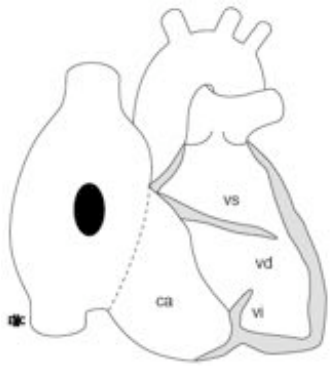
CHU Reims



UNIVERSITÉ DE REIMS CHAMPAGNE-ARDENNE

Malformation d'Ebstein

Pr. Pierre Mauran
AMH-CHU Reims

M3C

Centre de Compétence
Malformations Cardiaques Congénitales Complexes
American Memorial Hospital - CHU Reims

VD normal

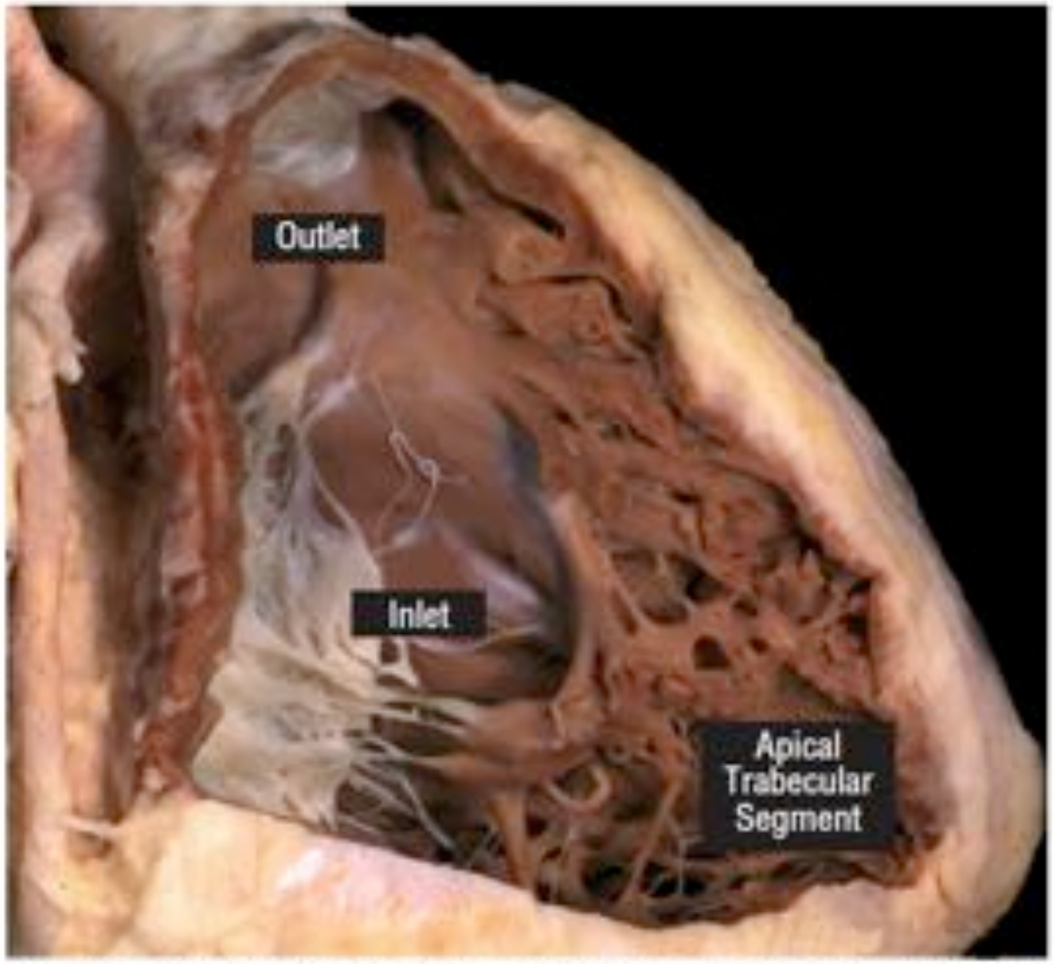
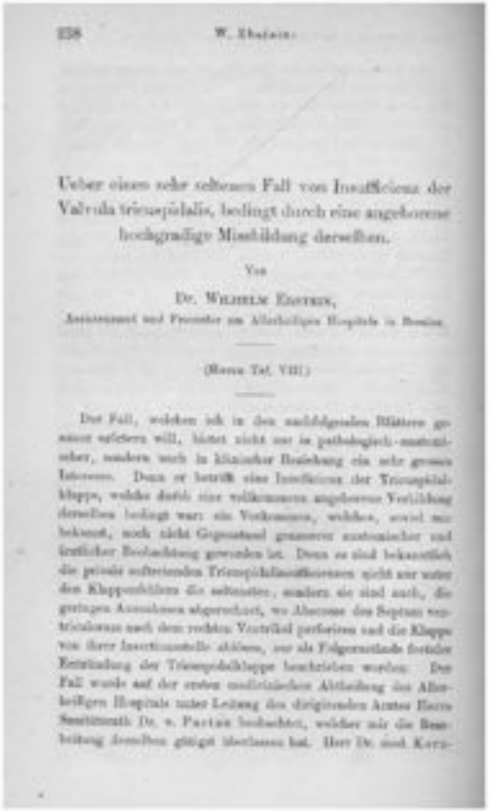


Figure 1. This window dissection shows the 3 components of a normal right ventricle. Courtesy of Professor Y. Ho.

Alonzo-González et al. The right heart in adults with congenital heart disease. Rev Esp Cardiol. 2010;63(9):1070-86

Wilhelm Ebstein 1866

Photograph shows Wilhelm Ebstein, ca. 1885.
(© Städtisches Museum; Göttingen, Germany)

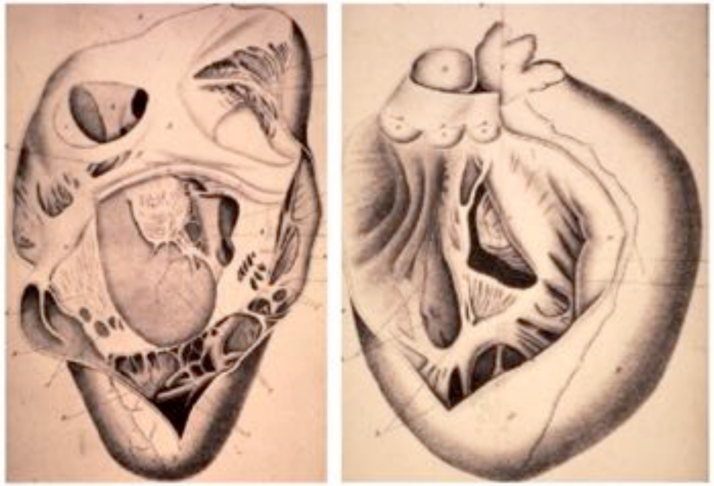


Ueber einen sehr seltenen Fall von Insufficienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben.

Von

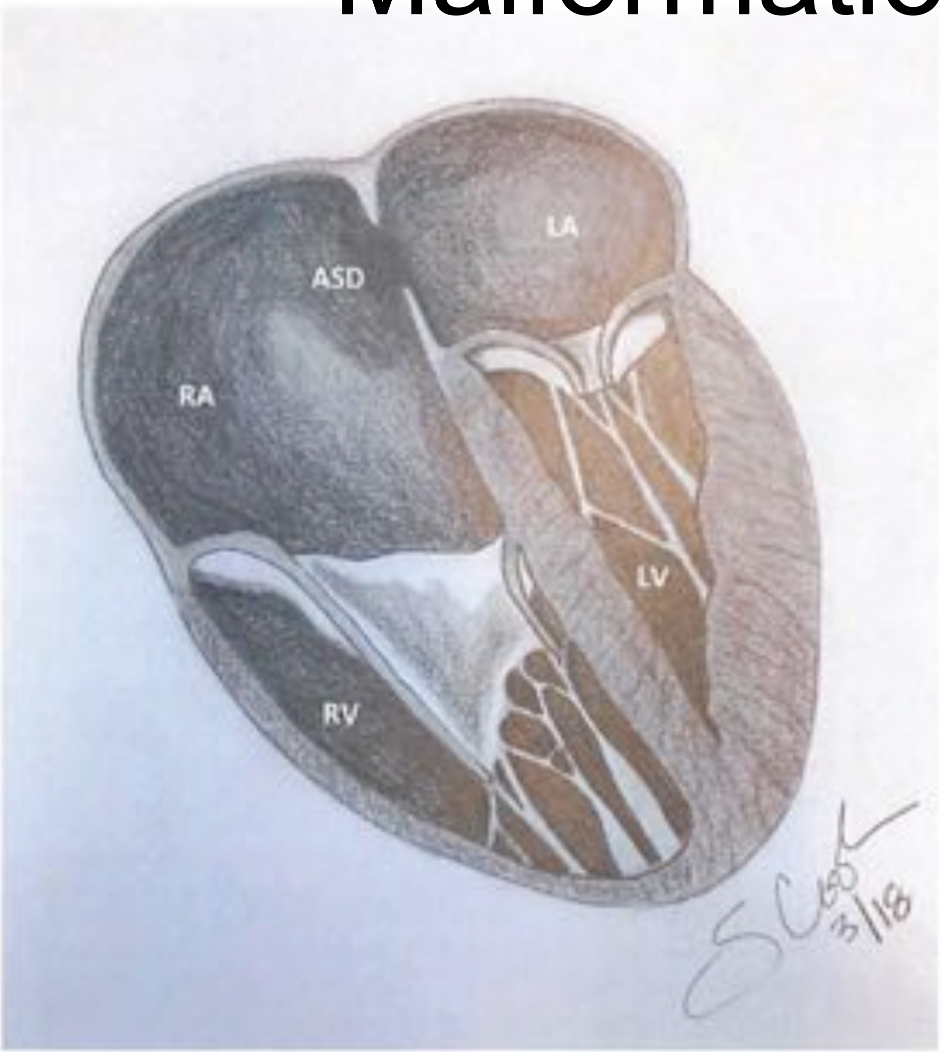
Dr. Wilhelm Ebstein,

The title page from Ebstein's original paper.
(Courtesy of BIU Santé.) Available from: http://www.biusante.parisdescartes.fr/histmed/medica/page?epo0270&p=1.

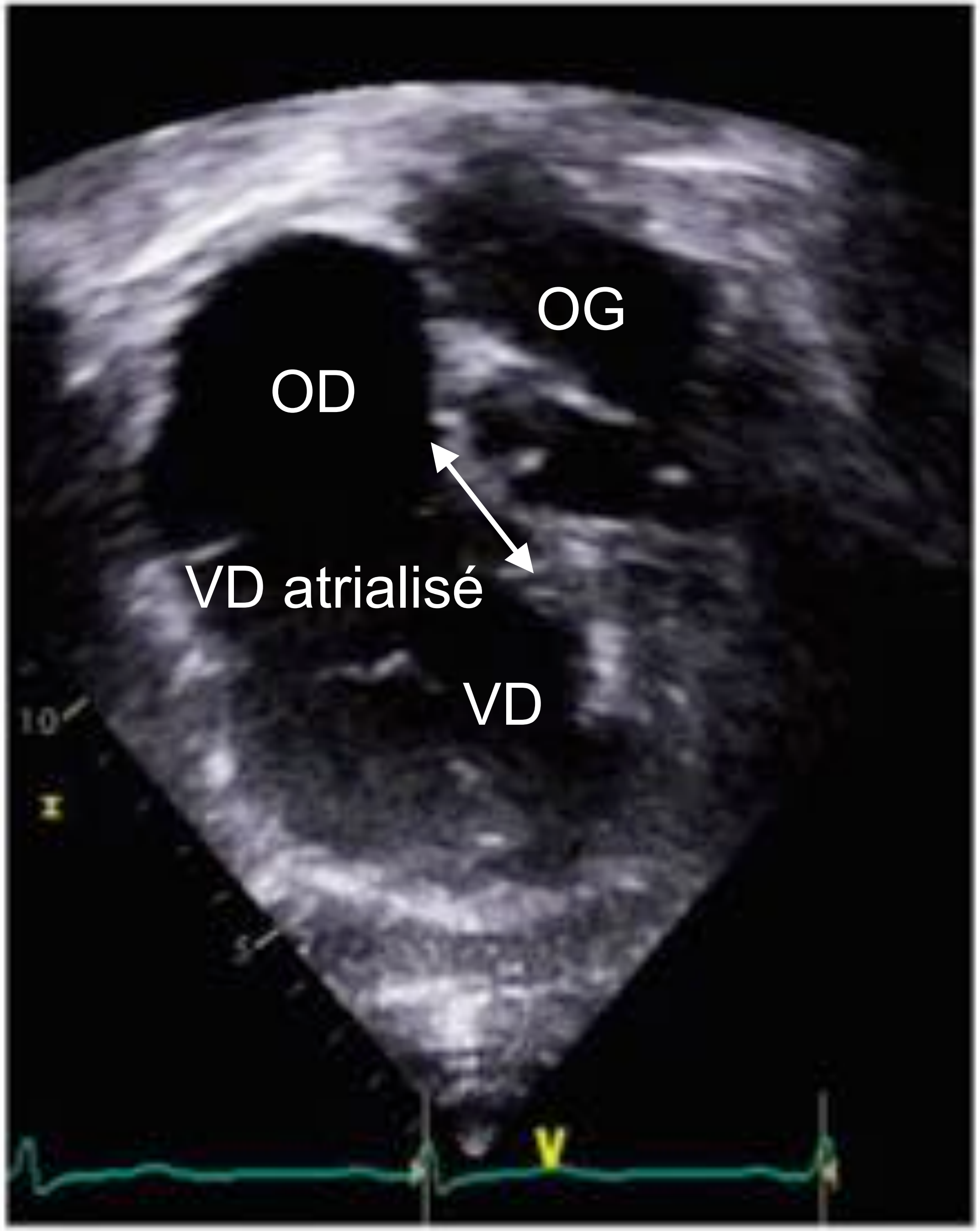


M. Mazurak et al. The two anomalies of Whilelm Ebstein. Tex Heart Inst J. 2017;44(3): 198-201

Malformation d'Ebstein



- Défaut de délamination du feuillet septal de la tricuspide
- Feuillet antérieur en « voile de navire »
- Déplacement de l'anneau fonctionnel tricuspide vers la pointe du VD
- Atrialisation d'une partie de la chambre d'admission du VD
- Dilatation de l'anneau tricuspide et de l'OD


OG
OD
VD atrialisé
VD

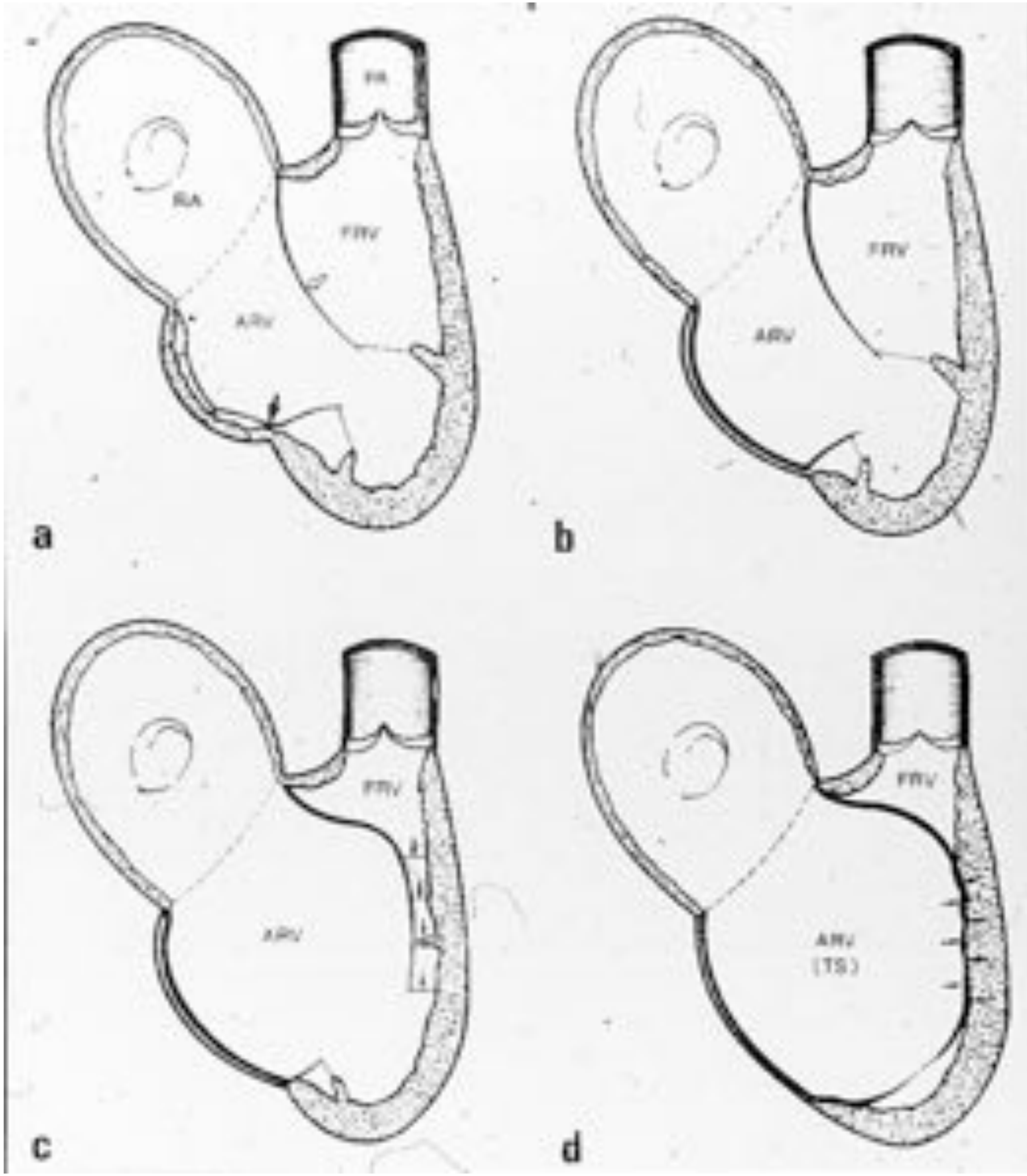
Historique

- 1866 : publication de W. Ebstein
- 1927 : « anomalie d'Ebstein »
- 1951 : Soloff *et al.* diagnostic par cathétérisme cardiaque
- 1962 : Ch. Barnard RVT par bioprothèse

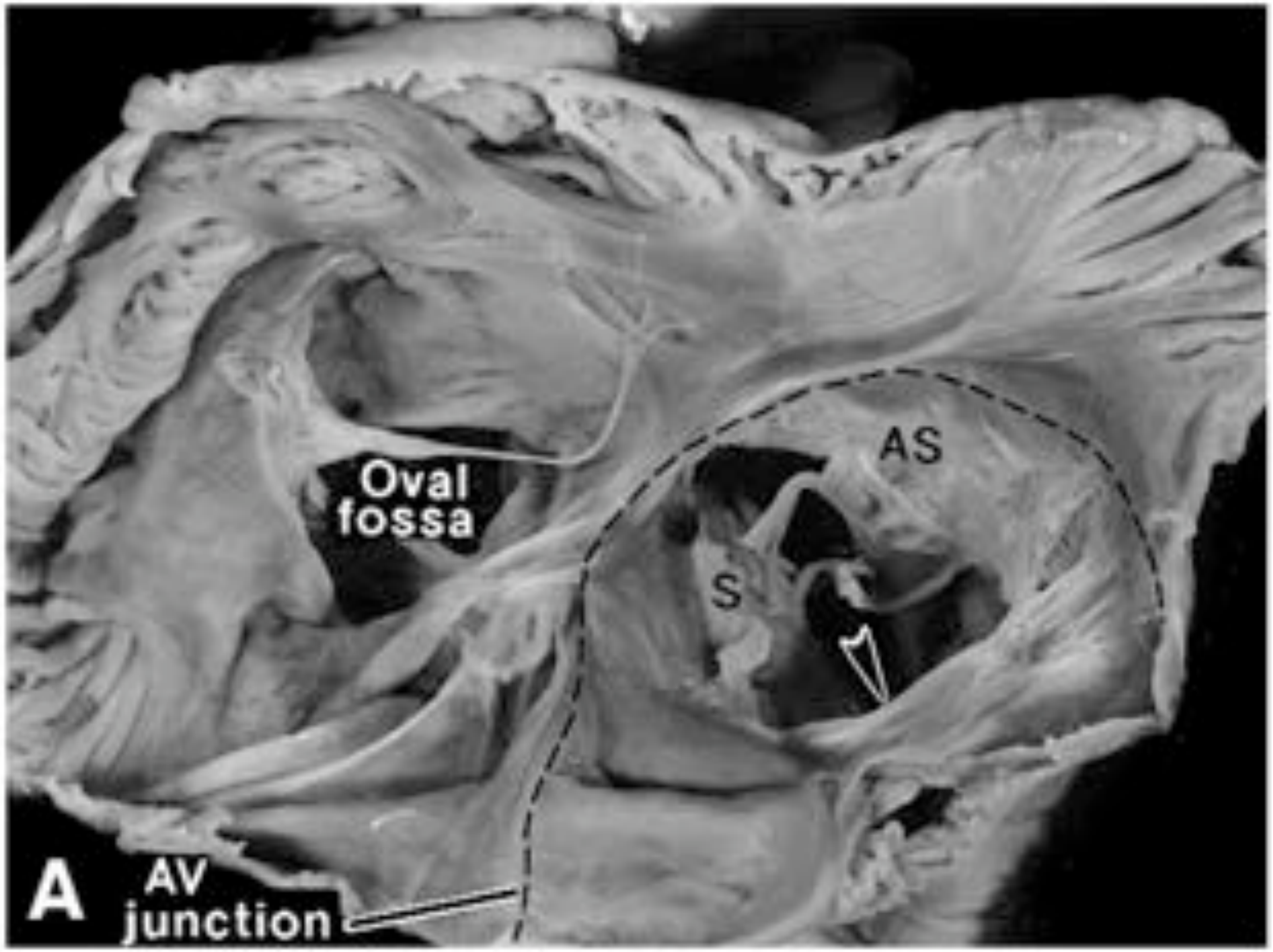
Classification de Carpentier *et al.* 1988

- Type A : volume du VD fonctionnel satisfaisant
- Type B : atrialisation d’une large portion du VD mais ample mouvement du feuillet antérieur de la tricuspide
- Type C : restriction sévère du mouvement de la valve antérieure pouvant créer une sténose infundibulaire
- Type D : atrialisation complète du VD, à l’exception d’un petit infundibulum

Ebstein



Malformation d'Ebstein type A



Ch. Schreiber *et al.* J Thorac Cardiovasc Surg 1999;117:148-55

Malformation d'Ebstein type C

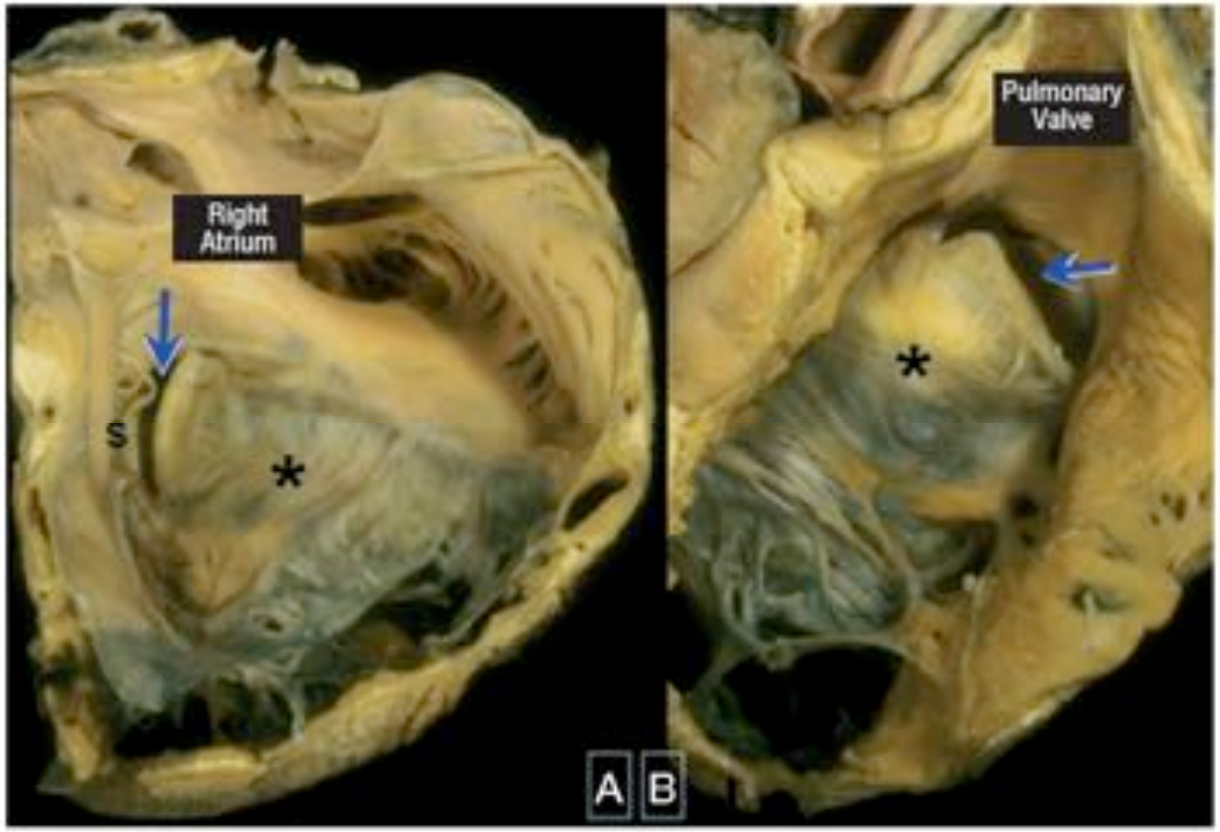


Figure 9. Ebstein malformation of the tricuspid valve viewed from the right atrium (A) and from the right ventricular outflow tract (B). In this heart the sail-like leaflet (*) has a limited free margin. This results in a key-hole aperture (arrow) between the anterior leaflet and the underdeveloped septal leaflet (s) that is the effective valvar orifice displaced toward the outflow tract. Courtesy of Professor Y. Ho.

R. Alonzo-González et al. The right heart in adults with congenital heart disease. Rev Esp Cardiol. 2010;63(9):1070-86

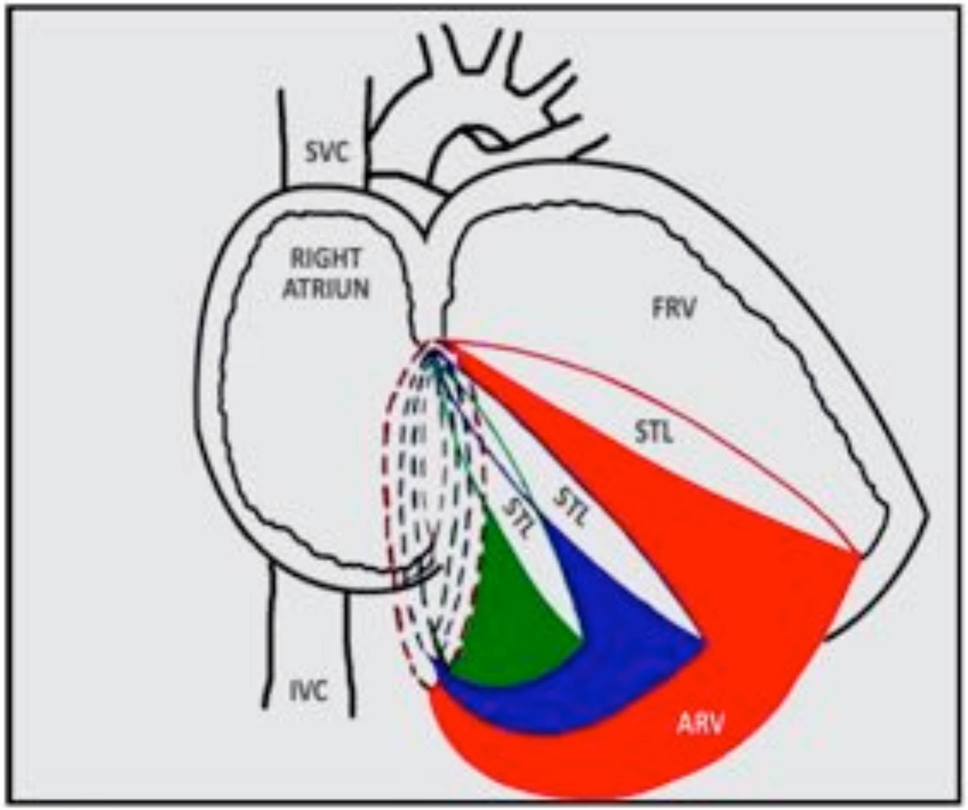


Fig. 4 - *Schematic representation of severity of Ebstein's anomaly. As the displacement of septal tricuspid leaflet increases, there is a corresponding increase in true tricuspid annulus dilatation and a decrease in the amount of functional right ventricle. Broken lines indicate true tricuspid annulus. Coloured region indicates the thinned out atrialized right ventricle. Black – normal; green – mild dilatation; blue – moderate dilatation; and red – severe dilatation.*

A.Malhotra *et al.* Braz J Cardiovasc Surg 2018;33(4):353-61

Grande variété des formes du VD et de la tricuspide

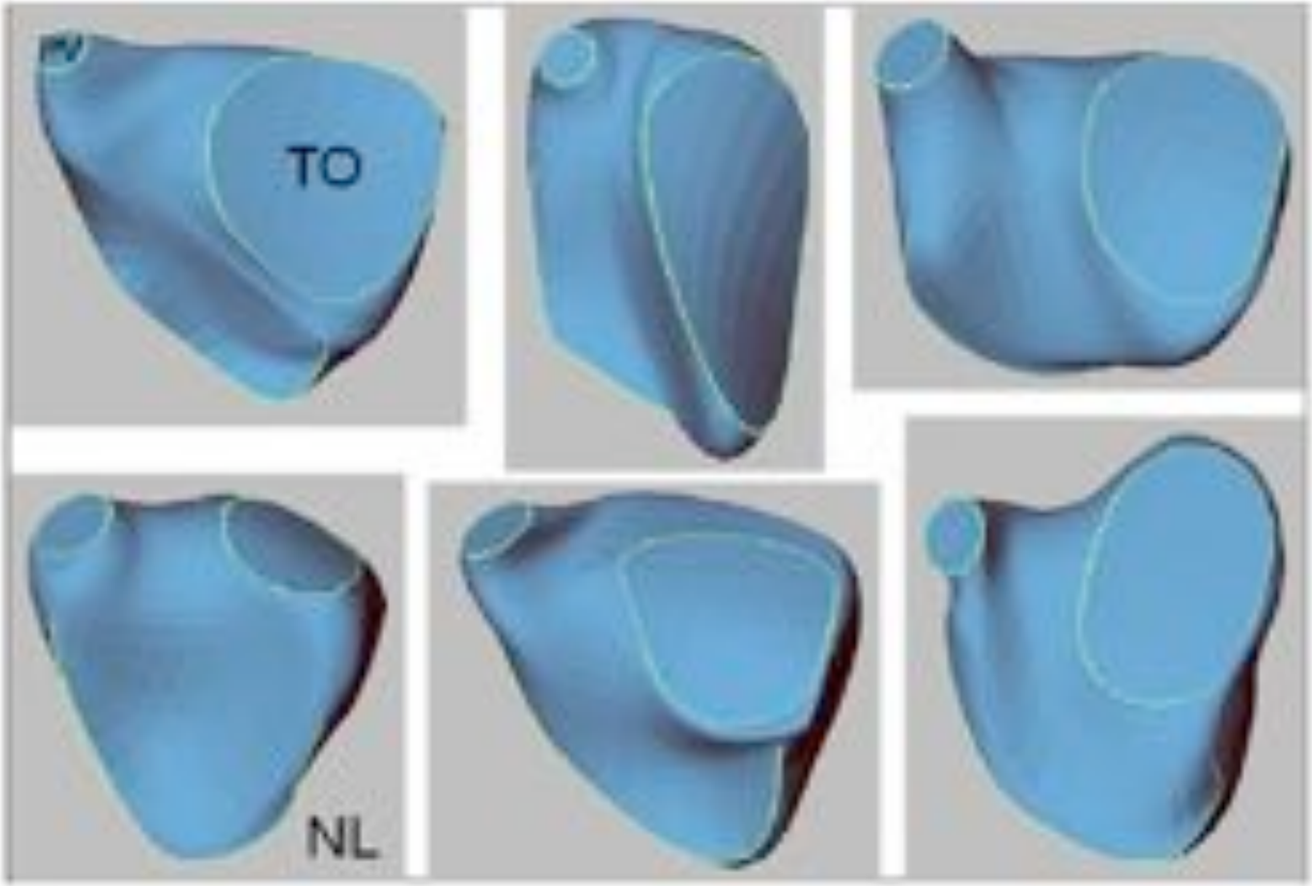


Fig. 7. There was wide shape variability in the right ventricle and tricuspid orifice (TO) of Ebstein's anomaly patients, as illustrated here by 3D reconstruction in five patients, compared with one normal (NL) subject (below left).

Ch.M.Lee *et al.* The shape and function of the right ventricle in Ebstein's anomaly.
Int J Cardiol (2012), doi:10.1016/ j.ijcard.2012.03.062

Ch.M.Lee *et al.* The shape and function of the right ventricle in Ebstein's anomaly.
Int J Cardiol (2012), doi:10.1016/ j.ijcard.2012.03.062

A B S T R A C T

Background: Ebstein's anomaly involves both the right ventricle (RV) and tricuspid valve.
Methods: The functional RV and tricuspid orifice were traced from magnetic resonance images in 29 adult Ebstein patients and 9 normal subjects and reconstructed for visualization and measurement of regional RV size, function, and shape at 20 cross sections, and inlet and outflow tract ejection fractions (EFs).
Results: The RV in Ebstein's had RV dilation (end diastolic volume index 179 ± 69 vs. 84 ± 22 ml/m^2 in normals, $p<0.001$) and global dysfunction (EF 45 ± 8 vs. $55 \pm 5\%$ in normals, $p<0.001$). Longitudinal contraction was preserved (26 ± 13 vs. 26 ± 4 mm in normals) and correlated more weakly with EF than short axis fractional shortening ($r=0.44$ vs. $r=0.71$, $p<0.05$ for both). The apical region in Ebstein's RV was enlarged, rounded and contributed more than normal to the global stroke volume. However this contribution correlated inversely with global EF. In contrast slices in the basal region had normal cross sectional area and their function correlated directly with global EF. Inlet EF was depressed ($46 \pm 8\%$ vs. 55 ± 6 in normals, $p=.002$); outflow tract EF was even more depressed (39 ± 14, $p=0.019$).
Conclusion: The three dimensional shape of the RV in adult Ebstein patients was demonstrated. The Ebstein's RV remodels in diverse regional patterns rather than following a shape continuum. Changes at the apex and base had opposing effects on function. Global EF was supported more by short axis than longitudinal contraction.

Ch.M.Lee *et al.* The shape and function of the right ventricle in Ebstein's anomaly.

Int J Cardiol (2012), doi:10.1016/ j.ijcard.2012.03.062

- Dilatation du VD :
  - VTD : 179±69 vs 84±22 mL/m^2
- Dysfonction du VD :
  - FE : 45±8 vs 55±5%
- Région apicale du VD
  - Dilatée et arrondie
  - Contribuant plus que normalement à la fonction globale
- Contraction longitudinale préservée :
  - TAPSE : 26±13 vs 26±4 mm
  - mal corrélée à la fonction globale (R^2 = 0,19 vs 0,50)
- Raccourcissement du petit axe
  - bien corrélé à la fonction globale (R^2 = 0,391 , p< 0,001)

S. Fratz *et al.* The functional right ventricle and tricuspid regurgitation in Ebstein's anomaly. Int J Cardiol (2012), doi:10.1016/j.ijcard.2011.12.081

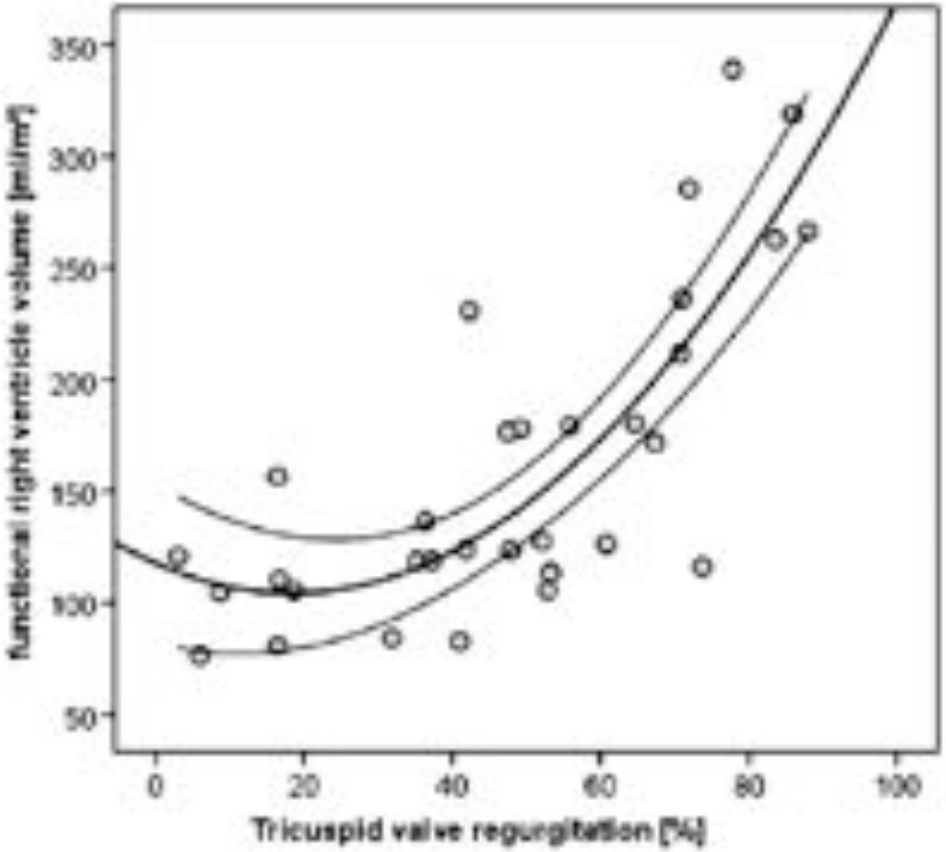


Fig. 1. Strong positive statistically significant correlation of functional right ventricle volume and tricuspid regurgitation (polynomial regression: $y = 118.6 - 1.6\,x + 0.04\,x^2$, $p < 0.001$, $R^2 = 0.65$). Depicted lines represent fitted regression equation with 95% confidence band.

Extended Glasgow Outcome Scale

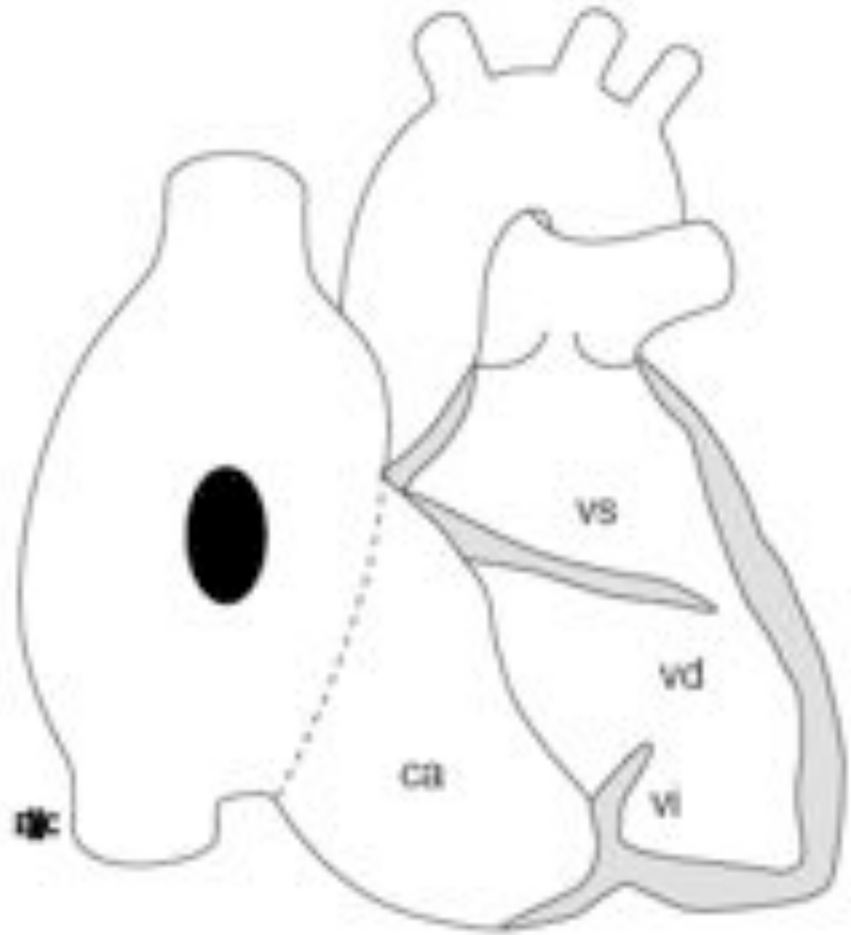
D.S.Celermajer et al. Ebstein's anomaly: presentation and outcome from fetus to adult. J Ann Coll Cardiol 1994;23:170–6.

Echographie **chez le nouveau-né**

Surface (OD + VD atrialisé) / Surface (VD fonctionnel + OG + VG)

- Grade 1 : < 0,5
- Grade 2 : 0,5 - 0,99
- Grade 3 : 1 - 1,49
- Grade 4 : ≥ 1,5

Malformation d’Ebstein



- Rare :
  - 1 à 2 / 100 000 naissances vivantes
  - 0,5 % des cardiopathies congénitales

- Traitement maternel pendant la grossesse par :
  - Lithium : risque multiplié par 400
  - Benzodiazépines

R. Cabrera *et al*. Identification of clinically relevant phenotypes in patients with Ebstein's anomaly. Clinical Cardiology. 2018;41:343–348.

N : 147

Characteristic	Frequency, n	%
Female sex	69	46.9
Age at first consult at institution, y		
Early childhood, 0–5	63	42.9
Childhood, 6–11	27	18.4
Adolescence, 12–17	30	20.4
Young adult, 18–26	9	6.1
Adulthood, 27–59	17	11.6
Old age, ≥60	1	0.7
Severity of disease (per Carpentier)		
Type A	33	22.4
Type B	41	27.9
Type C	45	30.6
Type D	28	19.0
Familial cases	7	4.8

Associations

- CIA ostium secundum ou FOP
- WPW

- CIV
- Sténose pulmonaire
- APSI
- T4F
- CoAo
- Anomalies mitrales

- Doubles discordances : 1/3 ont un Ebstein

R. Cabrera *et al*. Identification of clinically relevant phenotypes in patients with Ebstein's anomaly. Clinical Cardiology. 2018;41:343–348.

N: 147

Comorbidities	N	%
ASD	89	60.5
Ostium secundum	62	42.2
PFO	22	15.0
Ostium primum	5	3.4
WPW	39	26.5
Complete RBBB	26	17.7
RVOT obstruction	23	15.6
Pulmonary stenosis	11	7.5
Pulmonary atresia with VSD	6	4.1
Pulmonary atresia without VSD	5	3.4
Tetralogy of Fallot	1	0.7
SVT	21	14.3
Incomplete RBBB	21	14.3
PDA	19	12.9

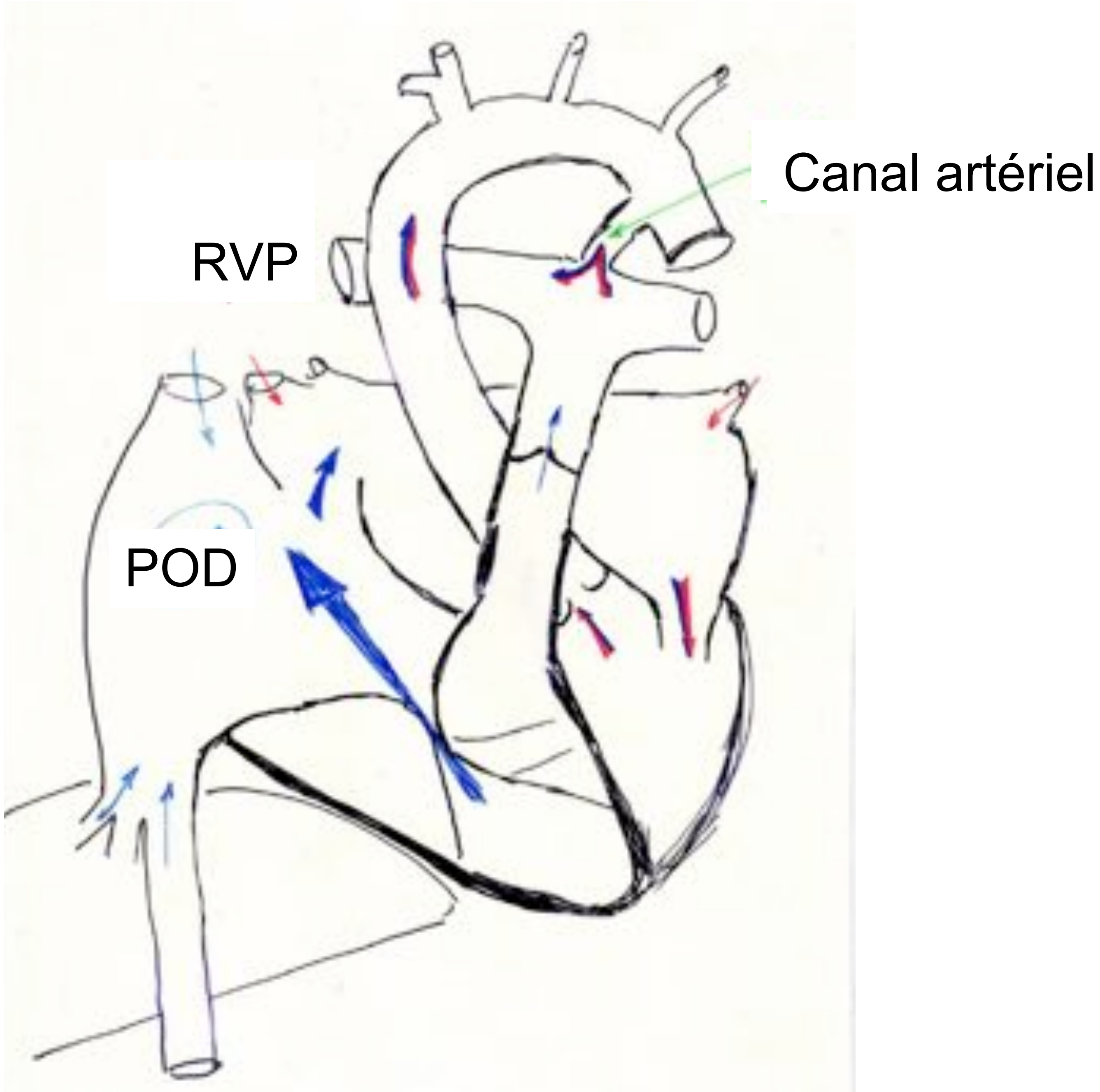
R. Cabrera *et al*. Identification of clinically relevant phenotypes in patients with Ebstein's anomaly. Clinical Cardiology. 2018;41:343–348.

N: 147

	N	%
Syndromic	14	9.5
Cognitive or developmental delay	3	2.0
Down syndrome	5	3.4
Kleefstra syndrome	1	0.7
1p36 deletion syndrome	1	0.7
Turner syndrome	1	0.7
VACTERL association	1	0.7
Congenital rubella syndrome	1	0.7
Scimitar syndrome suspicion	1	0.7

Physiopathologie

- L’hémodynamique est fonction
  - Des résistances pulmonaires
  - De l’importance de l’IT
  - De la fonction du VD
  - De l’existence d’une CIA
  - Du rythme cardiaque


Canal artériel
RVP
POD

Physiopathologie

- Chez le fœtus et le nouveau-né :
  - IT et RVP élevée =>
    - atrésie pulmonaire fonctionnelle ducto-dépendante
    - élévation de la pression atriale droite
  - FOP large ou CIA : la POD dépend du VG
    - Fœtus : décompression de l'OD : meilleur pronostic
    - Nouveau-né : shunt D-G => cyanose importante sans insuffisance cardiaque
  - Pas de CIA et FOP étroit : cœur en série
    - Fœtus : anasarque
    - Nouveau-né : insuffisance cardiaque droite et défaut de précharge du VG

Physiopathologie

- Chez le grand enfant et l'adulte :
  – IT et altération de la contractilité VD
    => Dilatation de l'OD et du VD
  – CIA :
    - Bon VD : shunt G-D
    - VD moyen : shunt G-D au repos et D-G (cyanose) à l'effort
    - Mauvais VD : Shunt D-G et cyanose
  – SIA intact :
    - Pas de cyanose
    - POD et insuffisance cardiaque fonction de l'IT et du VD
  – Arythmies

La présentation clinique

- Fœtus et Nouveau-Né
  - Anasarque
  - Cyanose et Insuffisance cardiaque
- Enfant et Adulte
  - Spectre étendu entre :
    - asymptomatique
    - cardiopathie cyanogène sévère
  - Symptômes principaux :
    - Arythmies
    - Fatigue, Dyspnée, Limitation à l’effort
    - Cyanose
  - Complications :
    - Embolies systémiques
    - Abcès cérébraux
    - Arythmies
    - Mort subite

Présentation clinique

- Cyanose inconstante
- Hépatomégalie inconstante
- Anomalies de l’auscultation :
  - Dédoublement large de B1 et/ou de B2
  - Clicks systoliques
  - B3 et/ou B4
  - Rythme à 3 ou 4 temps
  - Souffle systolique d’IT

ECG

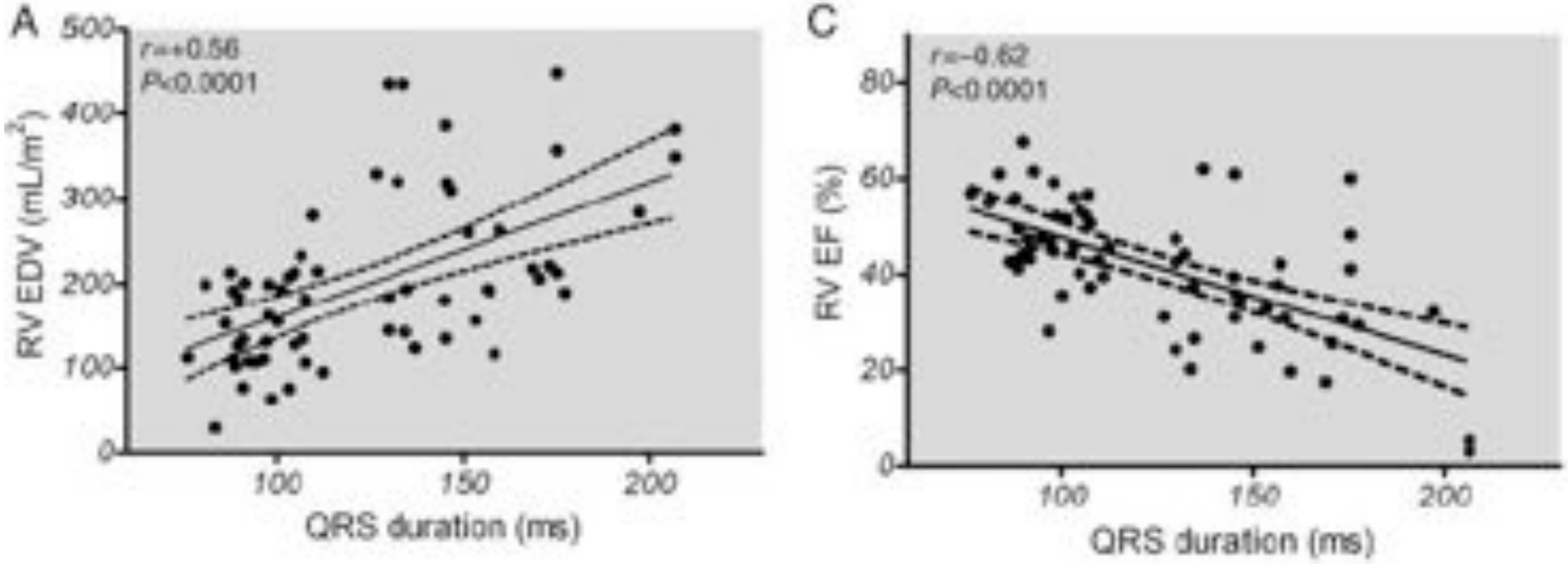
- HAD
- BAV1
- BBD polyphasique
- Ondes Q profondes
  - en D2,D3, aVF
  - De V1 à V4
- Aspect de pré-excitation ventriculaire
- Bas voltage des QRS
- Arythmies supra-ventriculaires ou ventriculaires

HAD, bas-voltage des QRS, BBD

ECG

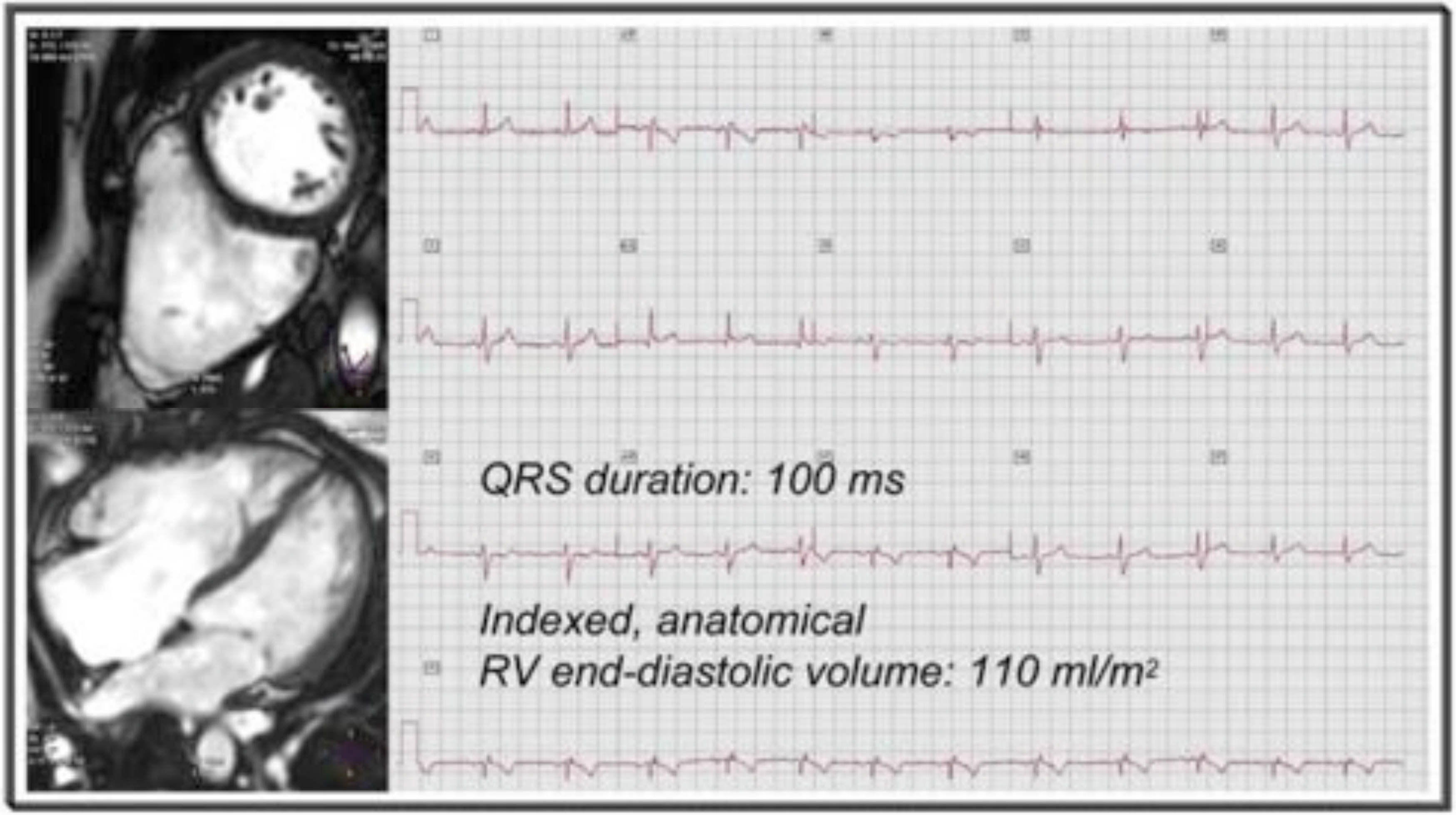
G.E.Assenza *et al.* European Heart Journal (2013) 34, 191–200

ECG : relation entre durée de QRS et fonction VD



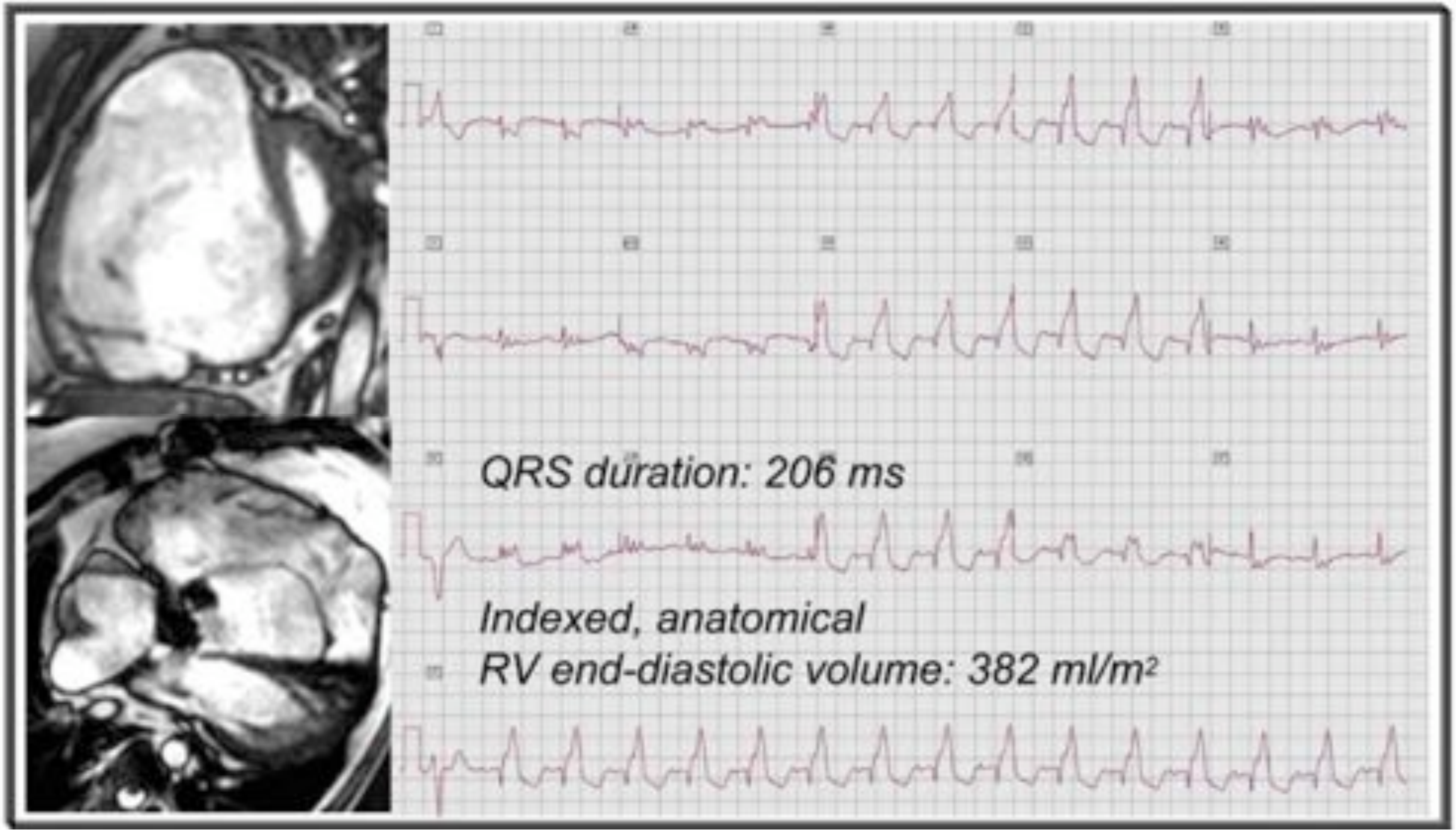
G.E.Assenza *et al.* European Heart Journal (2013) 34, 191–200

ECG

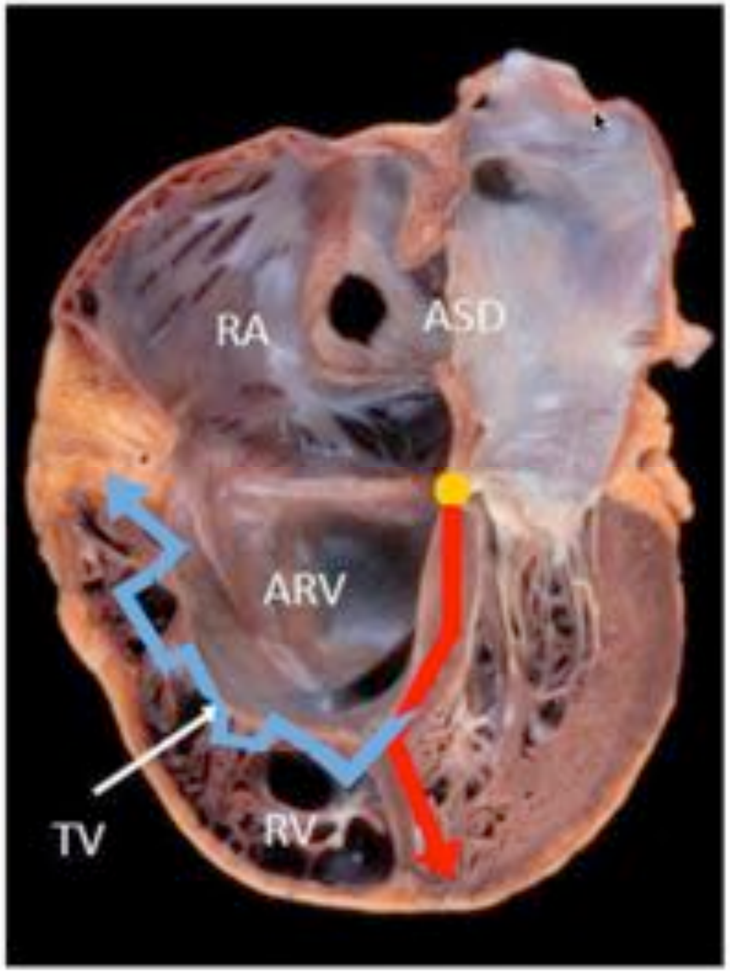


G.E.Assenza *et al.* European Heart Journal (2013) 34, 191–200

ECG



G.E.Assenza *et al.* European Heart Journal (2013) 34, 191–200



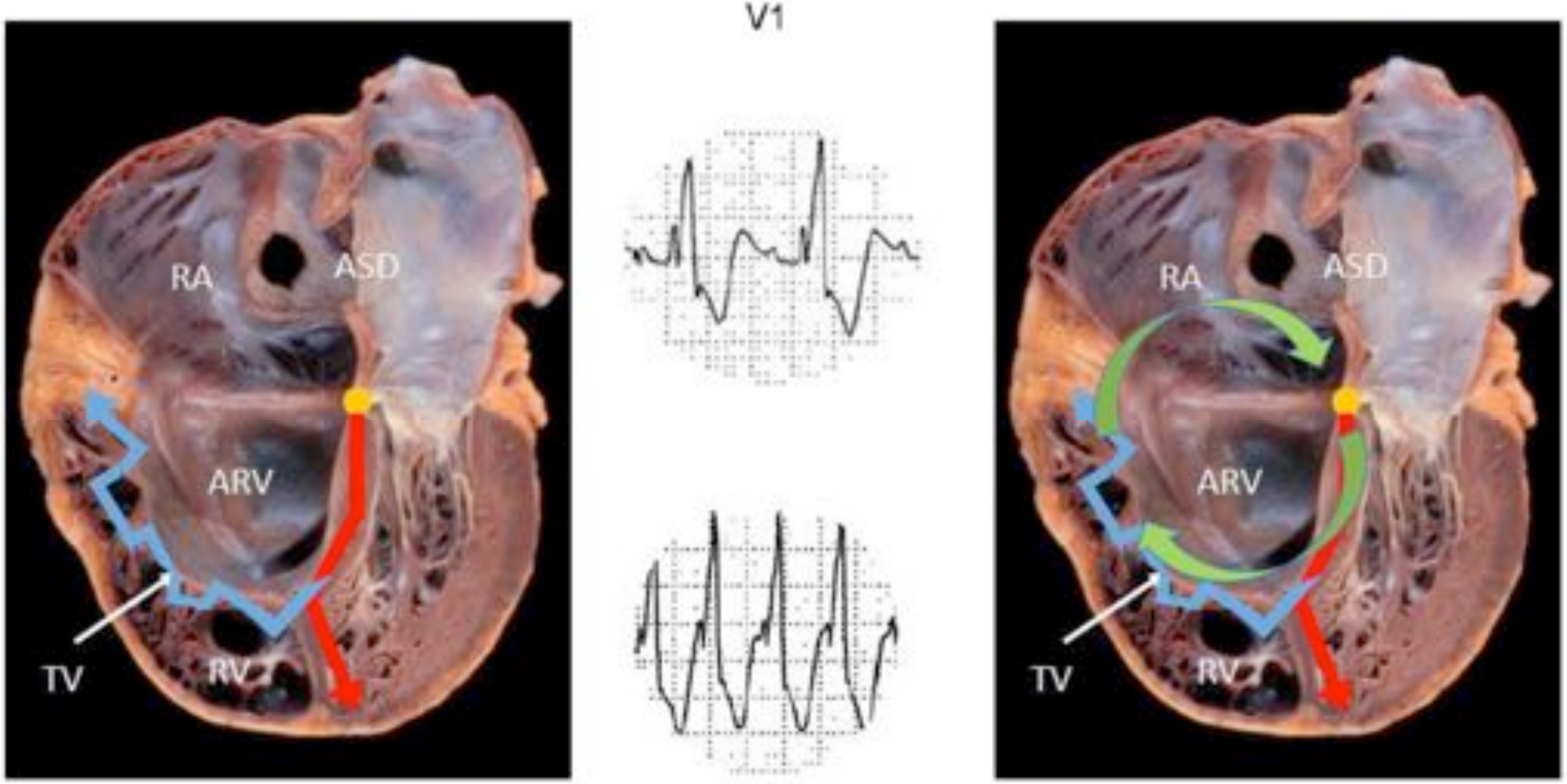
Voie accessoire et Ebstein

- 5-25% WPW
- VA droite et multiple > 50%
- VA maligne
- Taux de succès RF ≈ 80%
- Taux de récurrence ≈ 20%
- Complication 3-5%

Reich et al. JCE 1998

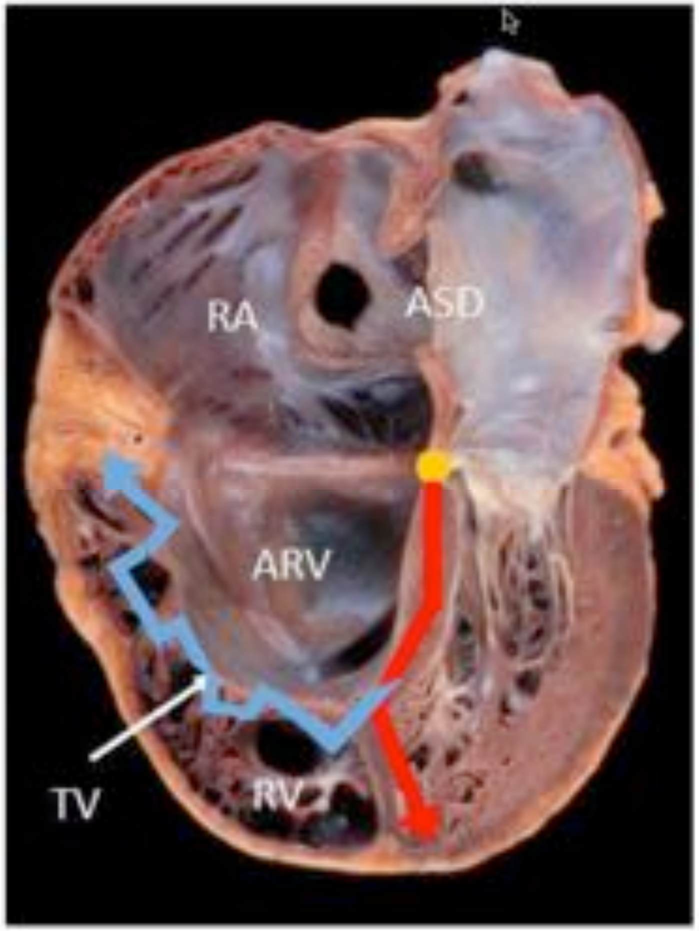
diapo de Alice Maltret

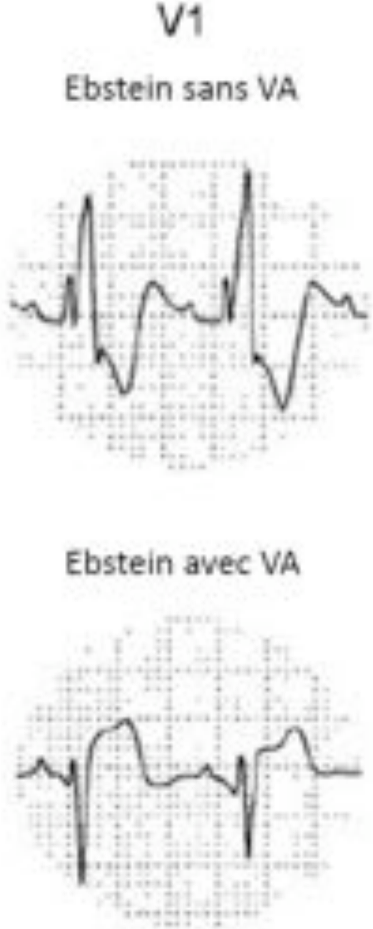
Ebstein sans voie accessoire



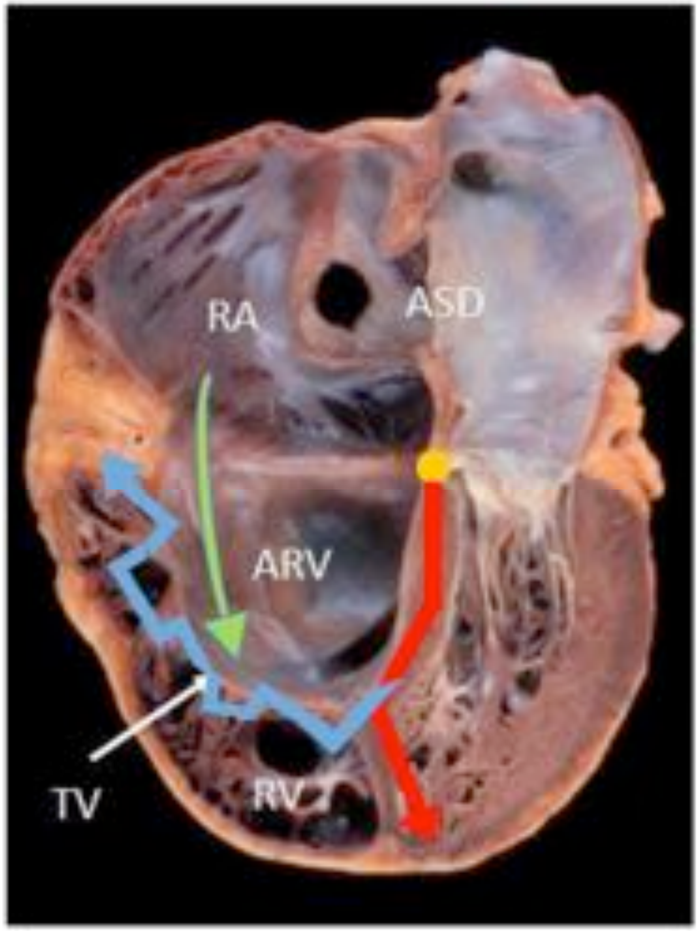
diapo de Alice Maltret

Sans voie accessoire





Avec voie accessoire



diapo de Alice Maltret

Radiographie thoracique

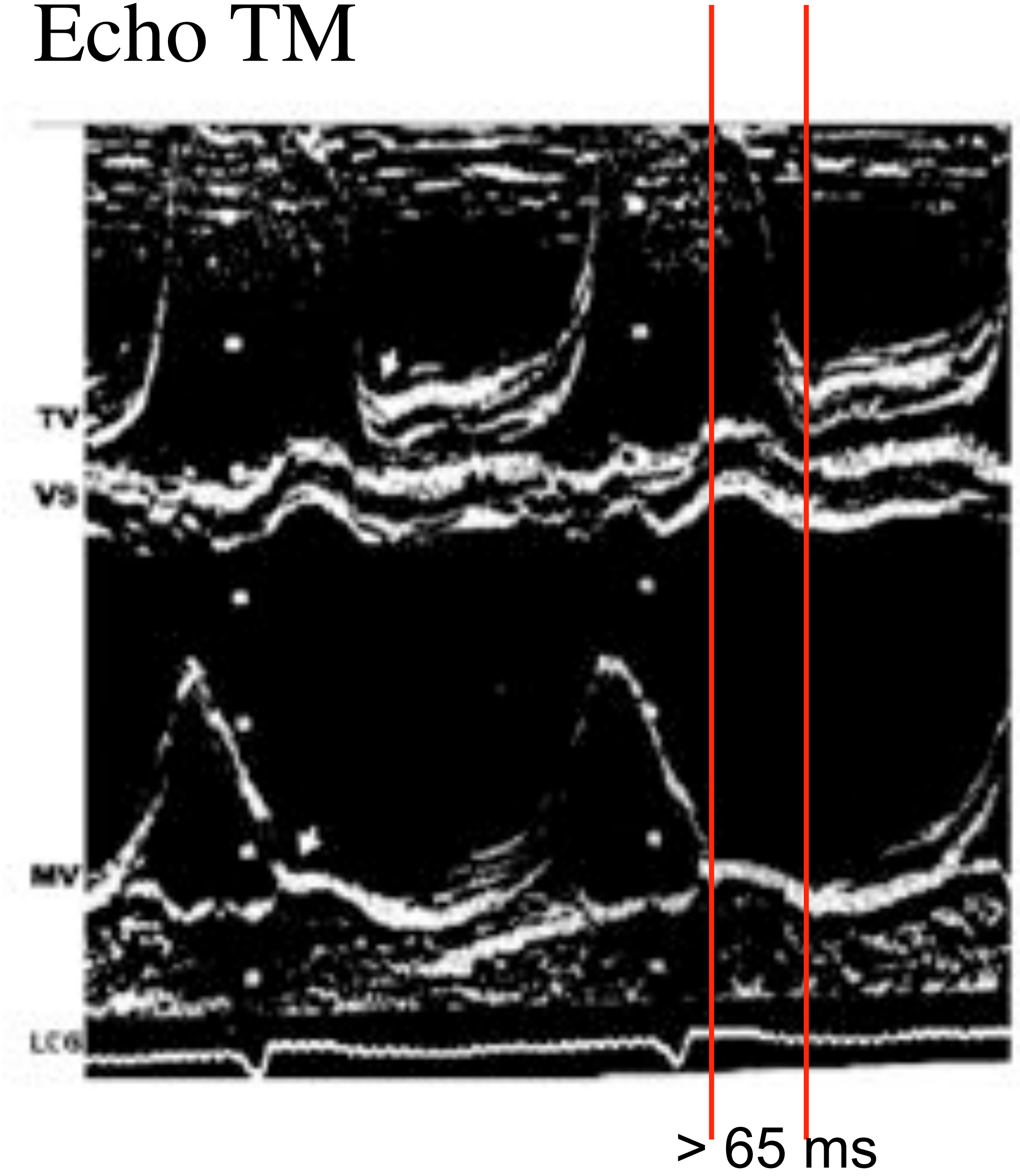
RCT > 0,65 : mauvais pronostic

Echocardiographie

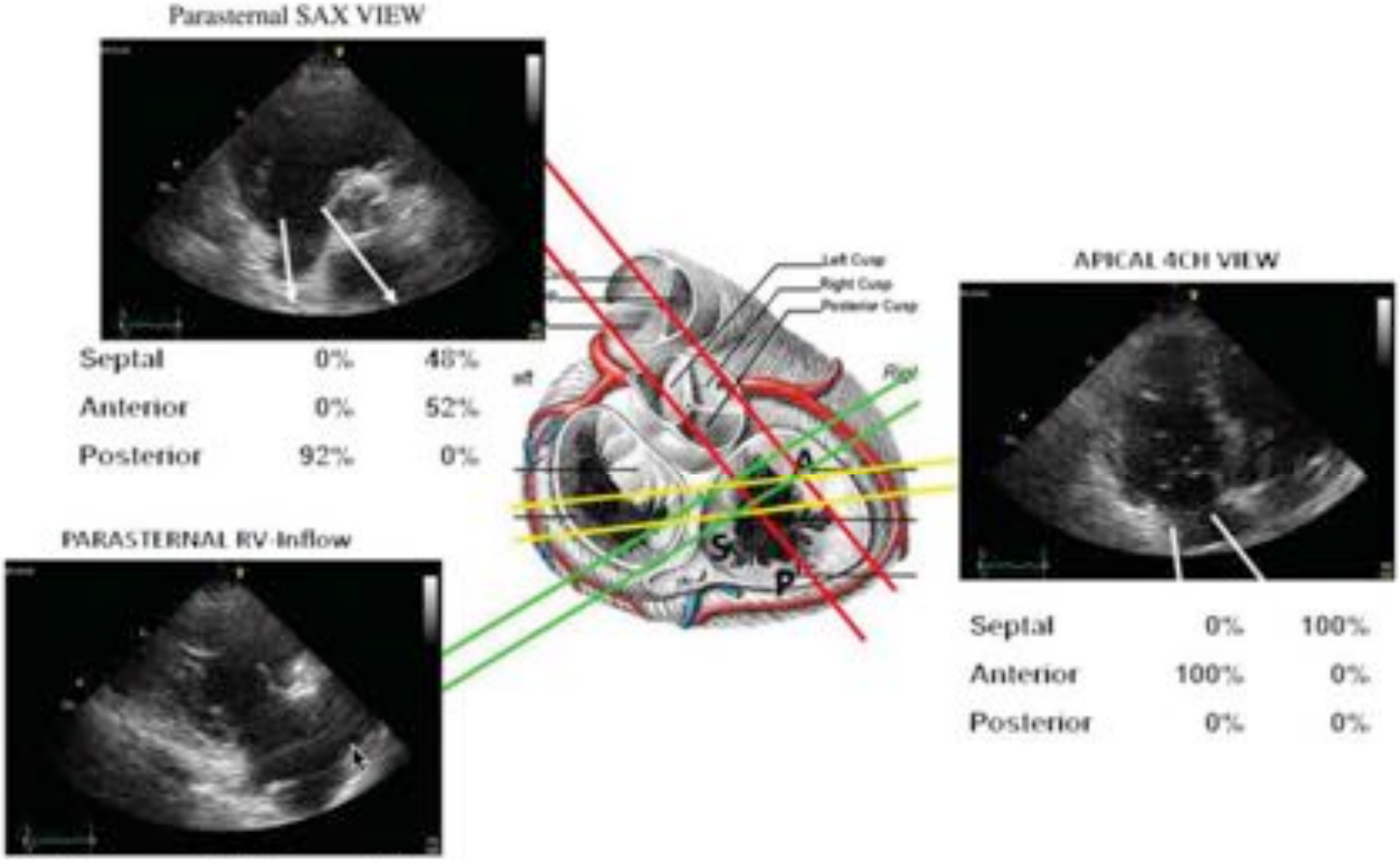
- Tricuspide :
  - Déplacement apical du feuillet septal de la tricuspide ≥ 8 mm/m^2
  - Attaches et mobilité des feuillets septal et postérieur
  - Dimension du feuillet antérieur
  - Abondance de l'IT
- VD :
  - Dimension de la portion atrialisée
  - Dimensions et fonction du VD résiduel
- Voie d'éjection droite et artères pulmonaires
- Septum interatrial
- Cavités gauches et fonctions VG
- Anomalies associées

Booker et Nanda. Echocardiography 2015;S32:177–S188

Echo TM



- Enregistrement simultané de Tric et Mit facile
- Amplitude Tric > Mit
- Retard de fermeture de Tric > 65 ms



d'après Anwar AM, *et al*. Assessment of normal tricuspid valve anatomy in adults by realtime three-dimensional echocardiography. Int J Cardiovasc Imaging. 2007;23:717–24.

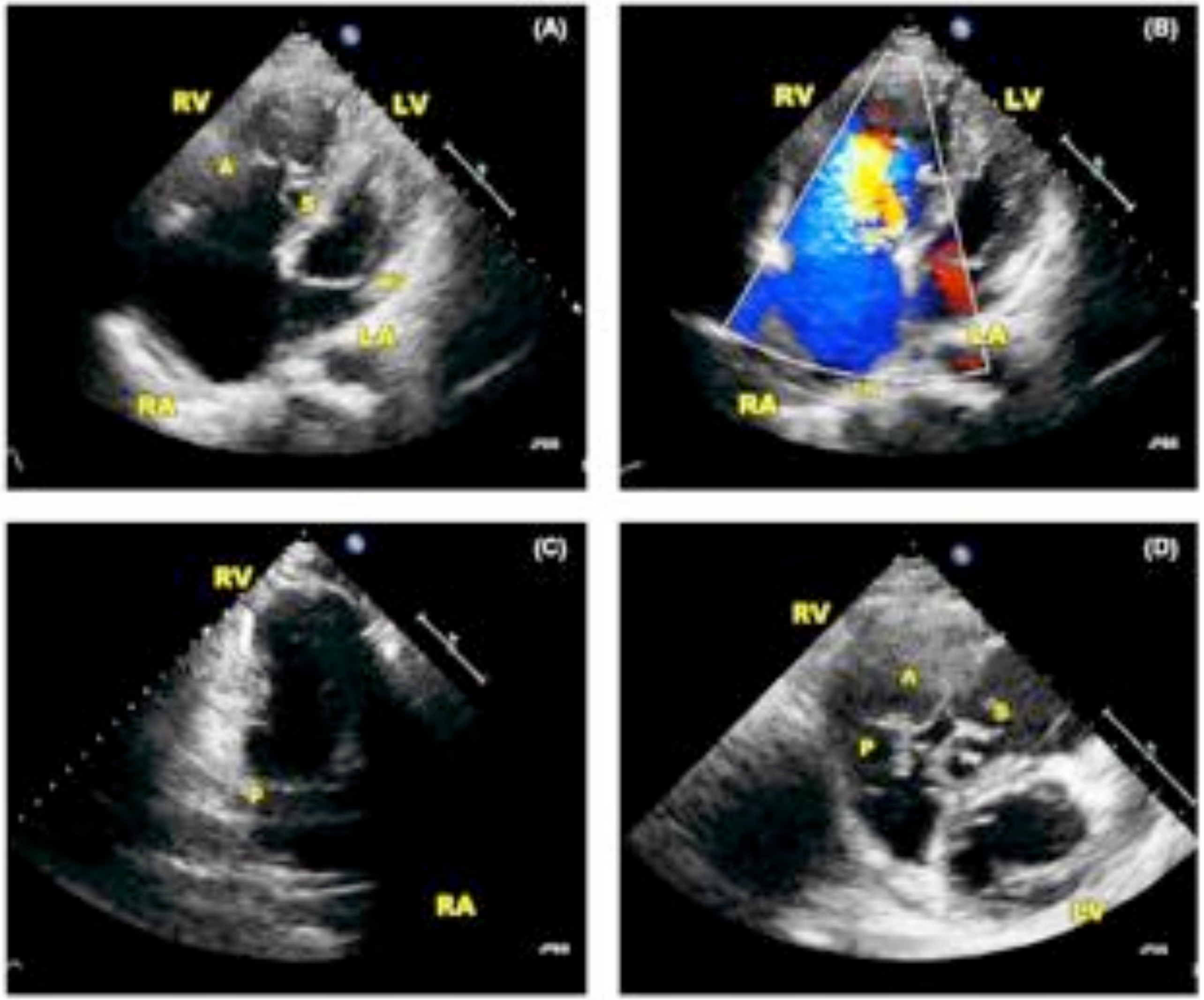
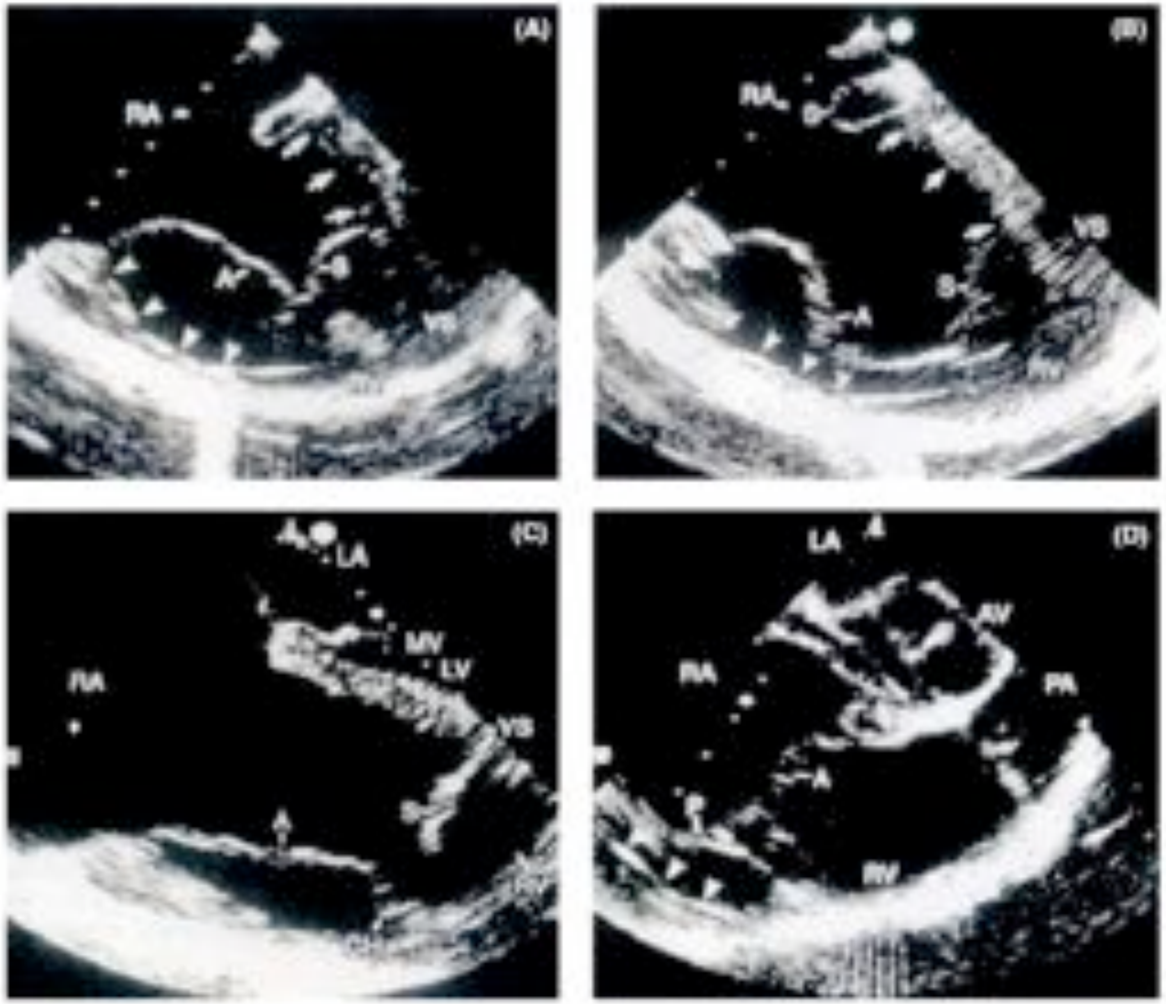


Figure 2. Ebstein's anomaly. Two-dimensional transthoracic echocardiography. Apparent apical displacement of the attachment of septal leaflet (S) of the tricuspid valve (TV) as compared to the mitral valve (MV) is seen in apical four-chamber view (**A**). This displacement is related to tethering of the septal leaflet to the inter-ventricular septum resulting in a bubble-like appearance. The septal leaflet in this patient and in almost all other patients with Ebstein's anomaly actually originates normally from the atrioventricular junction. **B.** Shows severe tricuspid regurgitation (TR). **C.** Posterior (P) leaflet of the TV can be seen in right ventricular two-chamber view. **D.** Left ventricular short-axis view at the level of mitral valve demonstrates all the three leaflets of the tricuspid valve. Movie clips S2A, S2B, S2C, and S2D. A = anterior tricuspid leaflet. (Reproduced with permission from Kapoor PM, Bhagatwala K, Karia N, Nanda NC: Echocardiographic Examination of the Tricuspid Valve. In: Nanda NC (Ed): *Comprehensive Textbook of Echocardiography*. New Delhi, India: Jaypee Brothers Medical Publishers (P) Ltd, 2013, pp. 989–1035.)

Booker et Nanda. Echocardiography 2015;S32:177–S188

ETO



Anwar AM, *et al*. Assessment of normal tricuspid valve anatomy in adults by realtime three-dimensional echocardiography.
Int J Cardiovasc Imaging. 2007;23:717–24.

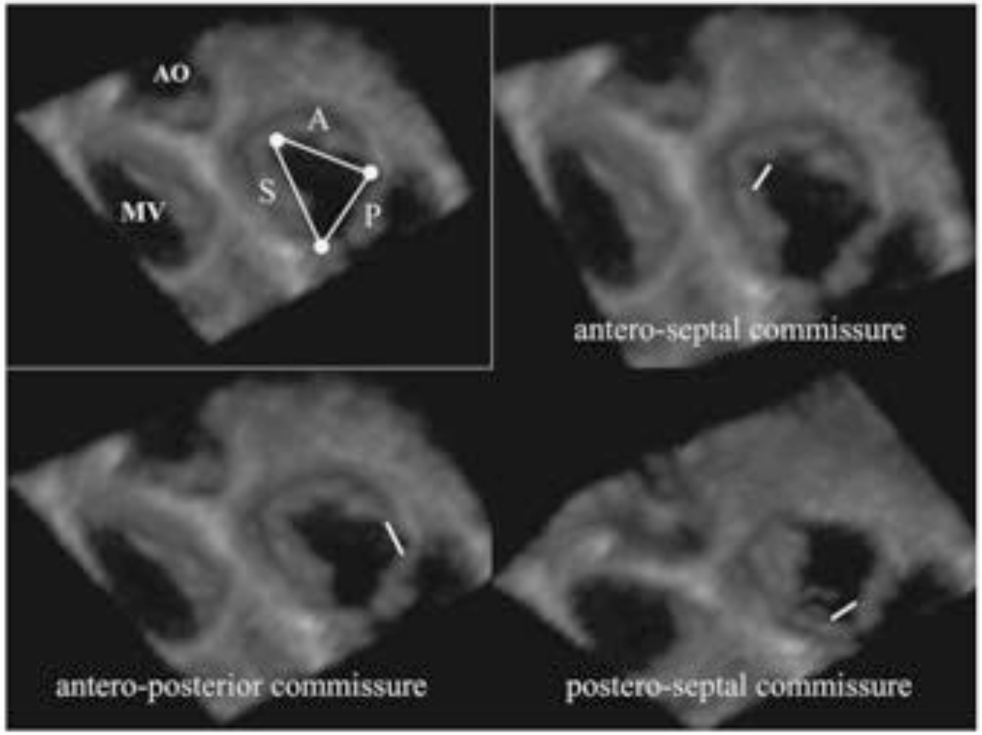


Table 2 Normal (absolute and index) values of tricuspid valve annulus (diameter and area), Tricuspid valve area and the width of the 3 commissures

Parameter	Absolute value	Index value
Tricuspid annulus diameter	4.0 ± 0.7 (cm)	2.2 ± 0.4 (cm/m^2)
Tricuspid annulus area	10.0 ± 2.9 (cm^2)	5.5 ± 1.6 (cm^2/m^2)
Tricuspid valve area	4.8 ± 1.6 (cm^2)	2.7 ± 0.9 (cm^2/m^2)
Antero-septal commissure	5.4 ± 1.5 (mm)	2.9 ± 0.8 (mm/m^2)
Postero-septal commissure	5.2 ± 1.5 (mm)	2.9 ± 0.7 (mm/m^2)
Antero-posterior commissure	5.1 ± 1.1 (mm)	2.8 ± 0.6 (mm/m^2)

L.P.Badano *et al.* Evaluation of the tricuspid valve morphology and function by transthoracic real-time three-dimensional echocardiography.
European Journal of Echocardiography (2009) 10, 477–484

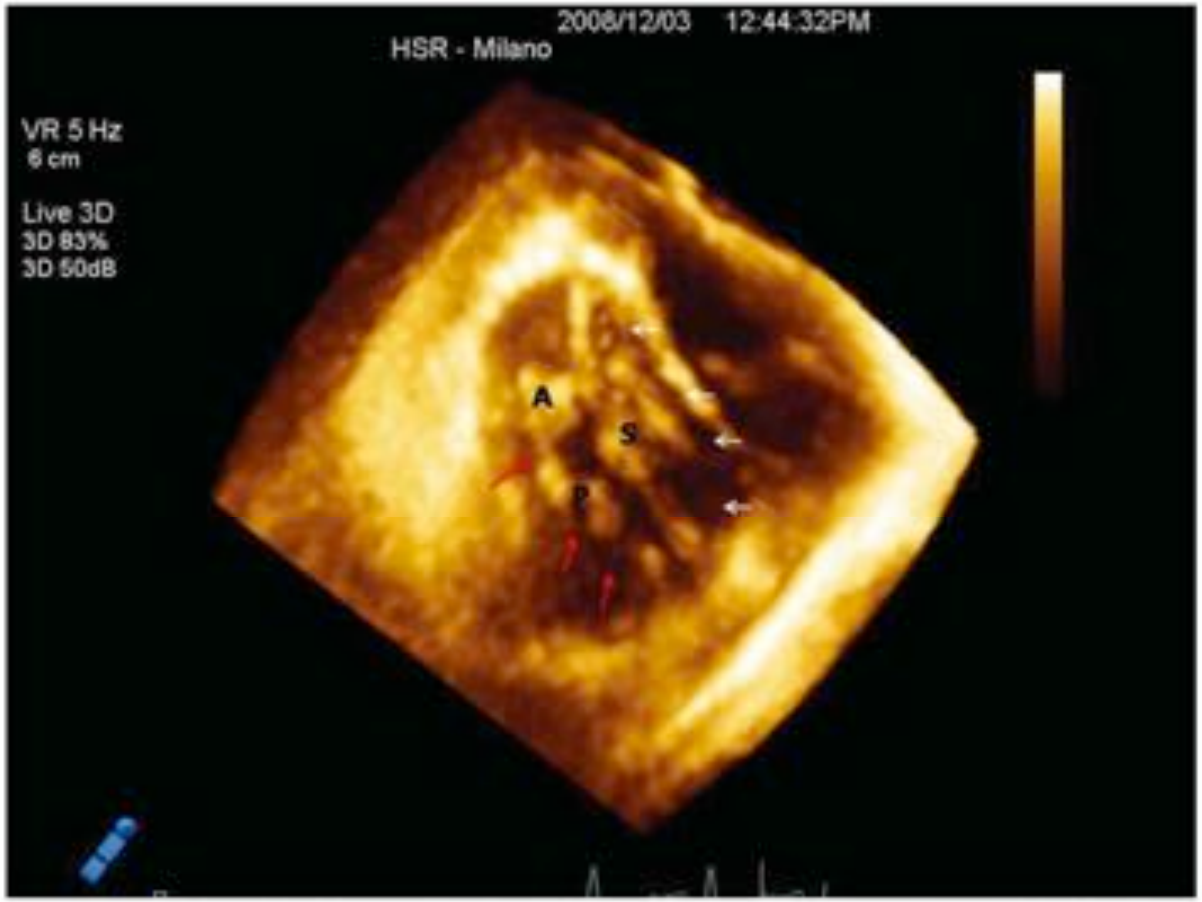
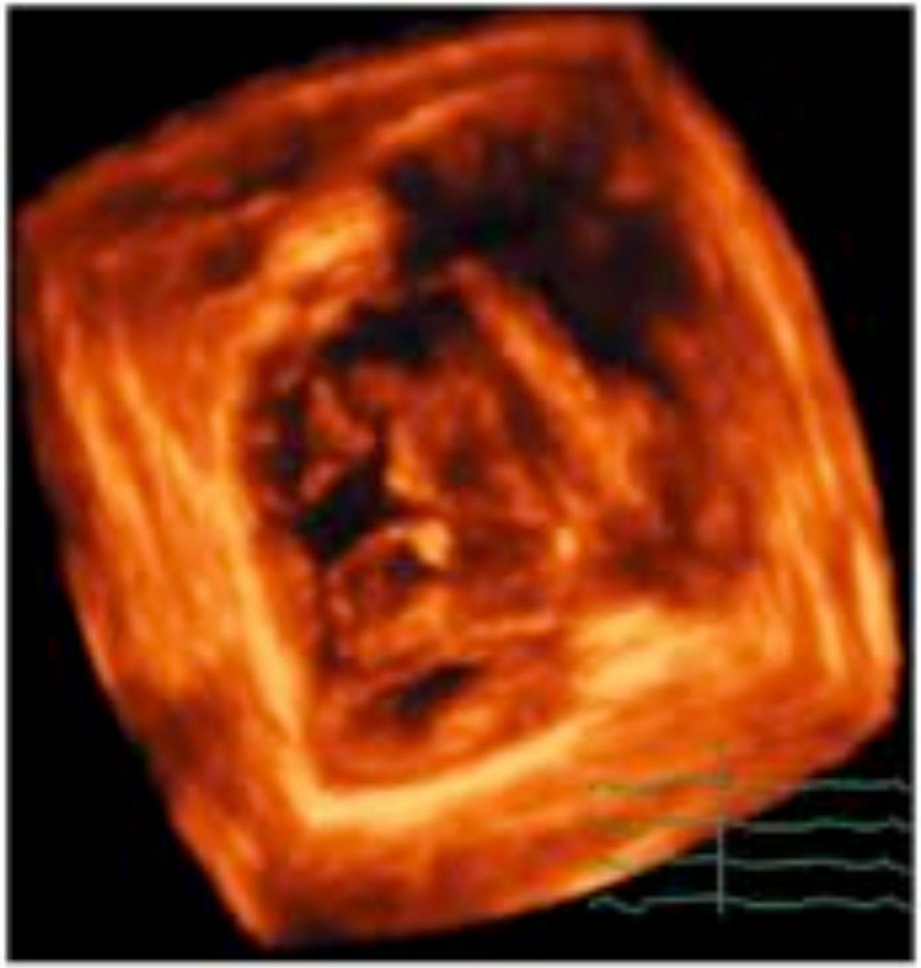
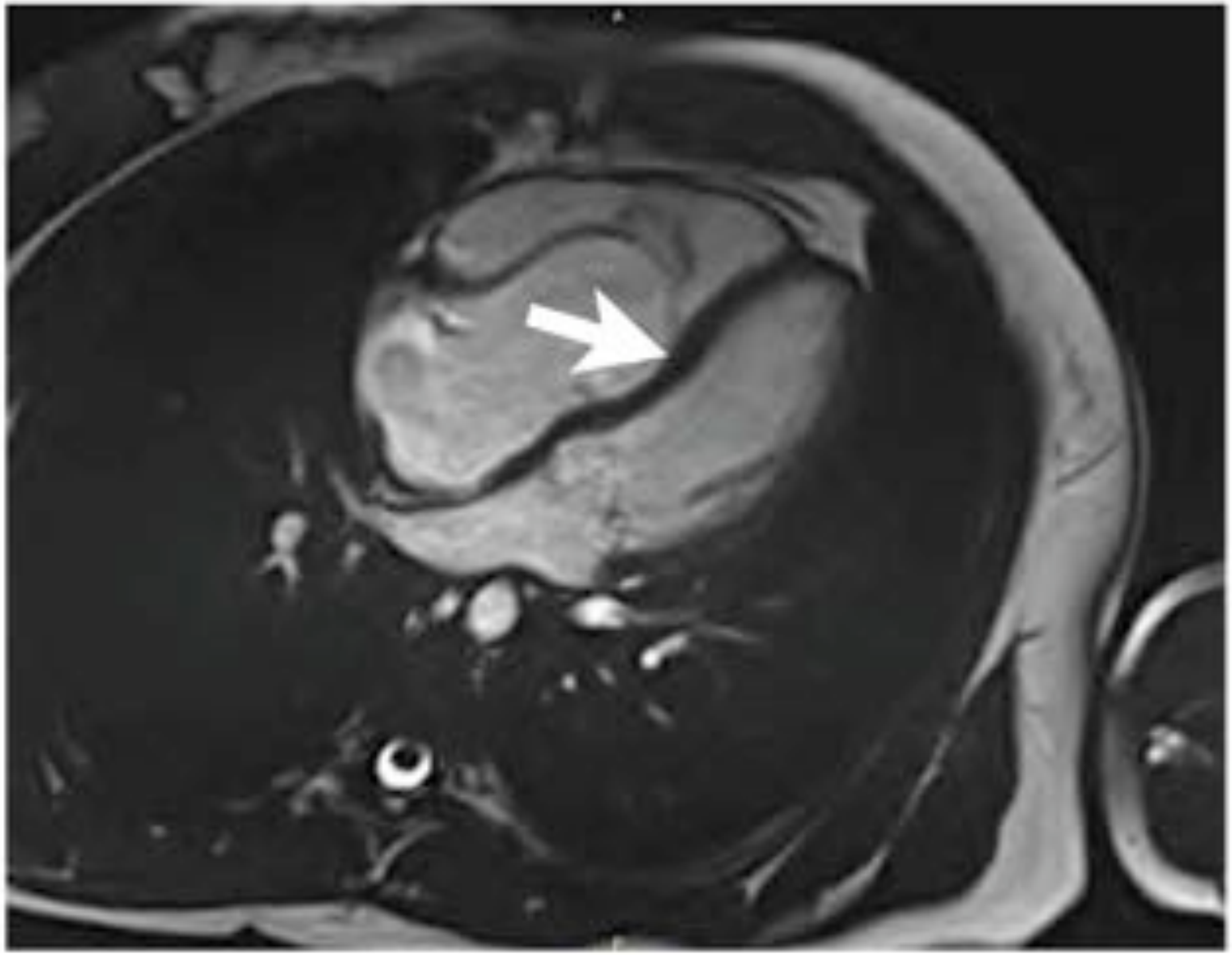


Figure 8 Ebstein's anomaly seen from the right ventricle. The deformed leaflets are clearly visible as well as the characteristic bubble-like aspect of septal (S, with arrows) and posterior (P, red arrows) leaflets as effect of tethering of the leaflet where the chordae are attached. The anterior leaflet (A) surface area can be visualized and the regions of ineffective leaflet coaptation easily identified.



Ph. Acar

IRM



Volumes

Fonction VD

Fonction VG

Le pronostic est fonction :

- du degré de dysfonction tricuspide
- du degré d’atrialisation du VD
- du volume et de la fonction du VD résiduel
- de la fonction du ventricule systémique
- des malformations associées
- des arythmies

Techniques chirurgicales

- Nouveau-né :
  - Blalock-Taussig-Thomas
  - Technique de Starnes (1991)

- Grand enfant et adulte :
  - Simple fermeture de CIA
  - Chirurgie tricuspide
    - Danielson (1979)
    - Carpentier et Chauvaud (1988)
    - Da Silva : Réparation en cône (2011)
    - Remplacement valvulaire tricuspide (1962) : à éviter

- Dérivation cavo-pulmonaire partielle : 1,5 ventricule
- Circulation de Fontan : DCPT

Technique de Starnes

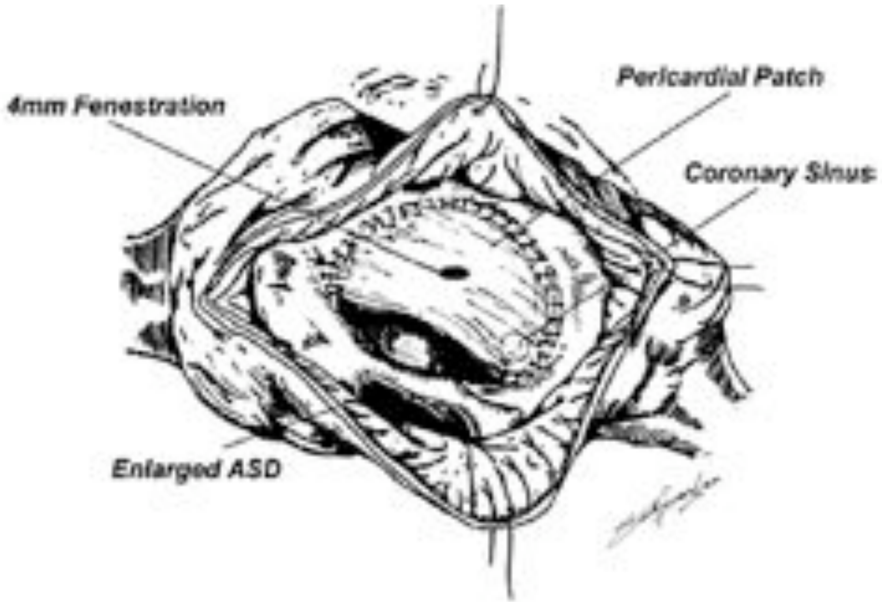


Figure 2 In the Starnes operation, the tricuspid valve is closed with a patch using autologous pericardium, which should be fenestrated unless there is antegrade egress to decompress the right ventricle. An atrial septectomy is performed, and a systemic to pulmonary shunt provides pulmonary blood flow (not shown). (Reprinted with permission from Knott-Craig and Goldenberg.[15])

- Fermeture de la tricuspide par un patch de péricarde fenestré

- CIA chirurgicale

- BTT

Technique de Danielson (1979)

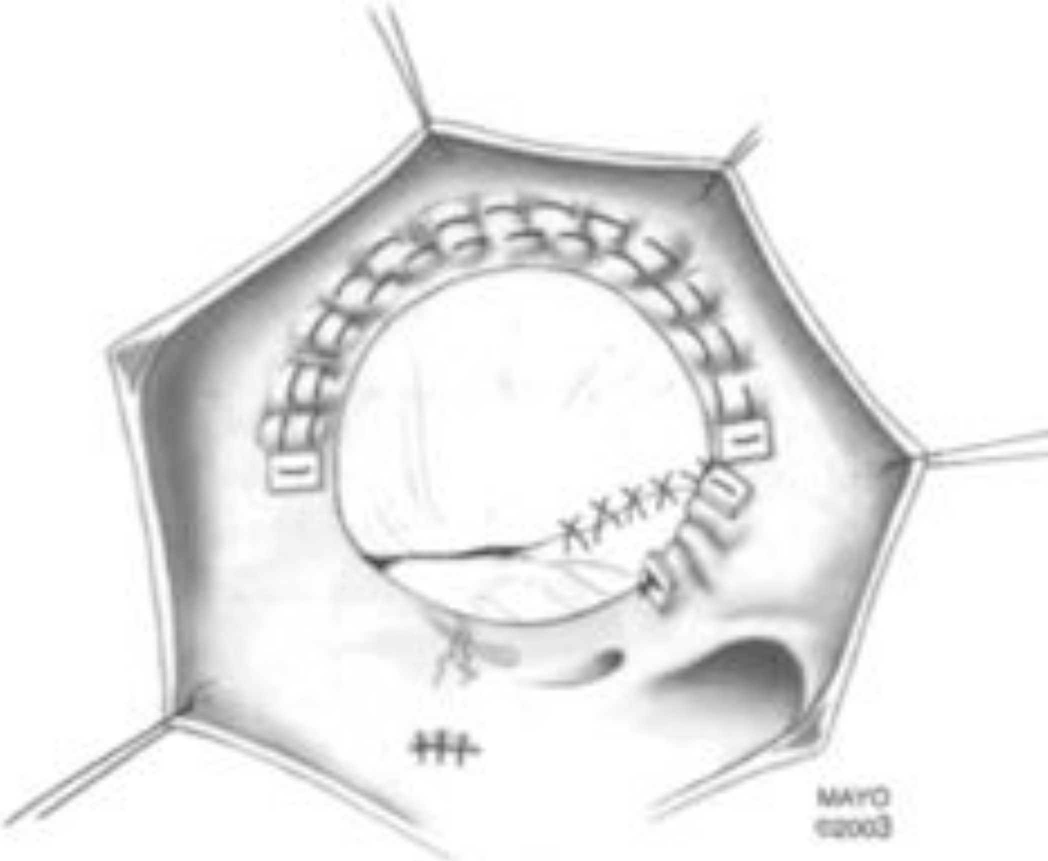
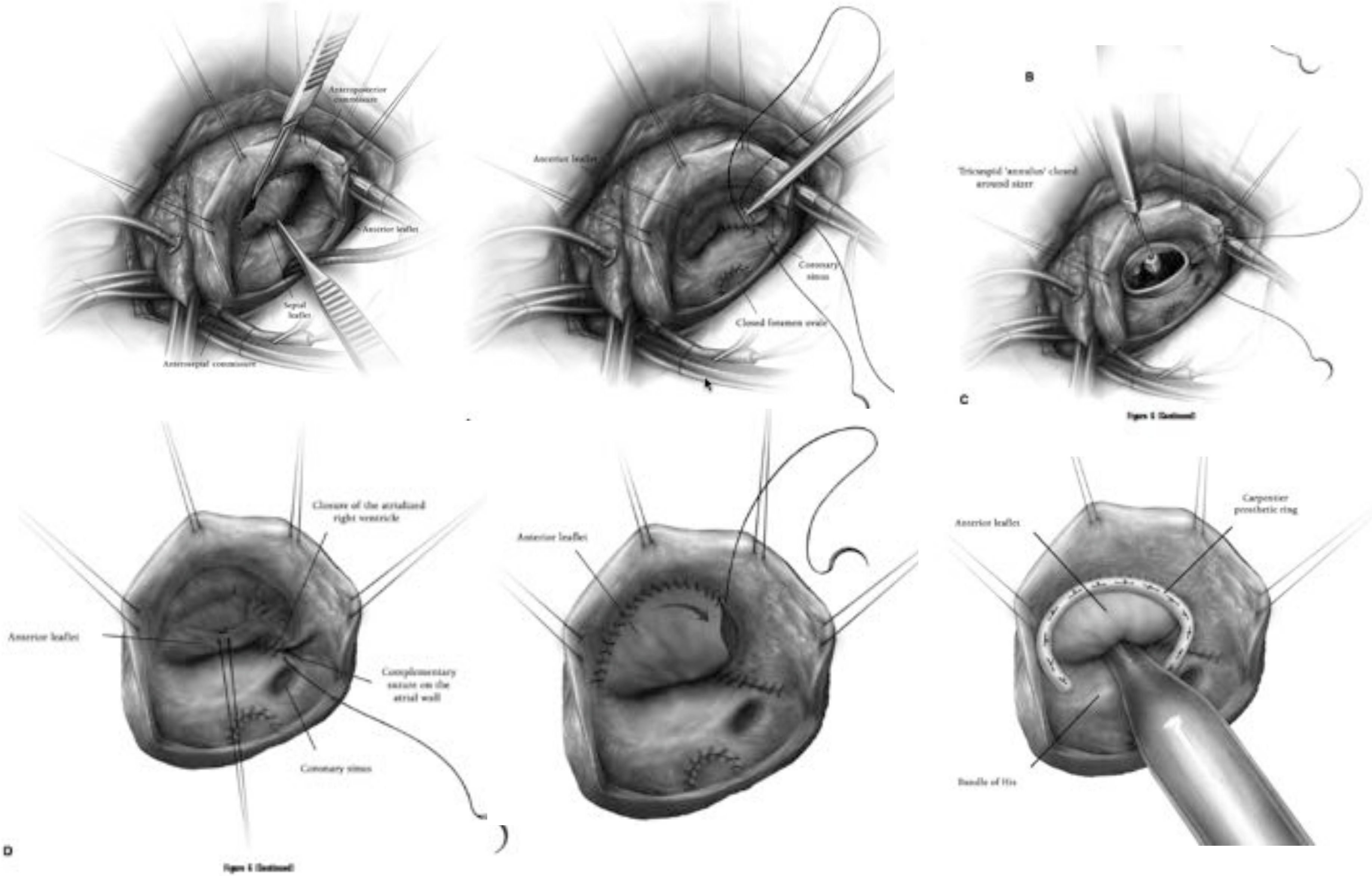


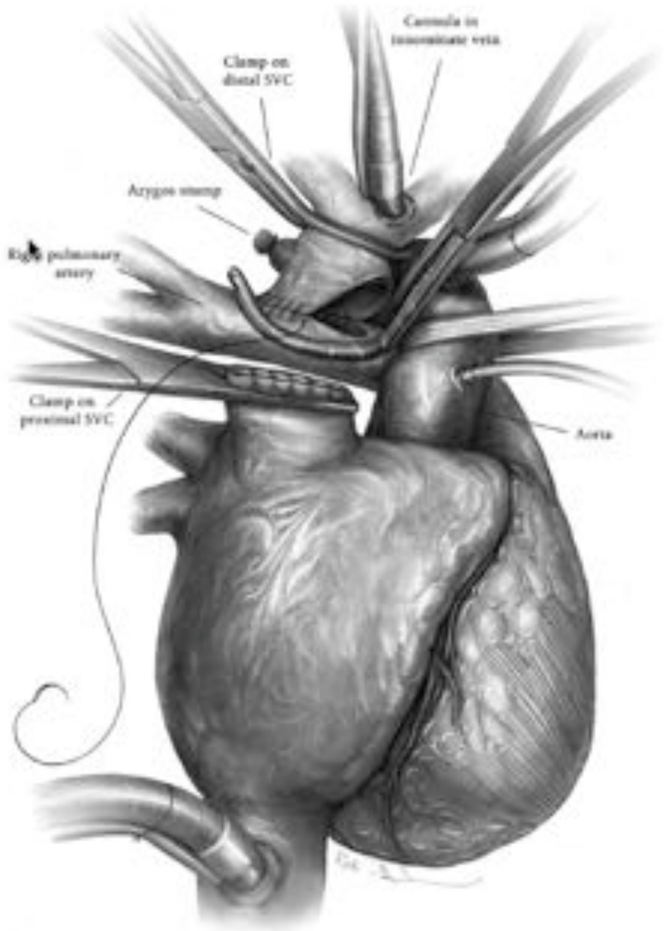
Figure 3: The Danielson Technique for Tricuspid Valve Repair in Ebstein Anomaly.
The anterior leaflet functions as a monocuspid valve.

Technique de Carpentier et Chauvaud



S. Chauvaud. Repair of Ebstein's anomaly. Oper Tech Thorac Cardiovasc Surg. 2004,9:194-207

Dérivation cavo-bipulmonaire (DCPP)



- Indications :
  - IT sévère
  - Mauvais VD
  - POD élevée
  - POG basse
  - VG mal empli

S. Chauvaud. Repair of Ebstein's anomaly. Oper Tech Thorac Cardiovasc Surg. 2004,9:194-207

José Pedro da Silva *et al.* , The cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and midterm results

J Thorac Cardiovasc Surg 2007;133:215-23

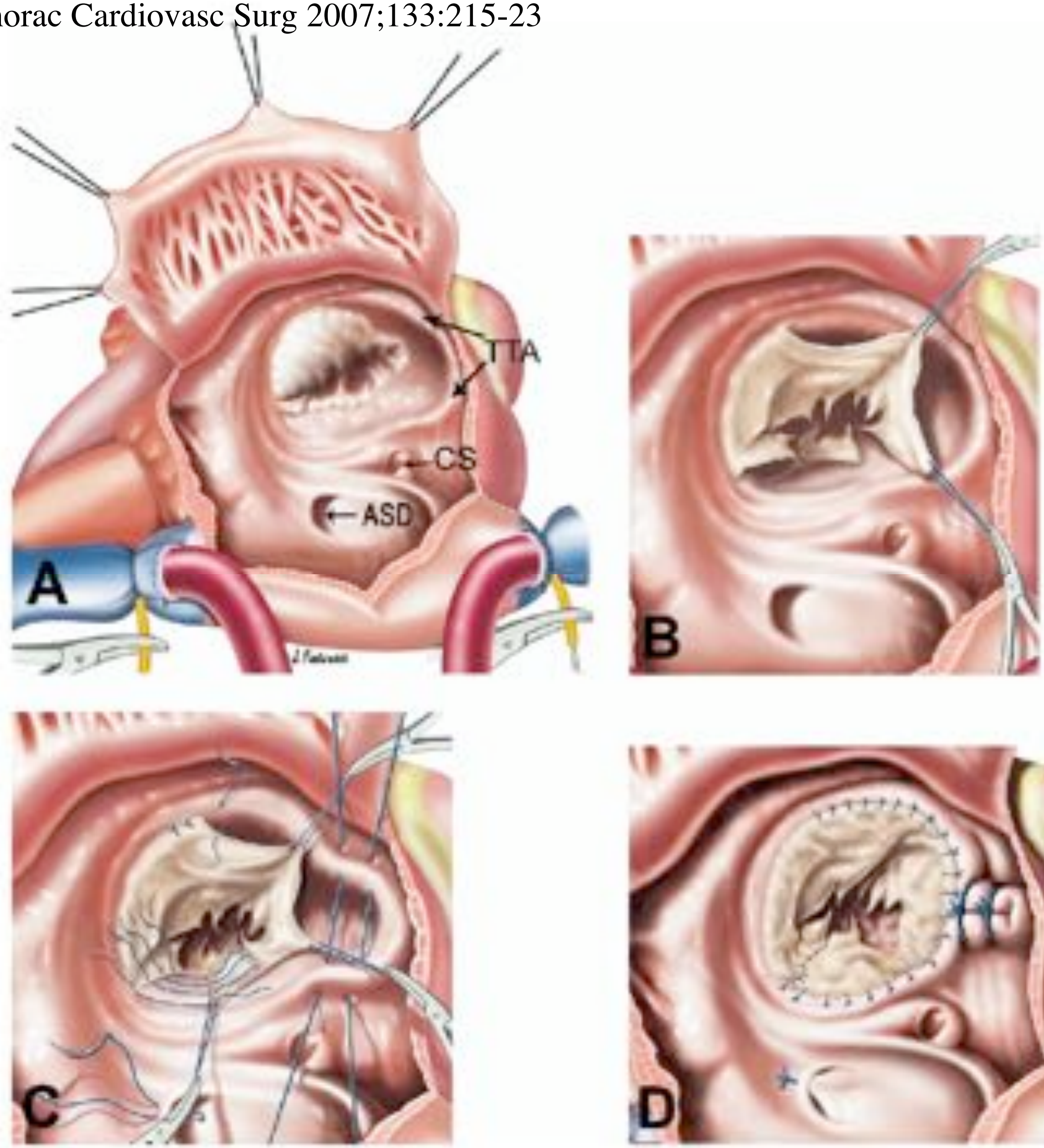


Figure 1. Operative steps for Ebstein's anomaly repair. A, Opened right atrium showing displacement of the tricuspid valve. *TTA*, True tricuspid annulus; *ASD*, atrial septal defect; *CS*, coronary sinus. B, Detached part of the anterior and posterior leaflet forming a single piece. C, Clockwise rotation of the posterior leaflet edge to be sutured to the anterior leaflet septal edge and plication of the true tricuspid annulus. D, Complete valve attachment to the true tricuspid annulus and valved closure of the atrial septal defect.

José Pedro da Silva *et al.* , The cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and midterm results

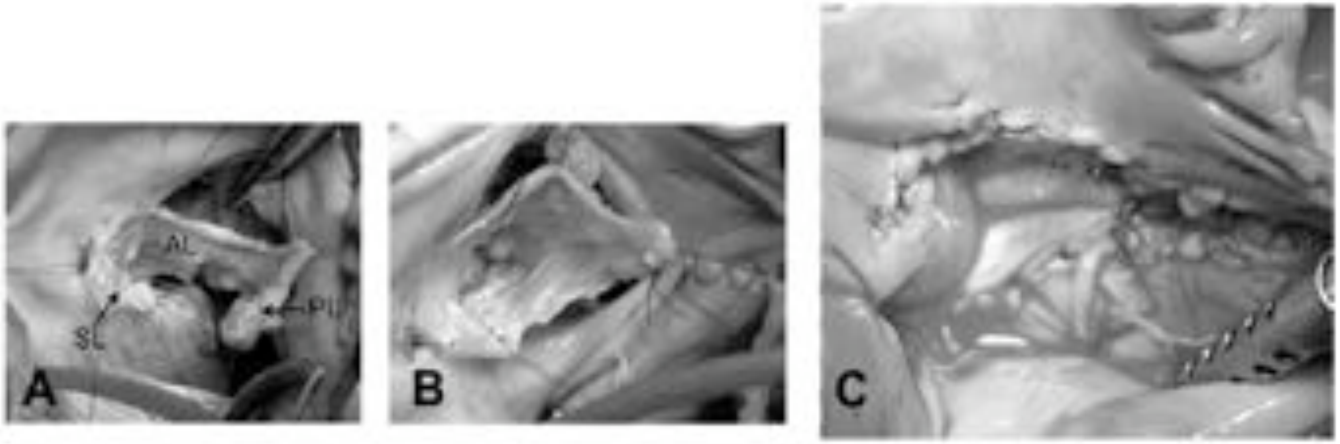


Figure 2. Surgical sequence of tricuspid valve repair with incorporation of the septal leaflet. A, Detached septal and posterior leaflets and part of the anterior leaflet of the tricuspid valve. *AL*, Anterior leaflet; *PL*, posterior leaflet; *SL*, septal leaflet. B, Septal leaflet sutured to the posterior and anterior leaflets in a cone shape and the plicated annulus. C, Final aspect after filling the right ventricle with saline.

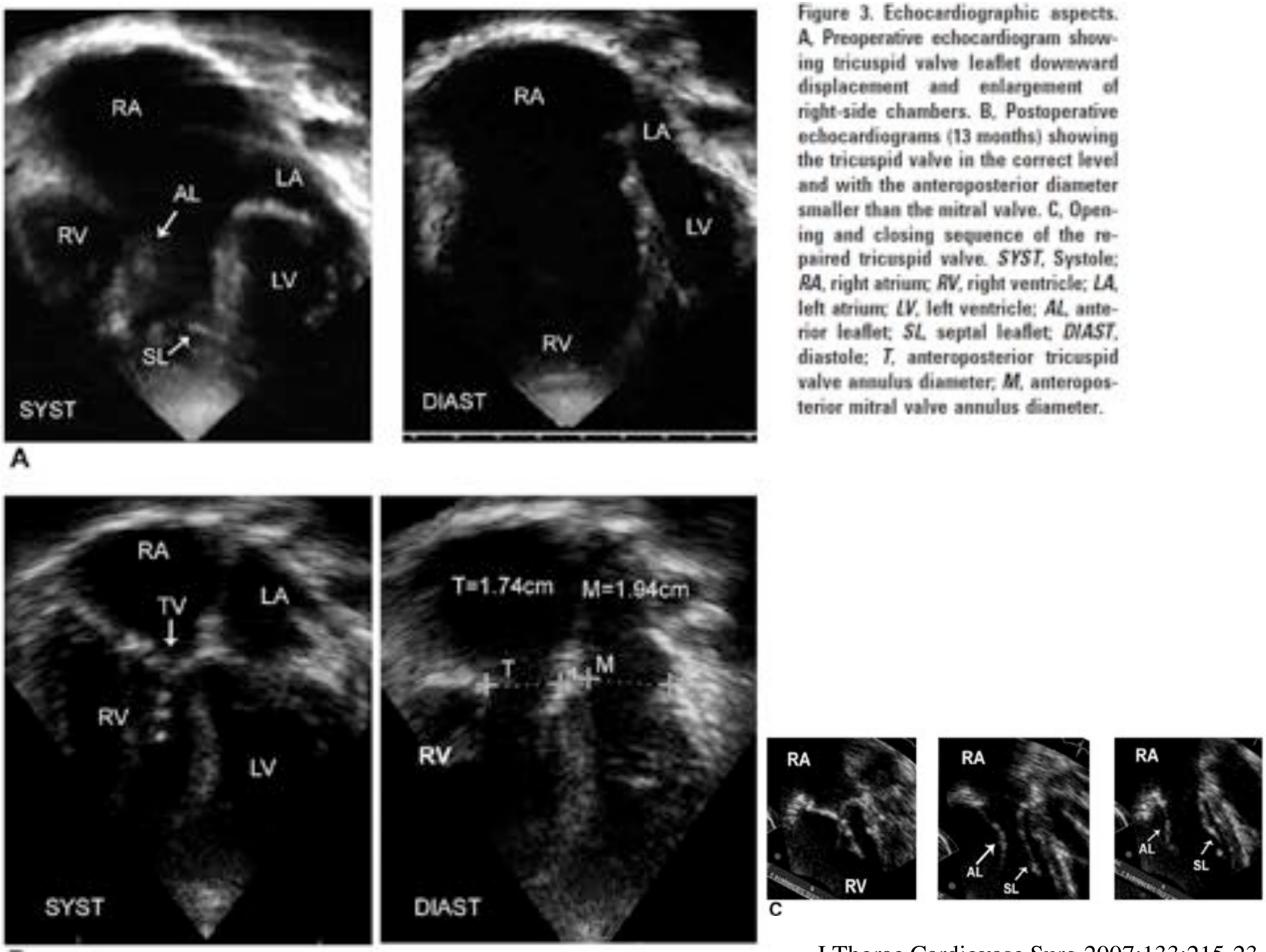


Figure 3. Echocardiographic aspects. A, Preoperative echocardiogram showing tricuspid valve leaflet downward displacement and enlargement of right-side chambers. B, Postoperative echocardiograms (13 months) showing the tricuspid valve in the correct level and with the anteroposterior diameter smaller than the mitral valve. C, Opening and closing sequence of the repaired tricuspid valve. *SYST*, Systole; *RA*, right atrium; *RV*, right ventricle; *LA*, left atrium; *LV*, left ventricle; *AL*, anterior leaflet; *SL*, septal leaflet; *DIAST*, diastole; *T*, anteroposterior tricuspid valve annulus diameter; *M*, anteroposterior mitral valve annulus diameter.

J Thorac Cardiovasc Surg 2007;133:215-23

TABLE 2. Early postoperative echocardiographic results and recent functional class of heart failure compared with preoperative data

	Preoperative	Postoperative	*P* value
Tricuspid insufficiency, n			<.0001
Grade 1	0	31	
Grade 2	1	5	
Grade 3	15	3	
Grade 4	24	0	
TTA AP diameter, mm (mean)	34.3 ± 6.5	15.1 ± 3.7	<.0001
Functional class (NYHA), n			<.0001
Class I	5	32	
Class II	8	5	
Class III	25	0	
Class IV	2	1	

NYHA, New York Heart Association; *TTA*, true tricuspid annulus; *AP*, anteroposterior.

José Pedro da Silva *et al.* , Cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and long-term results. Arq Bras Cardiol 2011; 97(3) : 199-208

Survie

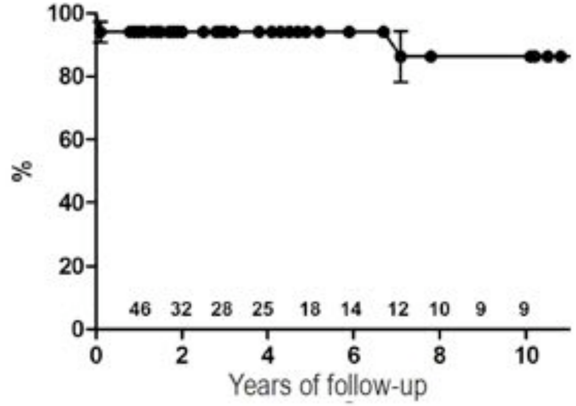


Figure 1 – *Actuarial survival curve estimated by Kaplan-Meier shows survival of 94.1 ± 3.2% from 1 month to 6 years of follow-up and 86.2 ± 8% after 7 years. are presented with standard error and the numbers indicate patients at risk in each period.*

IT

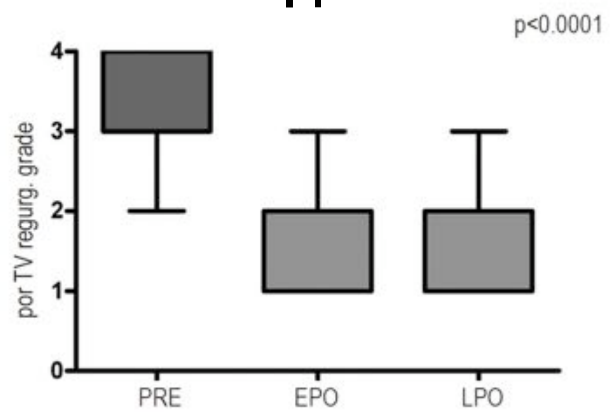


Figure 2 – *Tricuspid valve regurgitation: Comparison between the pre-operative (PRE), early postoperative (EPO) and long-term postoperative (LPO) periods. Mean ± 95% confidence interval. EPO – early postoperative, LPO - long-term postoperative, TV - tricuspid valve.*

VD

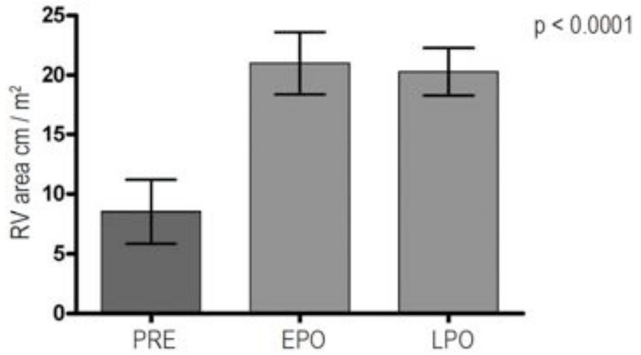


Figure 3 – *Functional area of the right ventricle, indexed by body surface area in cm2/m2: Comparison between measurements in the preoperative, early postoperative and long-term post-operative periods. Mean ± 95% confidence interval. RV - right ventricle, EPO – early postoperative, LPO - long-term postoperative.*

OD

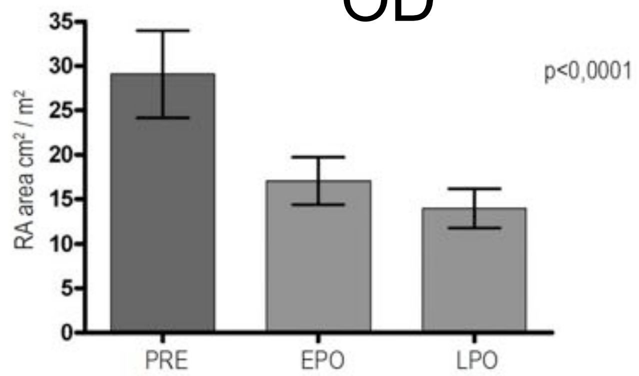


Figure 4 – *Right atrium area indexed by body surface area in cm2/m2: Comparison of measurements in the preoperative, early postoperative and long-term post-operative periods. Mean ± 95% confidence interval. RA - right atrium, EPO – early postoperative, LPO- long-term postoperative.*

K.M.Veen *et al*. Reconstructive surgery for Ebstein anomaly : three decades of experience.

European Journal of Cardio-Thoracic Surgery, 05 february 2019,
https://doi.org/10.1093/ejcts/ezz022

- 1988 - 2016
- 36 patients
- Age : 25,4±15,9 ans
- Sex ratio : 0,36
- Carpentier-Chauvaud : 21 (58%)
- Cone repair : 15 (42%)
- Décès : 1 précoce (3%), 2 tardifs
- Réinterventions : 6 patients
- Survie à 25 ans : 97±3%
- Sans réintervention à 25 ans : 80±8%
- Follow-up : probabilité de NYHA 3-4 < 10%

Key question

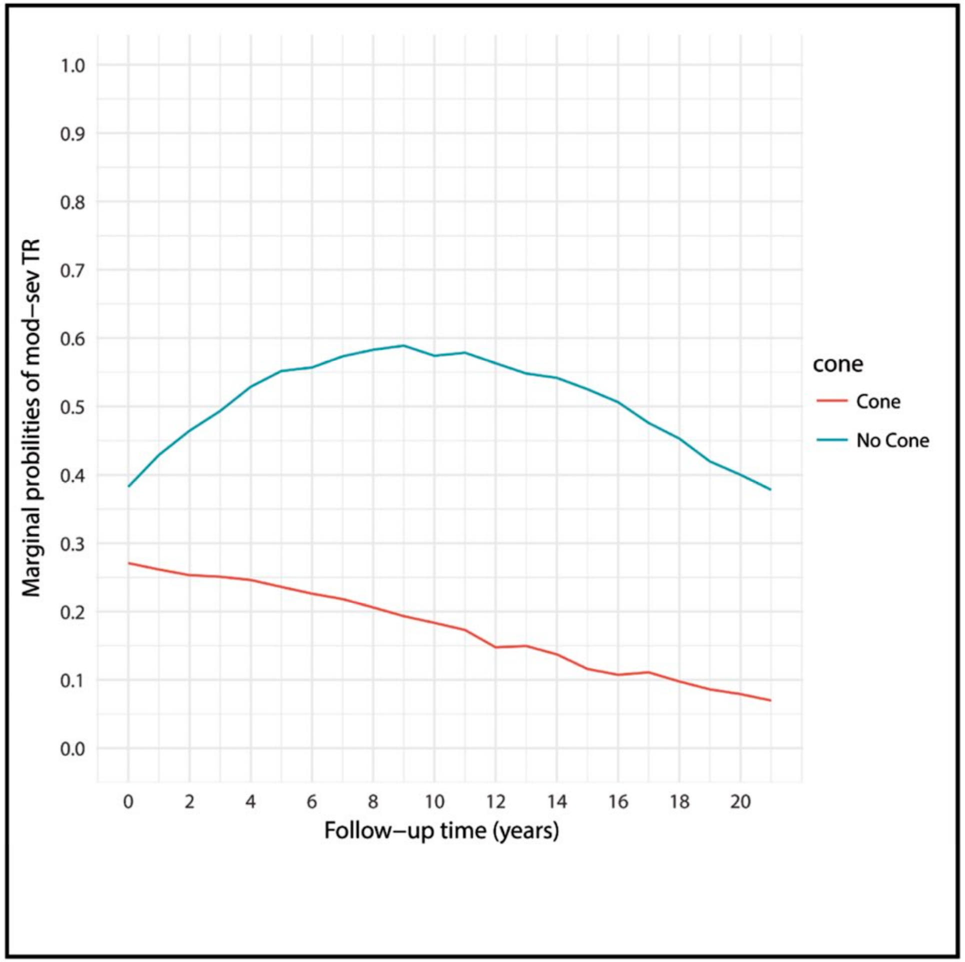
Is reconstructive surgery for an Ebstein anomaly associated with acceptable outcomes?

Key finding(s)

Hospital and late mortality rates were low.
Reoperation rates and valve function were acceptable.

Take-home message

Reconstructive surgery for Ebstein anomaly is a safe operation with a satisfactory long-term outcome.



Mortalité opératoire

- Danielson : 5,8%
- Carpentier et Chauvaud : 9%
- Da Silva (Cône) : 3,8%

A.Kühn *et al*. (Munich) : Tricuspid valve surgery improves cardiac output and exercise performance in patients with Ebstein's anomaly

Int J Cardiol (2011), doi:10.1016/j.ijcard.2011.11.033

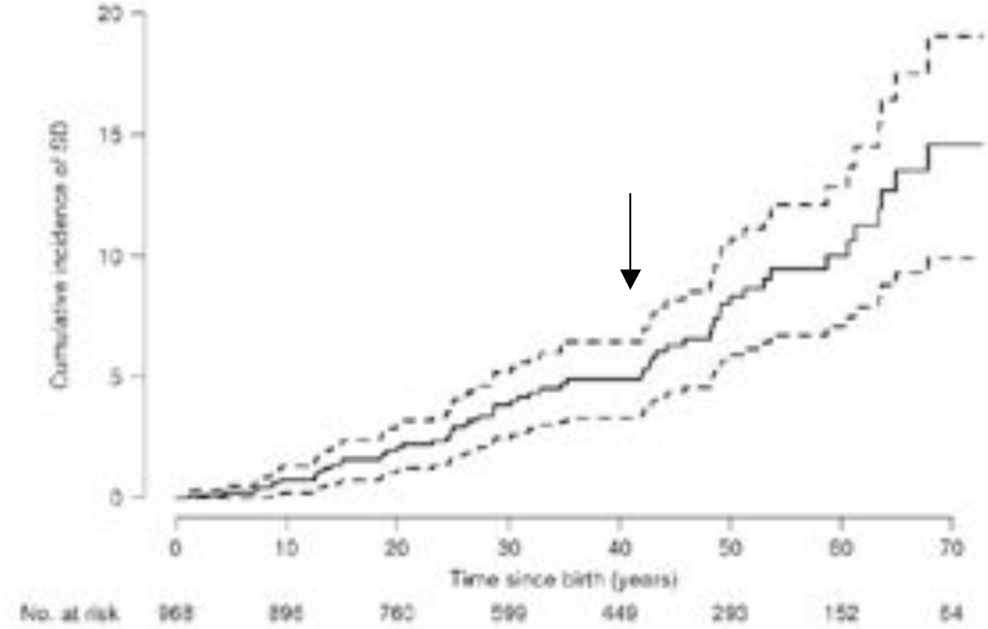
	Avant vs Après chirurgie	
Charge maximale :	1,87 vs 2,0 W/kg	p = 0,026
VO_2 max :	21 vs 22 mL/kg	p = 0,034
Débit cardiaque :	2,7 vs 2,9 l/min/m2	p = 0,035
VES pulmonaire :	29 vs 42 ml/m2	p = 0,005
FEVG :	61 vs 64 %	p = 0,083
FEVD :	50 vs 42%	p = 0,036
TAPSE :	23 vs 17 mm	p = 0,002
VTDVD :	124 vs 108 mL/m2	p = 0,034

Risque de mort subite

Cohorte de 968 patients Mayo Clinic

10 ans :   0,8%
50 ans :   8,3%
70 ans : 14,6%

Facteurs prédictifs : HR
TV : 6,37
Insuffisance cardiaque : 5,64
Chirurgie tricuspide : 5,94
Syncope : 2,03
Sténose pulmonaire : 3,42
Hb > 15 d/dL : 2,05



Take home figure The Kaplan–Meier curve demonstrating the incidence of sudden death from birth (dotted lines represent 95% confidence intervals). SD, sudden death.

Ch.H. Attenhofer Jost *et al.* European Heart Journal (2018) 39, 1970–1977

Indications chirurgicales : elles restent débattues

- Evident :
  - Symptomatique et bonne forme anatomique : chirurgie
  - Asymptomatique et mauvaise forme anatomique : abstention
- Plus difficile :
  - Asymptomatique et bonne forme anatomique : prévention des complications ?
  - Symptomatique et mauvaise forme anatomique : quelle chirurgie ?

d'après une diapo de Daniel Sidi

Indications chirurgicales

- Asymptomatique
  - sans dilatation de l 'OD : abstention
  - avec dilatation de l 'OD : Plastie préventive ?
  - avec shunt G-D atrial : Fermeture percutanée de la CIA
- Trouble du rythme: ablation ou plastie
- Symptomatique
  - Type A : plastie + fermeture de CIA
  - Type B : plastie + fermeture CIA ± DCPP
  - Type C : plastie + DCPP ± ouverture SIA ou DCPT
  - Type D : BTT ou DCPT
  - Mauvais VG : Transplantation

d'après une diapo de Daniel Sidi

ESC Guidelines for the management of grown-up congenital heart disease

European Heart Journal (2010) 31, 2915–2957

Table 14 Indications for intervention in Ebstein's anomaly

Indications	Class[a]	Level[b]
Indications for surgery		
• Surgical repair should be performed in patients with more than moderate TR and symptoms (NYHA class >II or arrhythmias) or deteriorating exercise capacity measured by CPET	I	C
• If there is also an indication for tricuspid valve surgery, then ASD/PFO closure should be performed surgically at the time of valve repair	I	C
• Surgical repair should be considered regardless of symptoms in patients with progressive right heart dilation or reduction of RV systolic function and/or progressive cardiomegaly on chest X-ray	IIa	C
Indications for catheter intervention		
• Patients with relevant arrhythmias should undergo electrophysiologic testing, followed by ablation therapy, if feasible, or surgical treatment of the arrhythmias in the case of planned heart surgery	I	C
• In the case of documented systemic embolism probably caused by paradoxical embolism, isolated device closure of ASD/PFO should be considered	IIa	C
• If cyanosis (oxygen saturation at rest <90%) is the leading problem, isolated device closure of ASD/PFO may be considered but requires careful evaluation before intervention (see text)	IIb	C

[a]Class of recommendation.
[b]Level of evidence.
ASD = atrial septal defect; CPET = cardiopulmonary exercise testing; NYHA = New York Heart Association; PFO = patent foramen ovale; RV = right ventricle; TR = tricuspid regurgitation; VSD = ventricular septal defect.

Grossesse et Ebstein

- Risques de
  - Arythmie
  - Insuffisance cardiaque
  - Embolie paradoxale
- Grossesse à haut risque si :
  - Cyanose
  - Arythmie
  - Insuffisance cardiaque droite

- Risque de cardiopathie congénitale fœtale : 4-6%

Ebstein anténatal

- Anatomie :
  - Valve tricuspide
  - VD fonctionnel
  - Voie d’éjection droite
  - Autre malformation
- Hémodynamique :
  - IT
  - Shunt inter-atrial (dimension du FOP)
  - Atrésie pulmonaire anatomique ou fonctionnelle ?
  - Flux ductal
  - Dilatation cardiaque
  - Anasarque
- Information
  - => IMG ?
  - => Organisation de la naissance en milieu cardiopédiatrique

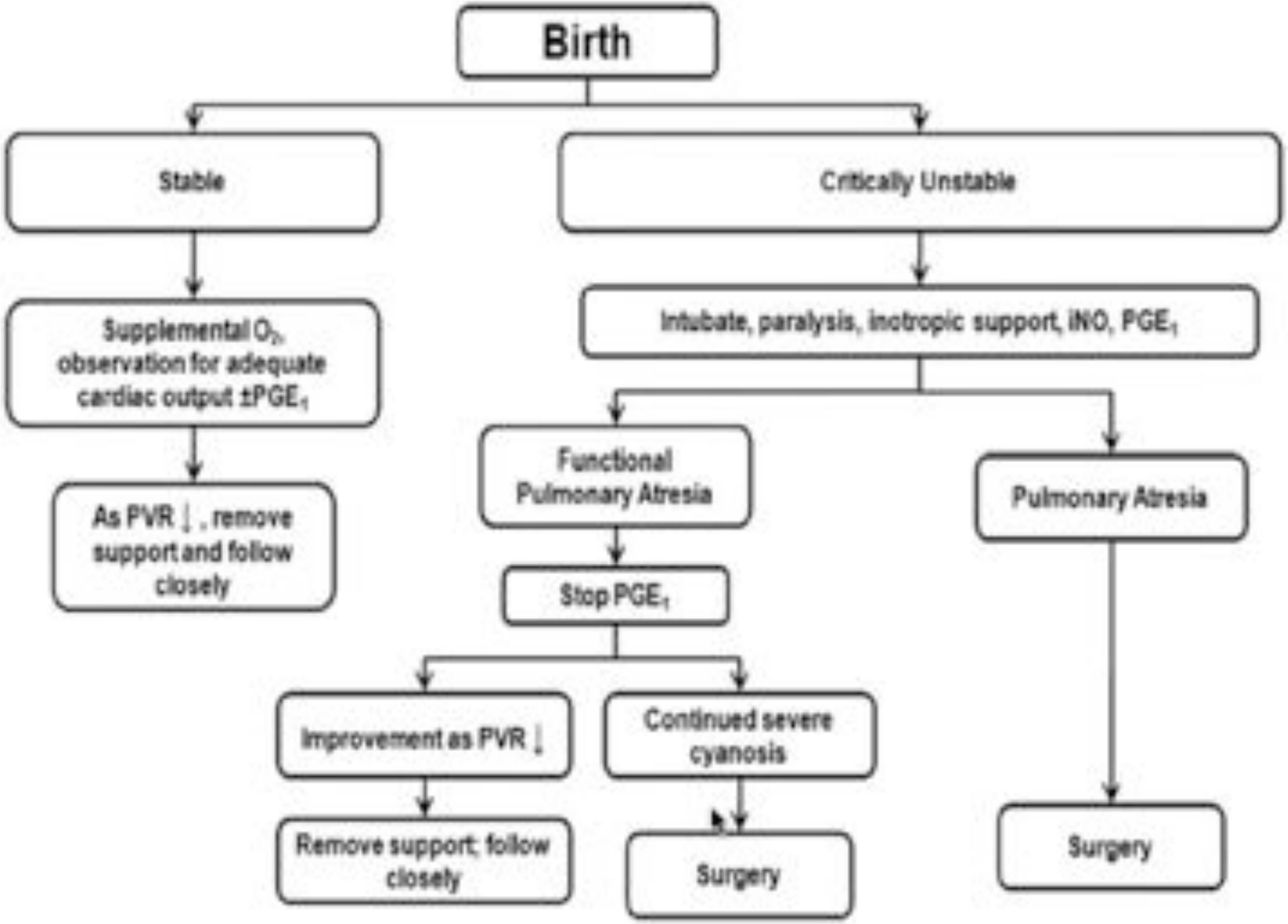


Figure 1. Algorithm for the initial management of Ebstein anomaly in neonates.

T.K. Susheel Kumar, Umar S. Boston, and Christopher J. Knott-Craig,
Seminars in Thoracic and Cardiovascular Surgery. 2017, 29 (3):331-337

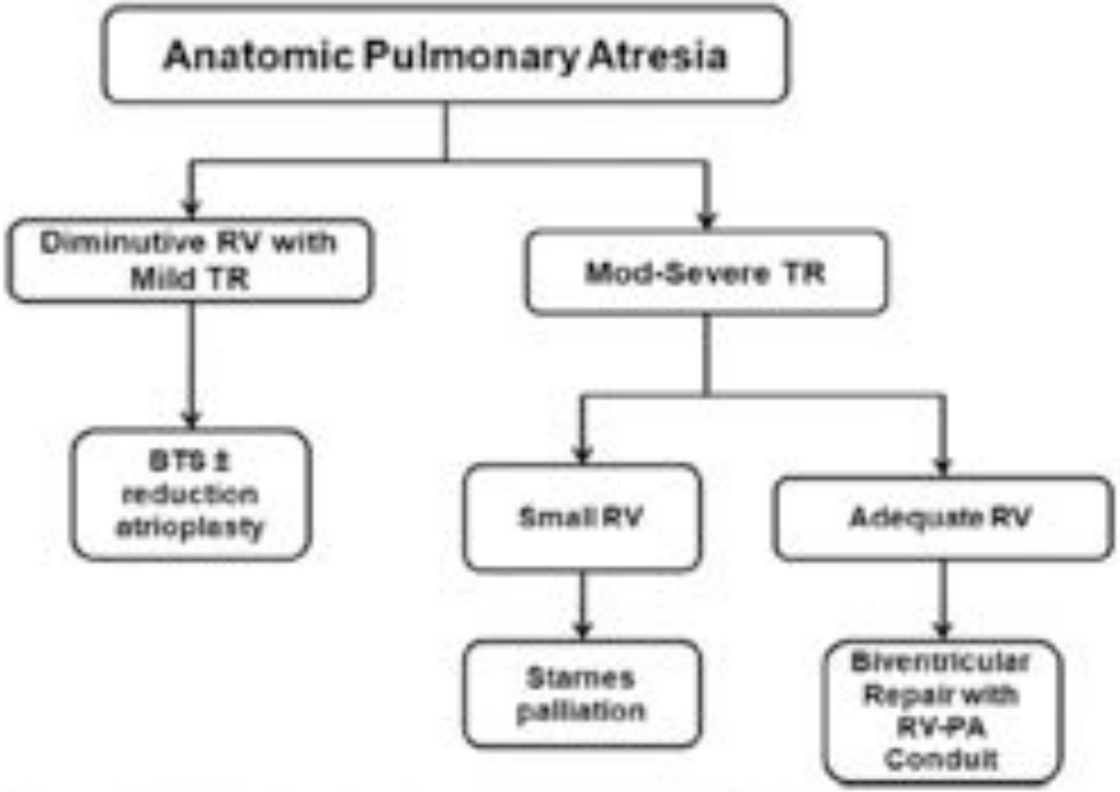


Figure 2. Algorithm for the management of Ebstein anomaly with anatomical pulmonary atresia. RV-PA, right ventricle to pulmonary artery.

T.K. Susheel Kumar, Umar S. Boston, and Christopher J. Knott-Craig,
Seminars in Thoracic and Cardiovascular Surgery. 2017, 29 (3):331-337

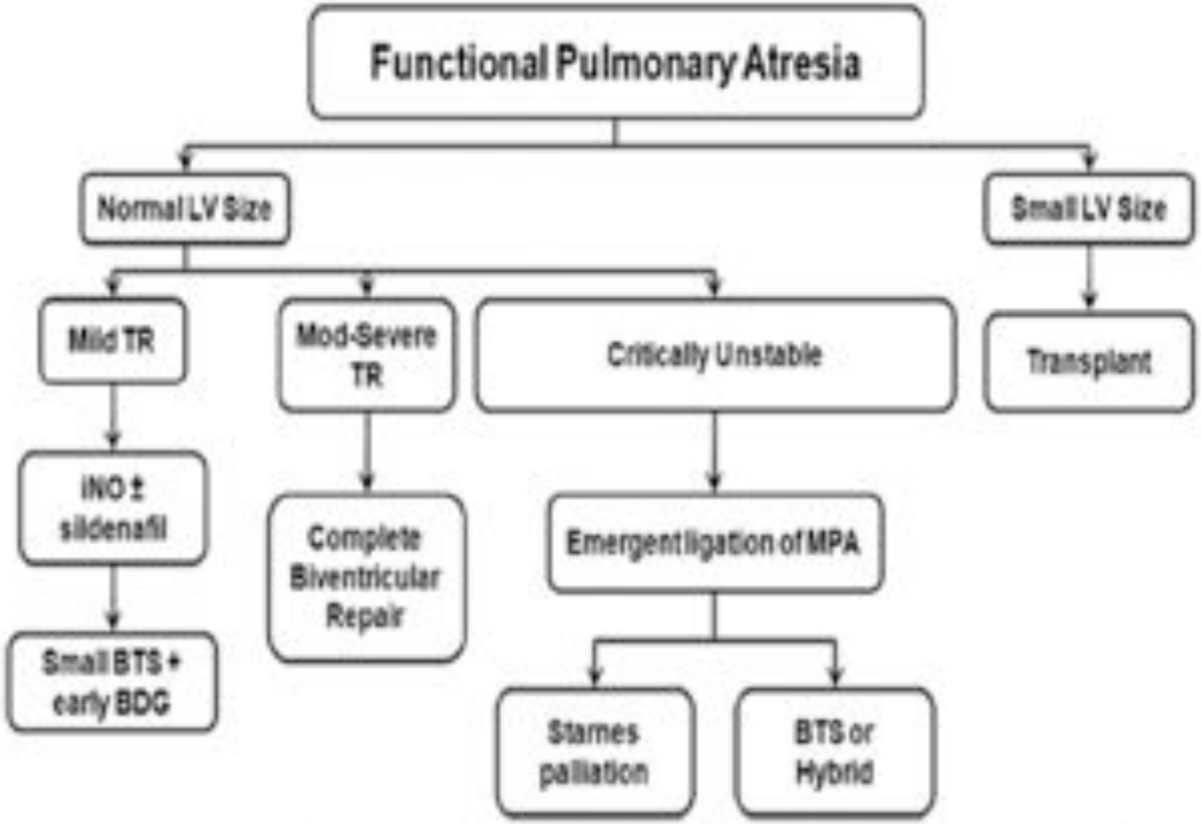


Figure 3. Algorithm for management of Ebstein anomaly with functional pulmonary atresia. BDG, bidirectional glen.

T.K. Susheel Kumar, Umar S. Boston, and Christopher J. Knott-Craig,
Seminars in Thoracic and Cardiovascular Surgery. 2017, 29 (3):331-337