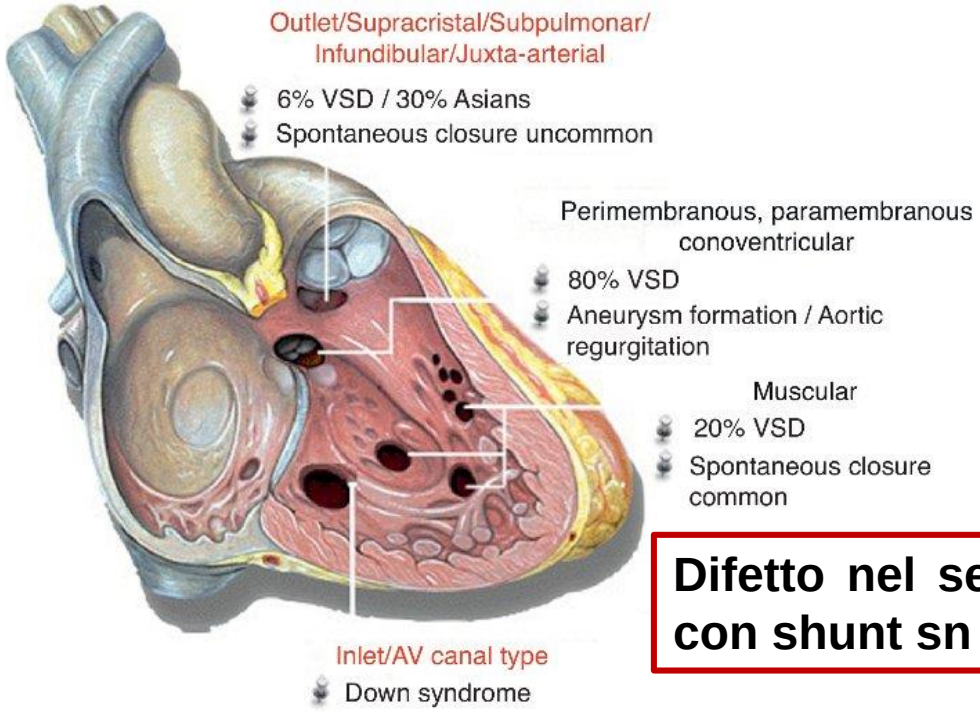
Shunt pretricuspidalico sn>dx:

1. **Dilatazione ventricolare destra**
2. **Qp:Qs > 1**
3. **Flusso in arteria polmonare (PASV)=stroke volume RV (RVSV)**

Shunt pretricuspidalico dx>sn:

1. **Dilatazione ventricolare destra**
2. **Qp:Qs < 1**
3. **Flusso in arteria polmonare (PASV)=stroke volume RV (RVSV)**

CARDIOPATIE CONGENITE: difetti interventricolari



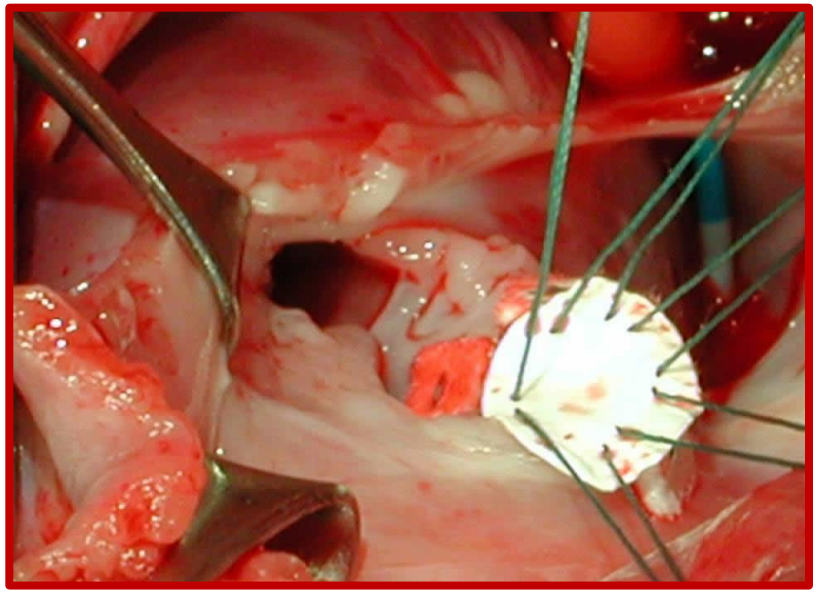
Difetto nel setto interventricolare con shunt sn >dx

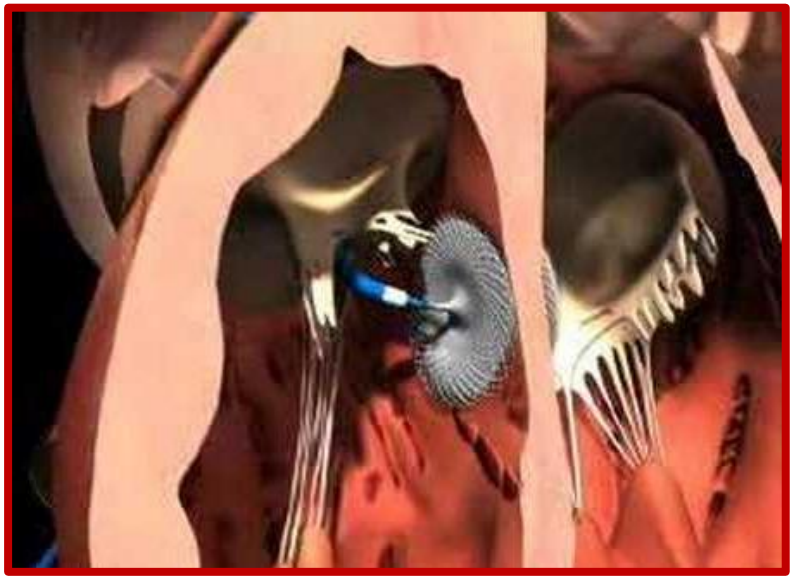
CARDIOPATIE CONGENITE: difetti interventricolari

Possibili reperti associati ai DIV:

- **Aneurismi del setto IV**
- **Ventricolo destro «a doppia camera»**
- **Prolasso delle cuspidi aortiche +/- insufficienza valvolare aortica**
- **Stenosi sottovalvolare aortica**

CARDIOPATIE CONGENITE: difetti interventricolari





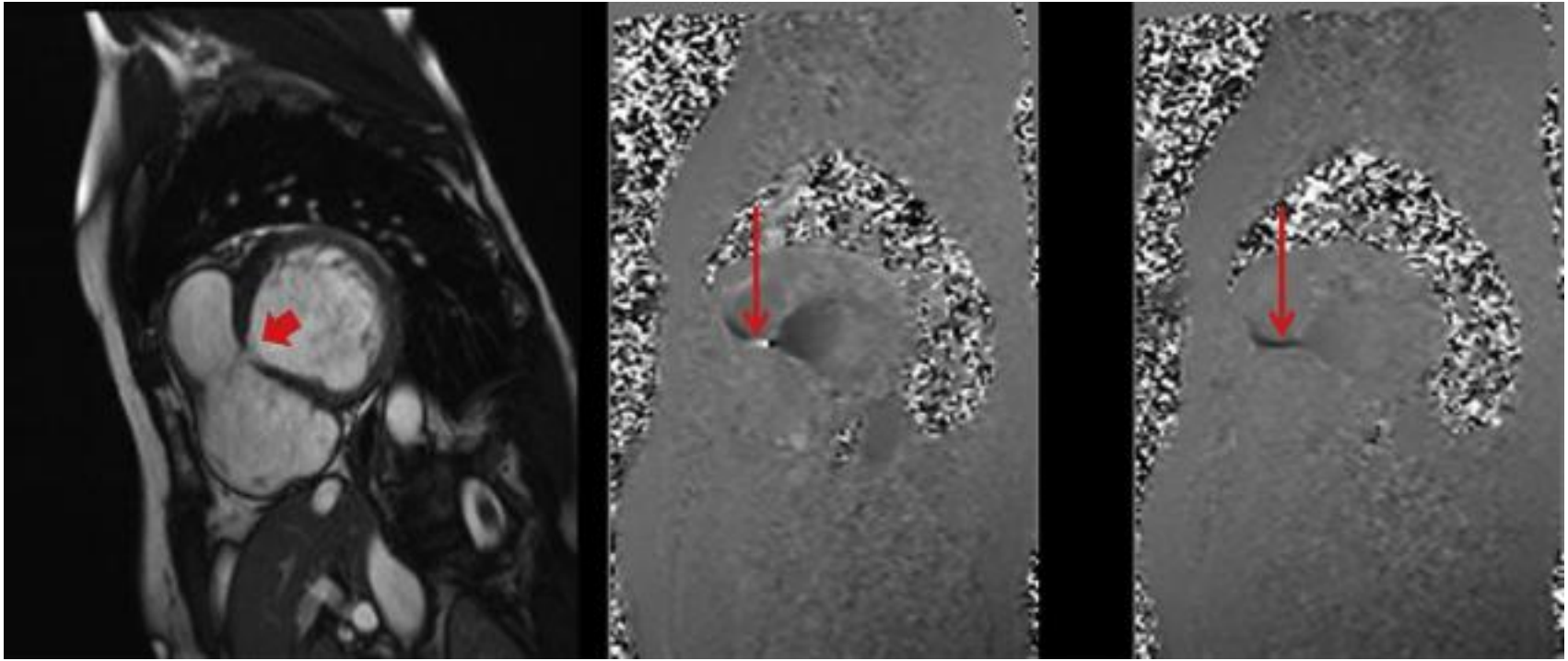
Correzione chirurgica o percutanea
Complicanze: DIV residuo, ostruzione RVOT

CARDIOPATIE CONGENITE: difetti interventricolari

DIV restrittivo:

- **Piccolo (<1/2 diametro valvolare aortico)**
- **Alta velocità sn > dx**
- **Vdx normale**
- **Pressioni polmonari normali**

CARDIOPATIE CONGENITE: difetti interventricolari



CARDIOPATIE CONGENITE: difetti interventricolari

DIV non restrittivo:

- **Grande**
- **Ipertrofia ventricolare destra e segni di sovraccarico pressorio ventricolare destro**
- **Possibile Eisenmenger S**

CARDIOPATIE CONGENITE: difetti interventricolari

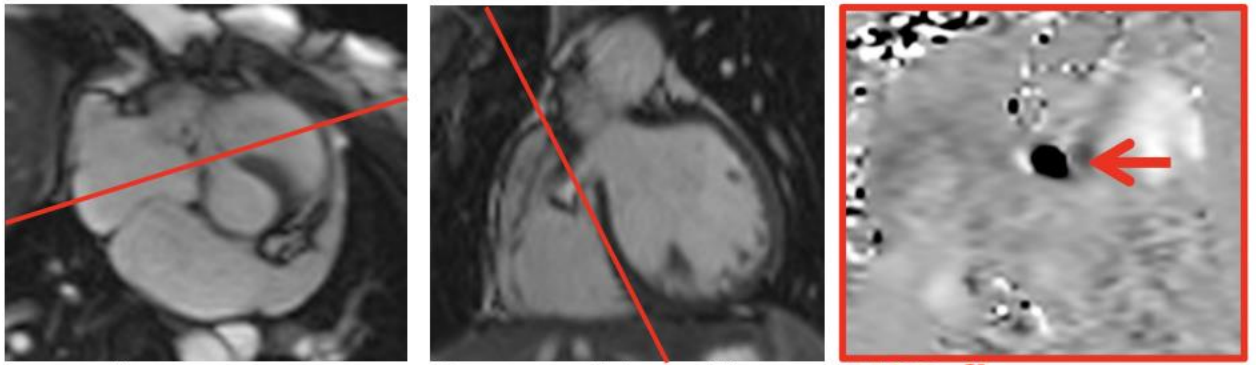
Shunt sn >dx:

- **Dilatazione Vsn**
- **QP:QS > 1**
- **LVSV = PASV e RVSV=AoSV**

Shunt dx >sn:

- **Dilatazione Vsn**
- **QP:QS < 1**
- **LVSV = PASV e RVSV=AoSV**

CARDIOPATIE CONGENITE: difetti interventricolari



Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE: difetti interventricolari

PROTOCOLLO DIV

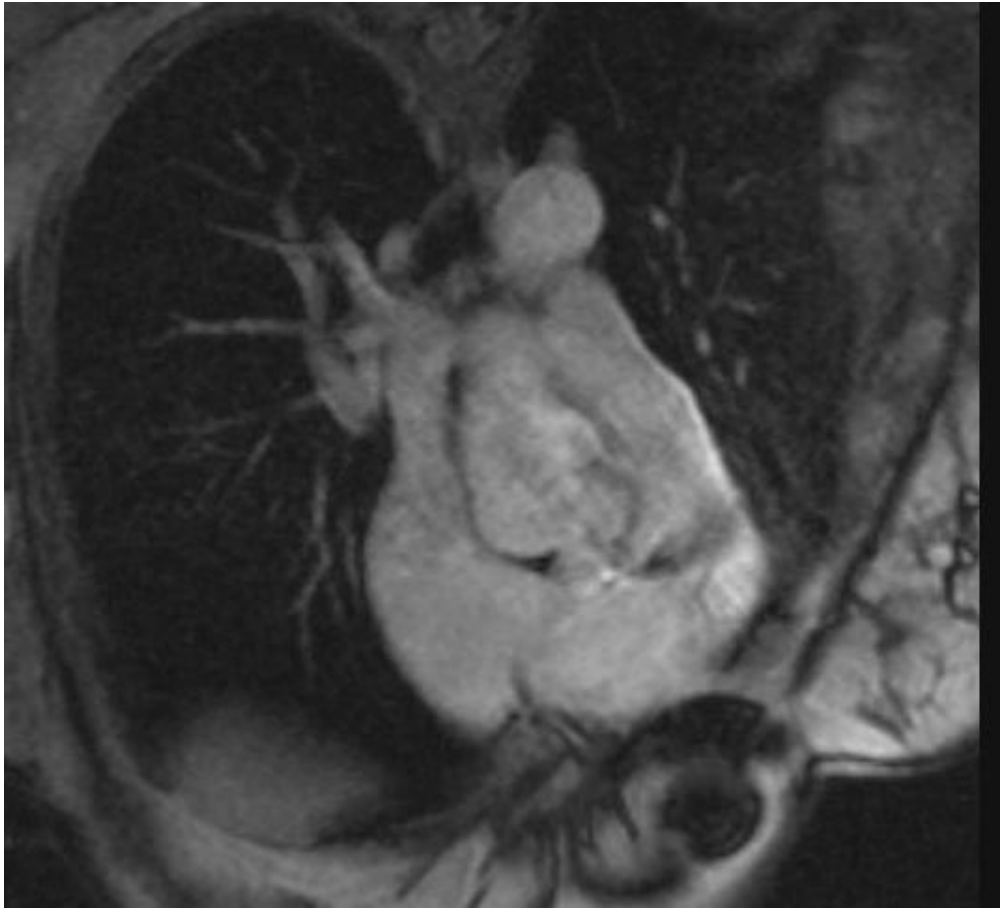
Standard imaging

1.Stacks anatomici
2.Cine VLA, HLA, LV stack, RV stack
3. Cine LVOT, RVOT
4.Cine valvola aortica
5.CP CMR: AAo, MPA

Eventualmente anche:

1. PC CMR: 1–3 slices parellele al piano del setto interventricolare, per ottenere una «en face view» del difetto e un through-plane velocity encoding
2. PC CMR: 1-3 slices in 4 camere e/o perpendicolari al SIV in un piano sagittale obliquo che includa completamente il SIV, con in-plane velocity encoding nella direzione del flusso del DIV

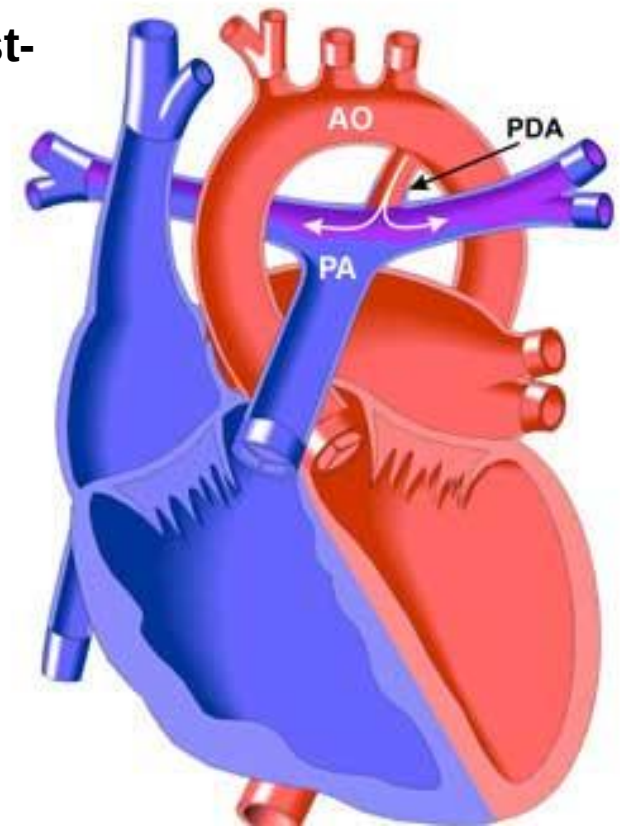
Key reporting elements: numero, tipo e posizione dei difetti, parametri ventricolari, Qp/Qs




H
R
L
A
F

CARDIOPATIE CONGENITE: pervietà del dotto arterioso

Mancata chiusura post-natale del dotto di Botallo



CARDIOPATIE CONGENITE: pervietà del dotto arterioso

- **Dilatazione Vsn**
- **QP:QS < 1**
- **RVSV = PASV e LVSV=AoSV**

- **Dilatazione Asn**
- **Dilatazione vene polmonari e aorta ascendente se dotto ampio**
- **Talora associato a Coartazione aortica**

CARDIOPATIE CONGENITE: pervietà del dotto arterioso

Chiusura:
- **chirurgica**
- **Percutanea (coil, occluder)**

Complicanze operatorie a lungo termine: shunt residuo

CARDIOPATIE CONGENITE: Pervietà del dotto arterioso

PROTOCOLLO PDA

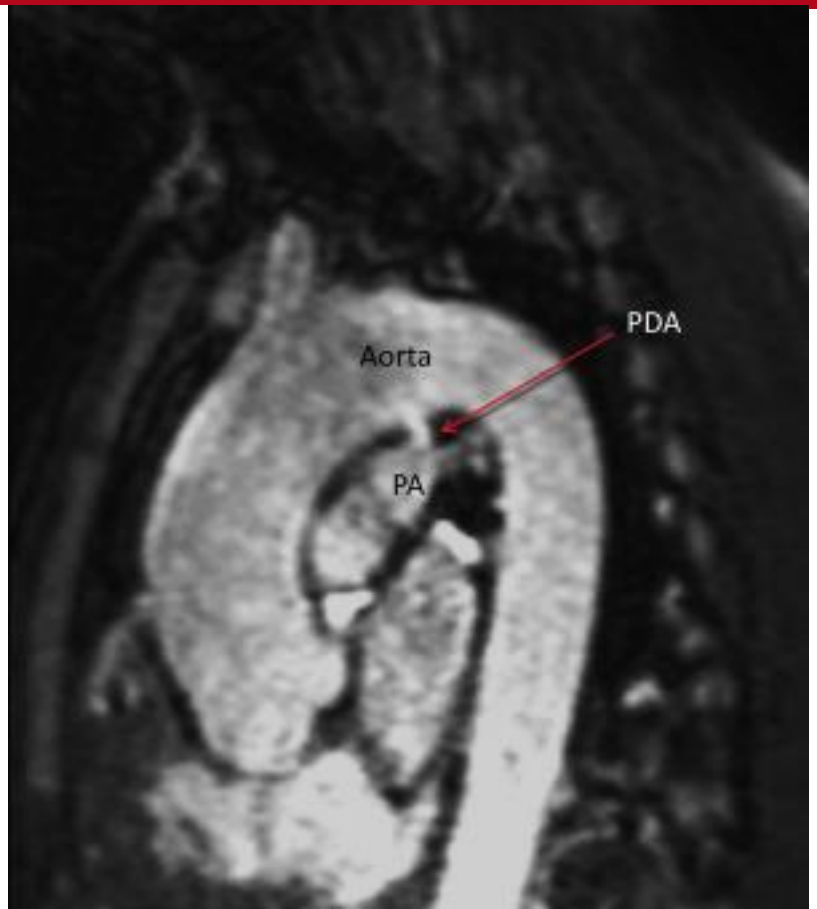
Standard imaging

1.Stacks anatomici
2.Cine VLA, HLA, LV stack, RV stack
3. Cine LVOT, RVOT
4.Cine arteria polmonare e arco aortico
5.Cine PDA
6.PC CMR: Aorta pre e post Pda, flusso v.aortica, flusso MPA, RPA e LP

Eventualmente anche:

MRA Aorta

Elementi essenziali: lunghezza, diametro e forma del dotto, parametri ventricolari, Qp/Qs

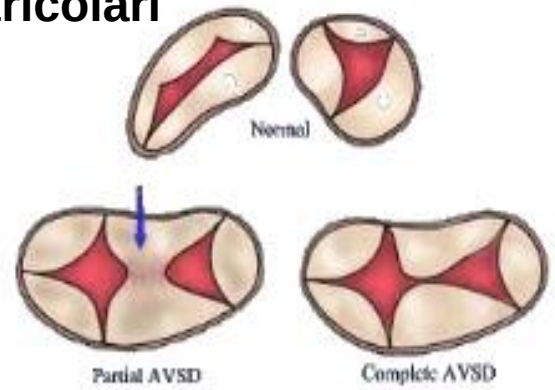


CARDIOPATIE CONGENITE: difetti del setto atrioventricolare

Mancata fusione del setto atriale e ventricolare, con residuo difetto confluente e anulus comune delle valvole atrio ventricolari

Types of AVSD:

- Complete AVSD
- Partial AVSD



Levels of shunting:

- Interatrial and interventricular shunt

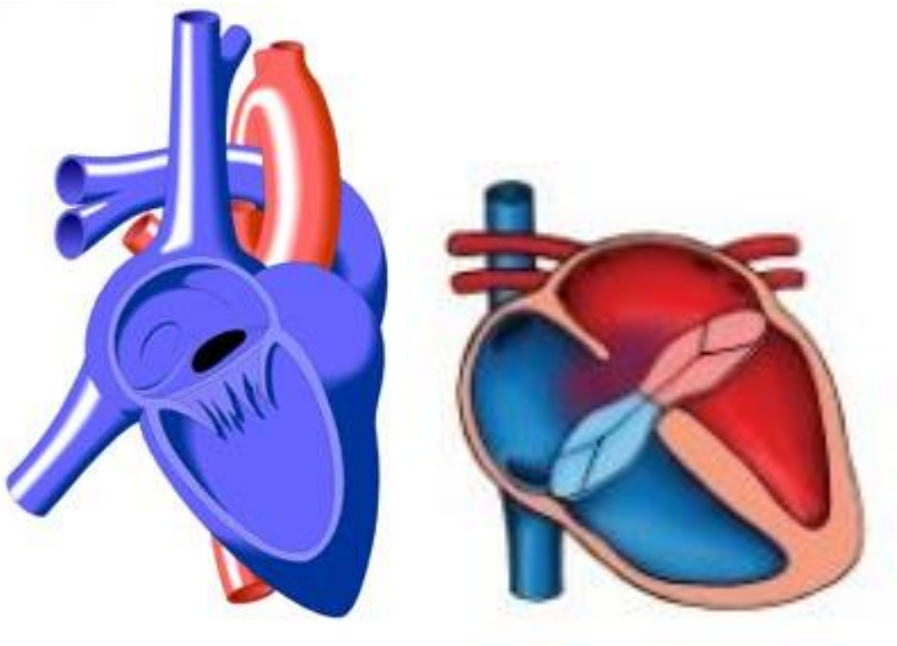
(not attached atrial or ventricular septal crest)

- Interatrial shunt

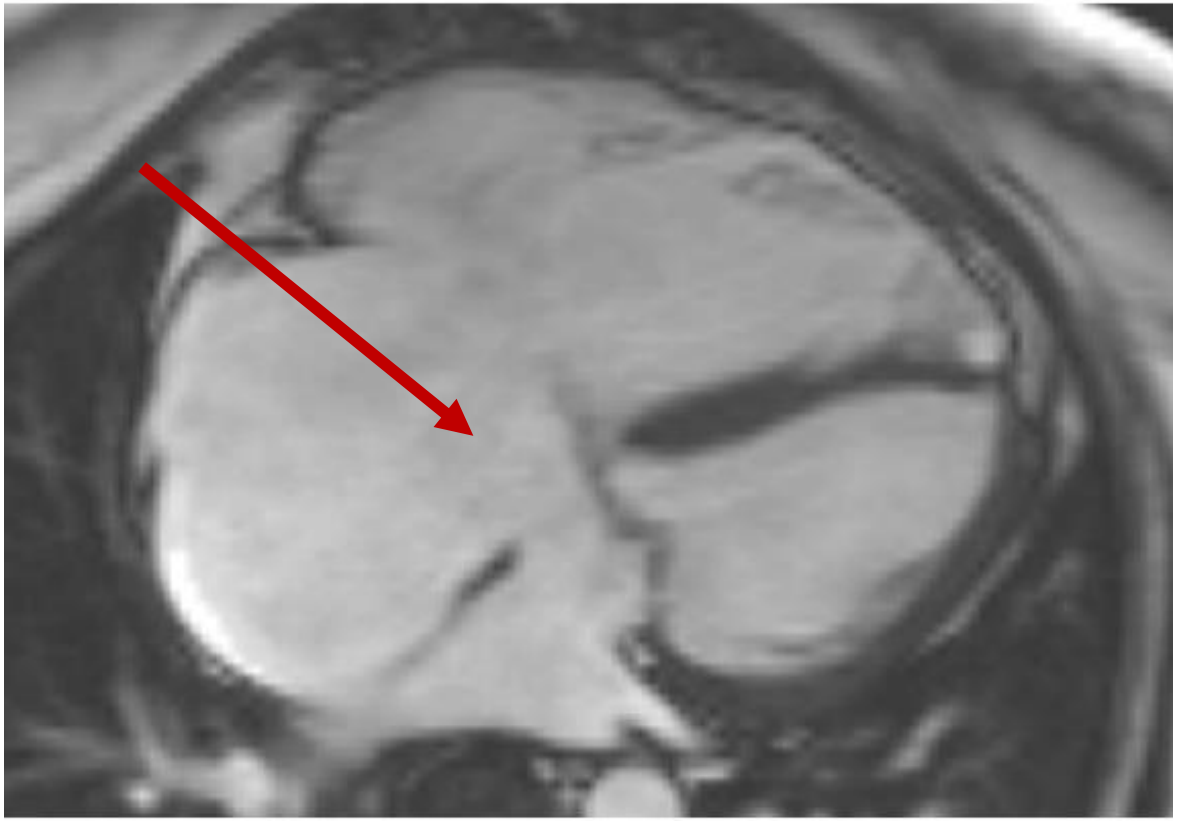
(attached to the ventricular septal crest)

CARDIOPATIE CONGENITE: difetti del setto atrioventricolare (parziali)

- Due orifizi separati per la valvola AV, con il lembo anteriore diviso in componente destra e sinistra e inserito direttamente sul bordo del SIV.
- Detto anche difetto interatriale “ostium primum”
- Spesso associato a Trisomia 21

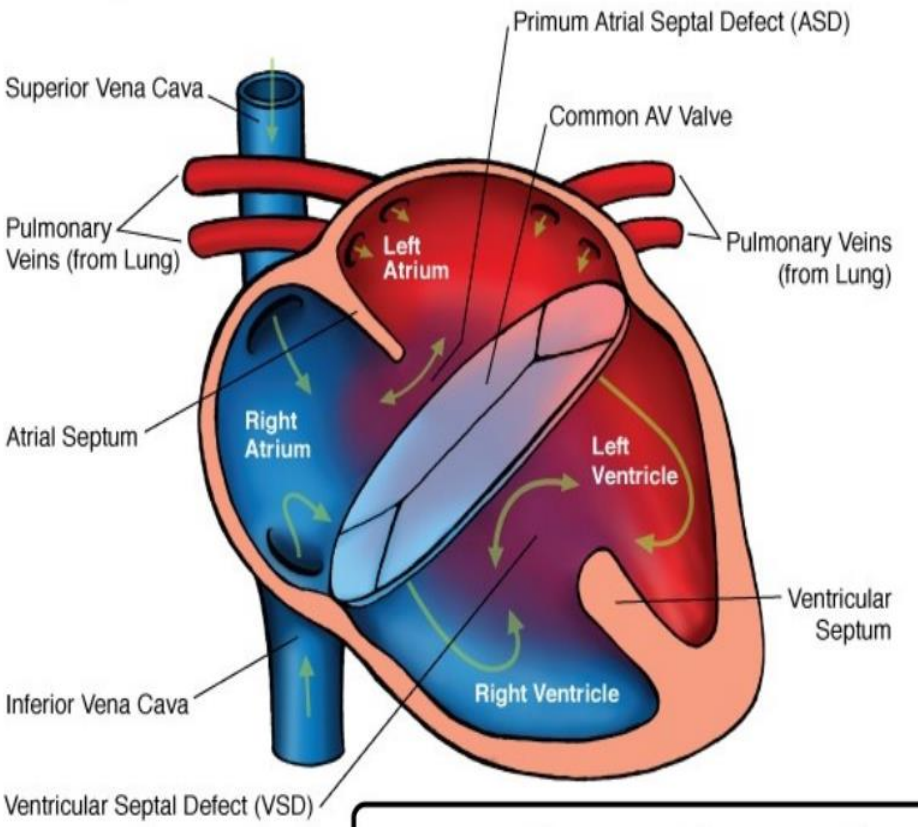


CARDIOPATIE CONGENITE: difetti del setto atrioventricolare (parziali)



CARDIOPATIE CONGENITE: difetti del setto AV completi -«canale AV»

- Il lembo anteriore della valvola AV è “free floating”, e si crea shunt interatriale e interventricolare
  - Frequente insufficienza valvolare



CARDIOPATIE CONGENITE: difetti del setto atrioventricolare

Intervento correttivo:

1. Banding AP come primo stage se iperafflusso polmonare
2. Chiusura chirurgica con patch dei difetti
3. Plastica della valvola AV
4. Chiusura PDA

Complicanze post operatorie:

1. DIA/DIV residui
2. Insufficienza/stenosi valvole AV
3. Ostruzione LVOT

CARDIOPATIE CONGENITE: difetti del setto atrioventricolare

Protocol	
1. **Anatomy stacks**	5. **Ao flow**
2. **VLA, HLA, LV stack, RV stack**	6. VSD flow
3. **LVOT, RVOT**	7. ASD flow
4. **MPA flow**	8. 3D whole heart

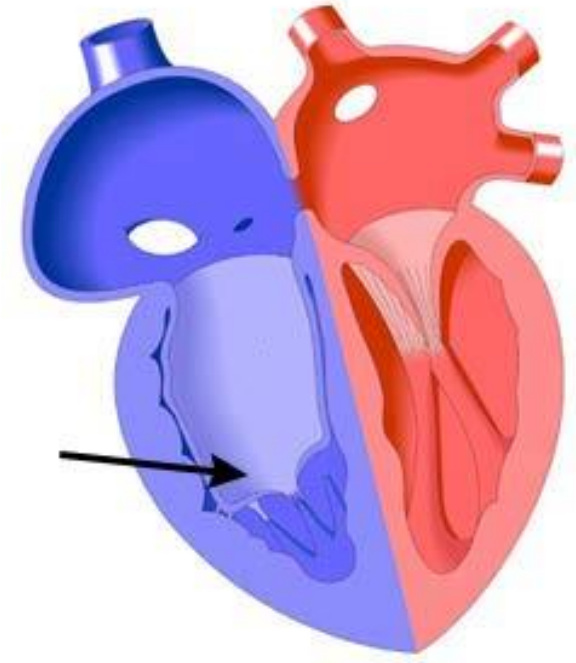
Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV , SV, EF, RWMA, note any LV dilation
   - RV: EDV, ESV, SV, EF, RWMA
2. **Presence and extent of ASD / VSD**
3. **Qp (MPA flow) : Qs (Ao flow)**
4. **Valve regurgitation or stenosis**

Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE: anomalia di Ebstein

- Dislocazione del lembo settale della tricuspide (>8mm/m2)
- Vari gradi di insufficienza tricuspidalica
- Atrializzazione e dilatazione dell'inflow del Vdx
- Si associa a: PFO/DIA (>50%), DIV, ostruzione RVOT
- Possibile riduzione riempimento Vsn dalla dilatazione Vdx



CARDIOPATIE CONGENITE: anomalia di Ebstein

Intervento correttivo:

1. Riparazione o sostituzione valvolare tricuspidalica
2. Chiusura PFO/DIA
3. Atrioplastica
4. Eventuale Glenn/Fontan nei casi più severi

Complicanze post operatorie:

1. IT residua
2. Insufficienza ventricolare destra
3. Compressione CDx dalla sutura

CARDIOPATIE CONGENITE: anomalia di Ebstein

Standard imaging

1. Cine CMR: stack assiale dal diaframma fino alla sommità dell'arco aortico
2. Modulo ventricolografia con slice continue multiple in 3CH Vdx e 4 CH
3. PC CMR: AAo, AP, v. tricuspide, mitrale

Eventualmente anche:

1. Cine CMR: stack continuo parallelo al piano della tricuspide funzionale per vedere la valvola «en face»
2. CE-MRA or 3D SSFP per l'albero vascolare toracico

Key reporting elements: descrizione della morfologia della tricuspide, del grado di insufficienza e stenosi, eventuale stenosi polmonare associata, parametri ventricolari, presenza di DIA, Qp/Qs

CARDIOPATIE CONGENITE: anomalia di Ebstein

Fratz et al. Journal of Cardiovascular Magnetic Resonance 2013, 15:51

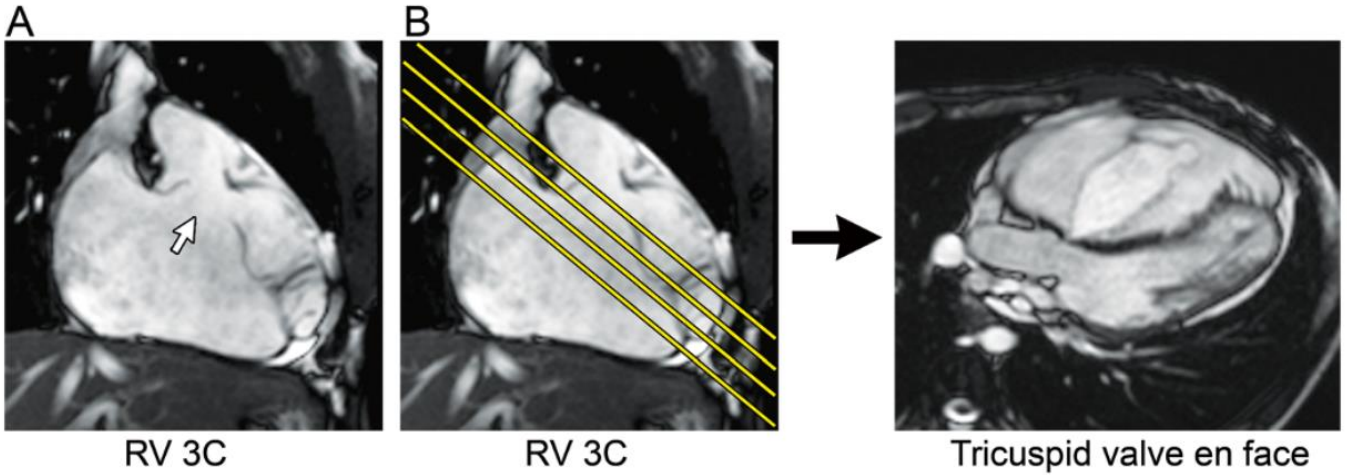
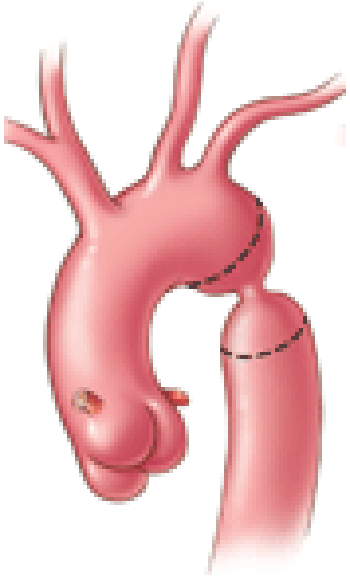


Figure 19 Ebstein anomaly imaging protocol. A. Right ventricular 3-chamber view (RV 3C) steady-state free precession cine image in a 22-year -old with Ebstein anomaly. In this example, the functional tricuspid valve plane is displaced and inflow is directed into the right ventricular outflow tract (white arrow). **B.** The RV 3C image is used to plan a stack of cine images to visualize the displaced tricuspid valve orifice *en face* for anatomic assessment or flow quantification.

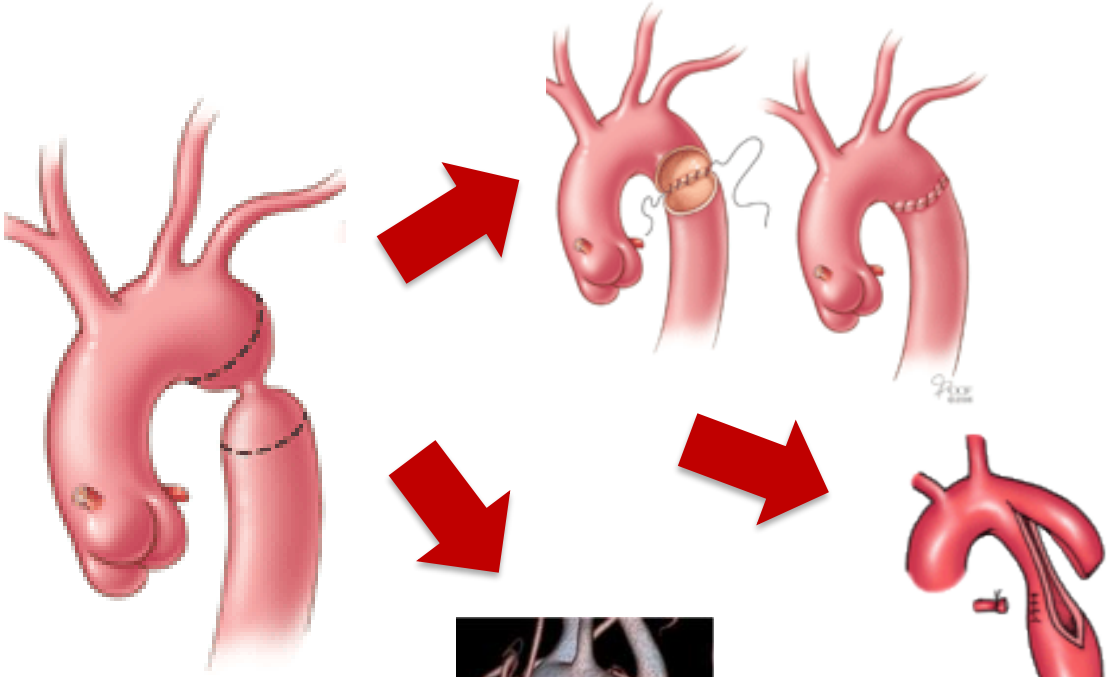
CARDIOPATIE CONGENITE: Coartazione aortica



Restringimento aortico a livello duttale, preduttale o post duttale
Collaterali aorto-sistemiche
Eventuale ipertrofia ventricolare sinistra

Associata a valvola aortica bicuspide/dilatazione aorta scendente/stenosi subaortica/ipoplasia dell'arco/DIV/anomalie v. mitralica (paracadute)

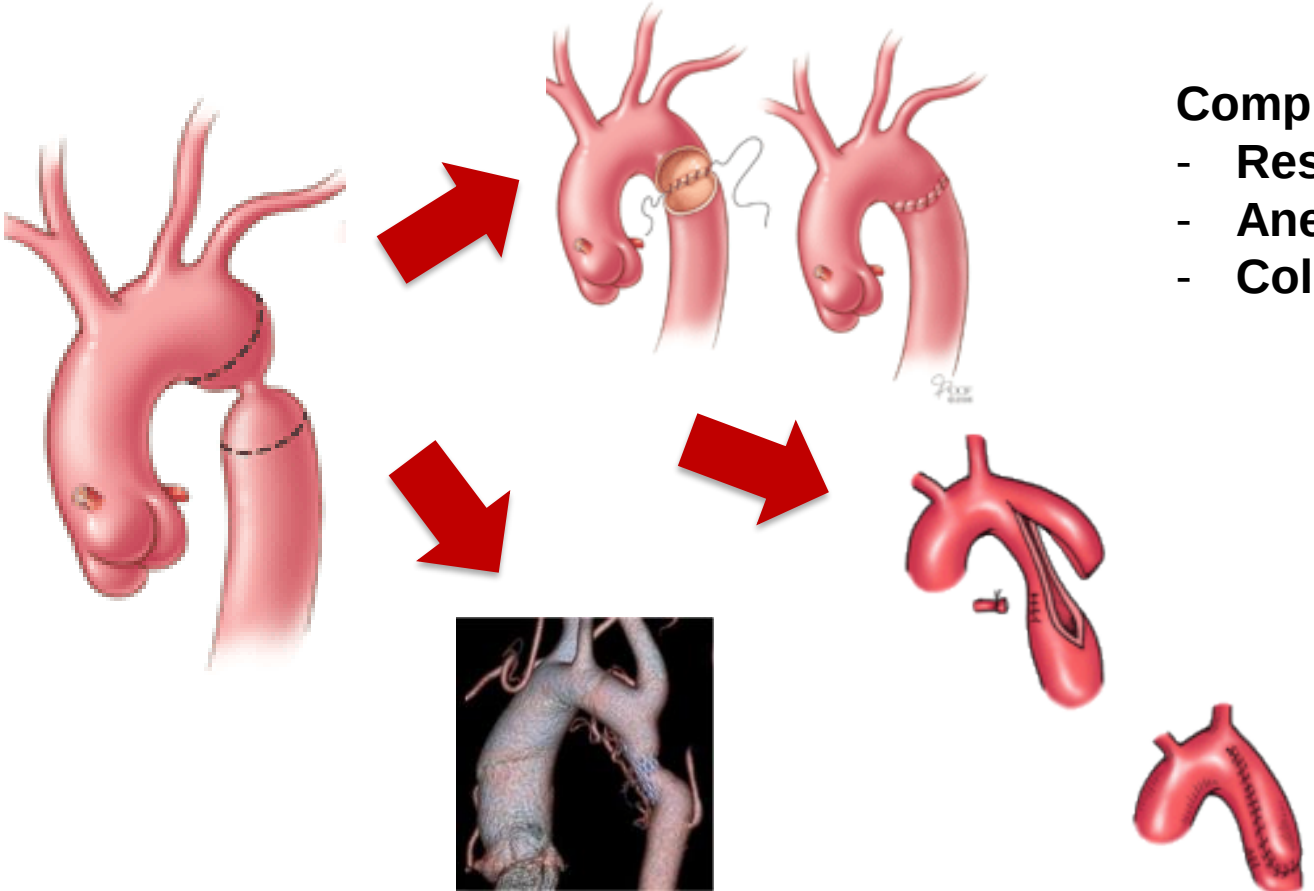
CARDIOPATIE CONGENITE:
Coartazione aortica



Interventi:

- **Anastomosi end to end**
- **Anastomosi con graft**
- **Patch**
- **Subclavian Flap**
- **Bypass**
- **Stent**

CARDIOPATIE CONGENITE:
Coartazione aortica



Complicanze
- **Restenosi**
- **Aneurismi**
- **Collaterali**

CARDIOPATIE CONGENITE: Coartazione aortica

Standard imaging

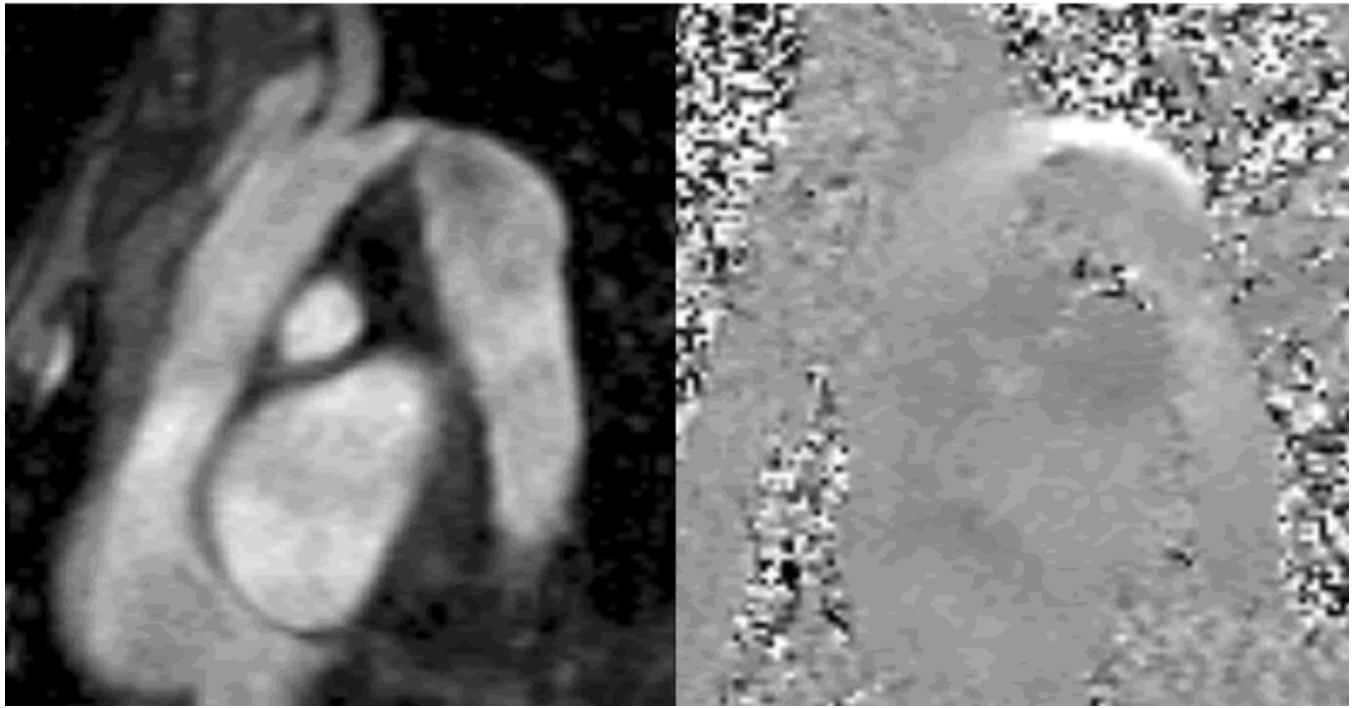
1. Cine CMR: long-axis arco aortico
2. Cine CMR: ventricolografia
3. CE-MRA or 3D SSFP per la vascolatura toracica
4. PC CMR: AAo, AP, aorta discendente a livello del diaframma

Eventualmente anche:

1. Spin echo: long-axis aorco aortico
2. Cine CMR: short-axis radice aortica
3. PC CMR: quantificazione delle collaterali aortiche (misurazione prossimale o appena distale alla coartazione e in aorta discendente a livello del diaframma)
4. LGE

Key reporting elements: posizione, dimensioni e severità della coartazione; lateralità dell'aroc aortico e tipo di branching; presenza di aneurismi, dissezioni o vasi collaterali; parametri ventricolari (NB massa); morfologia e funzionalità valvolare aortica (insufficienza/stenosi)

CARDIOPATIE CONGENITE:
Coartazione aortica



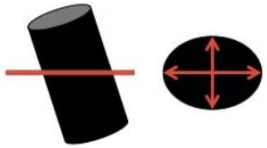
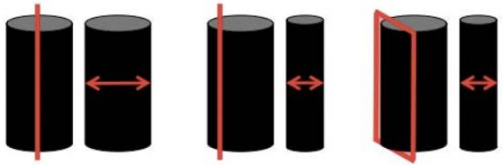
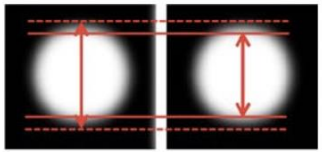
CARDIOPATIE CONGENITE:
Coartazione aortica

CARDIOPATIE CONGENITE:
Coartazione aortica

CARDIOPATIE CONGENITE:
Coartazione aortica

CARDIOPATIE CONGENITE: Coartazione aortica

Caveats of aortic measurements

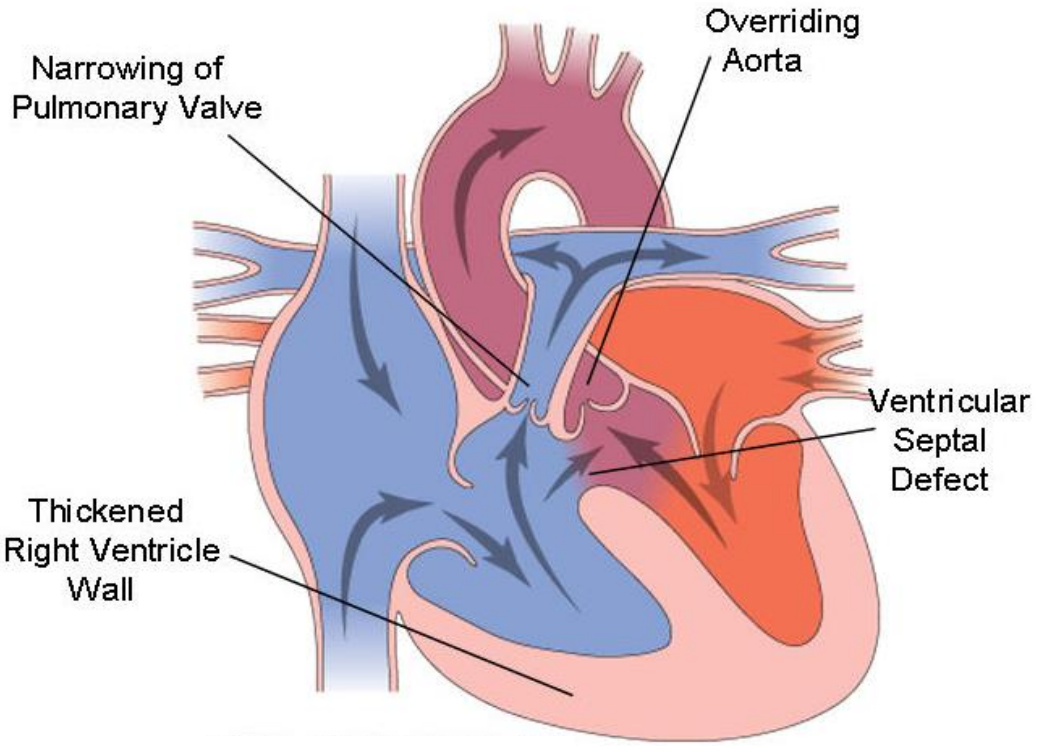
Transaxial	Overestimation due to non-orthogonal plane 
Oblique sagittal	Underestimation due to non-central or non-perpendicular plane 
Black Blood	Overestimation possible due to inclusion of aortic wall 
MRA	Over- / underestimation due to: • Acquisition not cardiac cycle specific • Motion artefacts, particular at aortic root /ascAo 
3D whole heart	Over- / underestimation due to: • Lower spatial resolution • Motion artefacts

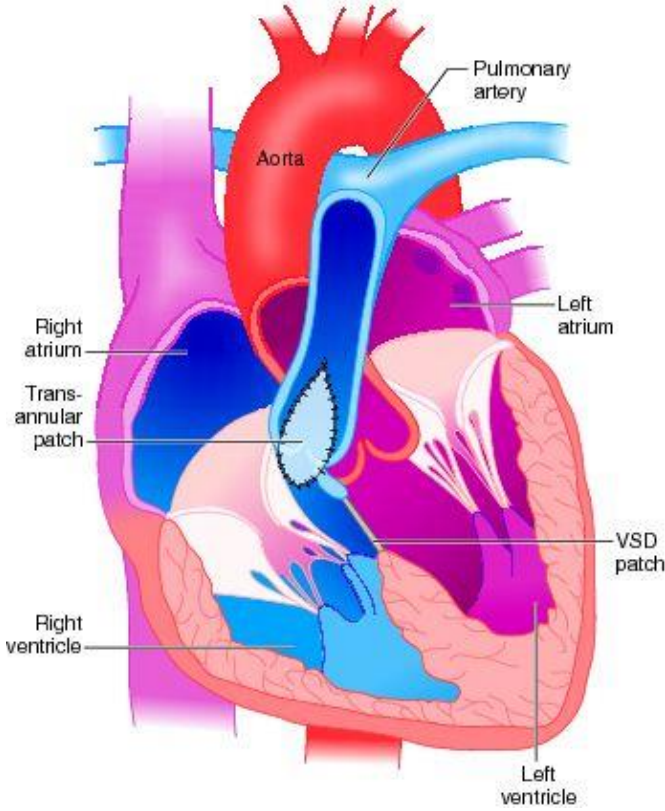
Key issues

1. **Aortic dimensions:**
   - Be aware of caveats of aortic measurements – see above
   - Diastolic measurements from cine images are preferred
   - Be clear in your report, which cardiac phase, orientation and sequence you used for measurements
2. **Severity of coarctation:**
   - Peak systolic flow is often underestimated by CMR - echocardiography superior to CMR
   - Diastolic prolongation of forward flow is a sign of significant coarctation
3. **Collateral flow:**
   - A decrease of <10% (prestenotic - descAo flow) is expected physiologically
   - An increase implies collateral flow rejoining the descending thoracic aorta
   - Abundant collaterals may reduce the gradient across the coarctation and mask the severity of the obstruction
4. **Aneurysms** of the circle of Willis or other cerebral vessels occur in up to 10% of patients with coarctation

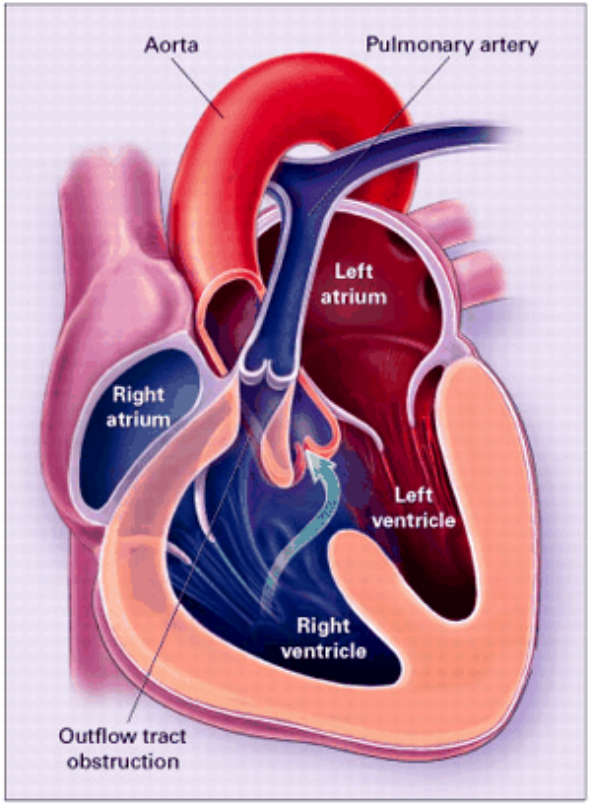
Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE:
Tetralogia di Fallot





CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta



To do list:

- Misura dei volumi, EF e massa biventricolari
- RVOT cine (2 views)
- Radice aortica +/- quantificazione IAo
- Quantificazione IP (RF)
- Valutazione AP dx e sn

CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta

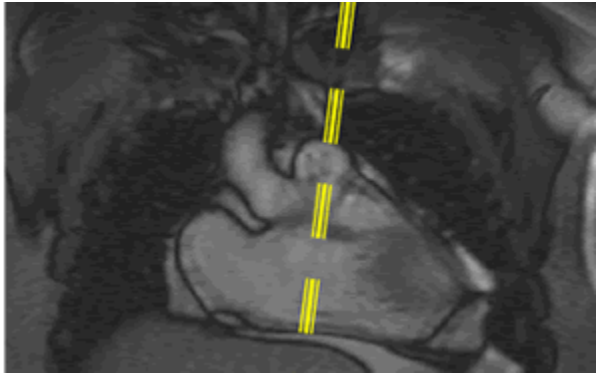
- **Multislice stack nei 3 piani : assiale, sagittale, coronale**
  - Half Fourier single shot TSE (assiale) + single shot SSFP (coronale + sagittale)



CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta

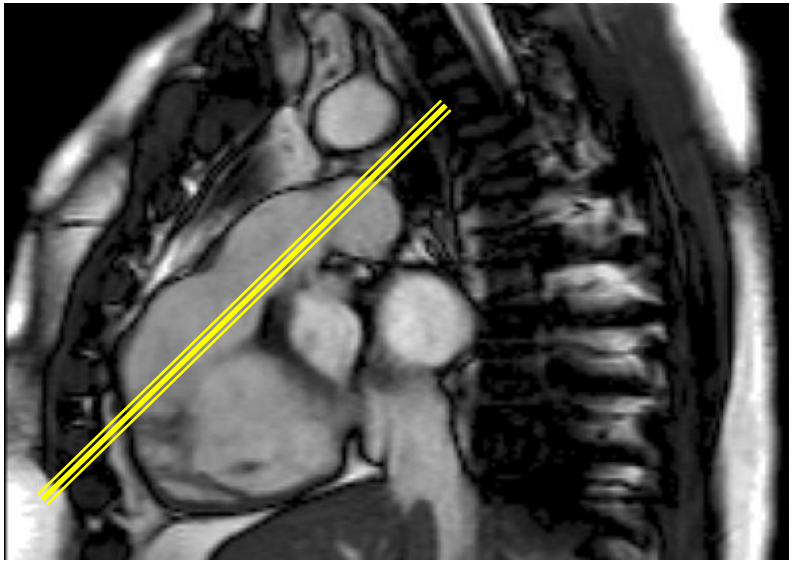
- **Scansioni guida**
- **Cine:**
  - 2Ch, 4Ch e SA
  - RVOT – 2 views
  - LVOT – 2 views

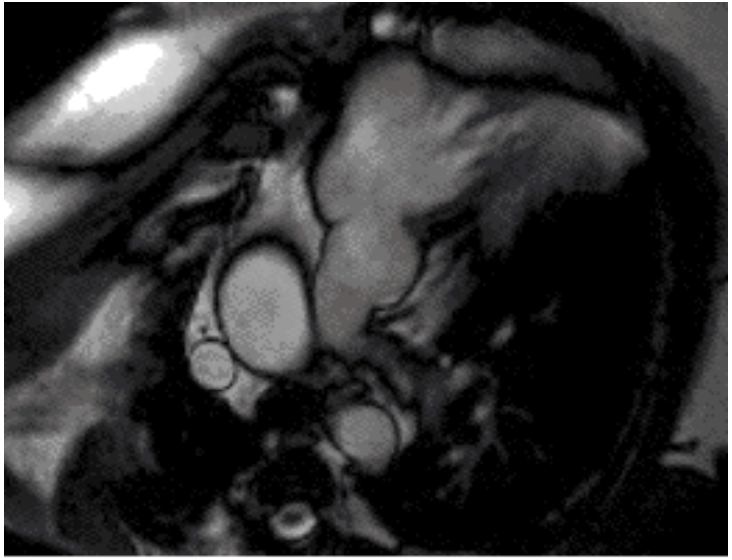
CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta



RVOT cine

CARDIOPATIE CONGENITE:
Tetralogia di Fallot corretta





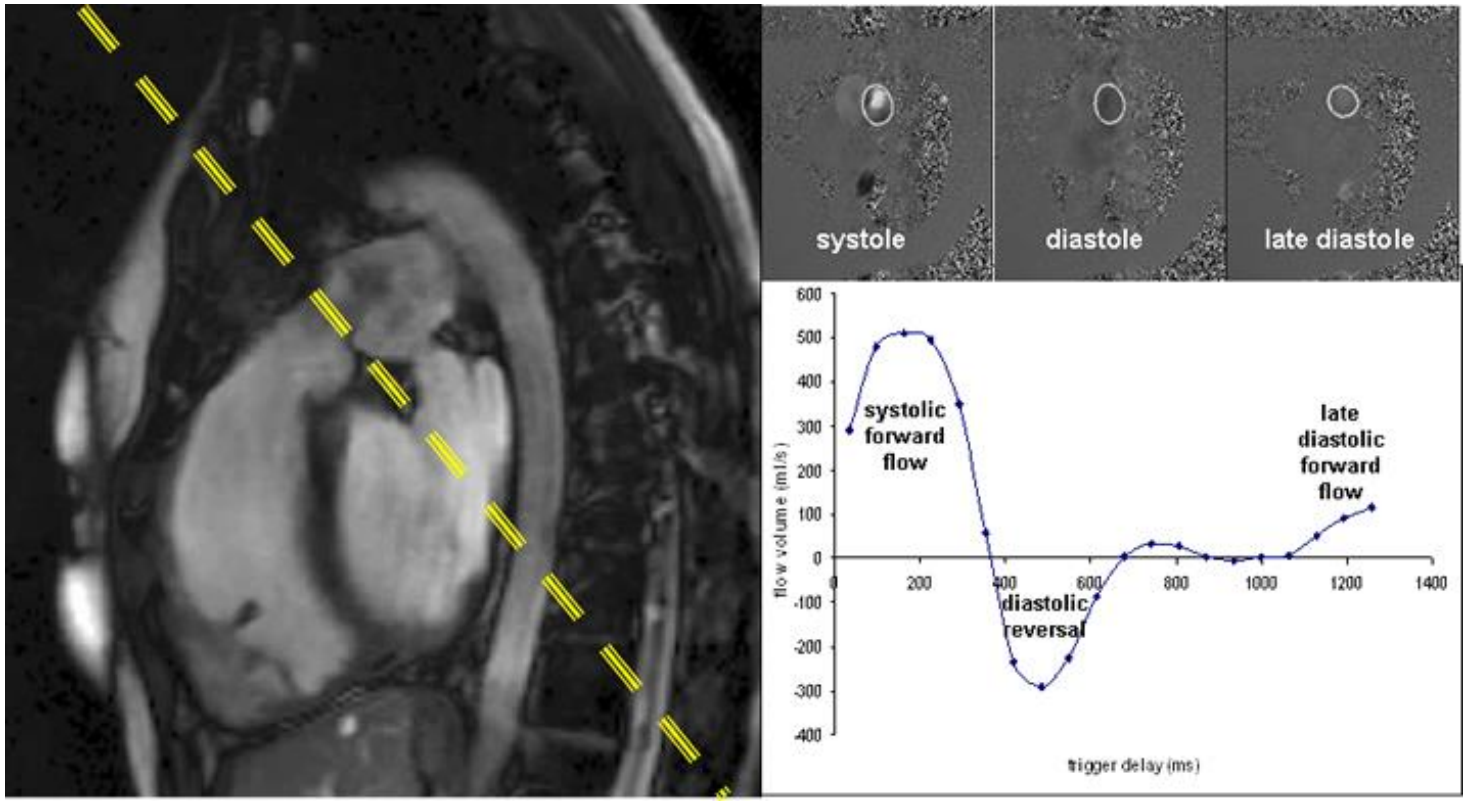
RVOT cine - 2

CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta

Stenosi infundibolare polmonare

CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta

- **Velocity mapping :** Pulmonary regurgitant fraction (PRF)



CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta

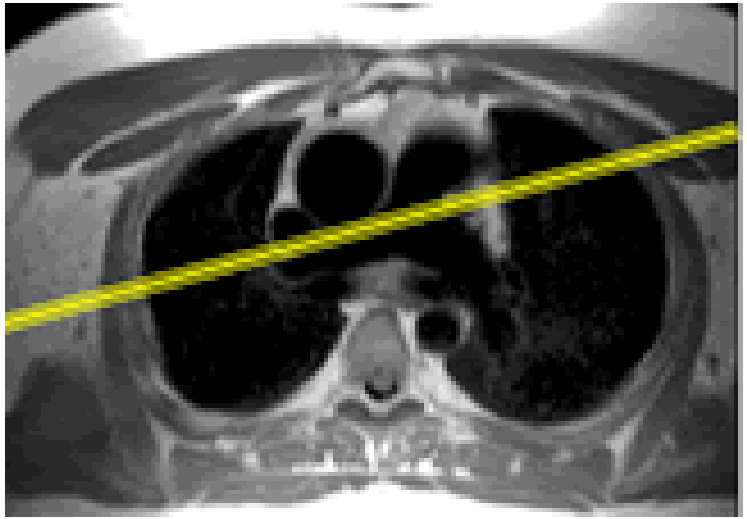
Calcolo PRF

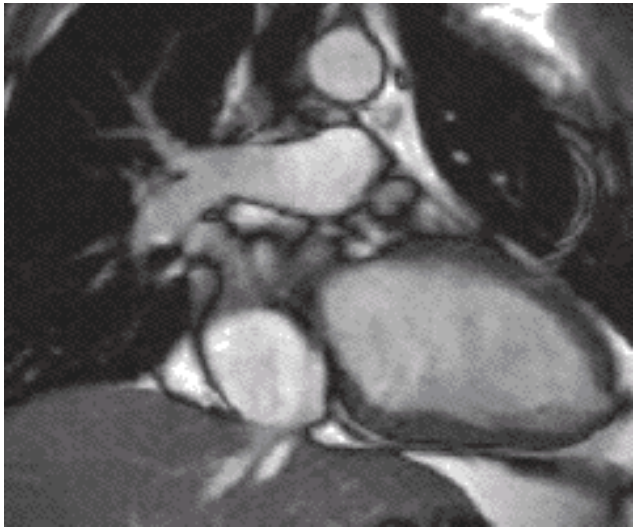
- 1) PRF = flusso retrogrado in AP/flusso anterogrado in AP*100 (es 25/75*100=33%)
- 2) oppure, se non ho altre insufficienze valvolari PRF = (RVSV-LVSV)/RVSV*100 ; es (75-50)/75*100= 33%

CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta

- Cine Ao per misurazioni radice aortica
- AP dx e sn se non chiaramente pervie in assiale
- Vdx “in and out”/ obliqua/2ch
- Ulteriori velocity mapping
    - 2 views per picco velocità AP
    - Ao e PA per Qp:Qs
    - Flusso APdX e Sn
- 3d angiography
    - MAPCA’s / pianificazione interventi (es impianto PV transcatetere)

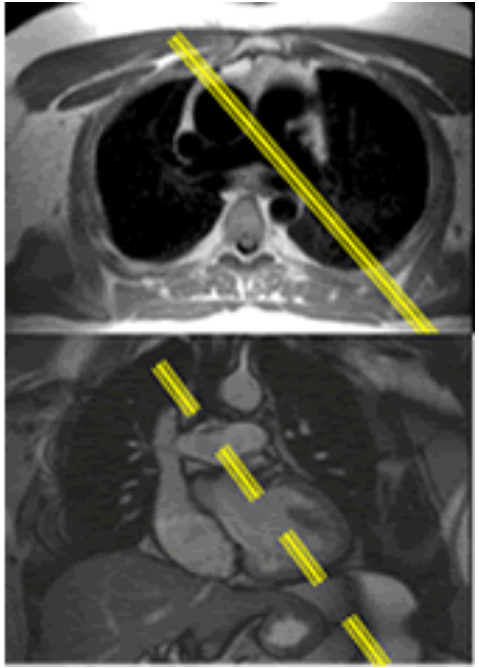
CARDIOPATIE CONGENITE:
Tetralogia di Fallot corretta





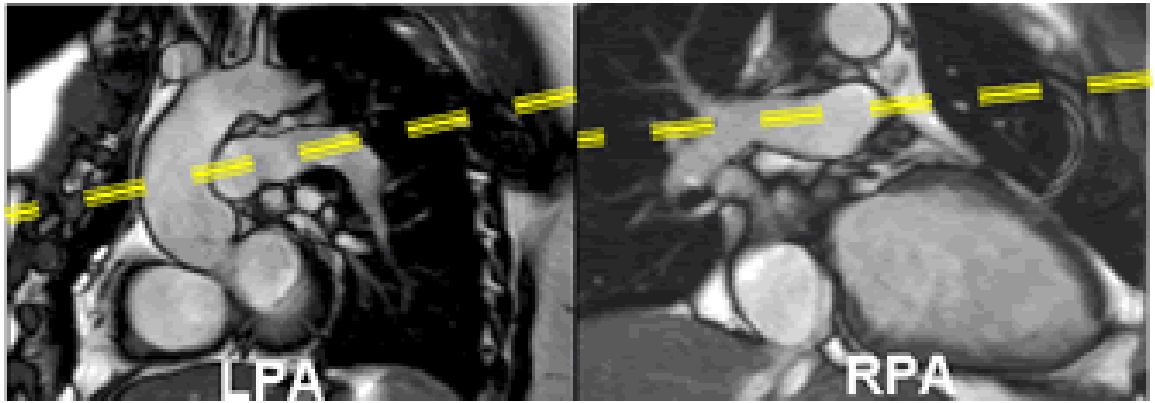
APdx

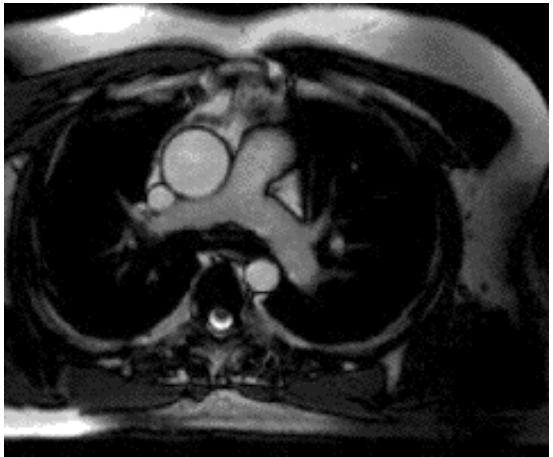
CARDIOPATIE CONGENITE:
Tetralogia di Fallot corretta



APsn

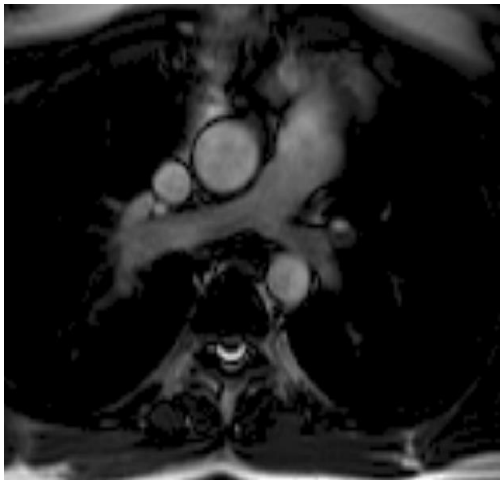
CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta



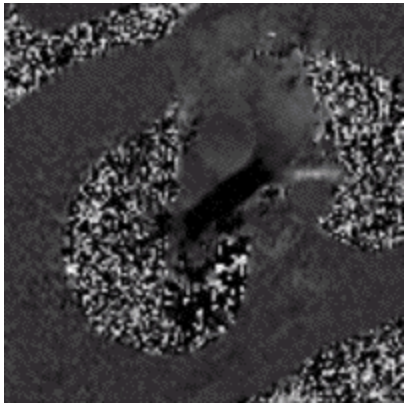


Biforcazione AP

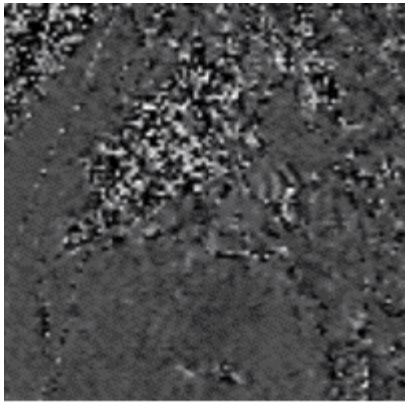
CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta



Cine



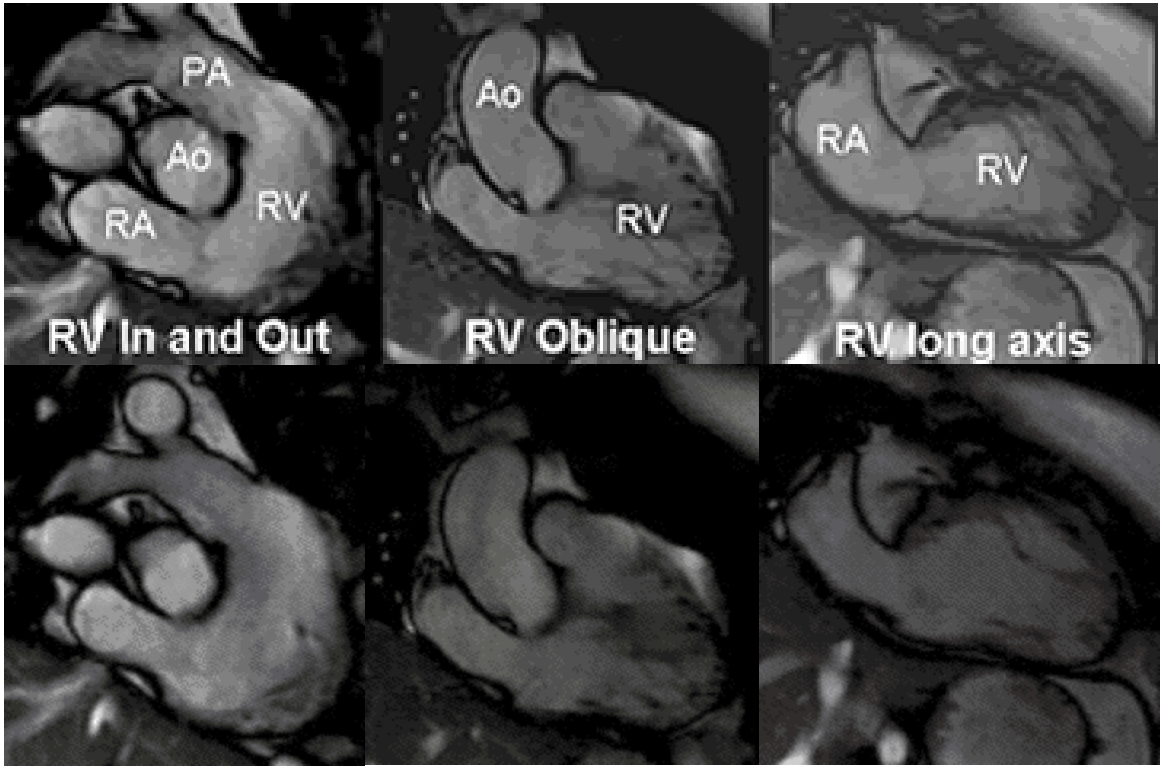
In-plane flow



Through plane flow

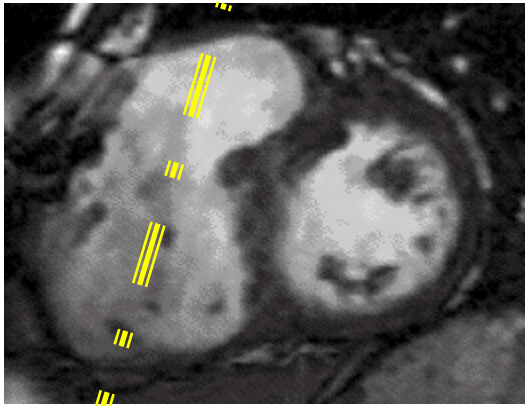
Stenosi AP sinistra

CARDIOPATIE CONGENITE:
Tetralogia di Fallot corretta

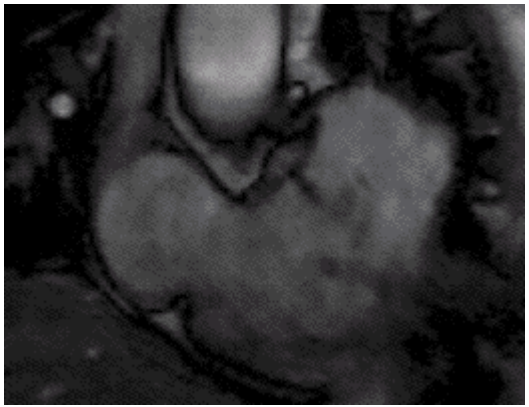


CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta

- Patch leak may be seen in:
  - LVOT view
  - RV in and out
  - RV oblique views
  - SA view



- If uncertain:
  - cross-cut a SA view where a jet core is suspected

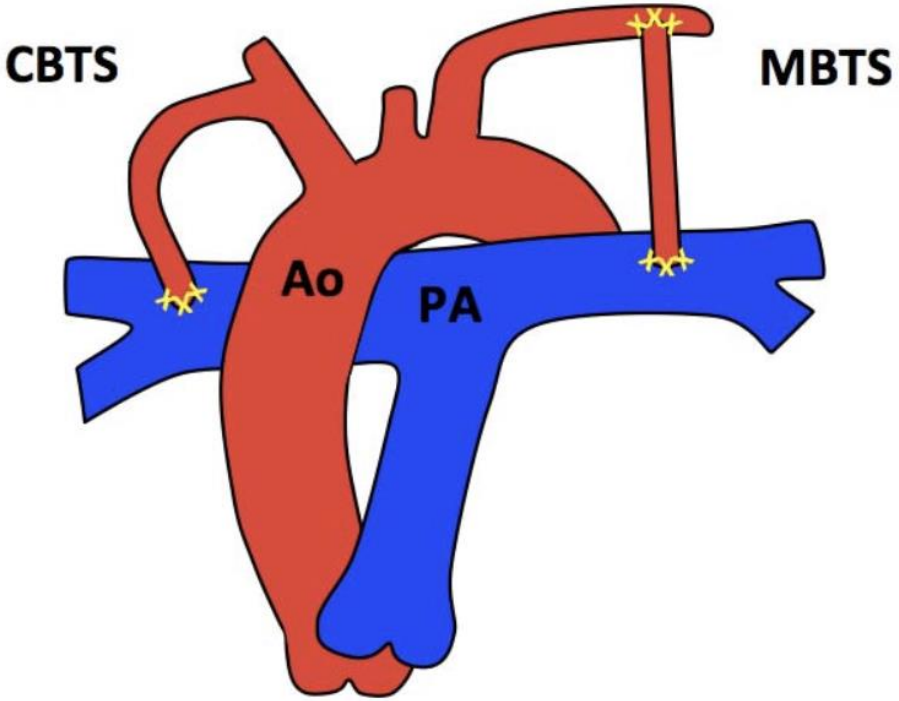


- Add NBH velocity:
  - Aorta and PA
  - Calculate Qp:Qs ratio
  - Stroke volume ratio may be relevant

CARDIOPATIE CONGENITE: Tetralogia di Fallot corretta

Acquisizioni 3D: utili anche per il planning di sostituzione valvolare polmonare percutanea (decorso coronarie)

CARDIOPATIE CONGENITE: Blalock-Taussig shunt

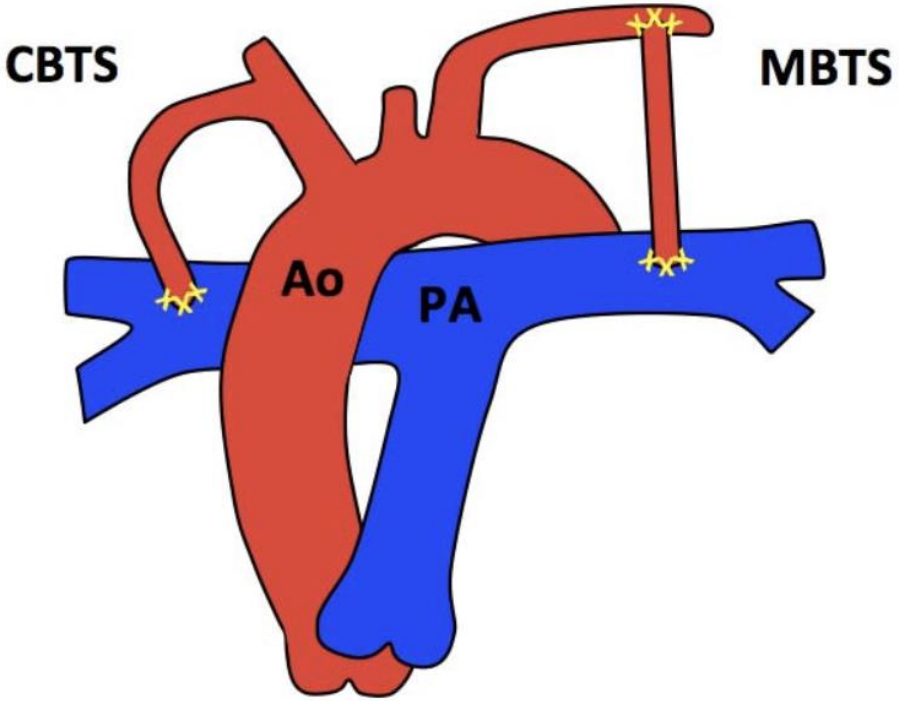


Findings
Classic BT Shunt (CBTS)
• Subclavian artery to PA
Modified BT Shunt (MBTS)
• Gore-Tex tube from subclavian artery to PA

Intervento palliativo nelle cc cianogene
Possibile bridge per Glenn/Fontan
Shunt talora piccolo > MRA sincronizzata con aorta
Possibile ischemia vertebrobasilare per furto della succlavia
Varianti: Waterston Pott, Central, Cooley

Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE: Blalock-Taussig shunt



Findings
Classic BT Shunt (CBTS)
• Subclavian artery to PA
Modified BT Shunt (MBTS)
• Gore-Tex tube from subclavian artery to PA

Complicanze post operatorie:

- **Stenosi shunt > stenting possibile**
- **Aneurismi**
- **Dilatazione PA**
- **Ipertensione polmonare**

Adattato da: ESC/EACVI CHD/CMR guide, 2014

CARDIOPATIE CONGENITE: BT shunt

PROTOCOLLO BT shunt

Standard imaging

1.Stacks anatomici
2.Cine VLA, HLA, LV stack, RV stack
3. Cine LVOT, RVOT
4.Cine arteria polmonare principali, sn, dx
5.PC CMR: valvola aortica, flusso Pas distale allo shunt

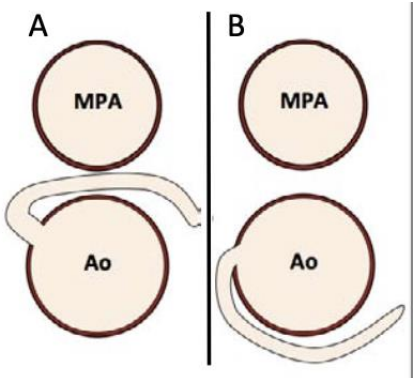
Eventualmente anche:

MRA

Elementi essenziali: lunghezza, pervietà e flusso nello Shunt, aneurismi, parametri ventricolari, Qp/Qs

CARDIOPATIE CONGENITE:
Anomalie dell'origine e decorso coronarici

- "Maligna" se decorso interarterioso (specialmente LCA dal seno di Valsalva destro)
- Cause di ischemia: compressione dinamica interarteriosa, origine "slit-like", bridging miocardico
- ALCAPA/ARCAPA: di solito associate ad anomalie della cinetica regionale, ev. insufficienza mitralica funzionale, ev. dilatazione ventricolare > diagnosi anche in età adulta



In figura: Origine anomala di LCA dal seno di valsalva destro:
A) Decorso interarterioso
B) Decorso retroaortico

CARDIOPATIE CONGENITE:
Anomalie dell'origine e decorso coronarici

CARDIOPATIE CONGENITE: Anomalie dell'origine e decorso coronarici

Protocollo MRI:

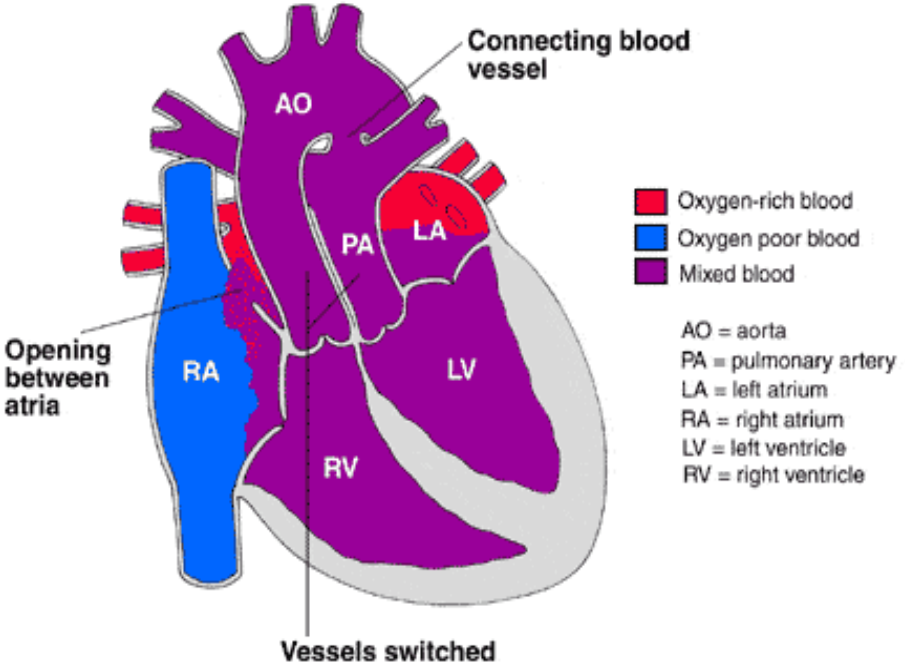
- 3DSSFP, tutta la regione cardiaca

Inserire nel report:

- Origine (alta/bassa/commissurale, dal seno coronarico opposto, fuori dai seni coronarici, osti separati di DA e Cx, ALCAPA, ARCAPA…)
- Decorso (interarterioso/anteriore o retroaortico/posteriore)
- Anomalie dell'anatomia intrinseca (ectasia, aneurismi, ipoplasia, decorso intramurale)
- Anomalie delle diramazioni
- Eventuale relazione con altre strutture cardiache

CARDIOPATIE CONGENITE: Trasposizione delle grandi arterie

- Origine dell'aorta dal Vdx e dell'arteria polmonare dal Vsn
- Emergenza neonatale, sempre corretta tempestivamente
- Fino agli anni '80, redirezionamento del flusso venoso con procedura di Mustard o Senning ("atrial switch")
- Poi correzione "anatomica" con "arterial switch"

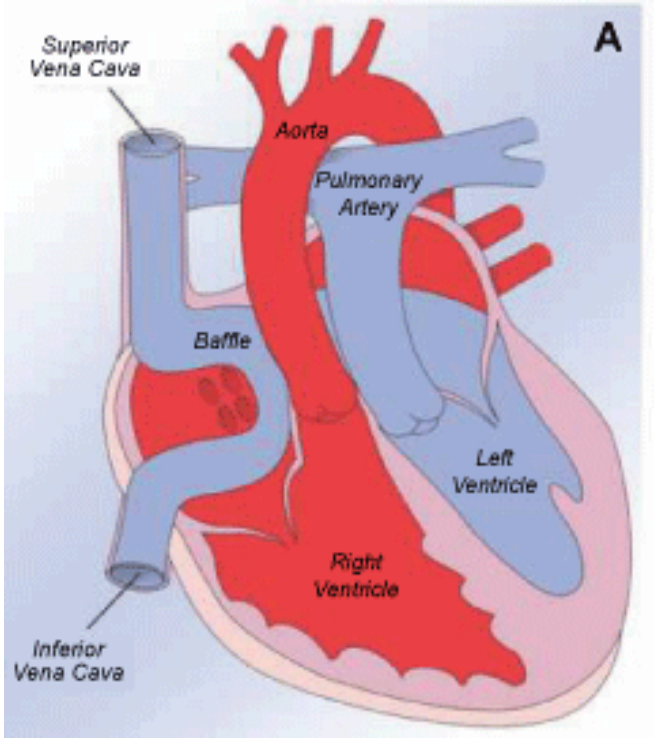


CARDIOPATIE CONGENITE: D-Trasposizione delle grandi arterie

- **Second most common cyanotic heart lesion with incidence of approximately 30/ 100k live births**
- **Approximate 5-7% of all CHD defects**
  Approximately 40-45% have a VSD (Complex TGA)
- **Over half of these with concomitant LVOT obstruction**
- **Rare associated lesions include arch anomalies and pulmonary venous return abnormalities**

Mustard and Senning procedure

- Un baffle (in pericardio o dacron) redireziona il flusso venoso cavale in atrio sinistro, così da inviarlo al ventricolo sinistro, il quale diventa sottopolmonare
- Il Vdx diventa sistemico



CARDIOPATIE CONGENITE: D-TGA s/p atrial switch

Standard imaging

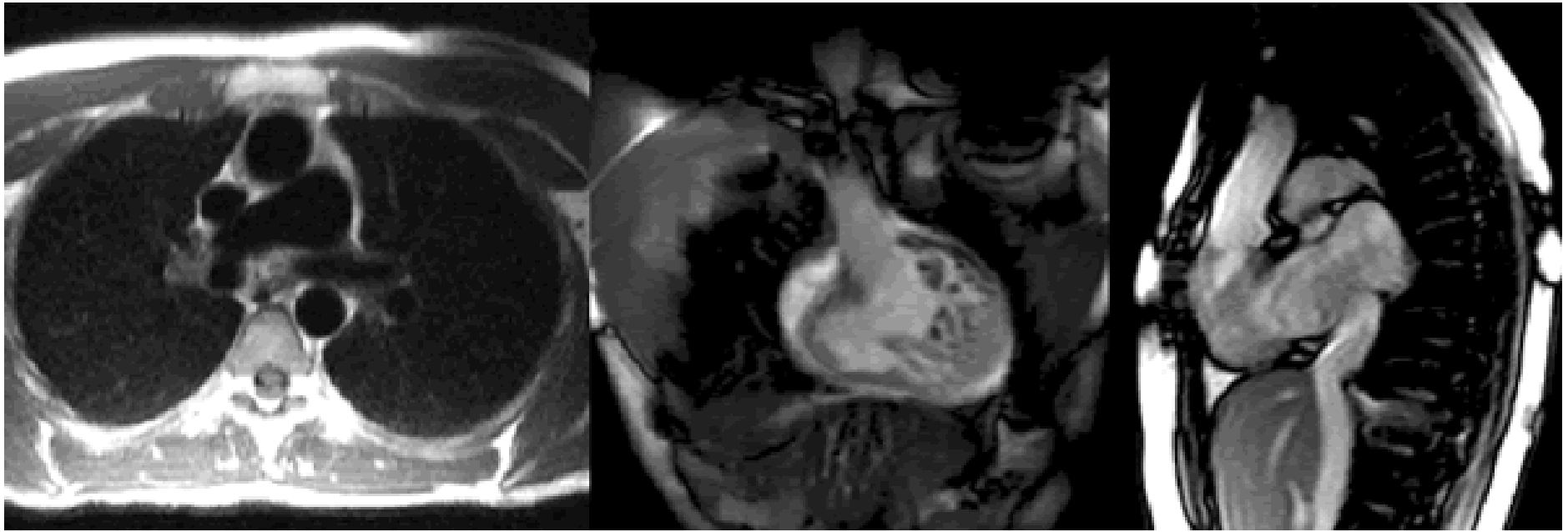
1.Cine CMR: axial stack from the mid-liver to the top of the aortic arch
2.Ventriculography module
3.Cine CMR: oblique planes to image the SVC and IVC pathways in long-axis
4.CE-MRA or 3D SSFP to image the thoracic vasculature, and systemic and pulmonary venous baffles
5.PC CMR: AAo, MPA, branch PAs, tricuspid and mitral valves

Additional case-specific/comprehensive imaging

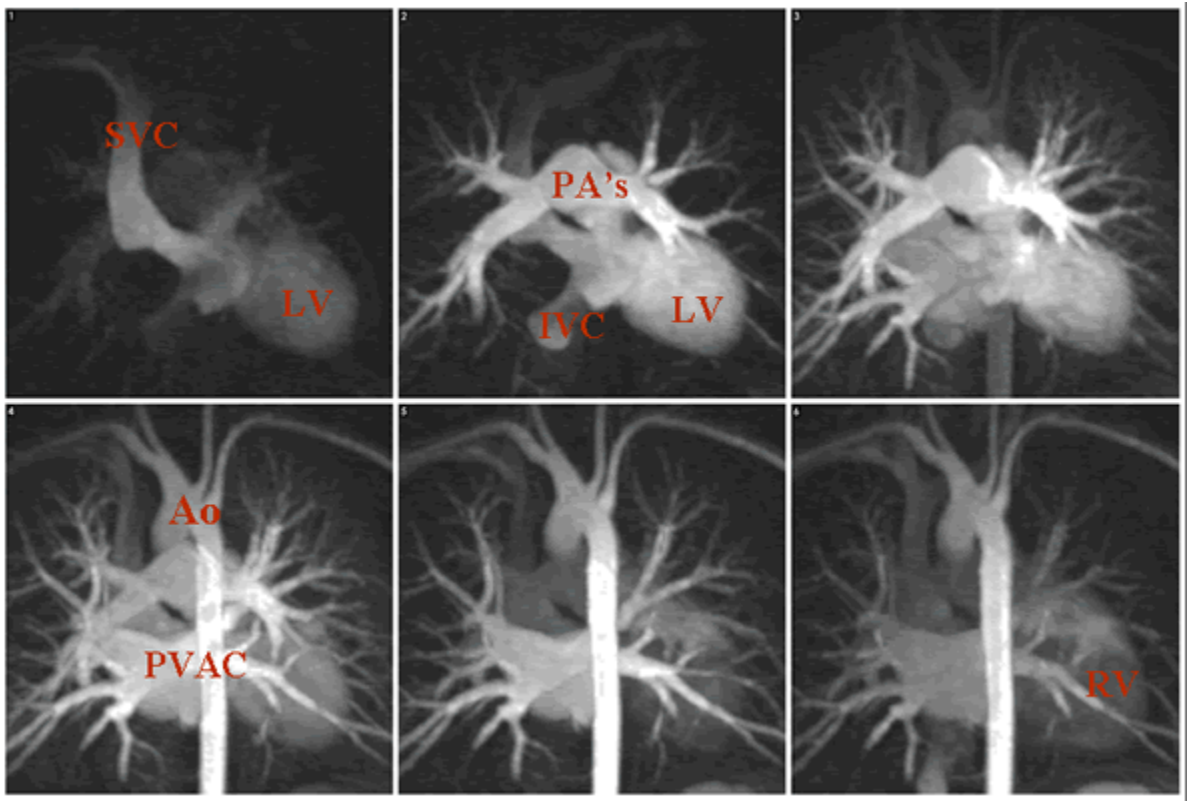
1.LGE module
2.PC CMR: SVC distal to the azygous vein and IVC when systemic venous obstruction is suspected.

Key reporting elements: location and severity of systemic and pulmonary venous pathway obstruction, atrial baffle leak, ventricular parameters, severity and mechanism of left or right ventricular outflow tract obstruction, tricuspid regurgitation, Qp/Qs, branch pulmonary artery flow distribution, SVC/IVC flow ratio as an indicator of systemic pathway obstruction

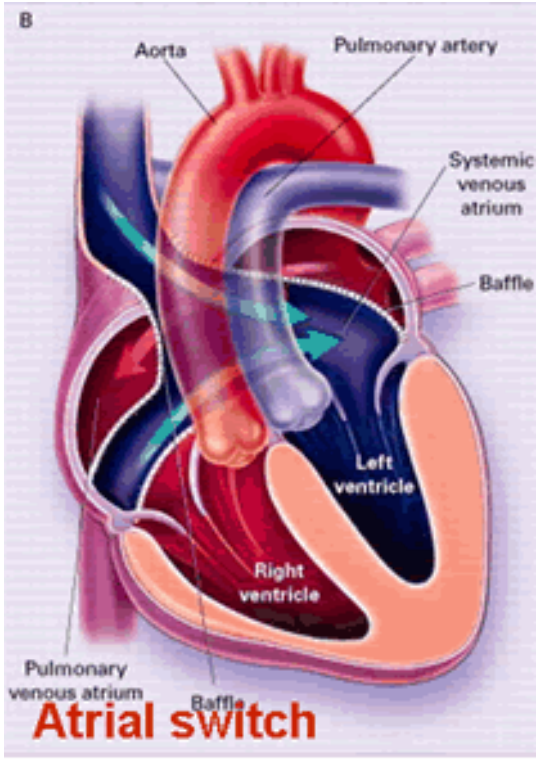
CARDIOPATIE CONGENITE: D-TGA s/p atrial switch



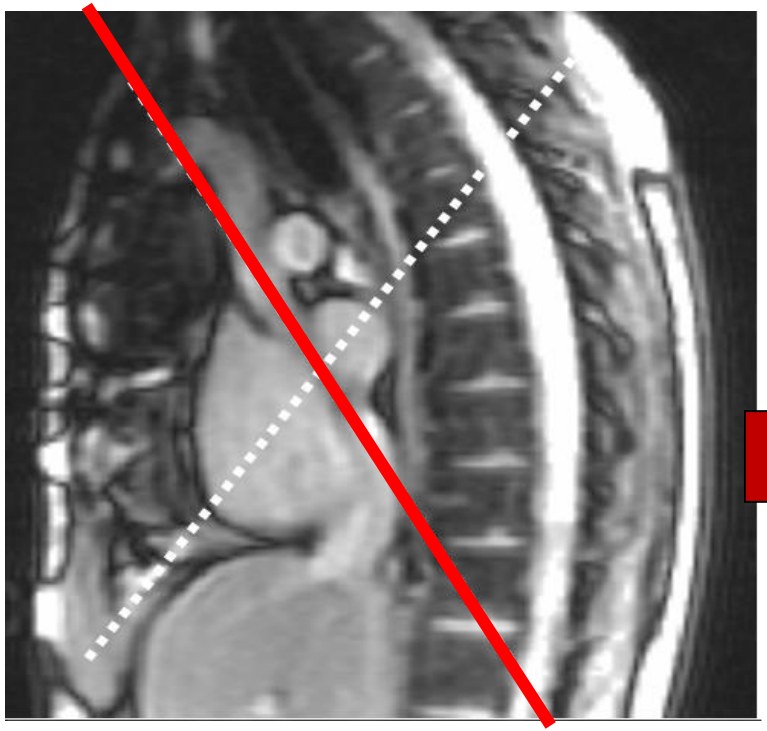
CARDIOPATIE CONGENITE:
D-TGA s/p atrial switch



CARDIOPATIE CONGENITE:
D-TGA s/p atrial switch

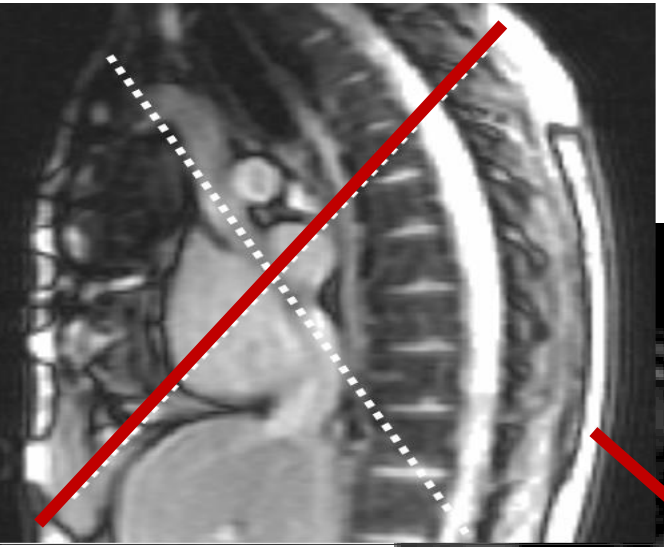


CARDIOPATIE CONGENITE:
D-TGA s/p atrial switch

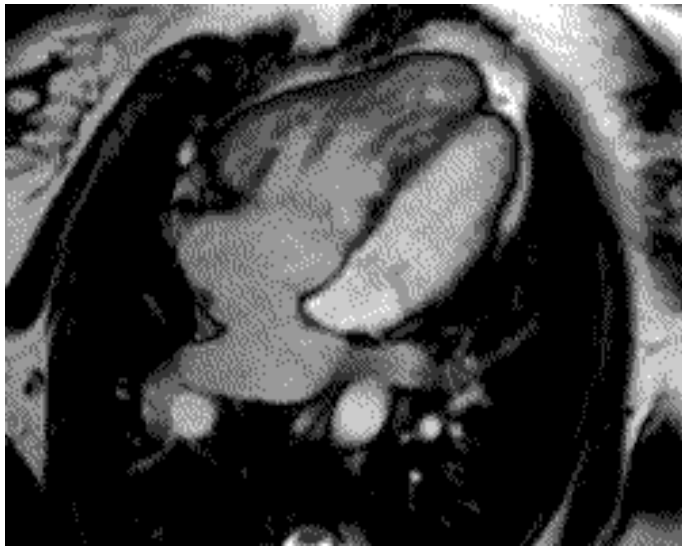


Multislice sagittale

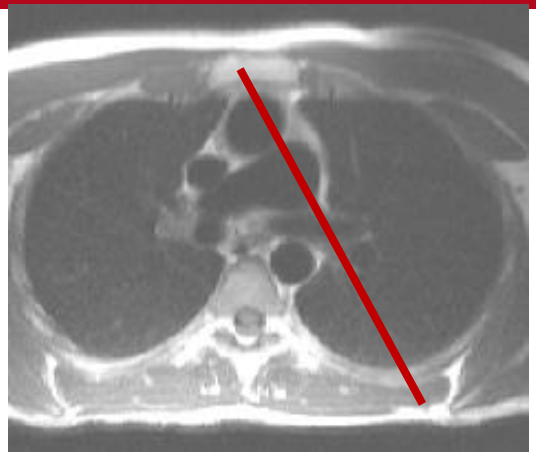
CARDIOPATIE CONGENITE: D-TGA s/p atrial switch

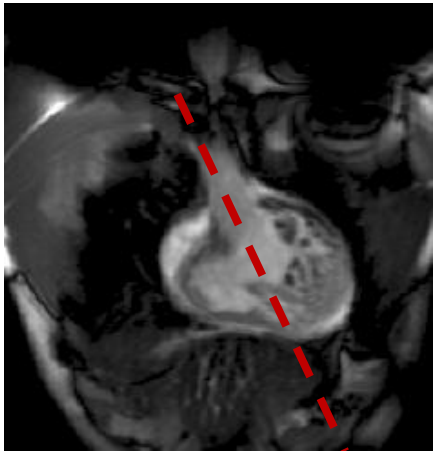


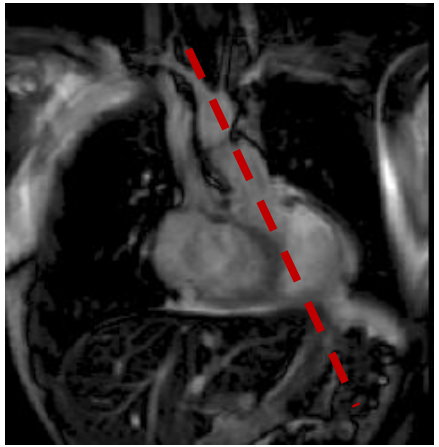
Multislice Sagittale e coronale



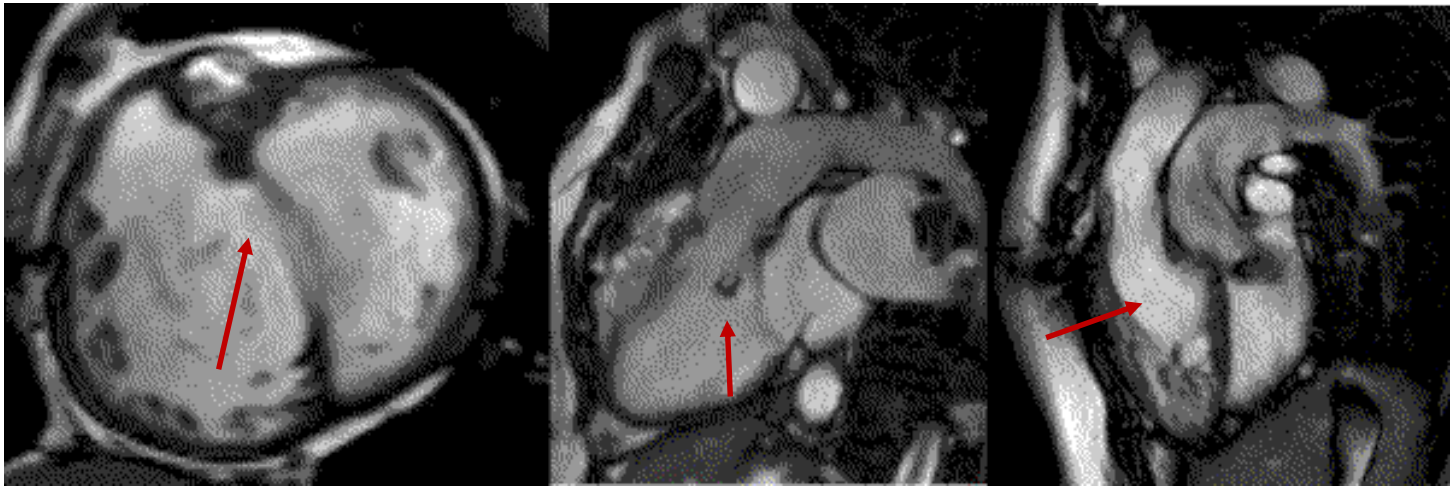
CARDIOPATIE CONGENITE:
D-TGA s/p atrial switch



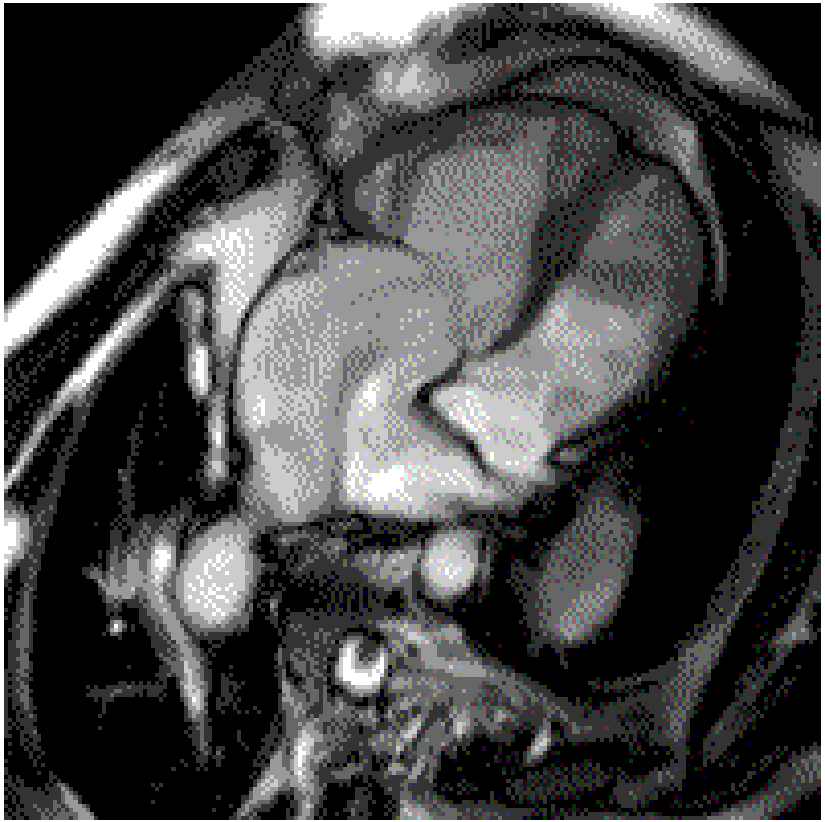




CARDIOPATIE CONGENITE: D-TGA s/p atrial switch



CARDIOPATIE CONGENITE: D-TGA s/p atrial switch

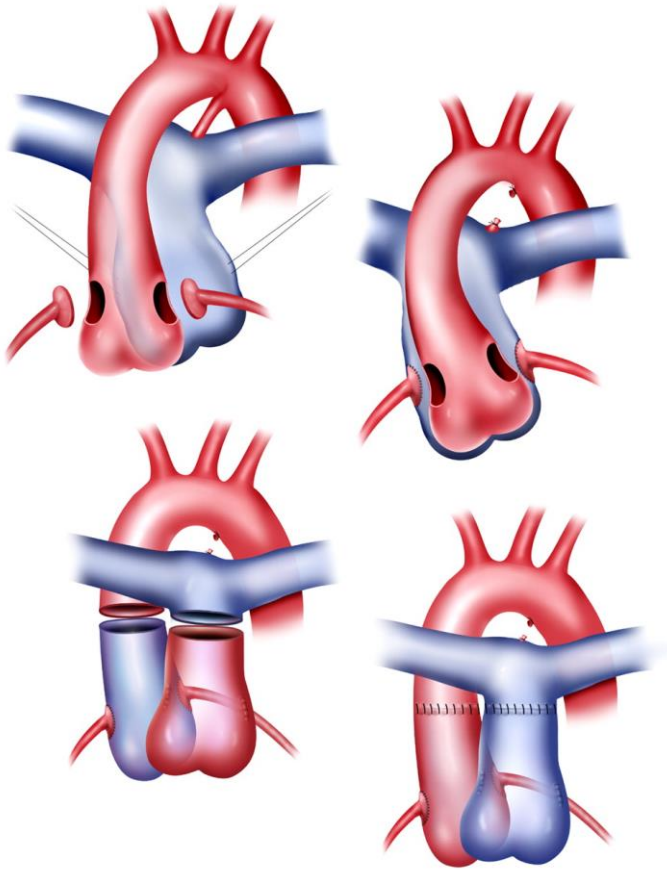


Disfunzione biventricolare

CARDIOPATIE CONGENITE: Trasposizione delle grandi arterie

Arterial switch

- Tecnica elettiva chirurgica per le TGA dagli anni ’80
- Resezione delle grandi arterie, manovra di Lecompte, reanastomosi all’efflusso ventricolare corretto, reimpianto coronarie nella neo-radice aortica



CARDIOPATIE CONGENITE: D-Trasposizione delle grandi arterie

Arterial switch

Majority survive to adulthood
Survival at 15 years ~ 88%
Freedom from reintervention is 82% at 15 years
Reoperation < 10%

TABLE 3 Post-Operative Sequelae Following the Arterial Switch Operation

Long-Term Post-Operative Sequelae	Incidence
Supravalvular pulmonary stenosis*	~10%
Supravalvular aortic stenosis*	~5%
Neoaortic root dilation	Nearly universal
Neoaortic regurgitation	Most (moderate or severe in <10%)
Asymptomatic coronary occlusion	2%–7%
Sudden cardiac death	<1%
Arrhythmia	2%–10%
Aortic dissection or rupture	Unknown

*Requiring intervention. Modified with permission from Wernovsky et al. (152).

CARDIOPATIE CONGENITE: D-Trasposizione delle grandi arterie

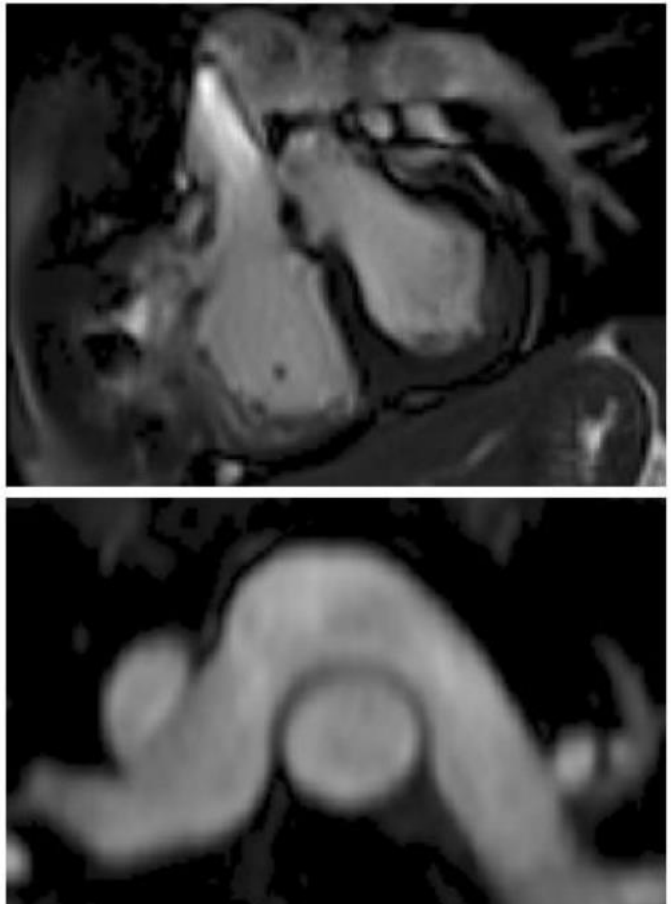
Arterial switch

Initial ASO mortality quite high due to coronary ischemia

• Incidence of myocardial ischemia is most prevalent in first 3 months

- Bimodal pattern of incidence

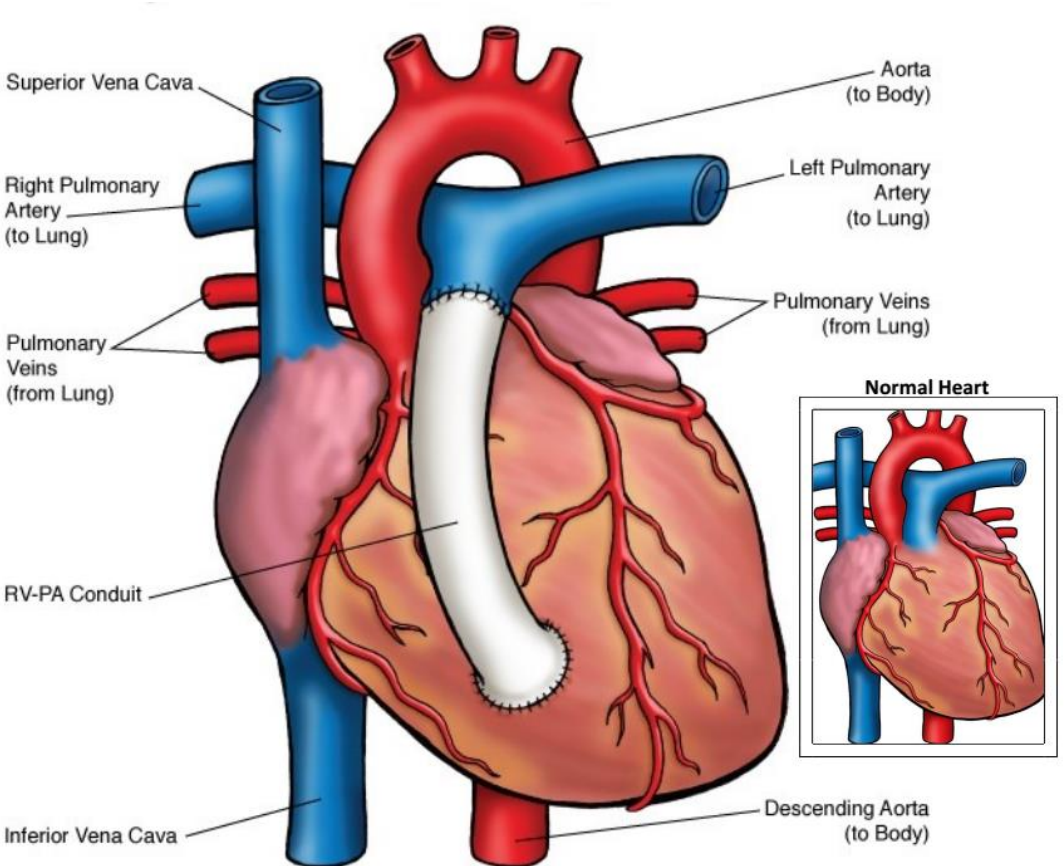
• Coronary obstruction present in 5% - 7% of ASO survivors



Arterial switch: protocollo

Axial Black Blood
• Cine imaging - VLA
- 4 Chamber - SAX
- RVOT
- Branch PA's
• Flow Quantification - Aorta
- MPA
- RPA/LPA - AVV
• 3D Whole Heart
• MRA
• Delayed Enhancement

CARDIOPATIE CONGENITE: Condotti Vdx - AP



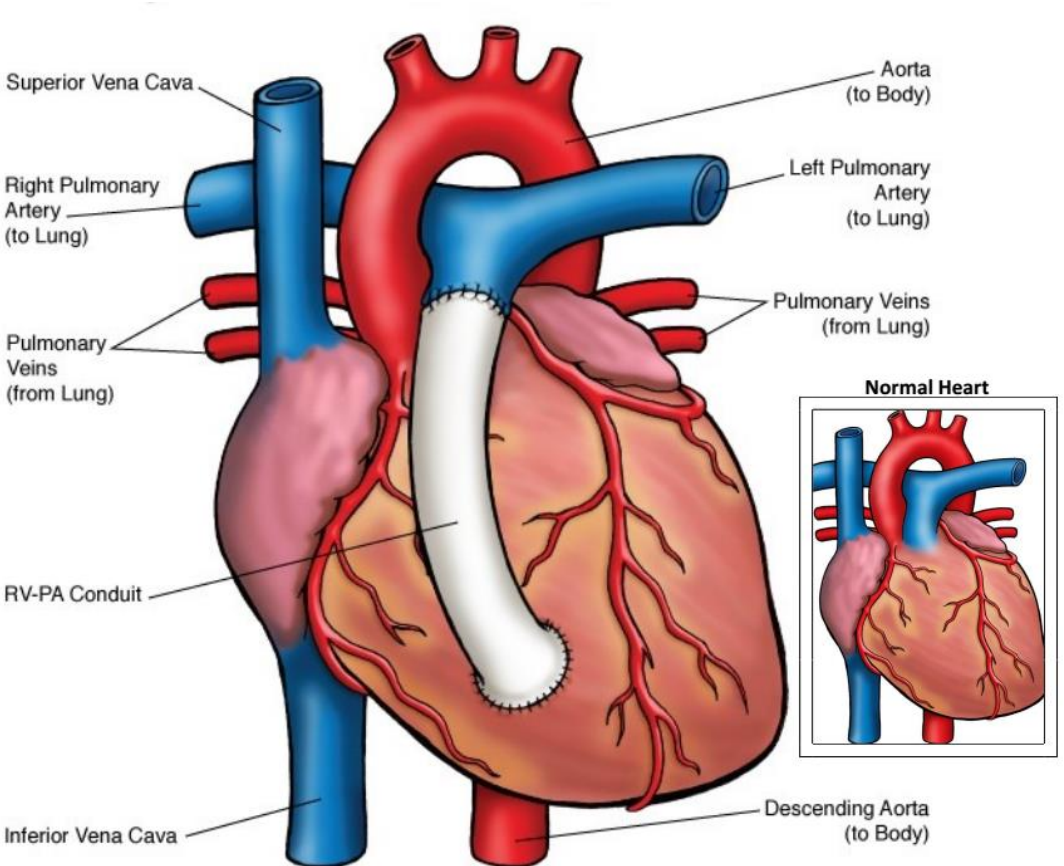
Rastelli

Permette correzione simultanea di dTGA/DORV + DIV + atresia polmonare/stenosi polmonare/sottovalvolare polmonare

Mantiene Vsn sistemico

Generalmente eseguito a 1-2 anni, insieme a BT shunt

CARDIOPATIE CONGENITE: Condotti Vdx - AP



Rastelli

Initial interventions

- Conduit RV-MPA
- Intra-ventricular baffle
  - VSD closure
  - Redirection of left ventricular outflow to anterior aortic valve

Late interventions

- Re-operation conduit
- VSD closure device

Post-operative complications

- Conduit or conduit valve stenosis / obstruction
- LVOT obstruction
- Residual VSD
- Residual ASD
- Branch PA stenosis

CARDIOPATIE CONGENITE: Condotti Vdx - AP

Rastelli

Protocol	
1. **Anatomy stack**	6. **MPA flow**
2. **VLA, HLA, LV stack, RV stack**	7. **AoV flow**
3. **LVOT, RVOT**	8. Branch PAs flow
4. **PAs**	9. 3D whole heart
5. **Conduit cross-cuts**	10. Coronal cine stack

Report

1. **Size** (corrected for BSA) and **function**
   - LV: EDV, ESV , SV, EF, RWMA, mass
   - RV: EDV, ESV, SV, EF, RWMA
2. **Conduit patency and proximity to sternum**
3. **LVOT obstruction**
4. **MPA & branch PA patency (and flow)**
5. **Residual ASD, VSD, Qp (MPA flow) : Qs (Ao flow)**
6. **Course of coronary arteries and likelihood of compression if percutaneous intervention to the conduit**

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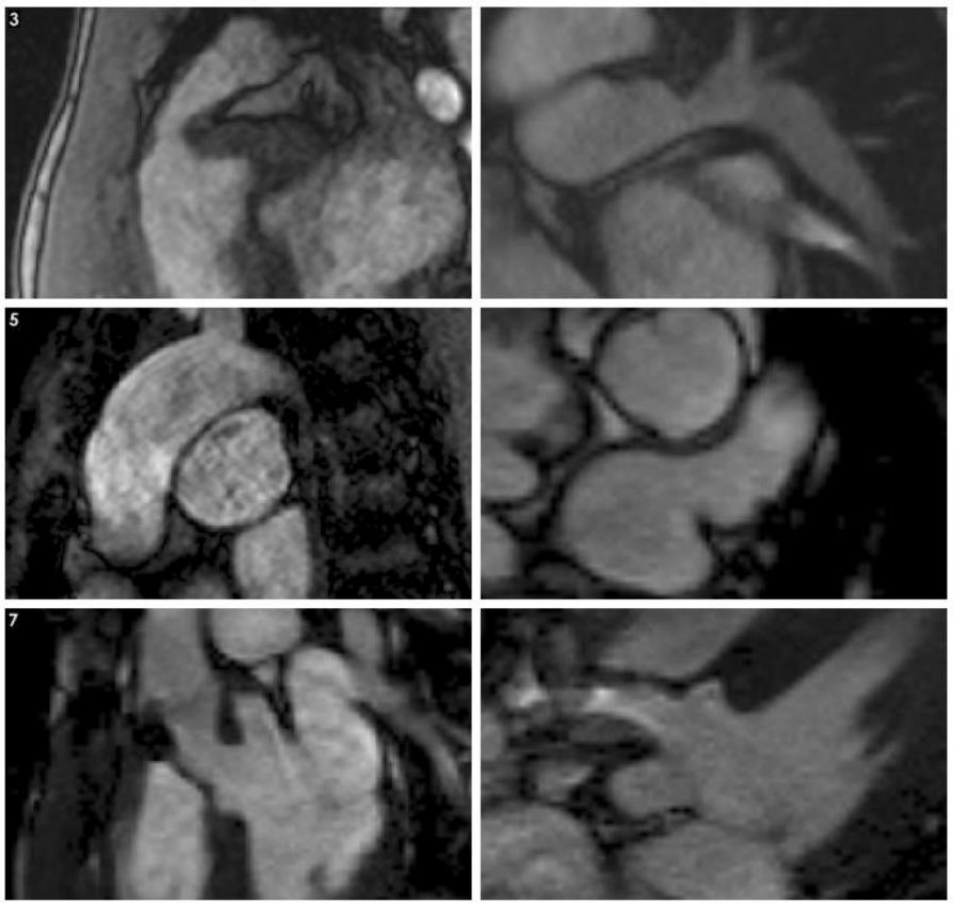
CARDIOPATIE CONGENITE: Condotti Vdx - AP



Rastelli

CARDIOPATIE CONGENITE: Cuori univentricolari

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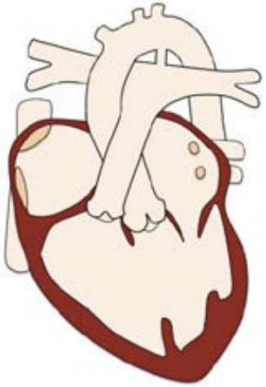
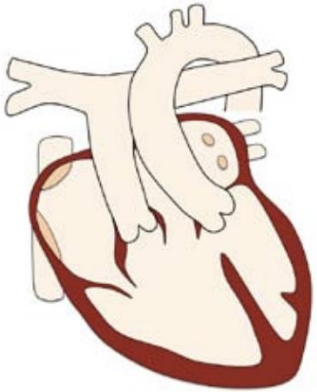
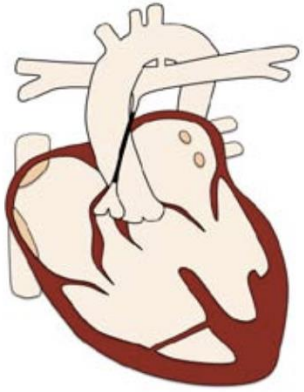


Tricuspid atresia

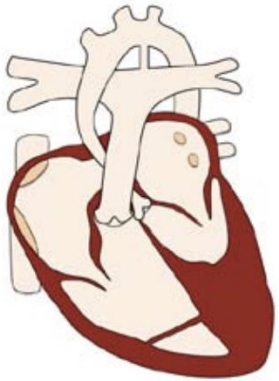


Double inlet ventricle

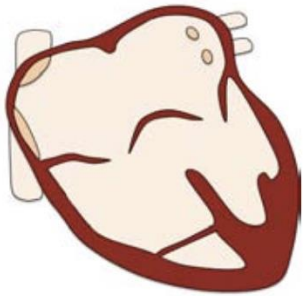


Pulmonary atresia

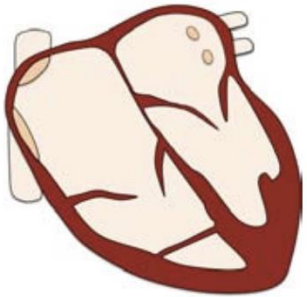
Pre-intervento



Hypoplastic left heart



Unbalanced AVSD



Ebstein anomaly

CARDIOPATIE CONGENITE: Cuori univentricolari

Adattato da: ESC/EACVI CHD/CMR guide, 2014

Palliative procedure

Stage 1 - Glenn procedure

Stage 2 - Fontan completion

- Total cavo-pulmonary connection (TCPC)
  - Lateral tunnel - intracardiac
  - Extracardiac
  - Atrio-pulmonary

Additional interventions

Atrial septostomy

- to maintain systemic venous return to heart

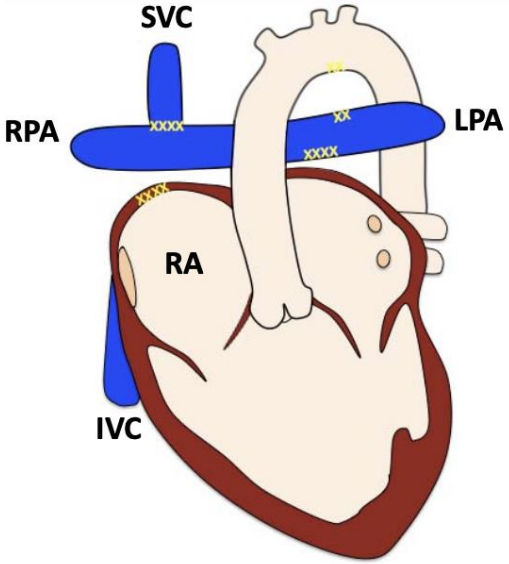
Arterial shunt

- if inadequate pulmonary blood supply

PA banding

- if excessive pulmonary blood supply

CARDIOPATIE CONGENITE: Cuori univentricolari



Initial interventions

- SVC detachment from RA
- Reconnection to RPA

Late interventions

- Usually proceeds Fontan
- Collateral vessels may require occlusion if significant desaturation

Post-operative complications

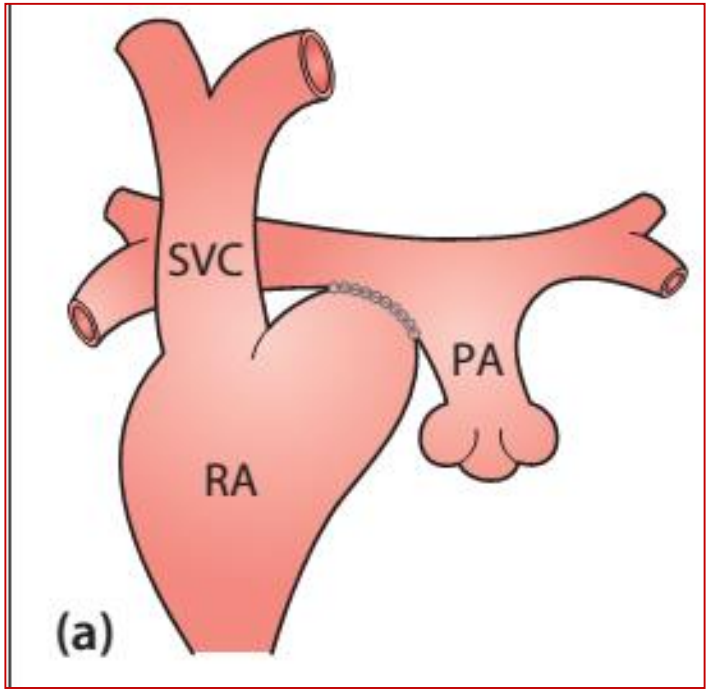
- Proximal insertion stenosis
- PA dilation
- Collateral formation (usually via azygous dilatation)

Intervento di Glenn

Adattato da: ESC/EACVI CHD/CMR guide, 2014

Classic Fontan (atriopulmonary)

L’atrio destro è connesso all’arteria polmonare grazie ad una anastomosi anteriore o posteriore



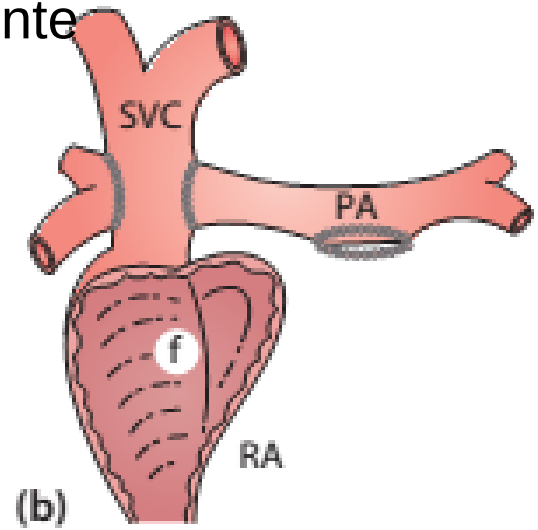
Classic Fontan (atriopulmonary)

Post-operative complications

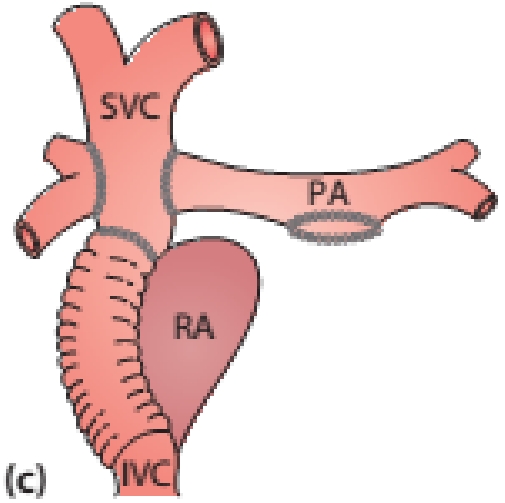
- Stenosis to systemic venous pathways
- Ascites (due to protein losing enteropathy)
- Deterioration of ventricular function
- Massively dilated RA
- Thrombus risk particularly high
- Pulmonary venous compression
- AV valve regurgitation
- No Glenn shunt as direct communication

Modified Fontan

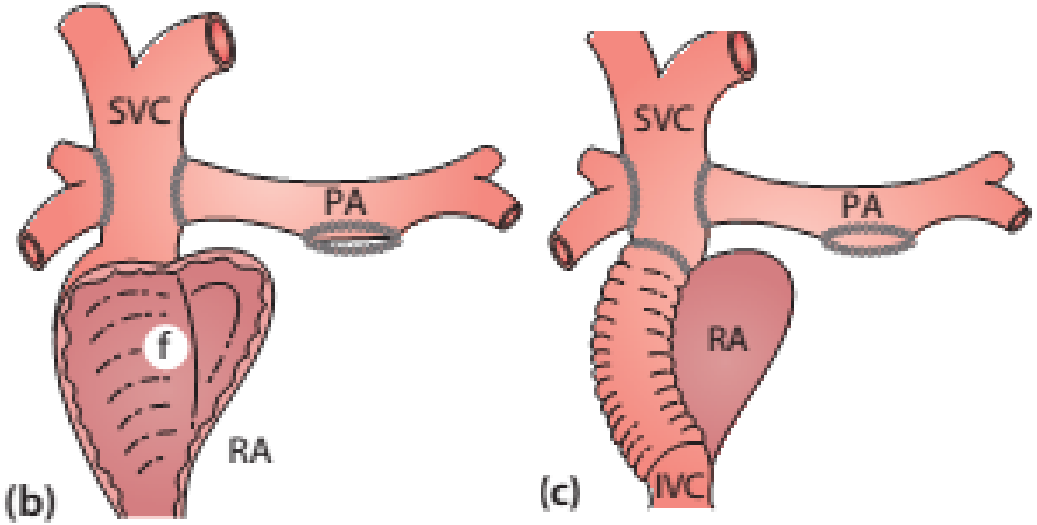
- Condotto intra-extra-atriale (eventualmente valvolato)



- Total cavopulmonary connection (le vene polmonari si connettono alla APDx; la VCS direttamente, la VCI con un tunnel intra- o extra-atriale)



Modified Fontan



Initial interventions

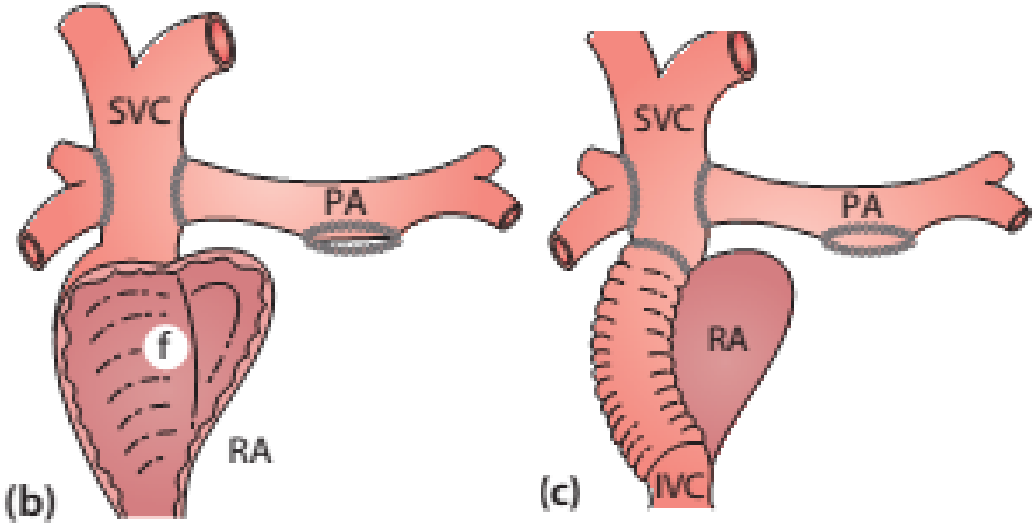
- Preceded by Glenn shunt or BT shunt

Late interventions

- Occasional fenestration closure
- Occlusion of systemic to pulmonary venous collaterals

CARDIOPATIE CONGENITE: s/p Fontan

Modified Fontan



Post-operative complications

- Stenosis to systemic venous pathways
- Ascites (due to protein losing enteropathy)
- Deterioration of ventricular function
- Thrombus always possible
- Pulmonary venous compression
- AV valve regurgitation

CARDIOPATIE CONGENITE: s/p Fontan

7 Fontan: Aspetti fisiopatologici peculiari

1. Bassa portata cronica (un singolo ventricolo per tutto il circolo)
2. Peggioramento della funzione ventricolare > aumento PTD e PAP
3. Insufficienza della valvola AV > aritmie atriali
4. Ostruzione ritorno venoso sistemico > aumento afterload > ascite, enteropatia proteinodisperdente, insufficienza ventricolare
5. Compressione della vena polmonare destra ad opera dell'atrio destro (Fontan AP) > aumento pressioni venose polmonari e in AP
6. Formazione di trombi

CARDIOPATIE CONGENITE: s/p Fontan

Fontan atriopolmonare

TCPC con condotto laterale

TCPC con condotto extracardiaco

CARDIOPATIE CONGENITE: s/p Fontan

Standard imaging

1. Cine CMR: axial stack from the mid-liver to the top of the aortic arch
2. Cine CMR: coronal or oblique stack to image the Fontan baffle in long-axis
3. Ventriculography module
4. CE-MRA or 3D SSFP to image the thoracic vasculature
5. PC CMR: AAo, native MPA, branch PAs, SVC, IVC

Additional case-specific/comprehensive imaging

1. Cine CMR: long-axis aortic arch plane
2. LGE module
3. PC CMR: AV valve, pulmonary veins

Key reporting elements: Fontan pathway, SVC, IVC, branch PA, pulmonary vein (compressed?), and aortic arch obstruction; Fontan baffle defects; presence of thrombus; ventricular parameters; valve regurgitation; aortopulmonary collaterals

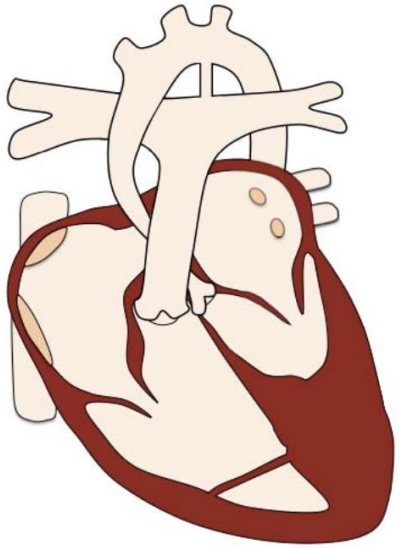
CARDIOPATIE CONGENITE: s/p Fontan

Dilatazione atriale destra con trombosi atriale

CARDIOPATIE CONGENITE: s/p Fontan

Dilatazione atriale destra con compressione vena polmonare inferiore destra

CARDIOPATIE CONGENITE: Hypoplastic Left Heart



Pre-operative findings

- Marked hypoplasia of the LV and ascending aorta
- AV and MV are atretic, hypoplastic, or stenotic
- PDA and / or ASD
- Double outlet RV in 25%

Associated findings

- Aortic coarctation

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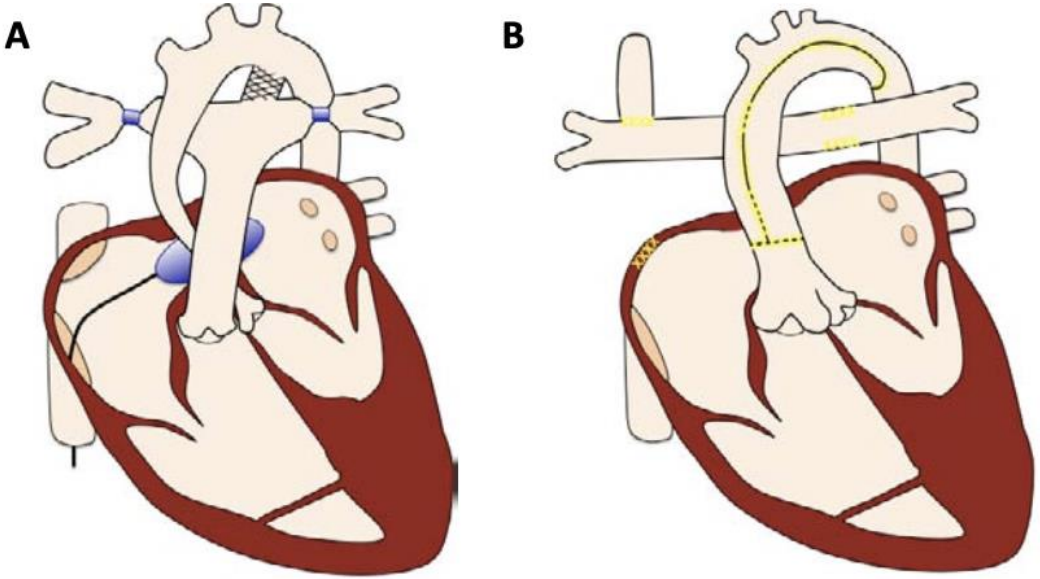
CARDIOPATIE CONGENITE: Hypoplastic Left Heart



Hybrid procedure (A)

- PDA stent, atrial septostomy, pulmonary banding

Norwood procedure (B)

Stage 1:
- MPA used to augment aorta
- RV utilised as a systemic ventricle
- PA to ascAo anastomosis to supply coronary circulation
- BT shunt or RV to PA shunt to supply PAs
- Atrial septectomy

Stage 2: Glenn procedure

Stage 3: Fontan completion

Damus-Kaye-Stancel anastomosis (augmented neo-aorta

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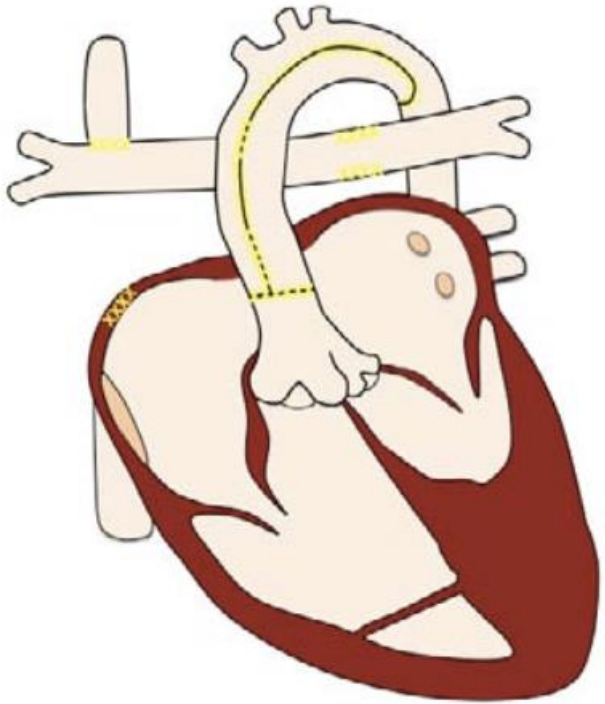
CARDIOPATIE CONGENITE: s/p Norwood

Norwood stadio I: Sano shunt

CARDIOPATIE CONGENITE: s/p Norwood



Complicanze post-operatorie:

Come per s/p Fontan, e inoltre

1. Ricoartazione aortica
2. Restringimento/chiusura DIA
3. Stenosi Apsn
4. Insufficienza del Vdx sistemico
5. Insufficienza tricuspidalica
6. Ischemia coronarica

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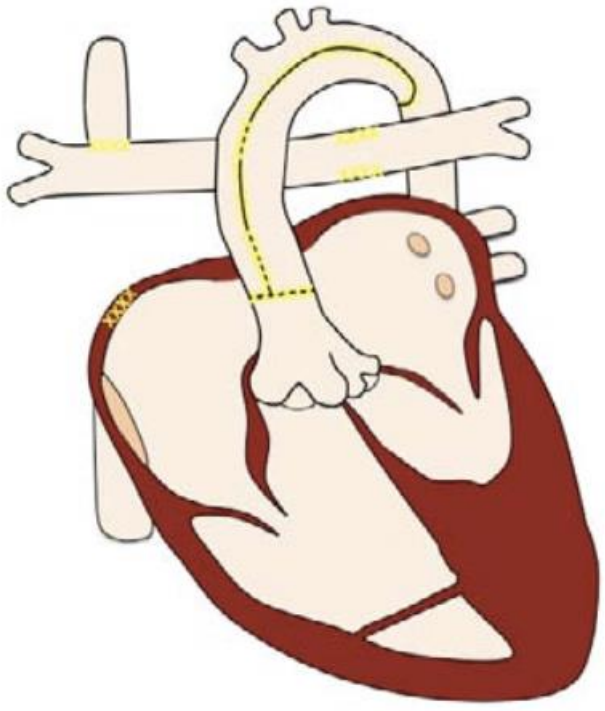
CARDIOPATIE CONGENITE: s/p Norwood



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Fisiopatologia:

Come per s/p Fontan, e inoltre

1. Ricoartazione o dilatazione neoaorta
2. Stenosi dei tronchi sovraaortici
3. Le coronarie originano dall'aorta originaria, ipoplastica, e anastomizzata alla neo aorta
4. Tricuspide > valvola AV sistemica > IT probabile
5. Vdx sistemico e plurimi interventi > prognosi peggiore a lungo termine

CARDIOPATIE CONGENITE: s/p Norwood

Standard imaging

1. Cine CMR: axial stack from the mid-liver to the top of the aortic arch
2. Ventriculography module
3. CE-MRA or 3D SSFP to image the thoracic vasculature and surgical shunts
4. PC CMR: AAo, native MPA, branch PAs

Additional case-specific/comprehensive imaging

1. LGE module
2. Cine CMR: long-axis aortic arch plane
3. Spin echo: axial plane to image the branch PAs and aortopulmonary shunt
4. PC CMR: SVC, IVC, tricuspid and mitral valves,
5. DAo at the level of the diaphragm, pulmonary veins

Key reporting elements: shunt, branch PA, pulmonary vein, and aortic arch obstruction; ventricular parameters; valve regurgitation; aortopulmonary collaterals; venous collaterals

Grazie per l'attenzione

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