

ANOMALIES CONGENITALES DE LA VALVE MITRALE

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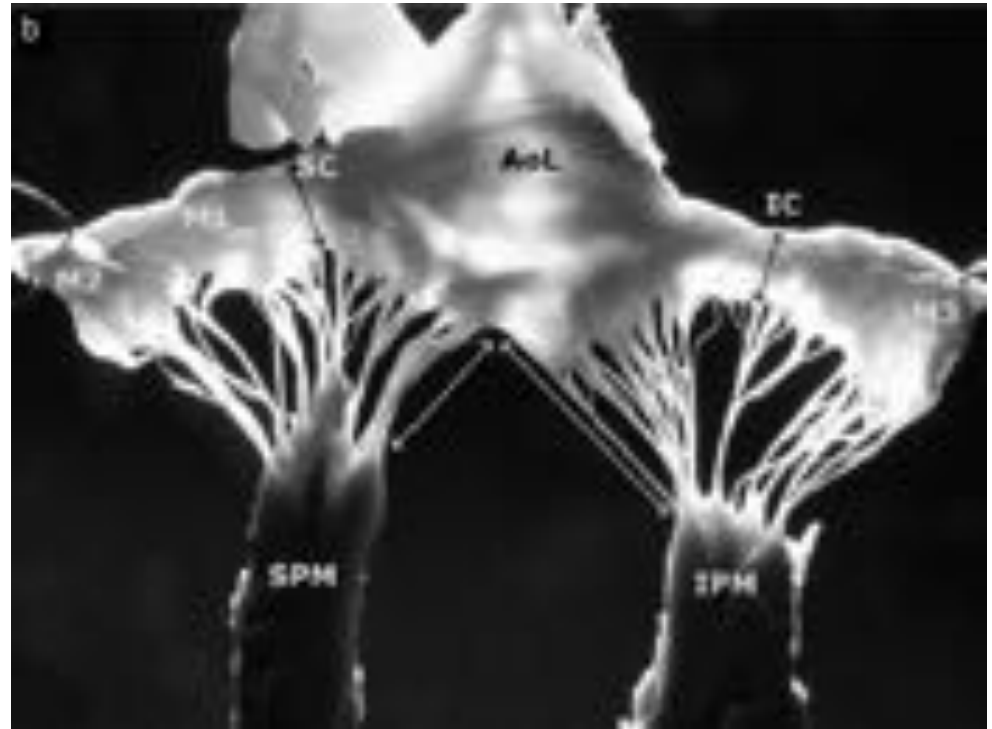
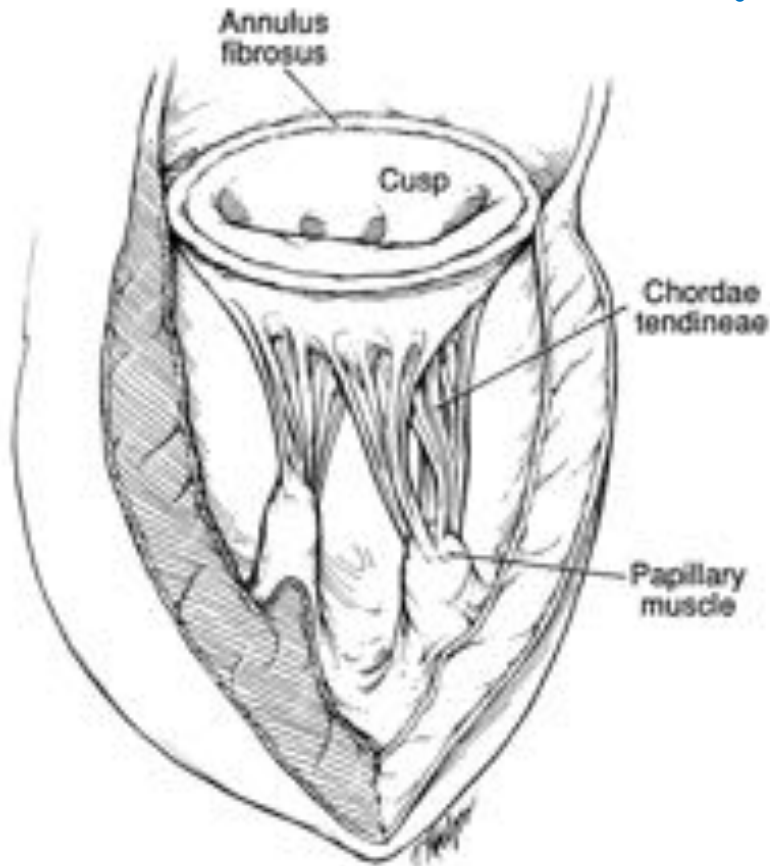
DIU Réanimation des Cardiopathies Congénitales

Mars 2016

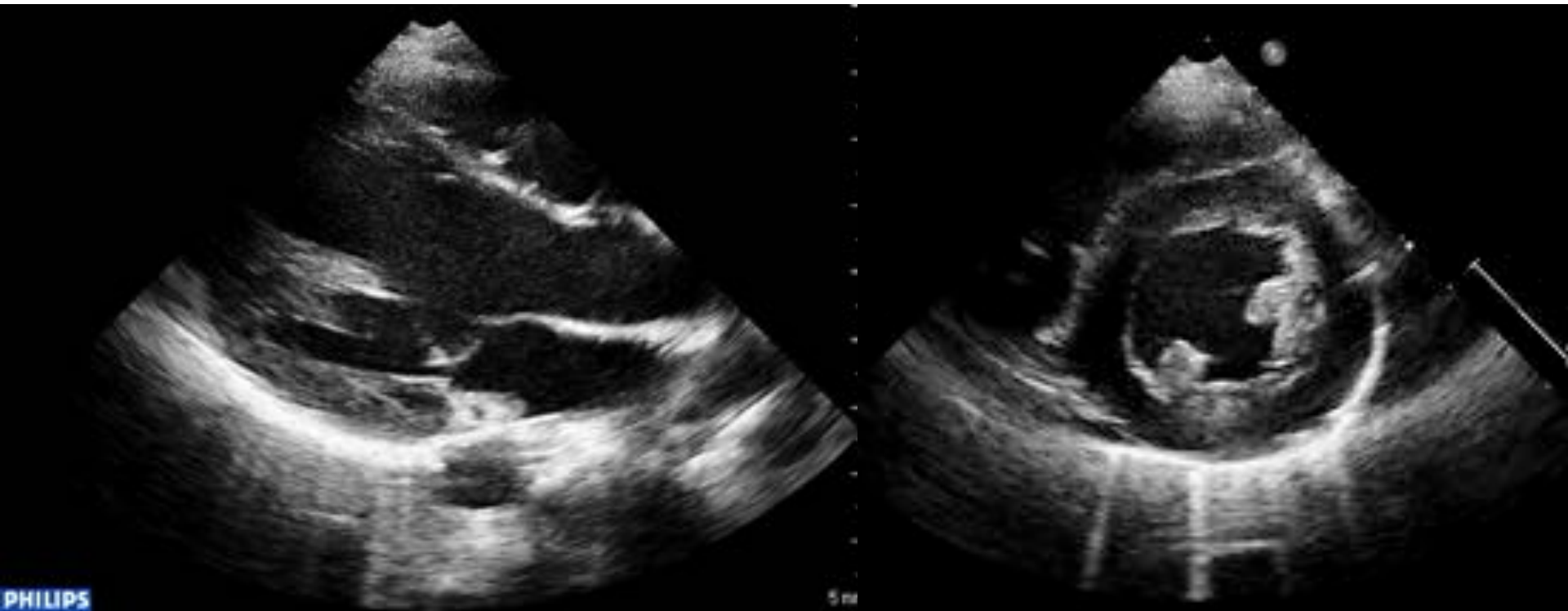
ANATOMIE NORMALE

Complexe valvulaire mitral

- Anneau
- Feuillet
- Cordages
- Muscles papillaires



MITRALE NORMALE



ANOMALIES CONGENITALES

- Prévalence: 0.5%

Banerjee et al. Am J Cardiol 1995.

- Rarement isolées

- Shone: 35%

- Anomalies du cœur gauche: 76%

- IM:72%, Sténose:13%, IM + Sténose: 15%

Fuller et al. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2009.

EPIDEMIOLOGIE

- Etude échocardiographique
- Sur 13400 enfants: 65 malformations mitrales
 - Parachute: 0.17%
 - Anneau supra-mitral: 0.15%
 - Cleft: 0.07%
 - DOMV: 0.05%

PHYSIOPATHOLOGIE

- Fuite

Dilatation progressive des cavités gauches et \searrow FEVG, arythmie atriale

→ Progression lente

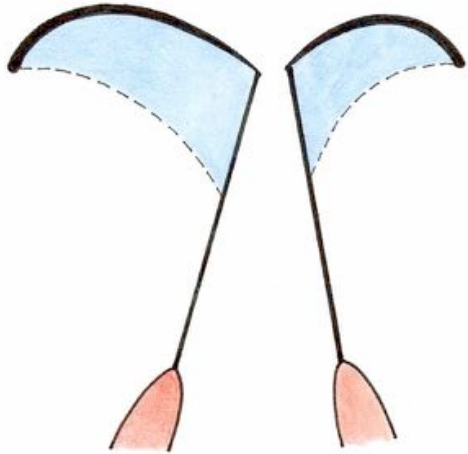
- Obstruction

HTAP post-capillaire + bas débit systémique

→ Formes néonatales sévères = chirurgie

→ Formes modérées = diurétiques le plus longtemps possible

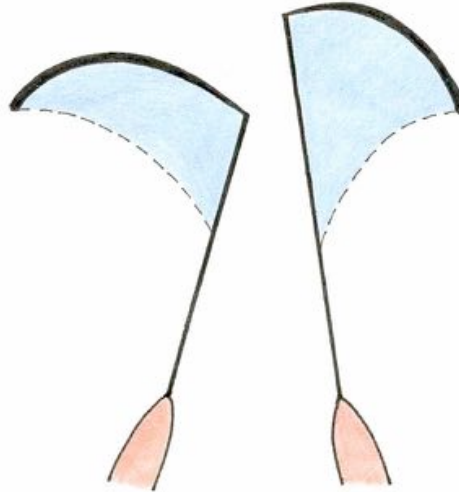
CLASSIFICATION FONCTIONNELLE (CARPENTIER)



1. Mobilité normale

- Dilatation de l'anneau
- Cleft
- Double orifice

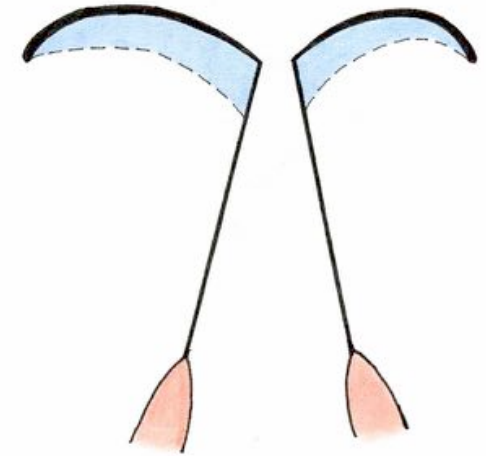
⇒ **IM**



2. Mobilité exagérée

- Prolapsus

⇒ **IM**



3. Mobilité restreinte

Muscles papillaires normaux

- Ring

- Ebstein

Muscles papillaires anormaux

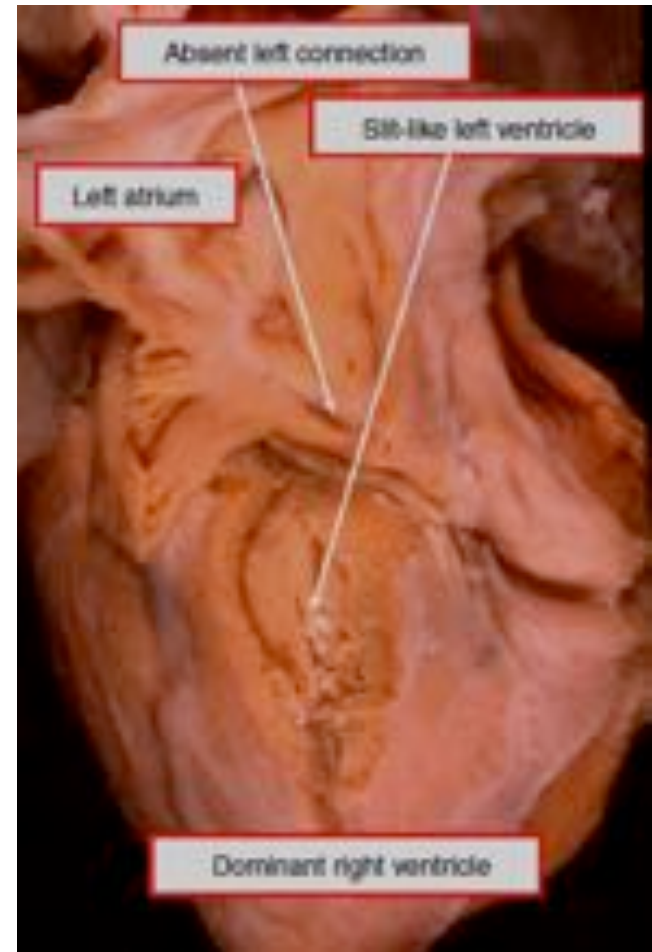
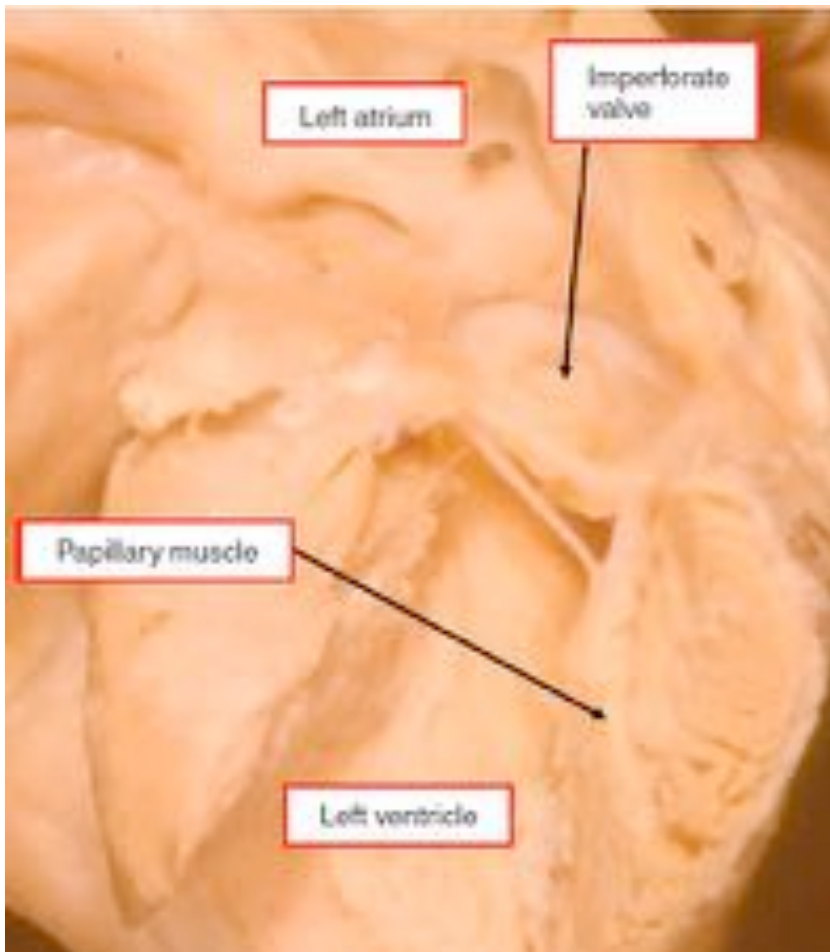
- Parachute

- Hamac

⇒ **IM et/ou Sténose**

ANOMALIES DES FEUILLETS

1) Imperforation de la valve mitrale



ANOMALIES DES FEUILLETS

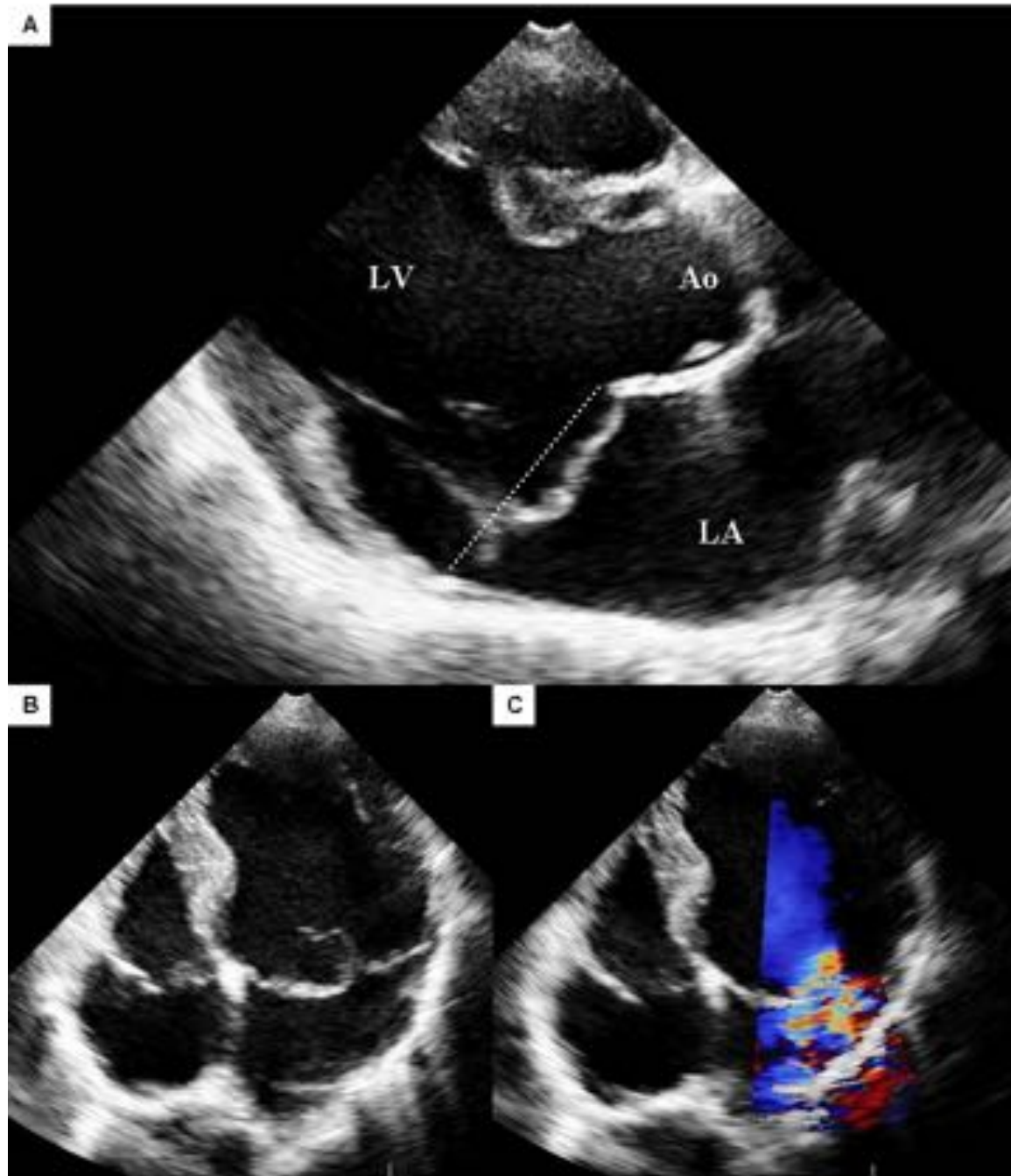
2) Prolapsus valvulaire mitral

- Diagnostic échographique:
 - Coupe parasternale grand axe
 - Déplacement d' un ou des deux feuillets > 2mm
 - ± épaissement des feuillets

Sattur et al. Exp Clin Cardiol 2010.

- 0.7% des adolescents
- Incidence ↗ avec l' âge

2) Prolapsus valvulaire mitral



2) Prolapsus valvulaire mitral

Etiologies

Atteintes du collagène et du tissu élastique

- Marfan +++

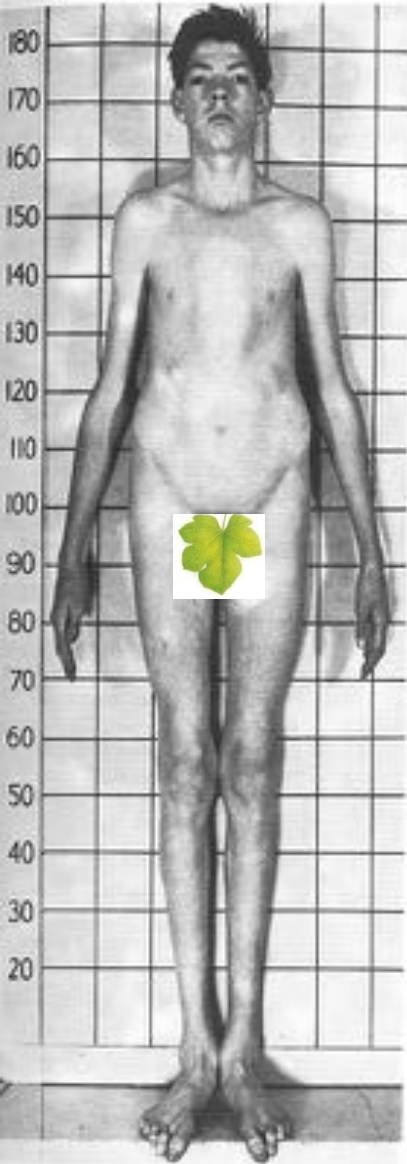
→ mutations gènes codant fibrillin-1

- Ehler-Danlos

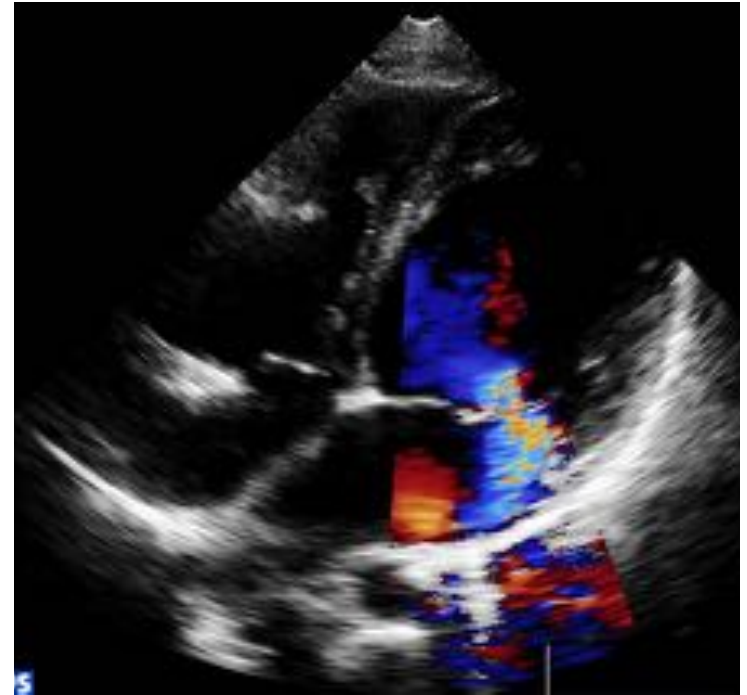
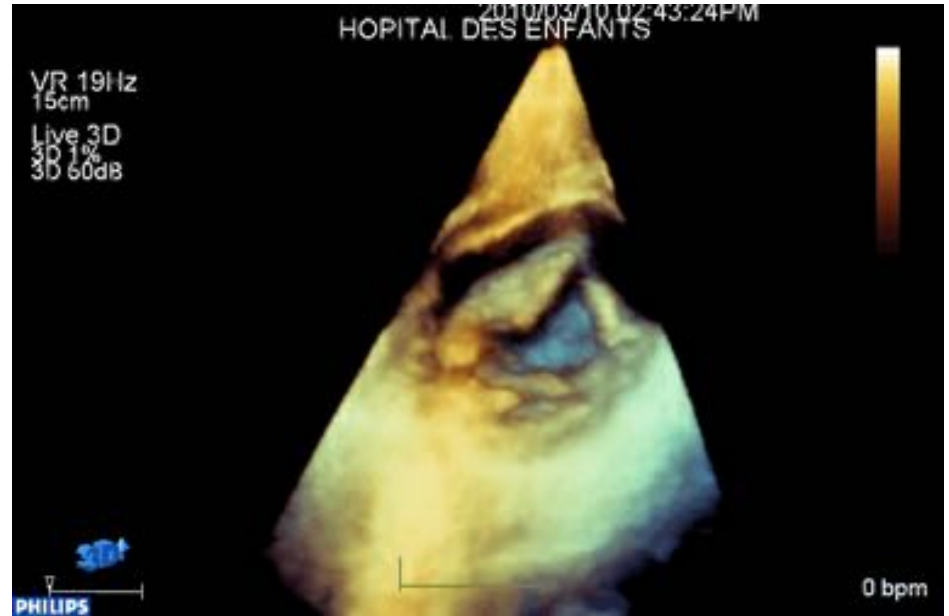
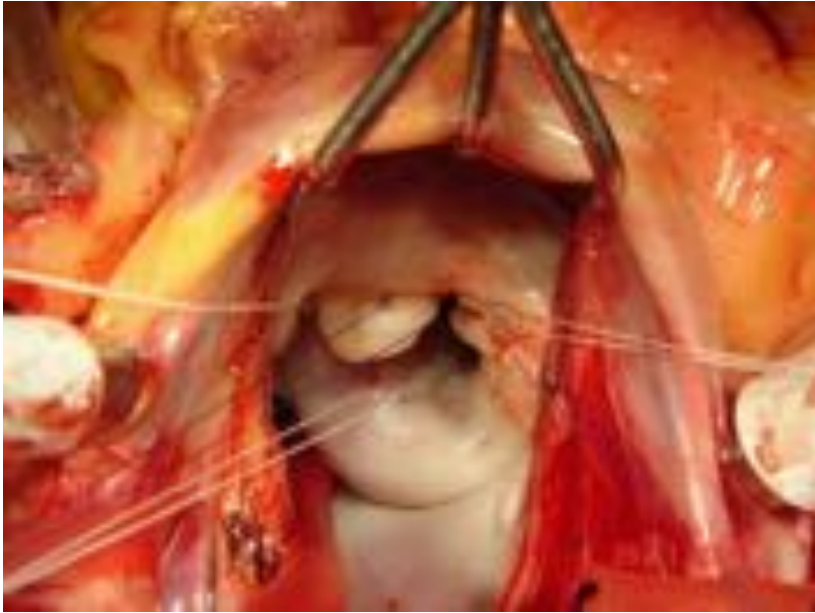
- Ostéogénèse imparfaite

- Cutis laxa

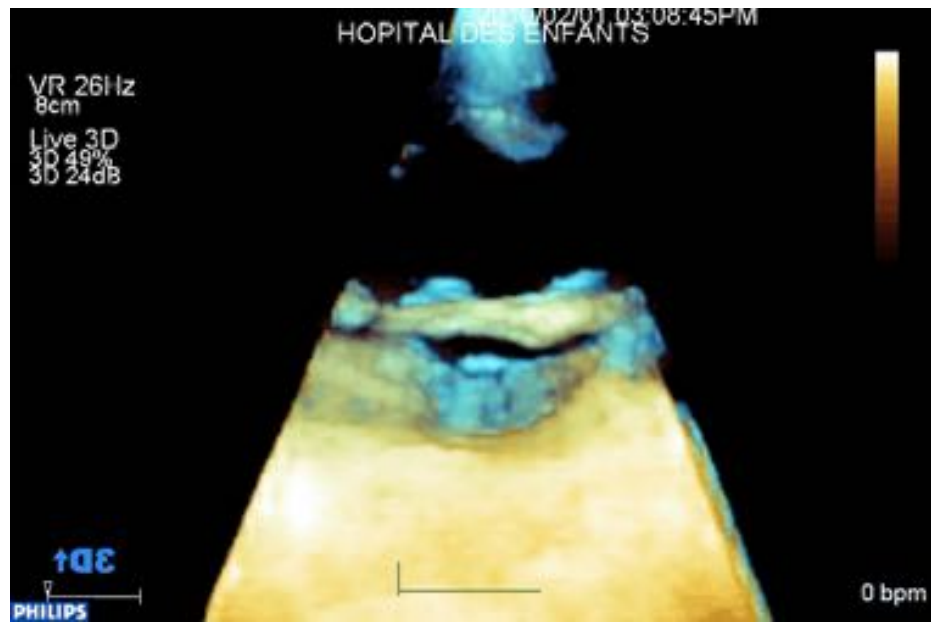
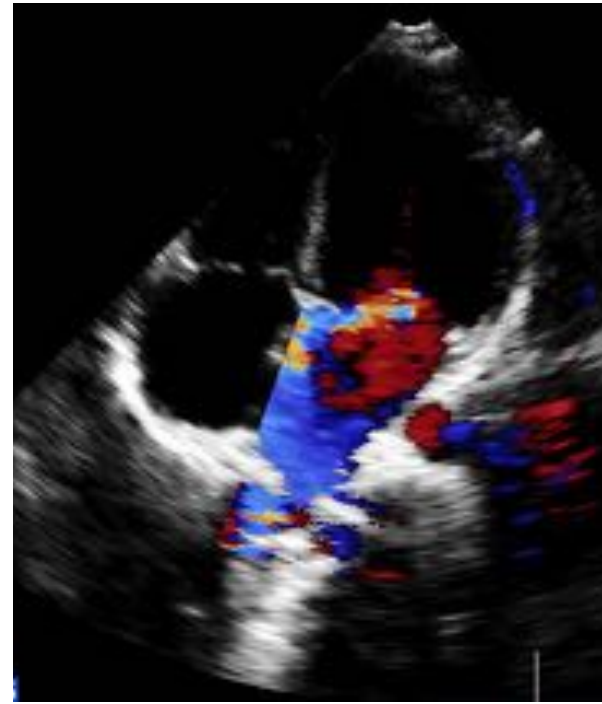
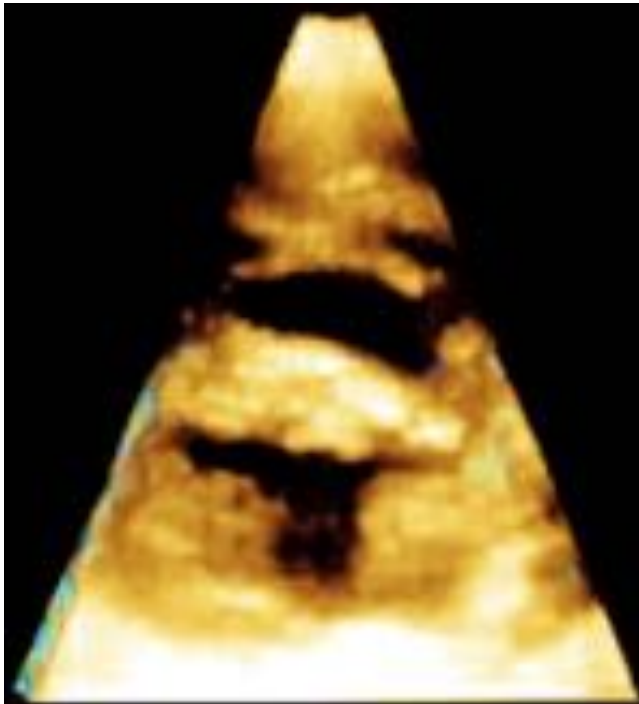
- Pseudoxanthoma elasticum



3) Fente mitrale antérieure isolée

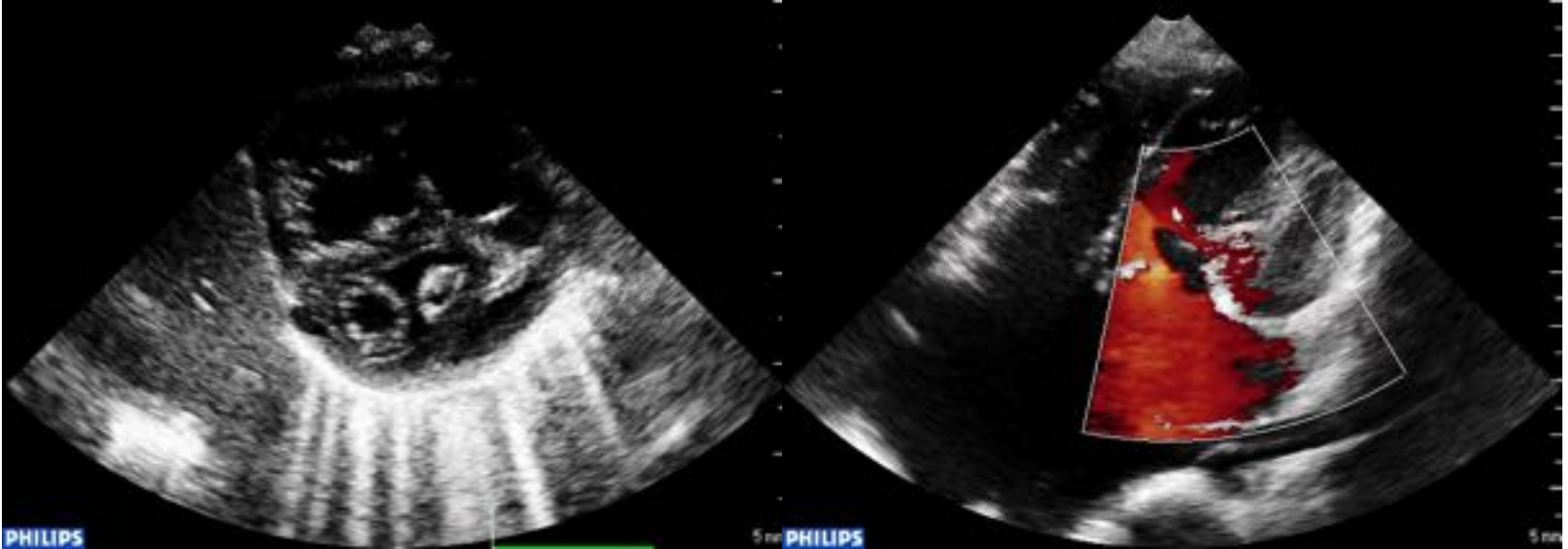
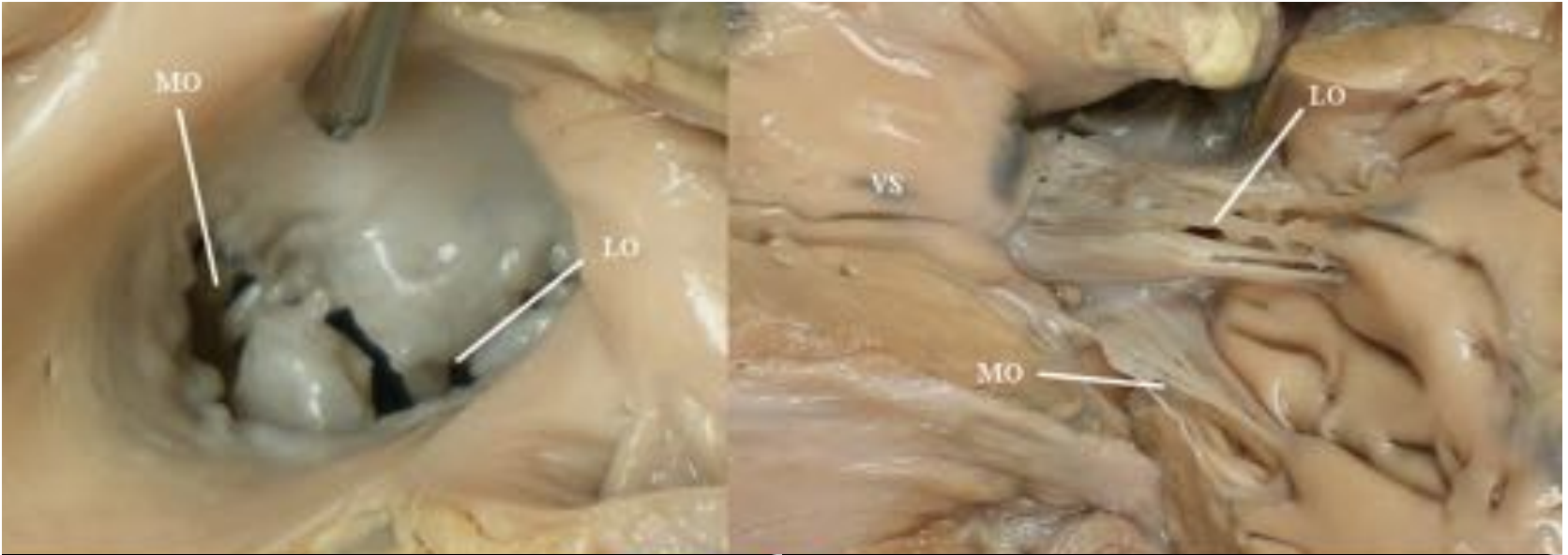


3) Fente mitrale postérieure isolée



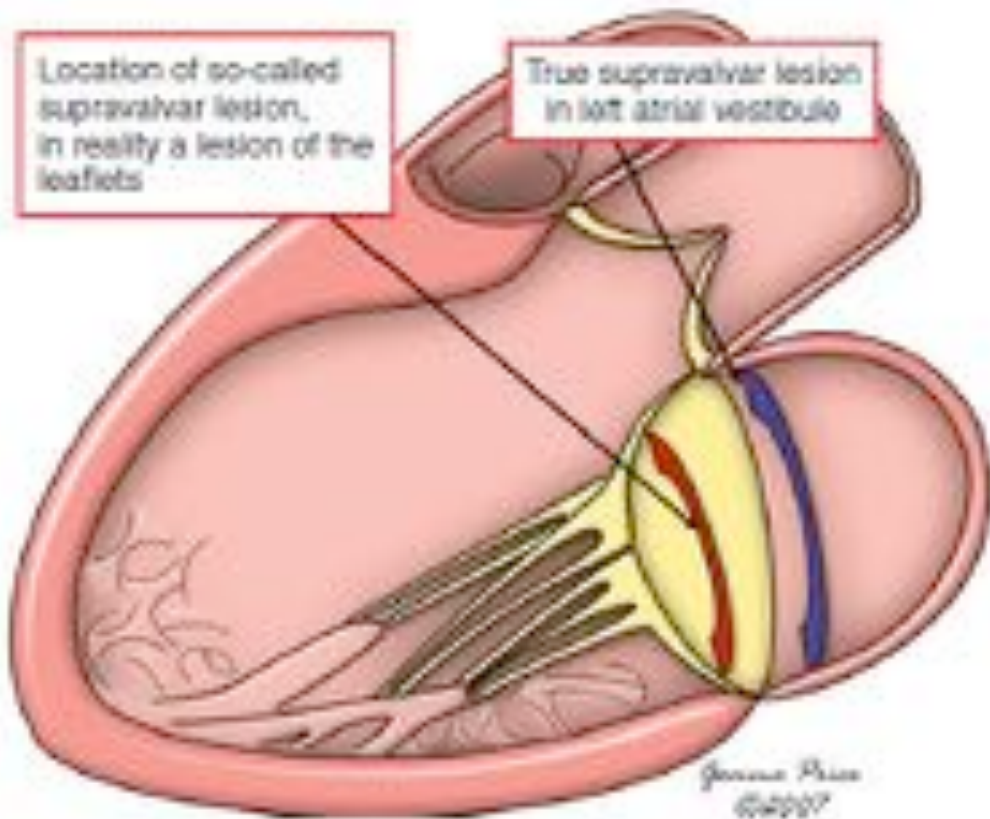
ANOMALIES DES FEUILLETS

4) Double orifice mitral



ANOMALIES DES FEUILLETS

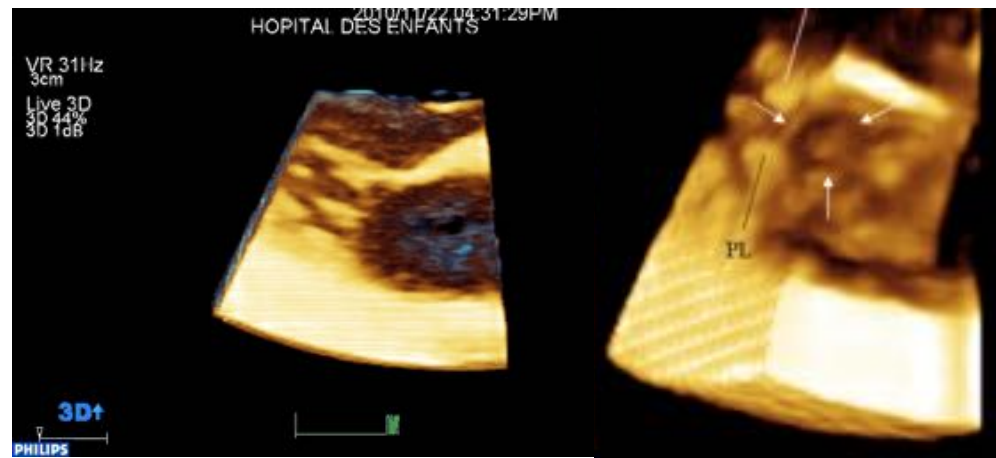
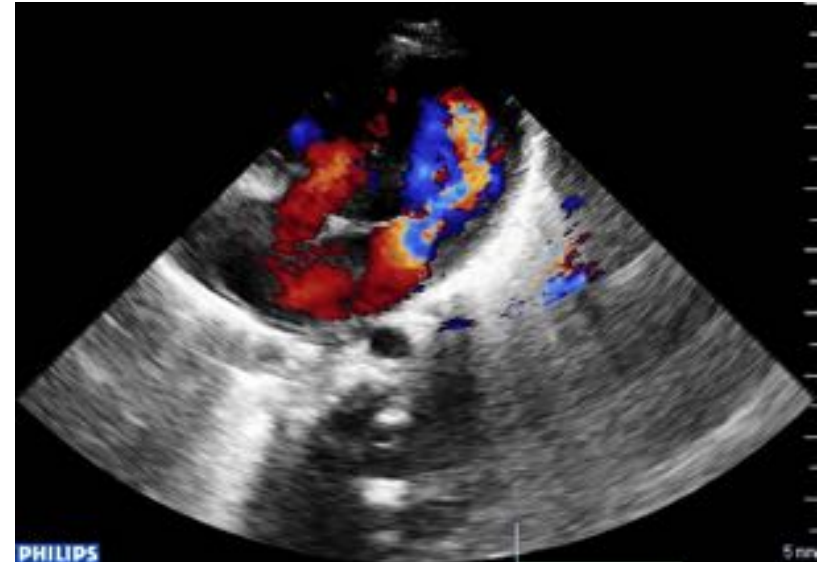
5) Mitral ring



Anneau supra-mitral

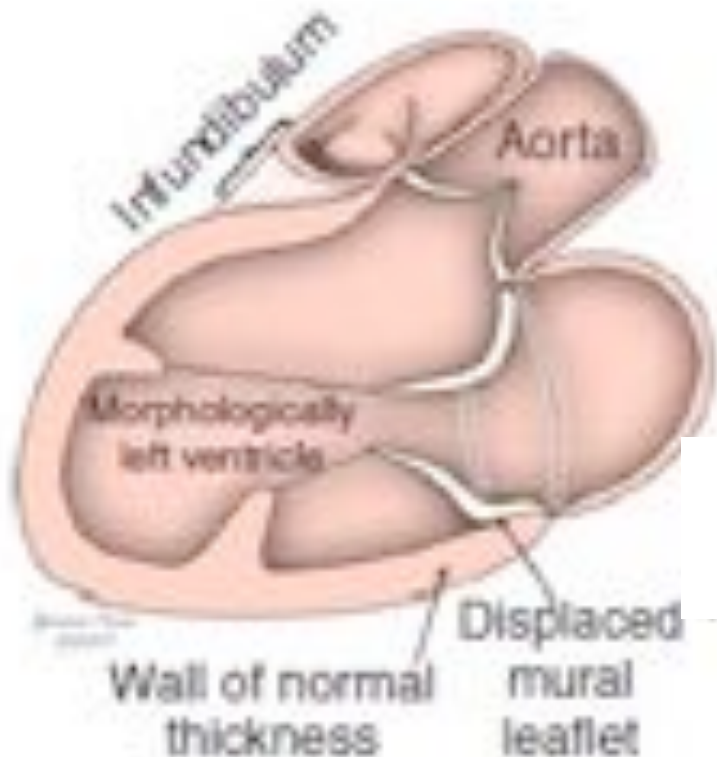


Anneau intra-mitral

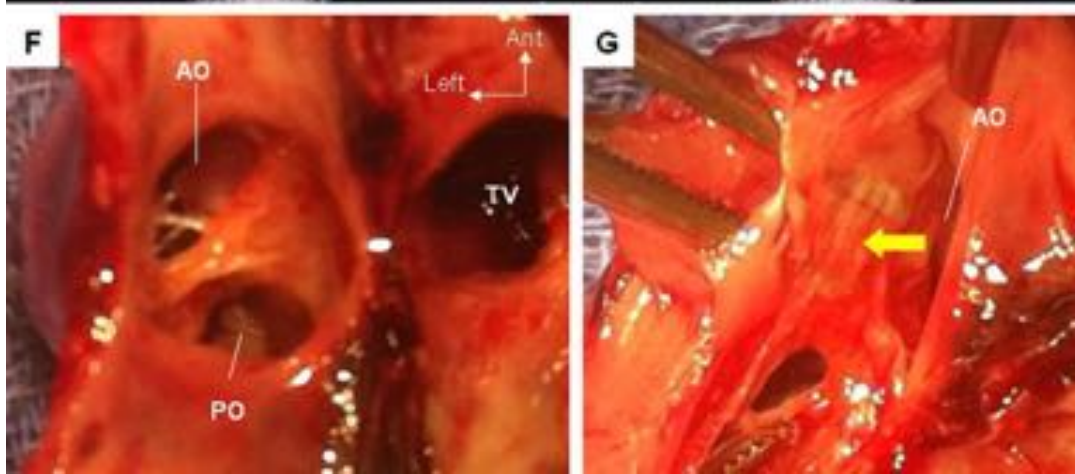
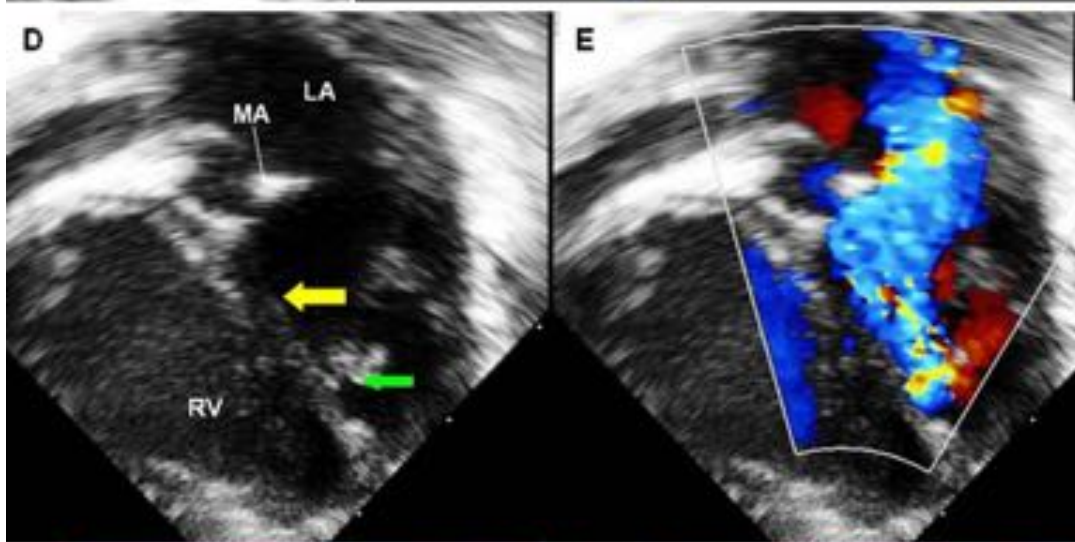
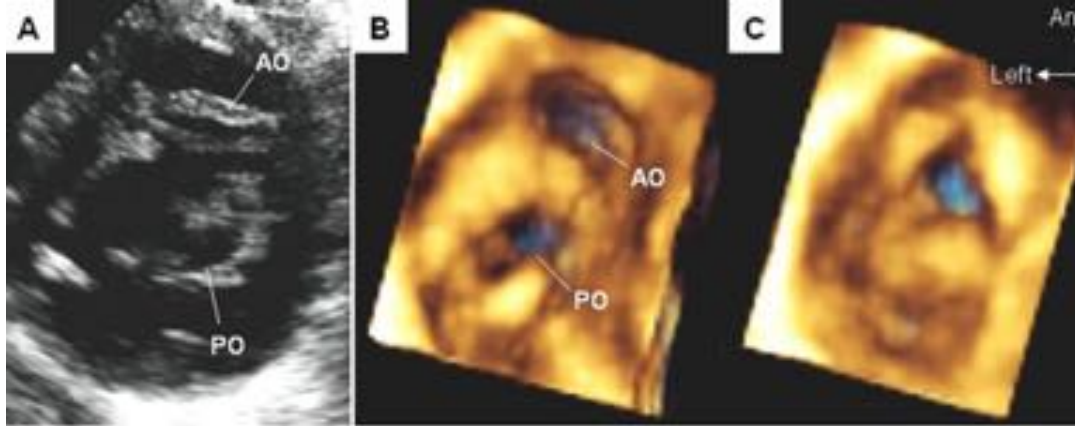


ANOMALIES DES FEUILLETS

6) Ebstein de la valve mitrale



Ruschhaupt et al. Am J Card 1976.



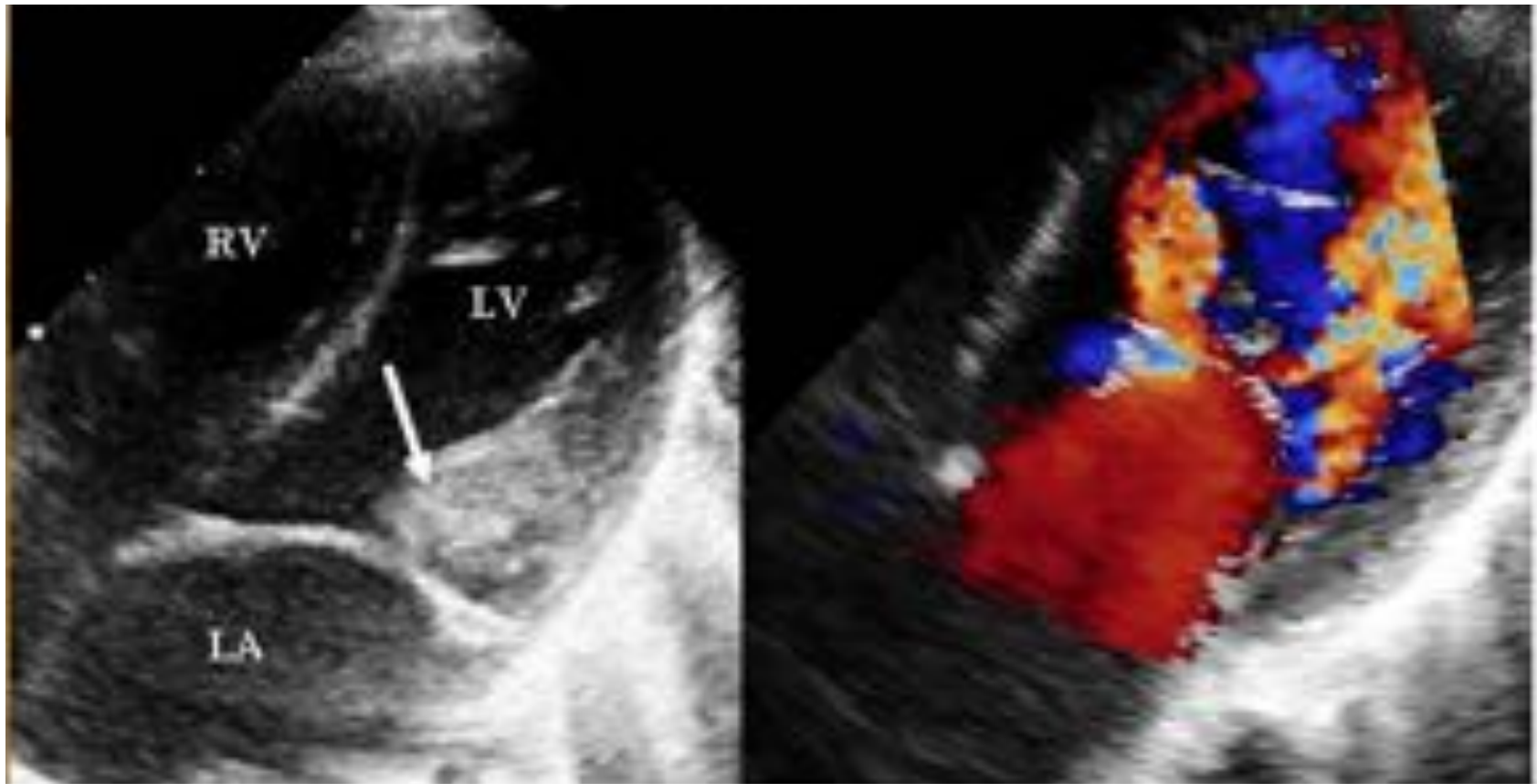
J2 de vie

ANOMALIES DES CORDAGES

1) Arcade / Hamac

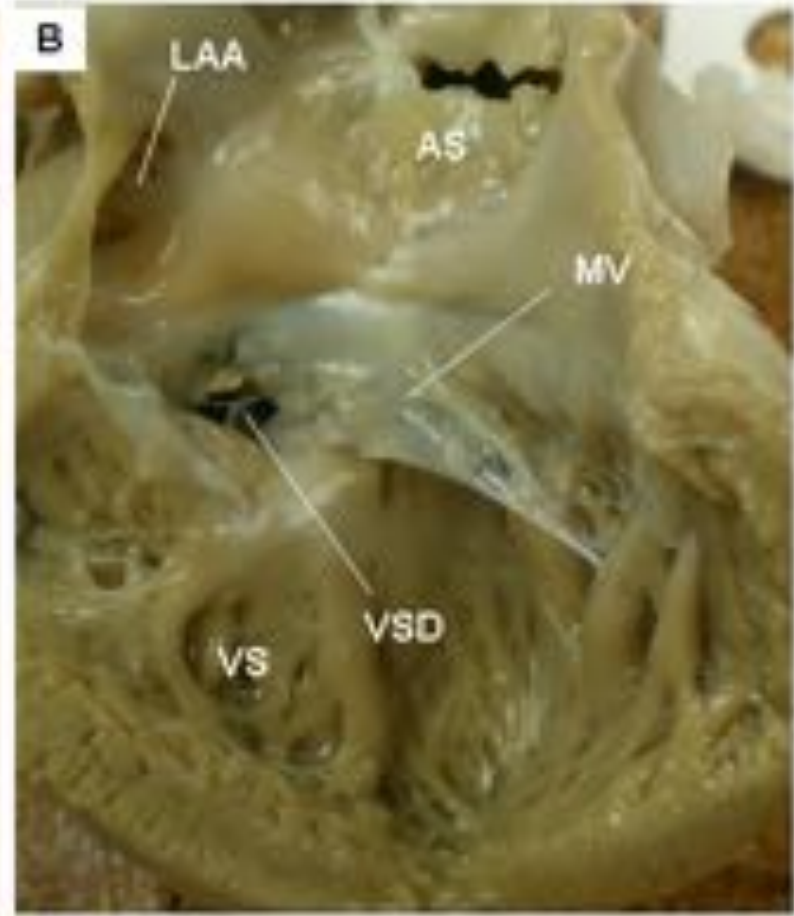
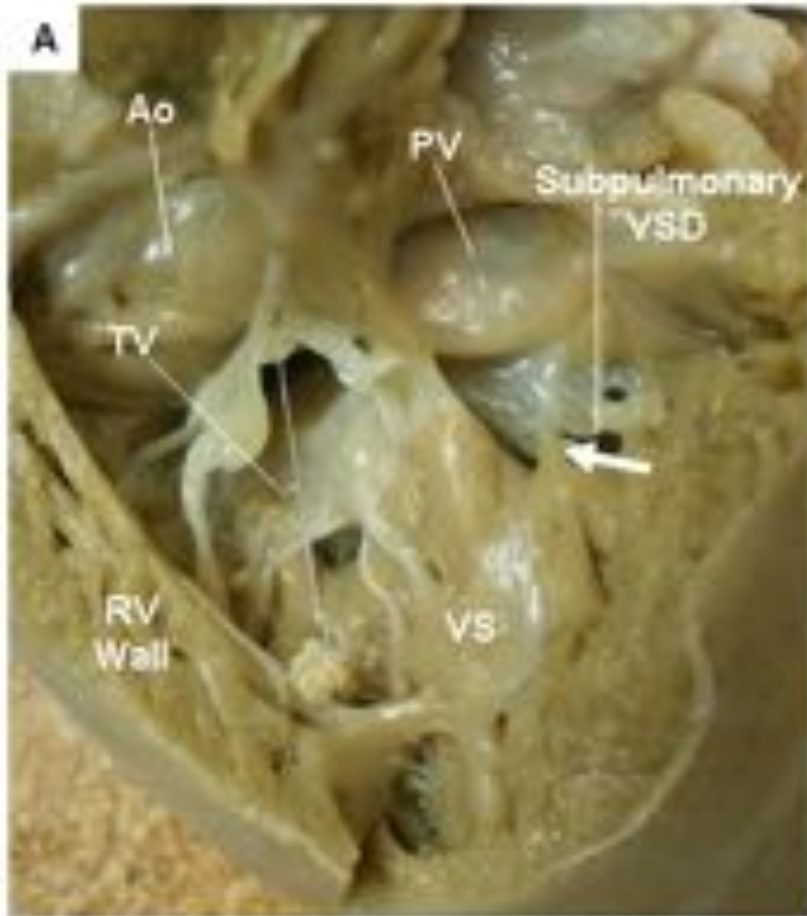


1) Arcade / Hamac



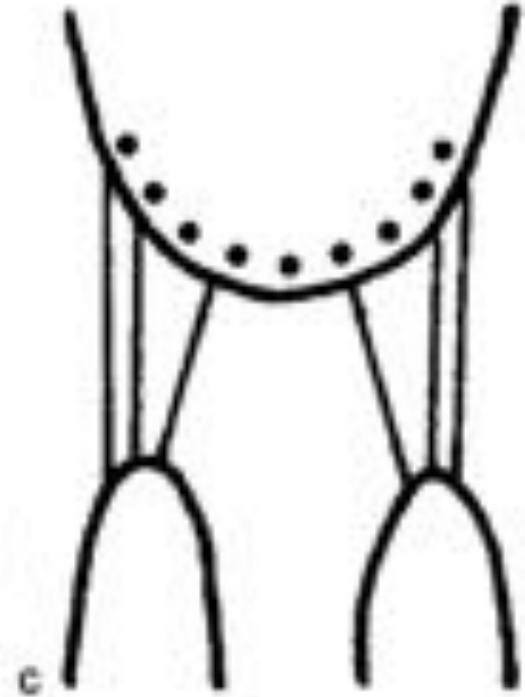
ANOMALIES DES CORDAGES

2) Straddling mitral



ANOMALIES DES PILIERS

Parachute / Parachute-like



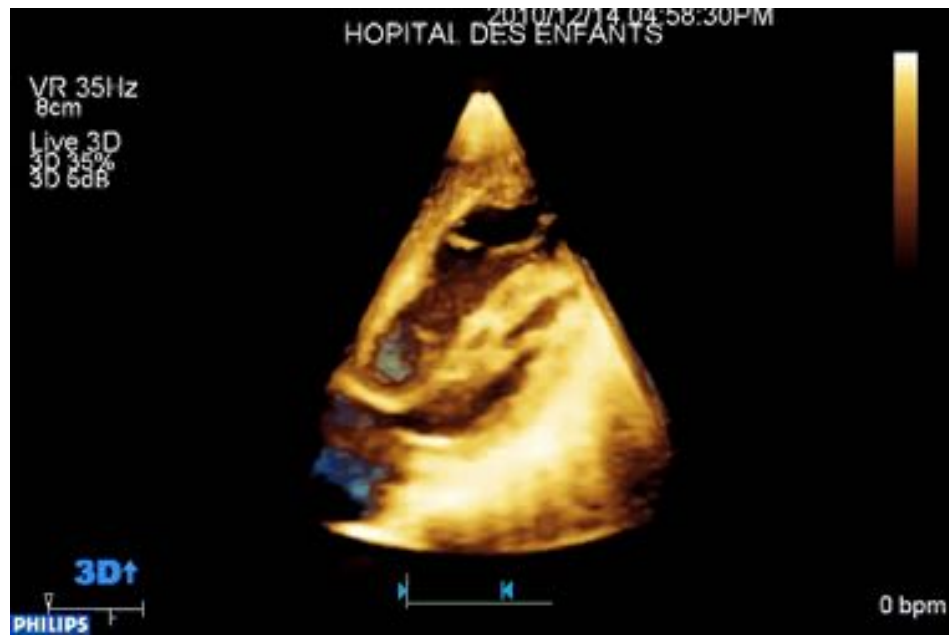
Parachute mitral valve



Parachute-like asymmetric mitral valve



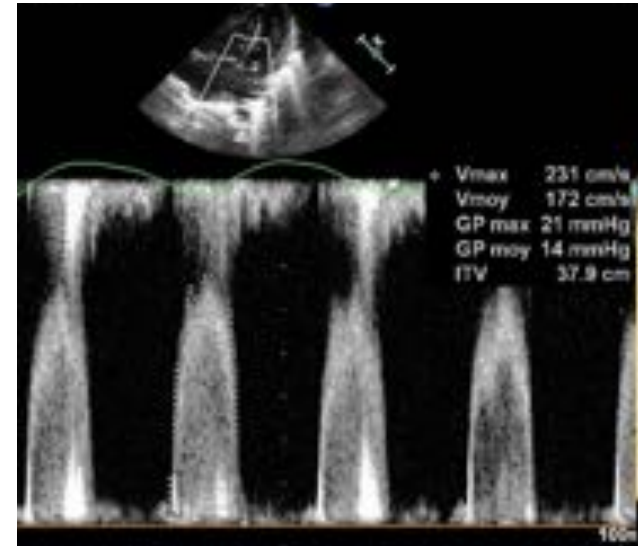
Parachute / Parachute-like



Sténose: quand intervenir ?

gradient moyen transmitral :

10 – 15 mmHg = sténose modérée
> 15 mmHg = sténose sévère



- Moore (Circulation 1994): **11.5 mmHg** ± 4.4 [n=85]
- Serraf (Circulation 2000): **12.6 mmHg** ± 7 [n=72]
- Henaine (Ann Thorac Surg 2010): **16.8 mmHg** ± 4.4 [n=29]
- Hoashi (Ann Thorac Surg 2010): **13.5 mmHg** ± 2.6 [n=20]

Insuffisance mitrale: quand intervenir ?

- Pas de diamètre VG seuil chez l'enfant...
(Valeur objective: DTS VG > 2 Z score)
- Mais une ↗ rapide du DTS VG traduit une ↘ de la fonction VG

EN CONCLUSION

- Pathologies rares
- Evaluation échographique indispensable pour le chirurgien
- Formes complexes = plusieurs anomalies mitrales associées