Anomalies d'EBSTEIN
2026

Dr Xavier Iriart
Bordeaux-Pessac
FRANCE

Maladie d'Ebstein

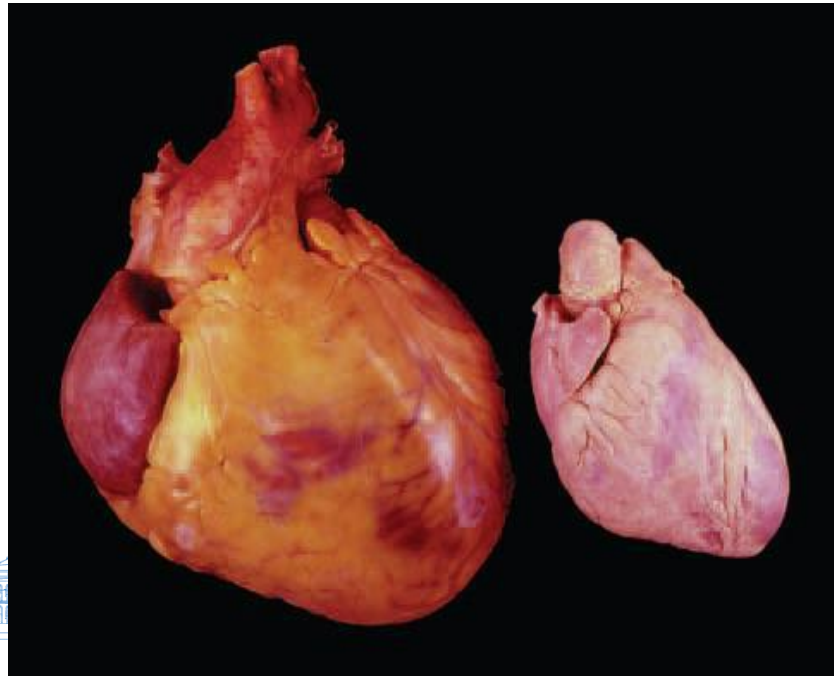
- Anatomie de la valve tricuspide
- La maladie d'Ebstein
  - Définition et anatomie
  - Nomenclature et classifications
  - Physiopathologie
  - Anomalies ventriculaires droites
  - Anomalies ventriculaires gauches
- Histoire naturelle et facteurs de pronostic
- Aspects cliniques
- Examen complémentaires
- Prise en charge

Wilhelm Ebstein, 1866,
Professor of Medicine
Gettingen, Germany

Anomalie d'Ebstein

- Willem Ebstein en 1866
- patient de 19 ans
  - insuffisance tricuspide
  - Cyanose
  - Dyspnée
  - Palpitations
  - turgescence jugulaire
  - cardiomégalie

Wilhelm Ebstein, 1866,
Professor of Medicine
Gettingen, Germany



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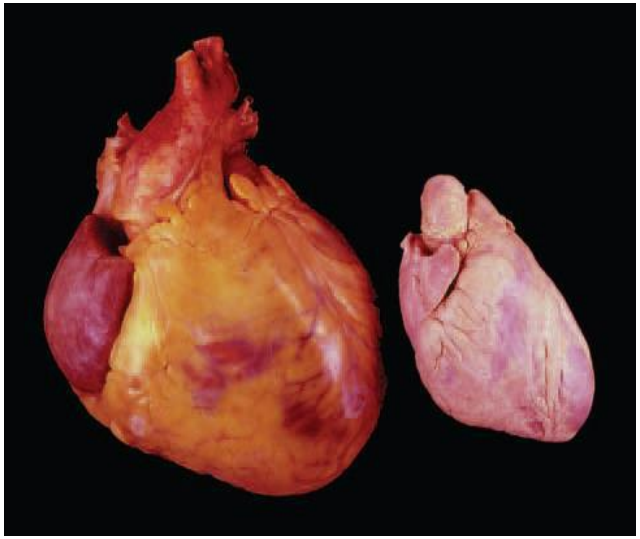
Description autopsique initiale

Wilhelm Ebstein, 1866, Professor of Medicine Gettingen, Germany



feuillet tricuspide antérieur élargi et fenestré

feuillets inférieur et septal hypoplasiques, épaissis, adhérents à la paroi du ventricule droit

Atrialisation, dilatation, amincissement de la paroi VD

OD très dilatée + FOP

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Données épidémiologiques

- < 1% cardiopathies congénitales

- 1/ 210 000 naissances vivantes

(mots fœtales in utero)

- Embryofoetopathie aux sels de lithium

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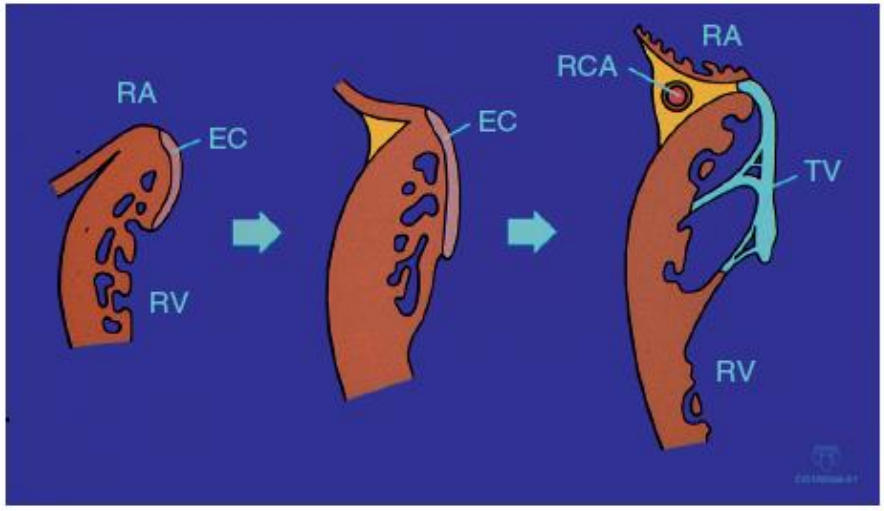
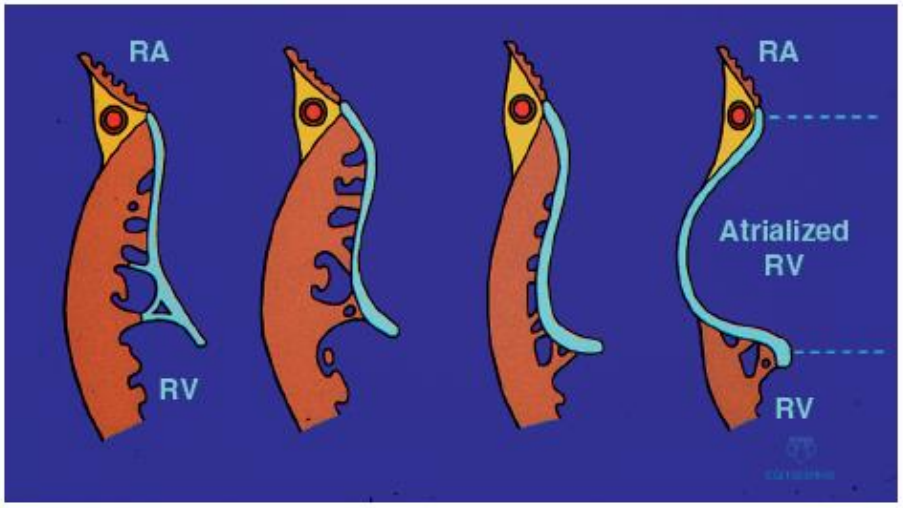
Données embryologiques

- Formation des feuillets par délamination
- Expansion de la lumière ventriculaire entre myocarde compacté et myocarde trabéculé



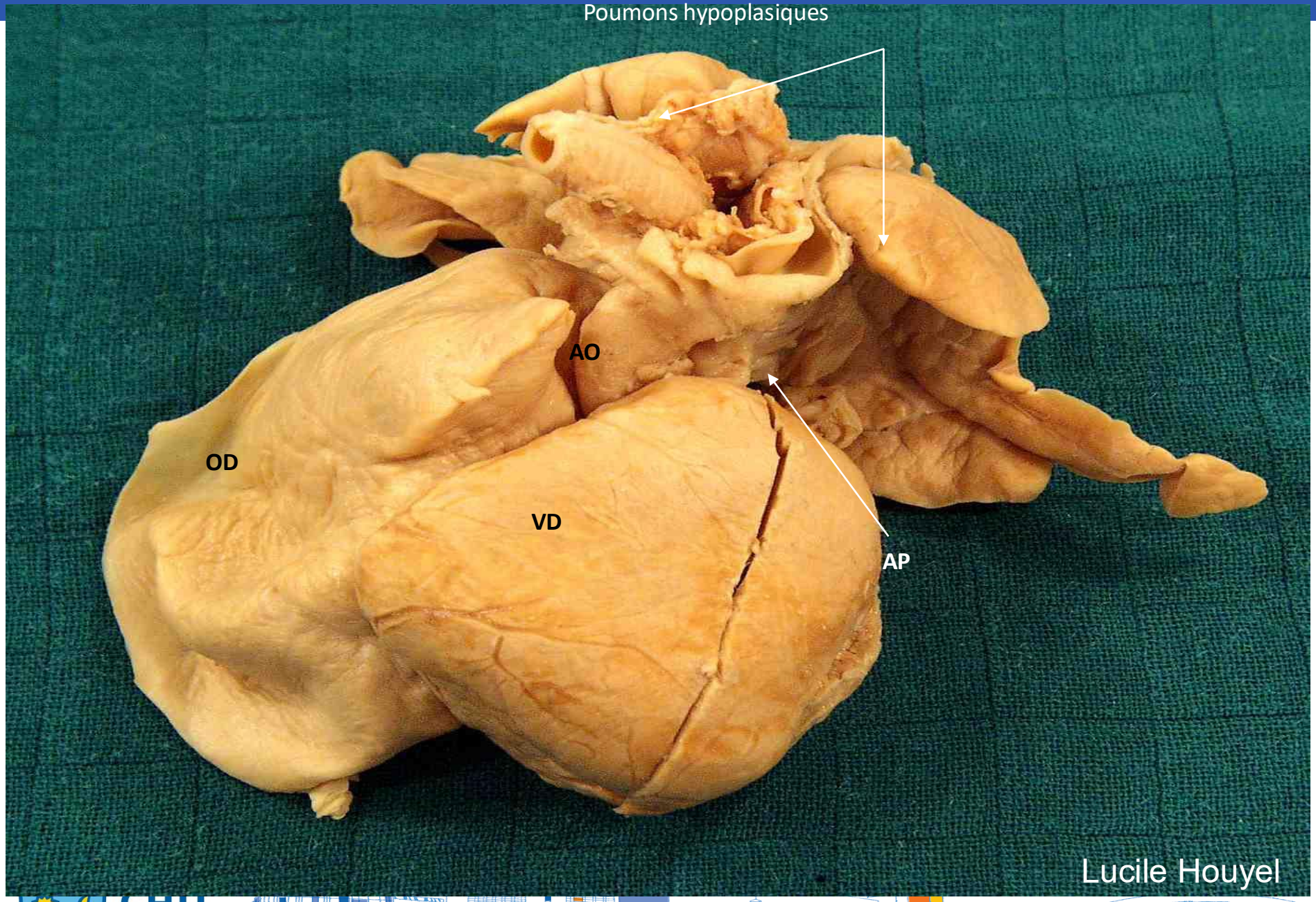
Tricuspide normale

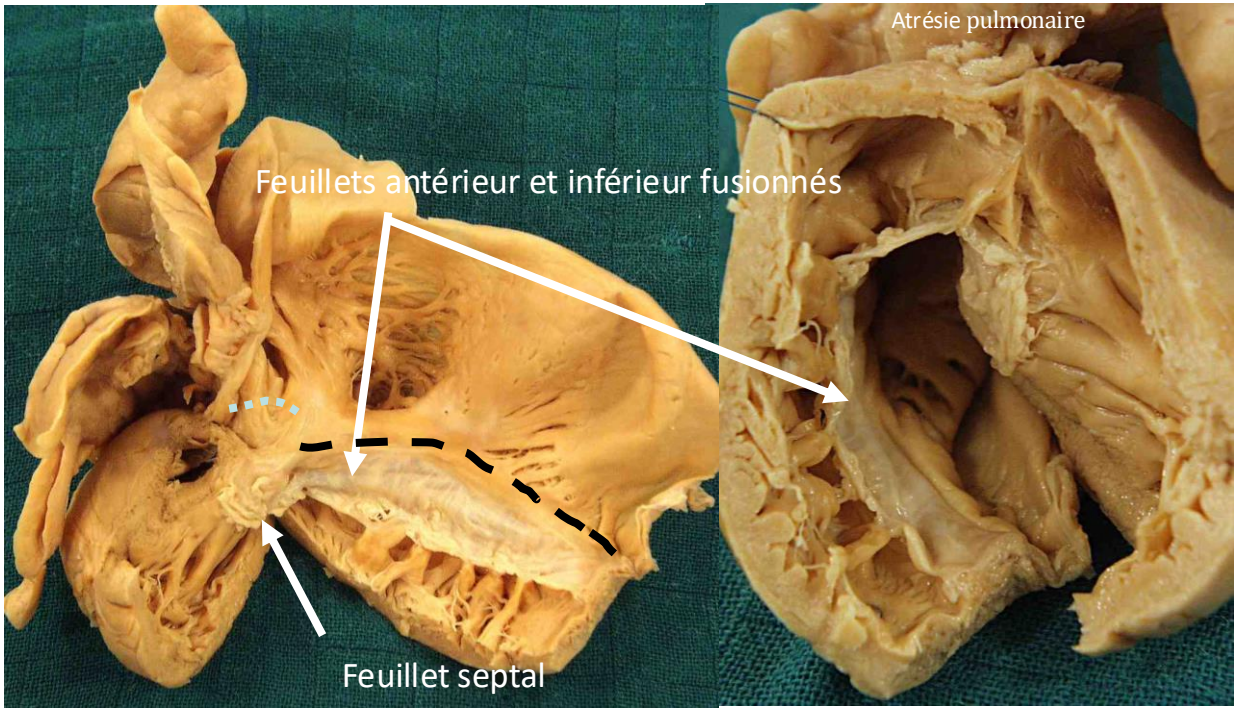


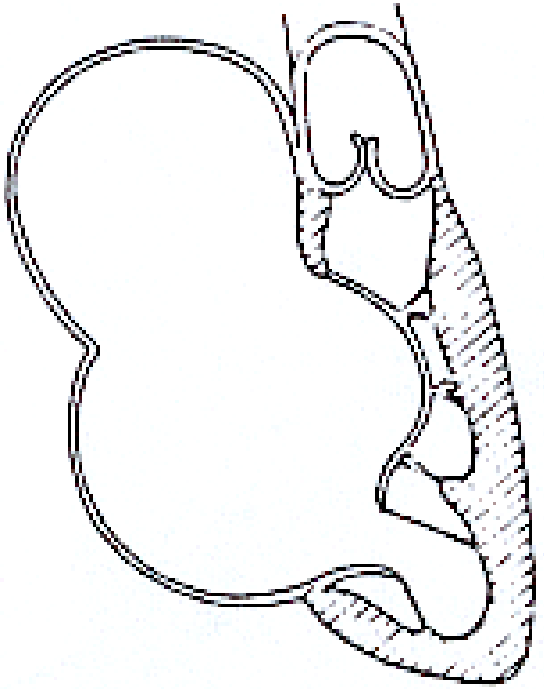
Ebstein

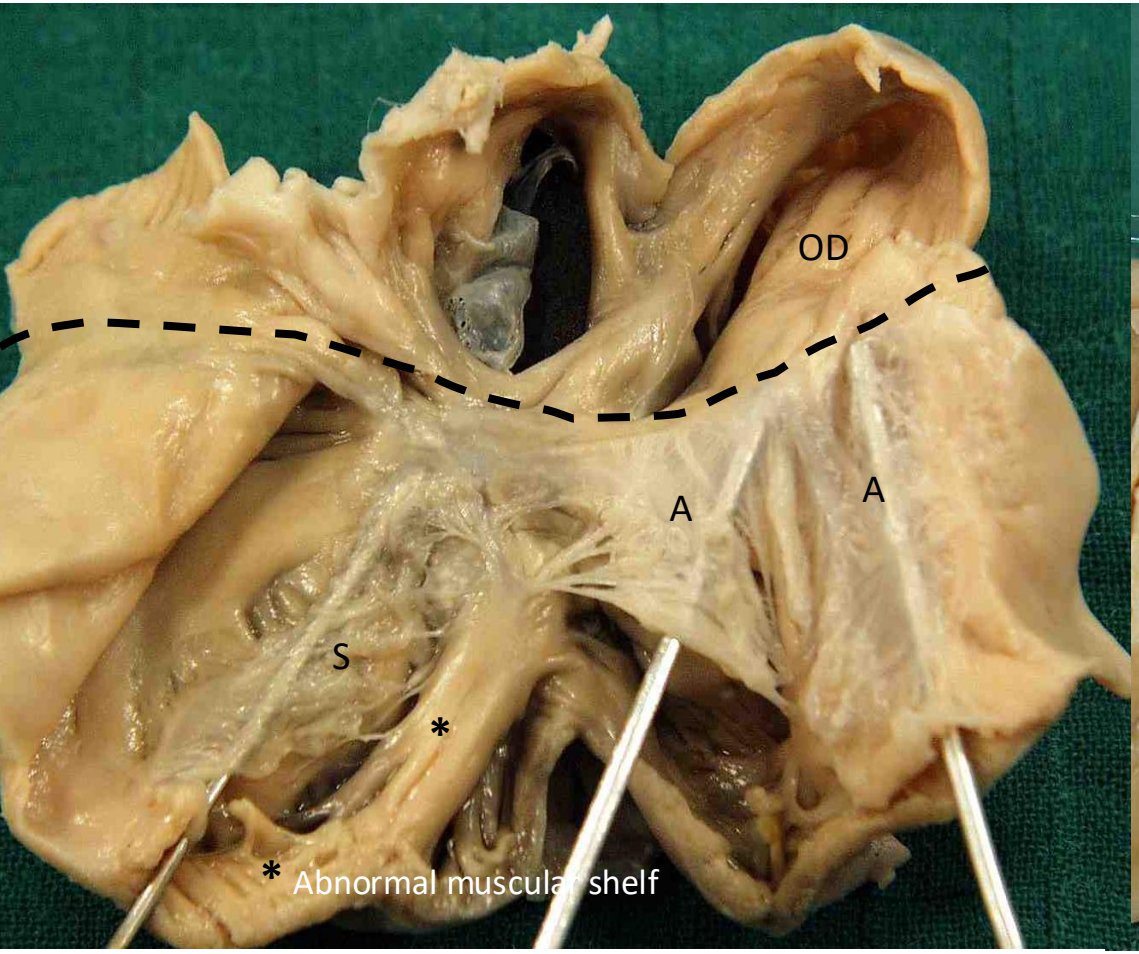
Oosthoek et al. Anat Embryol 1998

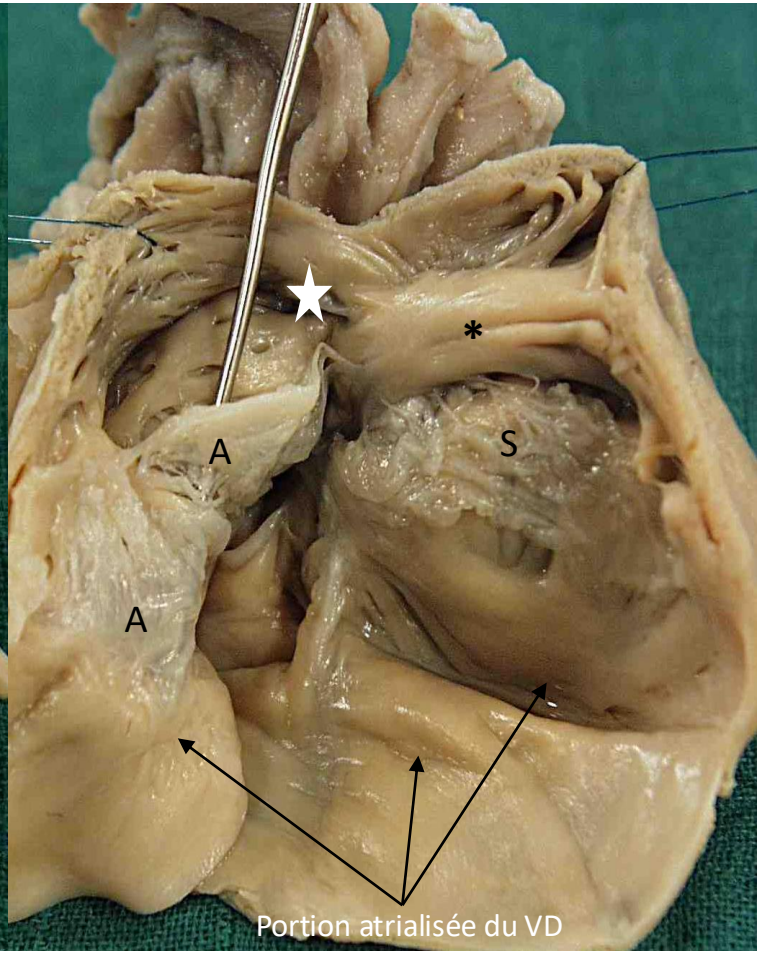
Poumons hypoplasiques




Feuillets antérieur et inférieur fusionnés
Feuillet septal
Atrésie pulmonaire







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Lucile Houyel

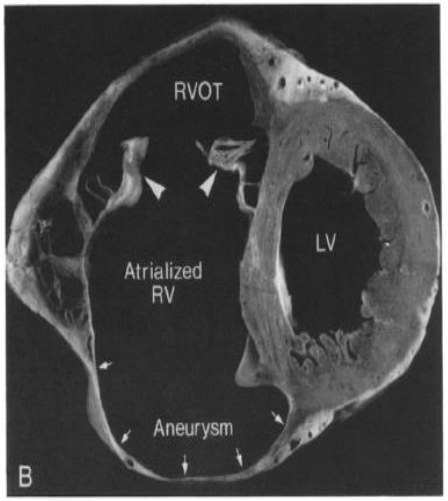
Anomalie du ventricule droit

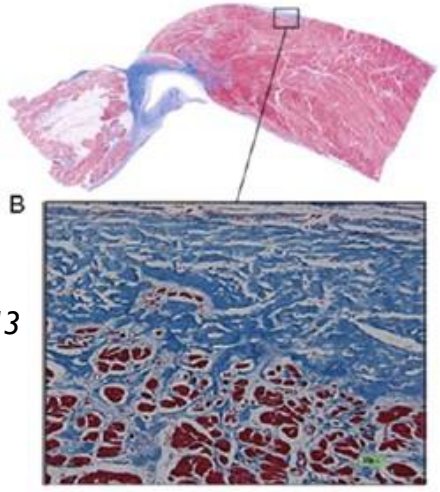
- Myocardiopathie du ventricule droit
  - Anatomie
  - histologie
  - fonction

anormales

- Dilatation de la jonction AV (incompétente)
- Chambre d'admission VD atrialisée
  - endocarde épais, fibreux et lisse
  - trabéculations musculaires anormales
  - paroi postérieure anévrismale très mince (démusculariséé)
- VD fonctionnel tripartite (variable)
  - Infundibulum
  - apex VD
  - cavité antérolatérale

Forme de croissant





Edwards WD. Prog Pediatr Cardiol. 1993
Assenza I G, Valente AM, Geva T, et al. Eur Heart Journ. 2013

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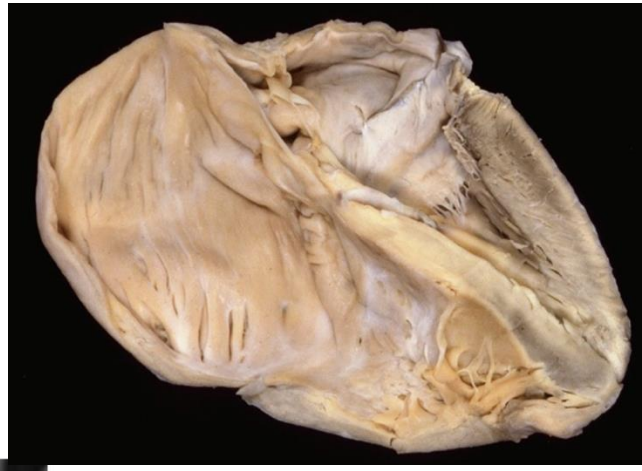
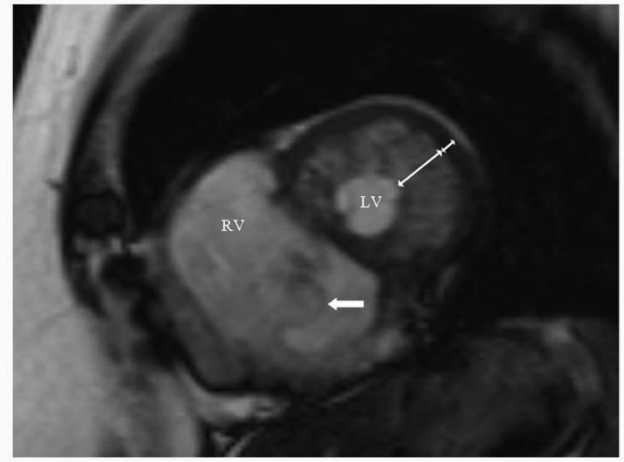
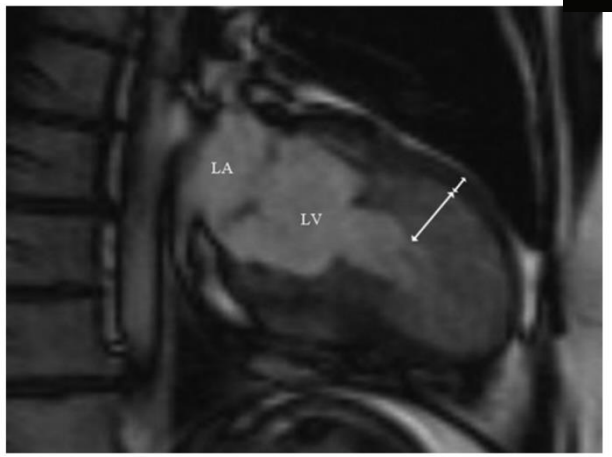
Anomalie du VG

- Anomalies de l'endocarde (épais) et fibrose endo-myocardique (risque de dysfonction VG)
- Anomalies génétiques (MYH7): non compaction



Courtesy of A cook UCL London





Gerlis L, 1993; Attenhofer 2005
Lee A, Cook AC et al 1994
Postma A 2011; Bettinelli A 2013

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Anomalies associées

CIA (50-70%)

CIV

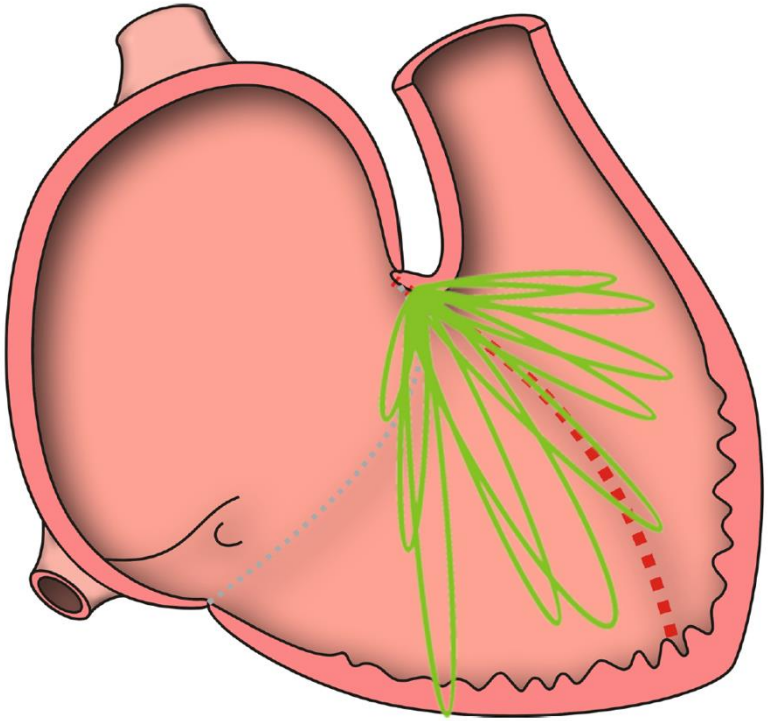
STÉNOSE/ATRÉSIE PULMONAIRE

DOUBLE DISCORDANCE

WPW (30%)

Déplacement rotationnel de l'orifice valvulaire tricuspide

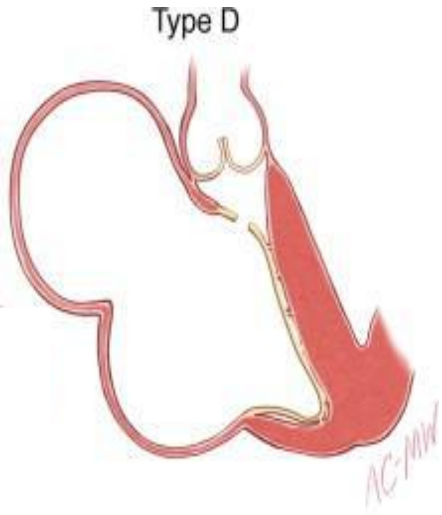
Rotational Displacement in Ebstein's Malformation



Adapted from Schreiber C, Cook A et al J Thorac Cardiovasc Surg. 1999

Spectre anatomique et clinique

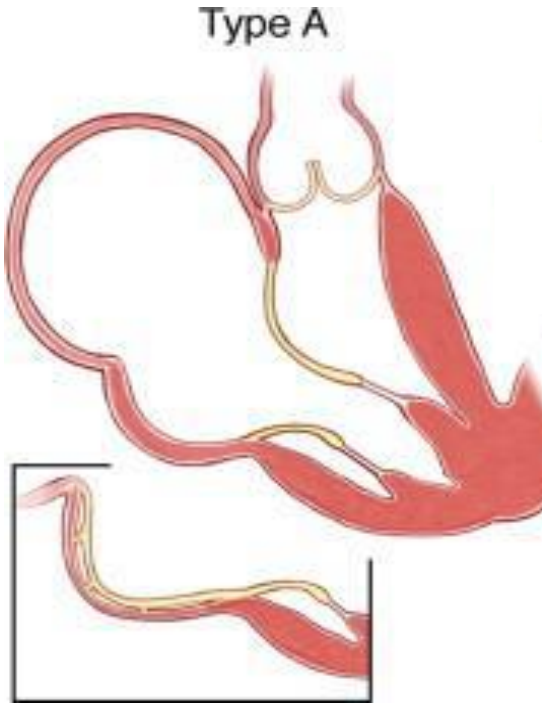
- Variable: stade d'arrêt du développement tricuspide



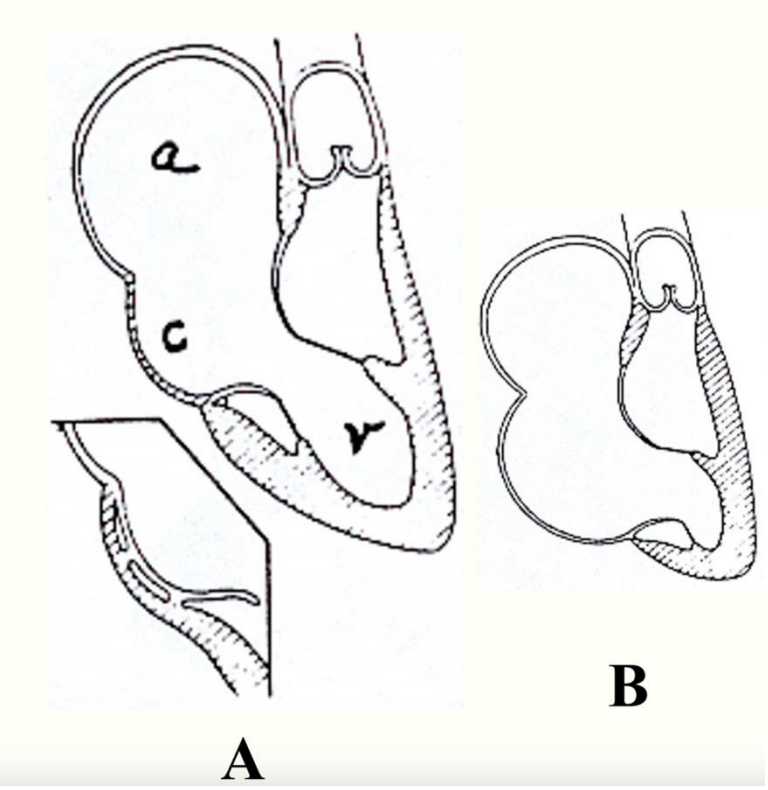
- Formes précoces = formes sévères:
  - feuillet inférieur absent, feuillet ant visible +/-non démusculariséé. Feuillet septal non délaminé
  - piliers non individualisables, pas de cordages, et le feuillet antérieur inséré sur un « éperon musculaire anormal VD non développé
  - ouverture tricuspide antérieure et dirigée vers la gauche
  - infundibulum VD fonctionnel (développée à partir de la voie d'éjection embryonnaire)
  - VD faussement "dilaté", cavité fctelle hypoplasique. La partie "dilatée" non fonctionnelle (atrialisée)

Spectre anatomique et clinique

- formes anatomiquement moins sévères = plus tardive
  - ouverture apicale
  - feuillets mieux formés, mais peu mobiles
  - cordages courts, petits piliers épars,
  - délamination incomplète du feuillet septal définissant l'anomalie d'Ebstein.



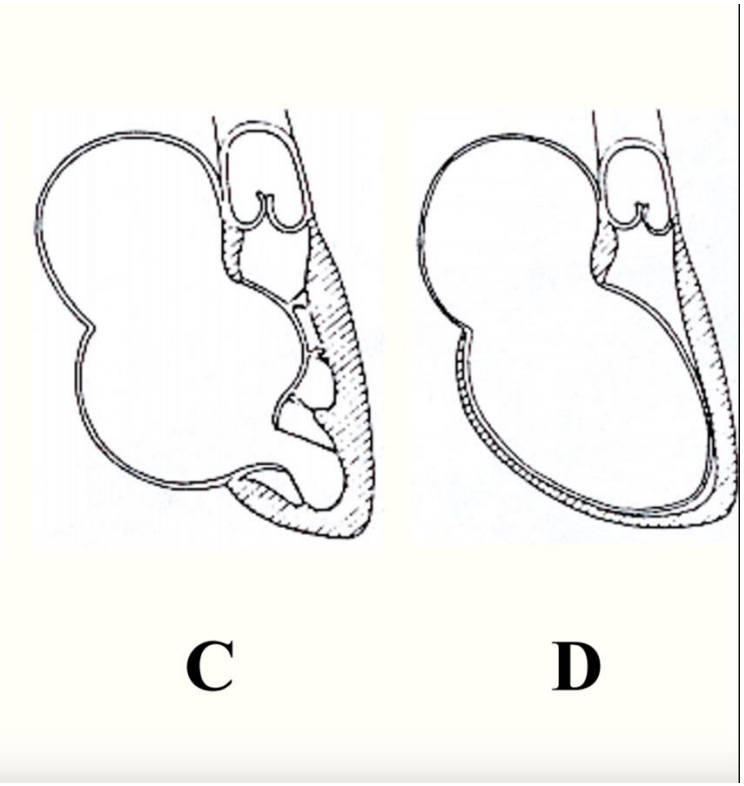
Classification de Chauvaud/Carpentier



Classification de Carpentier

- IT = restriction des mouvements valvulaires, cordages du bord libre courts, tractés par dilatation du VD, inclusion du pilier dans la paroi
- Type A : déplacement minime de la valve septale, petite chambre atrialisée
- Type B : ↑ déplac. ↑ ch. atrialisée, ↓ mvmts. valve ant. à cordages courts

Classification de Chauvaud/Carpentier



- Type C : absence de valves sept. et post., ↑↑↑ch.atrialisée
  anévrysmale, mvmts valve ant. très limités
  trabéculations entre valve ant et infundibulum VD
  → sténose infundibulaire

- Type D : valves accolées à la paroi ventriculaire ne laissant qu'une toute petite chambre de chasse
  chambre atrialisée / VD quasi anévrysmal = Uhl

Physiopathologie

- Altération fonctionnelle (volume + fonction) VD
- VD atrialisé = réservoir passif et distensible (limitation précharge VD fonctionnel pendant systole atriale)

Dysfonction atrio-ventriculaire

- IT sévère
  - Diminution flux antérograde
  - Diminution précharge VG.
  - dilatation progessive OD (+ CIA)

Forme fœtale grave

Forme néonatale
(atrésie anat ou fctelle)

Forme classique
± limitation d'effort
± cyanose d'effort
± arythmie

Aspects cliniques

Eventail clinique large

Symptômes cardinaux

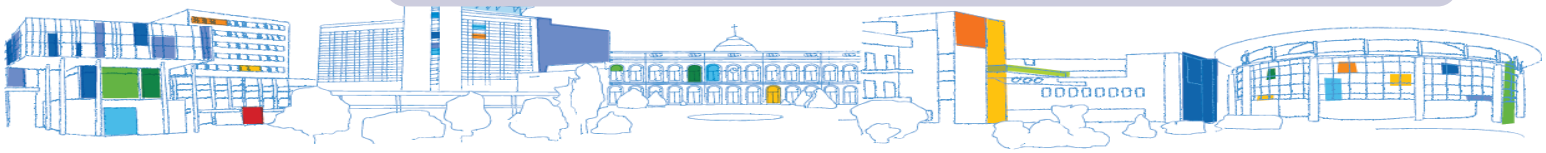
- cyanose
- Insuffisance cardiaque
- Arythmie et mort subite

forme néonatale sévère

forme classique enfant, adolescent, adulte

- Limitation fonctionelle (VO2)
- Rechercher cyanose d'effort
- arythmie

forme tardive: insuffisance cardiaque tardive de la personne âgée



Forme néonatale

Déterminer flux antérograde à travers la valve pulmonaire

Atrésie fonctionnelle (IT, fonction VD)

Atrésie anatomique

Perfusion pulmonaire antérograde

Flux dans CA canal artériel

IP postnatale (ou même parfois anténatale): risque de défaillance hémodynamique par la création d'un shunt circulaire

Comprendre les stratégies de réanimation postnatale

O2 ++++

NO

Prostine

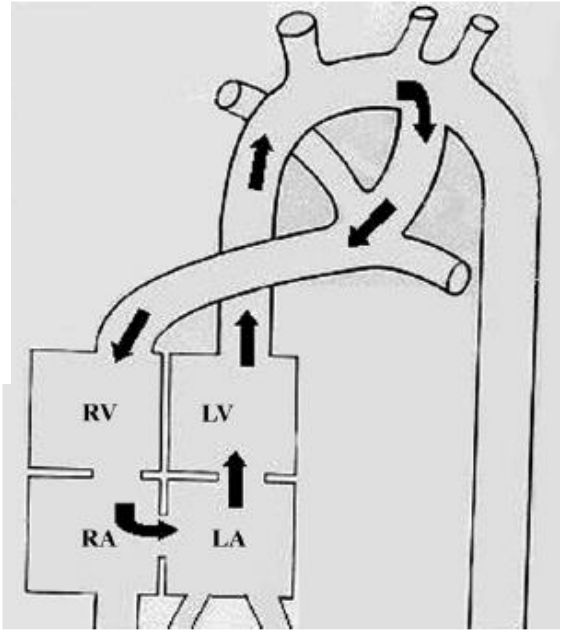
-ECMO

Outcomes and Predictors of Perinatal Mortality in Fetuses With Ebstein Anomaly or Tricuspid Valve Dysplasia in the Current Era

A Multicenter Study

CLINICAL PERSPECTIVE

Ebstein anomaly and tricuspid valve dysplasia are rare congenital tricuspid valve malformations associated with high perinatal mortality. Previous literature has consisted of single-center series, often spanning several decades. We report a series of 243 fetuses with Ebstein anomaly or tricuspid valve dysplasia from 23 centers across North America in the recent era. Unfortunately, perinatal mortality remained high at 45%, with one-third of patients not surviving to neonatal hospital discharge. Independent risk factors for mortality included gestational age at diagnosis of <32 weeks, larger tricuspid valve annulus z-score, the presence of pulmonary regurgitation, and a pericardial effusion. The presence of pulmonary regurgitation, in particular, signifies circular shunt physiology, which often culminated in mortality. An understanding of this unique physiology in utero may help clinicians better counsel expectant parents, develop and pursue novel treatment and perinatal management strategies, and ultimately improve mortality for fetuses with this rare and complex disease.



Freud LR et al.Circulation 2015

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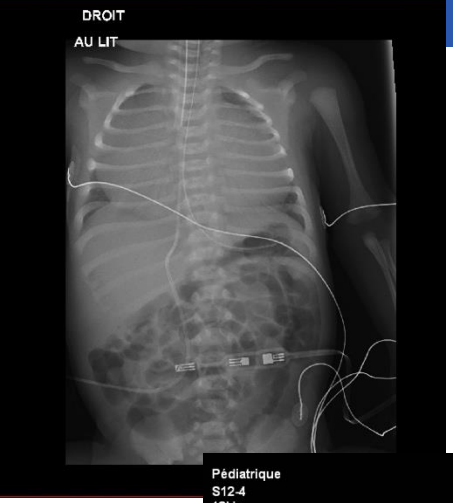
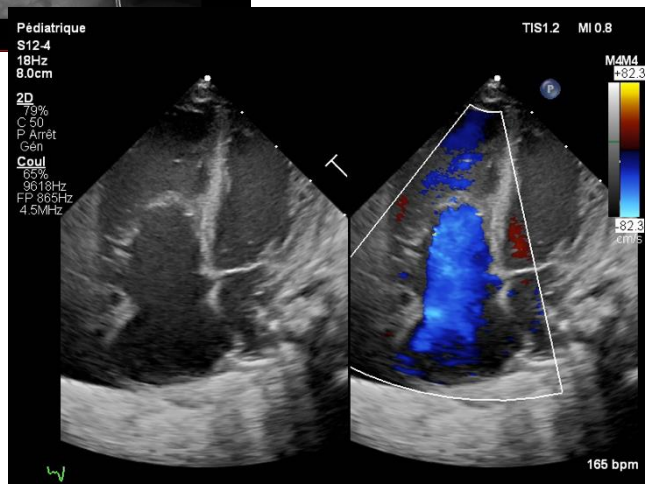
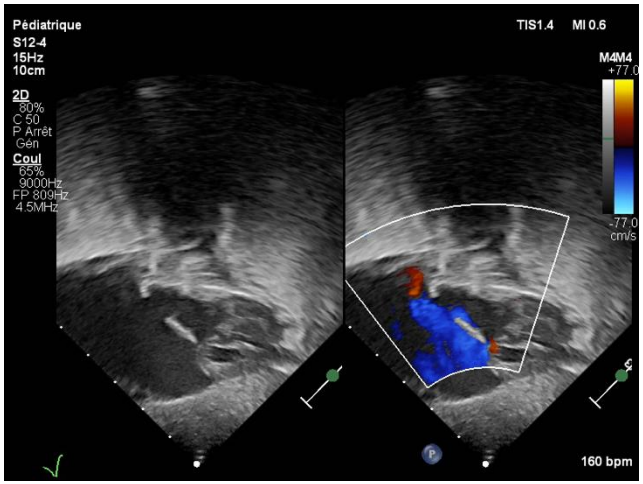
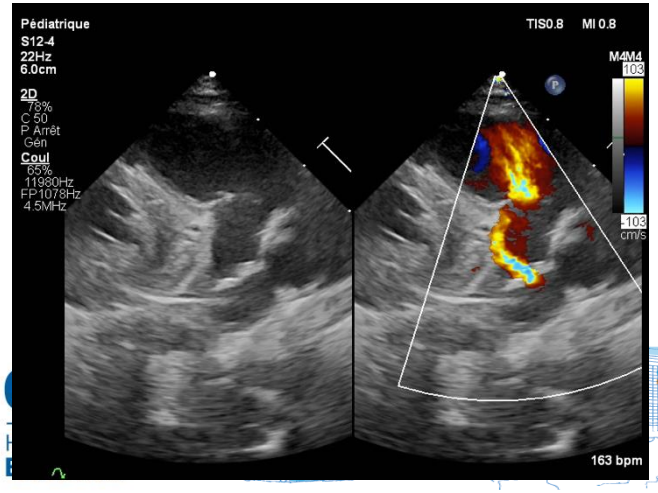


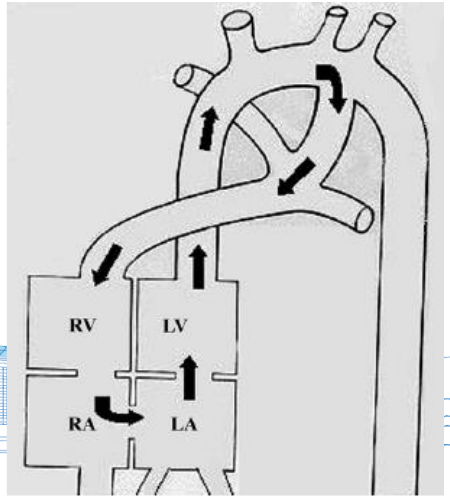
Neo natal assessment

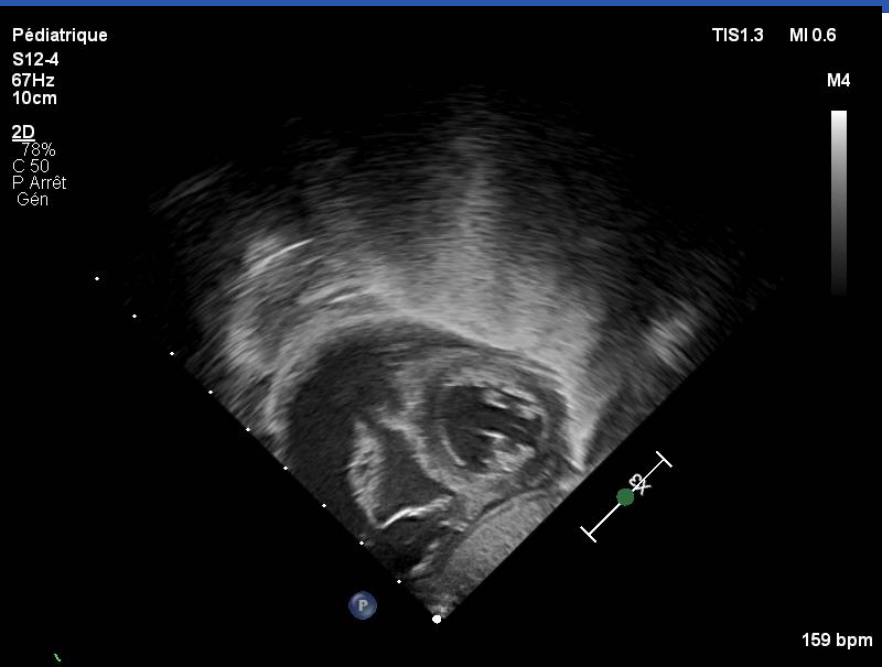
- Ebstein anomaly with circulatory shunt

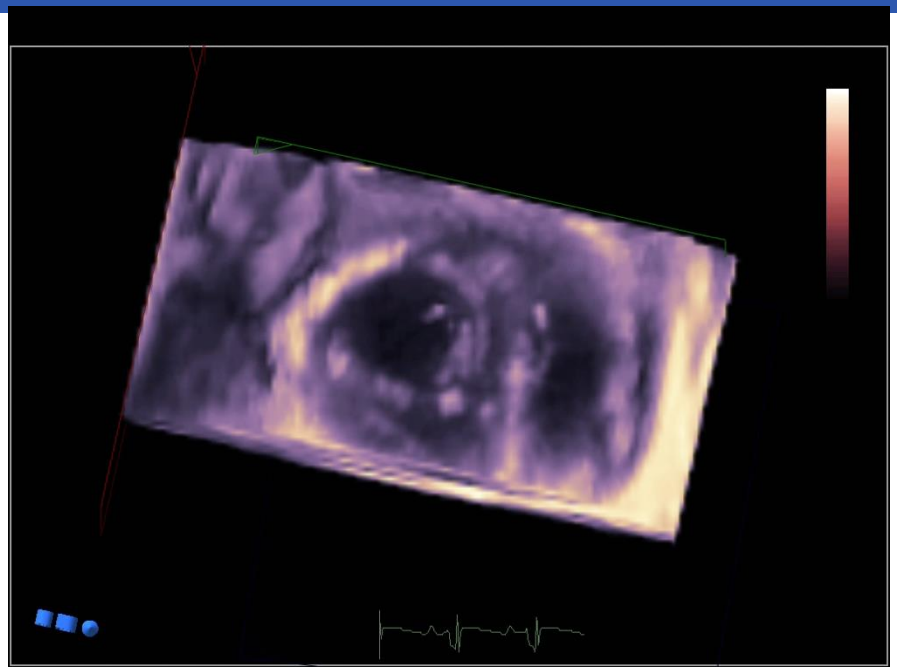


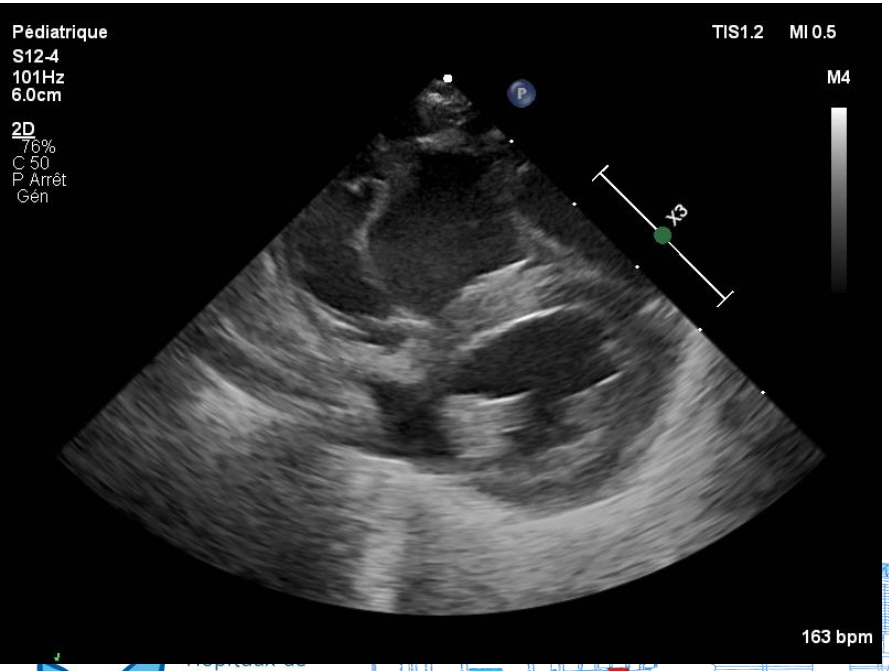






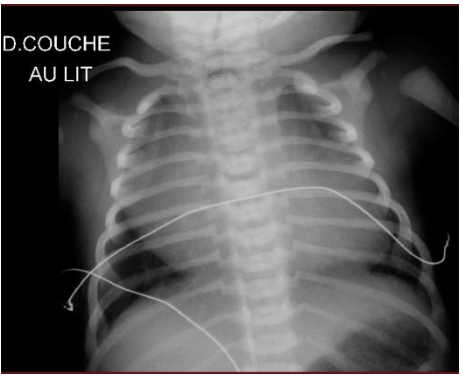

Pédiatrique
S12-4
67Hz
10cm
2D
78%
C 50
P Arrêt
Gén
TIS1.3
MI 0.6
M4
159 bpm

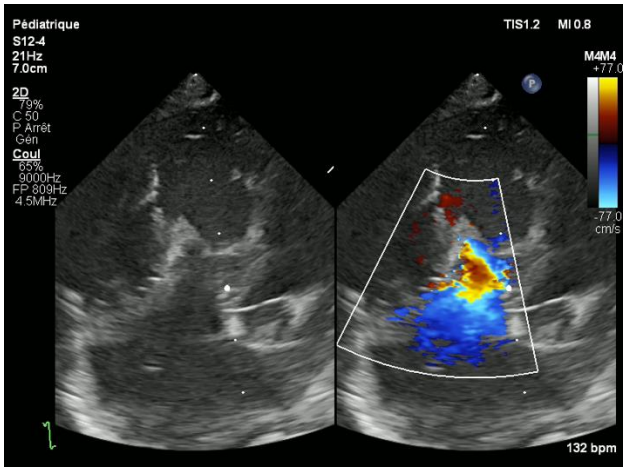


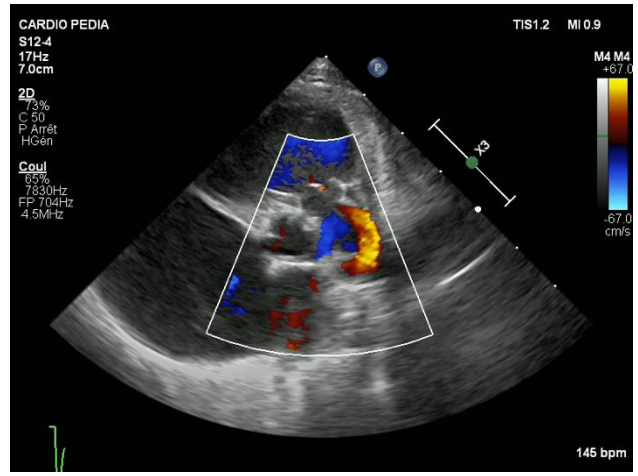

Pédiatrique
S12-4
101Hz
6.0cm
2D
76%
C 50
P Arrêt
Gén
TIS1.2
MI 0.5
M4
163 bpm

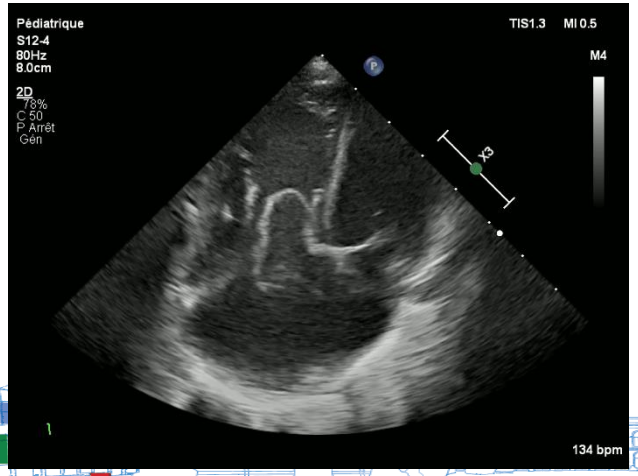
Neo natal assessment

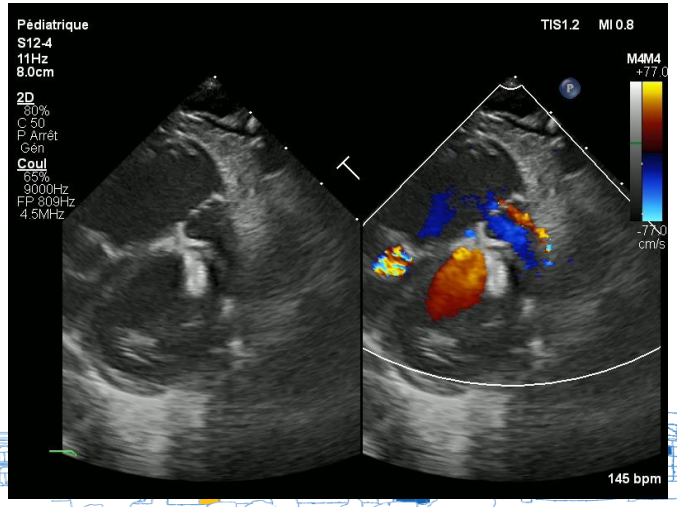
- Ebstein anomaly with no circulatory shunt











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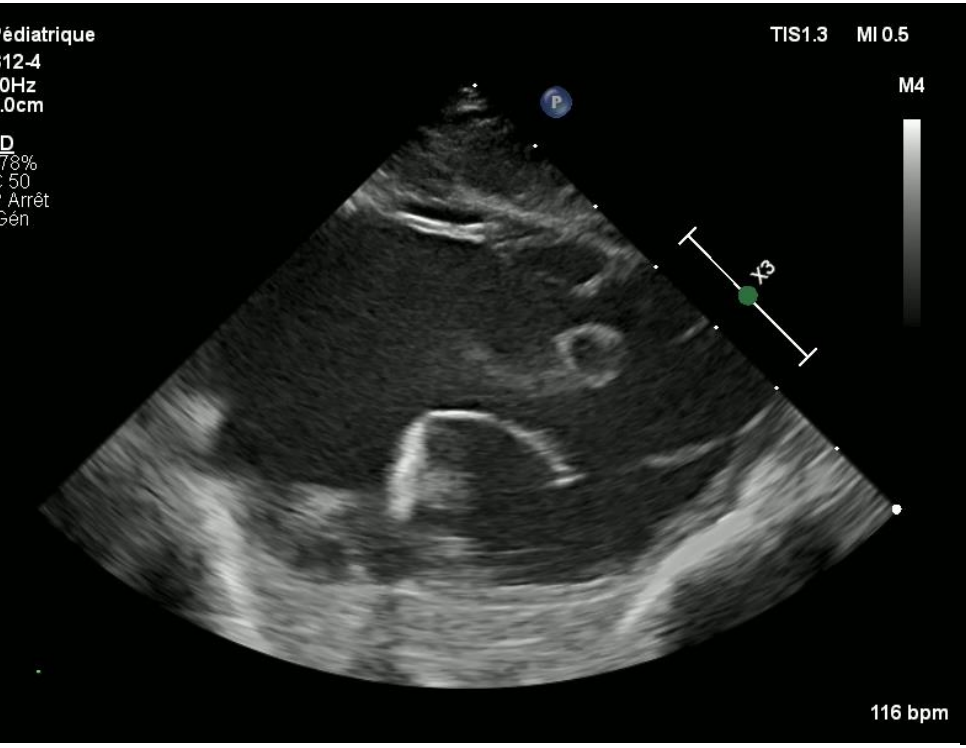

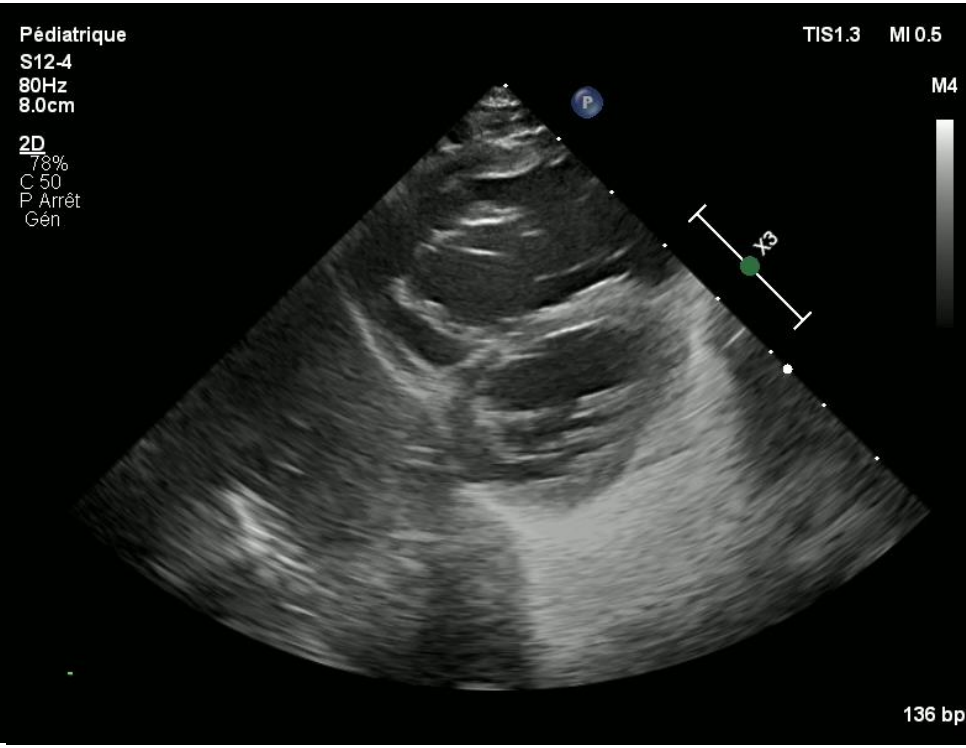

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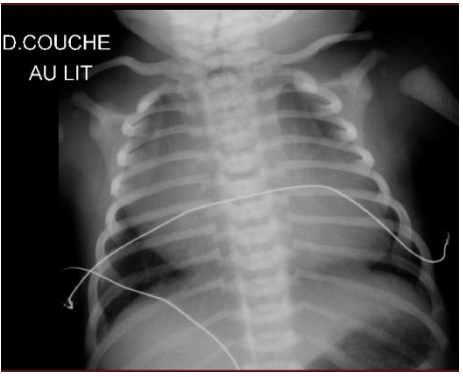
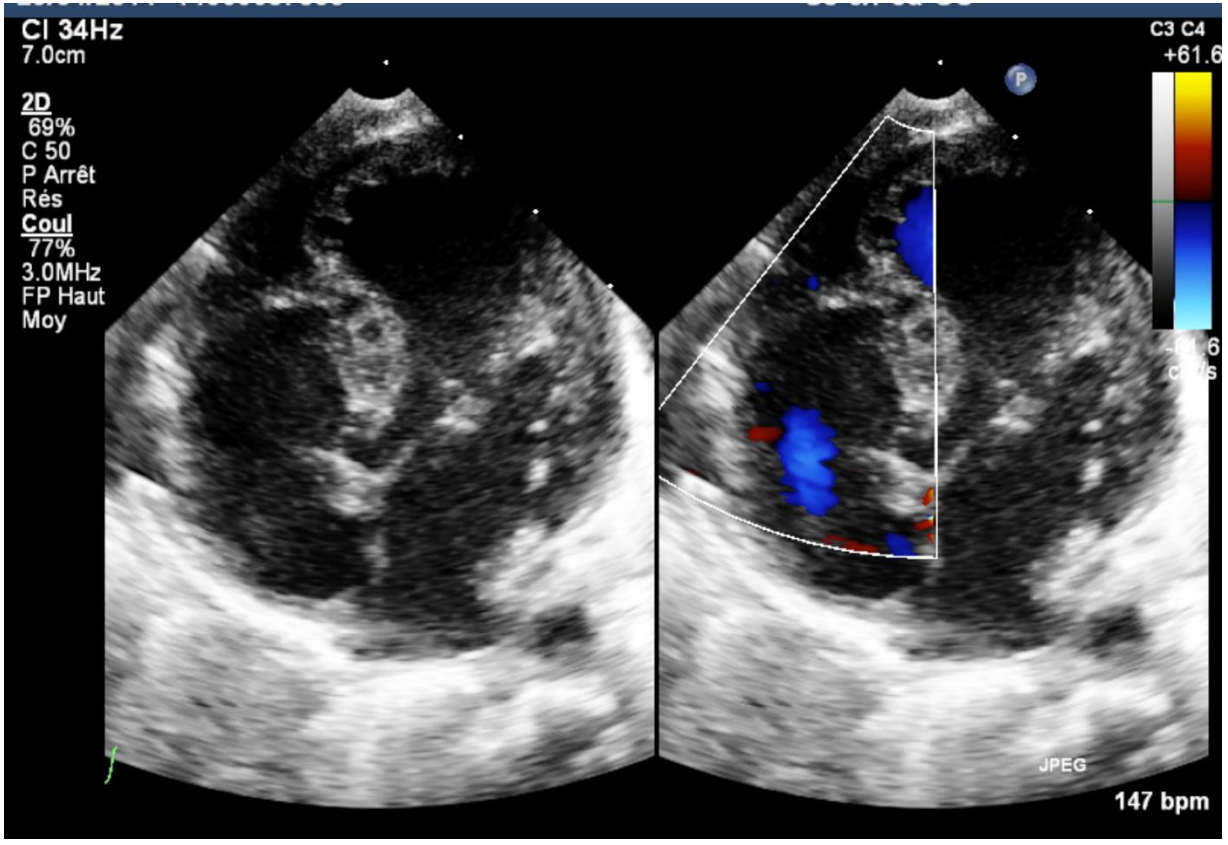
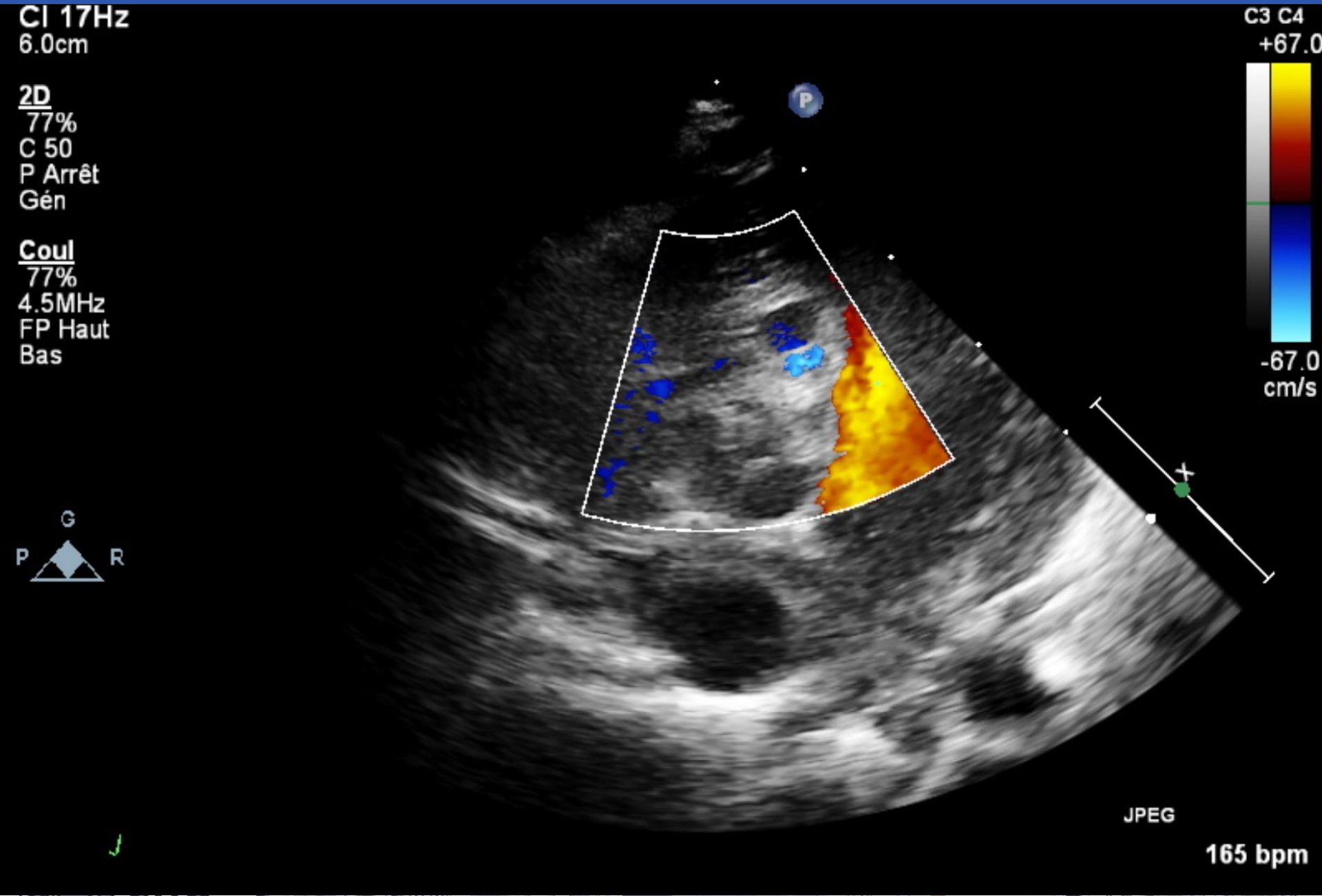
Neo natal assessment

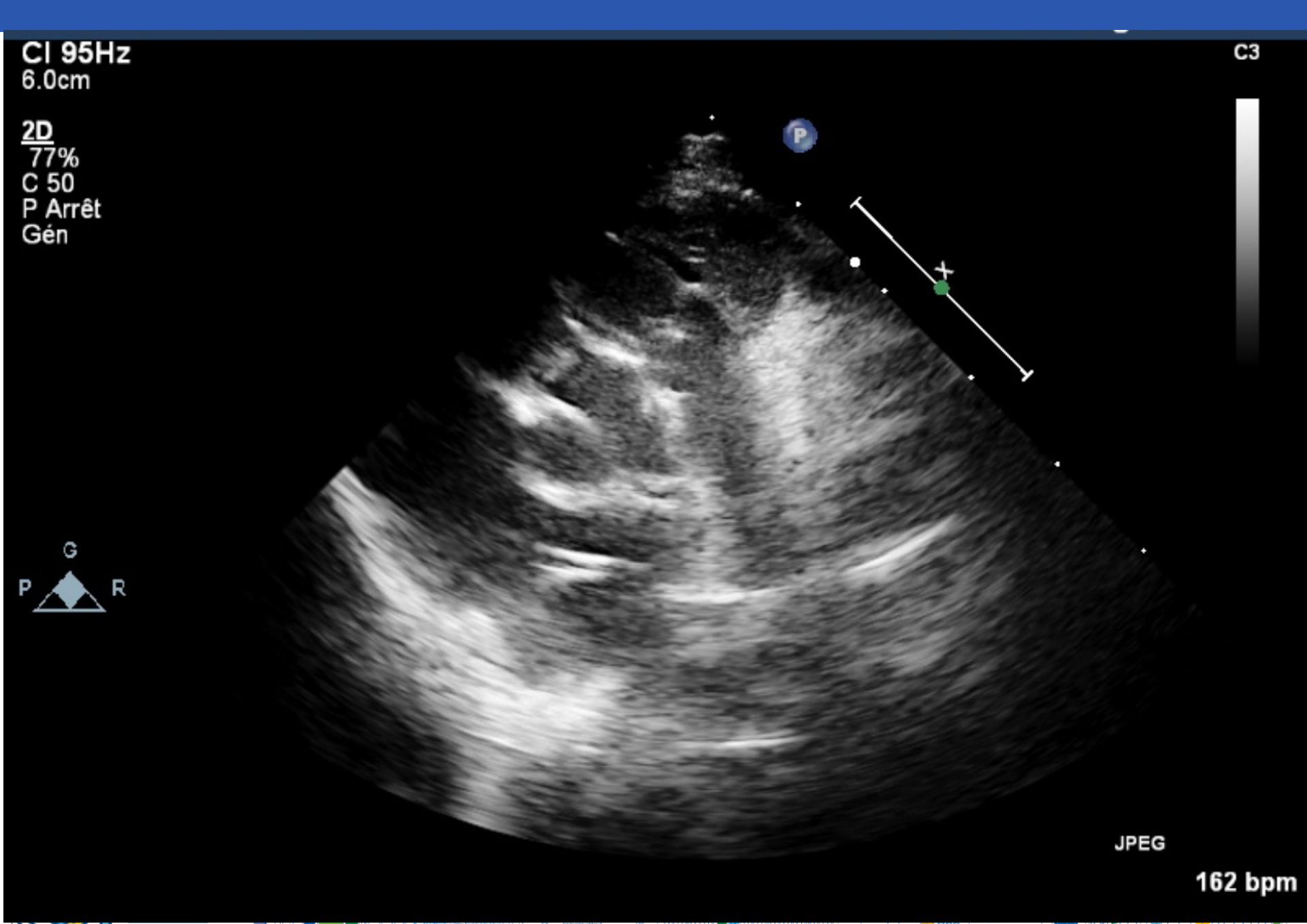
- Ebstein anomaly fonctionnal atresia





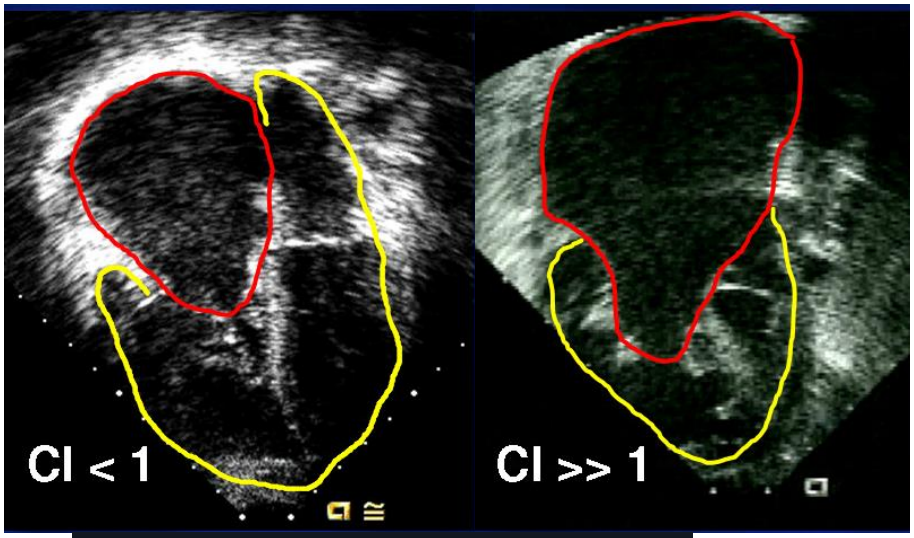
Pas d'IT
Pseudo AT + AP


CI 17Hz
6.0cm
2D
77%
C 50
P Arrêt
Gén
Coul
77%
4.5MHz
FP Haut
Bas
C3 C4
+67.0
-67.0
cm/s
G
P
R
JPEG
165 bpm


CI 95Hz
6.0cm
2D
77%
C 50
P Arrêt
Gén
C3
G
P
R
JPEG
162 bpm

Celermajer index

- Score pronostic pour les nouveau-nés



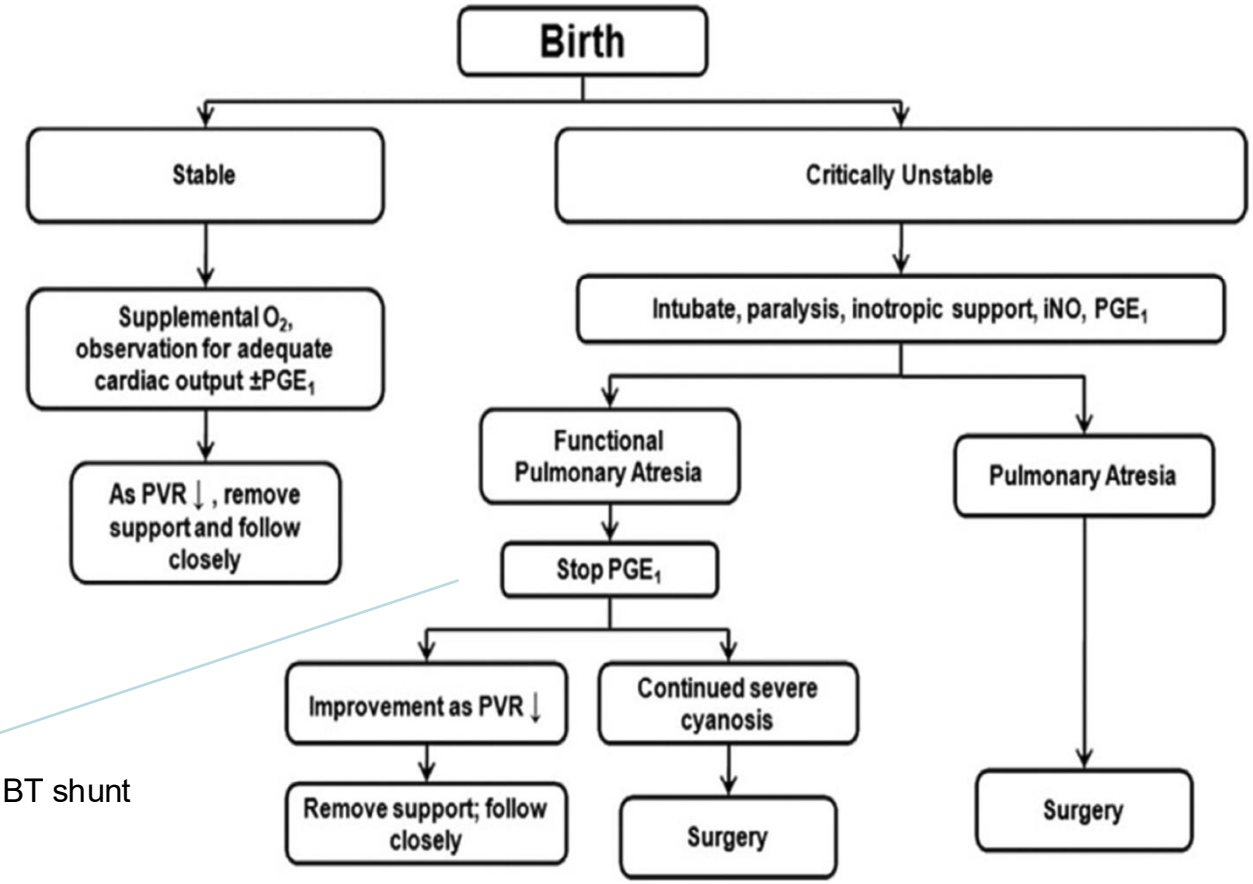
RA area + aRV area

fRV area+ LA area + LV area

Grade	ratio
1	< 0.5
2	0.5 ~ 0.99
3	1 ~ 1.49
4	> 1.5

GOSE score	ratio	Mortality (%)
1-2	< 1.0	8
3 (acyanotic)	1.1 ~ 1.4	10 (early) 45 (late)
3 (cyanotic)	1.1 ~ 1.4	100
4	> 1.5	100

Management néonatal



Don’t rush on the BT shunt

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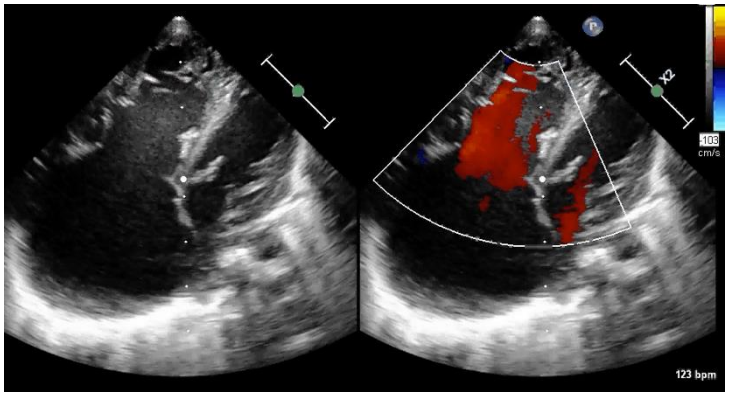
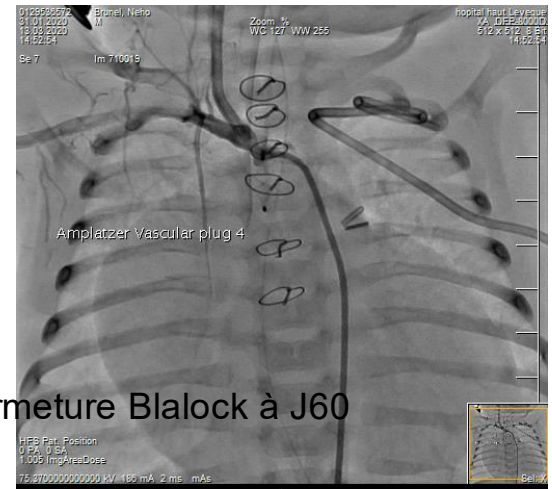
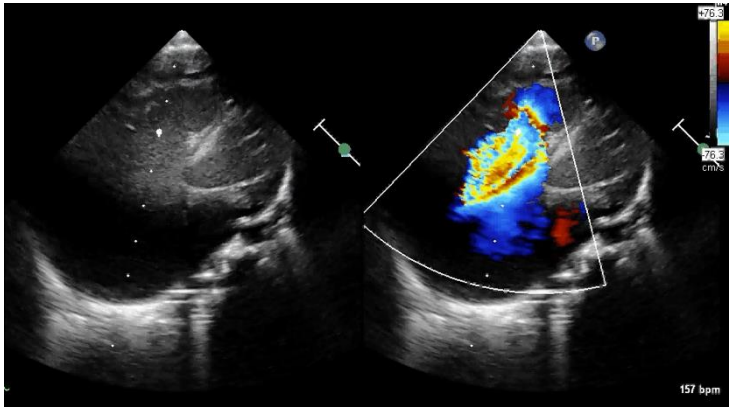
Formes potentiellement évolutives



Ducto dépendance néonatale (Blalock néonatal J12)

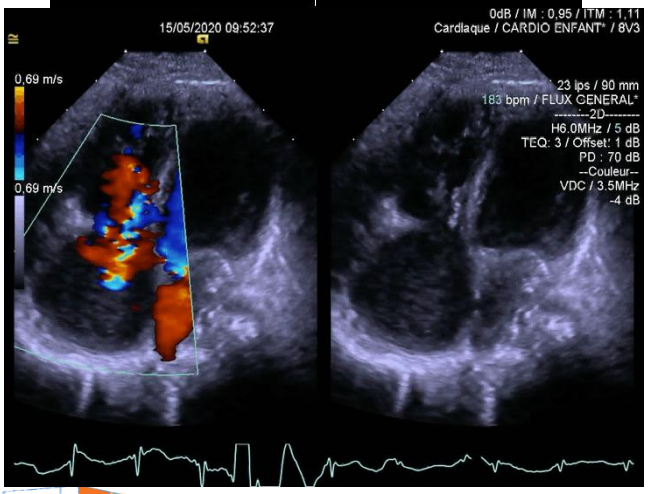


Fermeture Blalock à J60



Défaillance cardiaque PO

ETT M6



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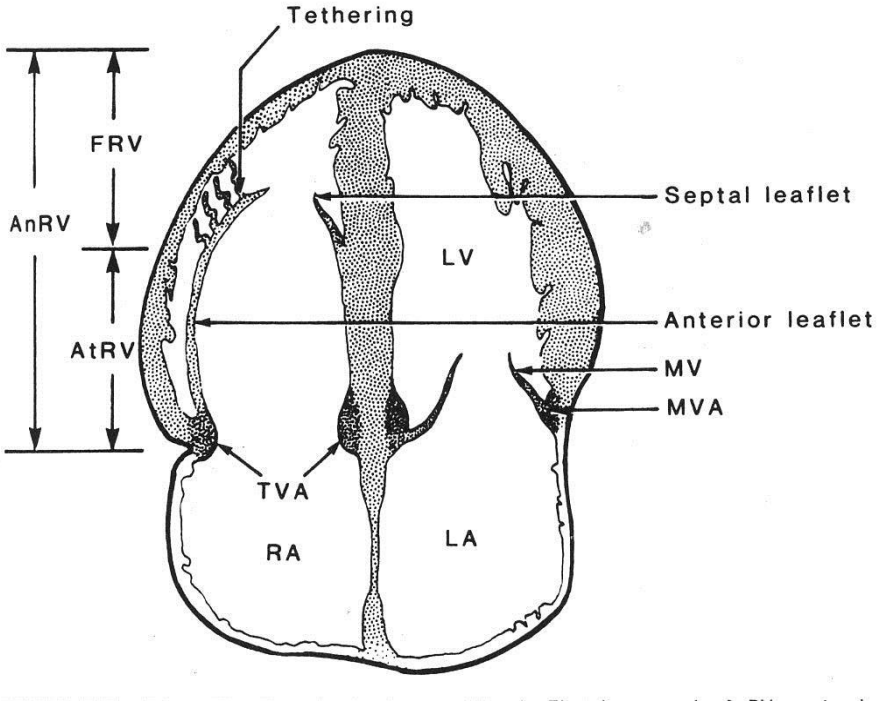
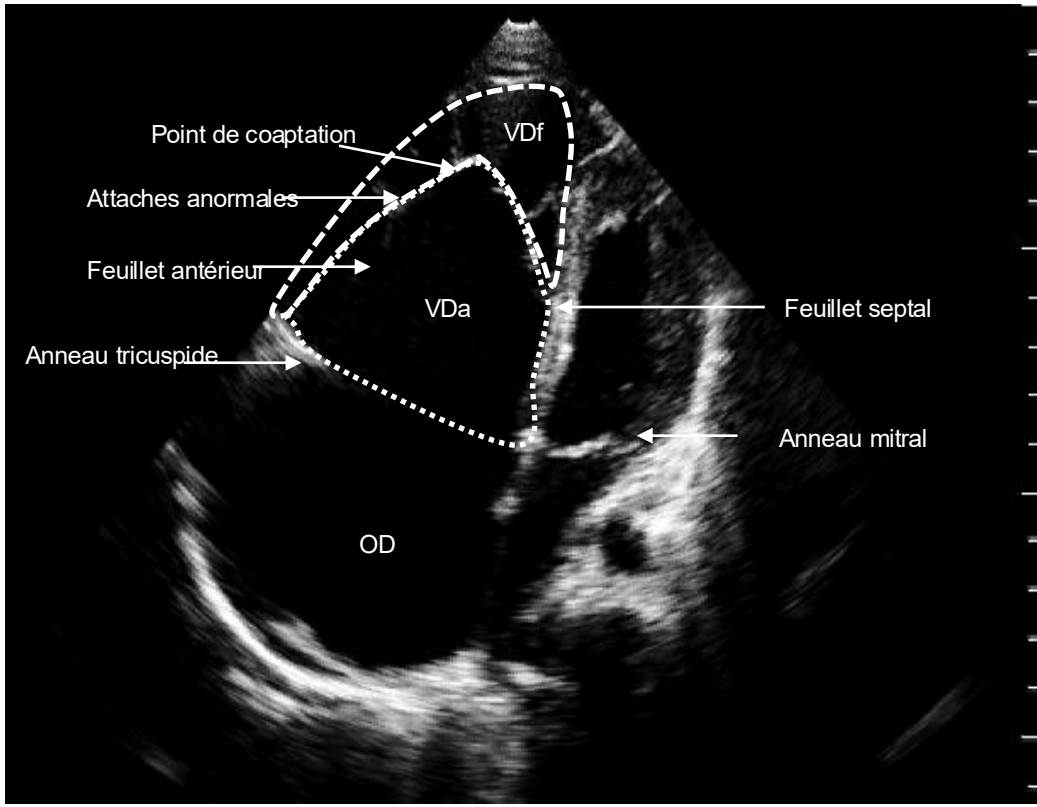
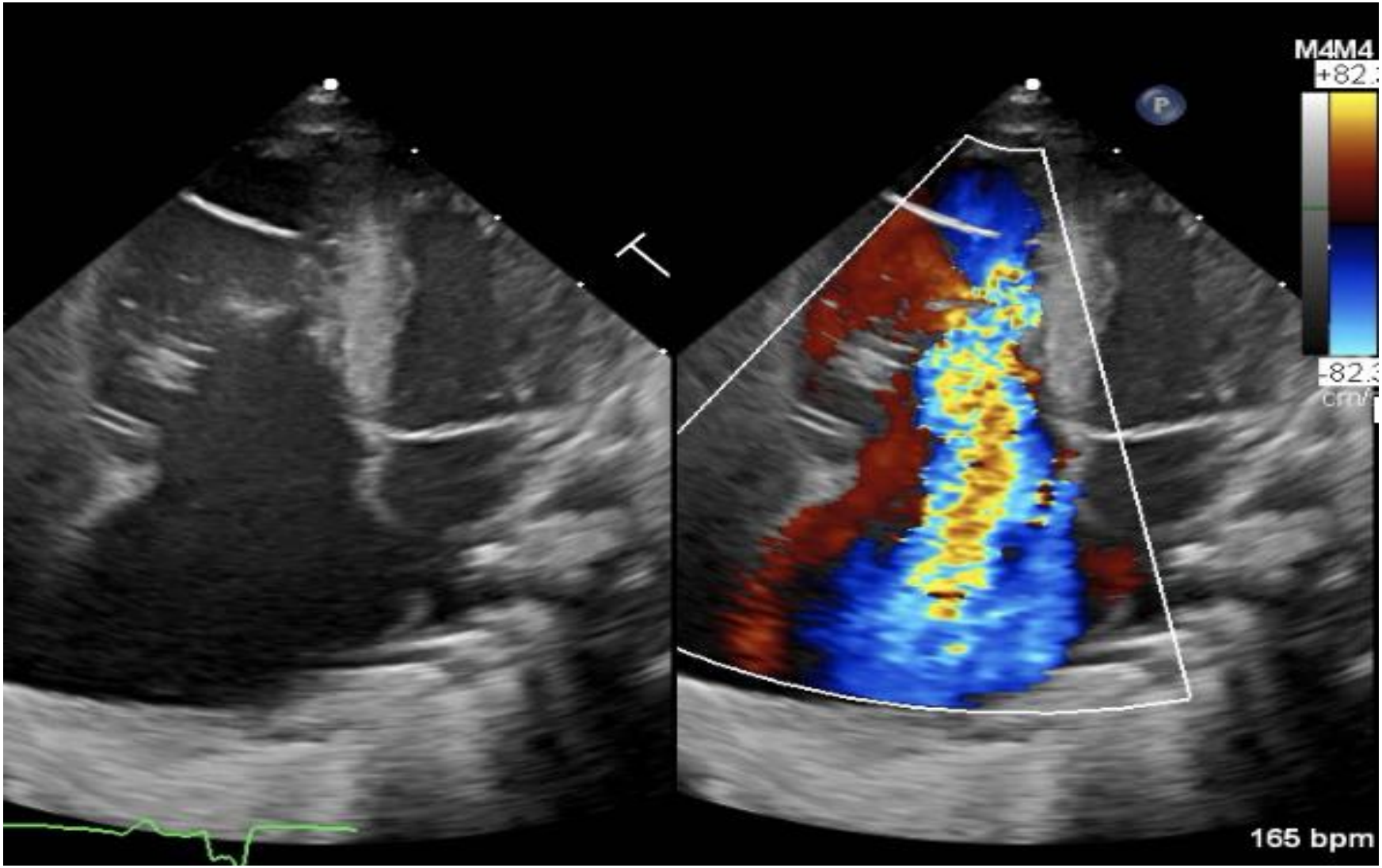

Tethering
FRV
AnRV
AtRV
Septal leaflet
LV
Anterior leaflet
MV
MVA
TVA
RA
LA


Point de coaptation
VDf
Attaches anormales
Feuillet antérieur
VDa
Feuillet septal
Anneau tricuspide
Anneau mitral
OD



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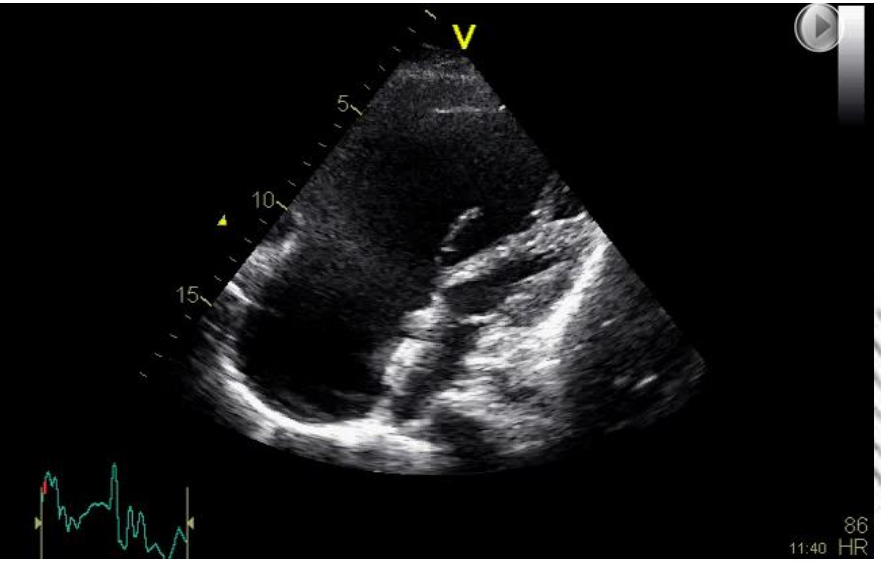

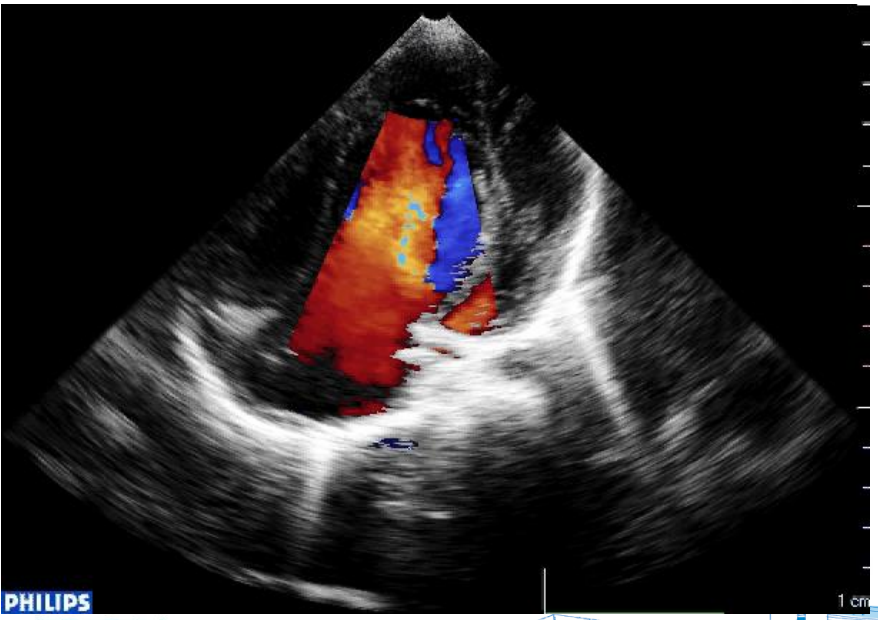

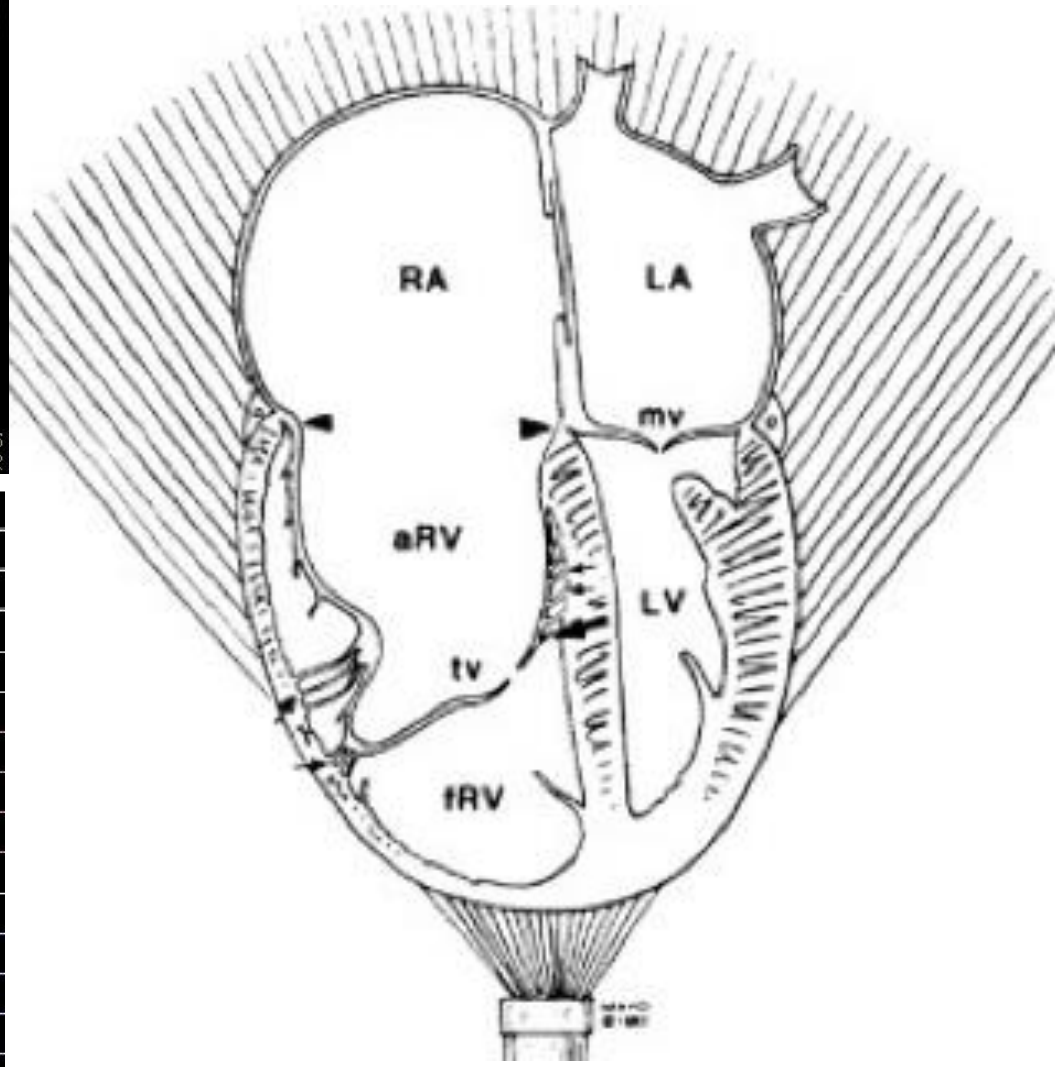

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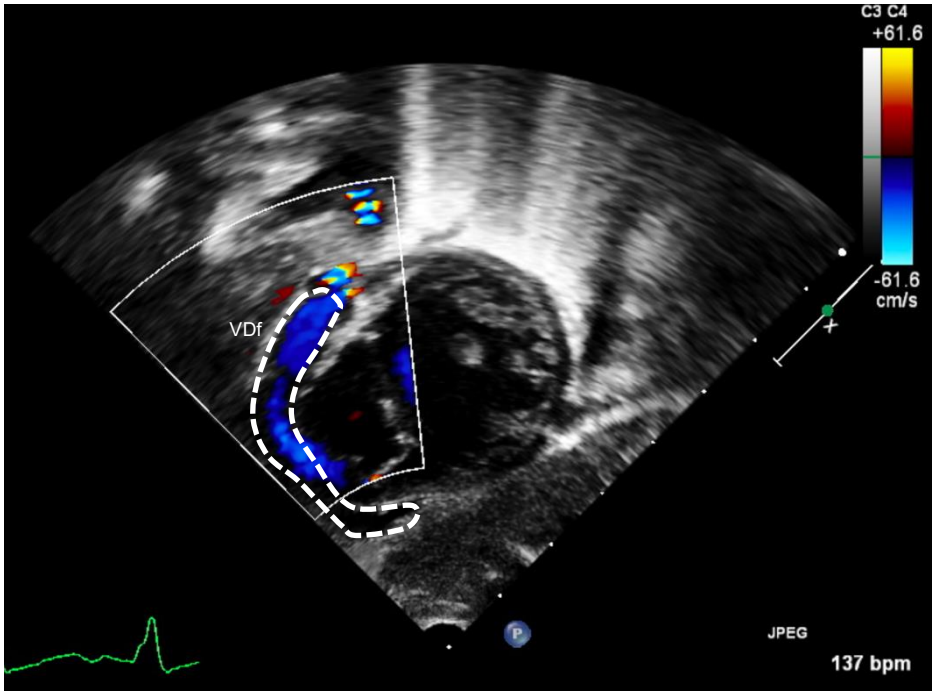

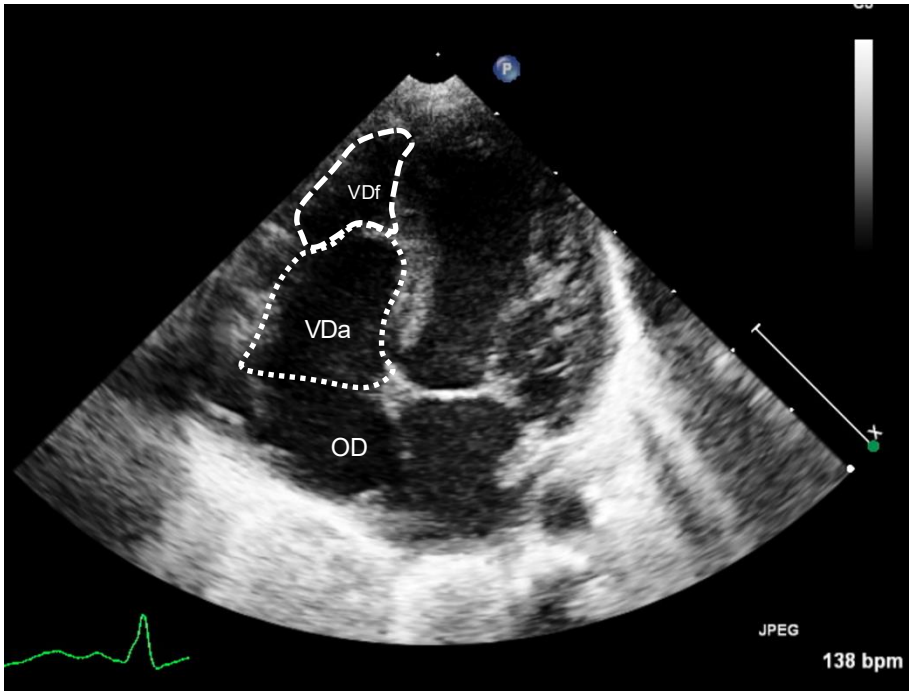

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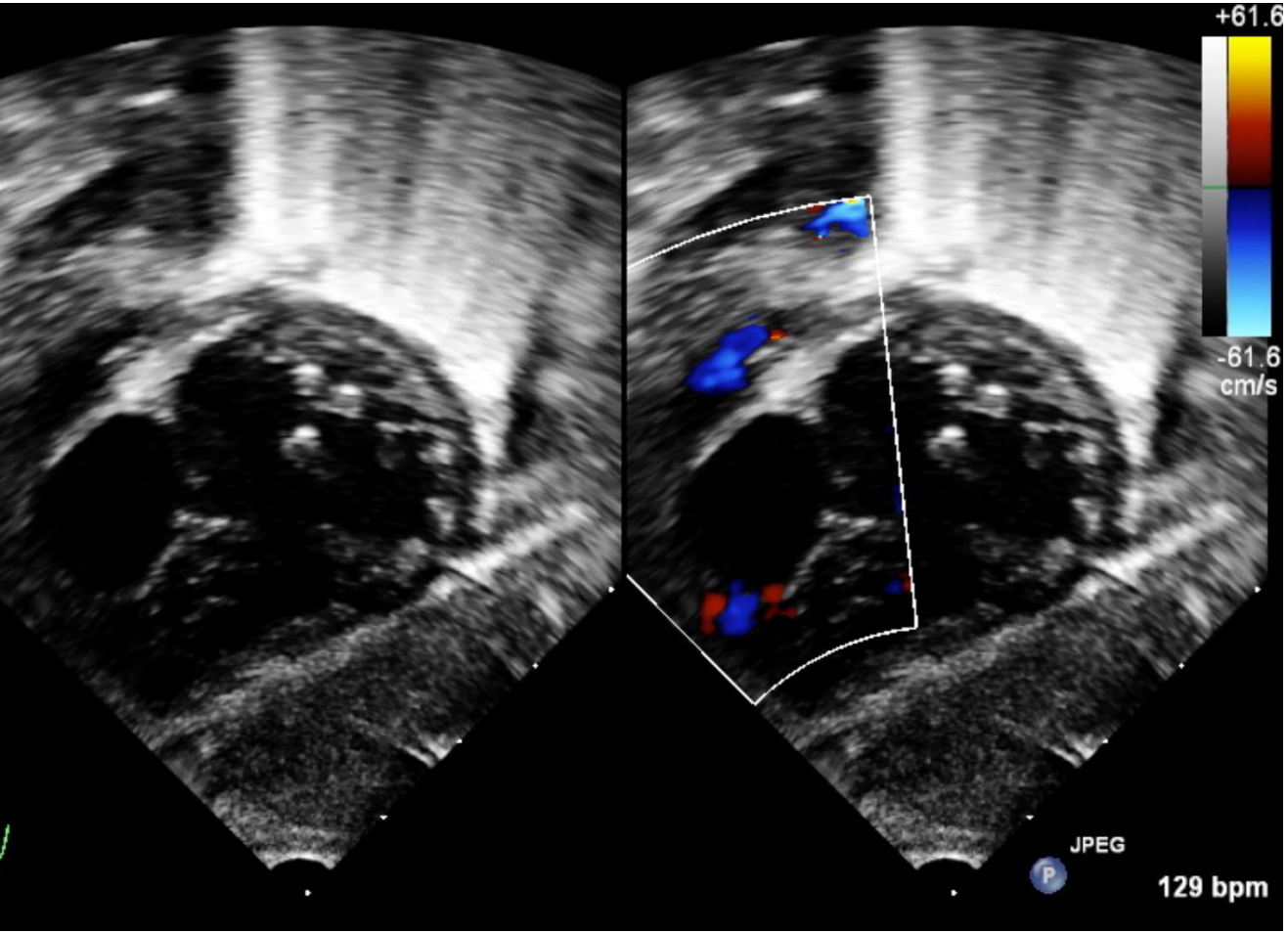

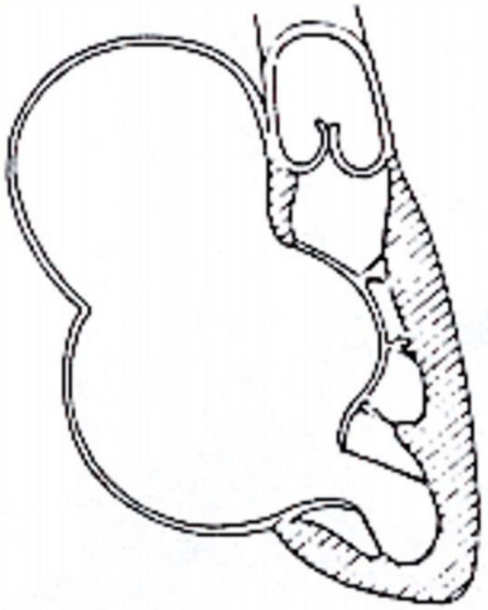

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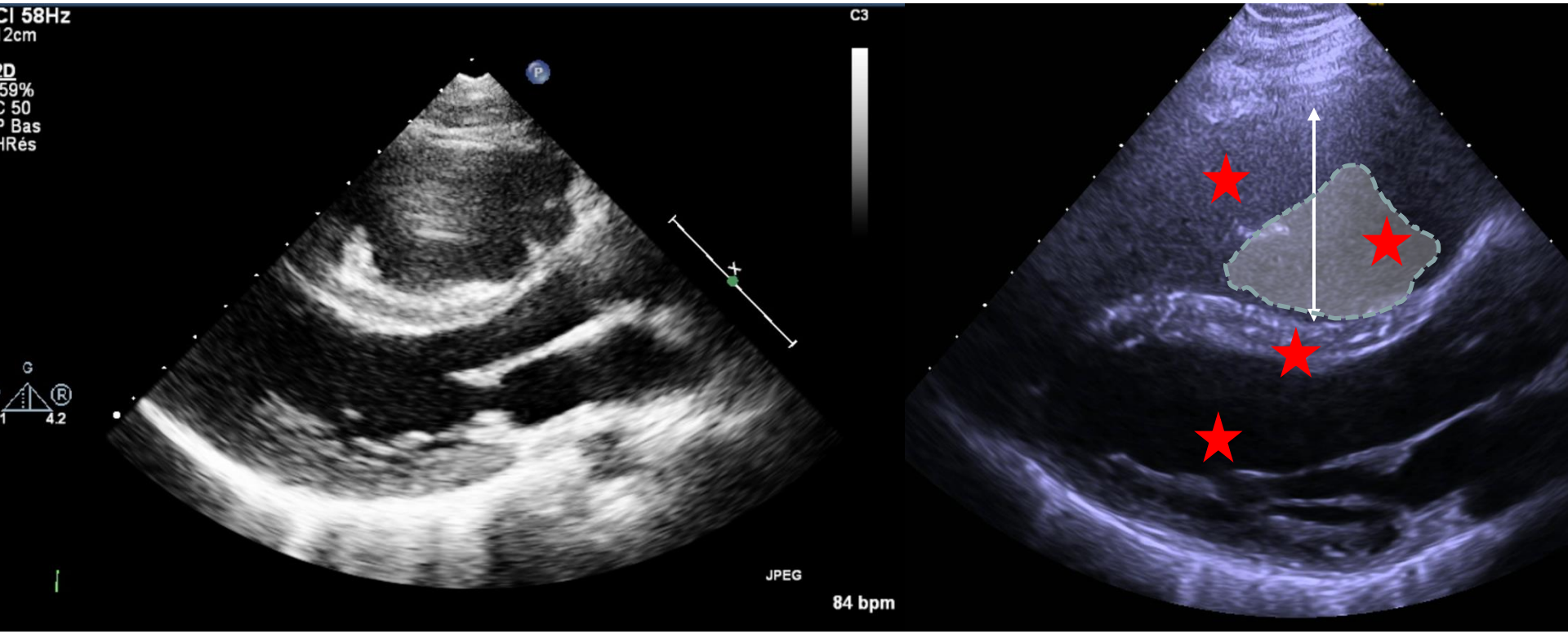
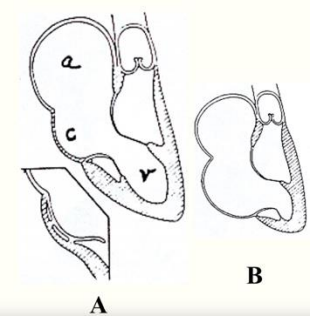
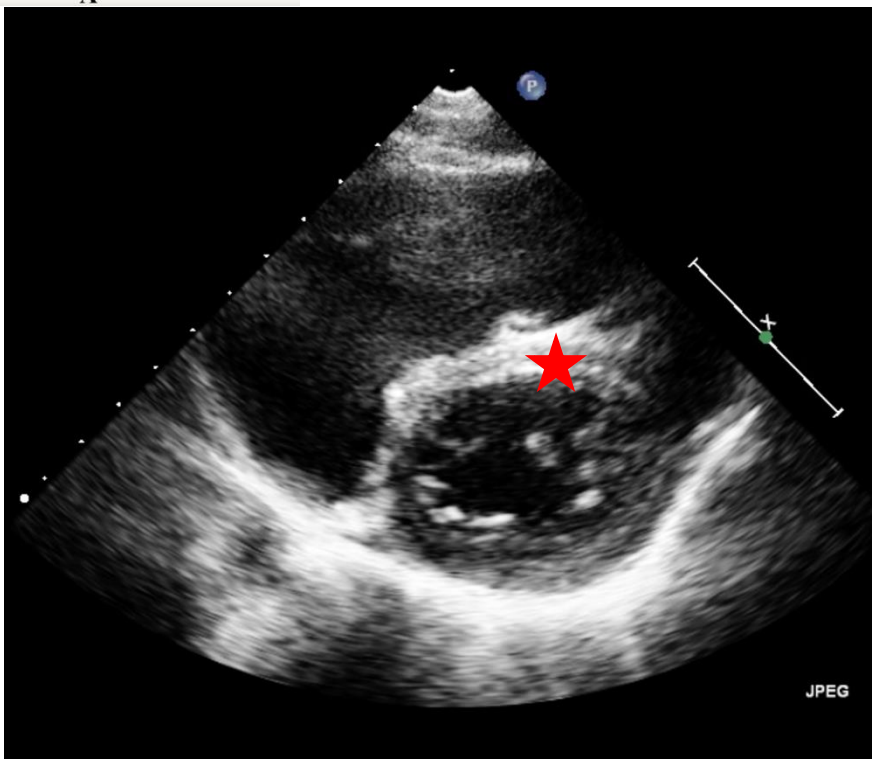
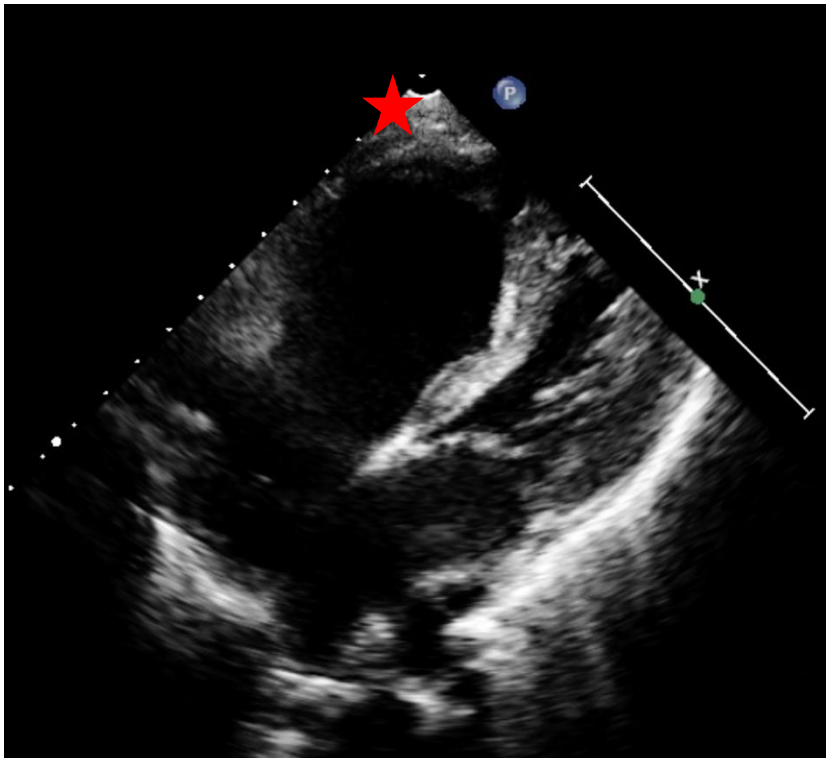
Jeune femme de 19 ans, mort subite récupérée sur super Wolf









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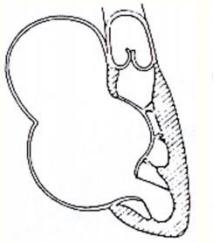

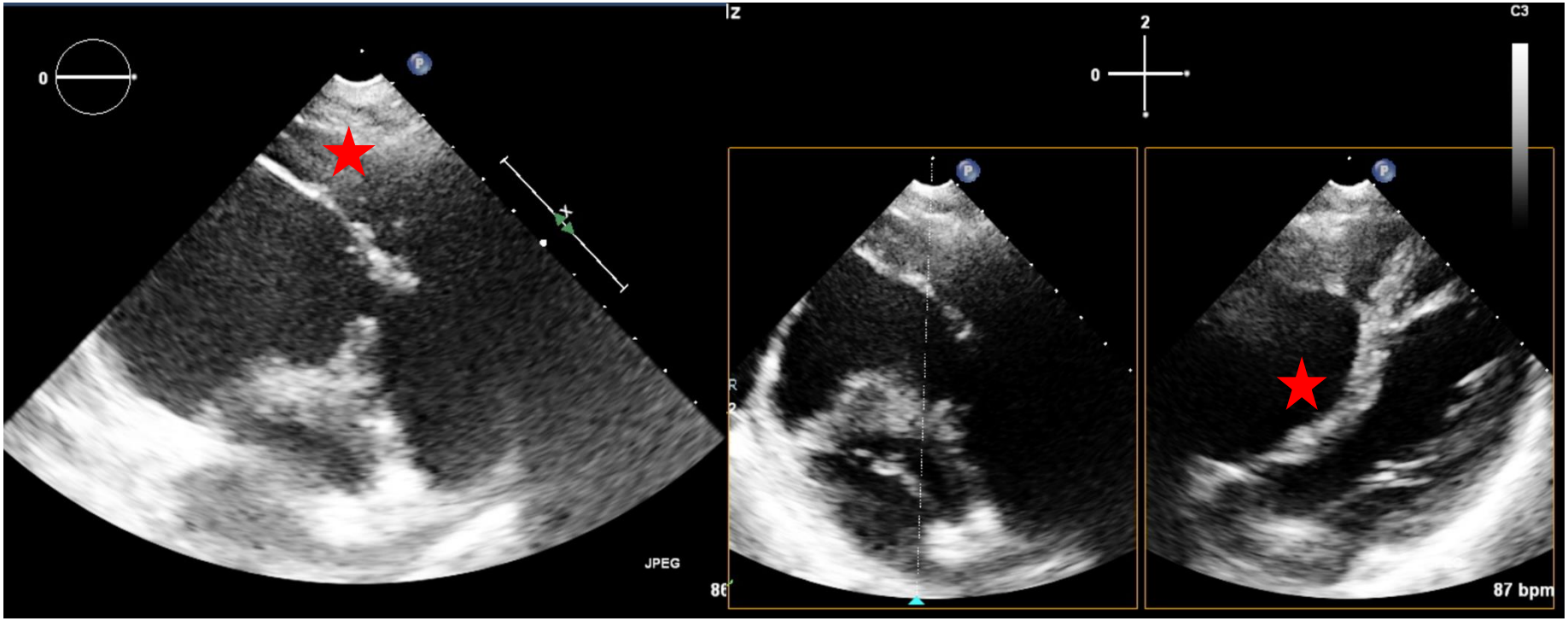

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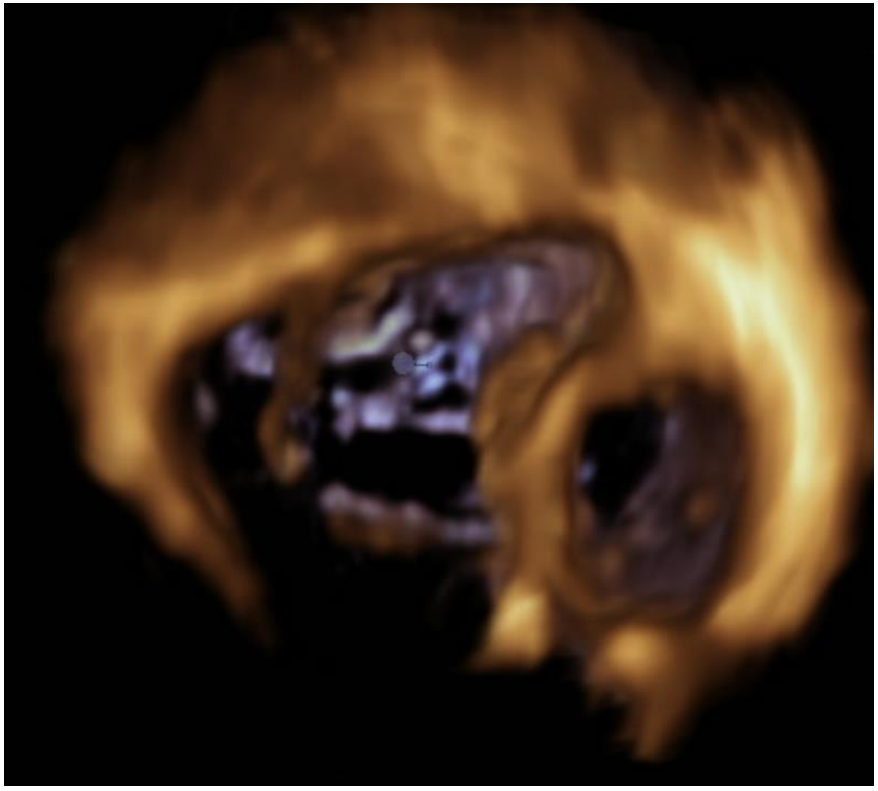

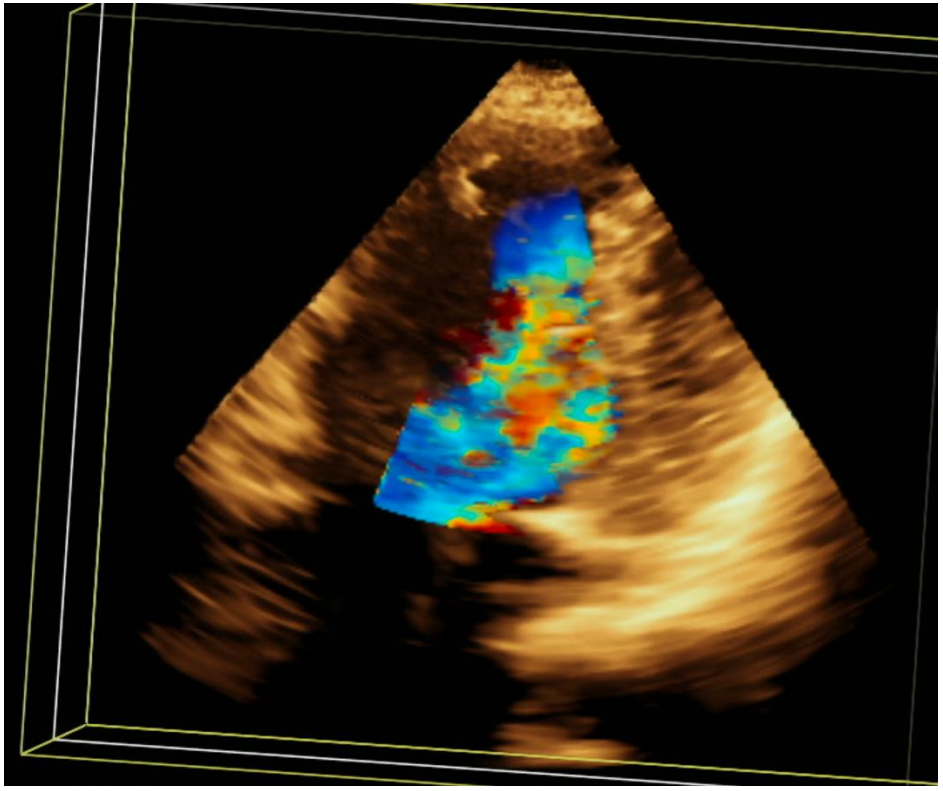

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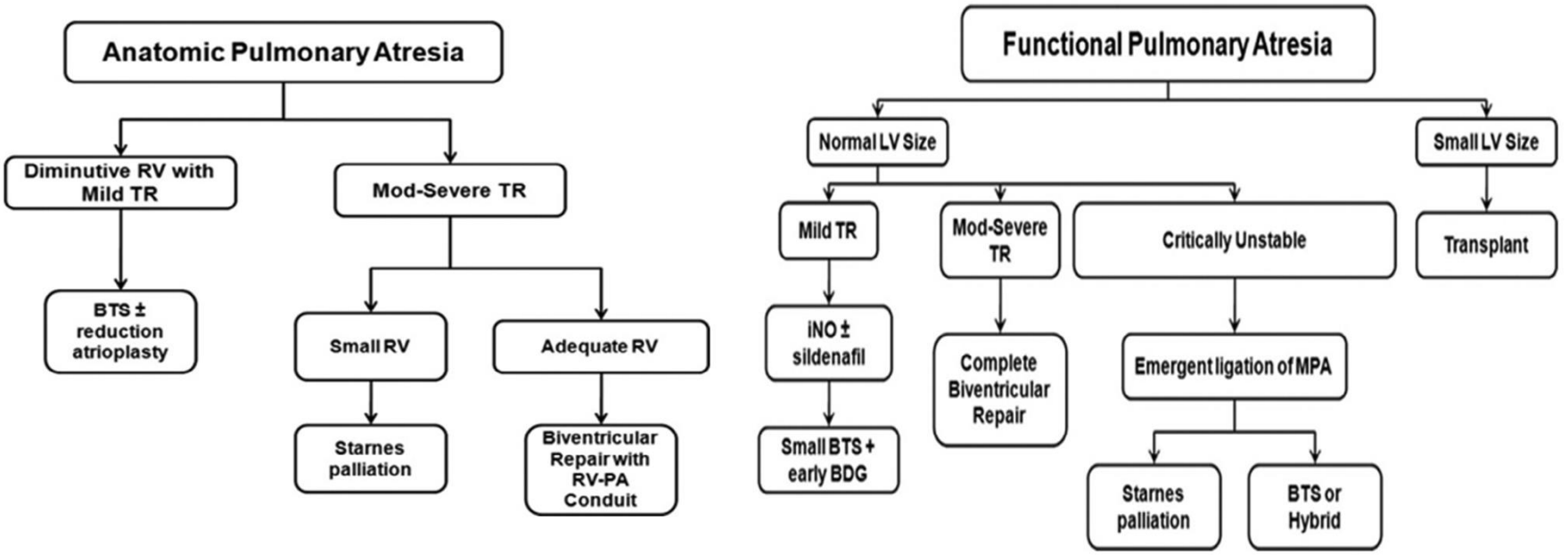
Management néonatal



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