IRM cardiaque et les cardiopathies congénitales

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Cours DU - Cardiologie pédiatrique
M3C- Necker-Enfant Malades

IRM cardiaque et les cardiopathies congénitales

IRM et Scanner

Scanner
-Irradiant
-Examen court
-Haute disponibilité

IRM
-Non irradiant
-Examen long
-Disponibilité limitée

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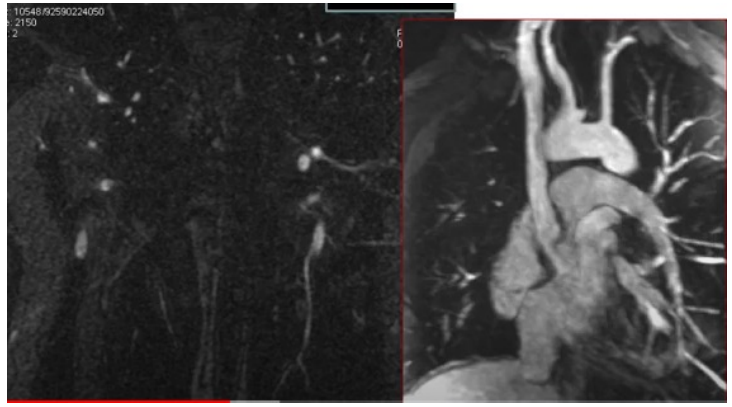
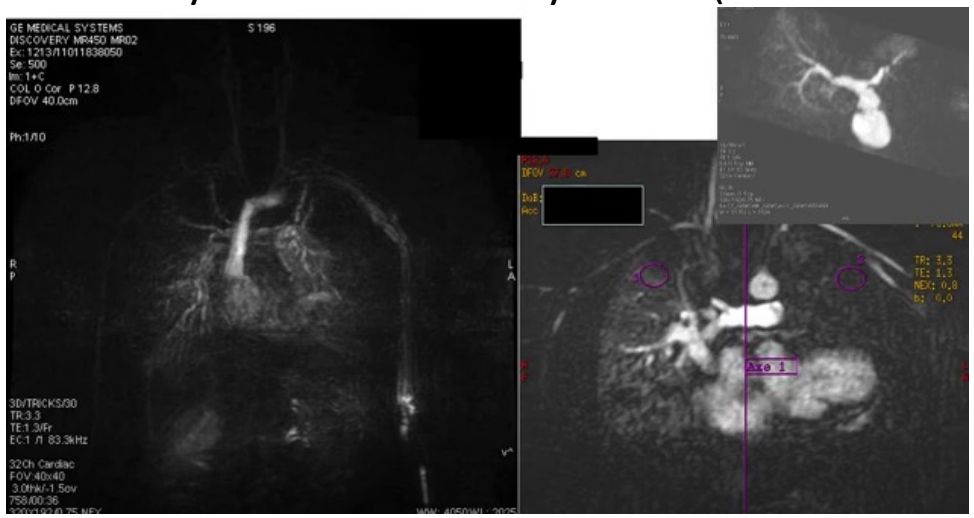
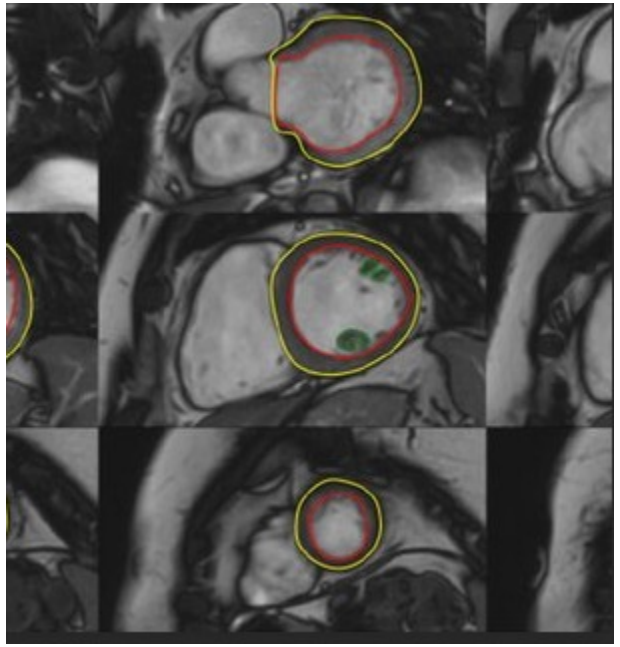
IRM cardiaque: indications

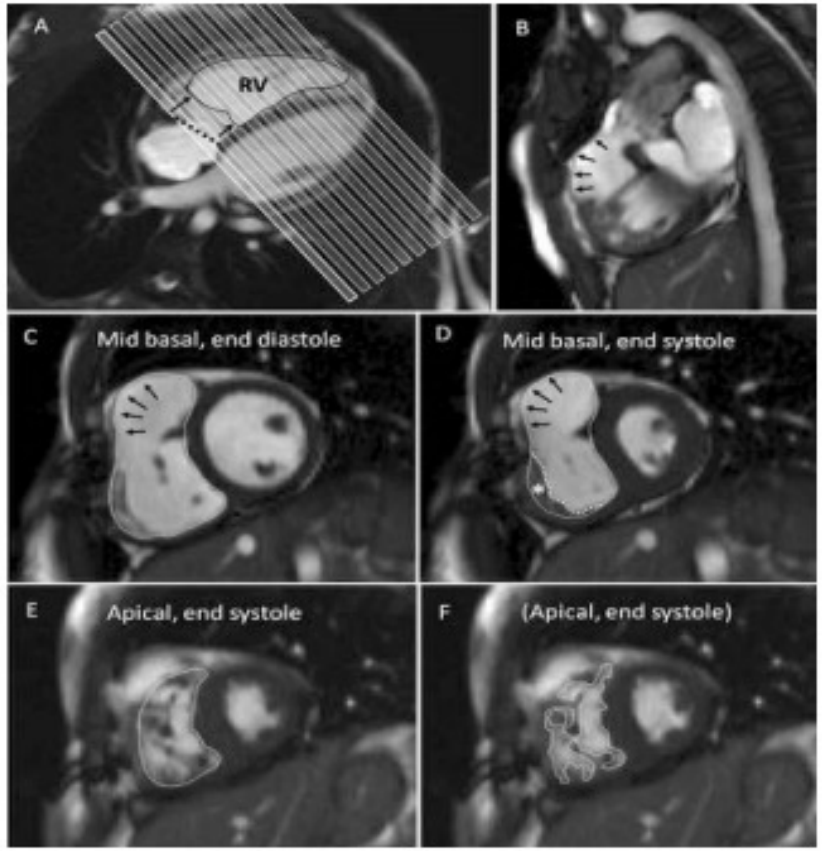


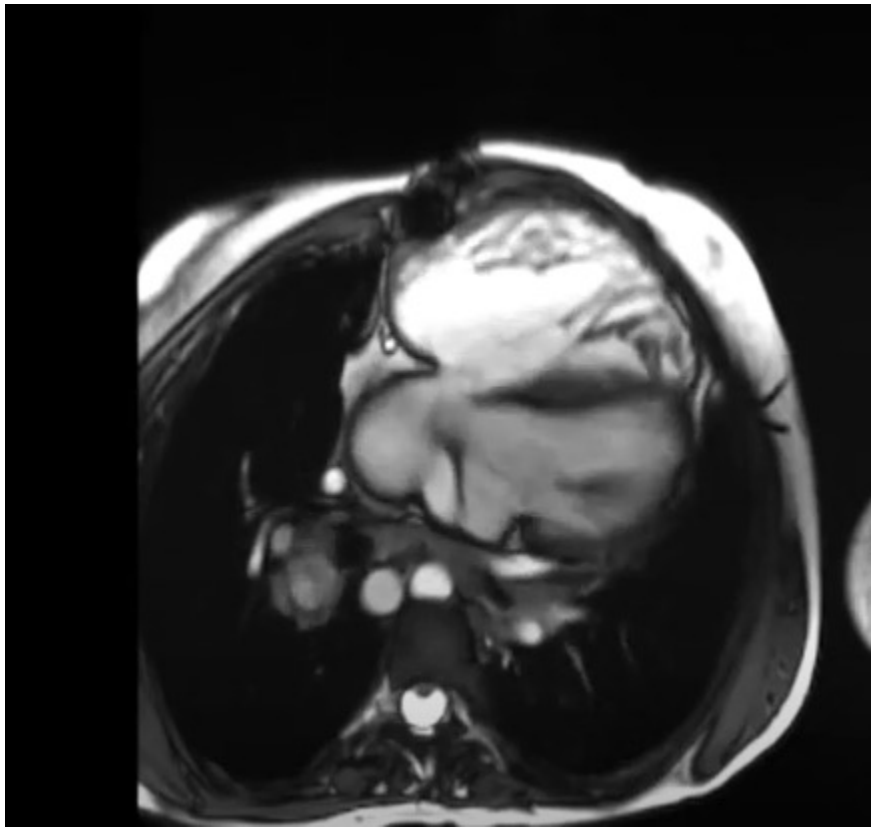
1. Morphologie et anatomie intracardiaque et extracardiaque
2. Fonction, masse
3. Fonction des Valves cardiaques
4. Masse intra-extracardiaque
5. Perfusion pulmonaire
6. Perfusion myocardique/réserve coronaire/réserve contractile
7. Analyse tissulaire du myocarde (inflammation/fibrose)



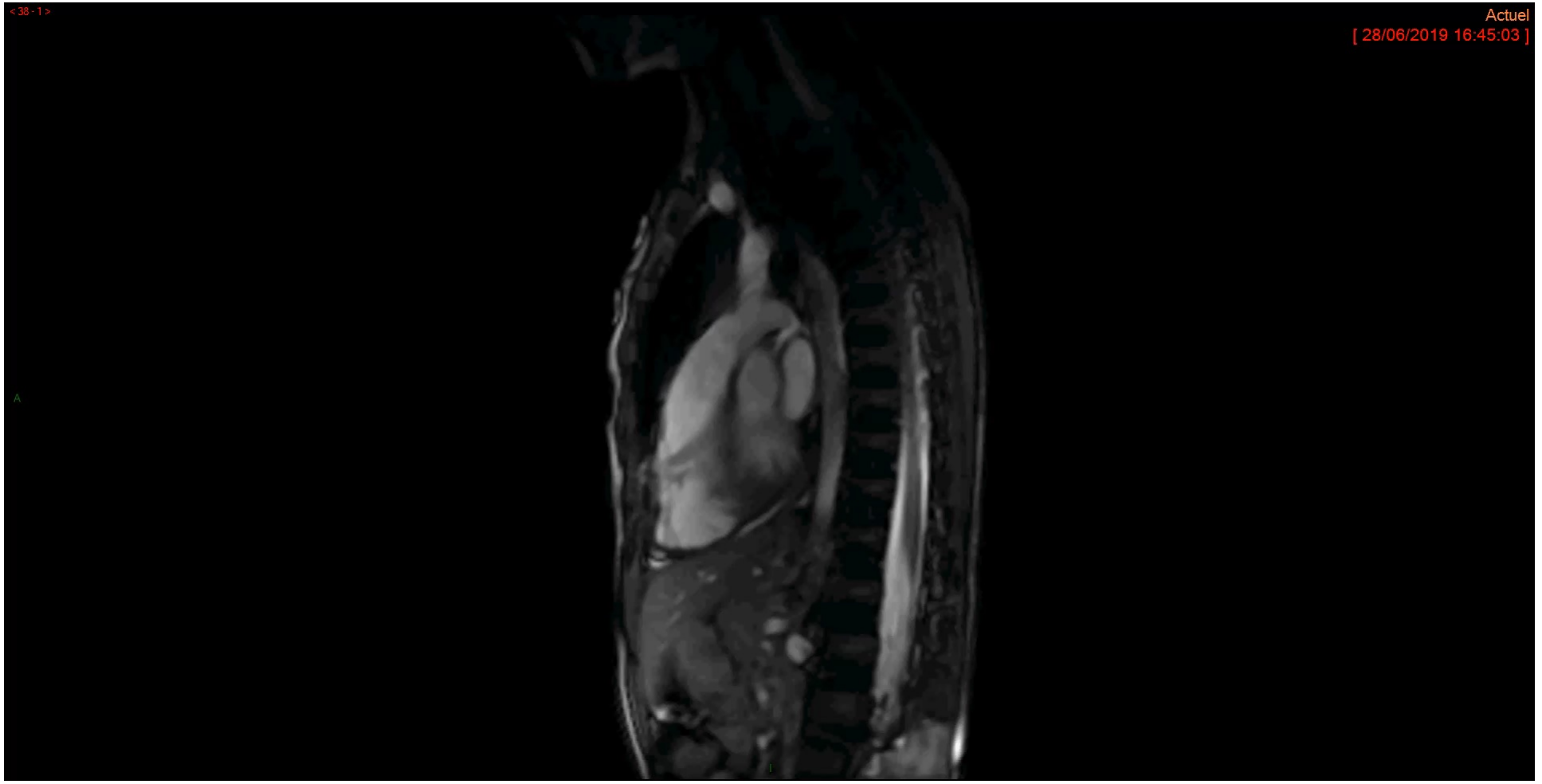
Volumes/Fonction: *Ventricule droit*



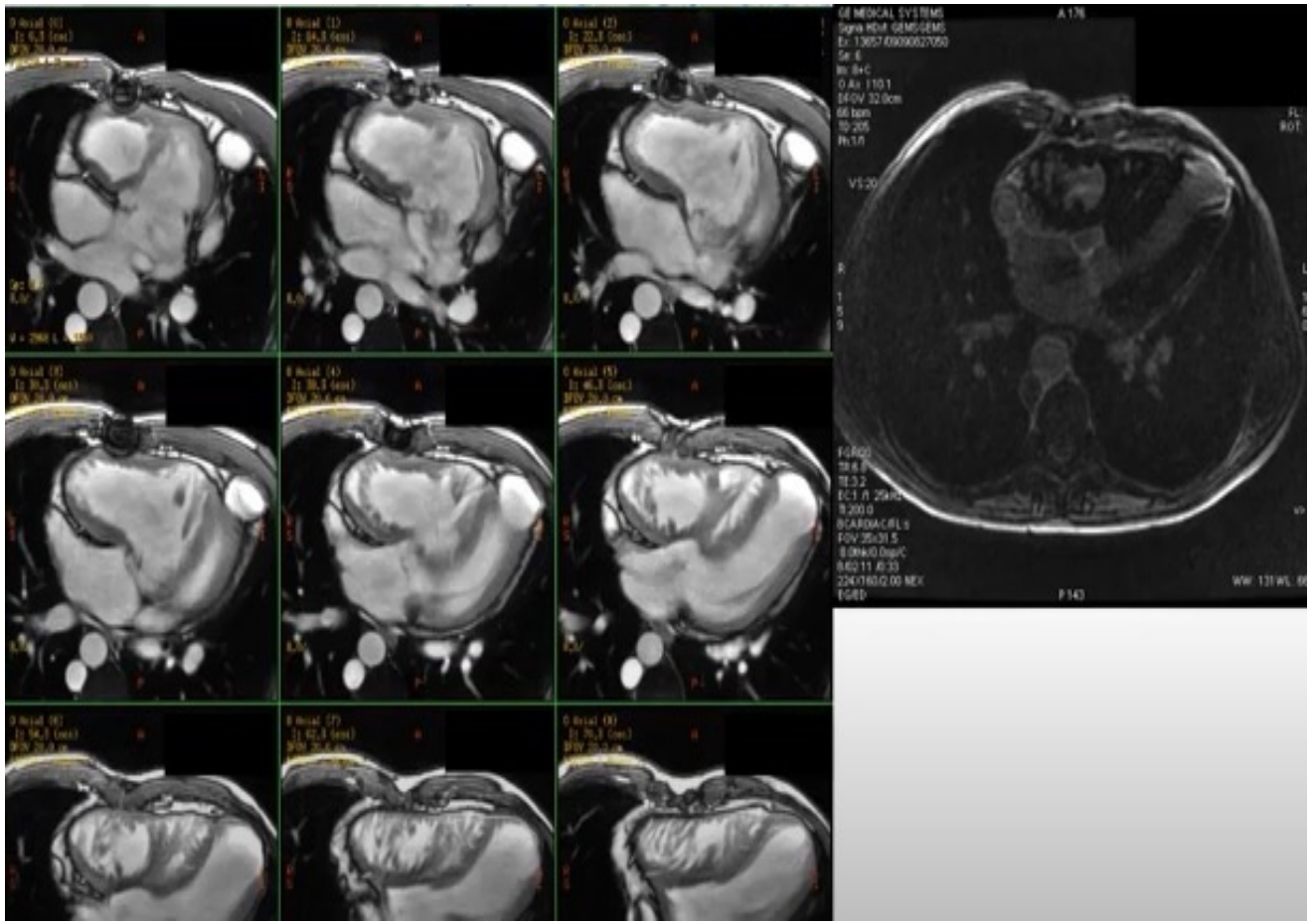


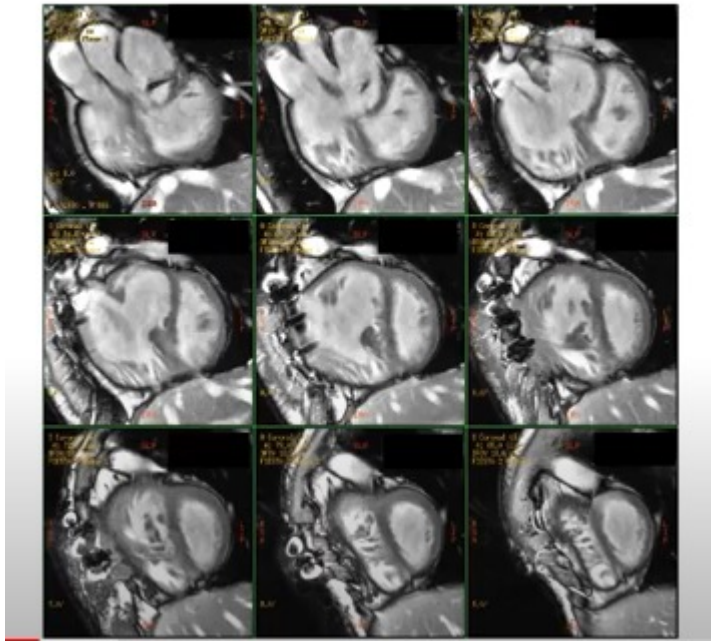


Volumes/Fonction: *Ventricule droit*



Volumes/Fonction: *Cardiopathie Complexe*





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Ventricule droit et DVDA

By MRI:

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
  - Ratio of RV end-diastolic volume to BSA ≥110 mL/m² (male) or ≥100 mL/m² (female)
  - *or* RV ejection fraction ≤40%

By MRI:

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
  - Ratio of RV end-diastolic volume to BSA ≥100 to <110 mL/m² (male) or ≥90 to <100 mL/m² (female)
  - *or* RV ejection fraction >40% to ≤45%

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Analyse tissulaire

Schultz JS, et al. Mayo Clin Proc. 2009 Analyse tissulaire Acute myocarditis Cardiovascular Magnetic Resonance

Native T_1 ↑	Native T_1 ↓
• *Oedema* e.g. tissue water↑ in acute MI, inflammation, pericardial effusion	• *Fat/Lipid overload* e.g. lipomatous metaplasia in chronic MI, Anderson-Fabry, lipoma
• *Increase of interstitial space* *e.g.* (replacement) fibrosis, scar, cardiomyopathy, amyloid deposition	• *Iron overload*

T_2 ↑	T_2 ↓
• *Oedema* e.g. tissue water↑ in acute MI, inflammation, pericardial effusion (T_2 more sensitive than native T_1 for oedema detection)	• *Fat/Lipid overload* • *Iron overload* (T_2*↓)

Edema

• T2-weighted image sensitivity detects tissue edema with the **long T2 of water bound protons as the contrast-generating mechanism**, resulting in a high signal intensity of edematous tissue.

Hyperemia and capillary leak (myocardial early gadolinium enhancement)

• Contrast-enhanced fast spin-echo T1-weighted MR is used to assess myocardial hyperemia and inflammation.

Necrosis and fibrosis (late gadolinium enhancement [LGE])

• Myocardial LGE reflects irreversible myocardial injury (i.e., necrosis and fibrosis

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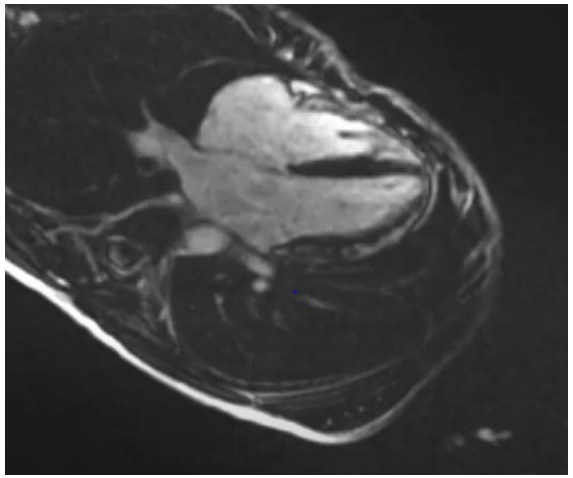
Myocardite



CMR Criteria of Myocardial Inflammation

Diagnostic Target		Original Lake Louise Criteria (Any 2 out of 3)	Updated Lake Louise Criteria (Any 2 out of 2)
Myocardial Edema		T2-weighted imaging Signal intensity↑	T2-weighted imaging Signal intensity↑ Relaxation time↑
Myocardial Injury	Hyperemia (Intra/extra cellular edema, capillary leak)	Early Gadolinium Enhancement	T1-weighted imaging Native (non-contrast) relaxation time ↑ Extracellular volume↑ Non-ischemic pattern LGE
	Myocardial Necrosis, scar	Late Gadolinium Enhancement	
Supportive Criteria		Pericardial effusion Systolic LV wall motion abnormality	Pericardial effusion High signal intensity of pericardium in LGE, T1, T2 mapping Systolic LV wall motion abnormality

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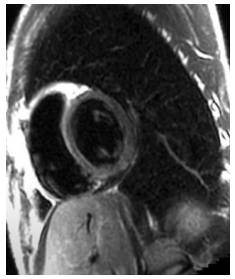
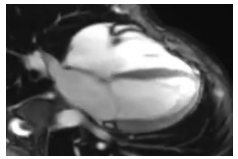
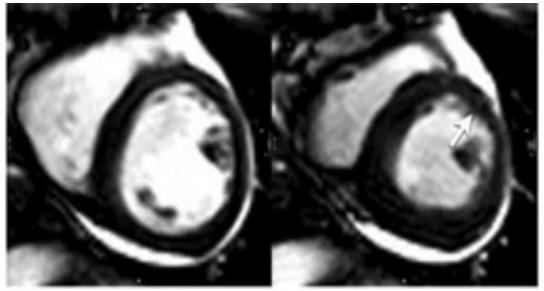
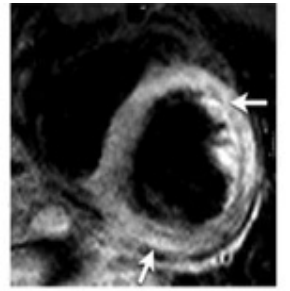
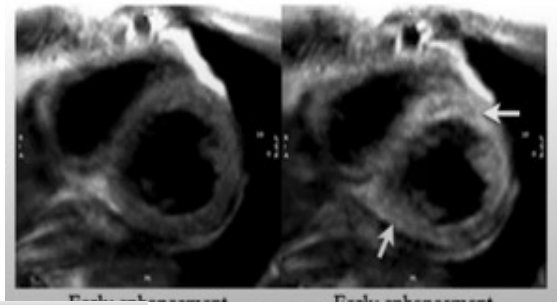
Myocardite

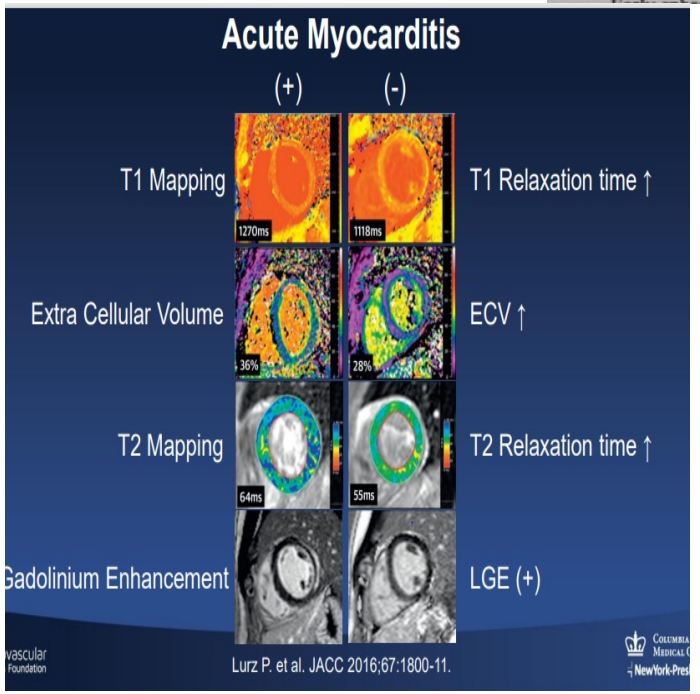












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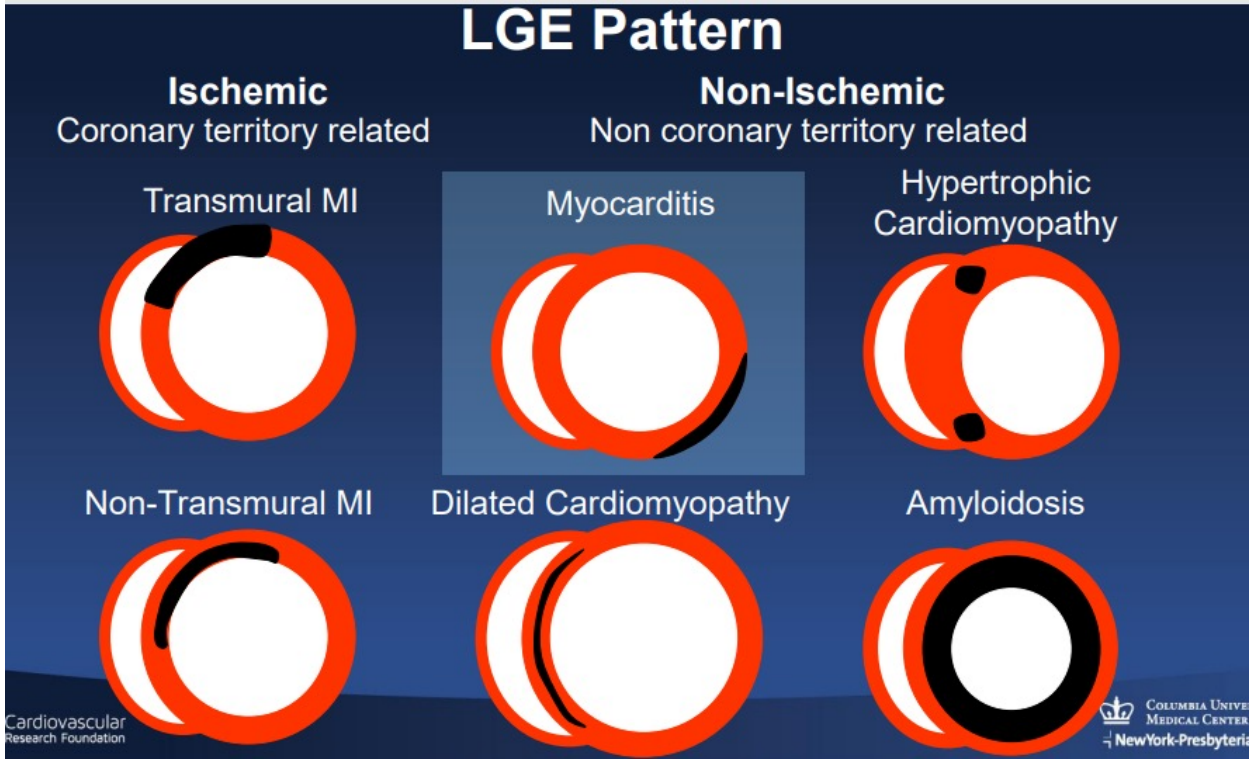
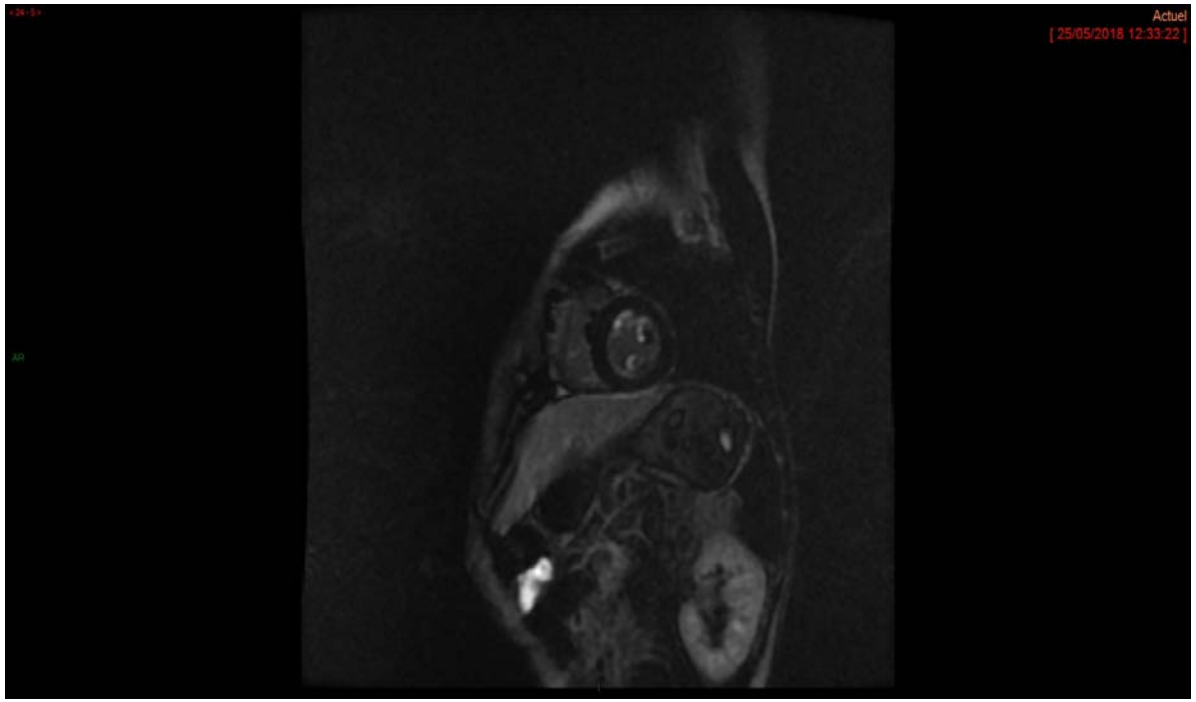
Rehaussement tardif

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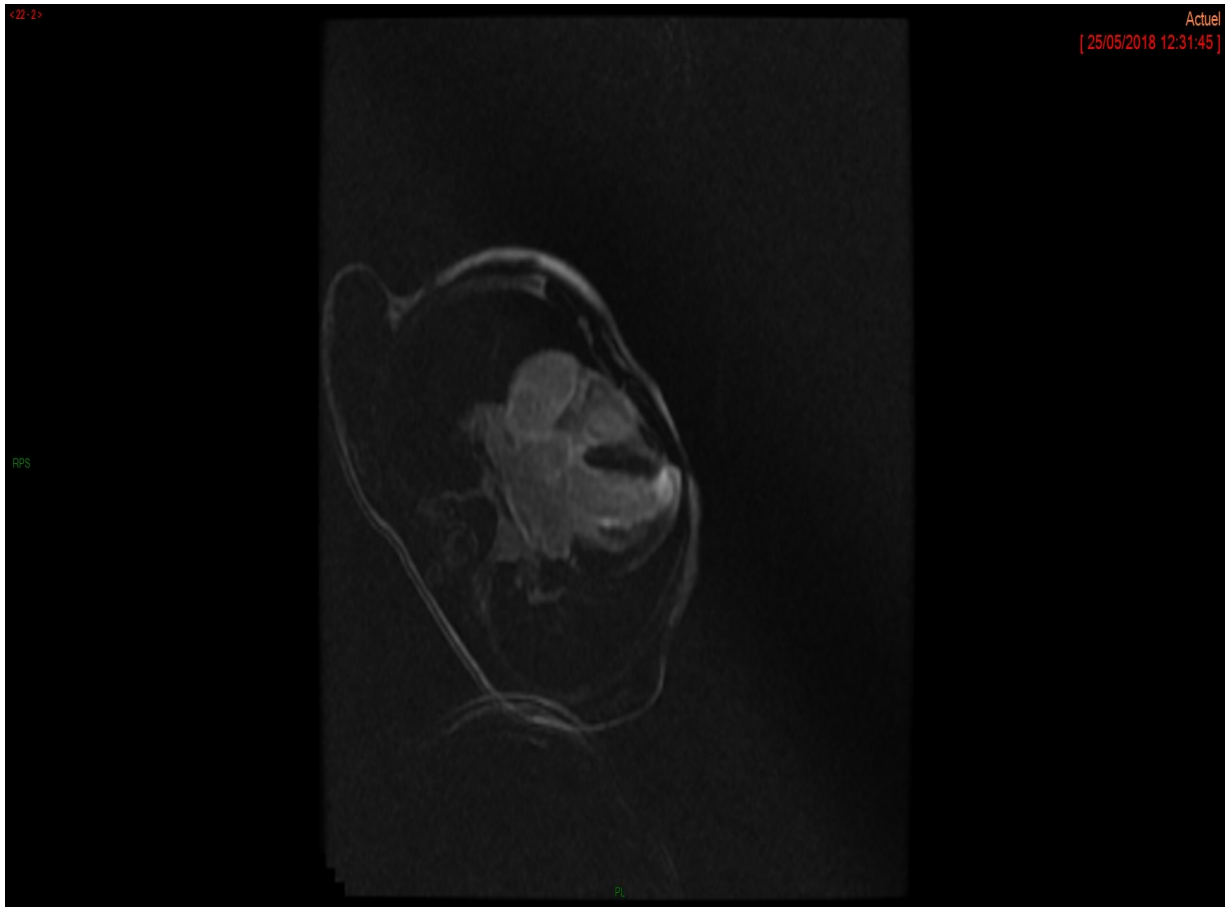
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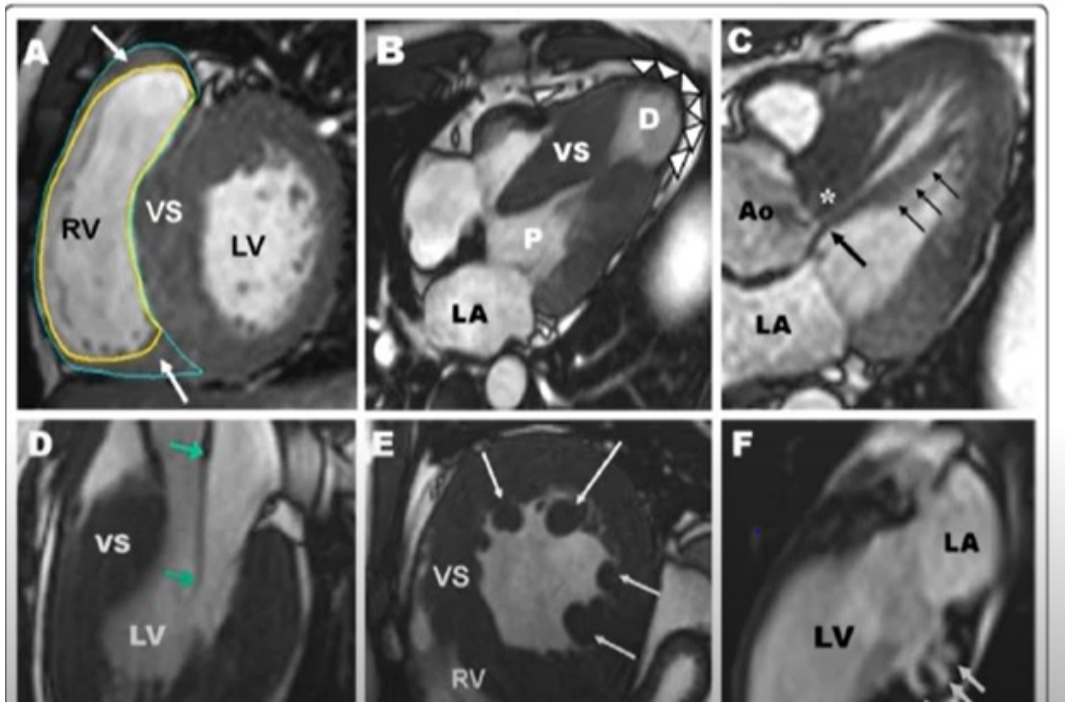
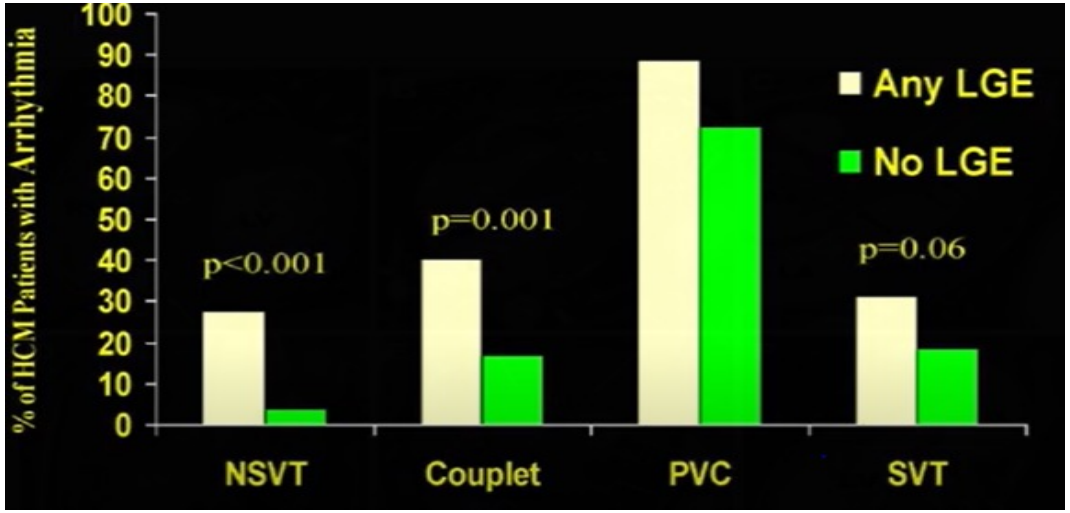
Cardiomyopathie hypertrophique





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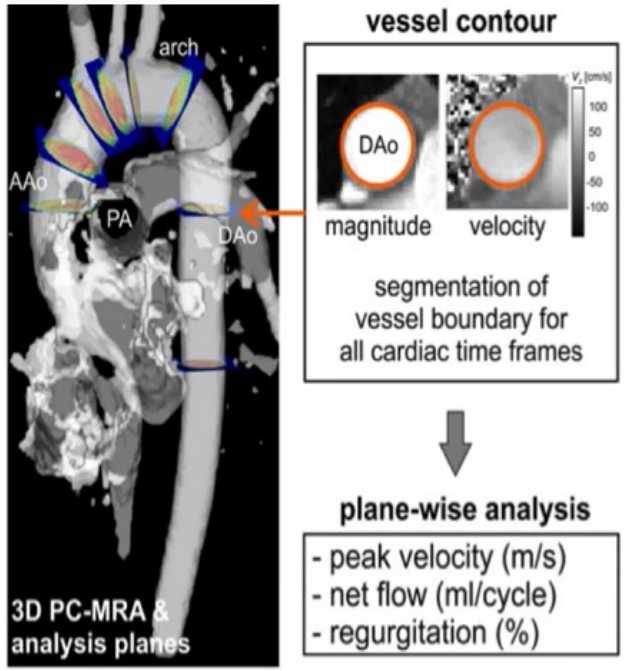
4DFlow



Phase contrast CMR with flow encoding in all 3 spatial directions that is resolved relative to all 3 dimension of space and of time along the cardiac cycle (3D+time= 4D)

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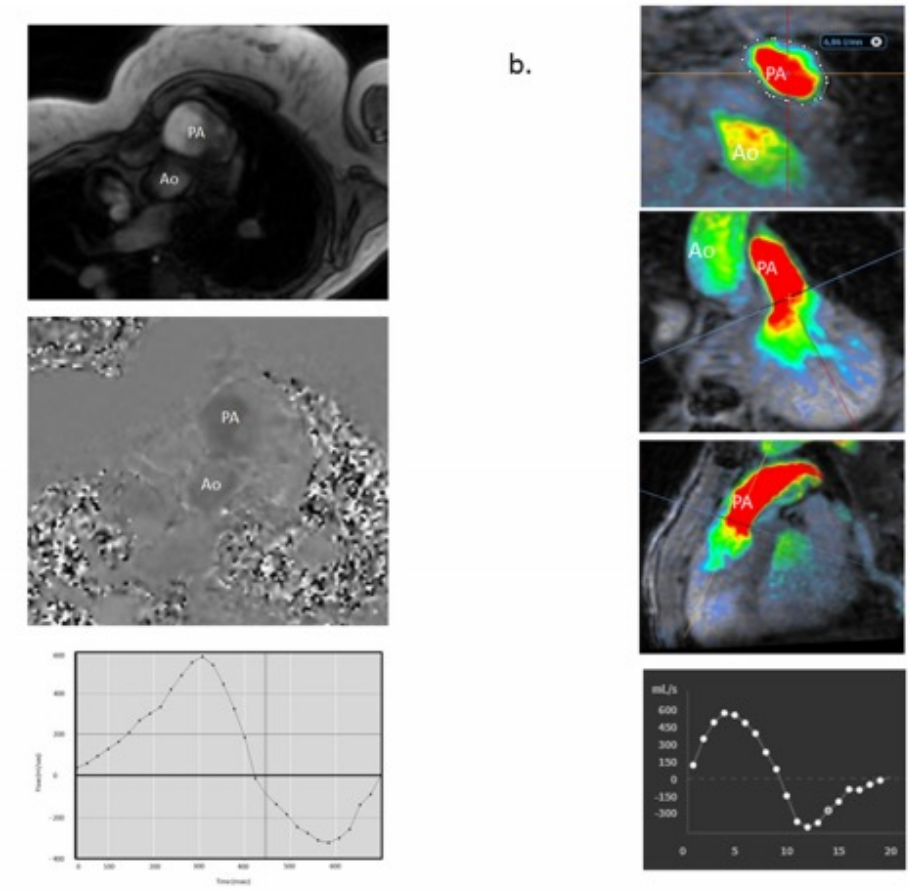
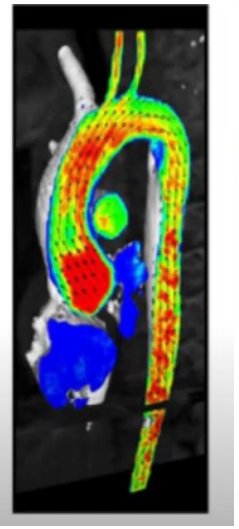
Phase contrast vs 4dflow



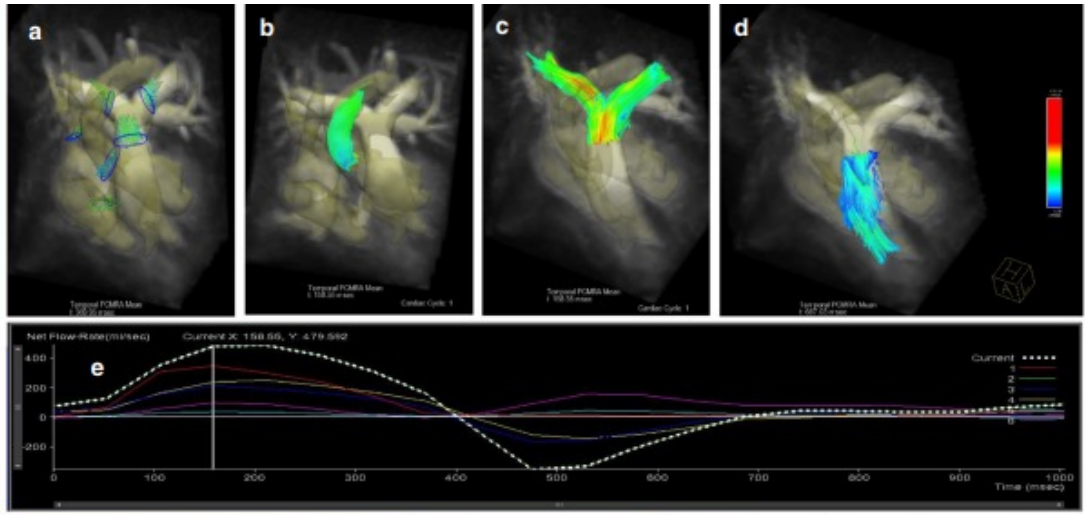
Isorni MA, Martins D, Ben Moussa N, Monnot S, Boddaert N, Bonnet D, Hascoet S, Raimondi F. 4D flow MRI versus conventional 2D for measuring pulmonary flow after Tetralogy of Fallot repair. Int J Cardiol. 2020 Feb 1;300:132-136

4Dflow

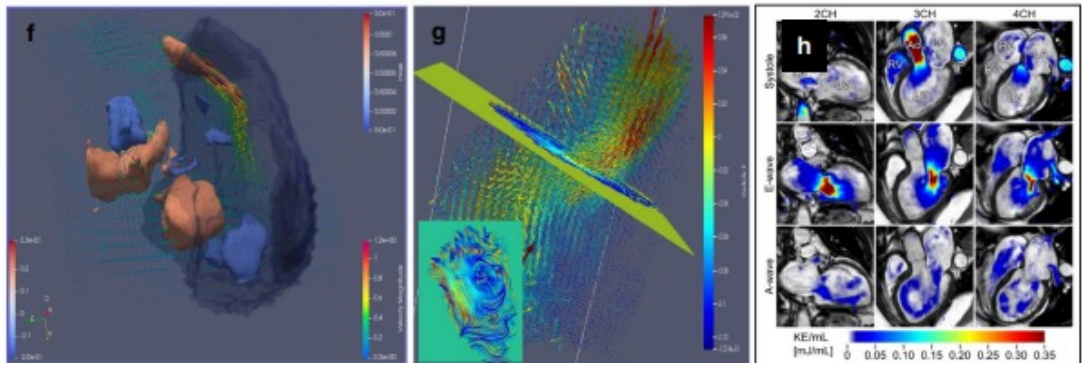
Vectors



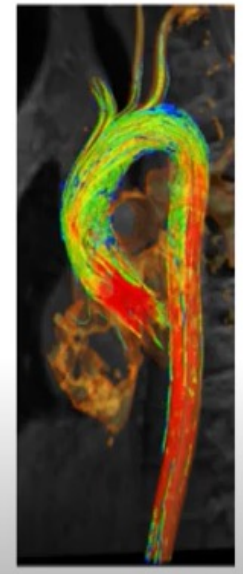
Flow analysis



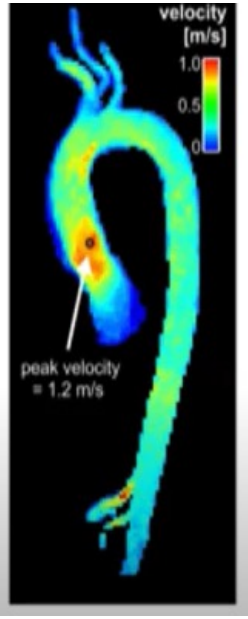
Novel applications



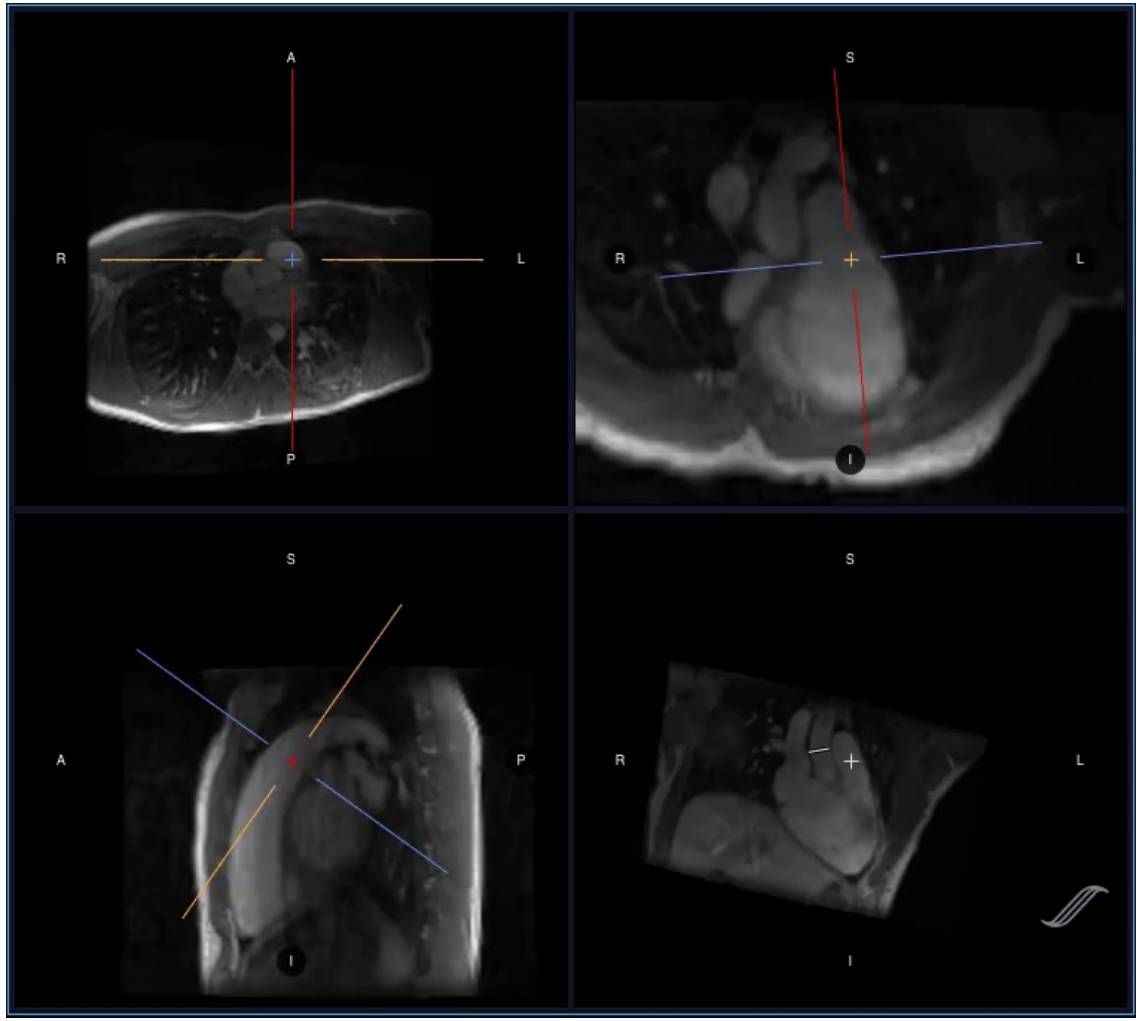
Streamlines



Velocities

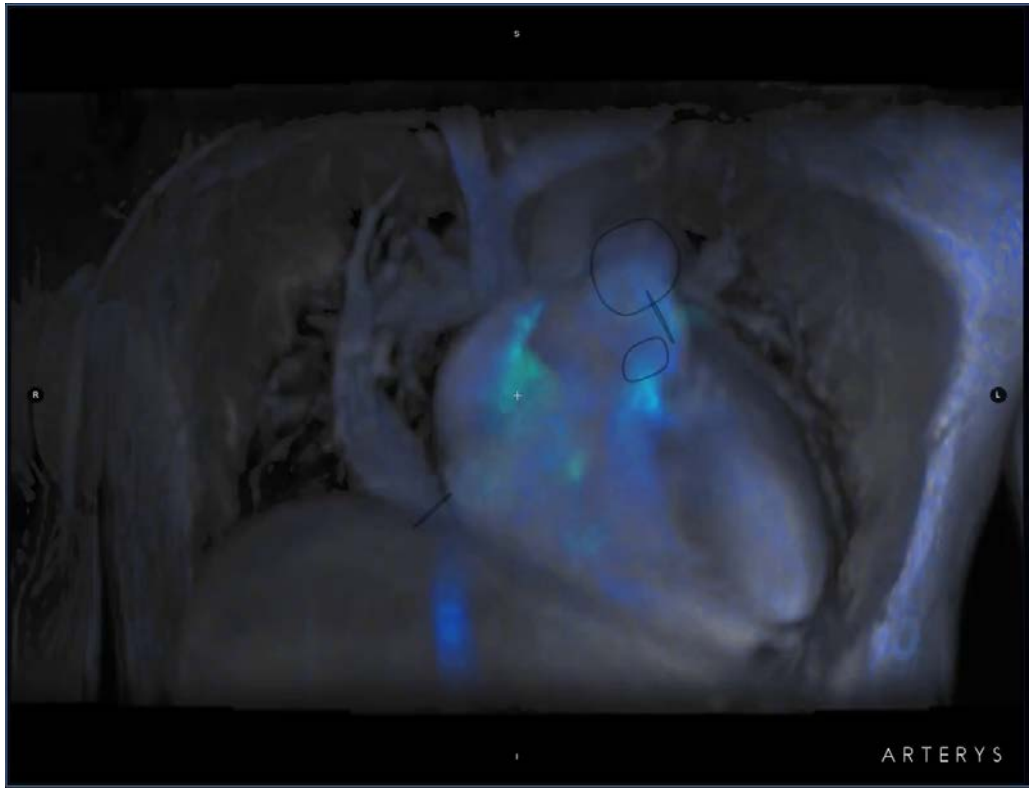


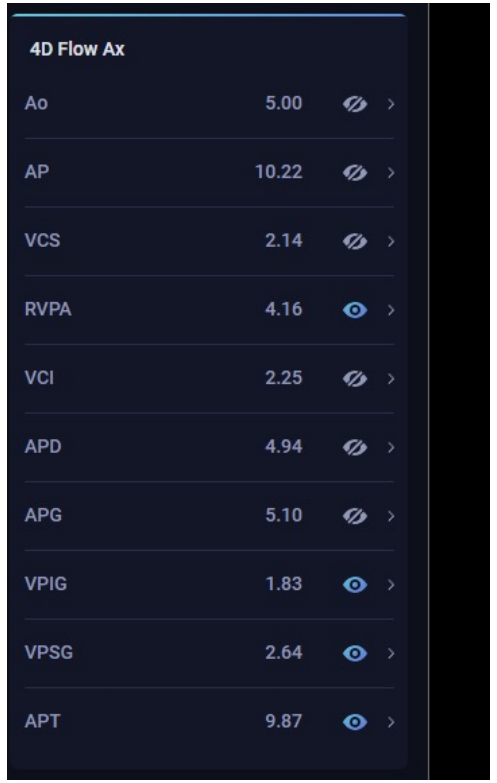
4Dflow



ARTERYS

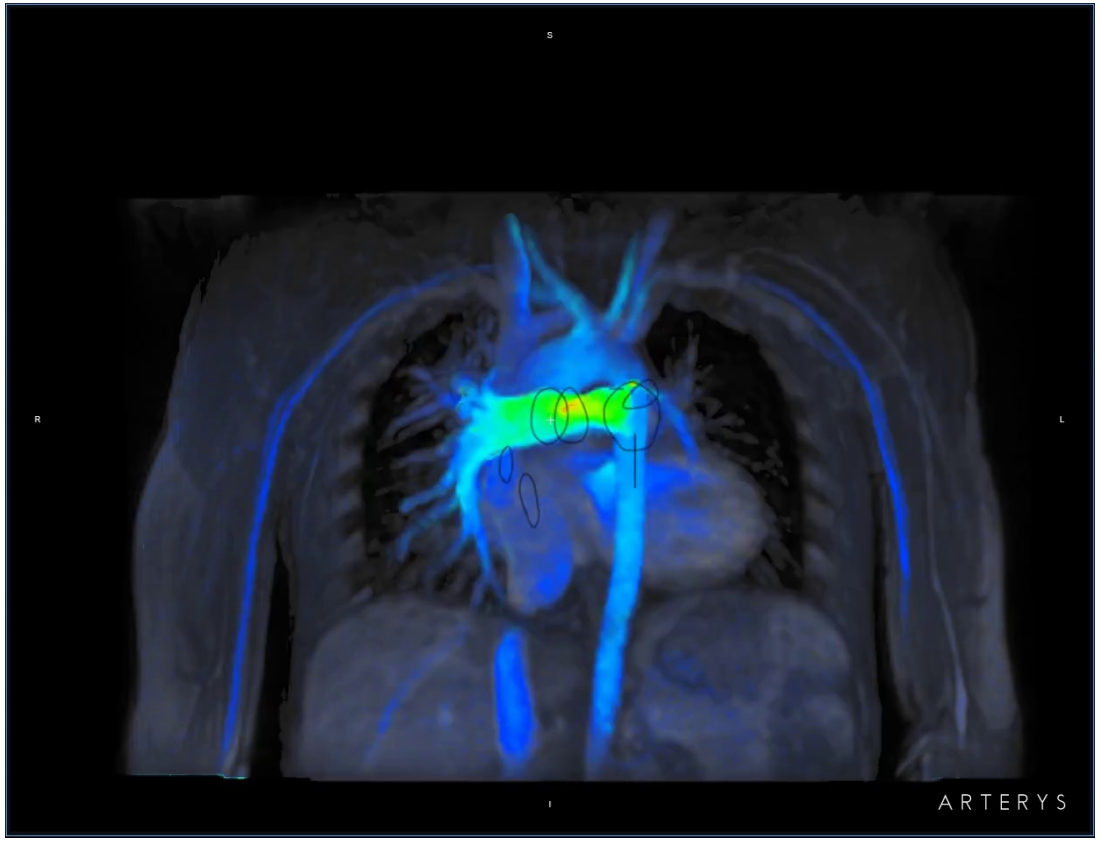
4Dflow exemples

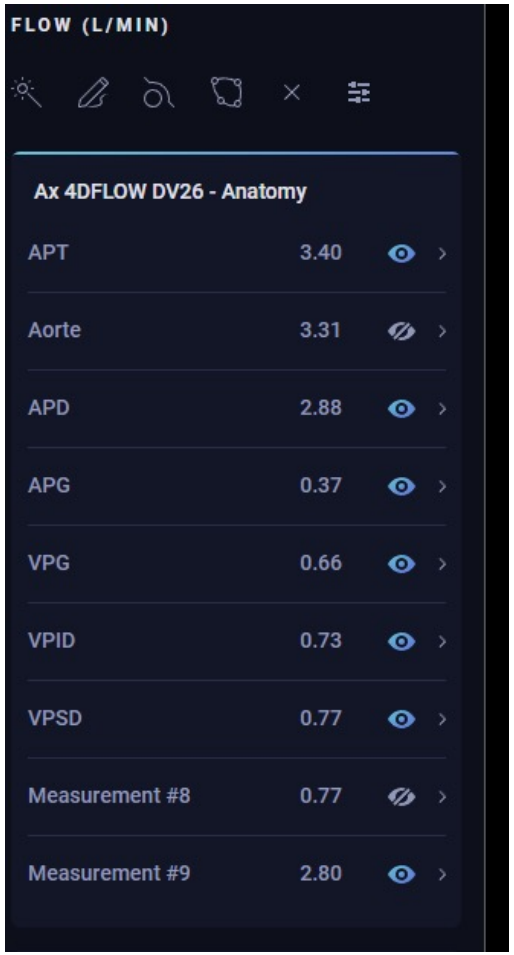




ARTERYS

4Dflow exemples





IRM cardiaque et les cardiopathies congénitales

Indications

Indications for CMR in ACHD patients

- Quantification of RV volumes, EF (including subpulmonary RV, systemic RV, and single ventricle)
- Evaluation of RVOTO and RV–PA conduits
- Quantification of PR
- Evaluation of PAs (stenoses, aneurysms) and the aorta [aneurysm, dissection, coarctation (CCT may be superior)]
- Evaluation of systemic and pulmonary veins (anomalous connection, obstruction, coronary venous anatomy pre-procedure, etc.)
- Collaterals and arteriovenous malformations (CCT may be superior)
- Coronary anomalies and CAD (CCT is superior for intramural course, slit-like course, acute angle take-off, myocardial bridging, and plaque assessment)
- Detection and quantification of myocardial ischaemia by CMR stress perfusion
- Evaluation of intra- and extracardiac masses
- Quantification of myocardial mass (LV and RV)
- Detection and quantification of myocardial fibrosis/scar (late gadolinium enhancement, T1 mapping) tissue characterization (fibrosis, fat, iron, etc.)
- Quantification of systemic and pulmonary blood flow to calculate Qp:Qs
- Quantification of perfusion distribution to the right/left lung
- Measurement of pulmonary blood flow in patients with multiple sources of blood supply (i.e. with major aorto-pulmonary collateral arteries)

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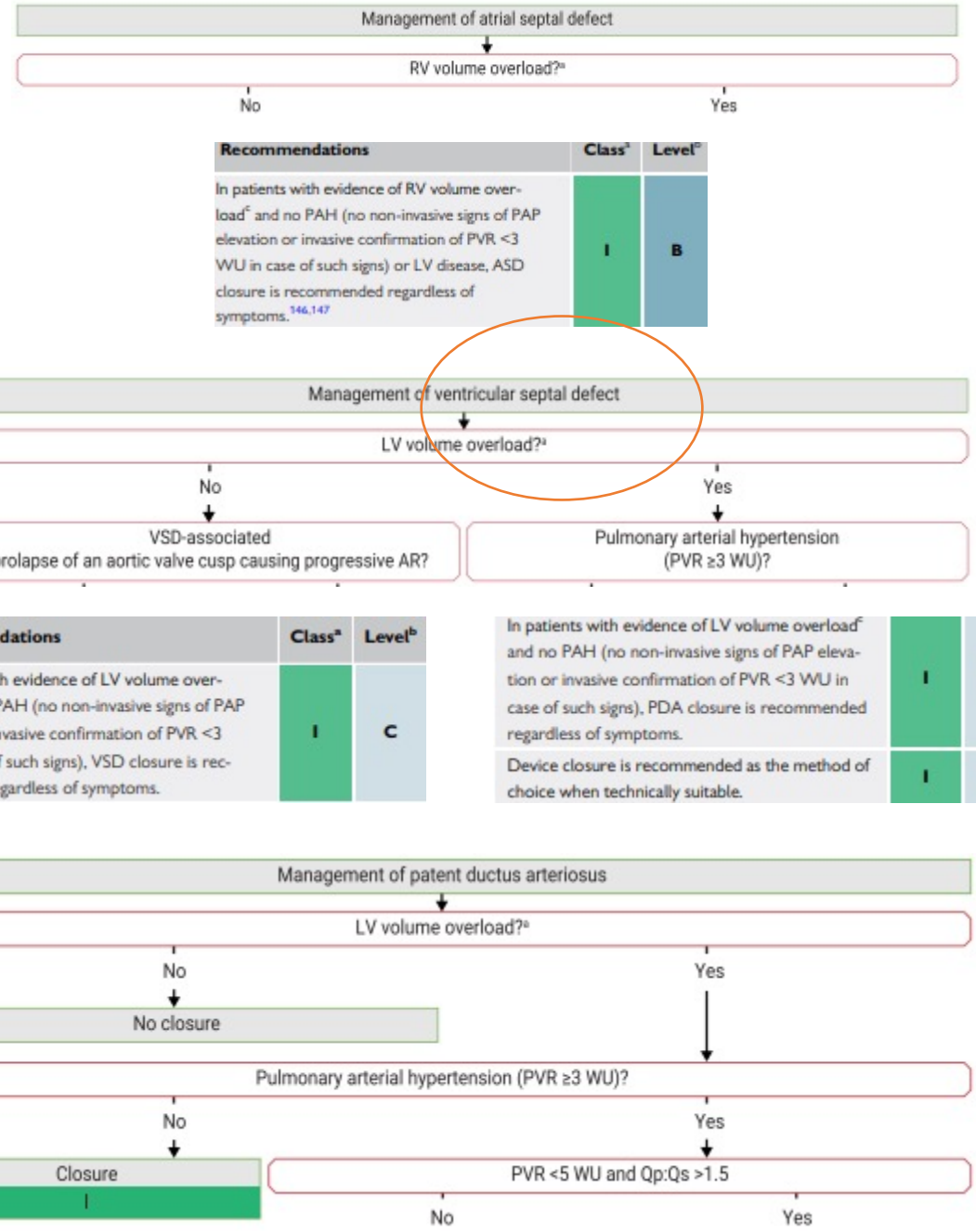
Quantification of systemic and pulmonary blood flow to calculate Qp:Qs

CIA, CIV et CAV



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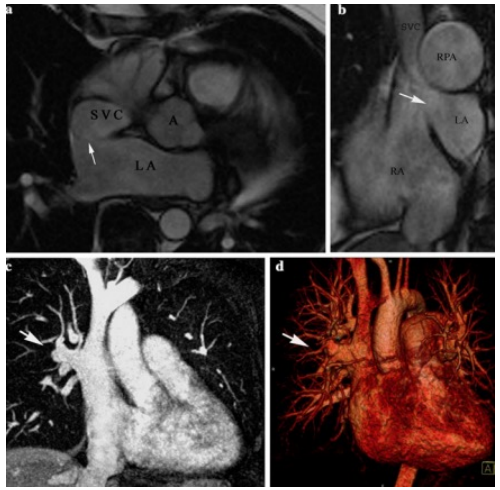
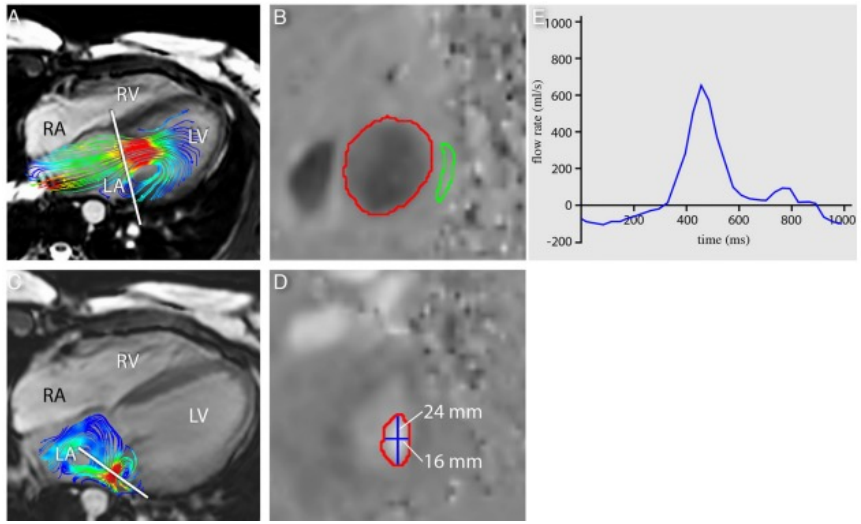


Prompona M, et al . MRI for detection of anomalous pulmonary venous drainage in patients with sinus venosus atrial septal defects. Int J Cardiovasc Imaging. 2011 Mar;27(3):403-12.



Calkoen EE, et al Characterization and quantification of dynamic eccentric regurgitation of the left atrioventricular valve after atrioventricular septal defect correction with 4D Flow cardiovascular magnetic resonance and retrospective valve tracking. J Cardiovasc Magn Reson. 2015 Feb 19;17(1):18.

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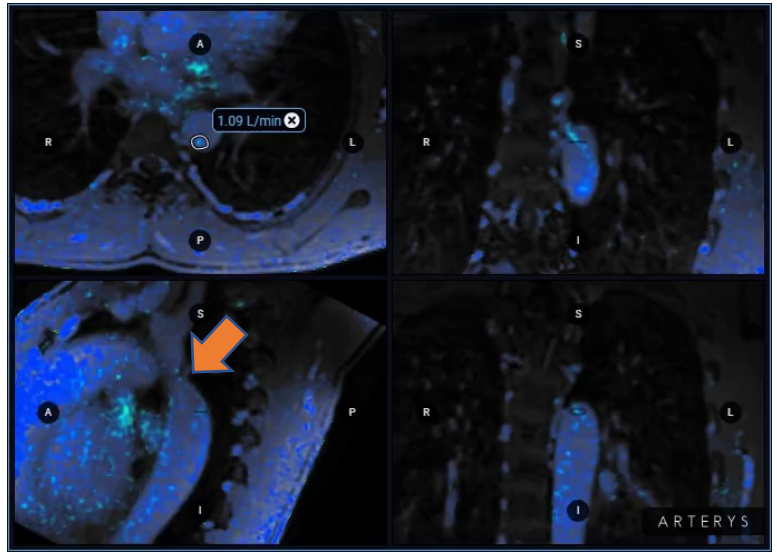
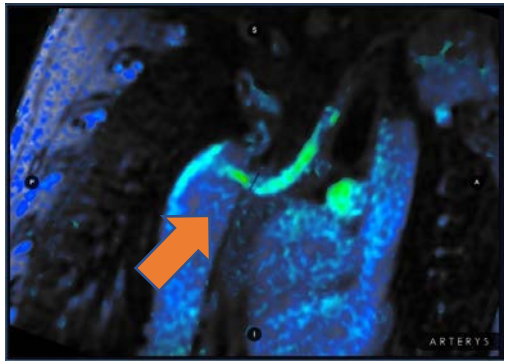
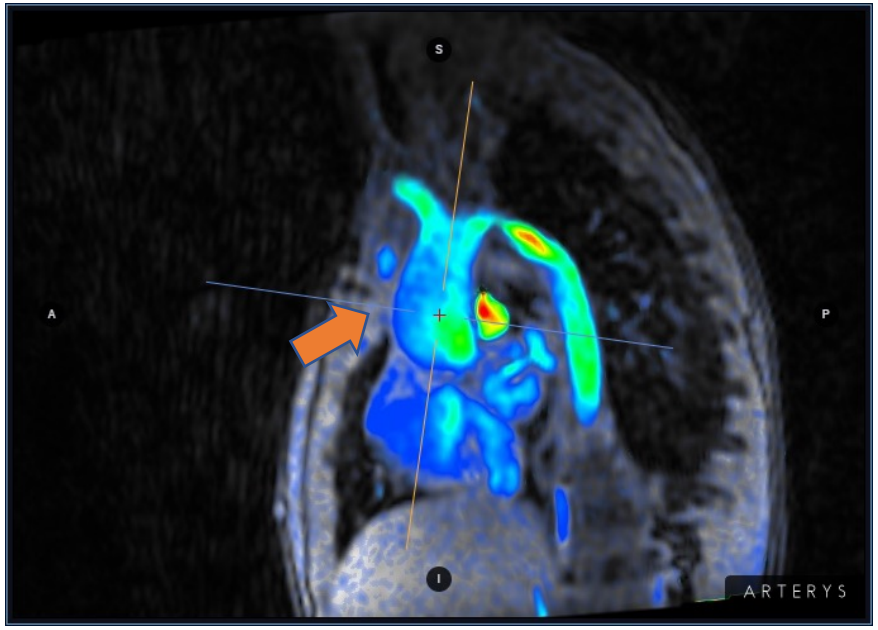
Evaluation of PAs (stenoses, aneurysms) and the aorta [aneurysm, dissection, coarctation (CCT may be superior)]

Coarctation aortique et collatérales aorto-pulmonaires

- Masse VG
- Vmax sur le rétrécissement
- Circulation collatéral
- Recoartaction
- Aneurisme







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Quantification of RV volumes, EF (including subpulmonary RV, systemic RV, and single ventricle)
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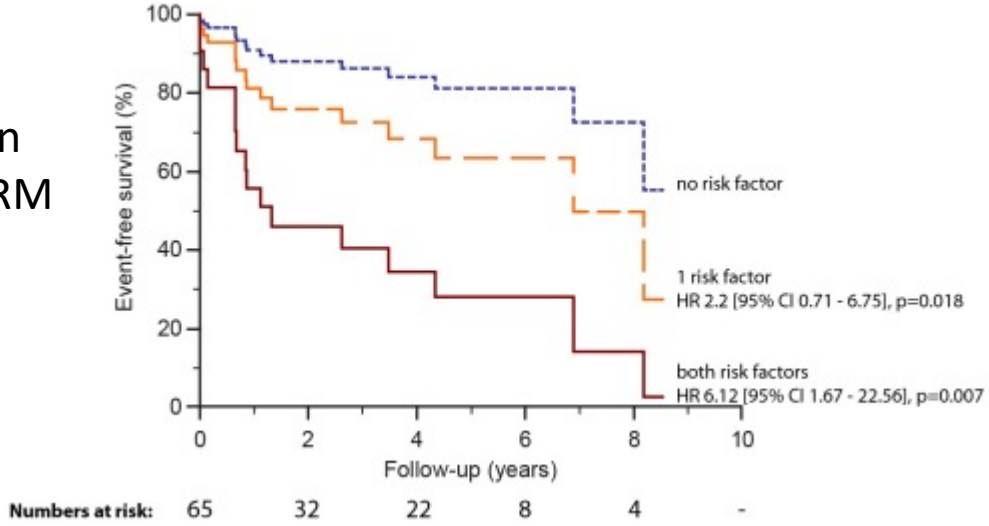
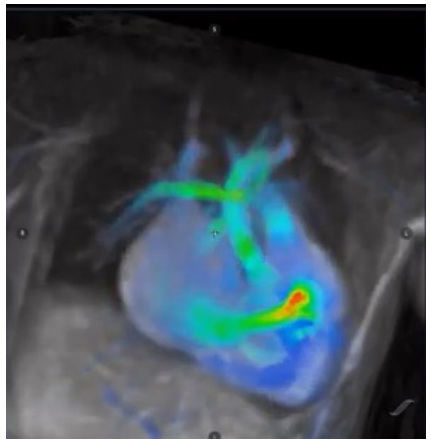
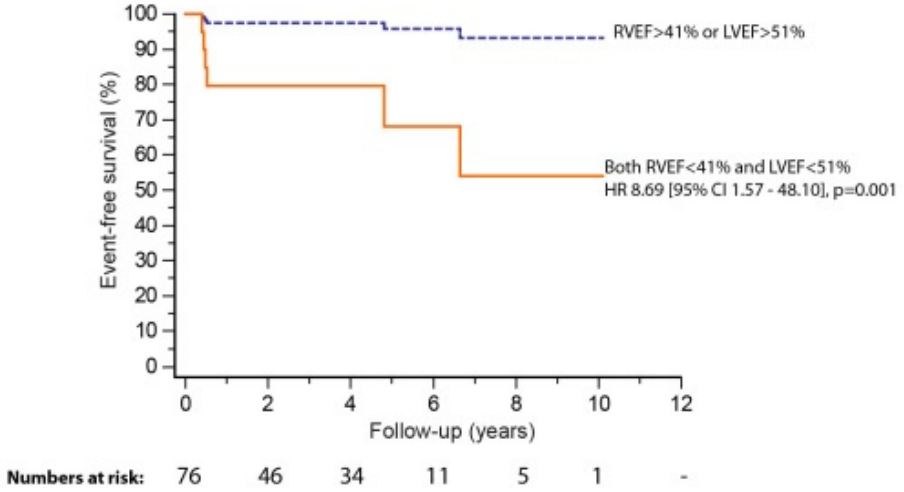
Ebstein

Pronostic: altération biventriculaire en IRM







Rydman, Riikka et al. “Major adverse events and atrial tachycardia in Ebstein’s anomaly predicted by cardiovascular magnetic resonance.” Heart 104 (2017): 37 - 44.

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Tétralogie de Fallot: principaux buts de l'imagerie post-réparation, évaluation anatomique et fonctionnel

RV volumes, function, and mass	LV volumes, function, and mass
RV systolic pressure	Size of the aortic root and ascending aorta
Regional RV wall motion abnormalities	Degree and mechanism of aortic regurgitation
Presence and degree of RVOT obstruction and aneurysm	Aortic arch sidedness and branching pattern
Assessment of the main and branch pulmonary arteries for stenosis	Origin and proximal course of the left and right coronary arteries
Differential pulmonary artery flow	Extent and location of

- Estimation **quantitative des volumes ventriculaires, masse, SV et fraction d'éjection**
- estimation quantitative de la **fuite pulmonaire et fuite tricuspide**
- estimation de **l'output cardiaque, QP/QS**
- Anomalies de **cinétique régionale** du ventricule droit
- **Anatomie de la voie d'éjection du ventricule droit**
- Anatomie des **artères pulmonaires**
- Estimation de **la perfusion pulmonaire**
- Estimation des **collatérales aorto-pulmonaires**
- **Culot aortique +/- fuite aortique**
- **Fibrose** (lésions séquellaires après chirurgie, patch au niveau du VD ou RVOT)

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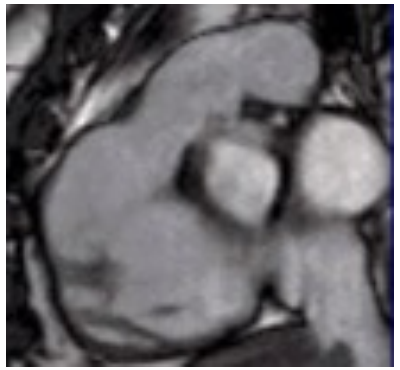
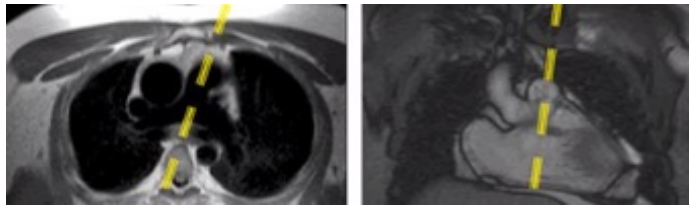
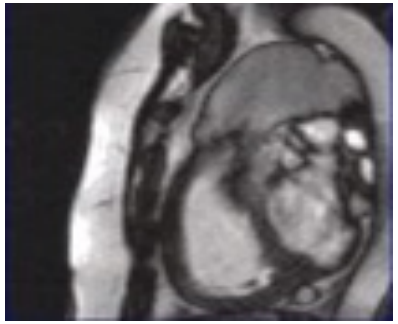
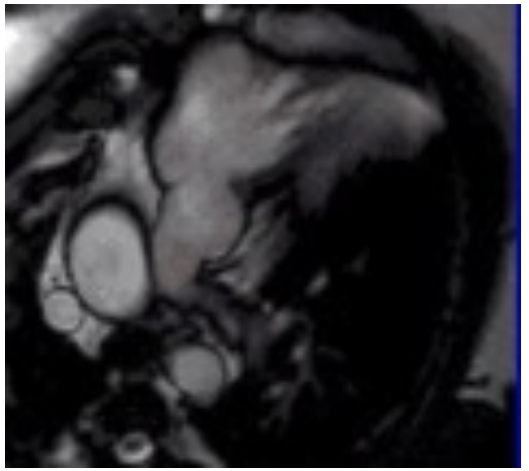
Tétralogie de Fallot: ventricule droit et voie d'éjection du ventricule droit, anatomie







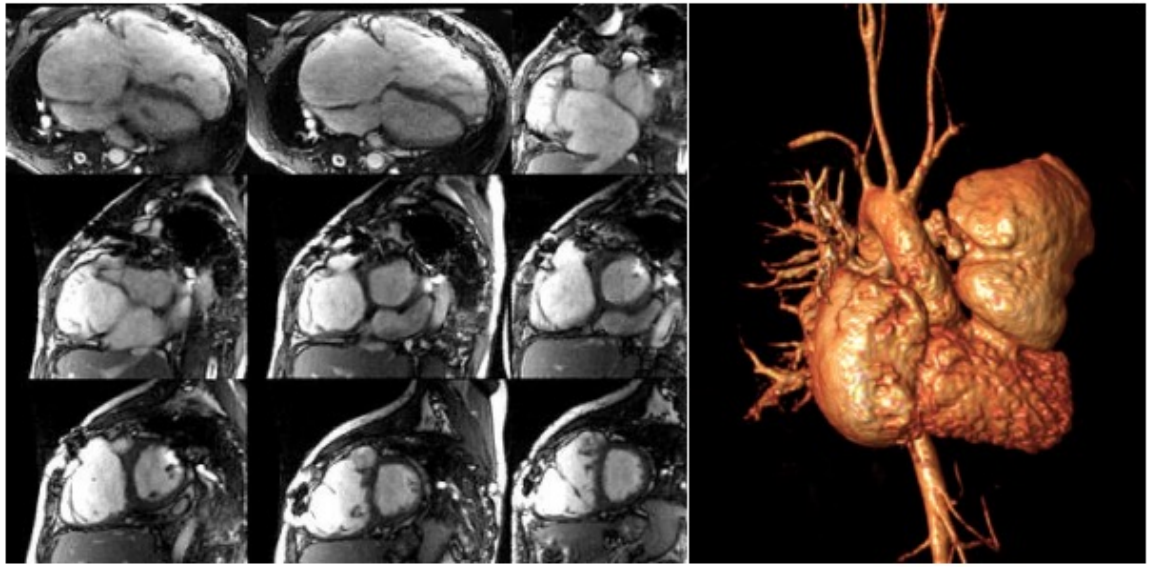


IRM cardiaque et les cardiopathies congénitales

Quantification of RV volumes, EF (including subpulmonary RV, systemic RV, and single ventricle)
Evaluation of RVOTO and RV–PA conduits
Quantification of PR
Evaluation of PAs (stenoses, aneurysms) and the aorta [aneurysm, dissection, coarctation (CCT may be superior)]

Tétralogie de Fallot: ventricule droit et voie d'éjection du ventricule droit, anatomie

	<2 y*	2–9 y	10–19 y	20–49 y	≥50 y
Echocardiogram	12 mo	12 mo	24 mo†	24 mo†	24 mo†
Cardiovascular MR	Not recommended routinely. Ordered to address specific questions not answered by echocardiography.		36 mo in stable patients.		
			12 mo if moderate (≥150 mL/m²) or progressive (increase of >25 mL/m²) RV dilatation or dysfunction (RVEF ≤48% or ≥6% decrease in EF) or nearing imaging criteria for PVR.		
Computed tomography	Not recommended routinely. Ordered when CMR is indicated but cannot be performed (eg, metallic artifacts or contraindications to CMR).				
Lung perfusion scan	If predicted RV systolic pressure ≥60% systemic or smallest branch PA diameter *z* score <−2.5. In patients ≥10 y of age consider CMR flow measurements.				
X-ray angiography	Not recommended routinely.‡ Ordered when noninvasive methods either cannot be performed or have failed to provide satisfactory diagnostic data.				Coronary angiography when clinically indicated.
Chest radiograph	Not recommended routinely. May be ordered for evaluation of stent integrity.				



Geva T. JCMR 2011

Valente AM, Geva T. Circ Cardiovasc Imaging. 2017

IRM cardiaque et les cardiopathies congénitales

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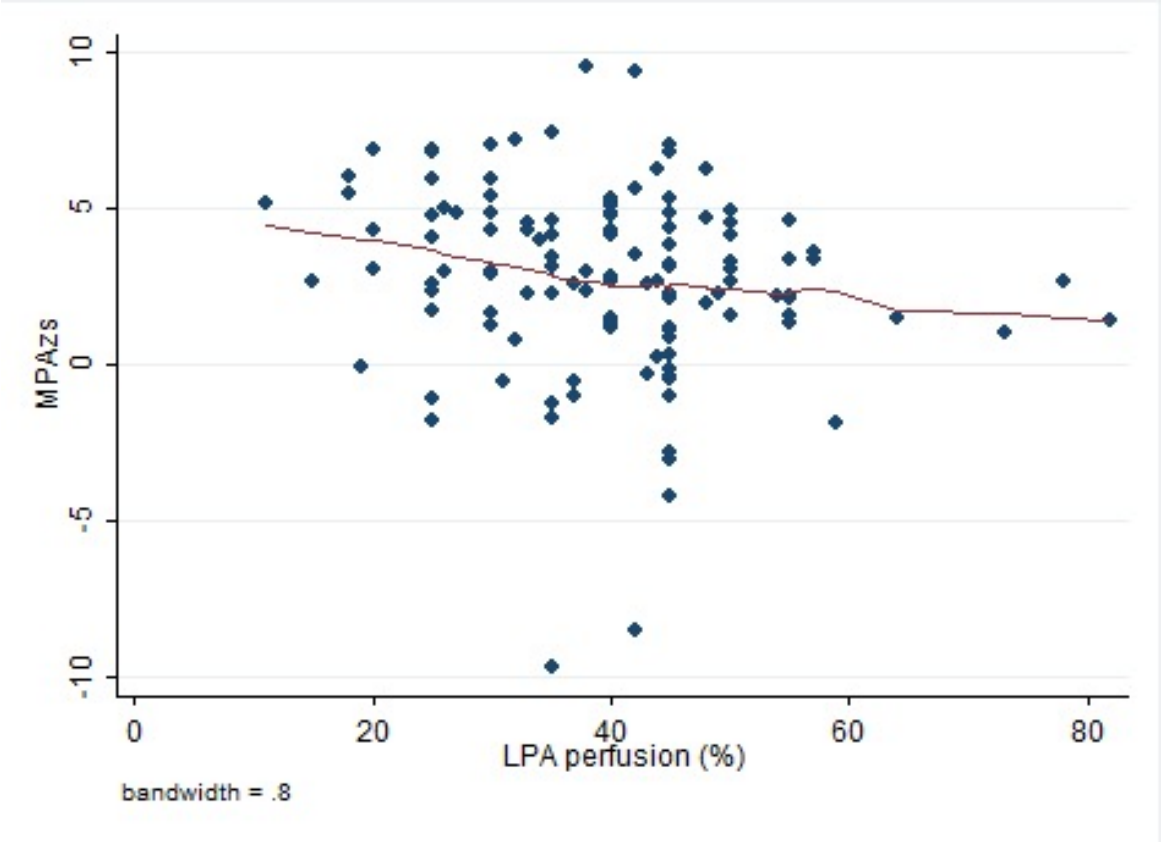
Tétralogie de Fallot: analyse fonctionnel des flux

Asymétrie de la perfusion pulmonaire dans le **48 %** des patientes→ **42%** hypoperfusion pulmonaire gauche



Panaioli et al Pulmonary Perfusion Asymmetry in Patients after Repair of Tetralogy of Fallot: a 4D Flow MRI-based Study. Congenital Heart Disease. 2021

IRM cardiaque et les cardiopathies congénitales

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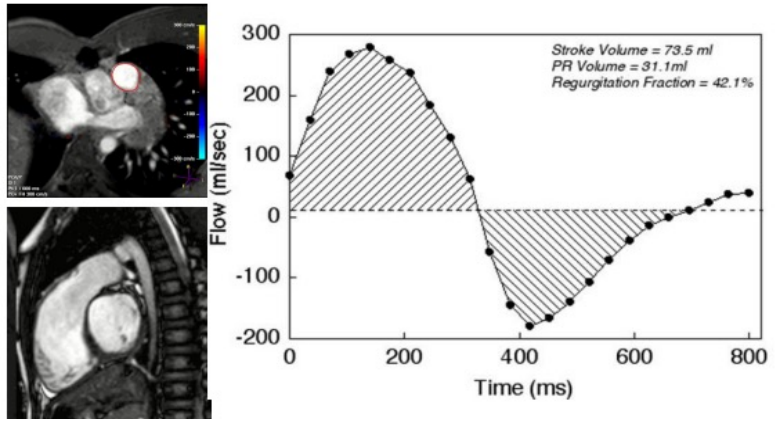
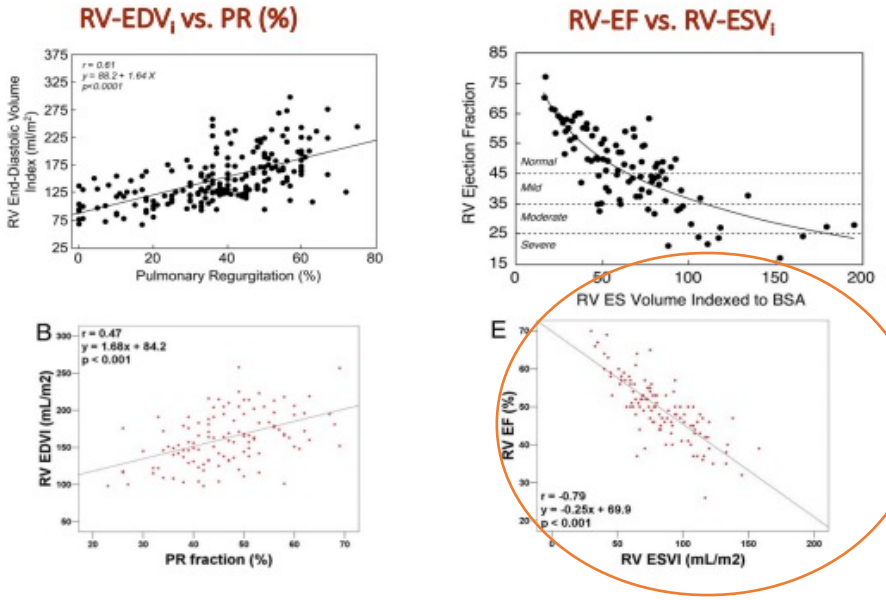
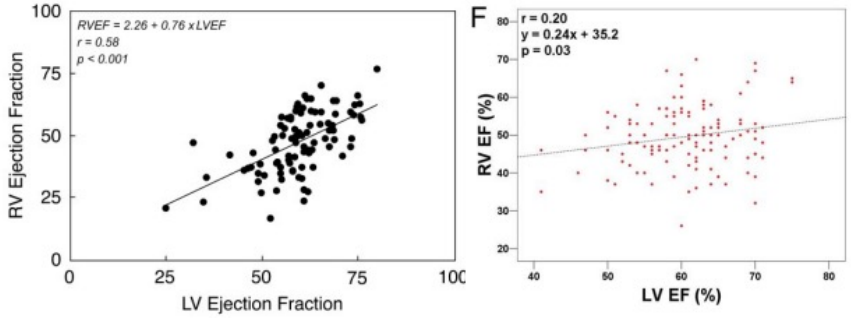
Tétralogie de Fallot: fuite pulmonaire







VTSVDi 80 ml/m2

Geva T. JCMR 2011
Geva T, et al. JACC 2004; Lee C, et al. JACC 2012

IRM cardiaque et les cardiopathies congénitales

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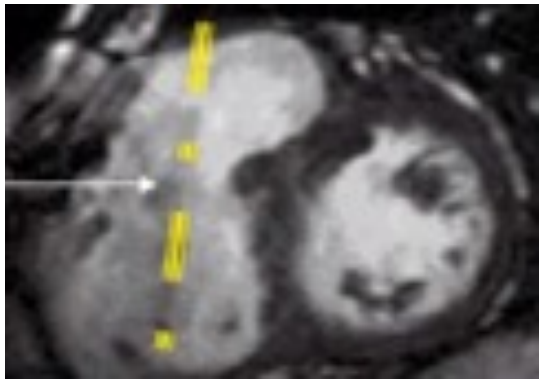
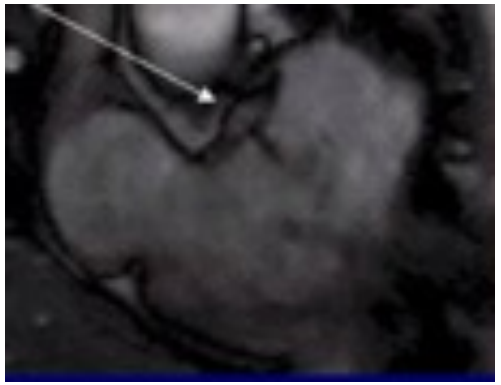
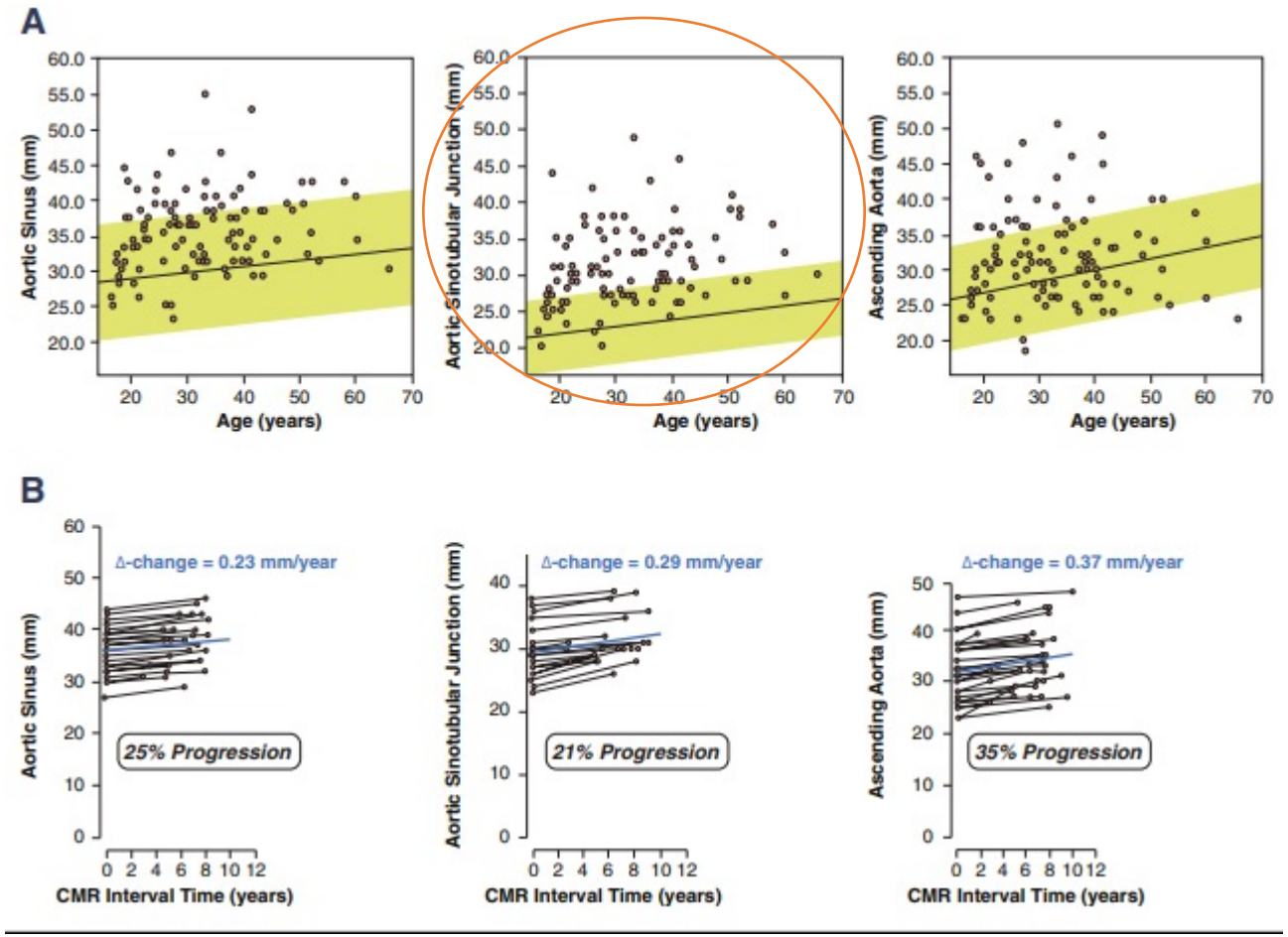
Tétralogie de Fallot: autres lésions résiduelles







B. Bonello, et al Aortic dilatation in repaired tetralogy of Fallot, JACC Cardiovasc. Imaging 11 (1) (2018) 150–152

IRM cardiaque et les cardiopathies congénitales

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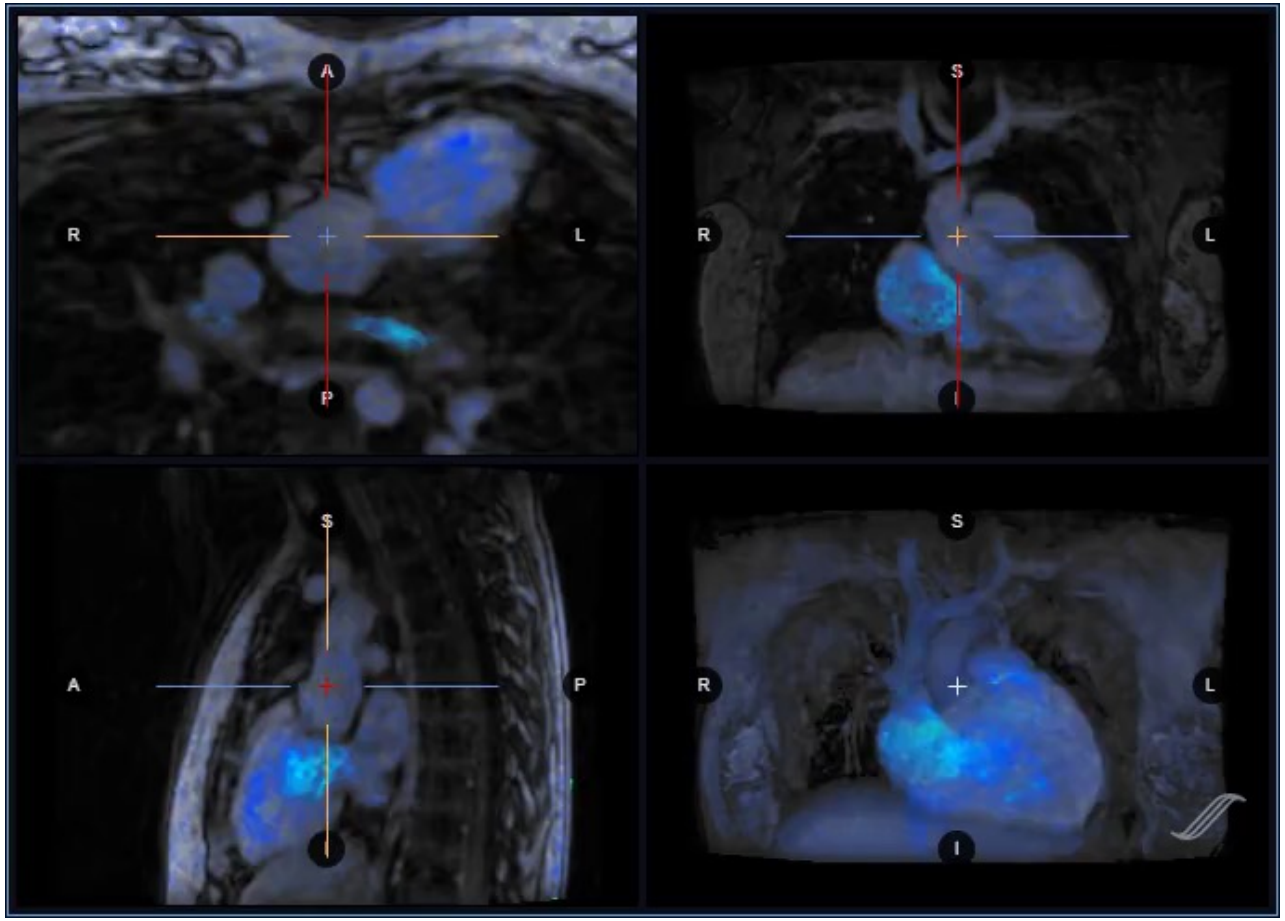
Tétralogie de Fallot: 4D flow synthèse



IRM cardiaque et les cardiopathies congénitales

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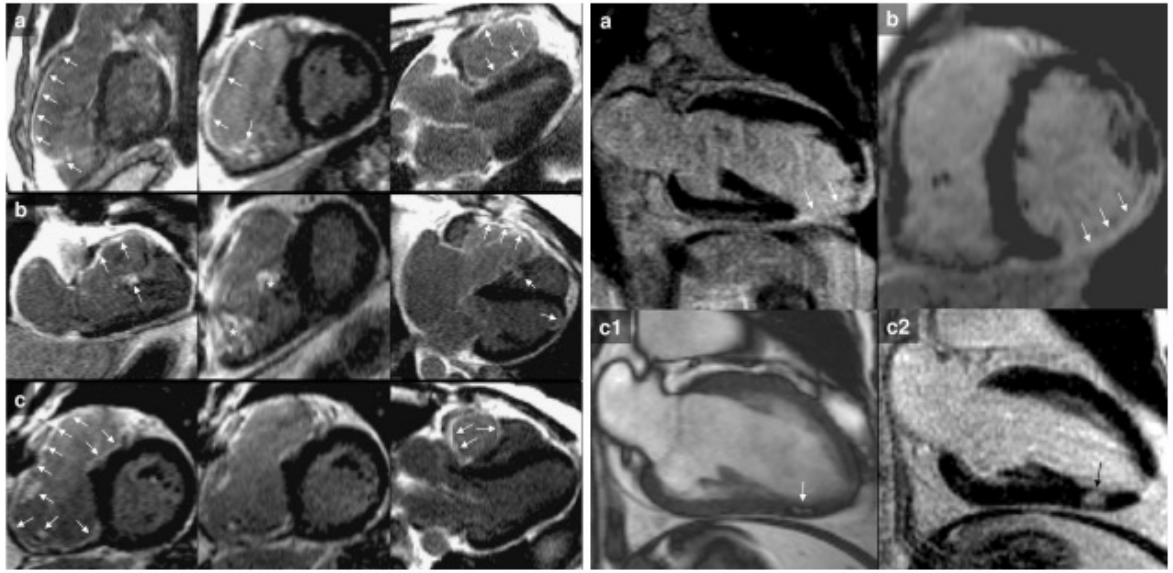
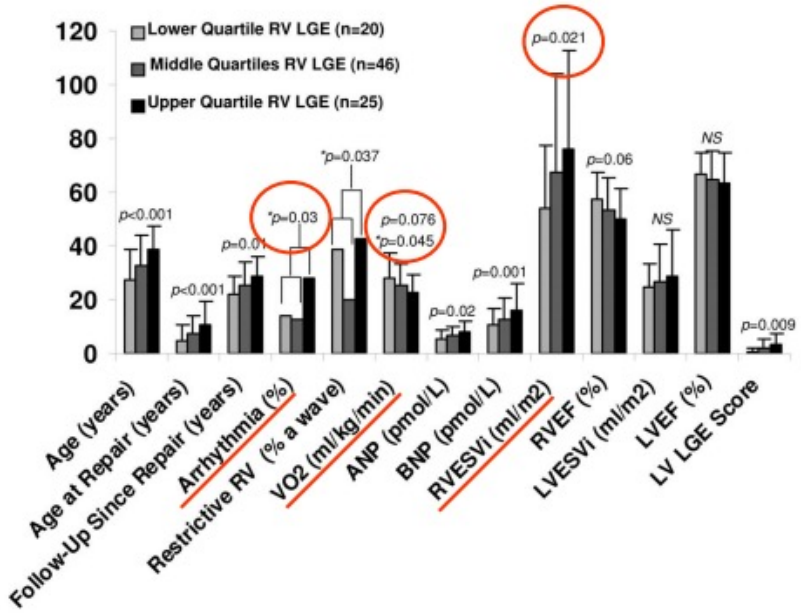
Tétralogie de Fallot: pronostic





Babu-Narayan, S. V. et al. Circulation 2006;113:405-413

IRM cardiaque et les cardiopathies congénitales

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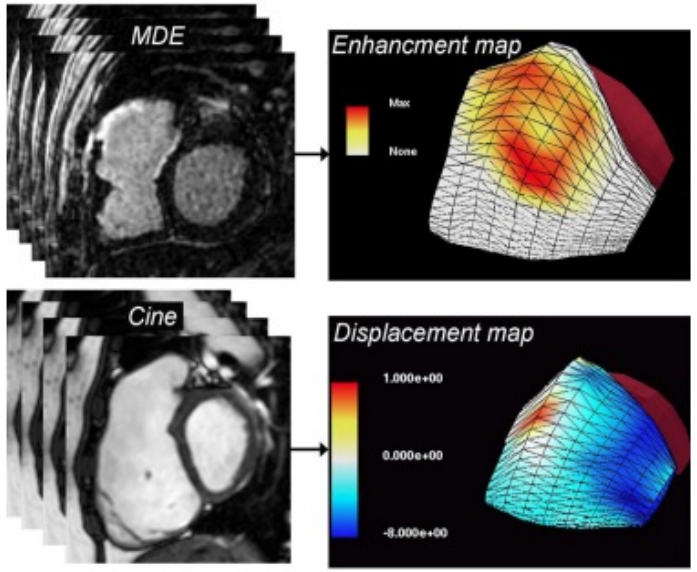
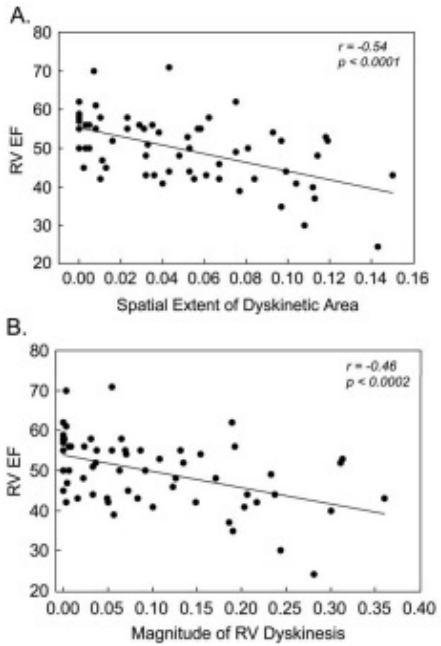
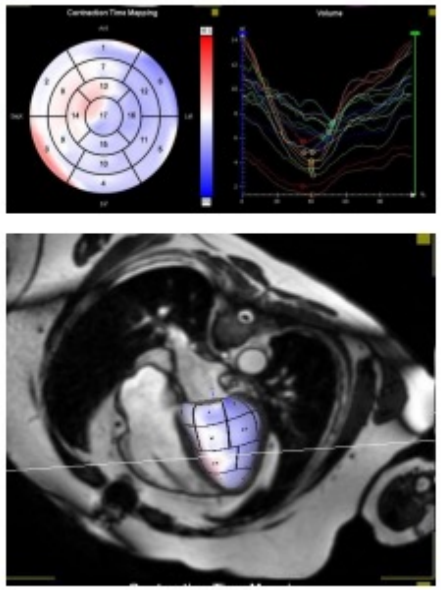
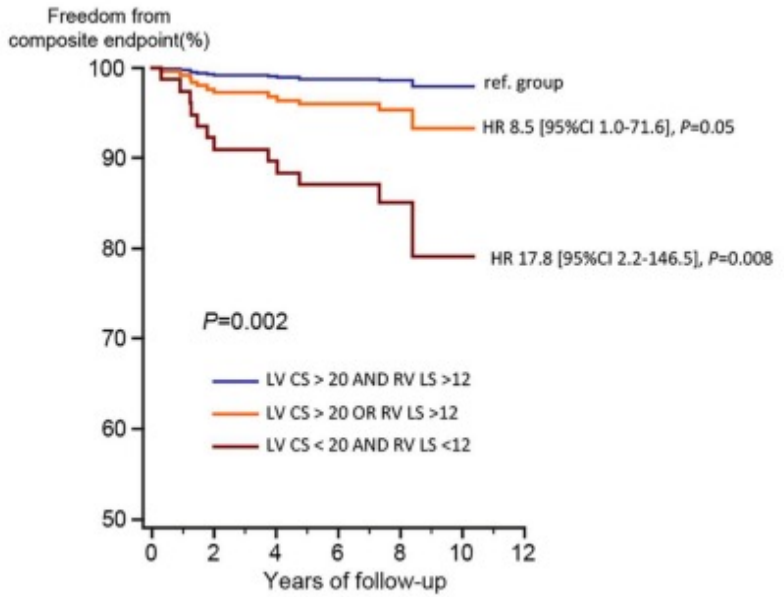
Tétralogie de Fallot: pronostic





Wald R., et al. Circulation 2009





Orwat S, Diller GP, et al., Baumgartner H; German Competence Network for Congenital Heart Defects Investigators. Myocardial deformation parameters predict outcome in patients with repaired tetralogy of Fallot. Heart. 2016 Feb;102(3):209-15

IRM cardiaque et les cardiopathies congénitales

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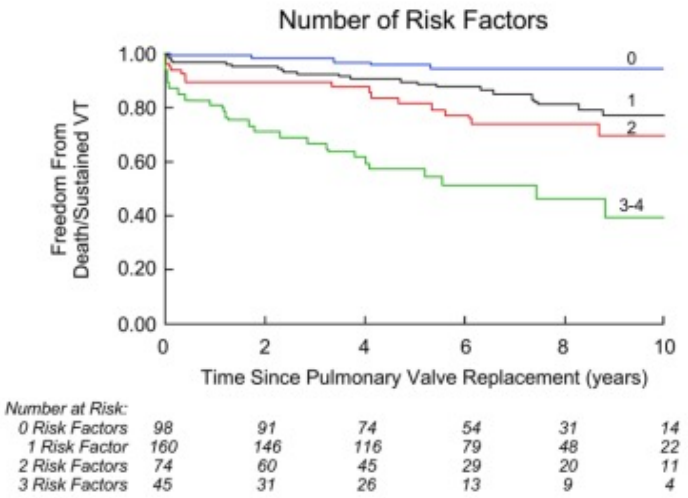
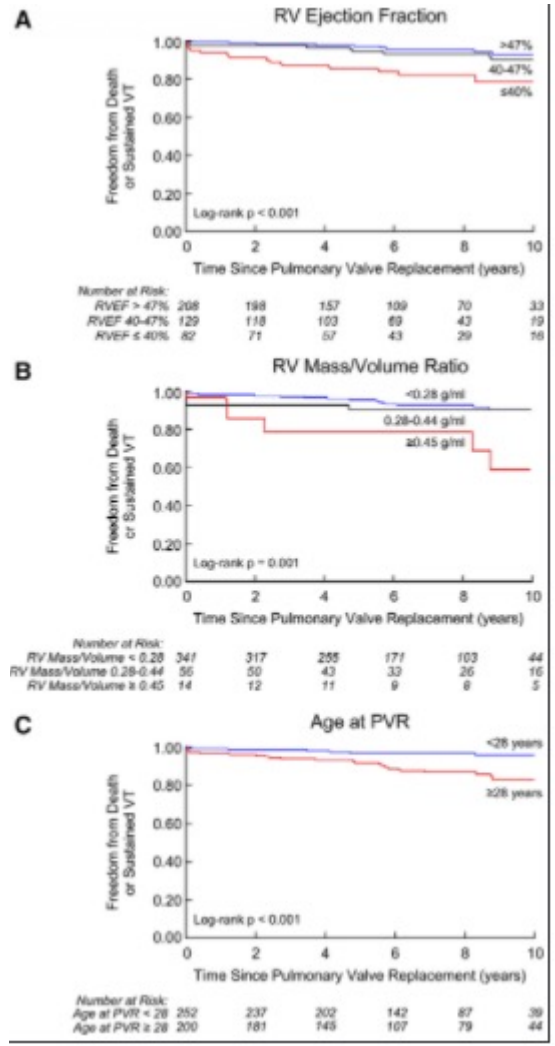
Tétralogie de Fallot: pronostic et therapie

- Age
- **Dysfonction** du ventricule droit
- **Hypertrophie** du ventricule droit
- PVD augmentée





Geva T, et al. Circulation. 2018 Nov 6;138(19):2106-2115

IRM cardiaque et les cardiopathies congénitales

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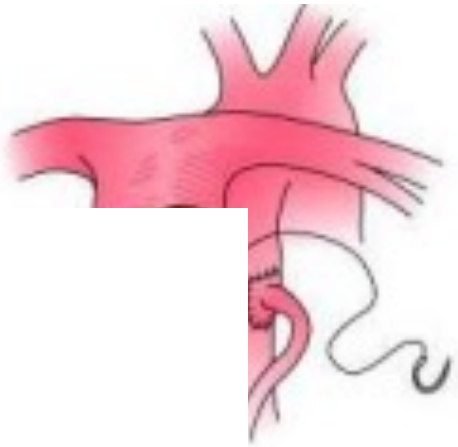
Tétralogie de Fallot: pronostic et thérapie

- Therrien RV EDVI 170 ml/m2 and RV ESVI 85 ml/m2
- Lee RV EDVI 163 ml/m2 or RV ESVI 80 ml/m2
- Oosterhof RV EDVI 160 ml/m2 and RV ESVI 82 ml/m2)
- Geva RV ESVI < 90 ml/m2
- Frigiola et al. aggressive policy: RV EDVI 150 ml/m2

PR> 30% et ≥ 2 des les critères suivantes (asymptomatique) :

1) **Volume télédiastolique du ventricule droit indexe > 150 ml/m2** ou Zscore >4
2) **RV/LV ratio télédiastolique volume >2**
3) **Volume télésystolique du ventricule droit indexé > 80 ml/m2**
4) **Fraction d'éjection du ventricule droit < 47%,**
5) **Fraction d'ejection du ventricule gauche < 55%**
6) **Aneurisme importante du VD**
7) Durée du QRS > 140 ms
8) Tachycardie ventricule soutenue
9) **Obstruction de la voie d'éjection du VD** avec une PVD > > 2/3 de la pression systémique
10) **Sténoses sévère de la branche pulmonaire** avec une réduction du flux < 30% (avec un traitement pas faisable par KT)
11) **Fuite tricuspide** ≥ grade moyen
12) **Shunt G-D** (CIA ou CIV QP/QS > 1.5)
13) **Fuite aortique sévère**
14) **Dilatation sévère de la racine aortique ≥ 5 cm**

Geva T, et al. Circulation. 2018 Nov 6;138(19):2106-2115



Catheter intervention (n = 54)	Number of procedures (%) 104 procedures 101 interventions	Number of patients
Relief supravalvular PS	80 (76.9%)	44
Balloon angioplasty[b]		27
MPA		5
RPA		8
LPA		5
Bilateral PA		9
Stent implantation		39
MPA only		1
RPA only		13
LPA only		11
Bilateral PA		13
MPA + RPA		1
Relief arch obstruction	15 (14.4%)	10
Balloon angioplasty	14	
Stent implantation	1	
Coronary artery intervention	2 (1.9%)	2
PTCA	1	
Stent implantation	1	
Neoaortic valve intervention	1 (1.0%)	1
TAVI	1	
Closure shunts	6 (5.8%)	5
Atrial septal defect	5	
Aorta-pulmonary collaterals	1	

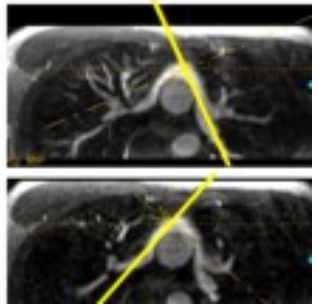
Reoperation (n = 83)	Number of procedures (%) 137 procedures 117 reoperations	Number of patients
RVOTO relief	68 (49.6%)	50
Neoaortic valve and root surgery	21 (15.3%)	15
Neoaortic valve plasty	1	
Neoaortic valve replacement	3	
Bentall operation	12	
David operation	2	
Switchback Ross	1	
Replacement ascending aorta	2	
Coronary revascularisation	8 (5.8%)	8
Ostial plasty	4	
CABG	4	
Relief arch obstruction	12 (8.8%)	12
LVOTO relief	4 (2.9%)	4
Miscellaneous[a]	24 (17.5%)	22

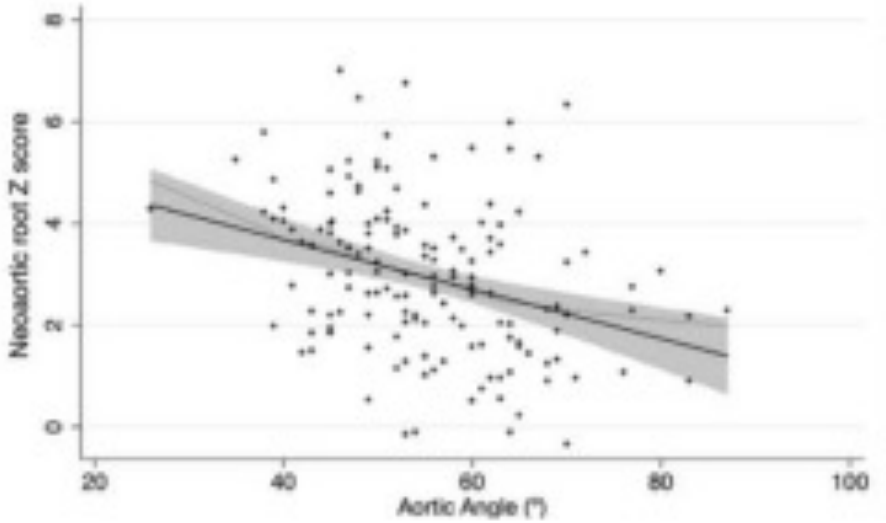
van der Palen RLF, et al Long-term outcome after the arterial switch operation: 43 years of experience. Eur J Cardiothorac Surg. 2021 May 8;59(5):968-977.

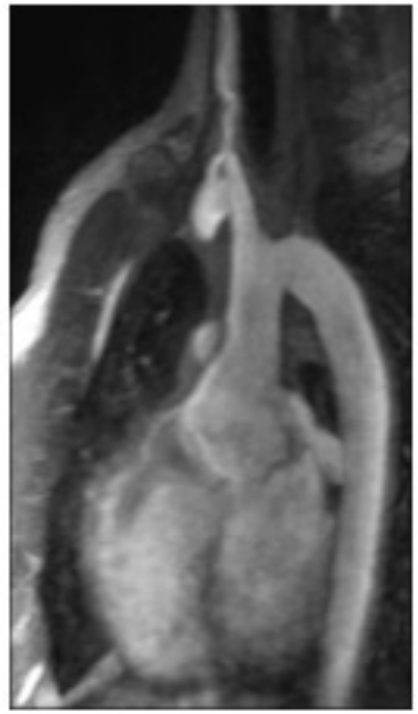
IRM cardiaque et les cardiopathies congénitales

Coronary anomalies and CAD (CCT is superior for intramural course, slit-like course, acute angle take-off, myocardial bridging, and plaque assessment)
Detection and quantification of myocardial ischaemia by CMR stress perfusion

Transposition des gros vaisseaux : switch artérielle, anatomie







Martins et al. Int J Cardiol 2019

IRM cardiaque et les cardiopathies congénitales

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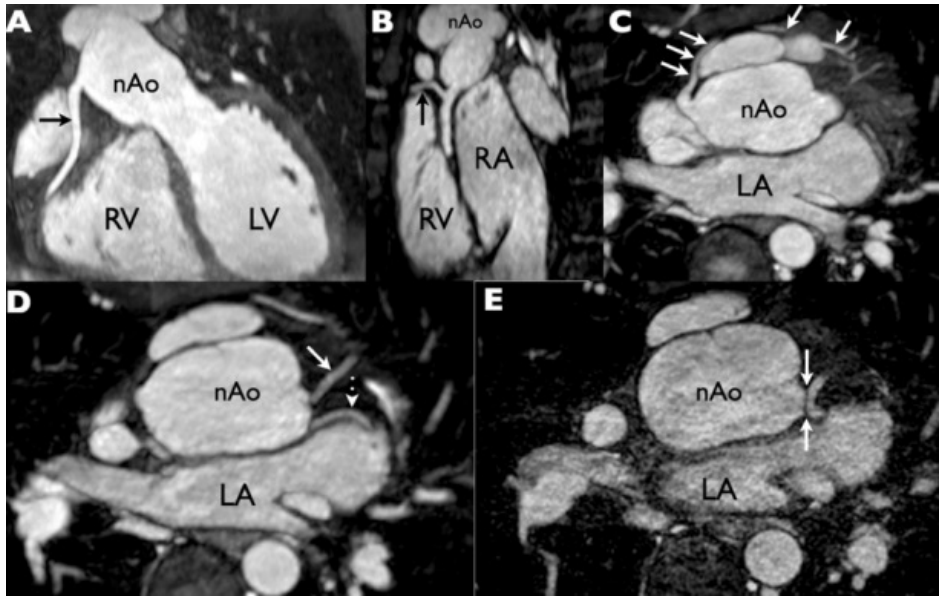
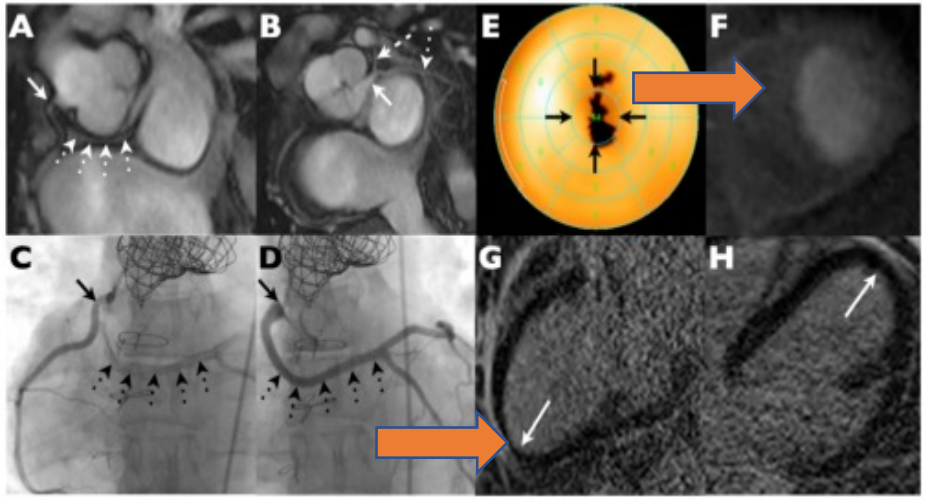
Transposition des gros vaisseaux : switch artérielle, ischemie





1) **Lésions des artères coronaires → 4/49 pts, 8%; Ou et al JTCS 2006**
2) **Perfusion normal dans 27/27 patients, FP dans 50% avec SPECT; Tobler, JSCMR et al 2014**
3) **Ischémie coronaire dans 2/145 pts; Kempny et al Intern. J Cardiology 2013**

Tobler D, et al. Evaluation of a comprehensive cardiovascular magnetic resonance protocol in young adults late after the arterial switch operation for d-transposition of the great arteries. J Cardiovasc Magn Reson. 2014 Dec 11;16(1):98.

IRM cardiaque et les cardiopathies congénitales

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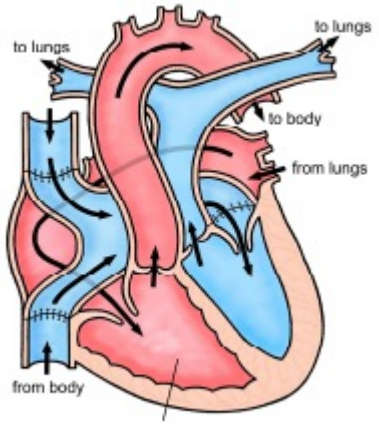
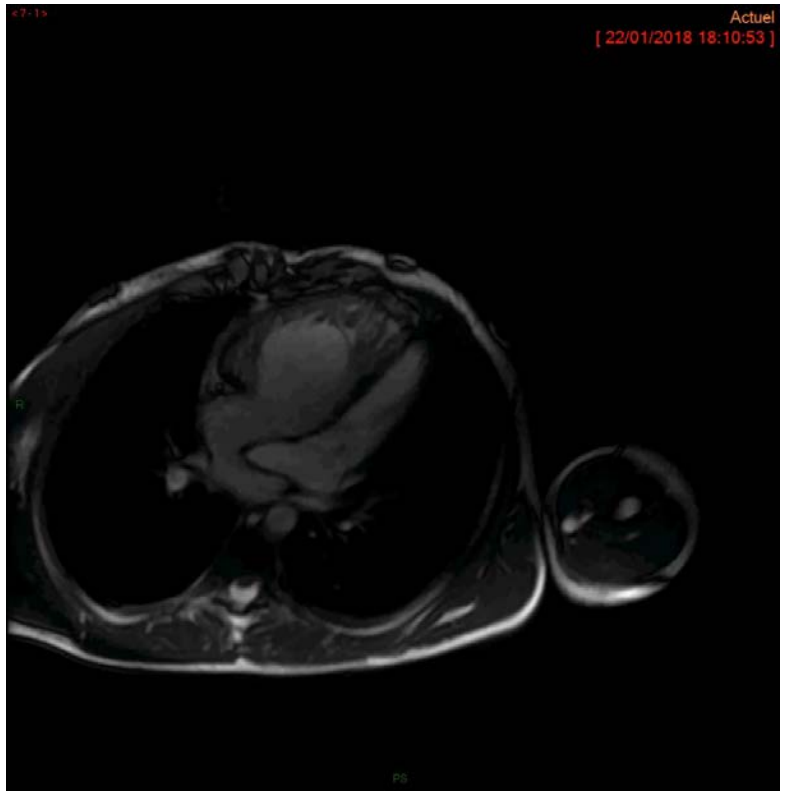
Detection and quantification of myocardial ischaemia by CMR stress perfusion

Transposition des gros vaisseaux : switch atrial, VD systémique

1) Ventricule droit systémique
2) Baffles auriculaire
3) Obstruction de la voie d'éjection gauche (sous pulmonaire)
4) Obstruction de la voie d'éjection droite (sous aortique)
5) Fuite tricuspide
6) Fibrose ventriculaire





IRM cardiaque et les cardiopathies congénitales

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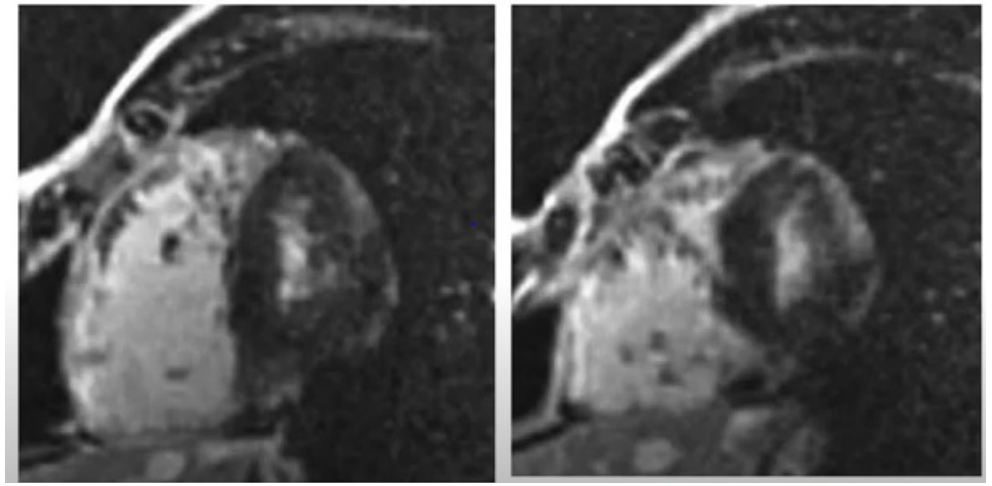

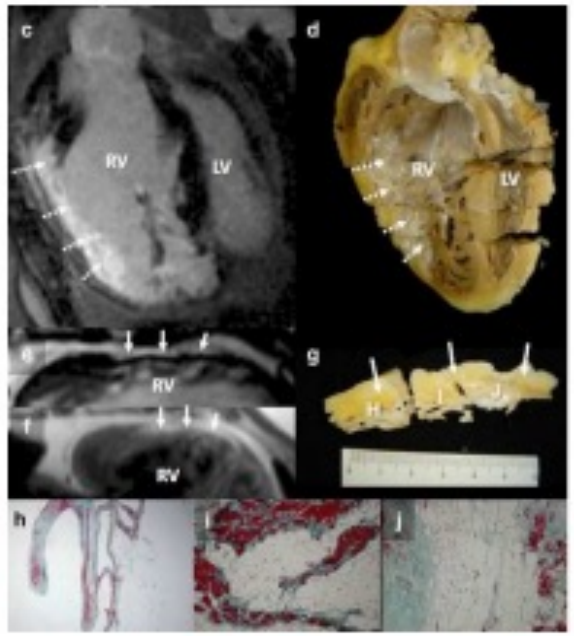

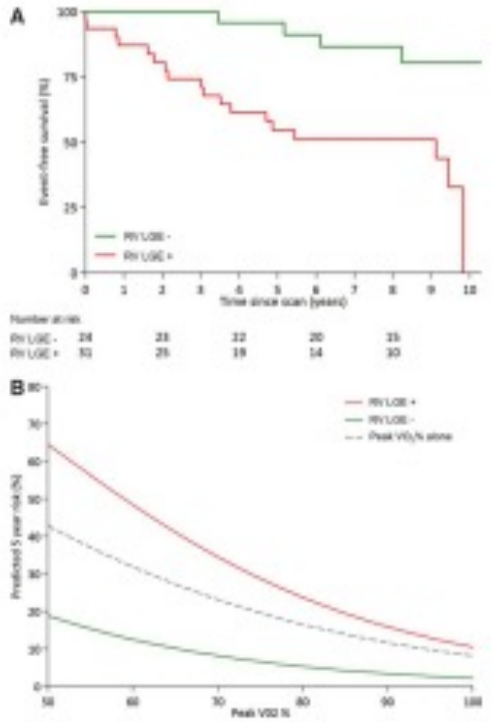

Rydmann et al. Circ Cardiovasc Im 2015

IRM cardiaque et les cardiopathies congénitales

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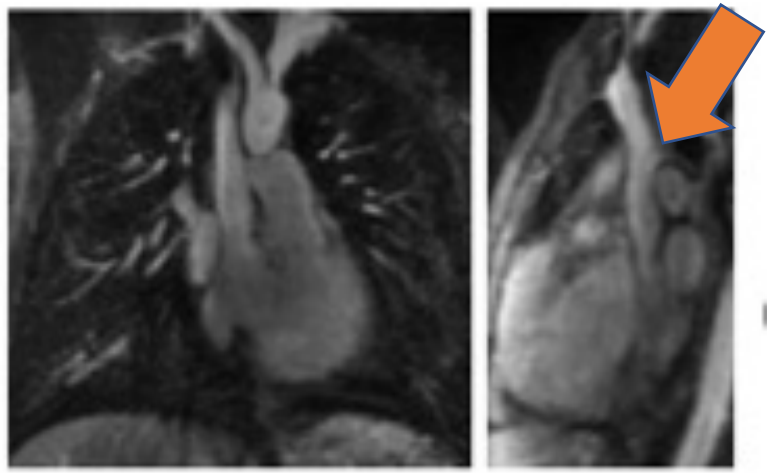
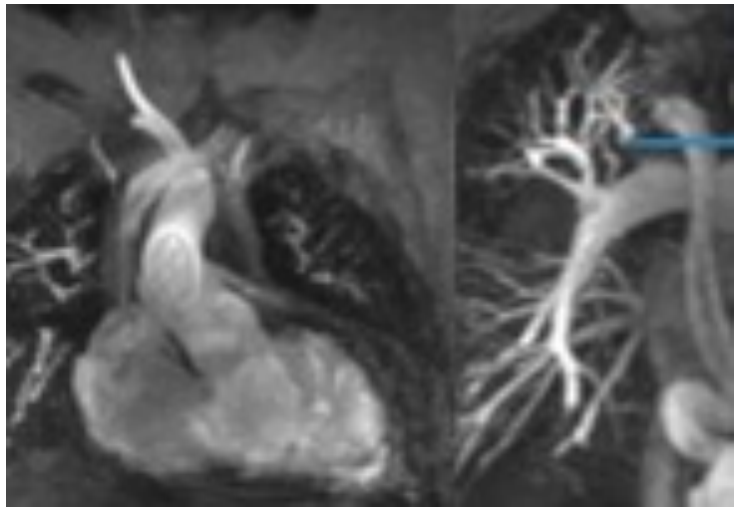
Transposition des gros vaisseaux : switch atrial

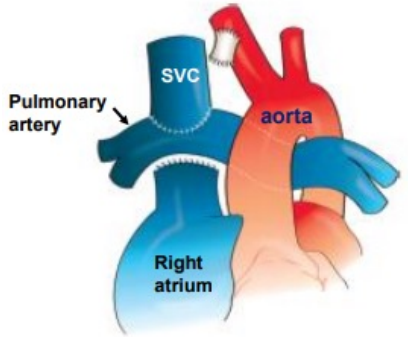


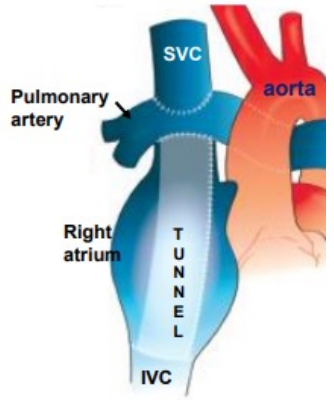


IRM cardiaque et les cardiopathies congénitales

Evaluation of systemic and pulmonary veins (anomalous connection, obstruction, coronary venous anatomy pre-procedure, etc.)

Ventricule unique et circulation du Fontan





1) Ventricule unique (fonction et morphologie)
2) Evaluation du montage
3) Anatomie des artères pulmonaires
4) Collatérales aorto-pulmonaires et fistule veine-veineuse
5) Obstruction de la voie d'éjection
6) Fuite de la valve AV ou systémique
7) Fibrose ventriculaire

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Evaluation of systemic and pulmonary veins (anomalous connection, obstruction, coronary venous anatomy pre-procedure, etc.)

Ventricule unique et circulation du Fontan

Grosse-Wortmann L, Al-Otay A, Yoo SJ. Aortopulmonary collaterals after bidirectional cavopulmonary connection or Fontan completion: quantification with MRI. Circ Cardiovasc Imaging. 2009 May;2(3):219-25

IRM cardiaque et les cardiopathies congénitales

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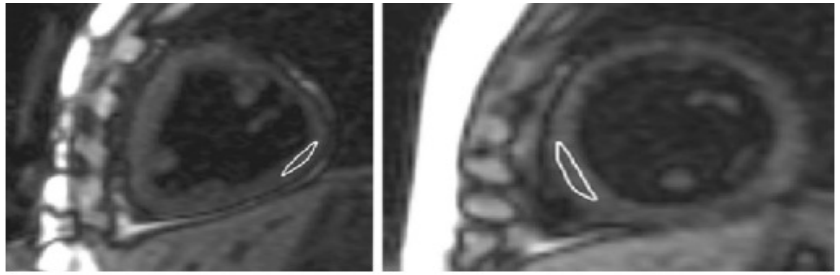
	Composite outcomes N = 22	No composite outcome N = 33	P Value
Volumetry and flows			
Mass indexed (g/m^2)	79.5 (22.8)	67.0 (20.6)	0.039
Mass/volume	0.55 (0.09)	0.57 (0.11)	0.397
EDVI (mL/m^2)	146.2 (50.0)	117.5 (36.2)	0.013
ESVI (mL/m^2)	87.2 (37.9)	63.6 (23.8)	0.006
SVSVI (mL/m^2)	58.9 (18.2)	53.9 (18.2)	0.239
SVEF (%)	41.9 (8.9)	46.4 (7.5)	0.045
APC flow (L/m^2)	0.81 (0.50)	0.51 (0.38)	0.017
Feature tracking			
Longitudinal strain (%)	− 12.4 (6.3)	− 15.4 (6.5)	0.299
Radial strain (%)	6.1 (1.6)	6.6 (1.3)	0.264
Circumferential strain (%)	− 21.5 (6.8)	− 25.0 (4.2)	0.029
STD-T2P	2.0 (0.9)	1.7 (0.9)	0.287
Myocardial fibrosis and scarring			
ECV freewall (%)	25.1 (4.4)	28.1 (8.9)	0.098
Native T1 freewall (ms)	1063 (62)	1026 (59)	0.029
LGE prevalence (%)	2 (9%)	6 (18%)	0.153
Coupling and wall and fiber stress			
VAC ratio	1.5 (0.6)	1.2 (0.4)	0.043
ESWS (kPA)	19.3 (4.1)	17.4 (3.6)	0.084
ESFS (kPA)	12.6 (1.9)	12.0 (1.7)	0.225



Pisesky A, et al. Adverse fibrosis remodeling and aortopulmonary collateral flow are associated with poor Fontan outcomes. J Cardiovasc Magn Reson. 2021 Nov 15;23(1):134.

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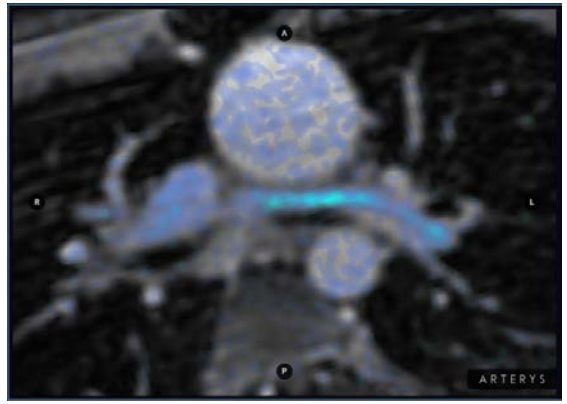

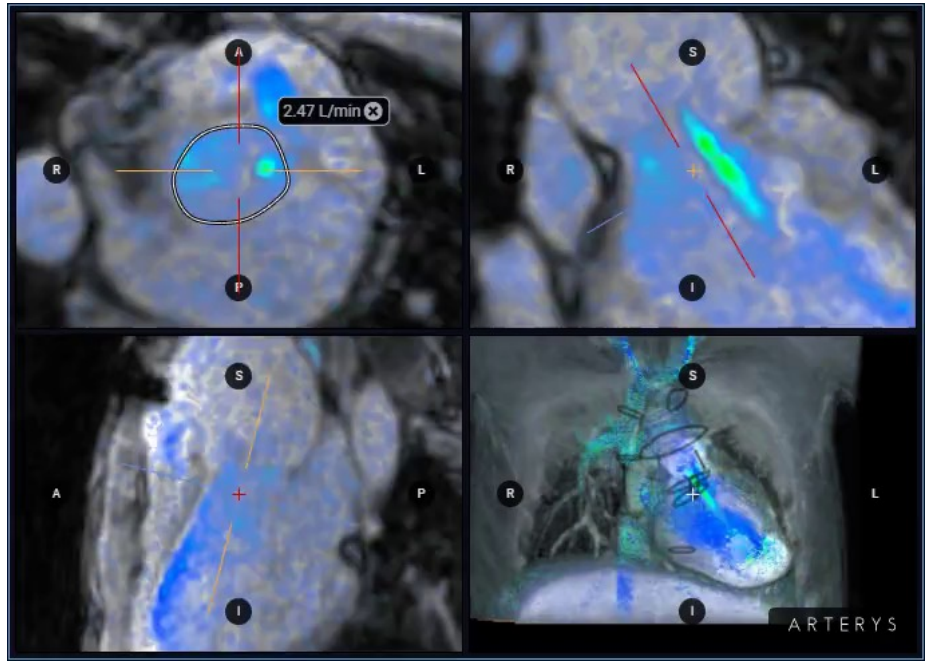

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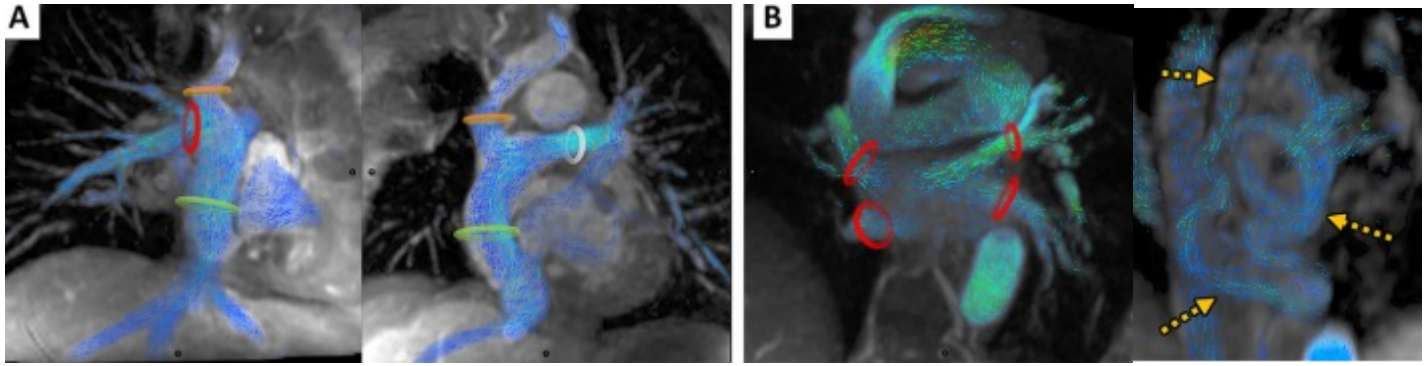
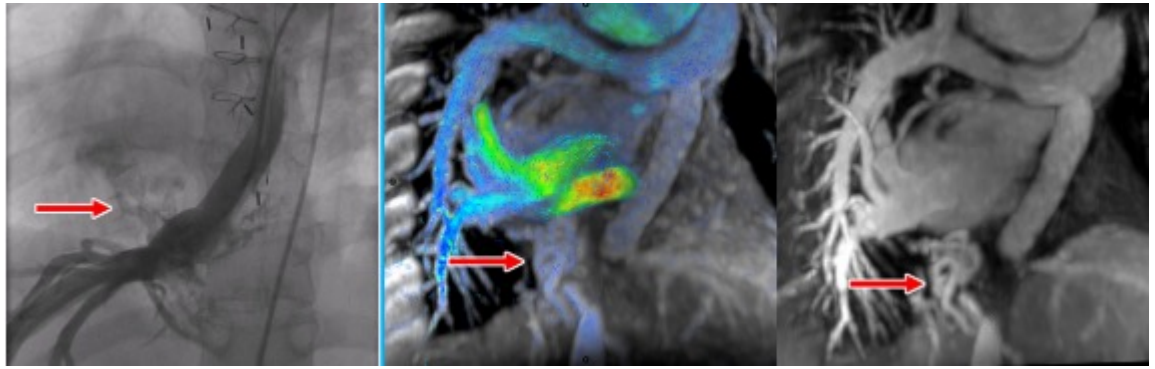




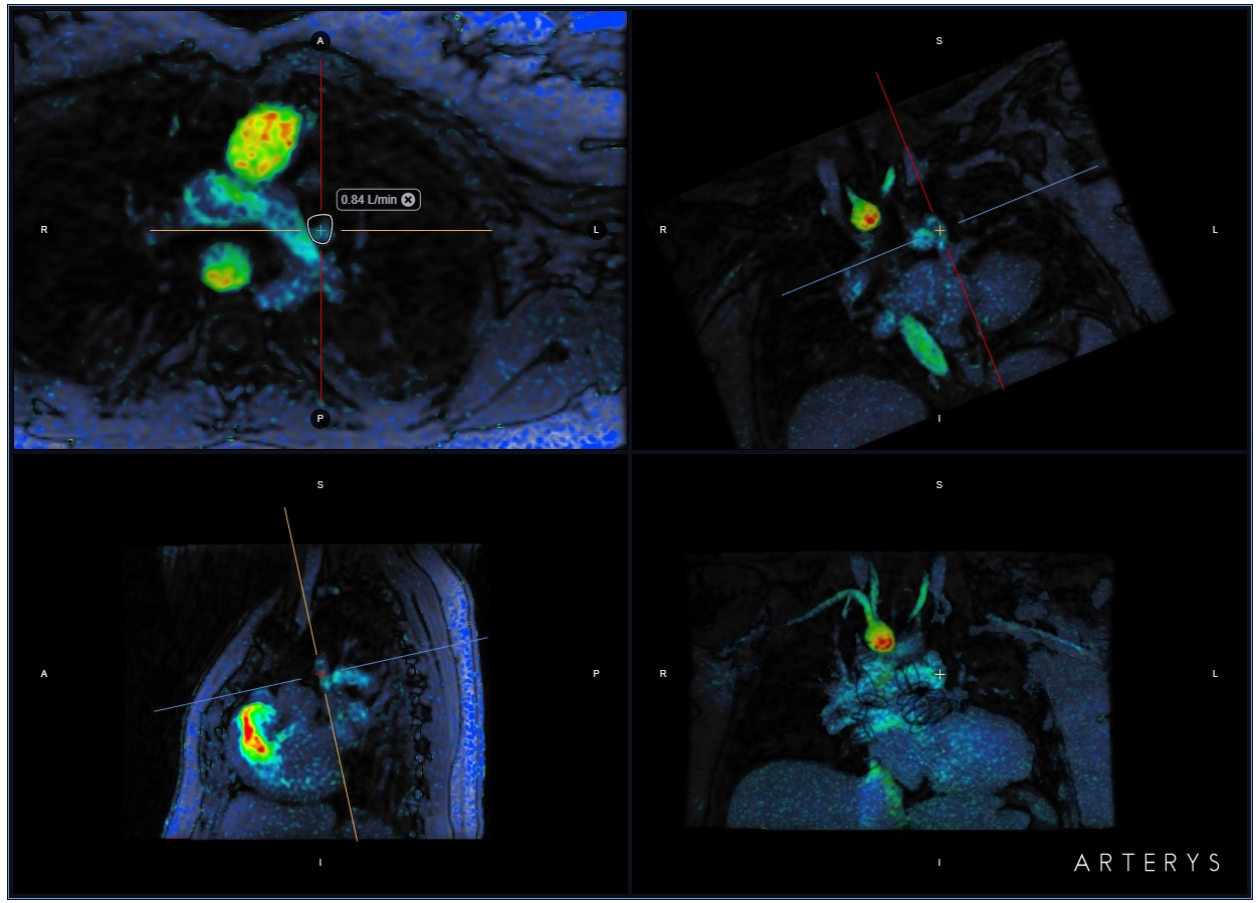
Raimondi F, et al Prevalence of Venovenous Shunting and High-Output State Quantified with 4D Flow MRI in Patients with Fontan Circulation. Radiol Cardiothorac Imaging. 2021 Dec 16;3(6):e210161.

Cas clinique

- Cardiopathie complexe, de type cardiopathie univentriculaire, avec atrésie de la valve AV gauche et malposition vasculaire. Dérivation cavo-pulmonaire totalw avec une fenêtre. Fenêtre spontanément fermée au cours du suivi.
- Limitation à l'effort marquée, se majorant progressivement.
- **Effort quasi maximal. Très nette limitation de la VO2 max. Désaturation à l'effort modérée, SV très précoce**. Pas de signe d'ischémie, 3 ESV à l'effort isolées monomorphes, profil TA normal.
- **Sat 89%**

Cas clinique

- **Circulation collatérale veno-veineuse du système hémi-azygos vers l'oreillette avec un débit estimé autour de 1 l/min.**
- **Kt**: obstacle sur le tube intracardiaque (2 mmHg) et **fistules veinocardiaques significatives**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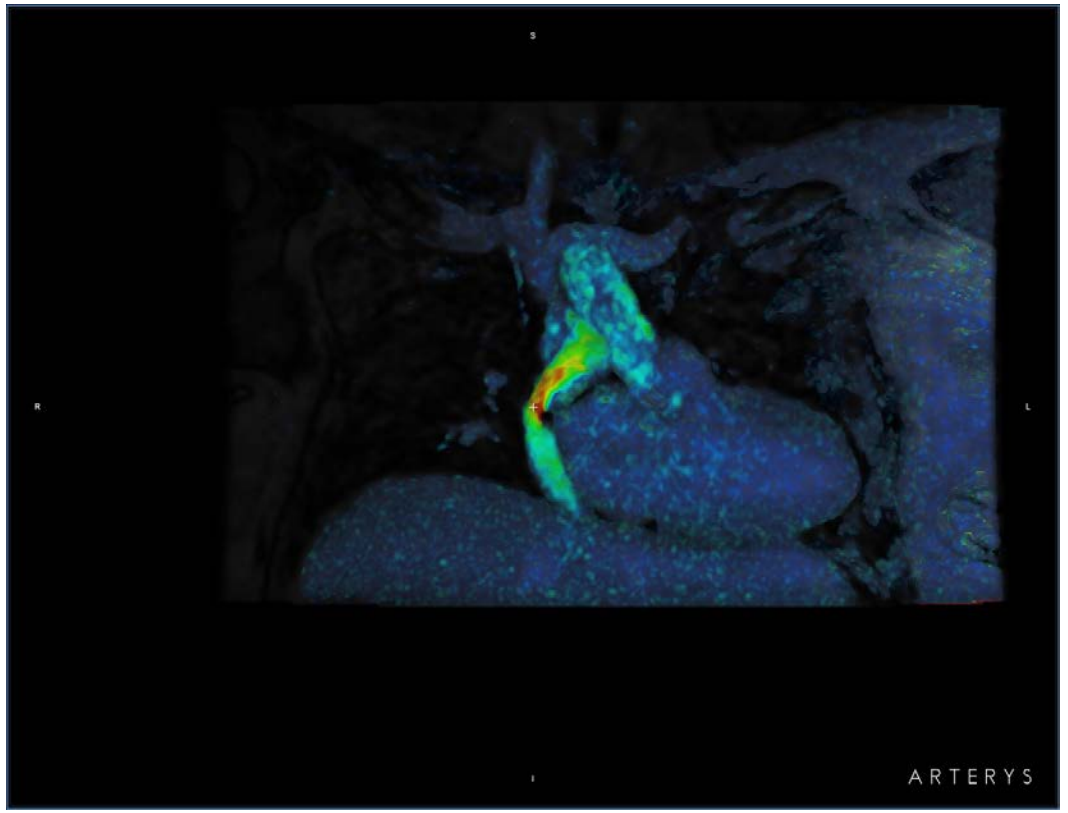
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Merci pour votre attention

Tétralogie de Fallot: *thérapie et IRM cardiaque*