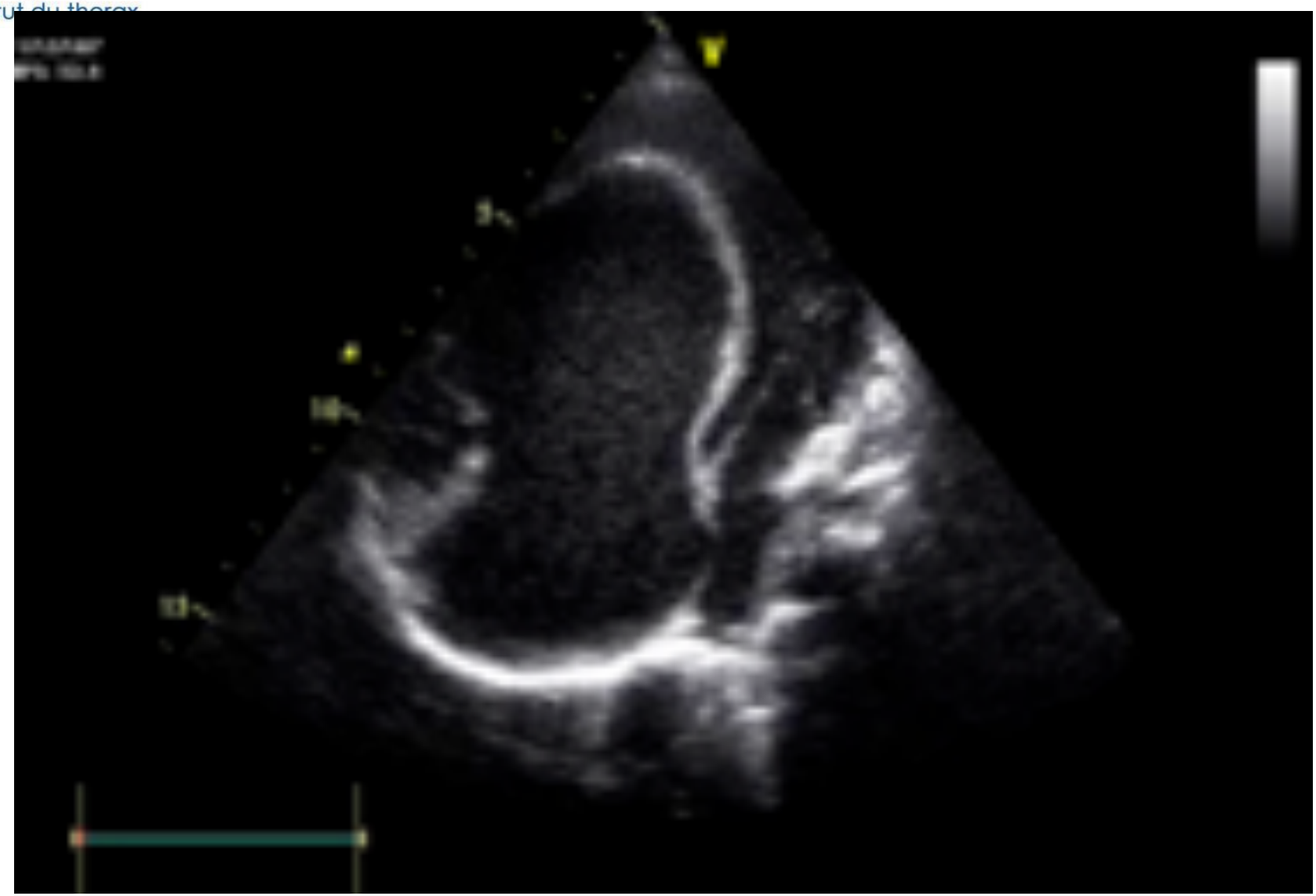
# Mme Pierrette D, 56 ans

- Souffle connu depuis l'enfance
- Cyanosée à l'effort
- 3 grossesses, sans souci
- Pas de suivi cardiologique depuis longtemps, car « elle a eu peur »
- Revient car plus essoufflée à l'effort depuis quelques mois



# Pour quelles raisons peut-elle être plus essoufflée?

- Elle doit avoir une IT sévère
- C'est à cause du shunt G-D de la CIA
- C'est l'âge!
- C'est la cyanose secondaire au shunt D-G

l'institut du thorax

# Maladie d'Ebstein

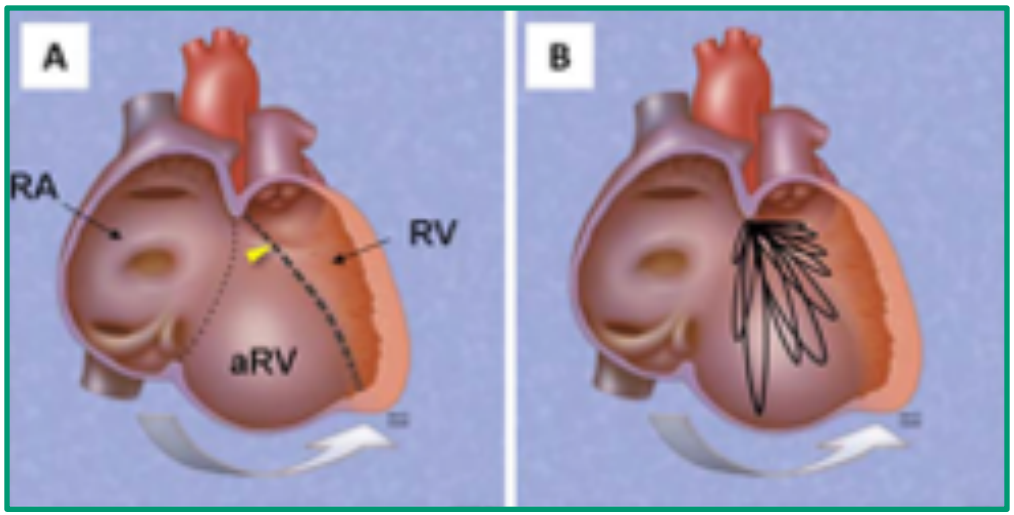
CENTRE HOSPITALIER UNIVERSITAIRE DE NANTES

- Anomalie rare, <1% des cardiopathies congénitales

**Concerning a very rare case of insufficiency of the tricuspid valve caused by a congenital malformation**

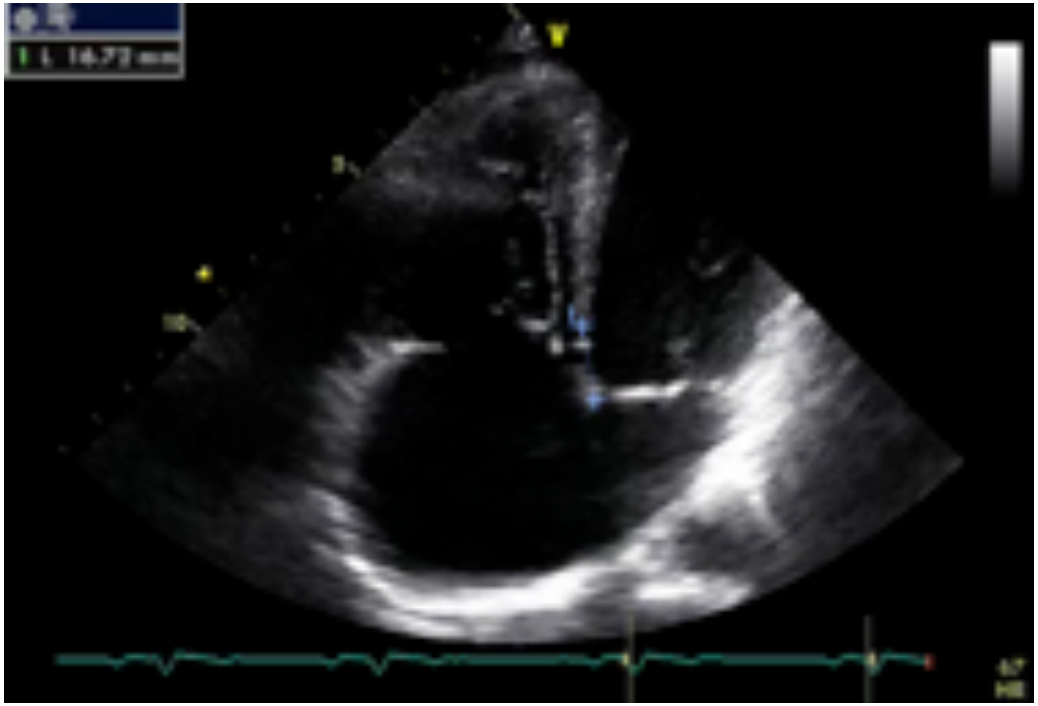
Ebstein W. *Arch Anat Physiol 1866*

- Maladie de la valve tricuspide et du VD:
  - ✓ Non délamination des feuillets septal et postérieur
  - ✓ Déplacement apical et vers RVOT de l'anneau fonctionnel
  - ✓ Dilatation de la portion atrialisée du VD
  - ✓ Anomalies du feuillet antérieur (fenestrations, attaches)
  - ✓ Dilatation de la jonction AV (anneau vrai)




Attenhofer Jost et al. *Circulation 2007*
Anderson et al. *Mayo Clin Proc 1979*
Qureshi et al. *Trends Cardiovasc Med 2018*

- Niveau de déplacement maximal: commissure feuillets S et P
- Quand >8 mm/m2 par rapport au feuillet antérieur mitral : maladie d'Ebstein



Attenhofer Jost et al. *Circulation 2007*
Anderson et al. *Mayo Clin Proc 1979*
Edwards. *Prog Pediatr Cardiol 1993*

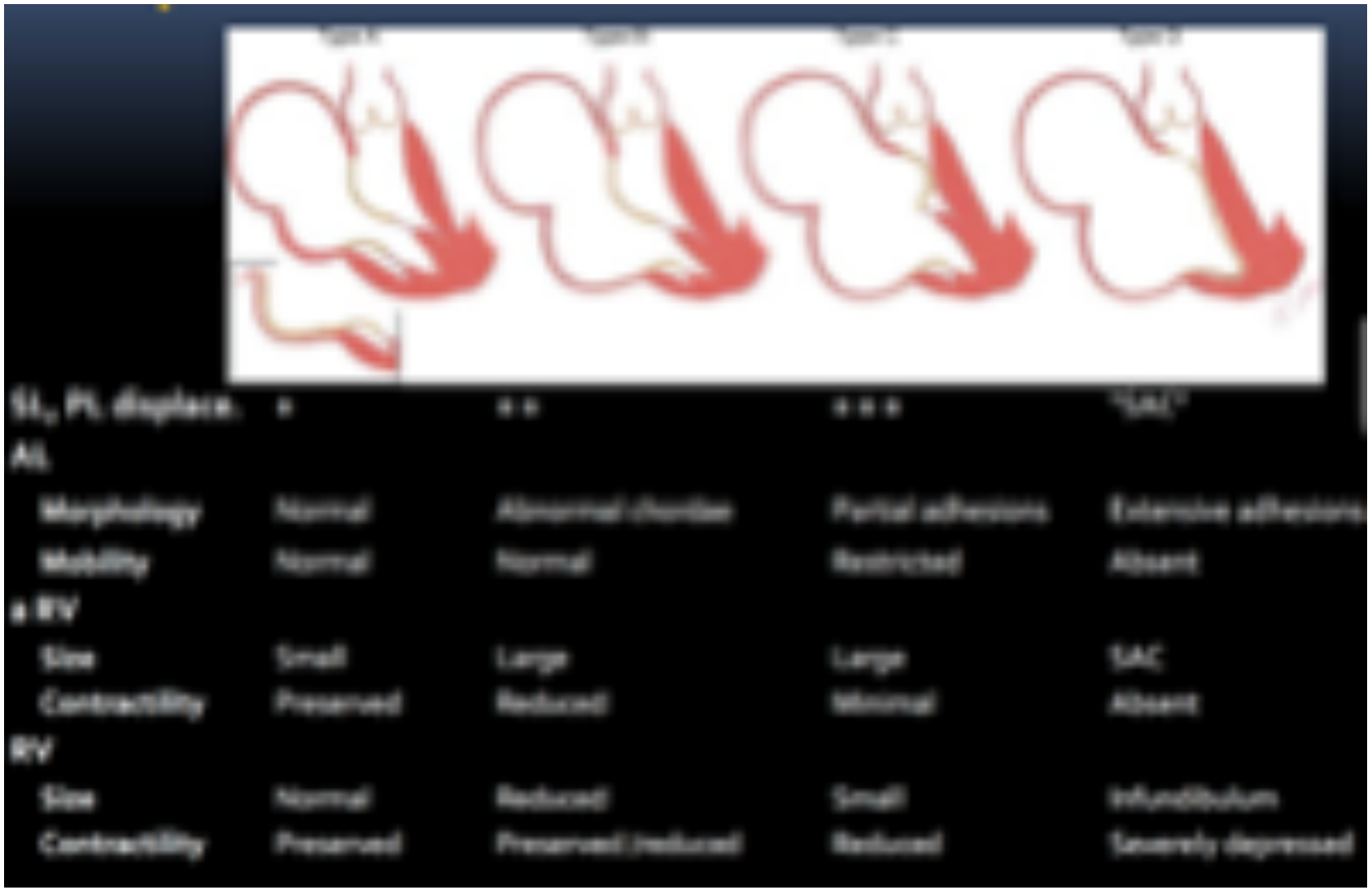
## Quelles complications en l'absence de réparation?

- Arythmies A/V
- Cyanose
- Embolie paradoxale
- EI
- Insuffisance cardiaque D
- Insuffisance cardiaque G
- Mort subite

## Imagerie multimodalités+++ (ETT/IRM) pour:

- Mobilités/Attaches du feuillet antérieur
- Voie pulmonaire
- Quantification de l'IT
- Fonction du VD
- Lésions associées
- NCVG

# Classification de Carpentier



| SL, PL displace. | + | + + | + + + | "SAC" |
|---|---|---|---|---|
| AL | | | | |
| Morphology | Normal | Abnormal chordae | Partial adhesions | Extensive adhesions |
| Mobility | Normal | Normal | Restricted | Absent |
| a RV | | | | |
| Size | Small | Large | Large | SAC |
| Contractility | Preserved | Reduced | Minimal | Absent |
| RV | | | | |
| Size | Normal | Reduced | Small | Infundibulum |
| Contractility | Preserved | Preserved (reduced) | Reduced | Severely depressed |

Carpentier et al. *J Thorac Cardiovasc Surgery 1988*

| | | |
|---|---|---|
| IIa | B-NR | 3. Electrophysiological study with or without catheter ablation can be useful in the diagnostic evaluation of adults with Ebstein anomaly and ventricular preexcitation but without supraventricular tachycardia (S4.3.4-3, S4.3.4-4). |
| IIa | B-NR | 4. In adults with Ebstein anomaly, electrophysiological study (and catheter ablation, if needed) is reasonable before surgical intervention on the tricuspid valve even in the absence of preexcitation or supraventricular tachycardia (S4.3.4-5). |

| | | |
|---|---|---|
| I | B-NR | 5. Surgical repair or reoperation for adults with Ebstein anomaly and significant TR is recommended when one or more of the following are present: HF symptoms, objective evidence of worsening exercise capacity, progressive RV systolic dysfunction by echocardiography or CMR (S4.3.4-6–S4.3.4-10). |
| IIa | B-NR | 7. Surgical repair or reoperation for adults with Ebstein anomaly and significant TR can be beneficial in the presence of progressive RV enlargement, systemic desaturation from right-to-left atrial shunt, paradoxical embolism, and/or atrial tachyarrhythmias (S4.3.4-11, S4.3.4-13, S4.3.4-14). |
| IIb | B-NR | 8. Bidirectional superior cavopulmonary (Glenn) anastomosis at time of Ebstein anomaly repair may be considered for adults when severe RV dilation or severe RV systolic dysfunction is present, LV function is |
| | | preserved, and left atrial pressure and LV end diastolic pressure are not elevated (S4.3.4-6, S4.3.4-15). |

Stout et al. *JACC 2018*