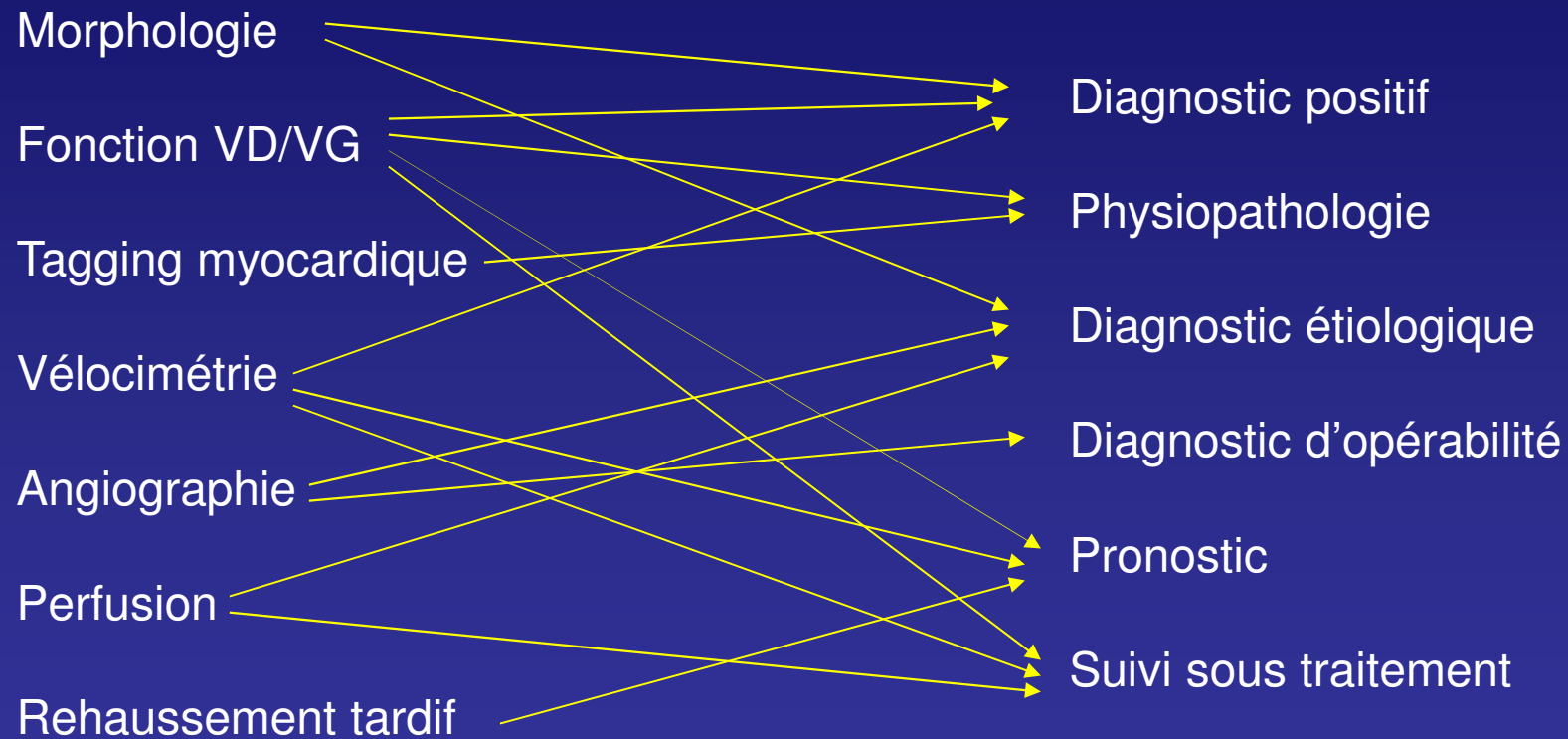


Hypertension artérielle pulmonaire : exploration IRM

Mathieu Lederlin
CHU Bordeaux

IRM

HTAP



Faut-il faire une IRM dans l'HTAP ?

ESC/ERS GUIDELINES

Eur Respir J 2009; 34: 1219–1263

DOI: 10.1183/09031936.00139009

Guidelines for the diagnosis and treatment of pulmonary hypertension

The Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS) endorsed by the International Society of Heart and Lung Transplantation (ISHLT)

N. Galiè, M.M. Hoeper, M. Humbert, A. Torbicki, J.-L. Vachiery, J.A. Barbera, M. Beghetti, P. Corris, S. Gaine, J.S. Gibbs, M.A. Gomez-Sanchez, G. Jondeau, W. Klepetko, C. Opitz, A. Peacock, L. Rubin, M. Zellweger and G. Simonneau

ESC/ERS GUIDELINES

N. GALIÈ ET AL.

and a specificity of 94–100%. While in PAH the ventilation/perfusion lung scan may be normal, it may also show small peripheral unmatched and mismatched defects in perfusion. Contrast-enhanced CT may be used as a complementary investigation but does not replace the ventilation/perfusion scan or traditional pulmonary angiogram. A caveat is that unmatched perfusion defects are also seen in PVOD.

7.1.7 High-resolution computed tomography, contrast-enhanced computed tomography and pulmonary angiography

High-resolution CT provides detailed views of the lung parenchyma and facilitates the diagnosis of interstitial lung disease and emphysema. High-resolution CT may be very helpful where there is a clinical suspicion of PVOD. Characteristic changes of interstitial oedema with diffuse central ground-glass opacification and thickening of interlobular septa suggest PVOD, additional findings may include lymphadenopathy and pleural effusion [64]. Pulmonary capillary haemangiomatosis is suggested by diffuse bilateral thickening of the interlobular septa and the presence of small, centrilobular, poorly circumscribed nodular opacities.

Contrast CT angiography of the PA is helpful in determining whether there is evidence of surgically accessible CTEPH. It can delineate the typical angiographic findings in CTEPH such as complete obstruction, bands and webs, and intraluminal irregularities as accurately and reliably as digital subtraction angiography [55, 56]. With this technique, collaterals from bronchial arteries can be identified.

Traditional pulmonary angiography is still required in many centres for the work-up of CTEPH to identify patients who may benefit from PEA [22]. Angiography can be performed safely by experienced staff in patients with severe PH using modern contrast media and selective injections. Angiography may also be useful in the evaluation of possible vasculitis or pulmonary arteriovenous malformations.

7.1.8 Cardiac magnetic resonance imaging

Cardiac magnetic resonance imaging provides a direct evaluation of RV size, morphology and function, and allows noninvasive assessment of blood flow including stroke volume, CO, distensibility of PA, and RV mass [57]. Cardiac magnetic resonance data may be used to evaluate right heart haemodynamics particularly for follow-up purposes. A decreased stroke volume, an increased RV end-diastolic volume, and a decreased LV end-diastolic volume measured at baseline are associated with a poor prognosis. Among the trial of prognostic signs, increased RV end-diastolic volume may be the most appropriate marker of progressive RV failure in the follow-up [58].

7.1.9 Blood tests and immunochemistry

Routine biochemistry, haematology and thyroid function tests are required in all patients, as well as a number of other essential blood tests. Serological testing is important to detect underlying CTD, HIV and hepatitis. Up to 40% of patients with IPAH have elevated anti-nuclear antibodies, usually in low titre [18]. Systemic side effects is the most important CTD to exclude because this condition has a high prevalence of PAH. Anti-centromere antibodies are typically positive in limited scleroderma as are other anti-nuclear antibodies including

dsDNA, anti-Ro, U3-RNP, Scl-70 and U1-RNP. In the diffuse variety of scleroderma, U3-RNP is typically positive. In individuals with systemic lichen erythematosus, anti-cardiolipin antibodies may be found. Thrombophilia screening including anti-phospholipid antibodies, lupus anticoagulant and anti-cardiolipin antibodies should be performed in CTEPH. HIV testing is mandatory. Up to 2% of individuals with liver disease will manifest PAH and therefore liver function tests and hepatitis serology should be examined if clinical abnormalities were noted. Thyroid disease is commonly seen in PAH and should always be considered, especially if abrupt changes in the clinical course occur [60].

7.1.10 Abdominal ultrasound scan

Liver cirrhosis and/or portal hypertension can be reliably excluded by the use of abdominal ultrasound. The use of contrast agents and the addition of a colour-Doppler examination may improve the accuracy of the diagnosis [61]. Portal hypertension can be confirmed by the detection of an increased gradient between free and occluded (wedge) hepatic vein pressure at the time of RHC [62].

7.1.11 Right heart catheterisation and vasoreactivity

RHC is required to confirm the diagnosis of PAH, to assess the severity of the haemodynamic impairment and to test the vasoreactivity of the pulmonary circulation. When performed at experienced centres, RHC procedures have low morbidity (1.1%) and mortality (0.05%) rates [63]. The following variables must be recorded during RHC: PAP (systolic, diastolic and mean), right atrial pressure, Pwoc and RV pressure. CO must be measured in triplicate preferably by thermodilution or by the Fick method, if oxygen consumption is assessed. The Fick method is mandatory in the presence of a systemic-to-pulmonary shunt. Superior vena cava, PA and systemic arterial blood oxygen saturations should also be determined. These measurements are needed for the calculation of PVR. Adequate recording of Pwoc is required for the differential diagnosis of PH due to left heart disease. In rare cases, left heart catheterisation may be required for direct assessment of LV end-diastolic pressure. A Pwoc >15 mmHg excludes the diagnosis of pre-capillary PAH. One of the most challenging differential diagnoses of PAH is heart failure with normal LV ejection fraction and diastolic dysfunction (see also section 1.1.9). In this population, Pwoc may be mildly elevated or at the higher end of the normal range at rest. Exercise haemodynamics or dobutamine challenge can show a disproportionate increase in Pwoc, although the relevance of the finding remains to be established. Coronary angiography may be required in the case of the presence of risk factors for coronary artery disease and angina or in case of testing for double lung transplantation or PEA in patients with CTEPH.

In PAH vasoreactivity testing should be performed at the time of diagnostic RHC to identify patients who may benefit from long-term therapy with calcium channel blockers (CCBs) (see also section 7.3.3) [65, 66]. Acute vasodilator challenge should only be performed with short-acting, safe and easy to administer drugs with no or limited systemic effects. Currently the agent most used in acute testing is NO (table 9) [66]. Based on previous experience [65, 67, 68] in epoprostenol or its analogue may also be used as an alternative (but with a risk of systemic vasodilator effects) (table 10).

7.1.8 Cardiac magnetic resonance imaging

Cardiac magnetic resonance imaging provides a direct evaluation of RV size, morphology and function, and allows noninvasive assessment of blood flow including stroke volume, CO, distensibility of PA, and RV mass [57]. Cardiac magnetic resonance data **may be** used to evaluate right heart haemodynamics particularly for follow-up purposes. A decreased stroke volume, an increased RV end-diastolic volume, and a decreased LV end-diastolic volume measured at baseline are associated with a poor prognosis. Among the triad of prognostic signs, **increased RV end-diastolic volume may be** the most appropriate marker of progressive RV failure in the follow-up [58].

Faut-il faire une IRM dans l'HTAP ?

Revue des Maladies Respiratoires (2010) 27, 141–150

REVUE GÉNÉRALE

Diagnostic et prise en charge de l'hypertension pulmonaire en 2009. Commentaires sur les nouvelles recommandations de l'European Society of Cardiology (ESC) et de l'European Respiratory Society (ERS)

Guidelines for the diagnosis and treatment of pulmonary hypertension

O. Sanchez^{a,*}, M. Humbert^b, O. Sitbon^b,
X. Jais^b, G. Simonneau^b



IRM = 0

IRM

HTAP

Morphologie

Fonction VD/VC

Tagging myoca

Vélocimétrie

Angiographie

Perfusion

Rehaussement tardif



Diagnostic positif

Physiopathologie

Diagnostic étiologique

Diagnostic d'opérabilité

Pronostic

Suivi sous traitement

IRM

Morphologie

Fonction VD/VG

Tagging myocardique

Vélocimétrie

Angiographie

Perfusion

Rehaussement tardif

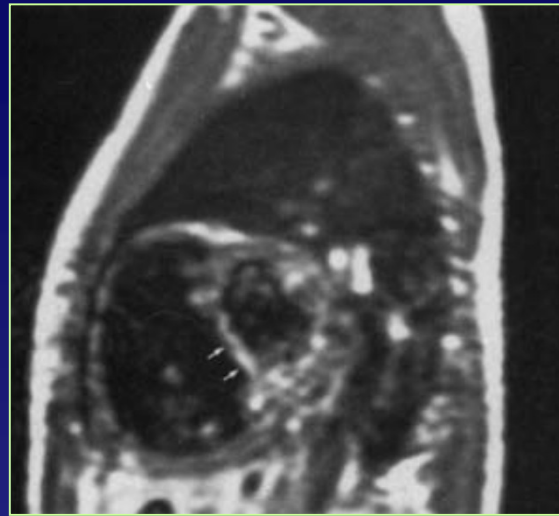
IRM

Morphologie

Fonction VD/VG

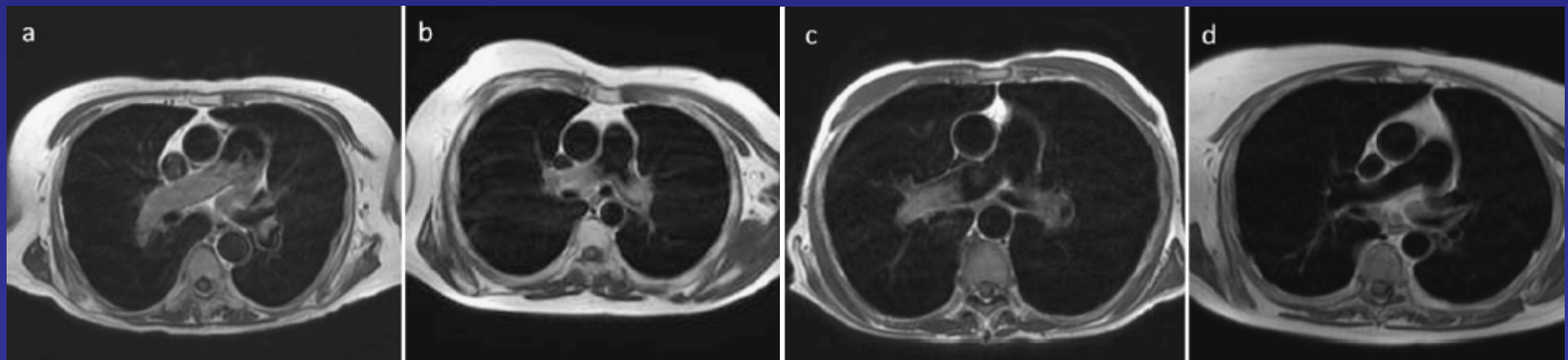
Tagging myocardique

Vélocimétrie



“Direct quantitation of right and left ventricular volumes with nuclear magnetic resonance imaging in patients with primary pulmonary hypertension”.

Boxt LM, JACC 1992



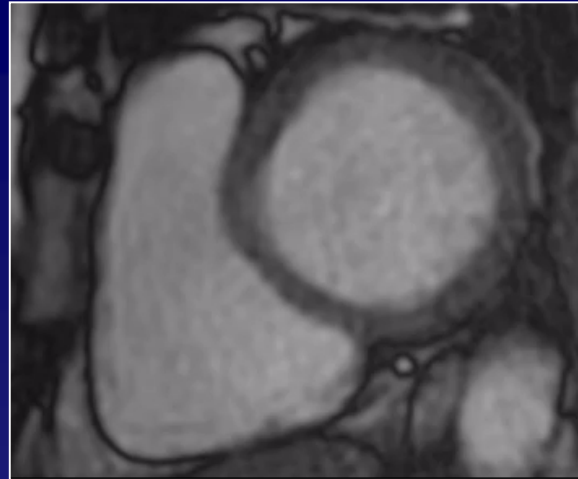
“Black blood MRI has diagnostic and prognostic value in the assessment of patients with pulmonary hypertension”.

Swift AJ, Eur Radiol 2012

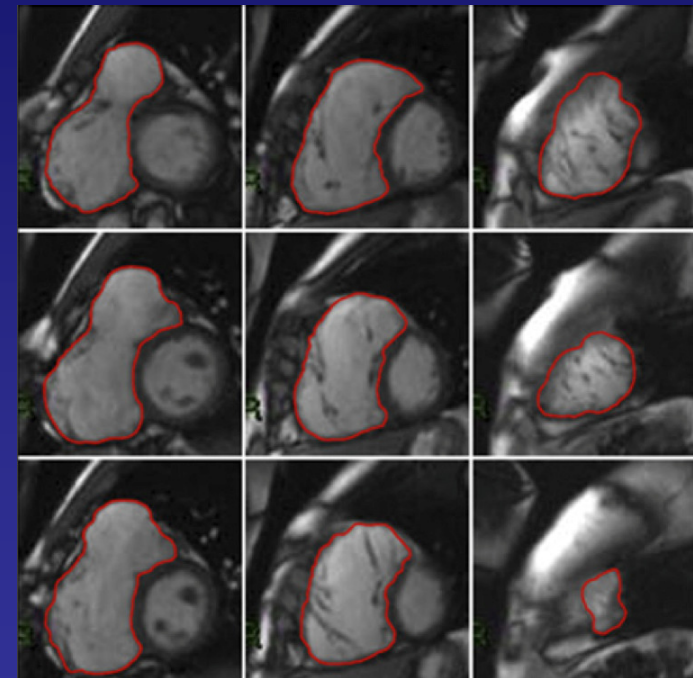
IRM

Morphologie

Fonction VD/VG



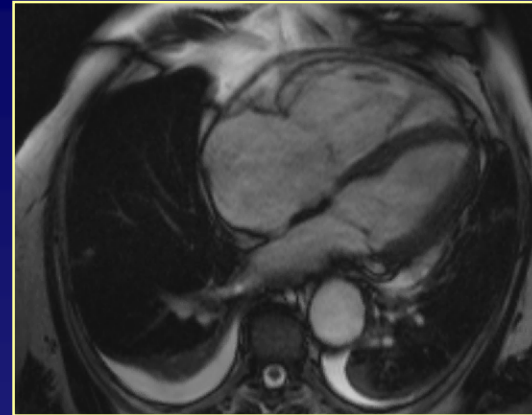
- séquences ciné SSFP
- segmentation radiale de l'espace k
- couverture de l'ensemble du VD



Right Ventricle - Absolute				
Cardiac Function			Normal Range (M) (MRI)	Units
Ejection Fraction	EF	63.9	47.00 ... 74.00	%
End Diastolic Volume	EDV	126.4	88.00 ... 227.00	ml
End Systolic Volume	ESV	45.6	23.00 ... 103.00	ml
Stroke Volume	SV	80.8	52.00 ... 138.00	ml
Cardiac Output	CO	4.77	2.82 ... 8.82	l/min
End Diastolic Volume	EDV	59.3	55.00 ... 105.00	ml/m ²
End Systolic Volume	ESV	21.4	15.43 ... 42.91	ml/m ²
Stroke Volume	SV	37.9	32.00 ... 64.00	ml/m ²
Cardiac Index	CI	2.24	1.74 ... 4.20	l/min/m ²

→ Volumétrie ventriculaire plus précise et plus reproductible qu'en échocardiographie
Mertens LL, Nat Rev Cardiol 2010

IRM



Morphologie

Fonction VD/VG

Tagging myocardique

Vélocimétrie

Angiographie

Perfusion

Rehaussement tardif

L'IRM permet de quantifier précisément :

- La dilatation du VD
- L'hypertrophie du VD
- La dysfonction systolique du VD
- Le défaut de remplissage du VG

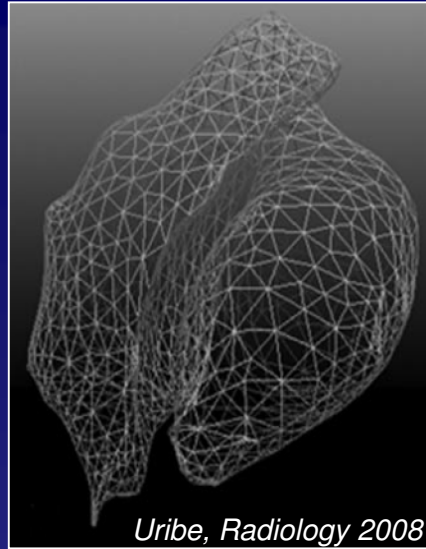
Meilleurs indices prédictifs de mortalité :

- Volume éjectionnel du VD $\leq 25 \text{ mL/m}^2$
- VTD VD $\geq 84 \text{ mL/m}^2$
- VTD VG $\leq 40 \text{ mL/m}^2$

IRM

Morphologie

Fonction VD/VG



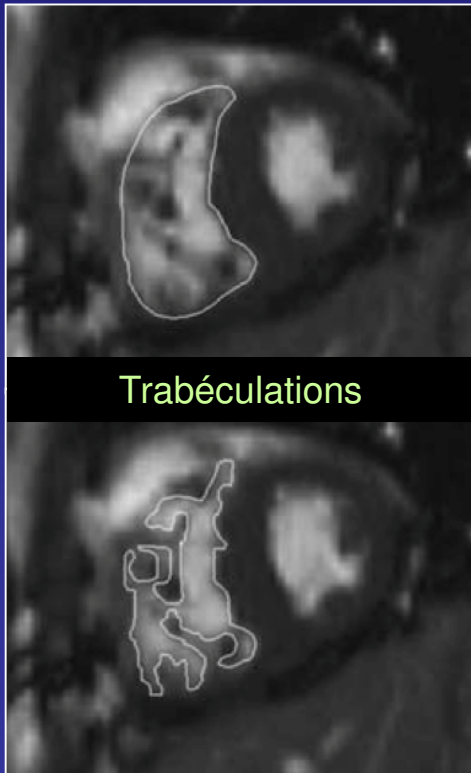
Evaluation du cœur droit à l'IRM

Paroi fine, géométrie complexe (VD moulé sur le VG)

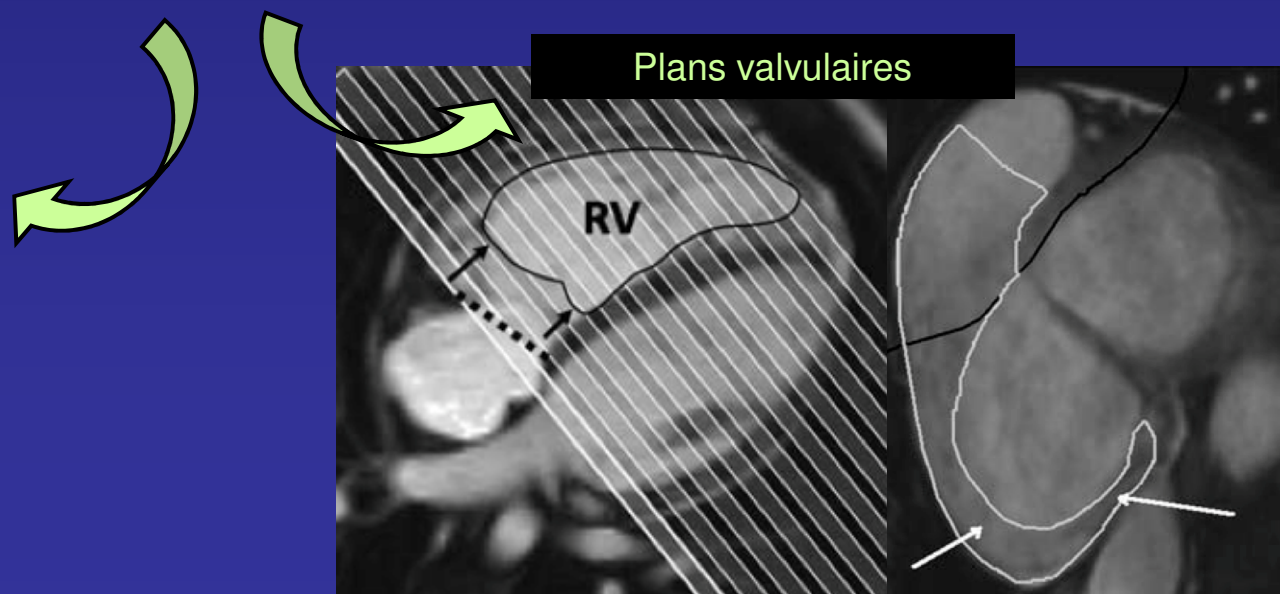
Même débit que VG avec 1/6 de sa masse

VTD plus important

Ejection débute plus tôt, finit plus tard, liée au raccourcissement longitudinal



Causes de variabilité des mesures



IRM

Morphologie

Fonction VD/VG

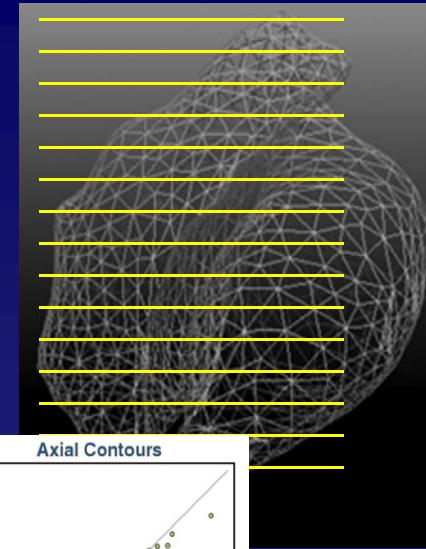
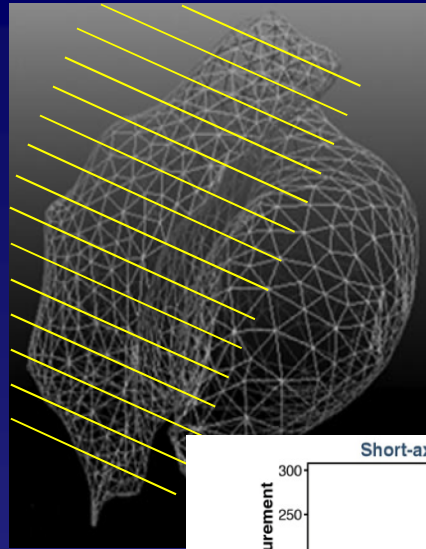
Tagging myocardique

Véloc

Angi

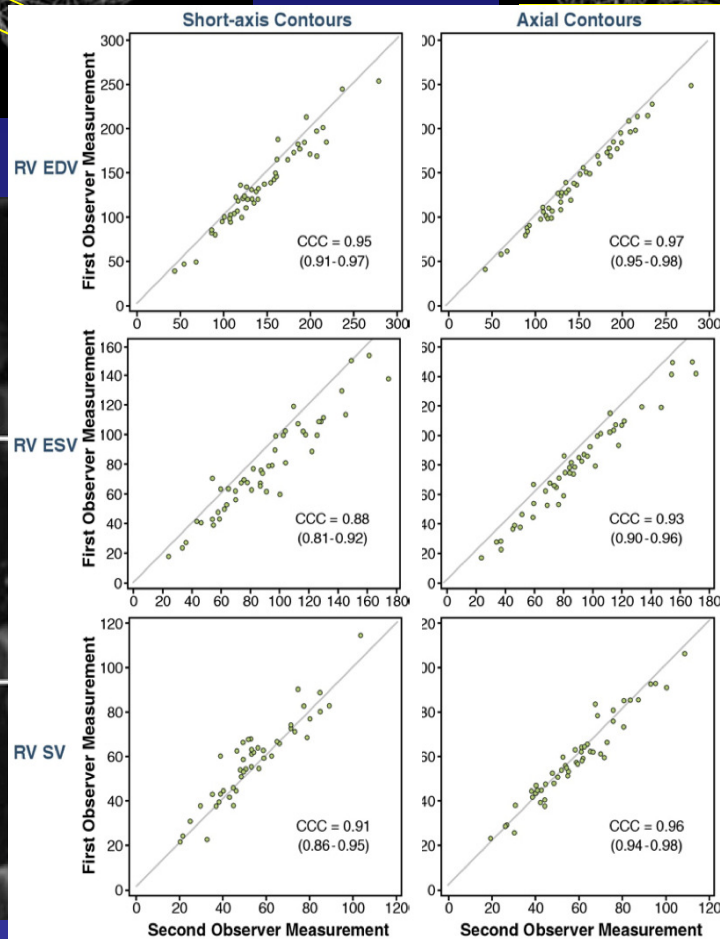
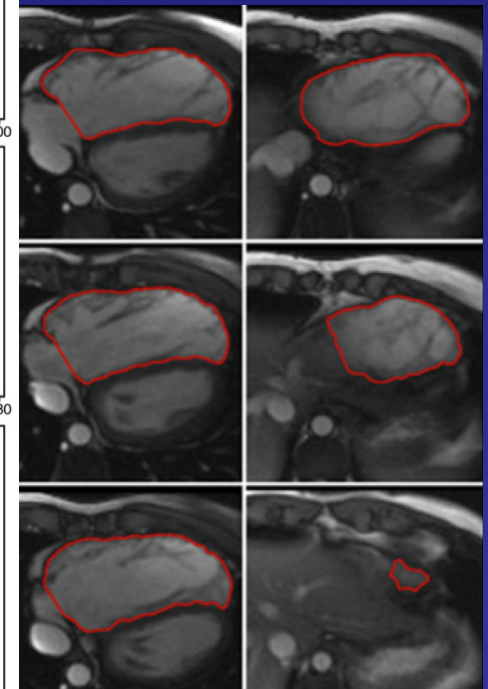
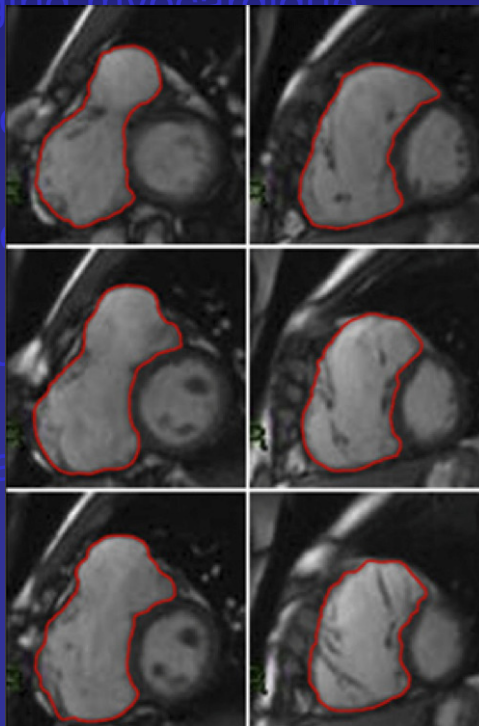
Perfu

Reha



Short axis

Axial plane



Clarke CJ, JACC Img 2012; 5:28-37

IRM

Autres applications des séquences SSFP

Morphologie

Fonction VD/VG

Tagging myocardique

Vélocimétrie

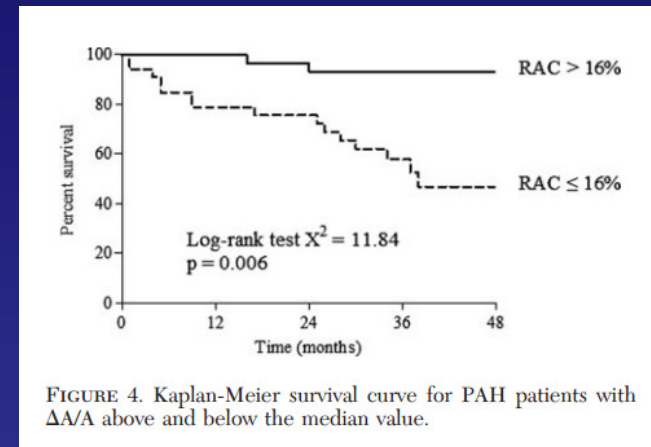
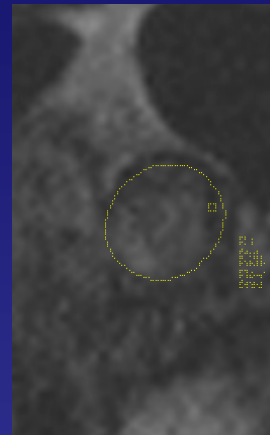
Angiographie

Perfu

Reha

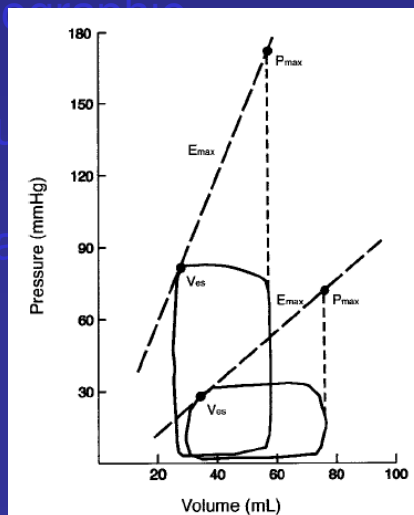
Etude de la distensibilité du TAP

La rigidité du TAP est un facteur prédictif de mortalité



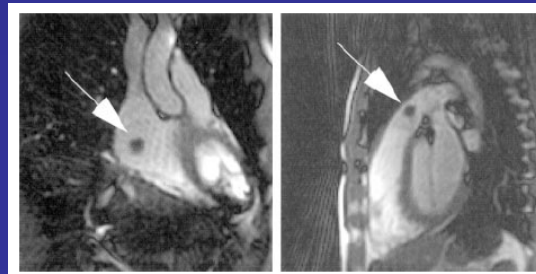
Gan CT, Chest 2007

Sanz J, JACC Cardiovasc Imaging 2009

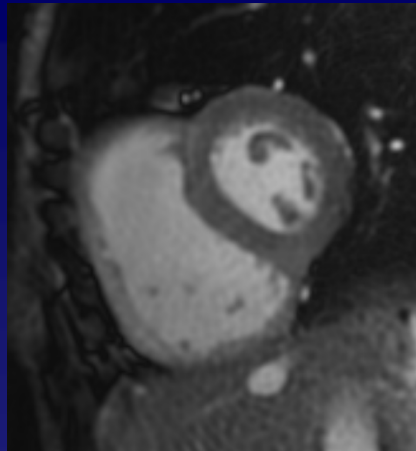


Etude de l'élastance du VD

Courbes pression-volume établies à partir de cathéter MR-guidés

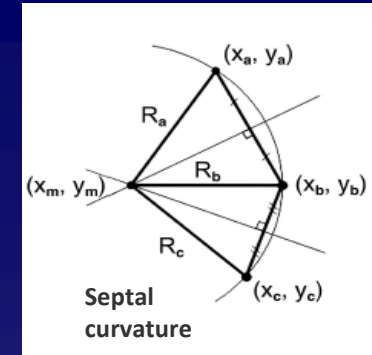


Kuehne T, Circulation 2004



Septum paradoxal

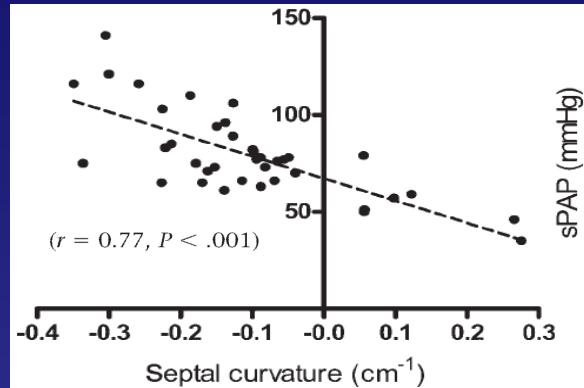
Relation linéaire entre la courbure septale et le gradient de pression transseptal (Loi de Laplace)



Roeleveld, Radiology 2005

Fonction VD/VG

Tagging myocardique



Detailed Calculation of Curvature

The three points on the septum are given by (a) x_a and y_a , (b) x_b and y_b , and (c) x_c and y_c (see Fig A1). The radius R of the circle, which passes through these three points, is to be calculated. The distance R_a between the middle (m) of this circle and the septal point (x_a, y_a) is

$$R_a = \sqrt{(x_a - x_m)^2 + (y_a - y_m)^2} \quad (A1)$$

Similar equations are written for R_b and R_c . Because the radius of the circle equals R at all locations, $R_a = R_b$. This yields Equation (A2), which is linear in x_m and y_m :

$$(x_a - x_b)^2 + (y_a - y_b)^2 - 2(x_c - x_b)(x_a - x_b) - 2(y_c - y_b)(y_a - y_b) = 2(x_c - x_b)(x_m - x_b) + 2(y_c - y_b)(y_m - y_b) \quad (A2)$$

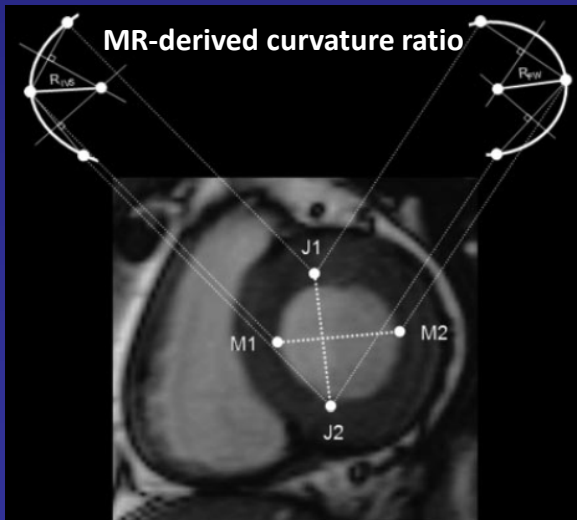
$$x_m = \frac{(x_a - x_b)^2 + (y_a - y_b)^2 - 2(x_c - x_b)(x_a - x_b) - 2(y_c - y_b)(y_a - y_b)}{2(x_c - x_b)} \quad (A3)$$

$$y_m = \frac{(x_a - x_b)(x_c - x_b) + (y_a - y_b)(y_c - y_b)}{x_c - x_b} \quad (A4)$$

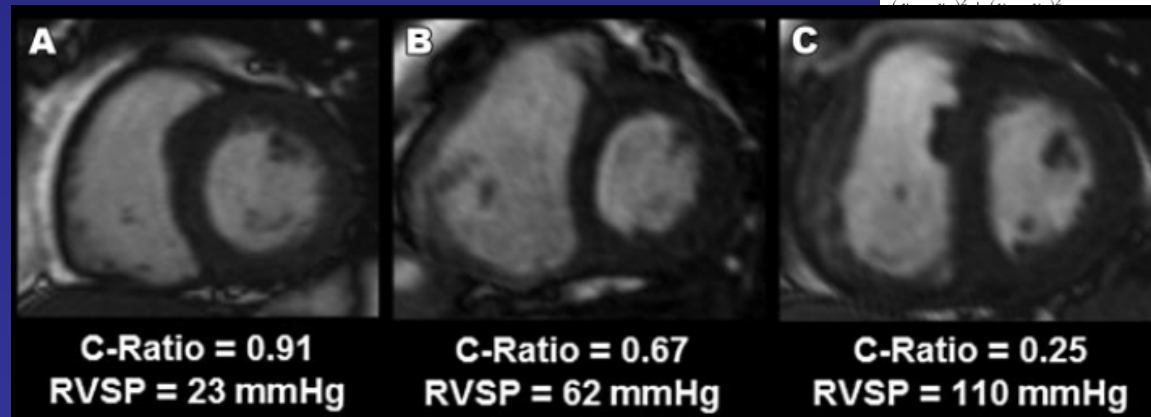
for x_m and y_m . The following equation is used to calculate the curvature R^{-1} :

$$R^{-1} = \frac{1}{R} = \frac{1}{\sqrt{(x_m - x_a)^2 + (y_m - y_a)^2}} \quad (A5)$$

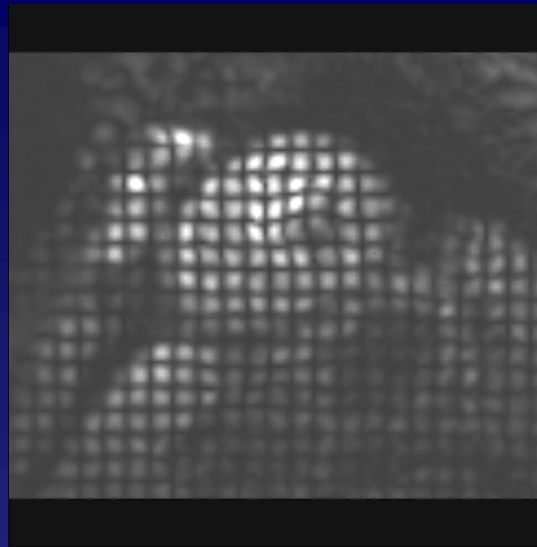
and y_b , and x_c and y_c were given in pixel numbers, then R needs to be multiplied by the pixel size in centimeters. The curvature equals R^{-1} . This procedure for calculating the curvature is easily programmed in Excel (Microsoft, Redmond, Wash), Matlab (release 13, version 6.5; MathWorks, Natick, Mass), or any programming language.



Dellegrottaglie, Radiology 2007



MR-derived curvature ratio : Se/Sp = 87/100 pour détecter une augmentation de la pression du VD



Etude de l'asynchronisme VD-VG

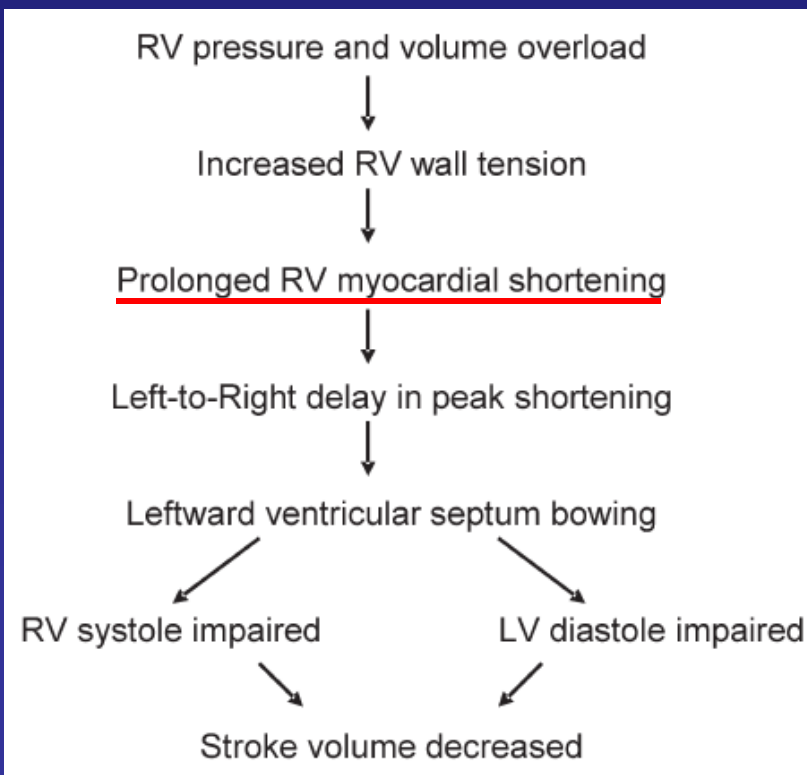
Tagging : étude de la contrainte pariétale des ventricules pour expliquer l'origine de l'asynchronisme VD-VG

Hypothèses testées :

~~contraction VD retardée ?~~

contraction VD prolongée ?

Marcus JT, JACC 2008



↳ **Resynchronisation ???**

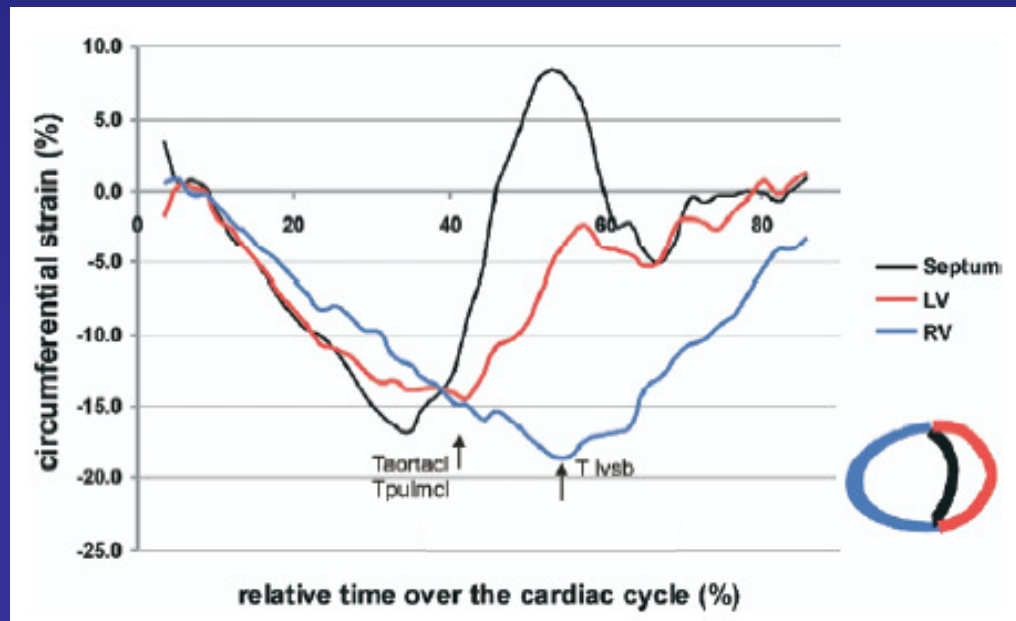


Figure 2

Circumferential Strain Over Time, PAH Patient

Circumferential strain curves over time after the electrocardiographic R-wave for the left ventricular (LV) and right ventricular (RV) free walls and the septum for 1 patient at basal level. The LV, RV, and septum start simultaneously with shortening (negative strain), but the RV reaches its peak later than the LV, by 12% of the cardiac cycle time. The closure times of aortic and pulmonary valves (T_{aortacl} and T_{pulmcl}) are coincident with the peak of LV shortening. The time of maximal leftward septal bowing (T_{lvsb}) is coincident with septal stretching (positive strain) and with the peak of RV shortening. The opening times of mitral and tricuspid valves ($T_{\text{mitr-op}}$ and $T_{\text{tric-op}}$) indicate the onset times of LV and RV filling. PAH = pulmonary arterial hypertension.

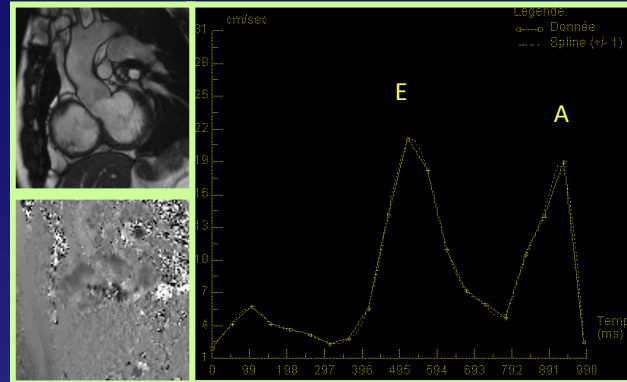
Figure 1

MRI Cine and Tagged Images

Three-chamber images (**top panels**), short-axis images (**middle panels**), and short-axis tagged images (**bottom panels**), at the time of aortic valve closure at trigger delay of 252 ms (**left**) and the time of peak right ventricular (RV) shortening at 341 ms (**right**). The 3-chamber images show that maximal leftward septal bowing occurs at 341 ms, well after aortic valve closure. In the tagged image at 341 ms, the distance of the tagging lines in the RV free wall show further shortening (**thick white arrows**), whereas the tagging lines in the left ventricular (LV) free wall show relaxation. MRI = magnetic resonance imaging.

IRM

Vélocimétrie tricuspide



Exploration fct° diastolique du VD :

$E/A < 1$: anomalie de la relaxation VD

$E/A > 2$: altération de la compliance VD

Kroft, Radiology 2000

Estimation PAP :

Mesure de la FR tricuspide

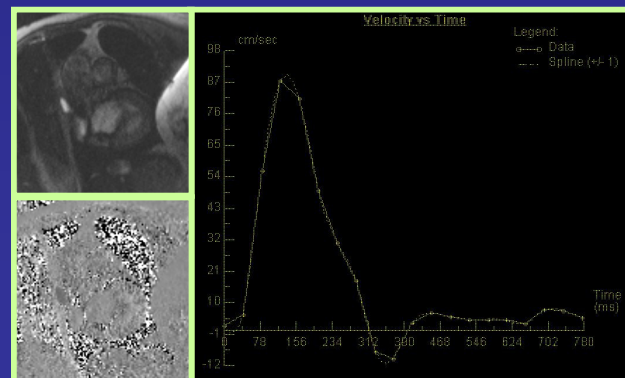
Vélocimétrie

Angiographie

Perfusion

Rehaussement tardif

Vélocimétrie AP



Exploration fct° systolique du VD :

Mesure du débit VD

Kondo, Radiology 1992

Estimation PAP :

Mesure des vitesses dans AP

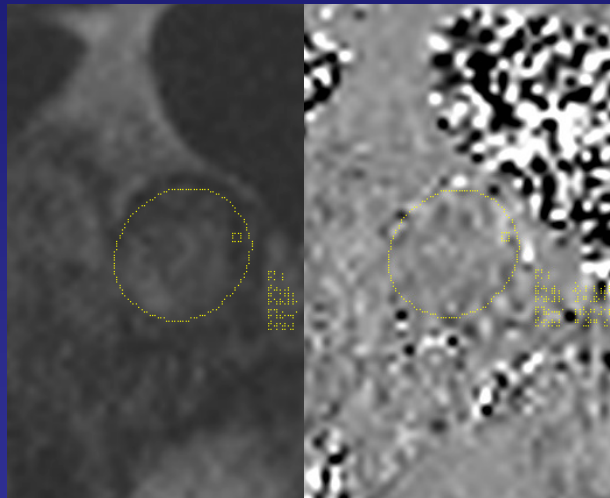
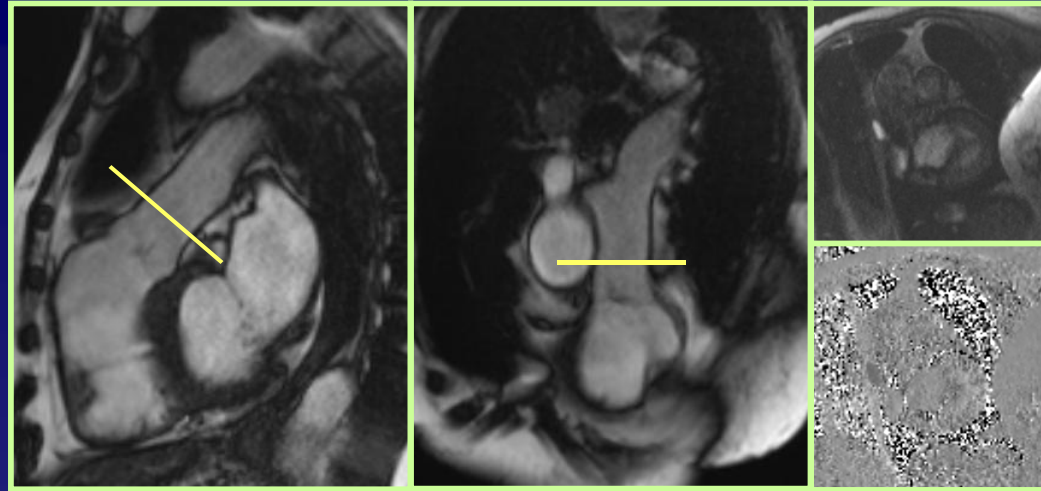
Tardivon, AJRCCM 1994

Mousseaux, Radiology 1999

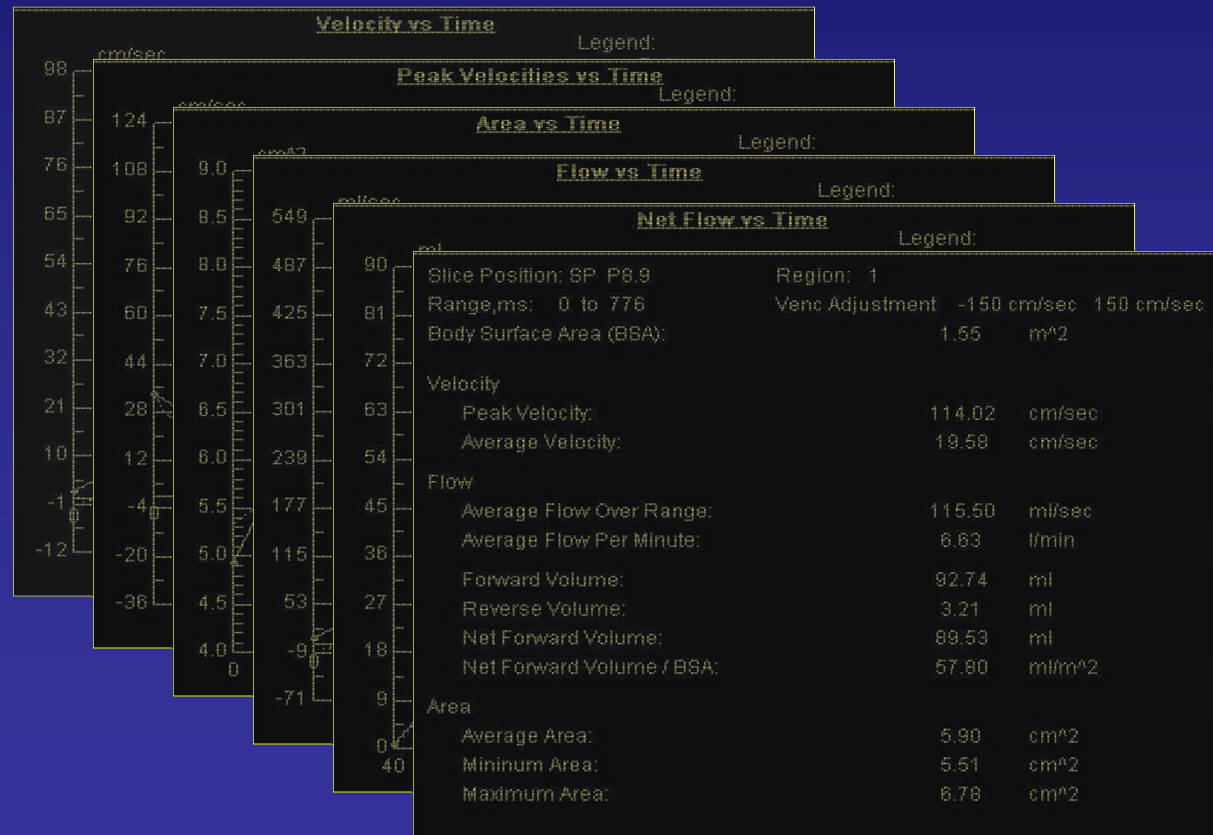
Laffon, J Appl Physiol 2003

IRM

Vélocimétrie AP



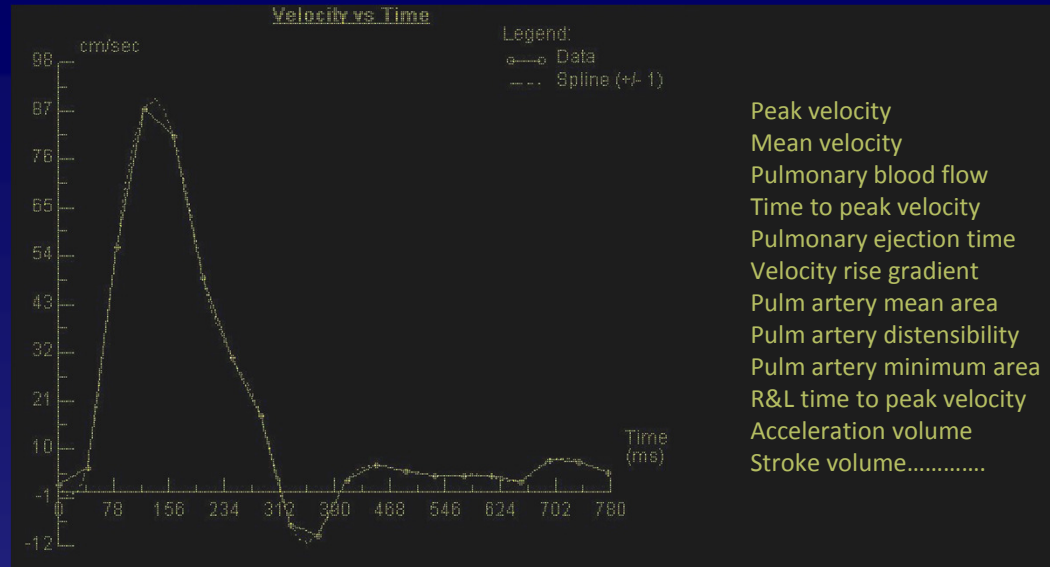
Rehaussement tardif



IRM

Morphologie

Fonction VD/VG



Phase-Contrast MR Imaging Data in Study Group and PAH Subgroup

Parameter	Patients	Patients	
Peak velocity			
Average velocity			
Maximum velocity			
Minimum velocity			
Average velocity during AT	38.4 (16.5)	16.9 (8.7)	<.001
Average velocity during ET	29.4 (12.4)	14.6 (6.8)	<.001

Correlation Coefficients between Pulmonary Pressures and Resistances

Parameter	PAP
Peak velocity	0.37*
Average velocity	0.73
Maximum velocity	0.61
Minimum velocity	0.67
Average velocity during AT	0.65
Average velocity during ET	0.52
AT (ms)	0.35*
ET (ms)	0.17†
AT/ET	0.28*
Average velocity during AT	-0.71
Average velocity during ET	-0.74

Limites de la vélocimétrie :

- Variabilité des mesures en fct° :
 - * degré d'inspiration (diminut° du débit cardiaque en insp. bloquée)
 - * acquisition sur l'ensemble du cycle ou non
- Reproductibilité insuffisamment évaluée

Futur :

- acquisit° en respiration libre
- 4D flow measurements...

IRM

Morphologie

Fonction VD/VG

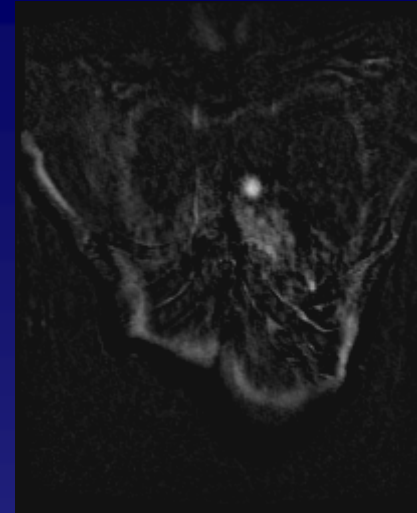
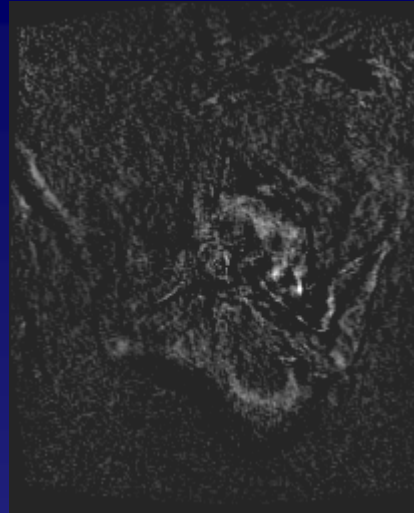
Tagging myocardique

Vélocimétrie

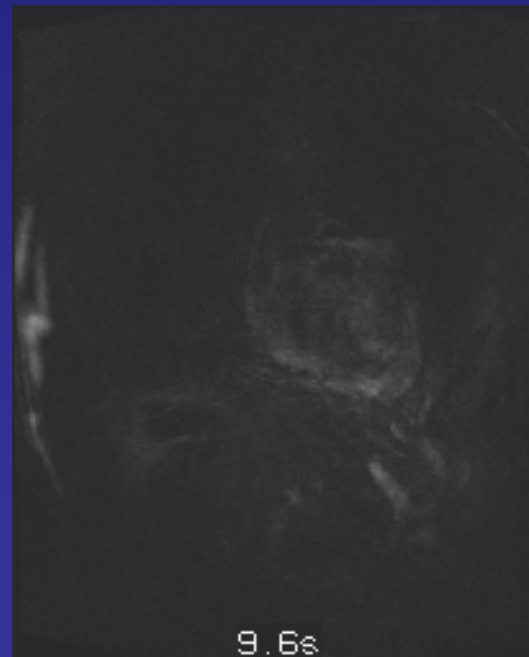
Angiographie

Perfusion

Rehaussement tardif



Angio-MR : précision jusqu'aux artères segmentaires



Angio 4D



Diagnostic HTAP primitive vs. HTPPE sur 29 patients :

- Diagnostic correct dans 90% des cas
- Concordance interobs : $\kappa = 0.70$

Nikolaou K, Radiology 2005

Angiographie

Perfusion

Rehaussement tardif

MRA vs. CTA (29 patients HTPPE) :

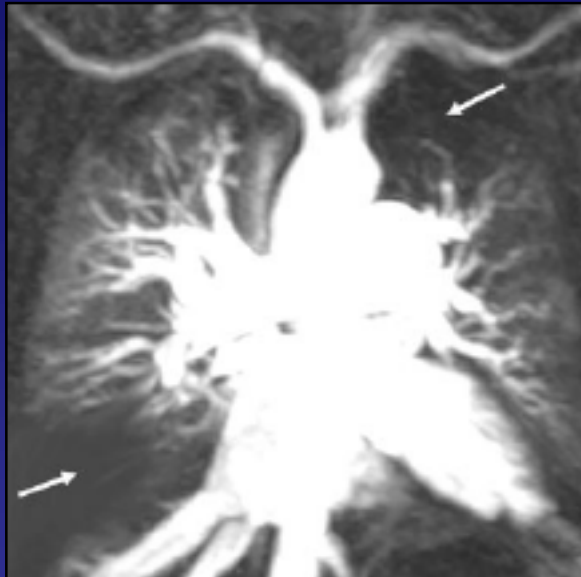
- CT=MR pour occlusions segmentaires
- CT>MR pour thrombi pariétaux, webs & bands, et artères sous-segmentaires

Ley S, Eur Radiol 2003

IRM

Les paramètres de perfusion pulmonaire (débit moyen, volume sanguin et temps de transit) sont significativement corrélés à la mPAP

Ohno Y, JMRI 2008



Perfusion

Rehaussement tardif

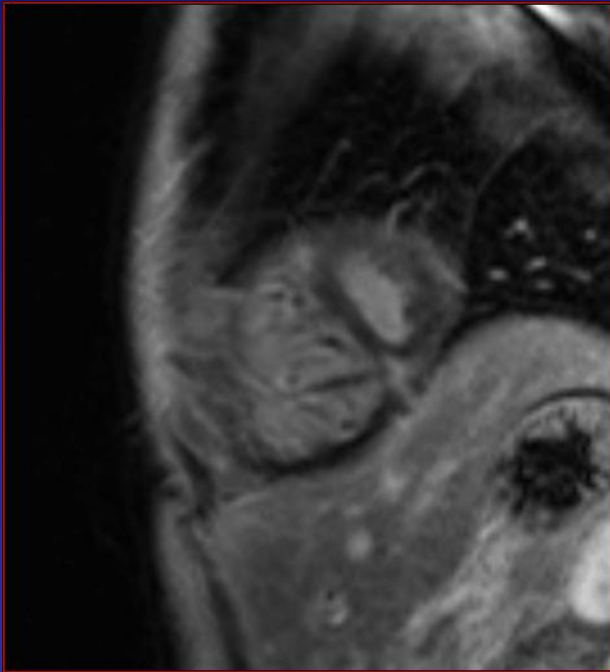
Seul paramètre significatif : augmentation du temps de transit moyen
Corrélation modérée entre PBV, MTT et mPAP

Ley S, Eur J Radiol 2007

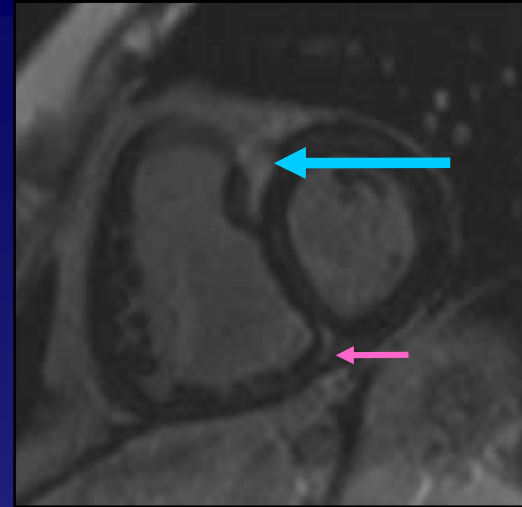
MR perfusion > CTA pour évaluer la réponse au traitement médical dans HTPPE

Ohno Y, JMRI 2012

IRM



Rehaussement tardif



Rehaussement tardif :

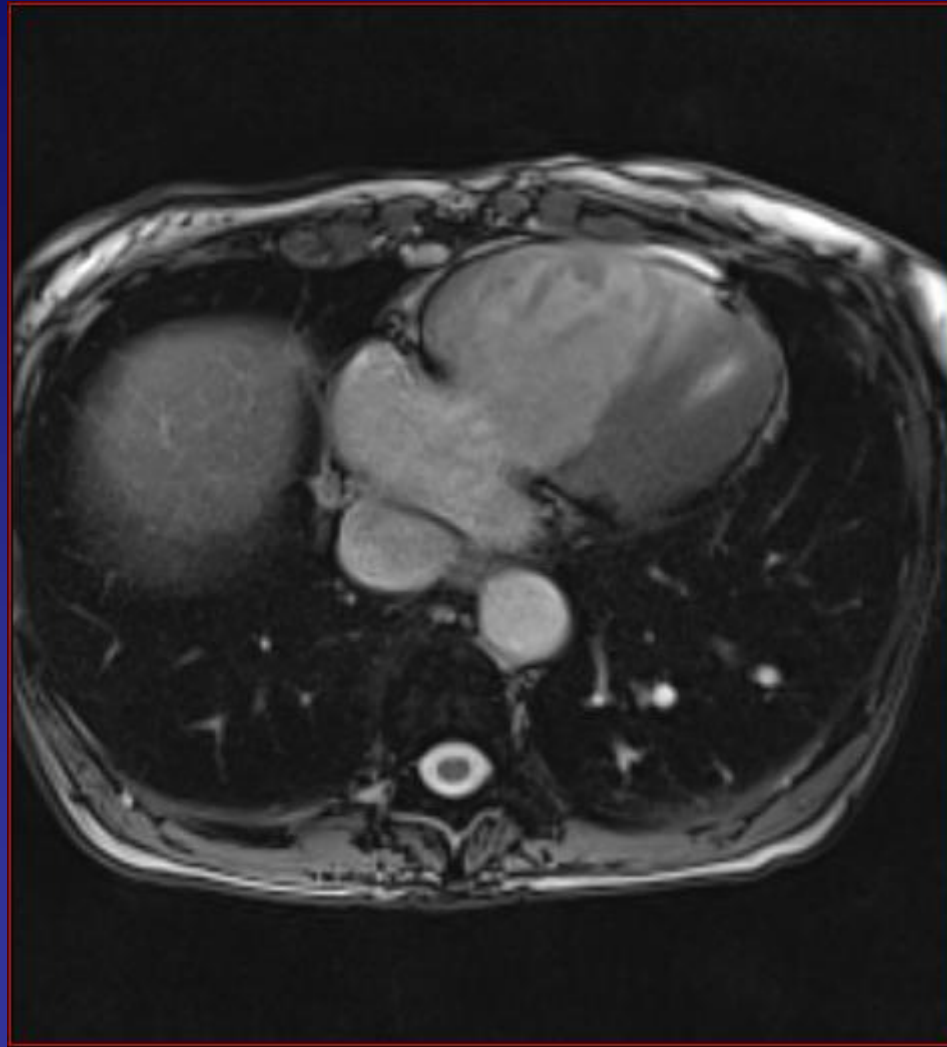
- Points d'insertion du VD sur le VG
- Septum interventriculaire

(= foyers de fibrose au sein du myocarde)

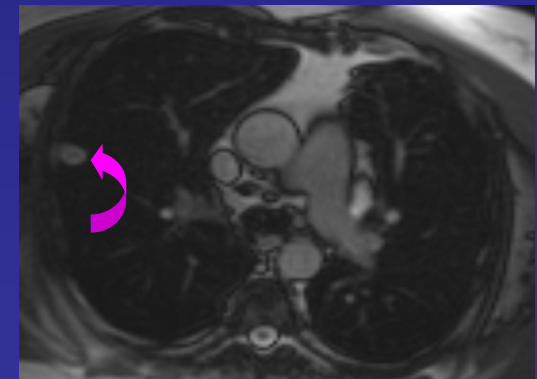
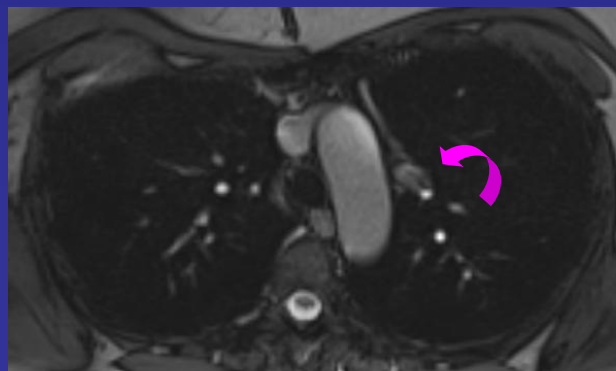
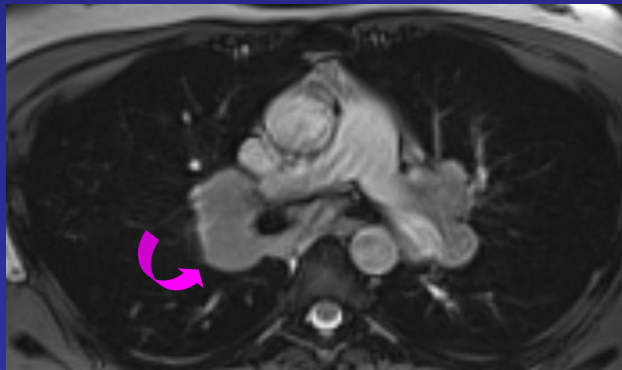
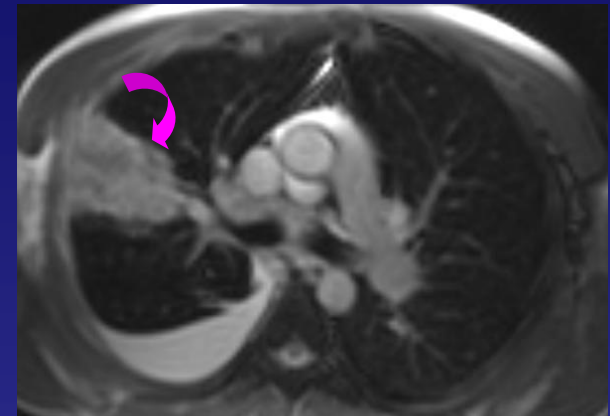
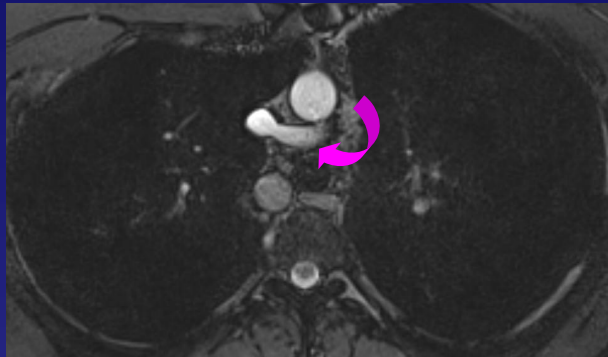
Corrélé avec mPAP, PVR, VTDVD...

Mais non spécifiques de l'HTAP

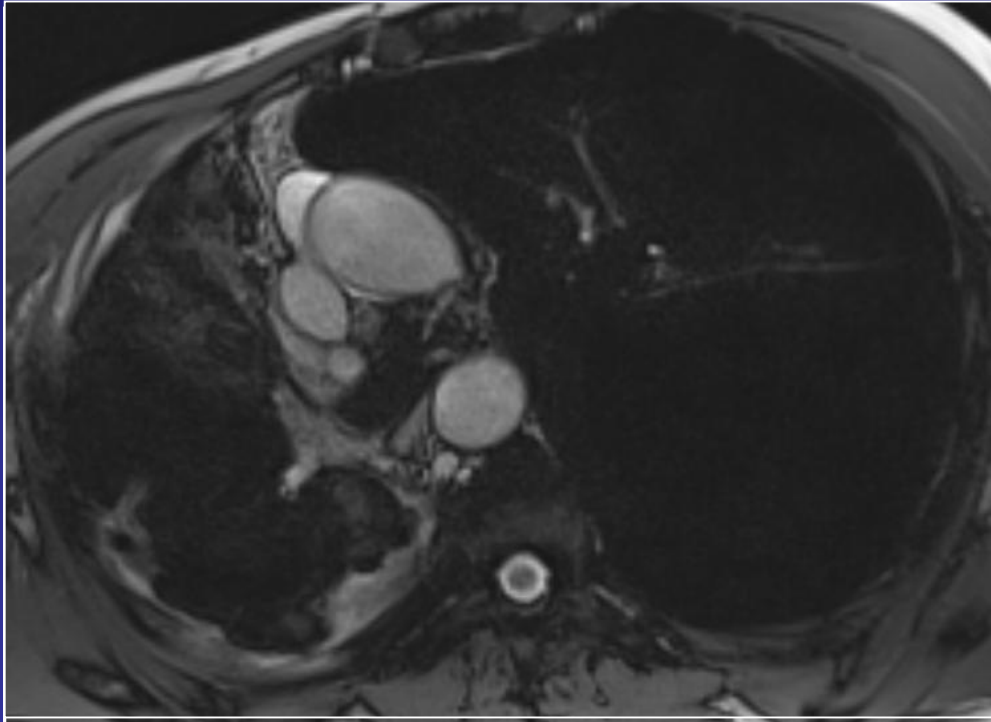
En pratique... IRM cardio-pulmonaire



En pratique... IRM cardio-pulmonaire



Pour la pratique



Bilan d'HTAP

Emphysème
centrolobulaire

Conclusion

IRM

HTAP

