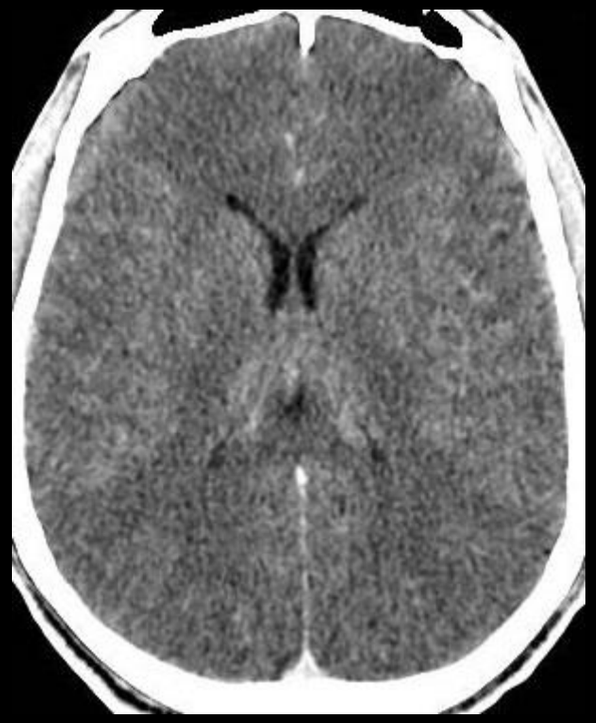
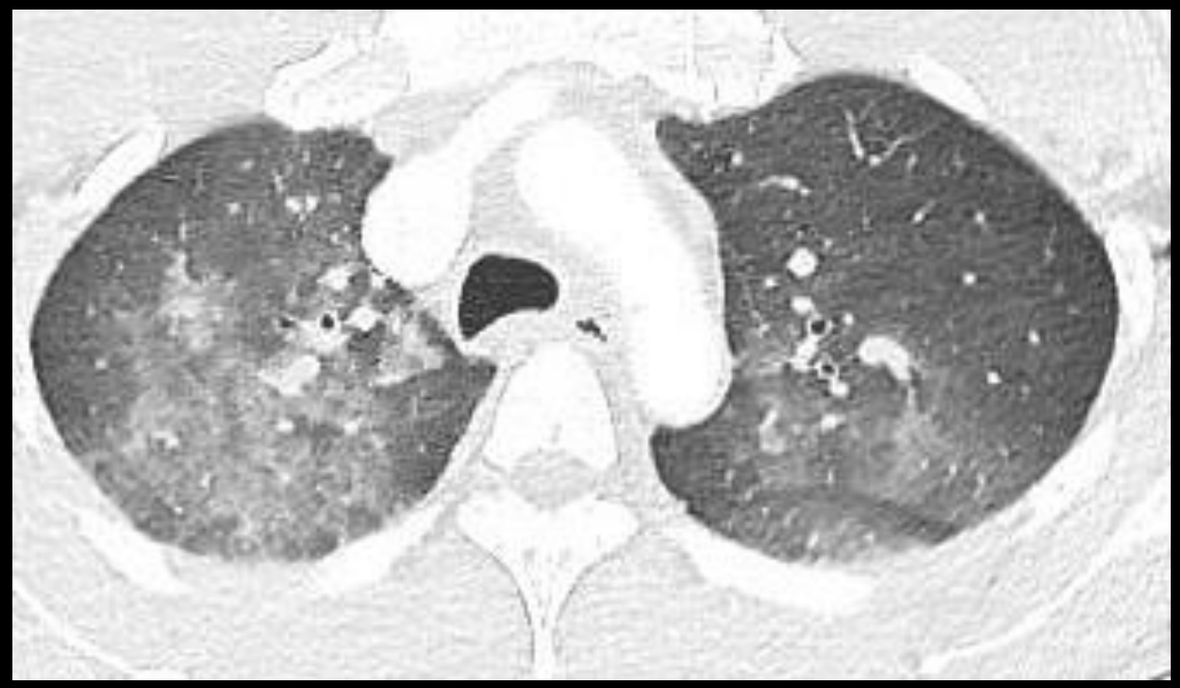
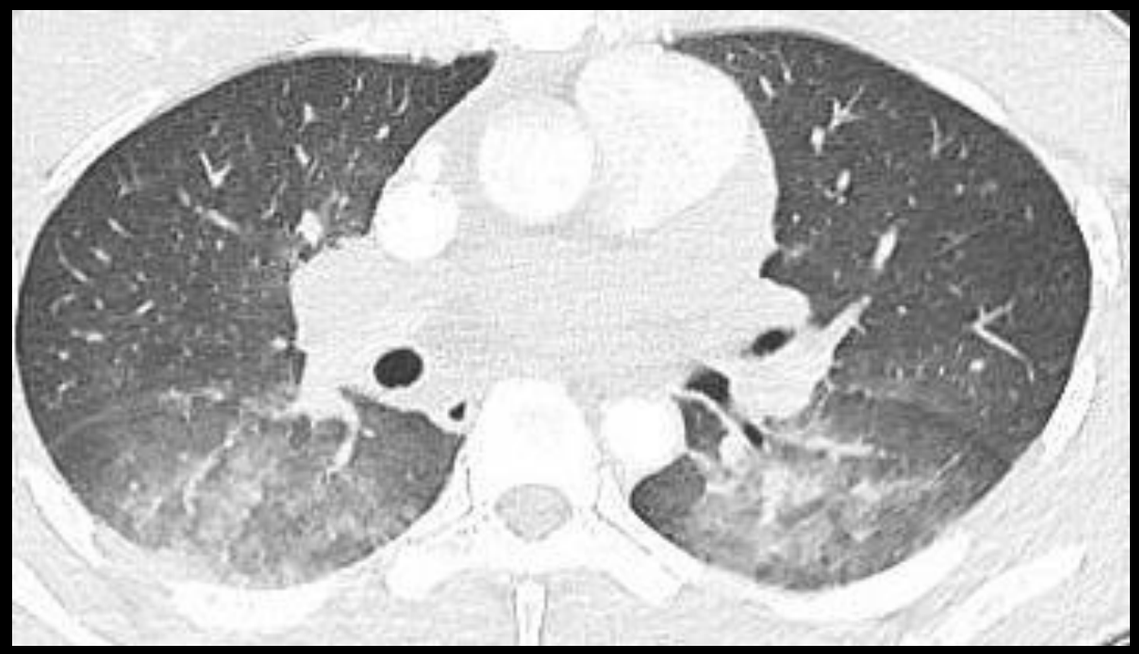
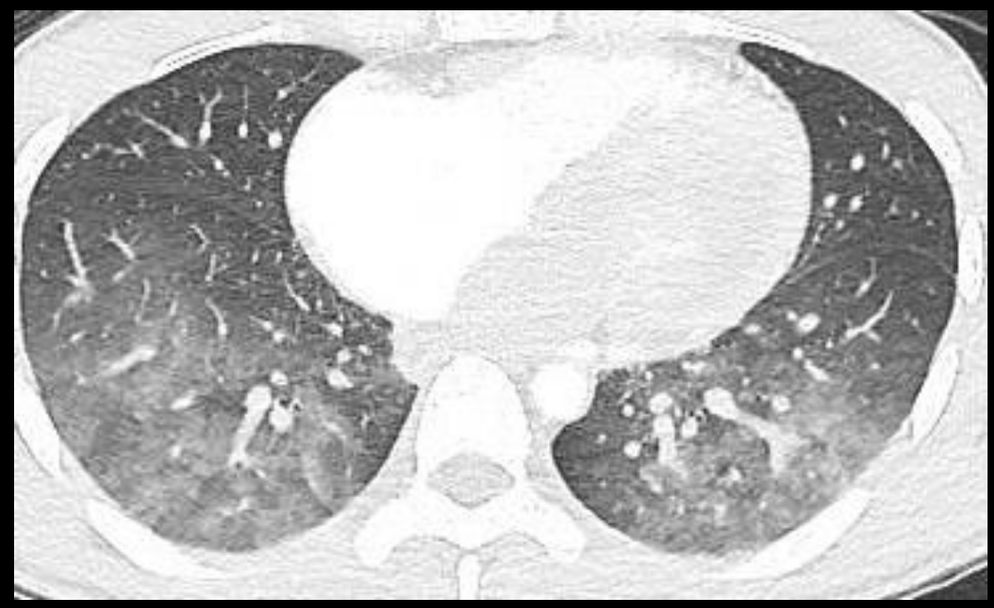
Patient de 31 ans, sans antécédent, originaire de Côte d'Ivoire, rentré d'Inde 4jours auparavant où il avait passé 3 ans, arrêt cardiaque devant des amis vers 17h, notion de céphalées le matin. 5' de no flow, 75' de low flow, plusieurs chocs électriques externes, TV réfractaire. Coronarographie normale, scanner vers 20h30

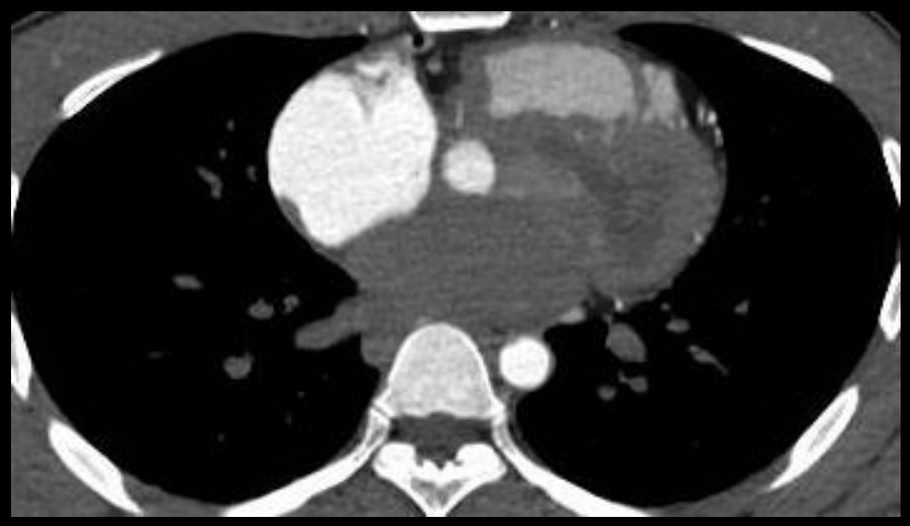


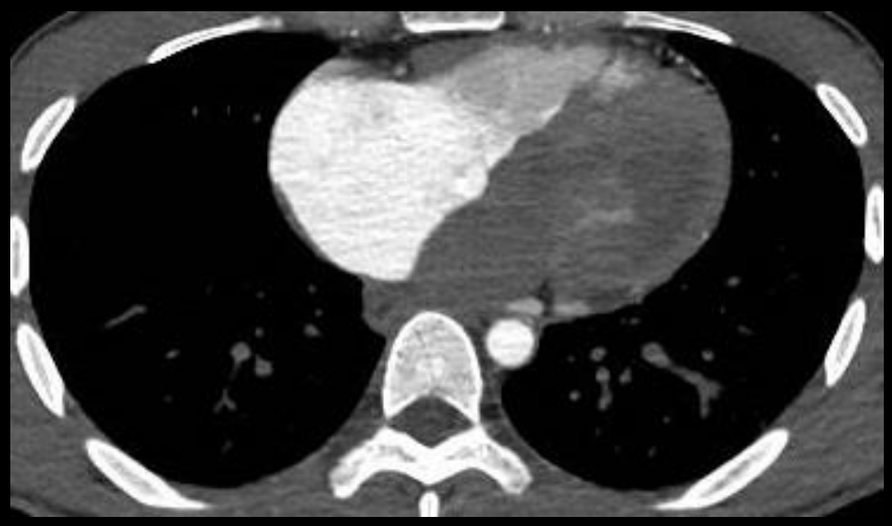


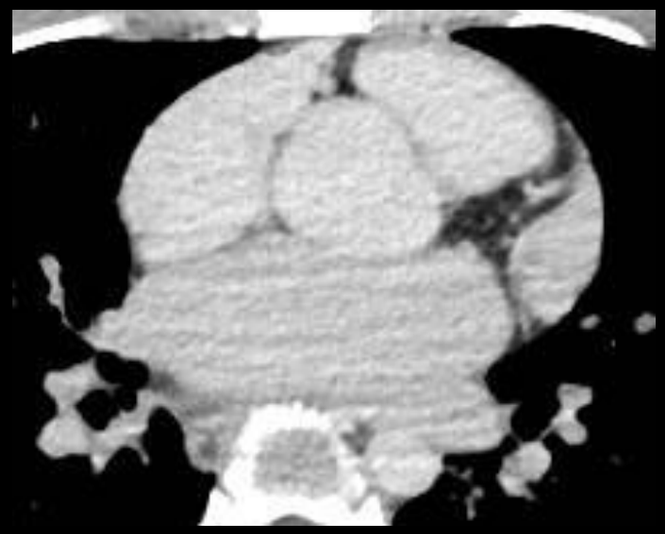


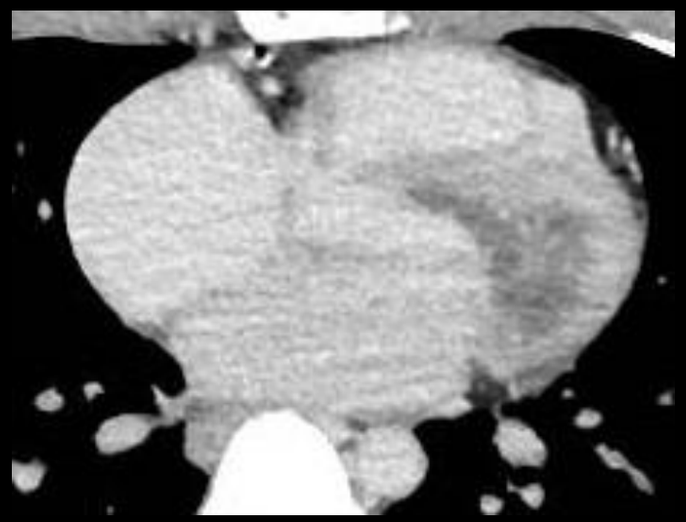


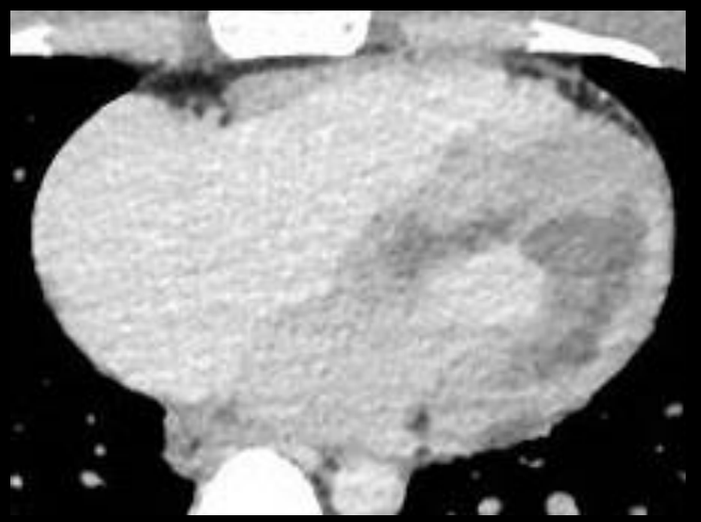
Alexia Savignac IHN

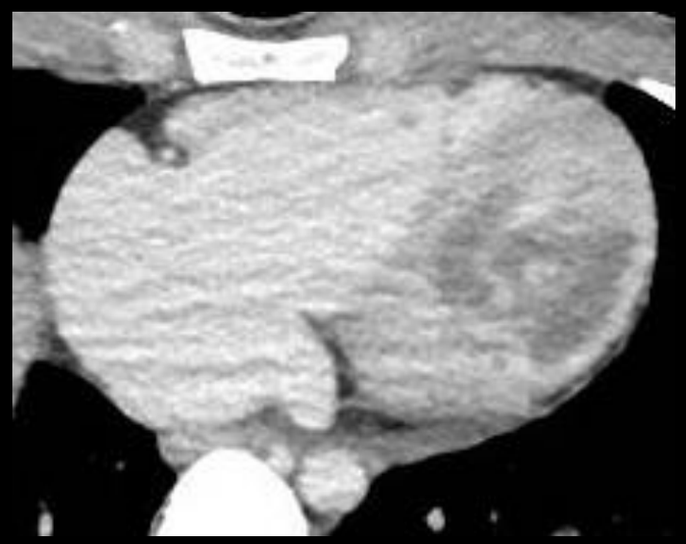


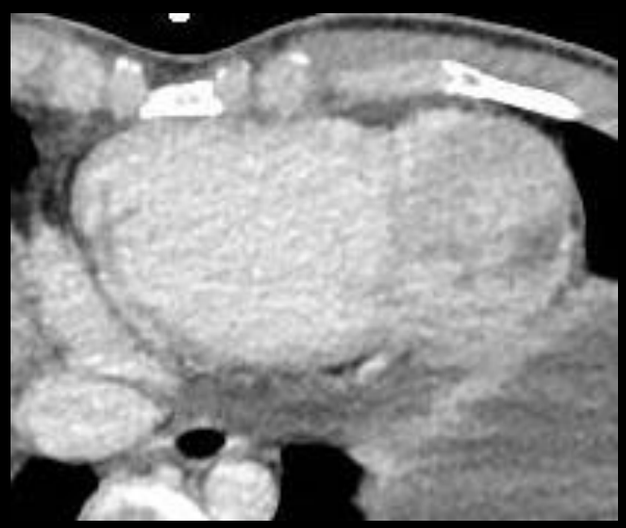


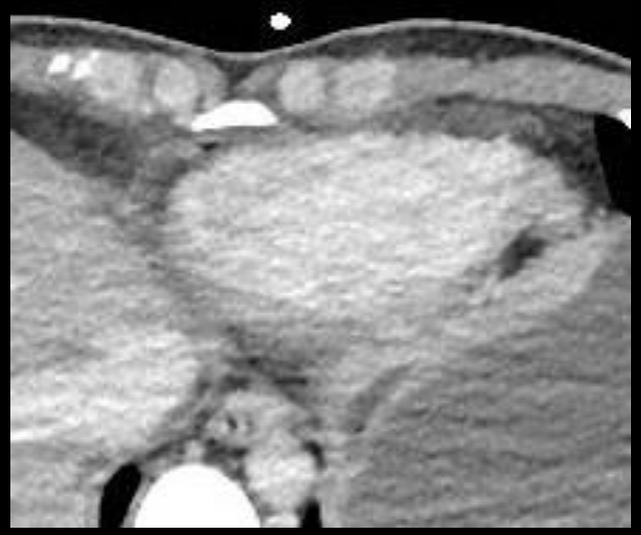


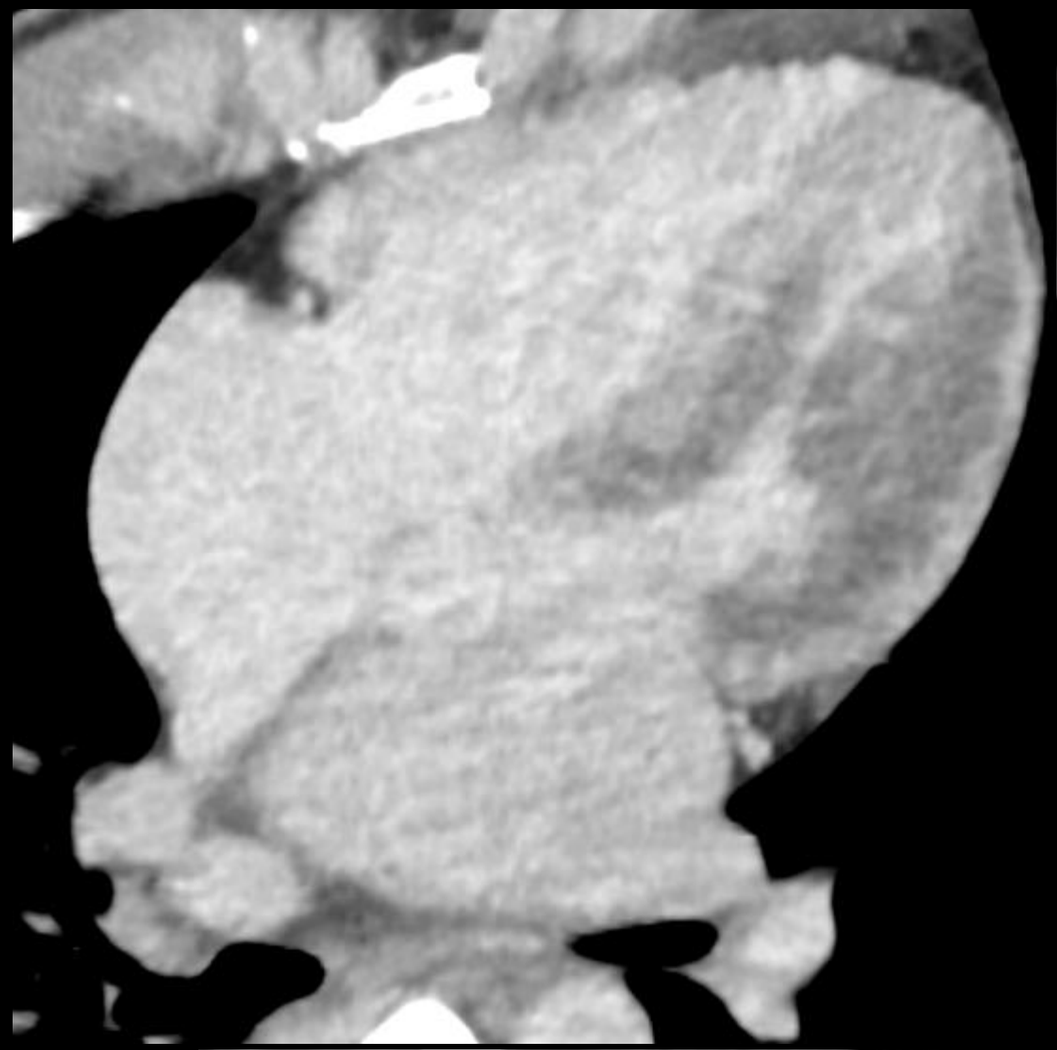


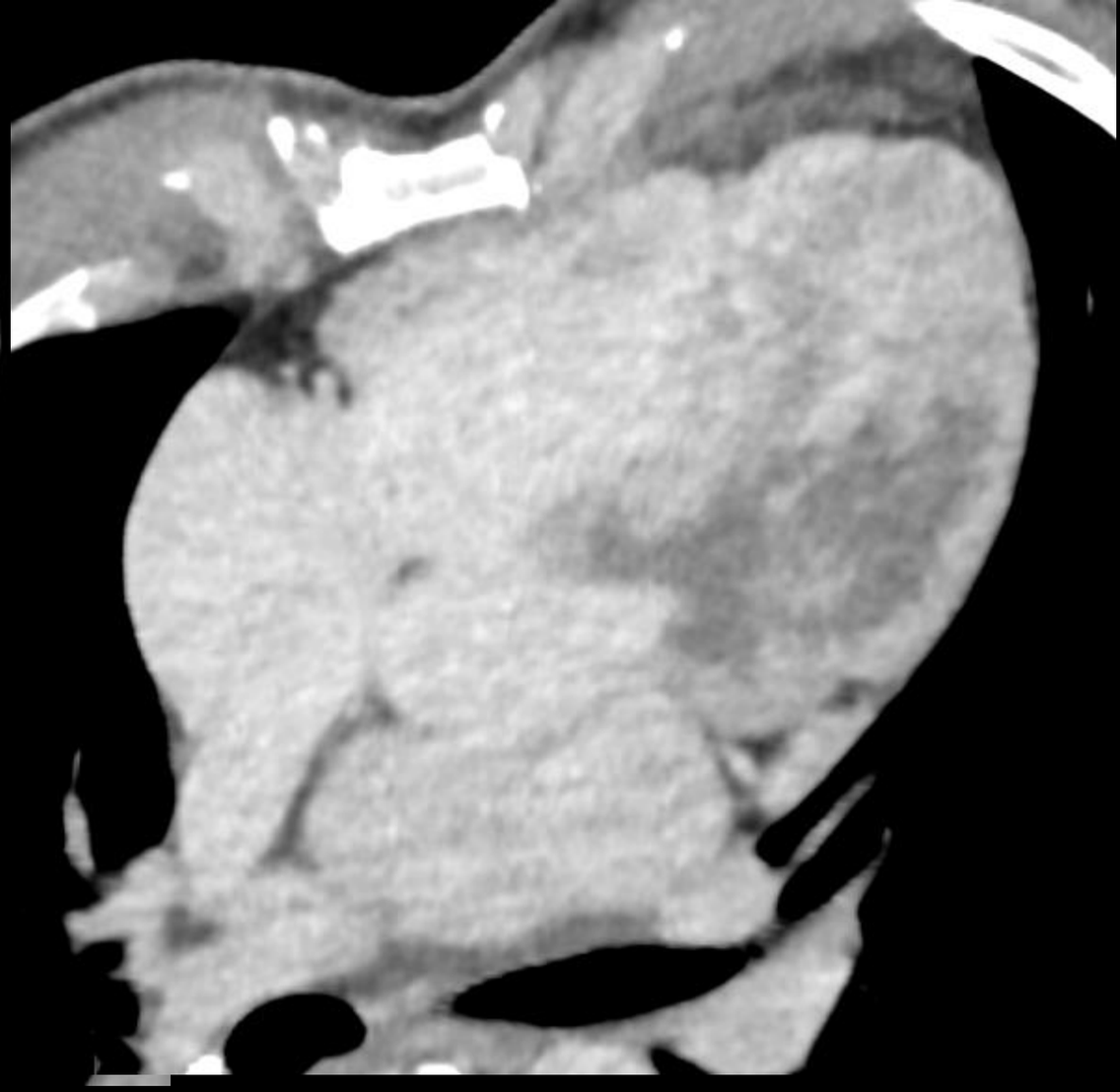


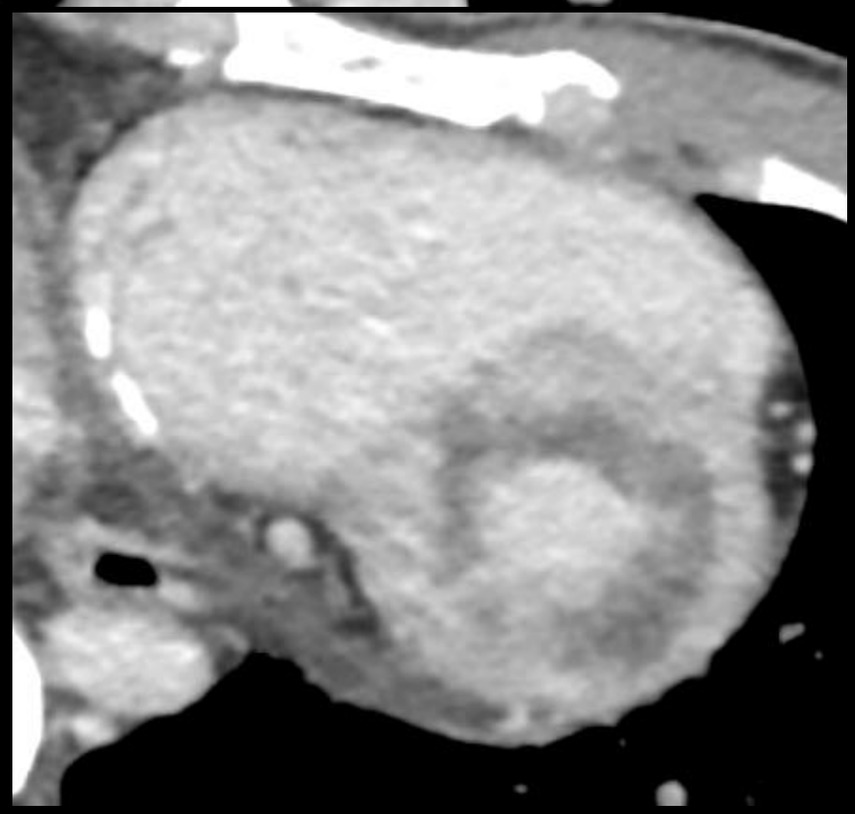


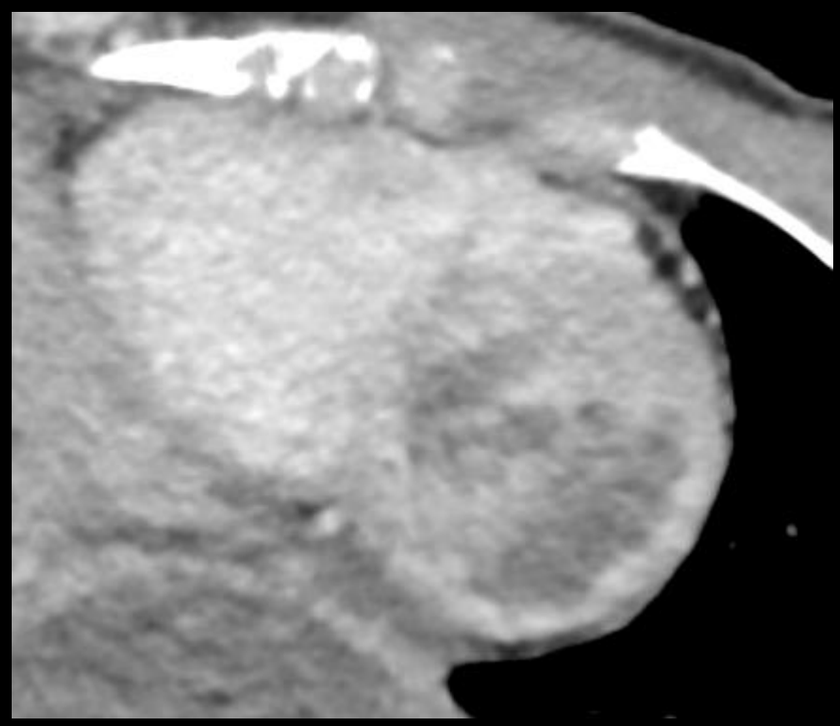


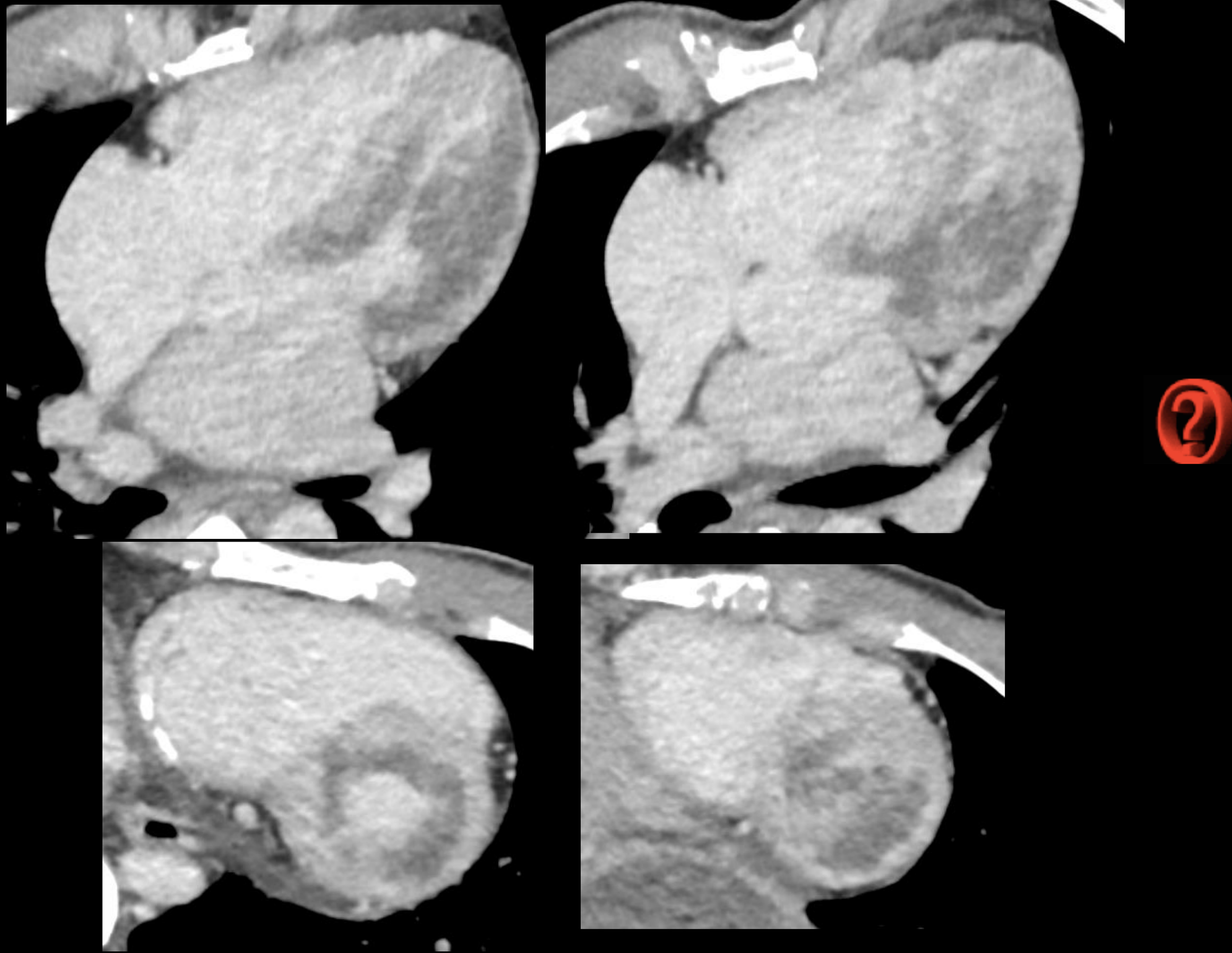






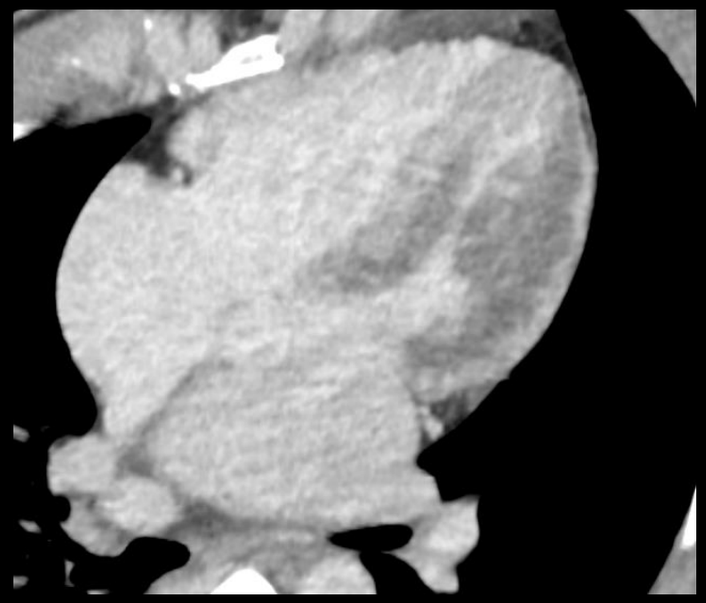






Pas de passage sans injection, contusions myocardiques ou plus

probablement infarctus massif sur bas débit



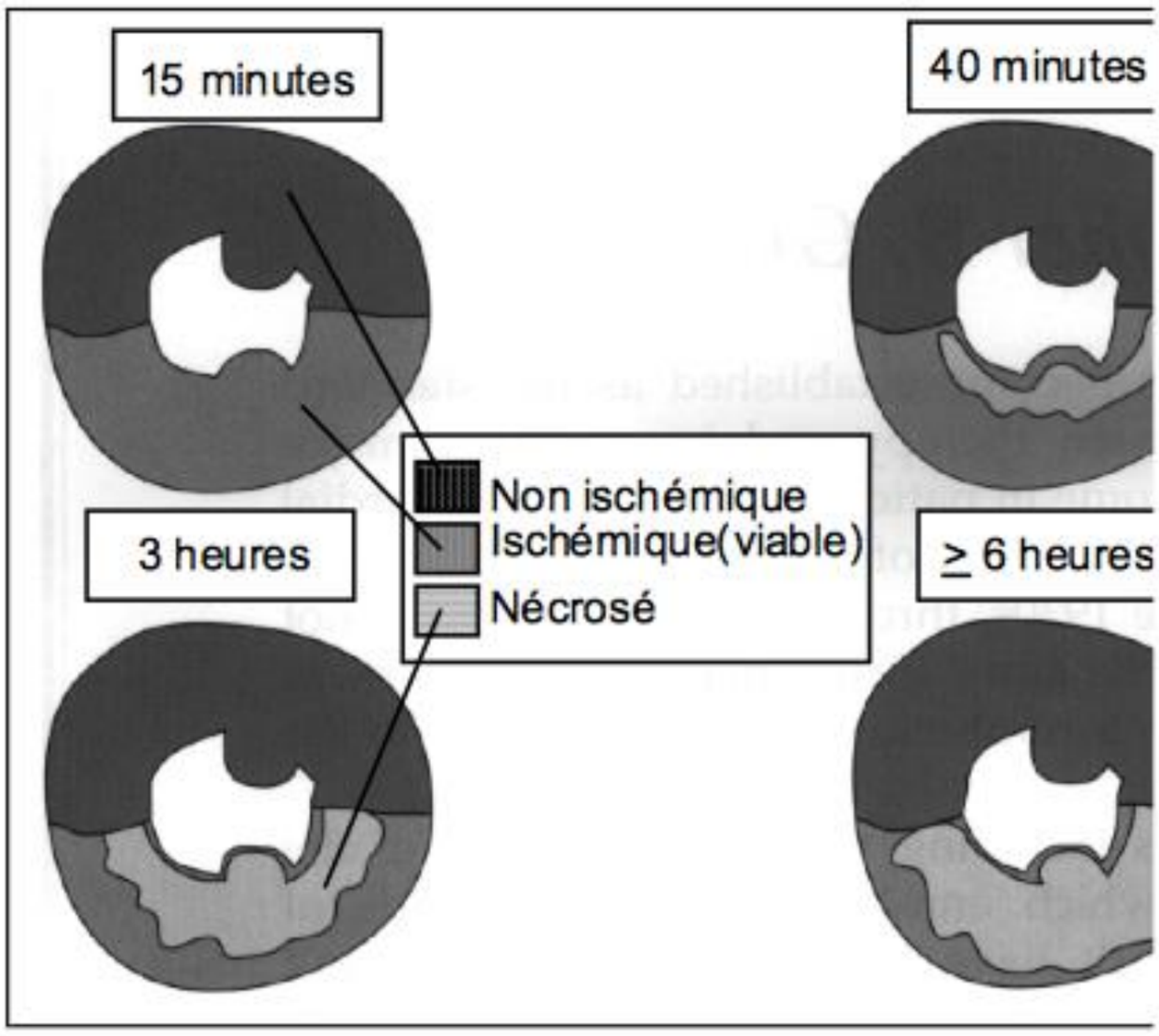
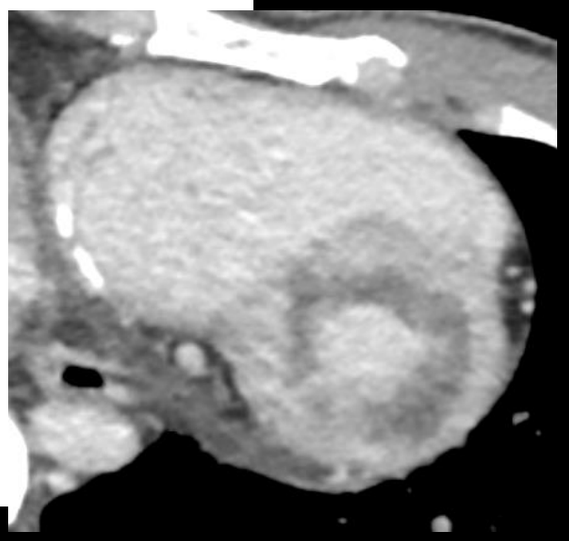


Figure 3
Schématisation de la progression de la nécrose en fonction du temps écoulé depuis l'occlusion de l'artère responsable de l'infarctus.

D'après Jennings et Reimer, Circulation 1983 ; 68 : suppl. 1 : 36.



ETT (tj en TV) :

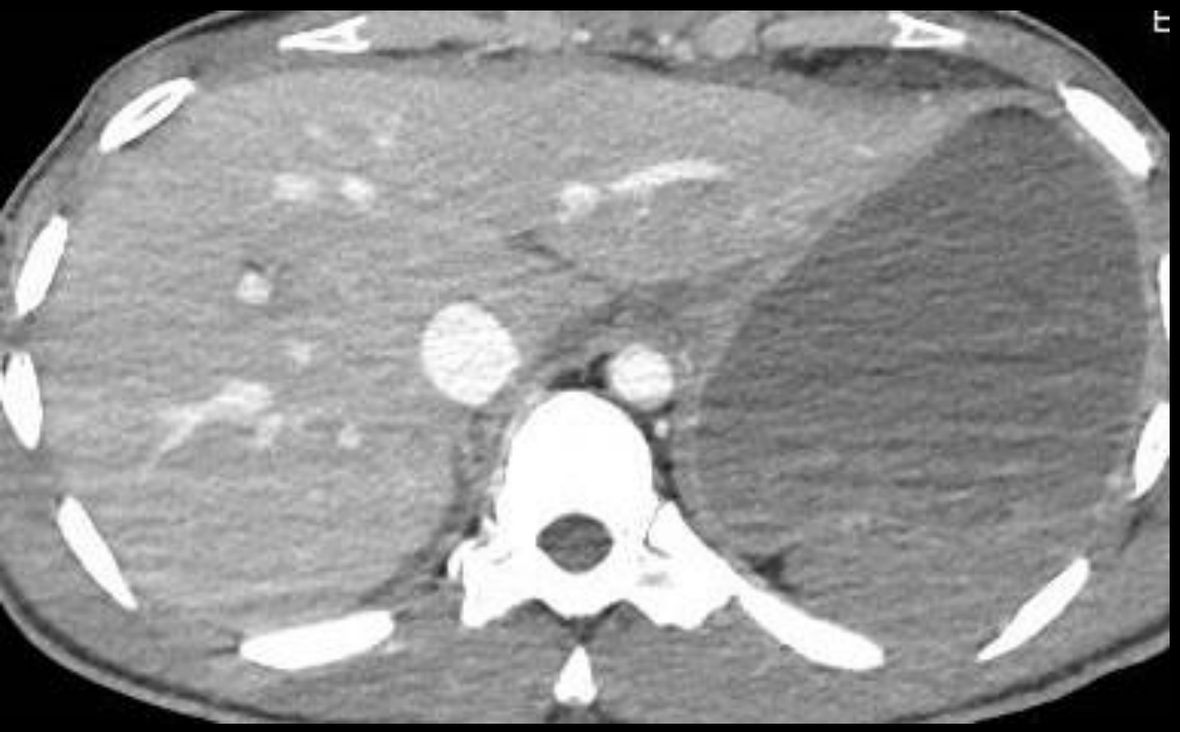
cardiomyopathie hypertrophie concentrique symétrique

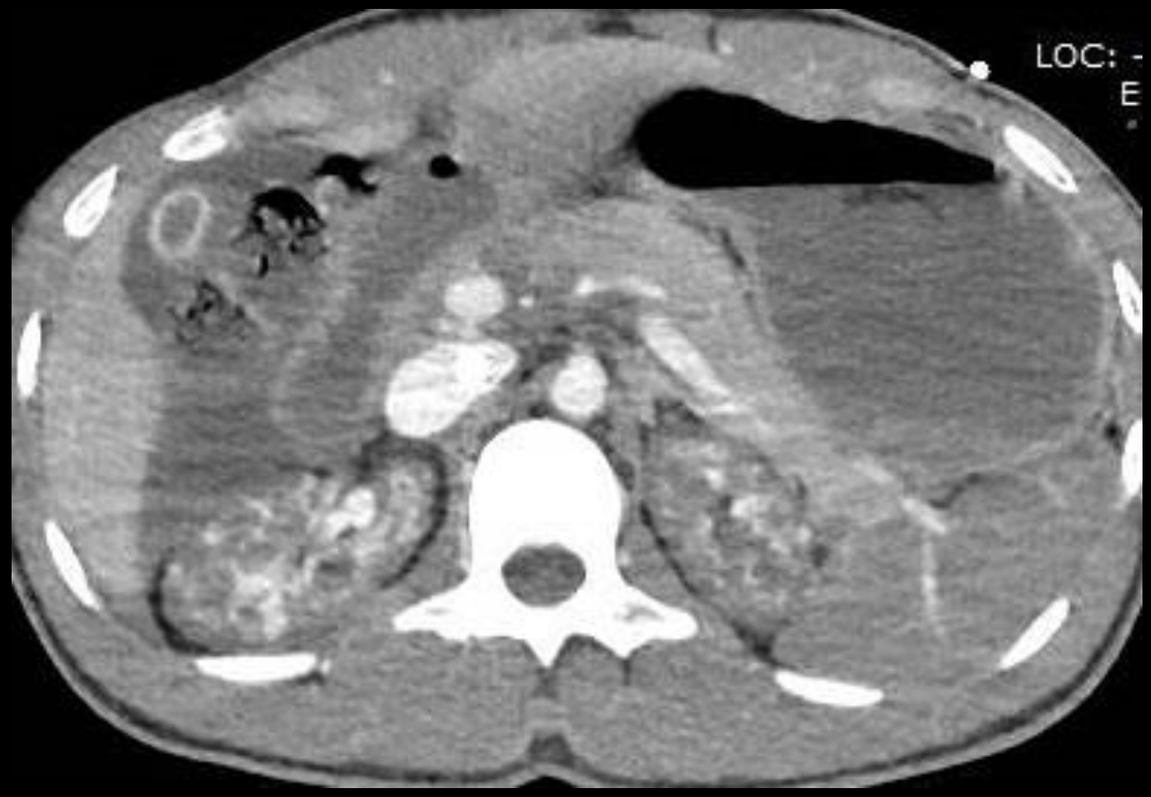
avec bourrelet septal de 25 mm d'épaisseur,

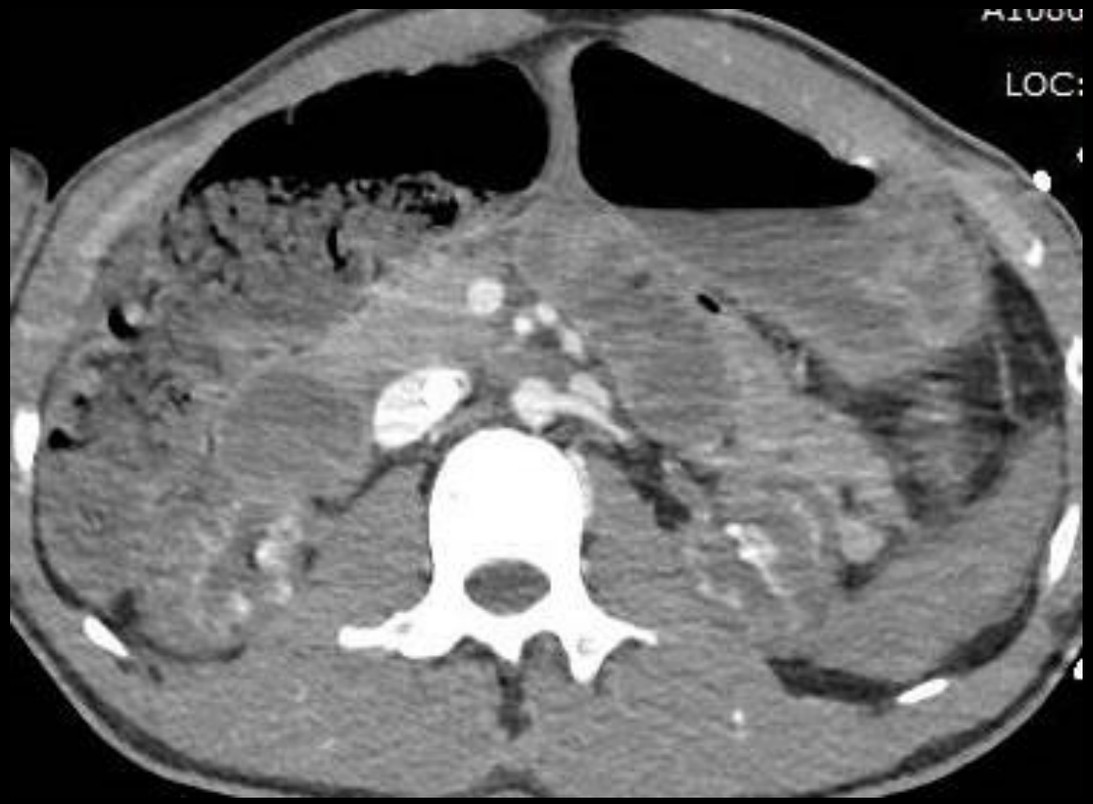
pas d'atteinte du VD

Temps portal

Temps tardif



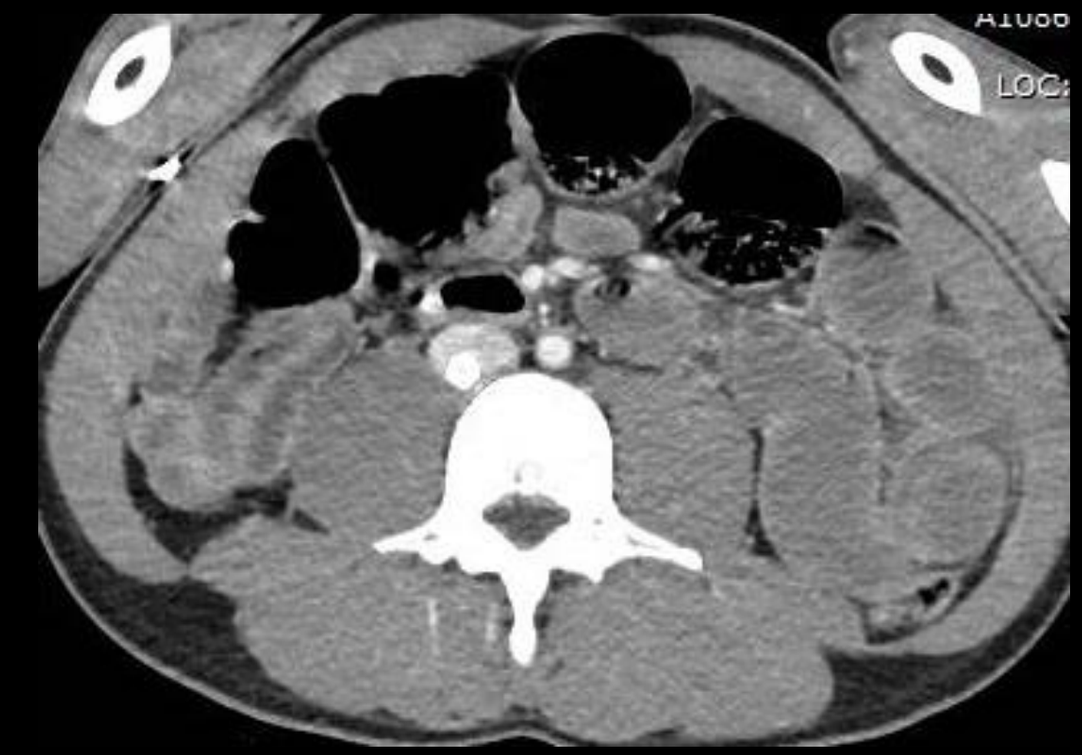


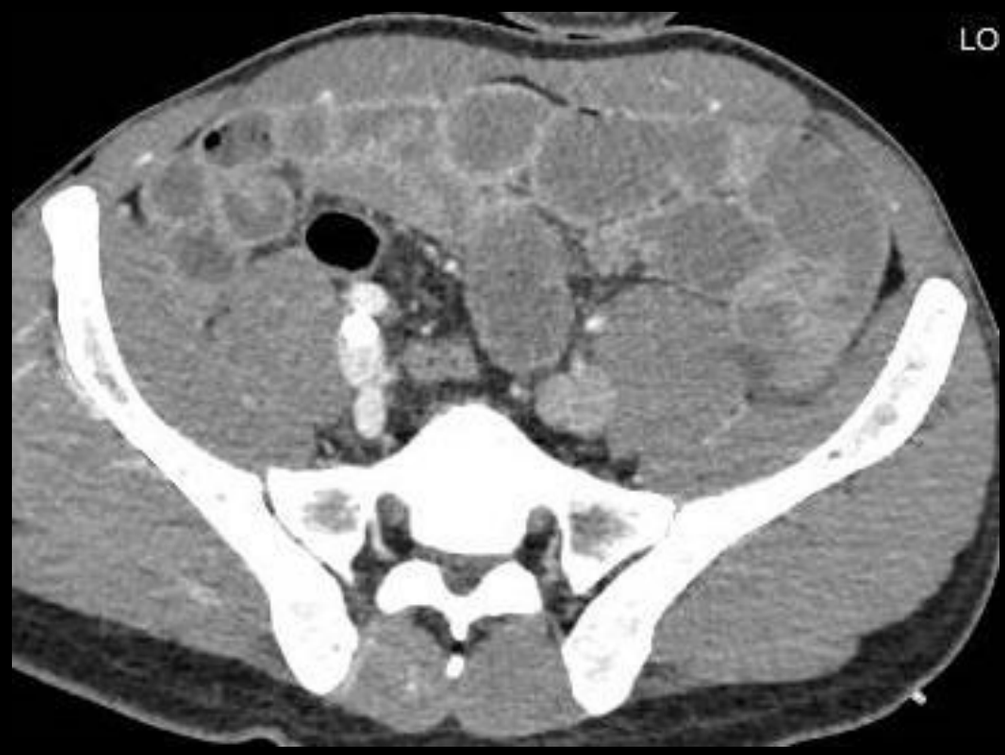


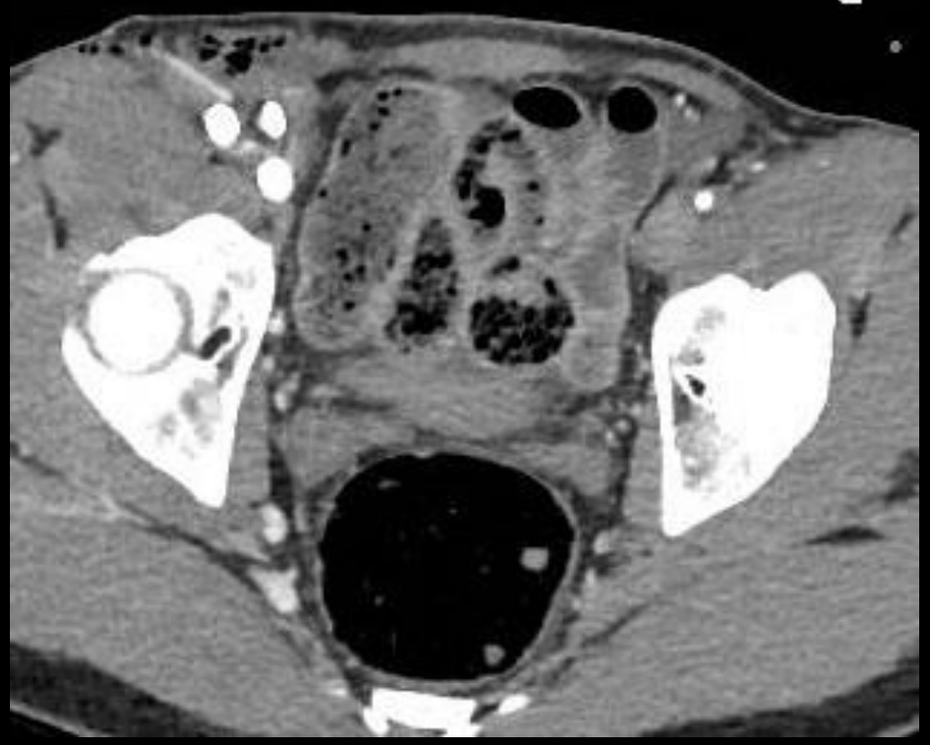
-défaut global de rehaussement des paros intestinales et des parenchymes pleins de l'abdomen

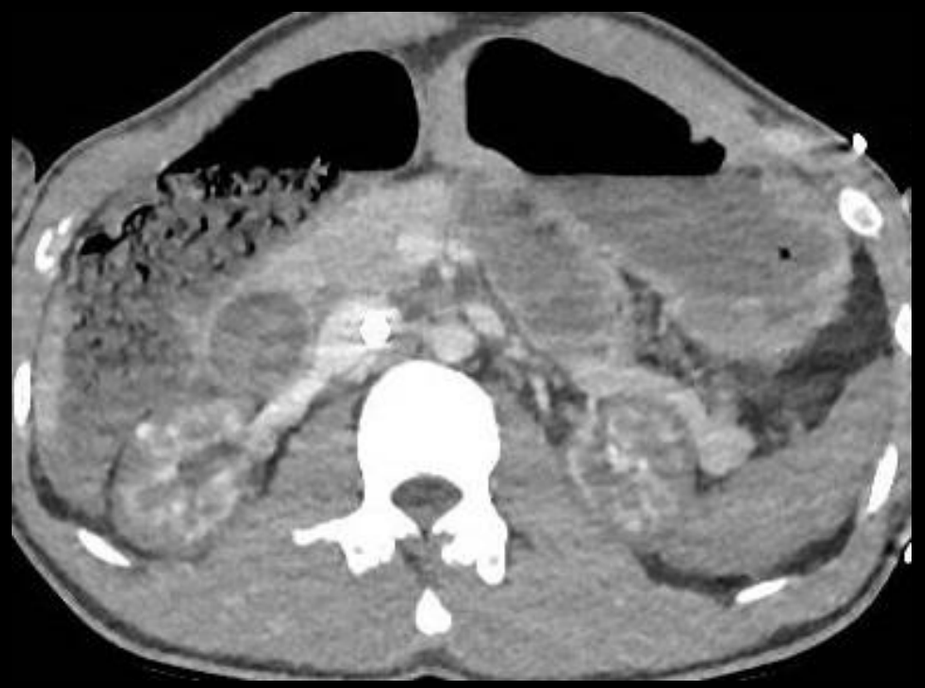
-œdème de la paroi vésiculaire

-ischémie rénale +++



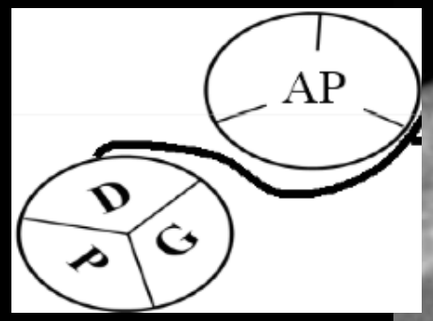


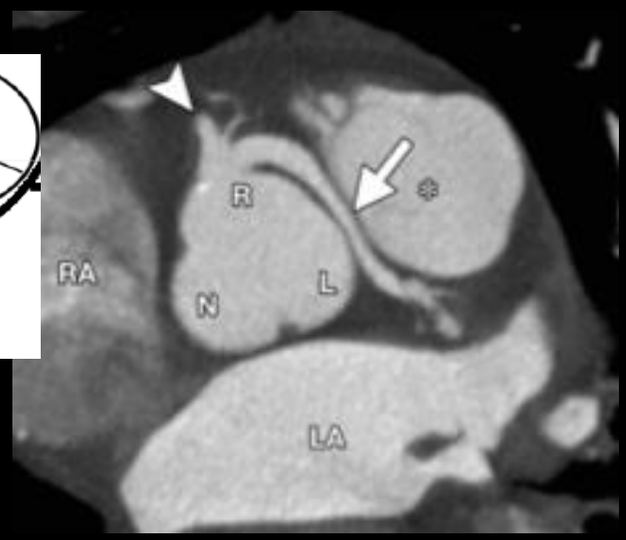


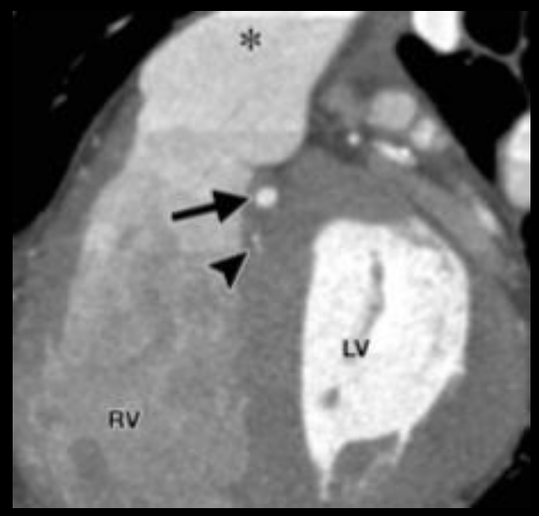


Causes de mort subite chez l'adulte jeune

- anomalies coronaires congénitales : trajet inter-artériel de la CG, ou trajet intramural



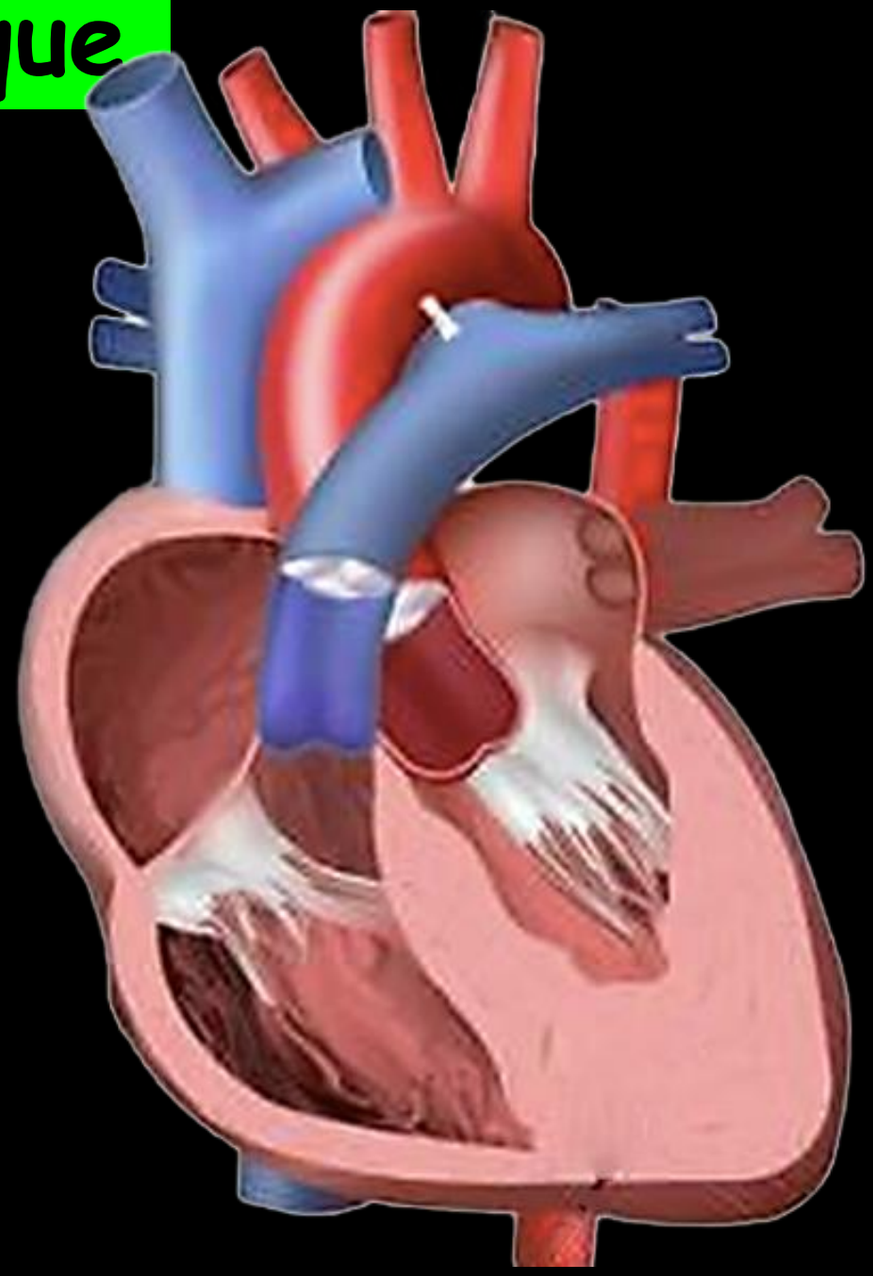




- cardiomyopathie héréditaire : CMH, dysplasie arythmogène ventriculaire droite, cardiomyopathie dilatée, cardiomyopathie non compactée

- mort subite souvent précipitée par un trouble du rythme (TV ou fibrillation) : stratification du risque (ATCD, ETT, CT, MR), pose de DAI en fonction

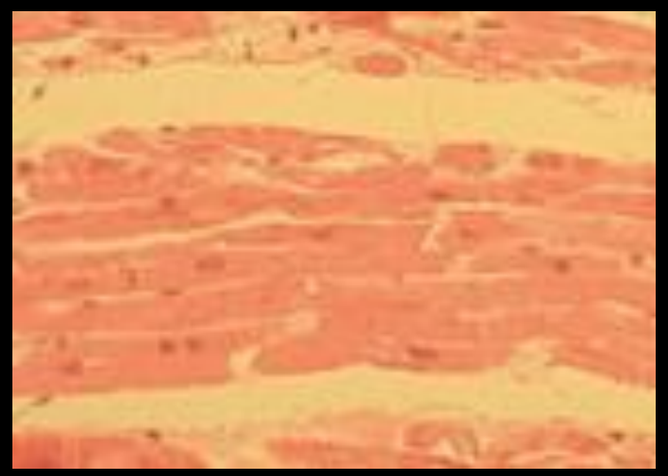
Cardiomyopathie hypertrophique

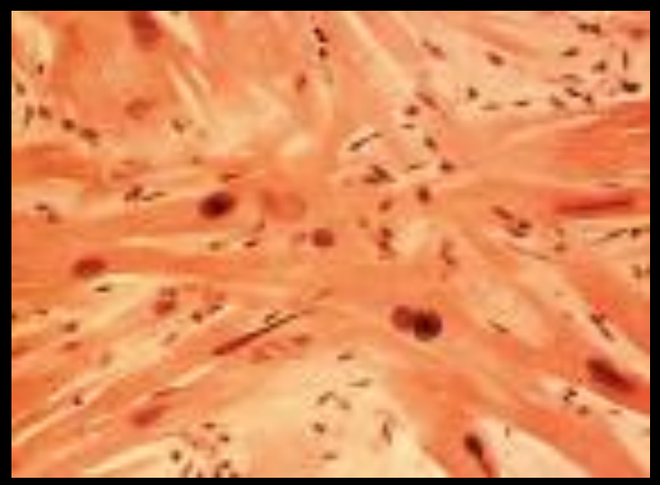


- maladie génétique cardiaque, autosomique dominante avec pénétrance et expression variables,

- mutation d'un des gènes codant les protéines sarcomériques => myocytes hypertrophiques désorganisés et fibrose interstitielle

- prévalence phénotypique 0.2%

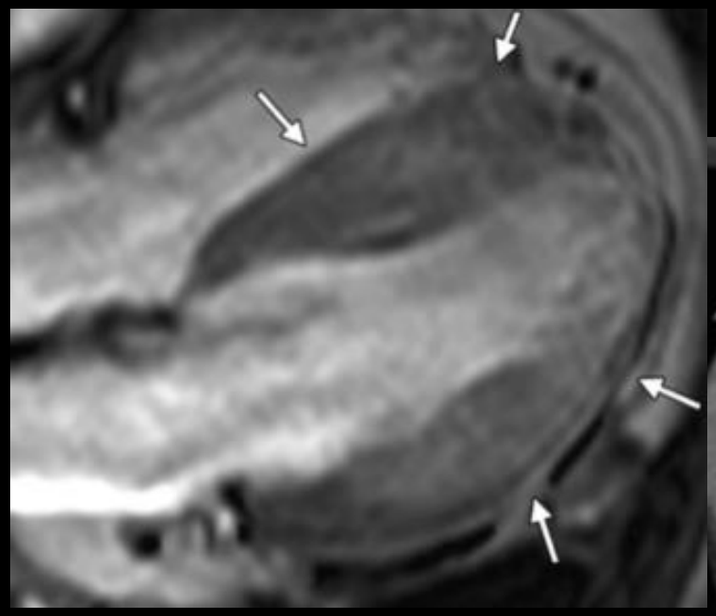


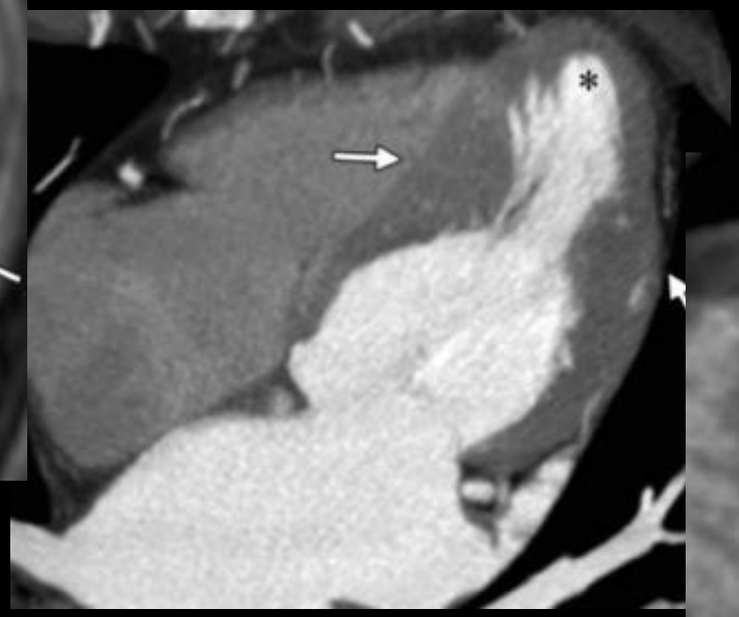


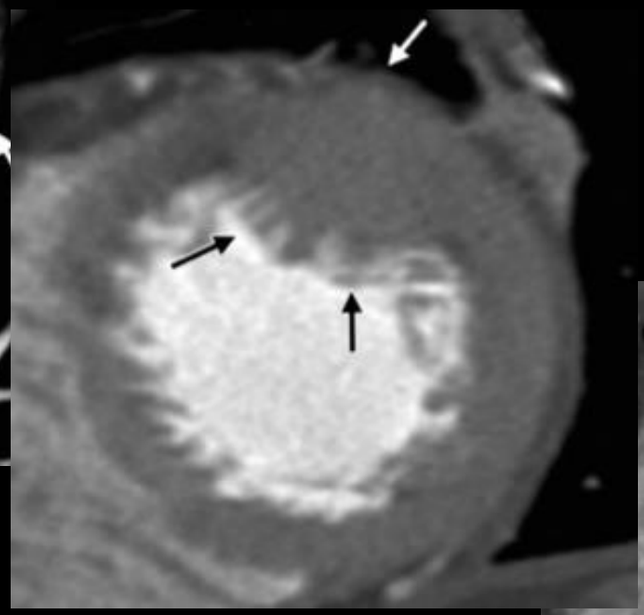
- cause la plus fréquente de mort subite chez l'adulte jeune

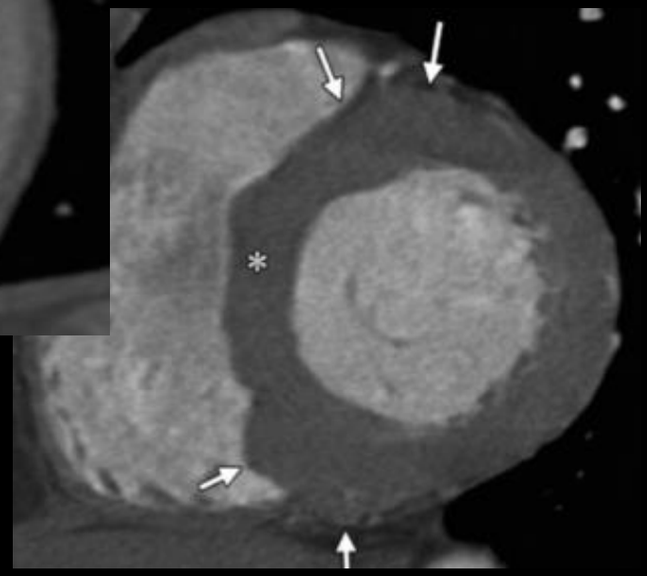
- hypertrophie diffuse ou segmentaire du VG : réduction du volume de la cavité ventriculaire et altération de la fonction diastolique

- asymptomatique, dyspnée, douleur thoracique, syncope, mort subite (risque annuel 1% adulte, 2 à 4% jeune)

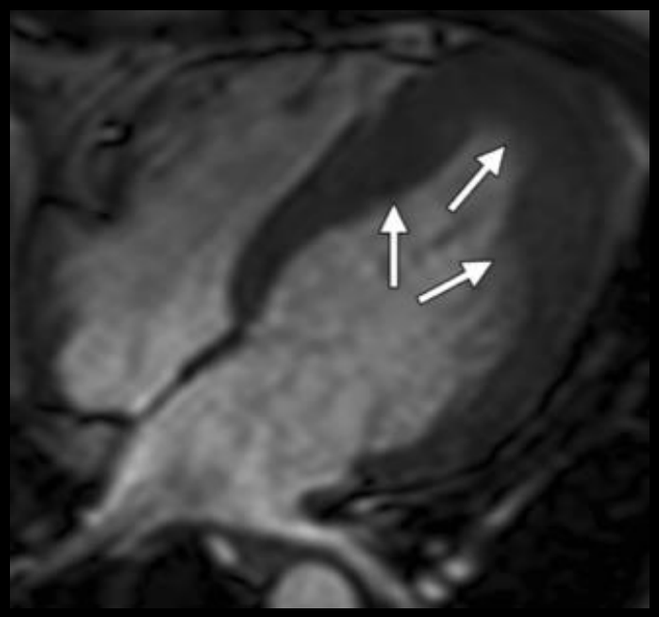








- augmentation de l'épaisseur de la paroi du VG mesurée en télé diastole ≥ 13mm chez l'homme, ≥ 12mm chez la femme et ≥ 15 mm chez le sportif de haut niveau



- forme asymétrique la plus fréquente, avec ou sans obstruction de la chambre de chasse du VG

- ttt médical (BB, ICa), DAI, +/- chirurgical (myomectomie chirurgicale ou alcoolisation septale)

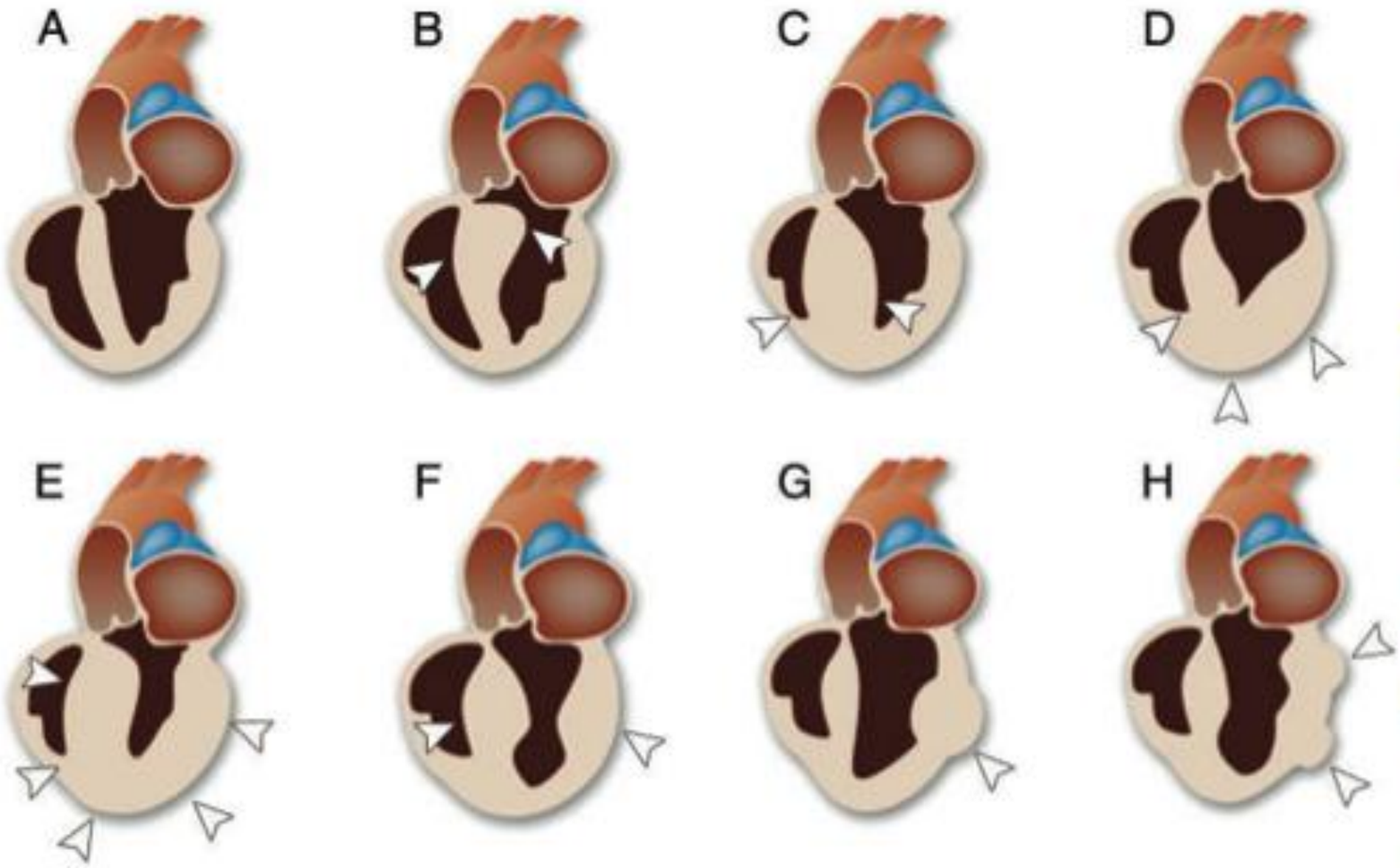
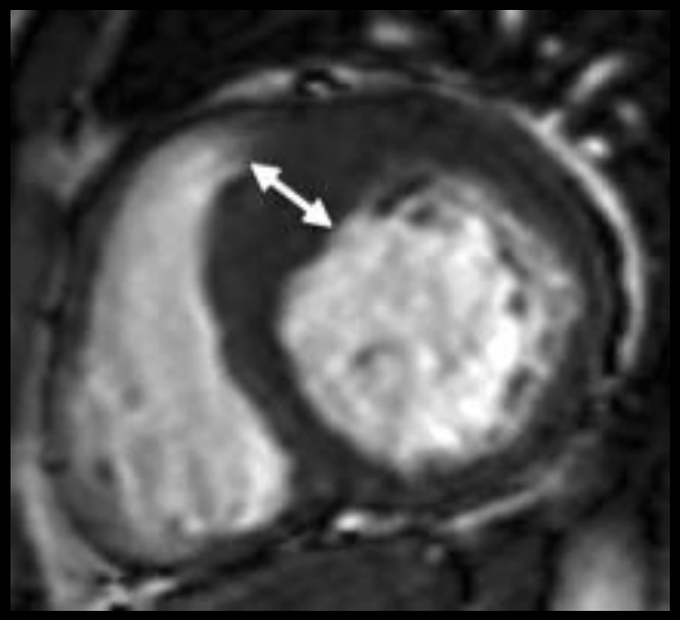
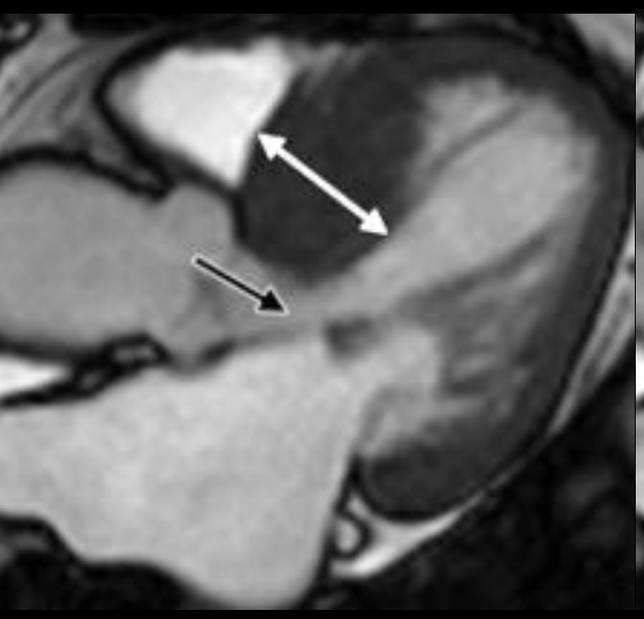
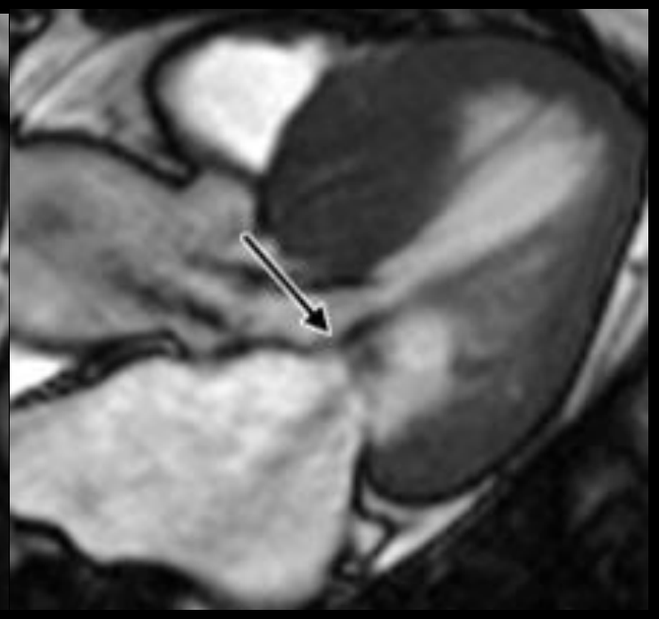


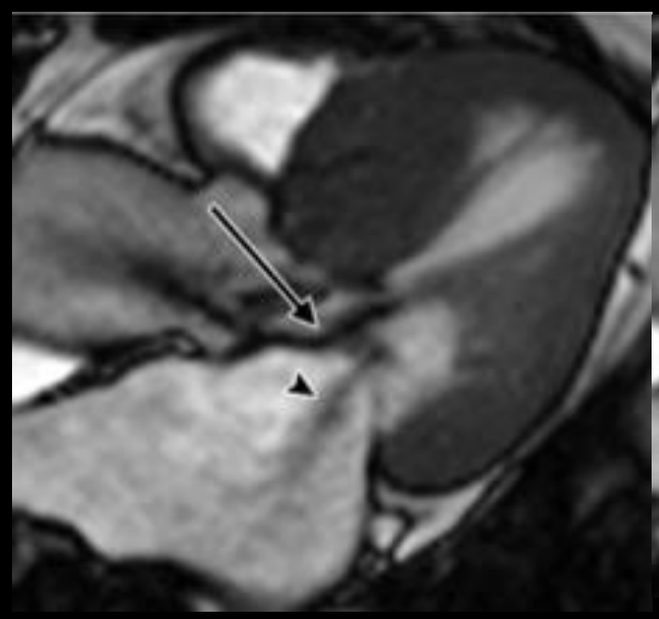
Figure 1. Drawings of the normal heart and the heart in the various phenotypes of HCM. The diagnostic criterion of HCM is that the maximal LV wall thickness is greater than or equal to 15 mm in the end-diastolic phase. *A,* Normal heart; *B,* asymmetric (septal) HCM with LVOT obstruction; *C,* asymmetric (septal) HCM without LVOT obstruction; *D,* apical HCM; *E,* symmetric HCM (concentric HCM); *F,* midventricular HCM; *G,* masslike HCM; *H,* noncontiguous HCM. The drawings of the various phenotypes of HCM show the areas of hypertrophy (arrowheads).

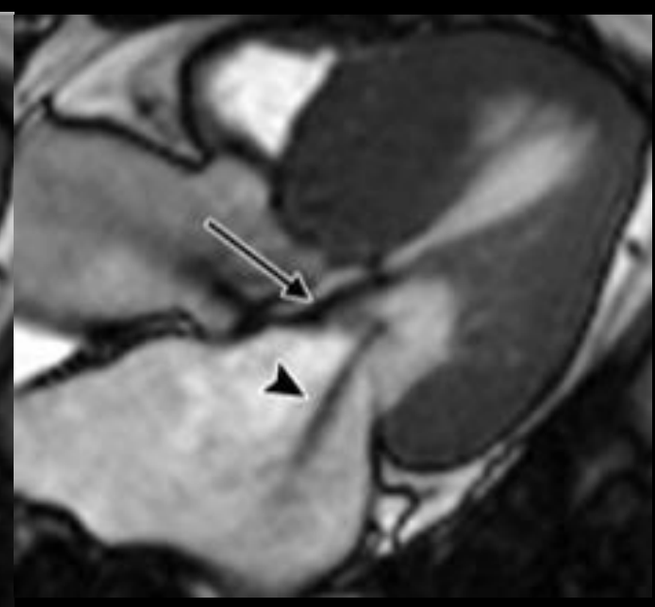


patient de 74 ans, hypertrophie septale (16 mm), obstruction de la chambre de chasse du VG, régurgitation mitrale

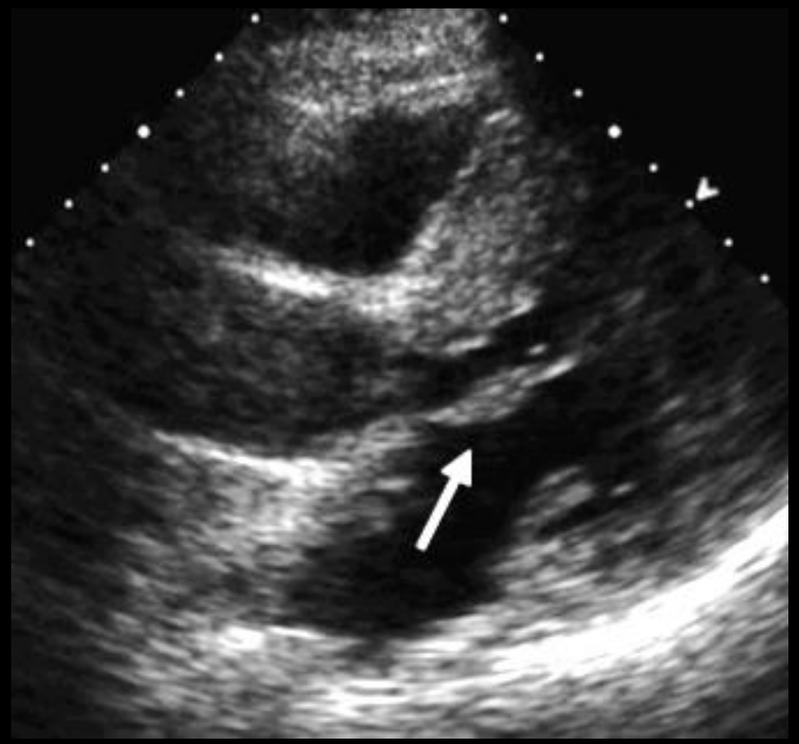


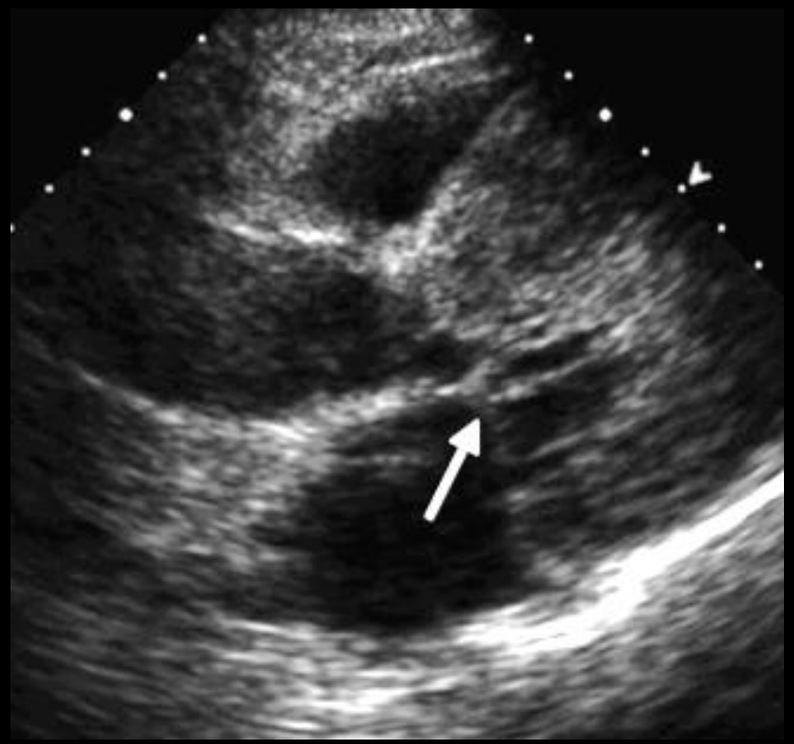




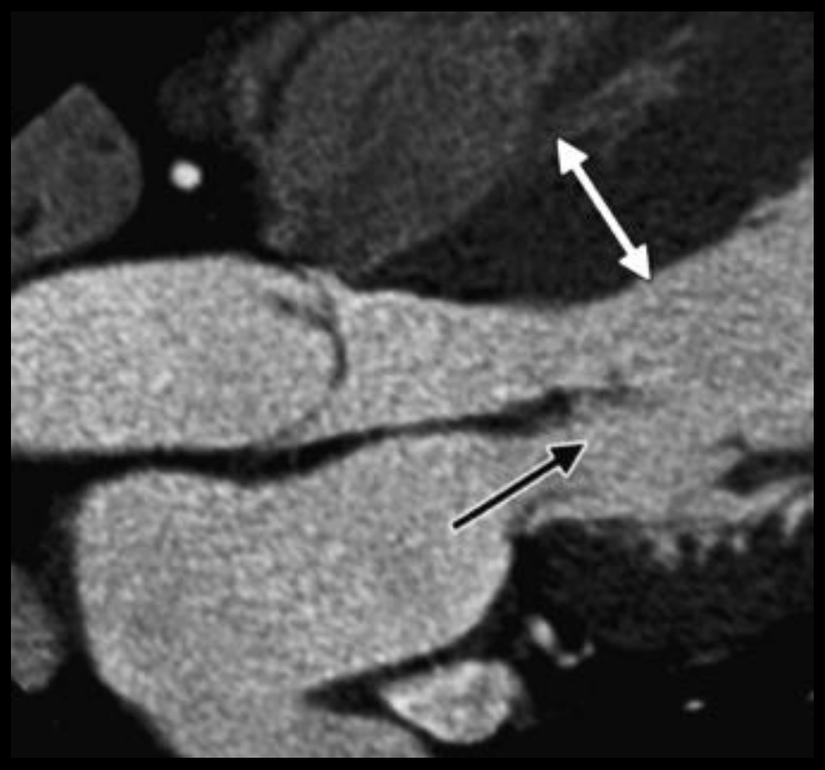


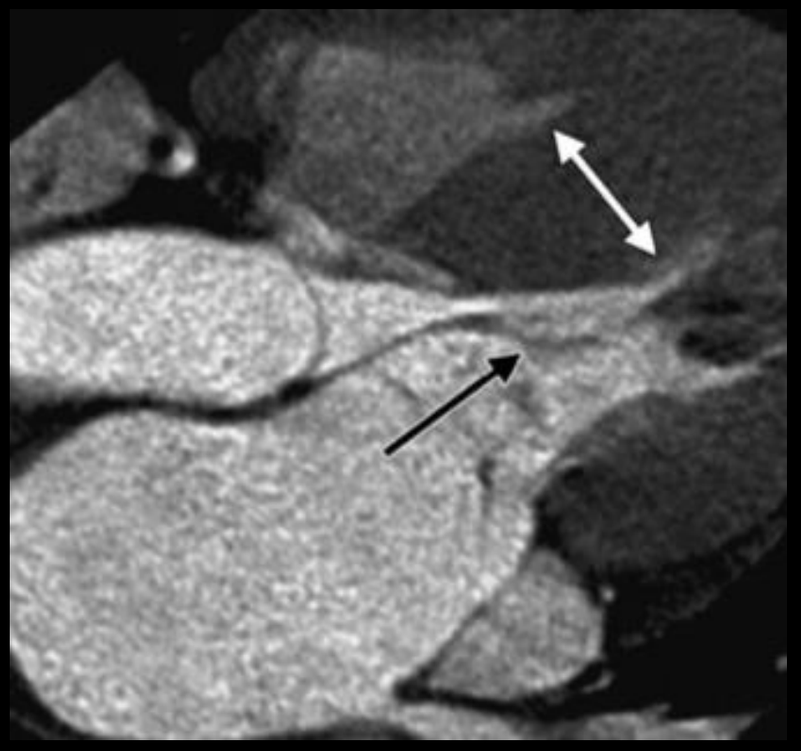
L’hypertrophie du septum interventriculaire empiète sur la lumière de la chambre de chasse du VG lorsque le muscle se contracte => accélération majeure du flux systolique => feuillet antérieur de la valve mitrale aspiré dans la CCVG => contribue encore à l’obstruction de la chambre de chasse + ouvre la valve à mi-systole et provoque une insuffisance mitrale méso-télésystolique

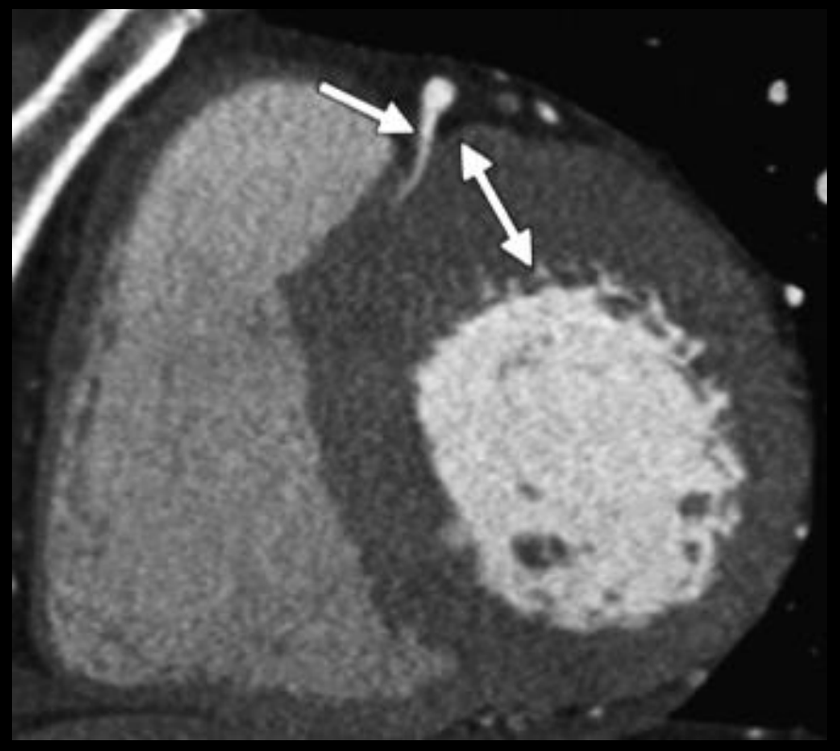




branche septale engorgée







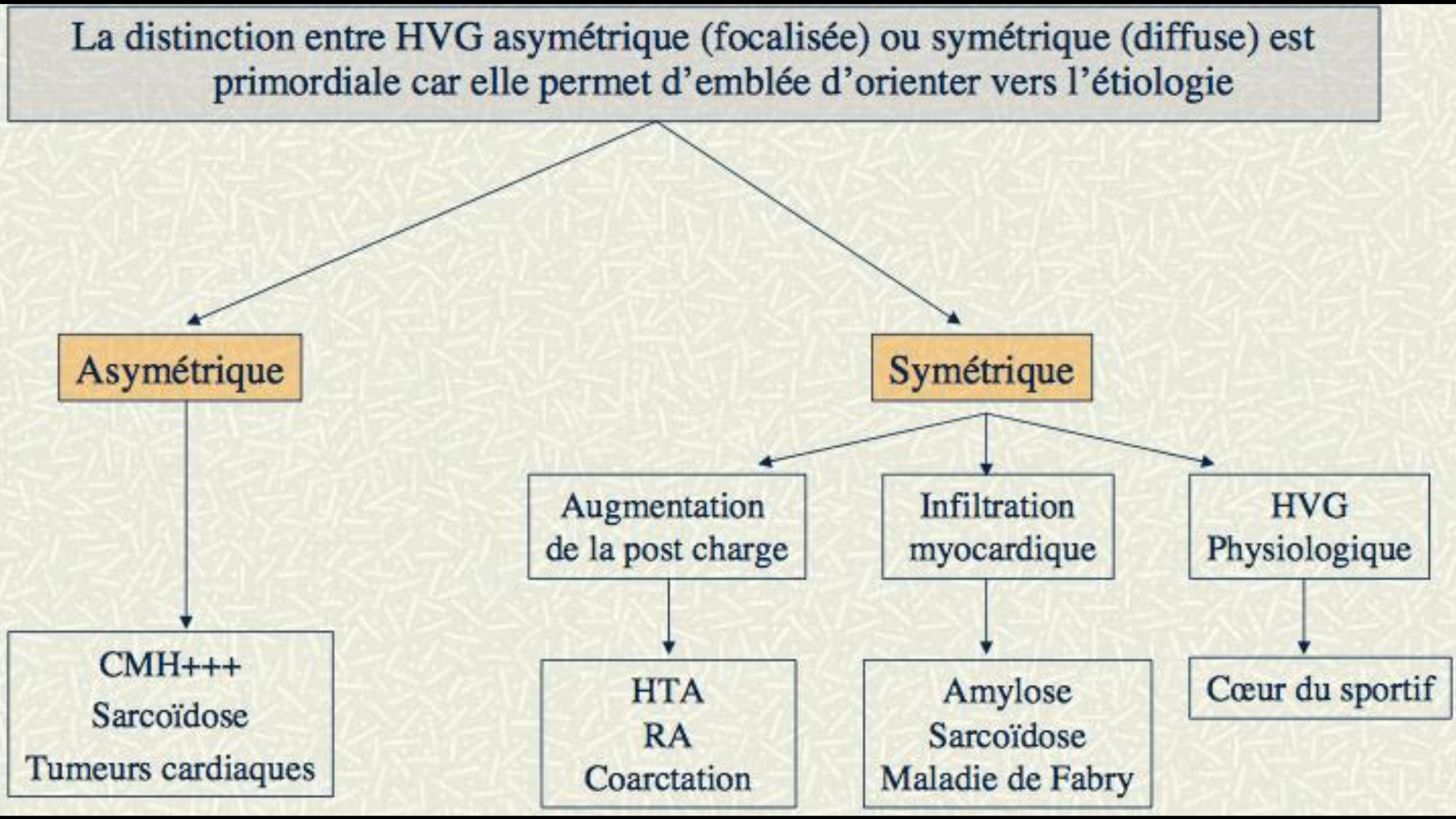
patient de 44 ans, dyspnée, hypertrophie septale
ETT en diastole et fin de systole : déplacement antérieur du feuillet mitral

Table 2
Relative Merits of Each Noninvasive Imaging Modality for the Assessment of HCM

Factor Assessed	Echocardiography*	Multidetector CT	MR Imaging
LV volume	+++	++	++++
LV hypertrophy	+++	++++	++++
Ejection fraction	+++	+++	++++
Regional function	+++	++	++++
LV filling pressure	+++	—	++
PA pressure	+++	—	+++
Dynamic obstruction	+++	+	+++
Mitral regurgitation	+++	—	++
Ischemia/CFR	+	—	++
Monitoring of therapy	+++	+	+++
Tissue characterization	++	+	++++
Preclinical diagnosis	++	+++	+++

Note.—The number of plus signs indicates the relative usefulness of the modality. — = modality is not useful, CFR = coronary flow reserve, PA = pulmonary artery.
*Data in this column are from Nagueh and Mahmarian (10).

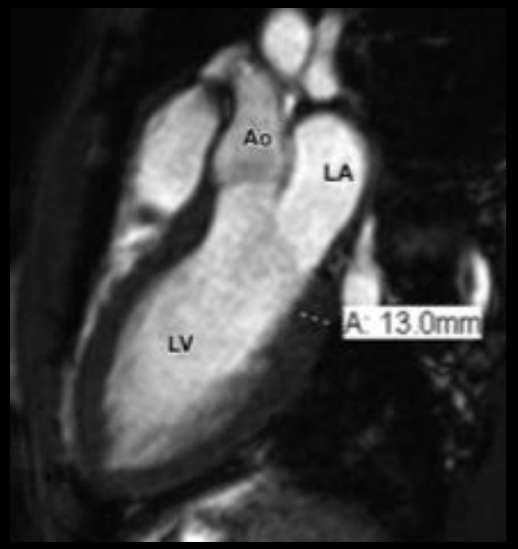
Diagnostic différentiel

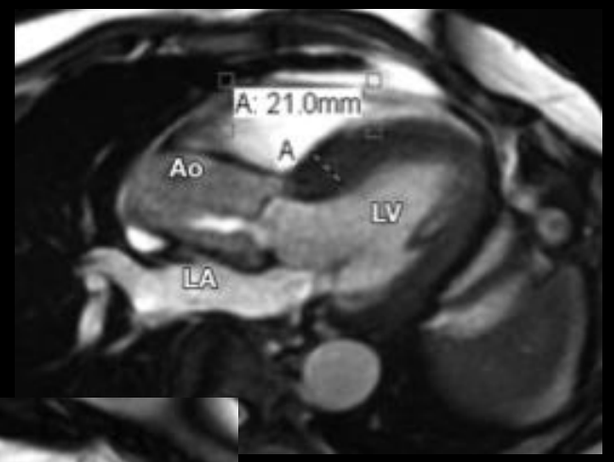


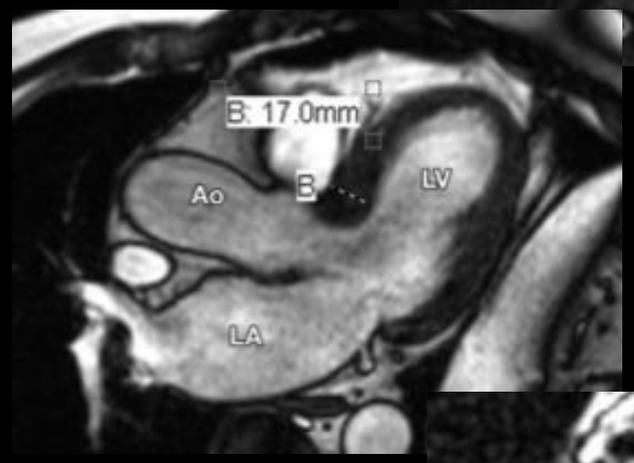
J Caudron et al., Rouen

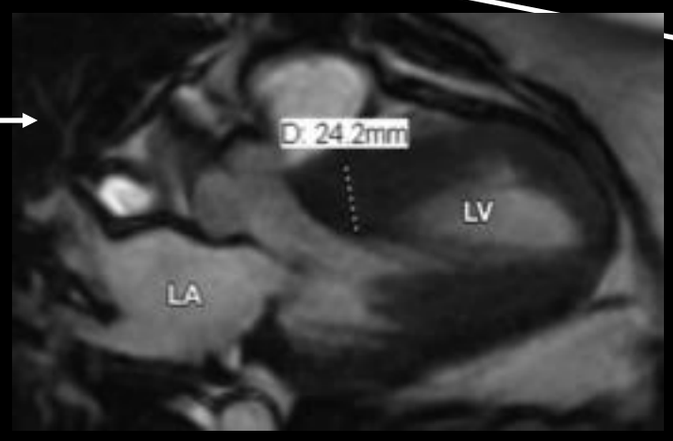
Diagnostic différentiel d'une HVG chez un adulte jeune

- coeur d'athlète (2aire à un entrainement intense prolongé)
- sténose aortique (bicuspidie le + svt)
- HTA
- amylose cardiaque
- maladie de surcharge









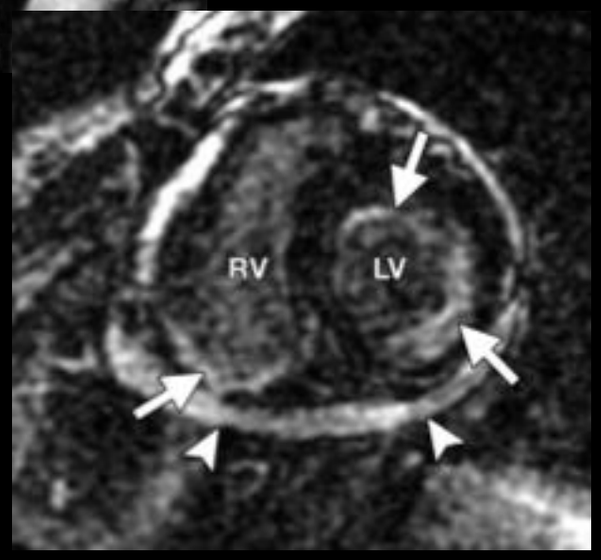
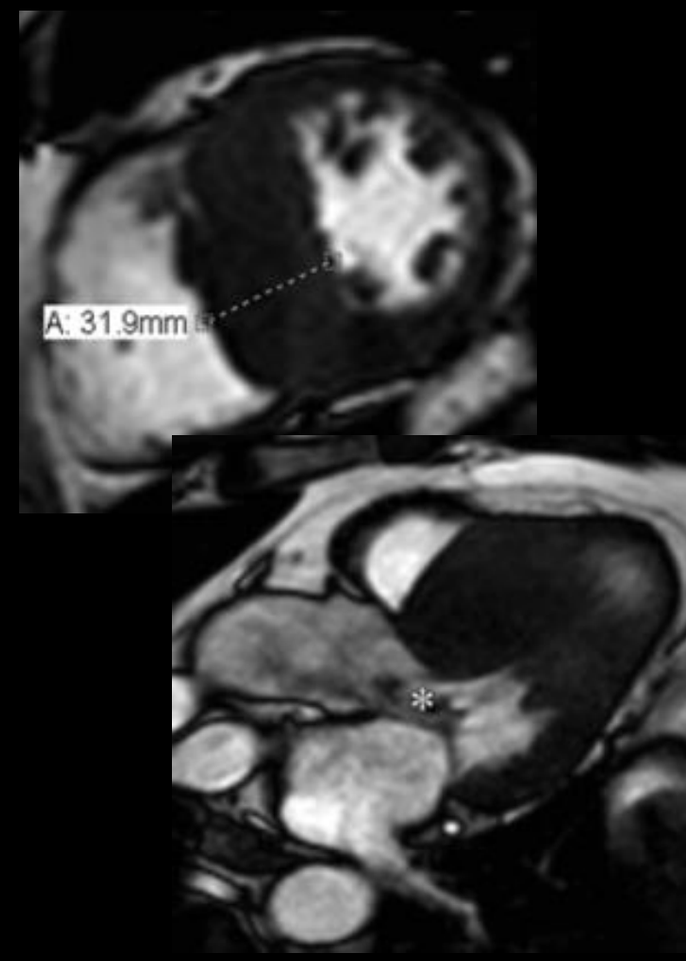


Table 1
Applications of Cardiac Imaging Modalities for Stratification of the Risk of Sudden Cardiac Death in Patients with HCM

Risk Factor	Echocar-diography	Multi-detector CT	MR Imaging
LV maximal wall thickness of 30 mm or more	+	+	+
Marked LVOT obstruction (LVOT gradient ≥ 30 mm Hg at rest)	+	−	+
LV dilatation and depressed ejection fraction	+	+	+
Fibrosis	−	−	+*
Perfusion defect	−	±	+
Reduced functional reserve flow	±	−	+

Note.—+ = modality can be used to assess the risk factor, − = modality is not useful for assessing the risk factor, ± = modality is available for assessing the risk factor, but the usefulness of the modality is still investigational and is not validated yet.
*Detection at DE MR imaging.



L'American College of Cardiology/ European Society of Cardiology a défini des **FDR de mort subite** :

- fibrillation ventriculaire, tachycardie ventriculaire spontanée soutenue ou non soutenue
- histoire familiale de mort subite, syncope inexpliquée, TA anormale à l'effort
- épaisseur du VG > ou = 30 mm

RadioGraphics

Hypertrophic Cardiomyopathy: Assessment with MR Imaging and Multidetector CT[1]

CME FEATURE

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Eun Ju Chun, MD • Sang Il Choi, MD • Kwang Nam Jin, MD • Hyon Joo Kwag, MD • Young Jin Kim, MD • Byoung Wook Choi, MD • Whal Lee, MD Jae Hyung Park, MD

RadioGraphics

Congenital and Hereditary Causes of Sudden Cardiac Death in Young Adults: Diagnosis, Differential Diagnosis, and Risk Stratification[1]

TEACHING POINTS

See last page