

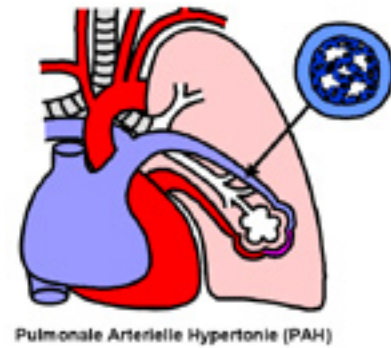
Hypertension artérielle pulmonaire

Cas Clinique

Pr Laila Haddour
RABAT

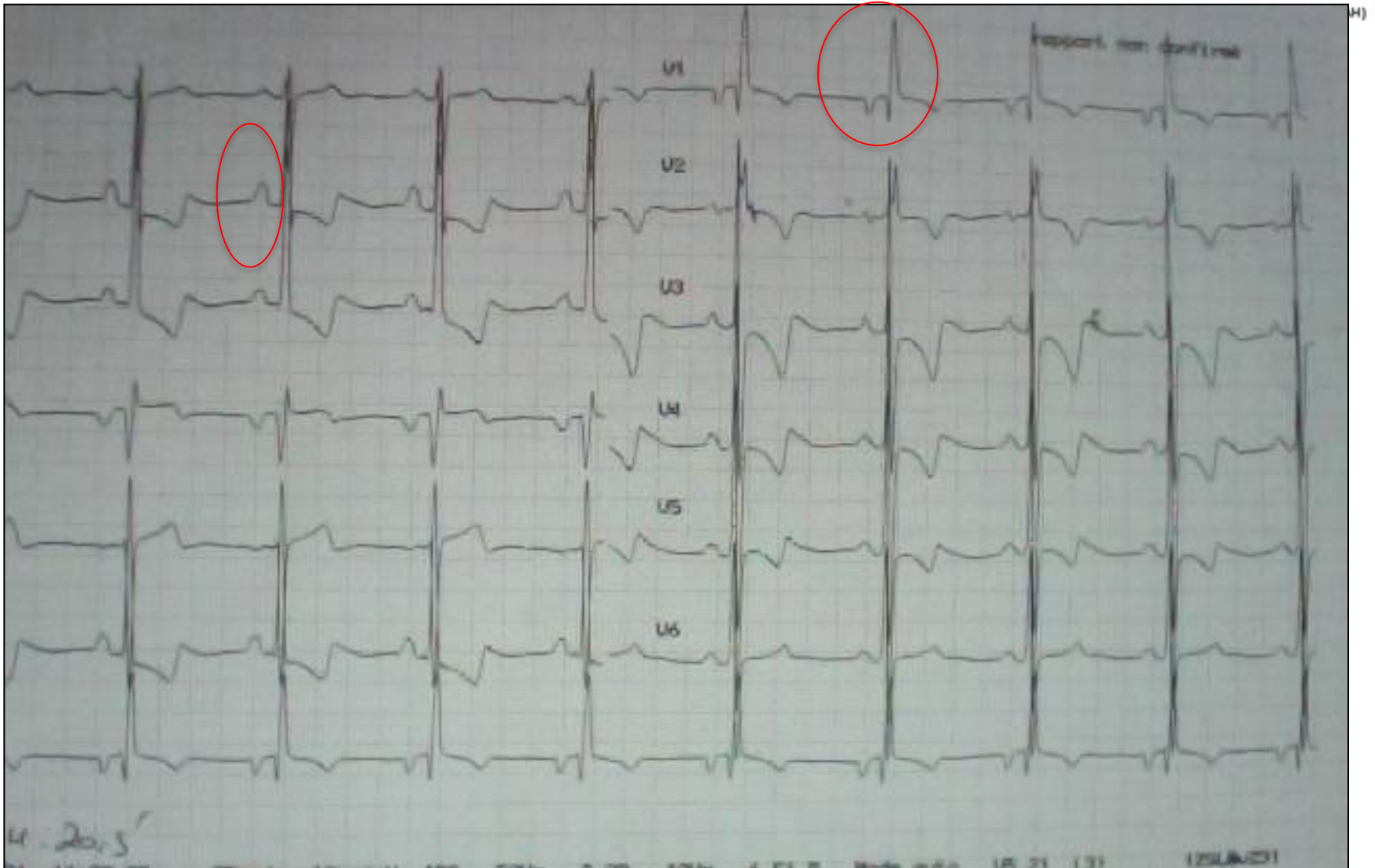
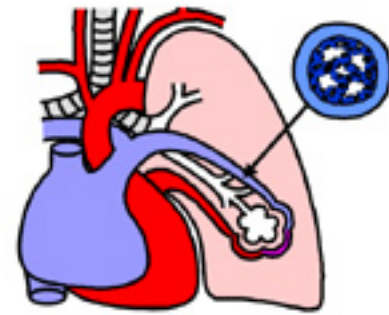
20e congrès national de la Société Marocaine de Cardiologie jumelé au 16ème
édition du PAFCIC (Pan African Course on Interventional Cardiology)
28 _31 octobre 2015 Marrakech

Mr M., 35 ans

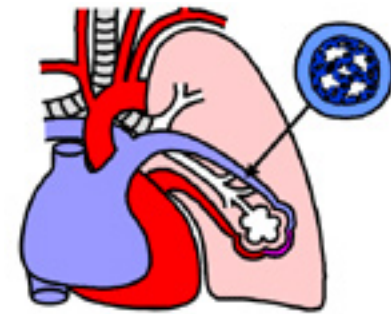


- Adressé en chirurgie cardiaque pour fermeture de CIA
- Dyspnée d'effort stade 2 NYHA depuis 10 ans
- A l'examen:
 - Pas de signe d'ICD
 - TA= 120/70 mmHg
 - FC=73PPM
 - SaO₂= 80%
 - Eclat B2 au FP

ECG



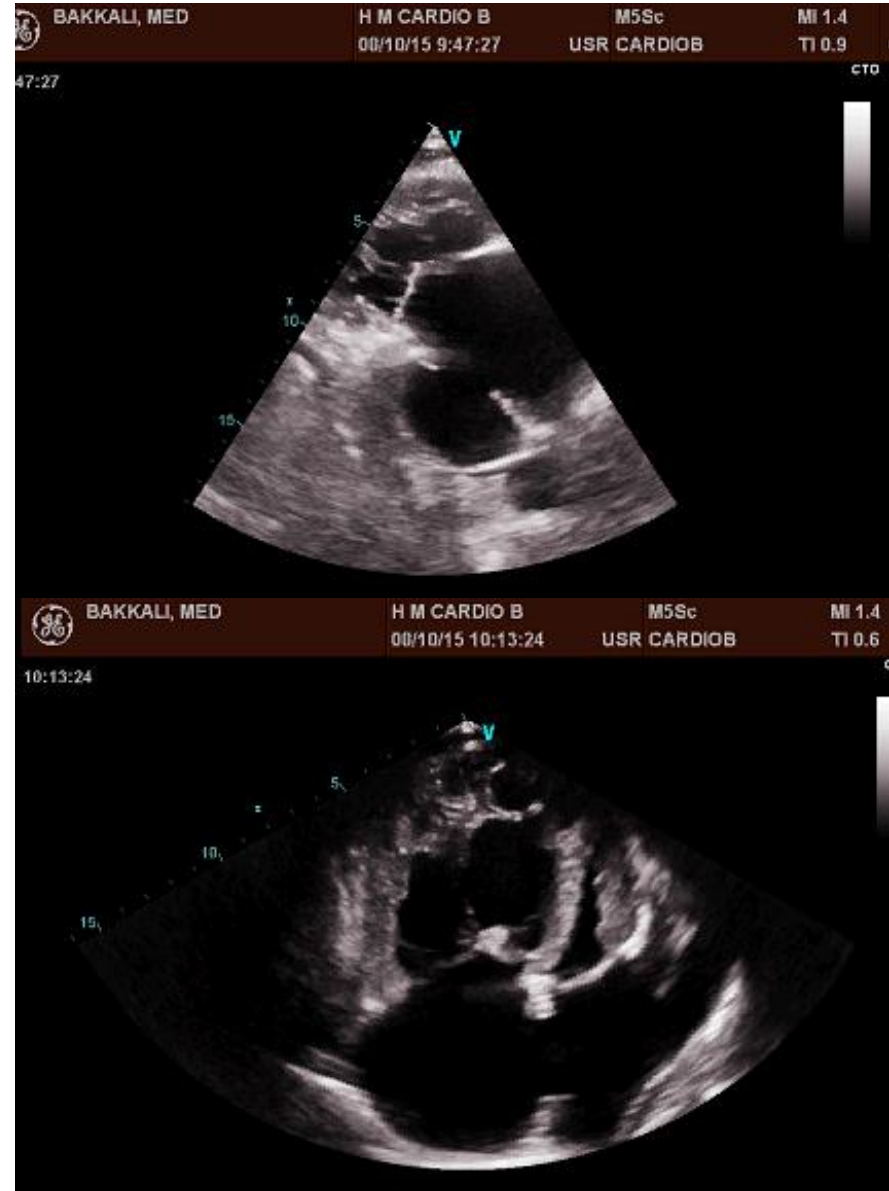
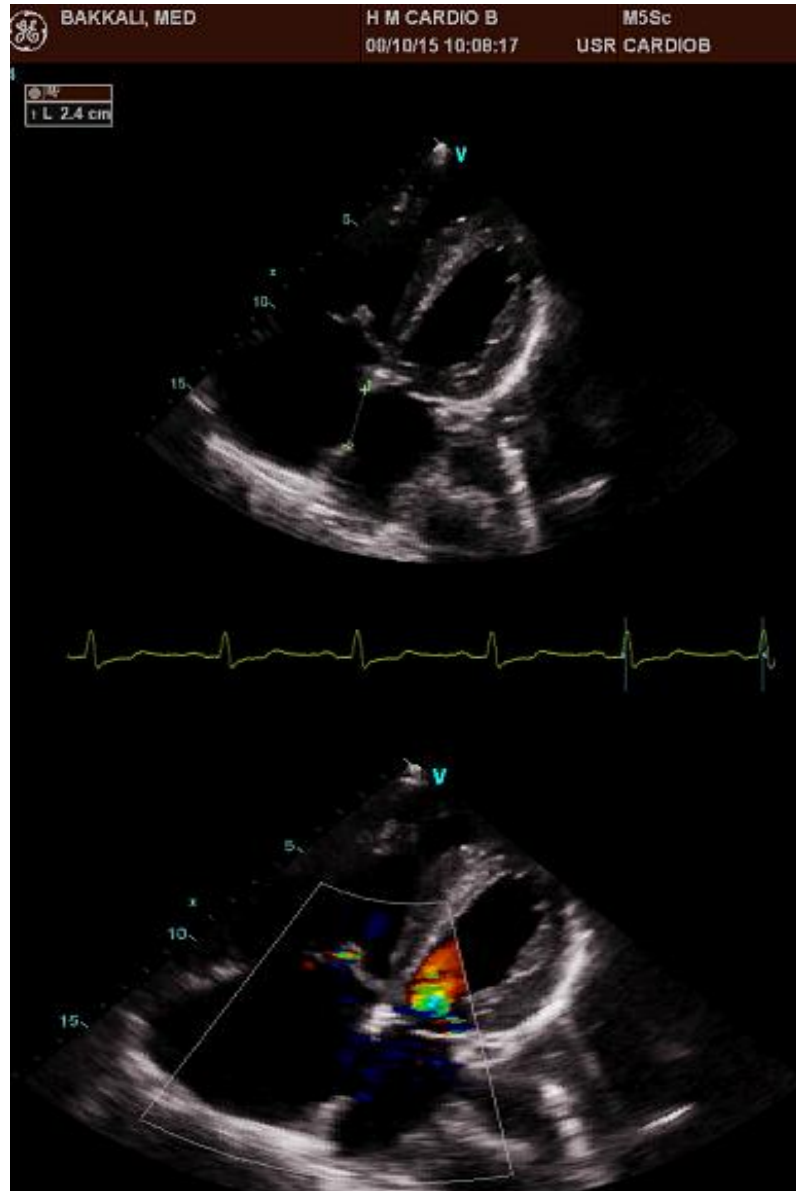
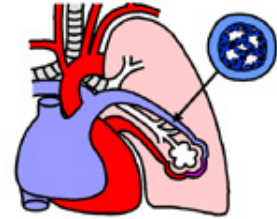
RADIO PULMONAIRE



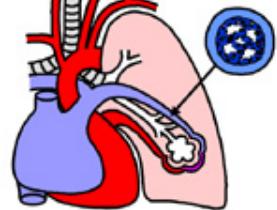
Pulmonale Arterielle Hypertonie (PAH)



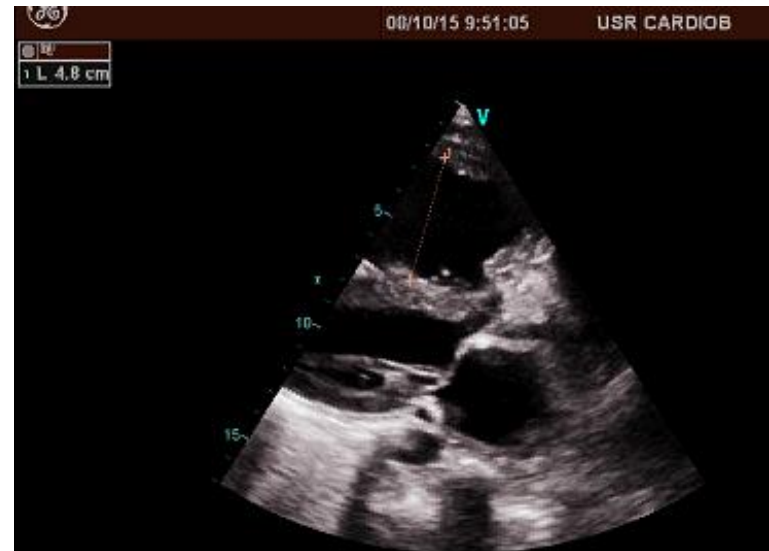
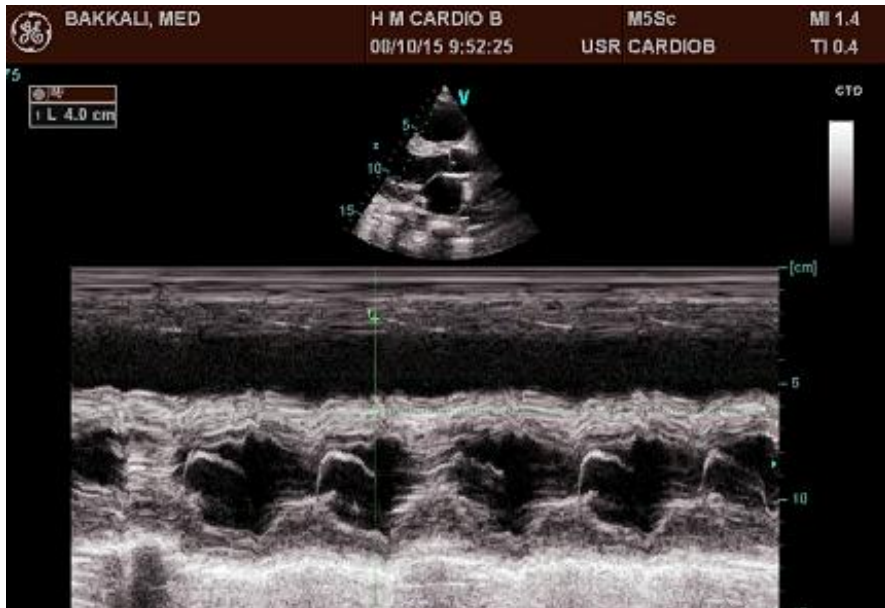
Echocardiographie



Echocardiographie

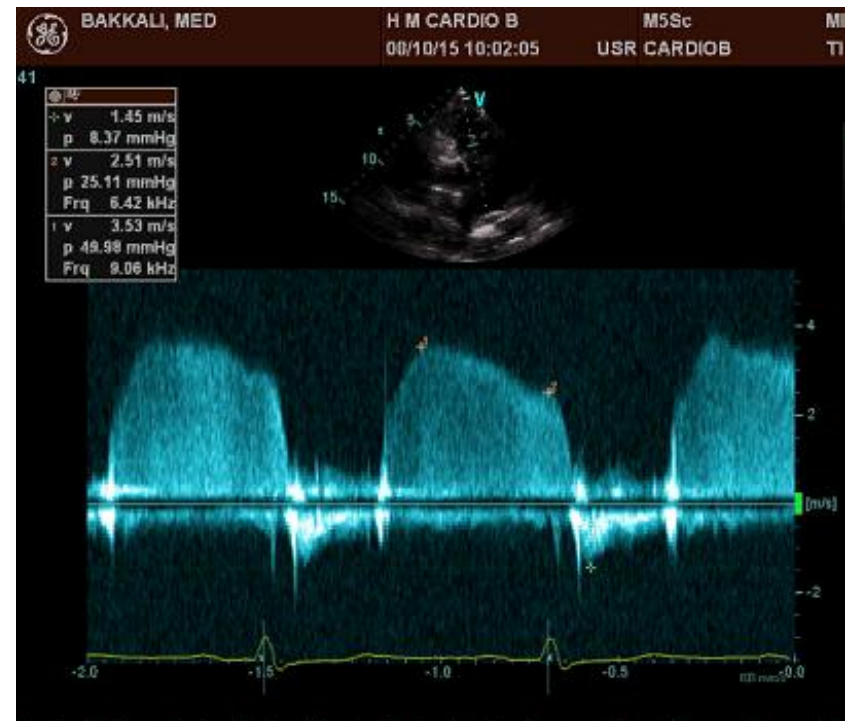
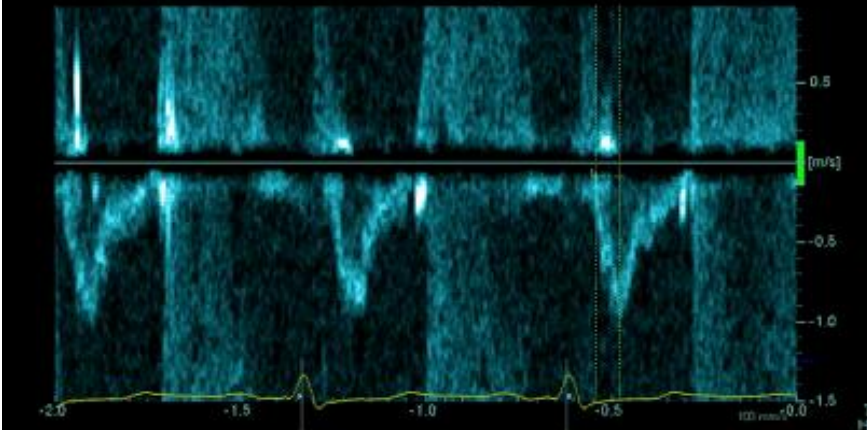
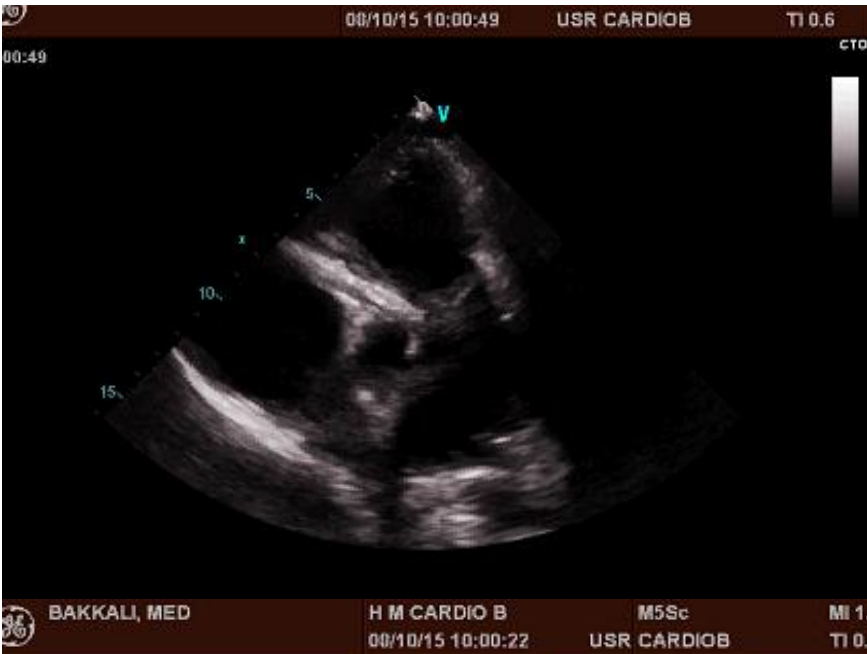
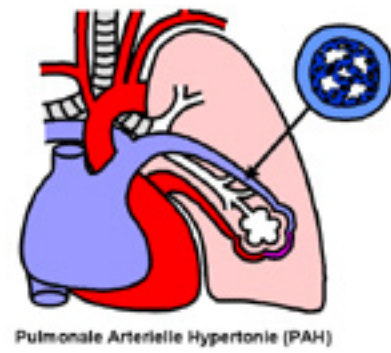


Pulmonale Arterielle Hypertonie (PAH)



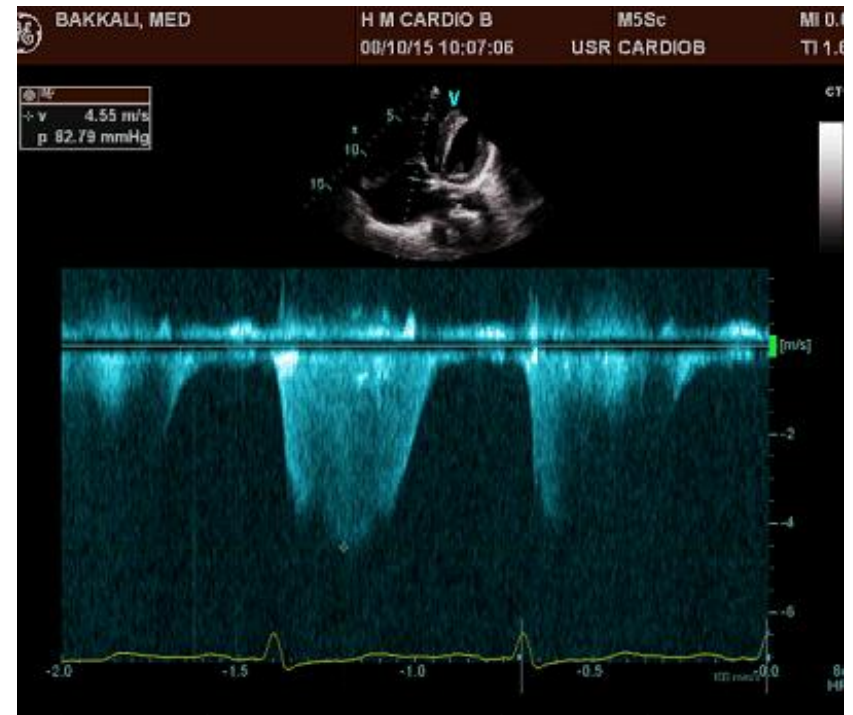
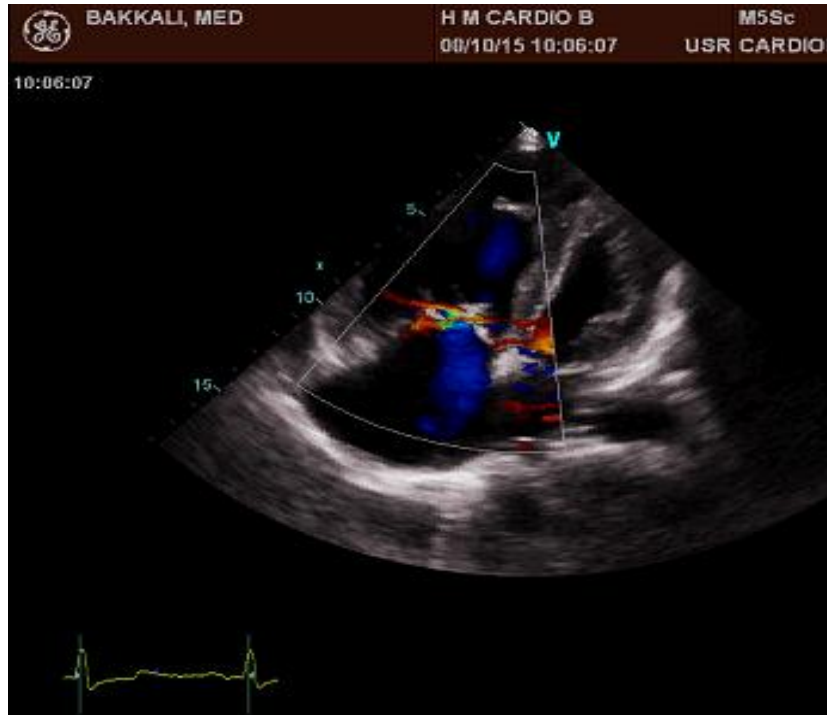
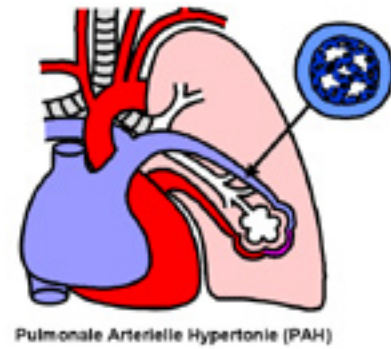
Ventricule droit dilaté

Echocardiographie



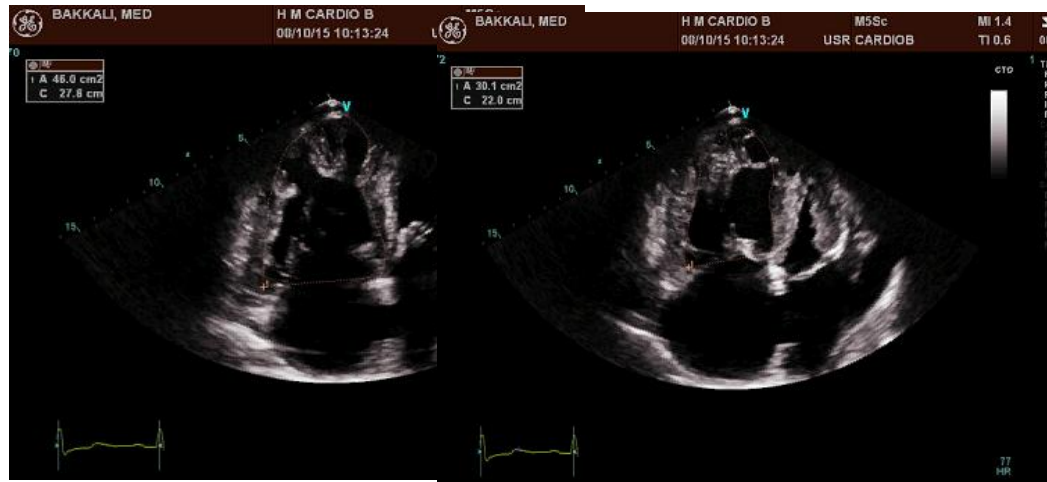
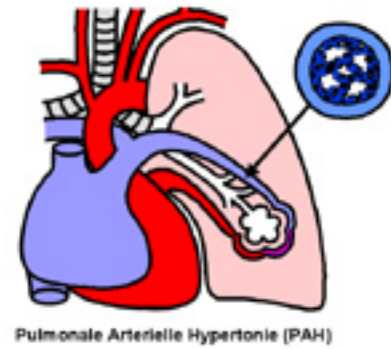
Flux pulmonaire et IP

Echocardiographie

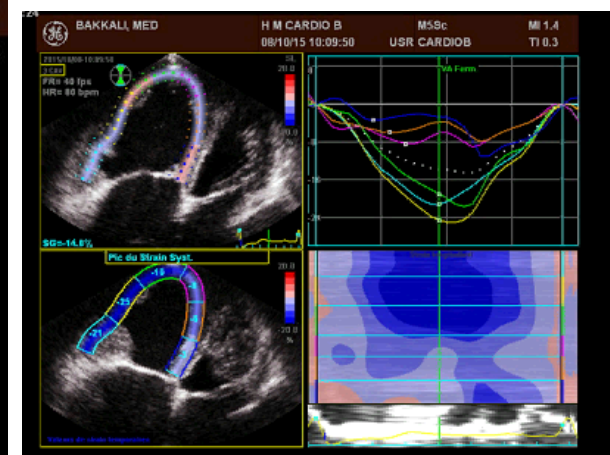


IT au D couleur et DC
 $V=4,6\text{m/s}$, $\rightarrow\text{PAPS}= 93\text{mmHg}$

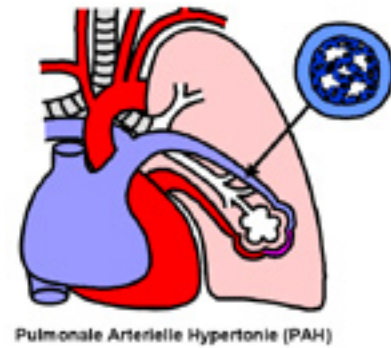
Echocardiographie



Fonction VD



BILAN PARACLINIQUE

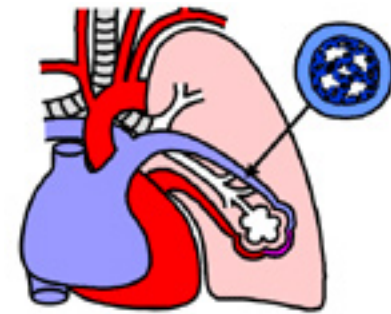


- **BIOLOGIE:**

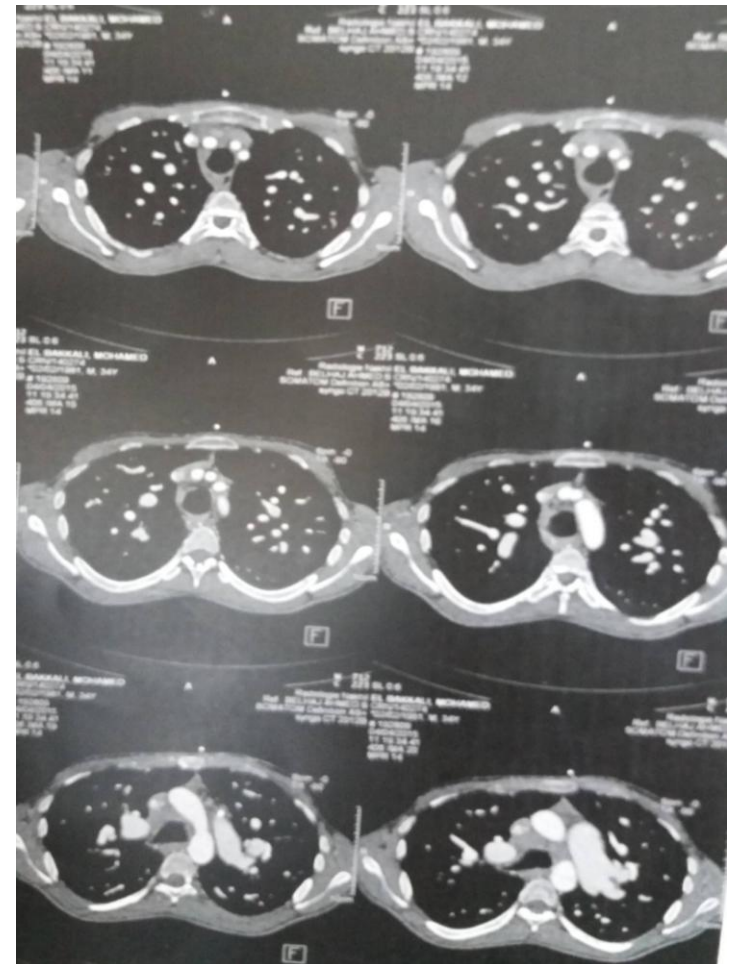
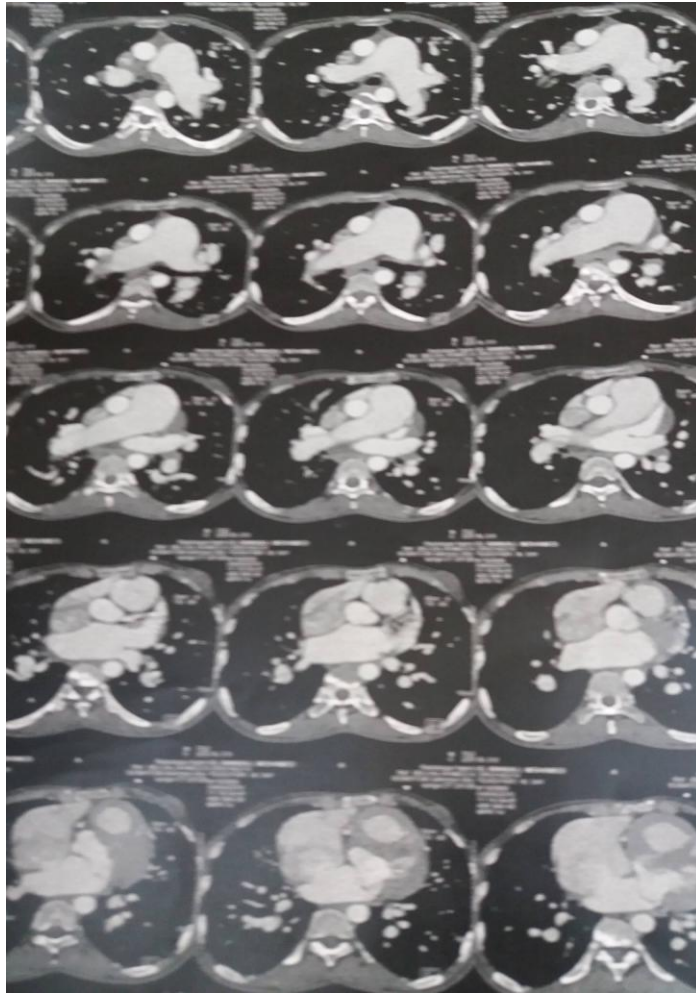
- NFS: Hb=18,6 g/100ml, Hte=57,7%, VGM=66,6
- TP spontané=50%
- fonction rénale: urée=0,24 créatinine=13
- TSH nle
- Sérologies HIV et hépatite virale B et C négatives

- **Echographie abdominale** normale
- **Scintigraphie pulmonaire** normale
- **Angioscanner thoracique**

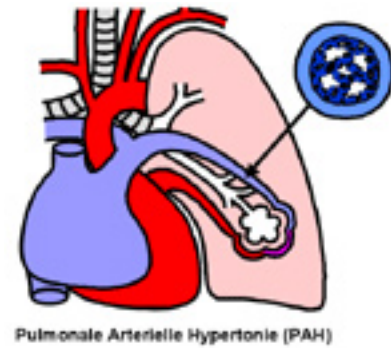
BILAN PARACLINIQUE



- **Angioscanner thoracique**, coupes millimétriques non encore faites pour éliminer une maladie veinoocclusive associée prévu ce mercredi



CATHETERISME DROIT

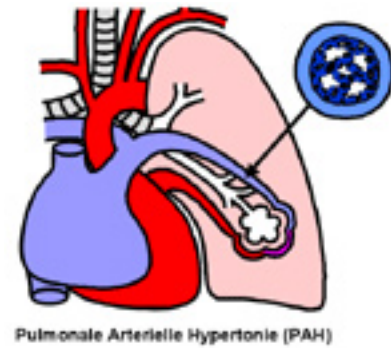


Fait le 13/04/2015 constantes hémodynamiques :

- FC=80/min , Rythme sinusal
- OD=7mmHg
- Aorte S/D mmHg
 - sous air 140/79/93
 - sous O2 et NO 140/50/70
- Artère pulmonaire
 - sous air 127/55/75 mmHg
 - sous O2 et NO 115/50/70
- Cap12mmHg
- Pas de réactivité artérielle pulmonaire
- RP/RS 0,8/1

→HTAP précapillaire présystémique aréactive à l'O2 et au NO

Mr M., 35 ans

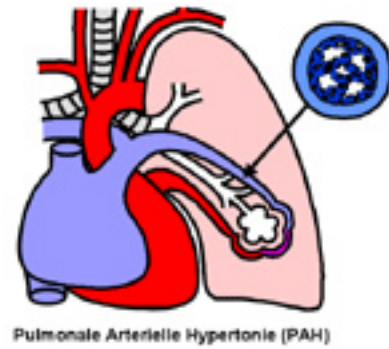


- Adressé en chirurgie cardiaque pour fermeture de CIA
- pas d'atcd cardiologique ou extracardiologique
- Symptôme: dyspnée stade 2 NYHA, SaO₂= 80%
- ECG,RX , ETT, angioscanner thoracique

→Diagnostic d'HTAP associée à une CIA ostium secundum de 24 mm de diamètre

Quel bilan?

Mr M., 35 ans



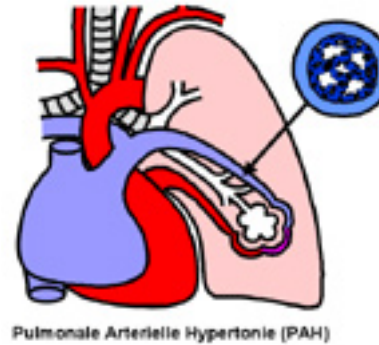
Classification ,Diagnostic positif ?

Diagnostic étiologique?

Stratification du risque?

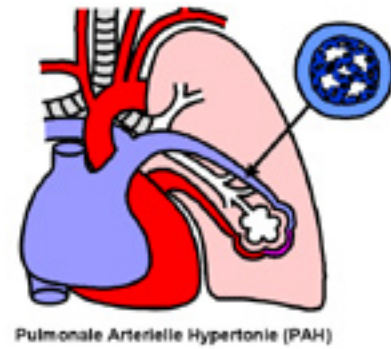
Traitement?

Bilan réalisé



- Clinique**: dyspnée stade 2 NYHA, SaO₂=80%, sous O₂ à 85%
- ECG**: RRS, axe droit, HAD, HVD
- Rx Pulmonaire**: grosses AP au niveau hilare, TAP dilaté
- ETT**: HTAP associée à une CIA OS de 24 mm de
- Biologie**: polyglobulie, microcytose
BNP=280 ng/l
GDS: non fait
- KT**: HTAP précapillaire aréactive,
- Angioscanner thoracique**(coupes millimétriques non encore faites):
- EFR**: Sd restrictif sévère
- Pléthysmographie**: DLCO non encore fait
- Scinti pulmonaire** normale

Stratification du risque



-Signes cliniques : ICD = 0

progression des

symptômes

syncope

0

classe fonctionnelle II WHO

-test de marche 6 min

-EE pic VO₂ non faite

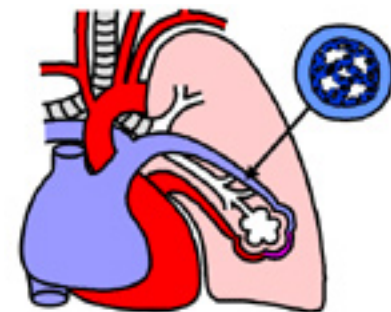
-BNP = 280 ng/l

-ETT surface OD : 32 cm²

-KT RP non données sur le compte rendu

IC

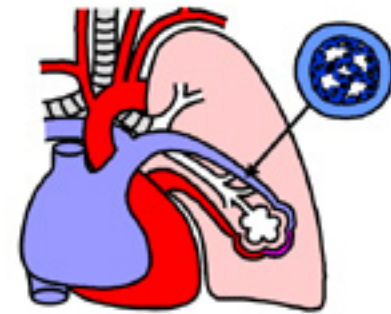
Stratification du risque



Pulmonale Arterielle Hypertonie (PAH)

Determinants of prognosis ^a (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope ^b	Repeated syncope ^c
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	Peak VO ₂ >15 ml/min/kg (>65% pred.) VE/VCO ₂ slope <36	Peak VO ₂ 1–15 ml/min/kg (35–65% pred.) VE/VCO ₂ slope 36–44.9	Peak VO ₂ <11 ml/min/kg (<35% pred.) VE/VCO ₂ ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/ml	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm ² No pericardial effusion	RA area 18–26 cm ² No or minimal, pericardial effusion	RA area >26 cm ² Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m ² SvO ₂ >65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m ² SvO ₂ 60–65%	RAP >14 mmHg CI <2.0 l/min/m ² SvO ₂ <60%

Traitement



Pulmonale Arterielle Hypertonie (PAH)

- Supplémentation en fer
- anticoagulant non donné car TP spontané à 50% , difficulté d'accès pour suivi INR, pas de signe IC ni de thrombose AP
- ttt spécifique HTAP non prescrit (pas de sécurité sociale): trop cher
- **Et surtout : ne pas fermer la CIA**
- suivi tous les 3 à 6

Recommendations	Class ^a	Level ^b
Bosentan is recommended in WHO-FC III patients with Eisenmenger syndrome	I	B
Other ERAs, PDE-5is and prostanoids should be considered in patients with Eisenmenger syndrome	IIa	C
In the absence of significant haemoptysis, oral anticoagulant treatment may be considered in patients with PA thrombosis or signs of heart failure	IIb	C
The use of supplemental O ₂ therapy should be considered in cases in which it produces a consistent increase in arterial O ₂ saturation and reduces symptoms	IIa	C
If symptoms of hyperviscosity are present, phlebotomy with isovolumic replacement should be considered, usually when the haematocrit is >65%	IIa	C
The use of supplemental iron treatment may be considered in patients with low ferritin plasma levels	IIb	C
Combination drug therapy may be considered in patients with Eisenmenger syndrome	IIb	C
The use of CCBs is not recommended in patients with Eisenmenger syndrome	III	C



2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

The Joint 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: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

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