

RV falen bij cardiomyopathie

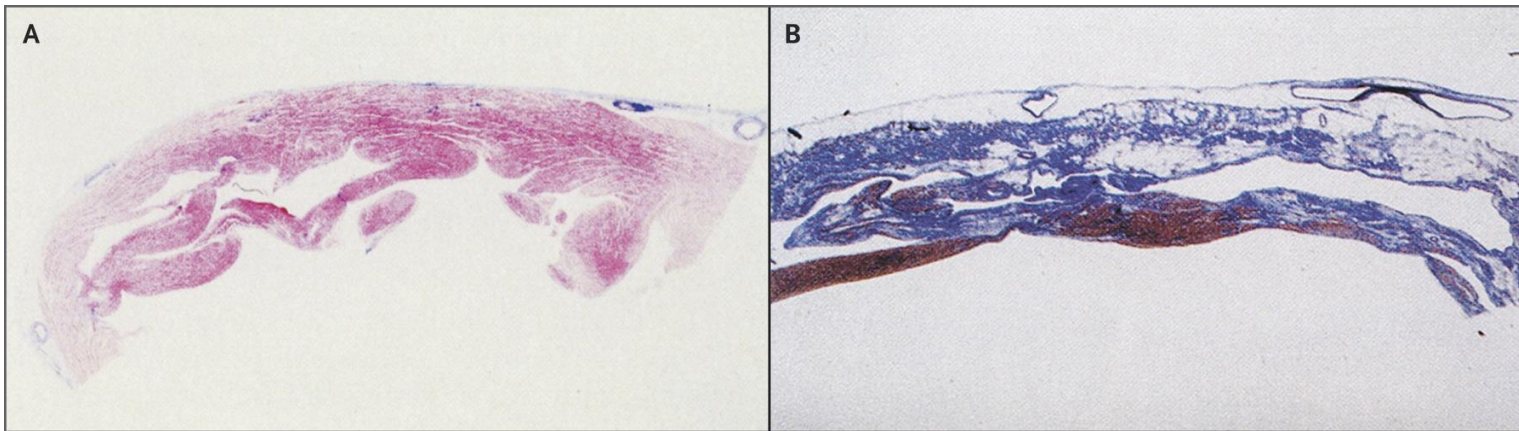
Dr. Anneline te Riele – cardioloog
UMC Utrecht

Disclosures

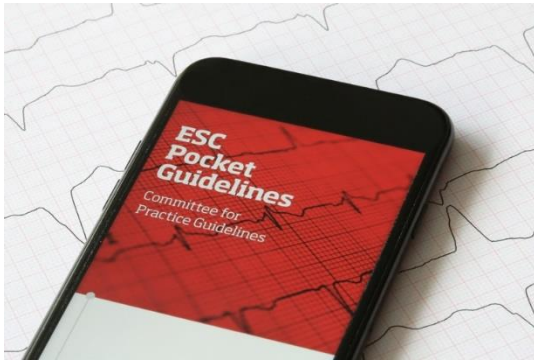
Potentiële belangenverstremgeling	
Voor presentatie mogelijk relevante relaties:	
Sponsoring of onderzoeksgeld	HORIZON 2020 IMPACT grant, ZonMW Off Road 2021
Honorarium of andere (financiële) vergoeding	Consultant voor Tenaya Therapeutics
Aandeelhouder	
Andere relatie, namelijk ...	

Aritmogene rechter ventrikel cardiomyopathie

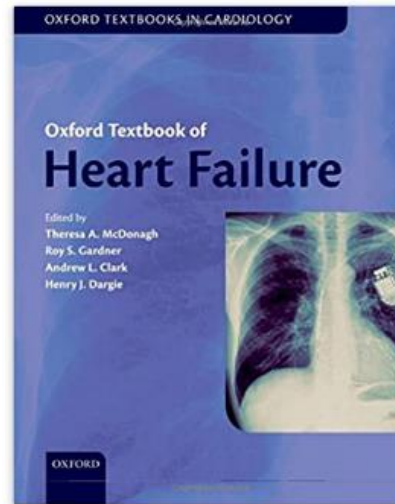
- Erfelijke hartspierziekte
- ~60% mutatie in cardiale desmosoom
- Progressieve vervetting en verlittekening van myocard
- (Rechtszijdig) hartfalen
- Ventriculaire ritmestoornissen



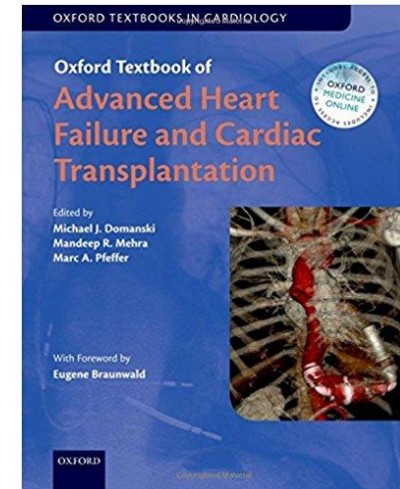
Wat zeggen de richtlijnen over RV falen?



No chapter on RV failure

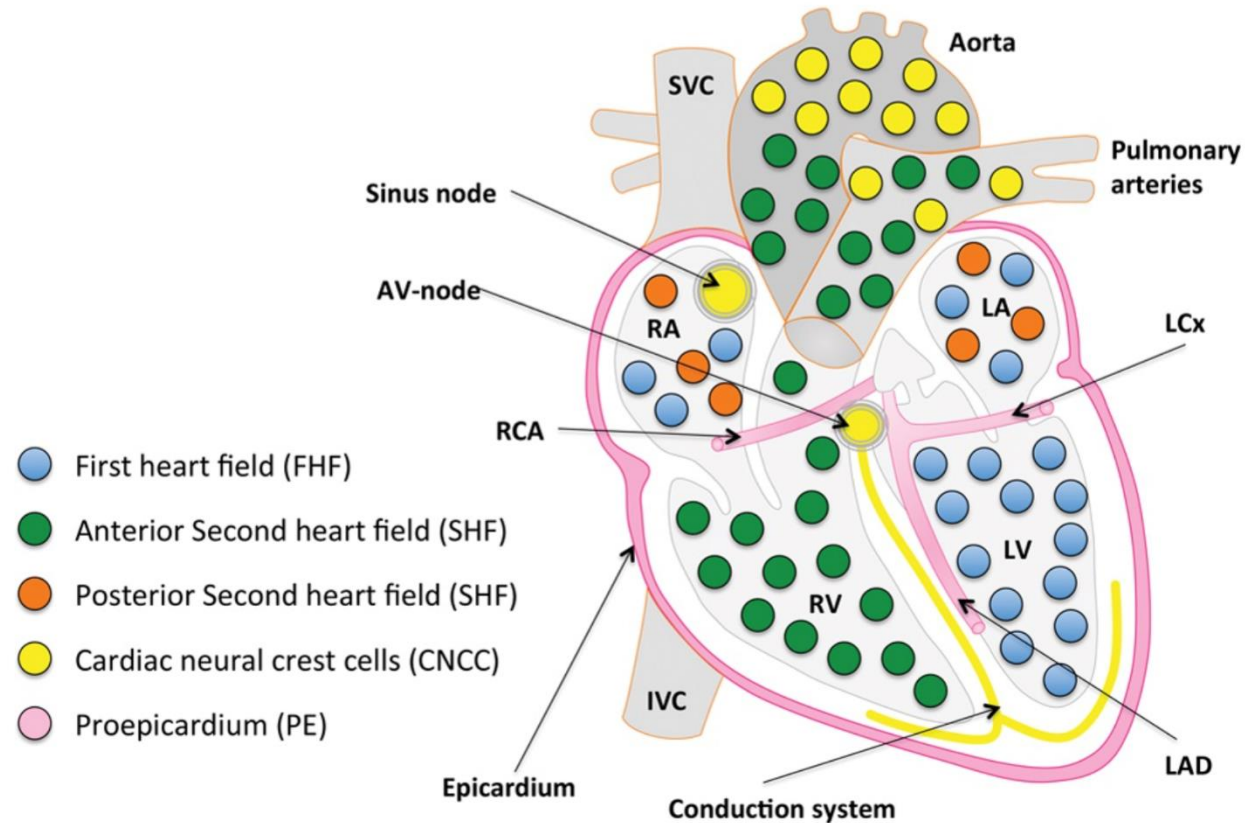


Right Ventricle Failure among co-morbidities



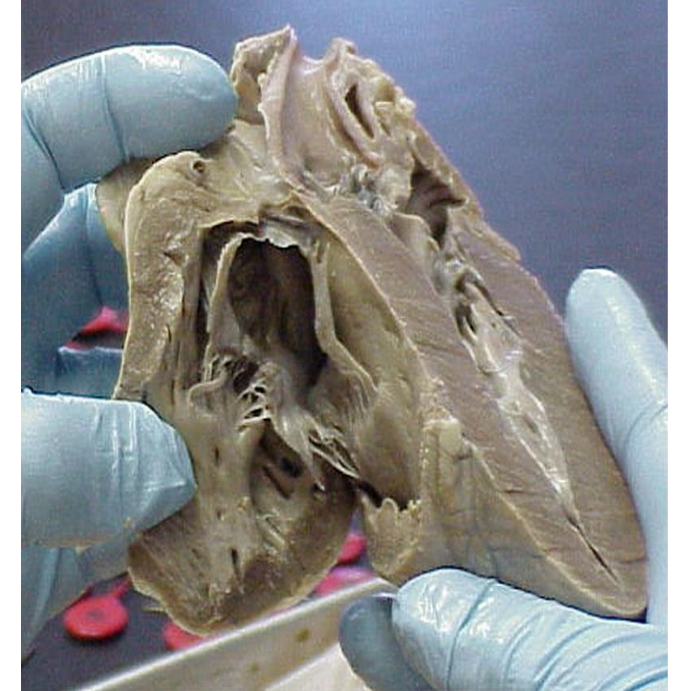
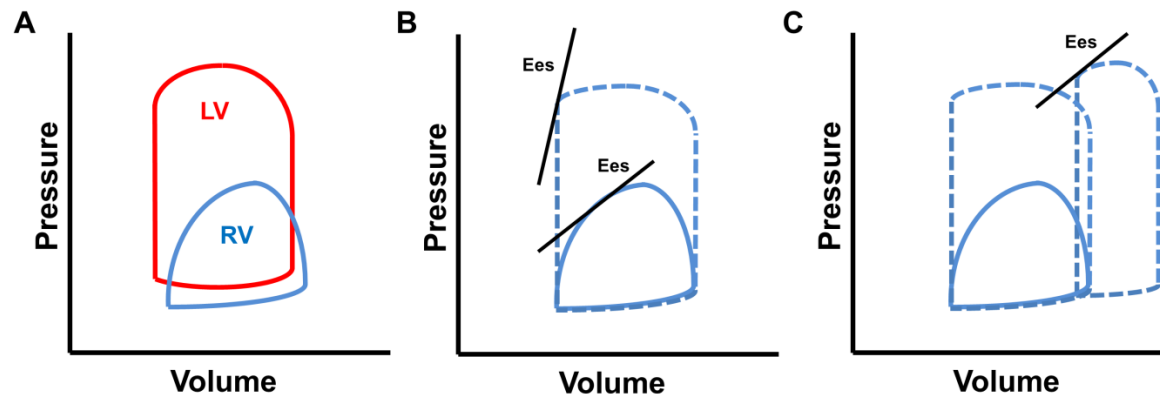
Chapter 20: RV Failure - anatomy, pathophysiology
Chapter 21: RV failure - management

De RV is geen “linker ventrikel aan de rechterkant”

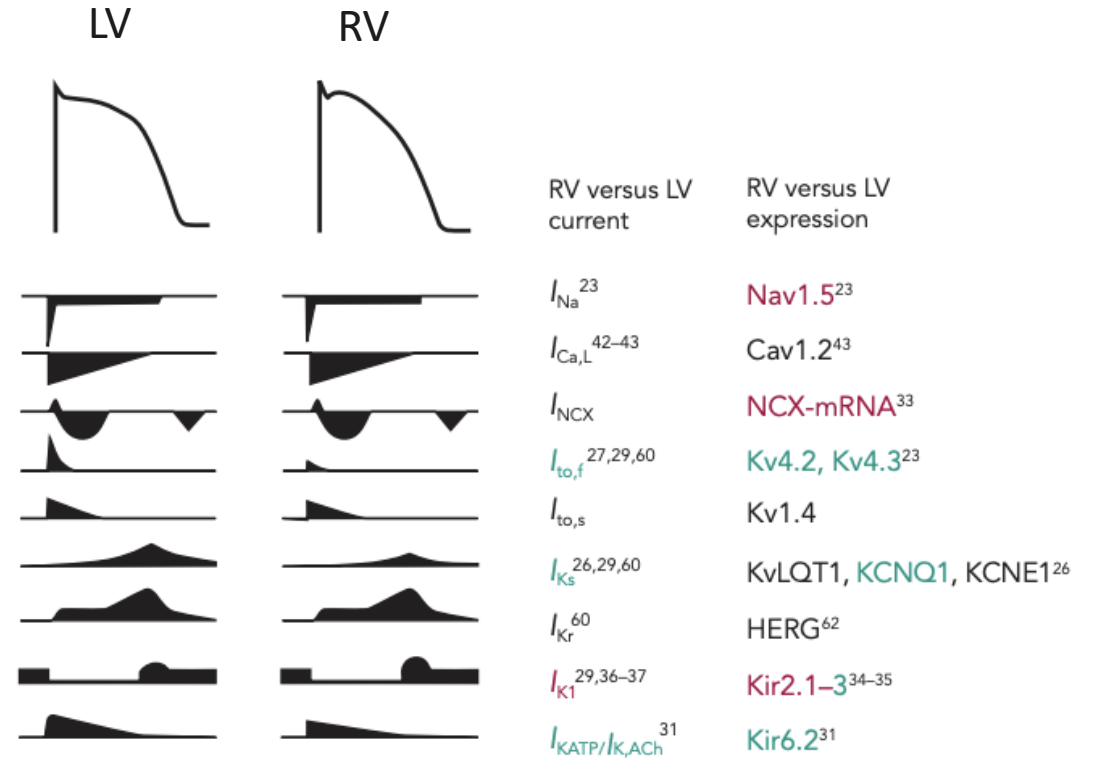
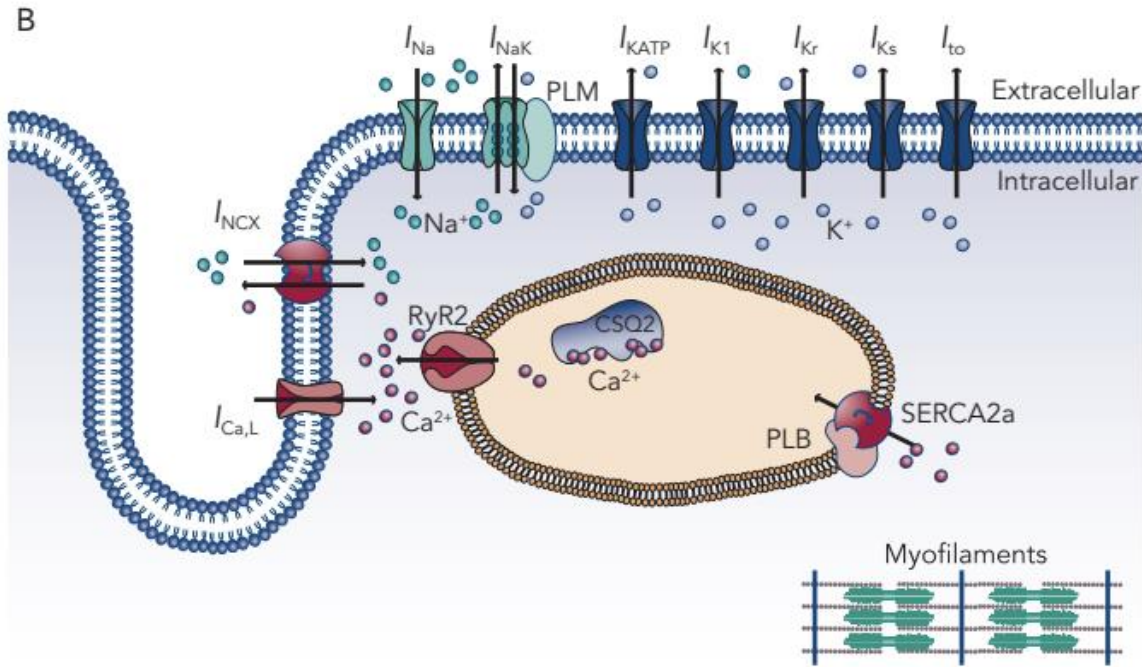


... Mechanisch en hemodynamisch

- RV heeft een hoge "surface-to-volume" ratio
 - Twee lagen cardiomyocyten (transversaal / longitudinaal)
 - Lagere drukken
 - Kortere isovolumetrische contractie en relaxatie
 - Minder gevoelig voor ischemie
 - Tot 40% van RV output is afhankelijk van septum
- RV falen bij "abnormal loading conditions" (druk / volume)



... Elektrofysiologisch



Ionic currents involved in the initiation and maintenance of an action potential (AP) and their chamber-specific differences. Green means up-regulation and red down-regulation in RV versus LV and black indicates no change between chambers. LV = left ventricle; RV = right ventricle. Underlying experimental data are summarised in Table 1.

Groepen van RV falen (niet "mutually exclusive")

Druk-overbelasting

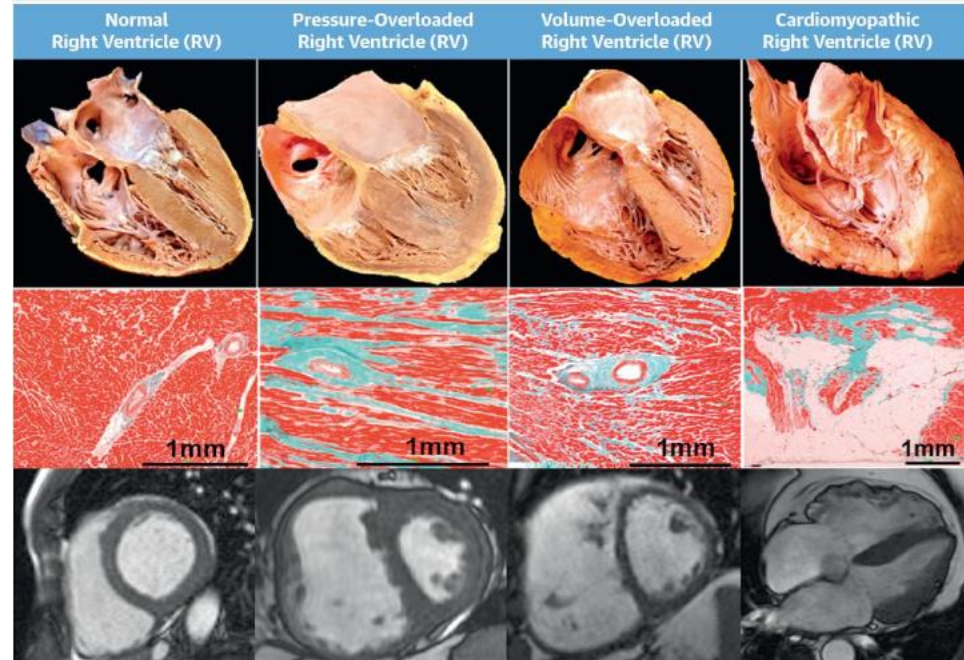
Pulmonale (arteriele) hypertensie
Longembolie

Volume-overbelasting

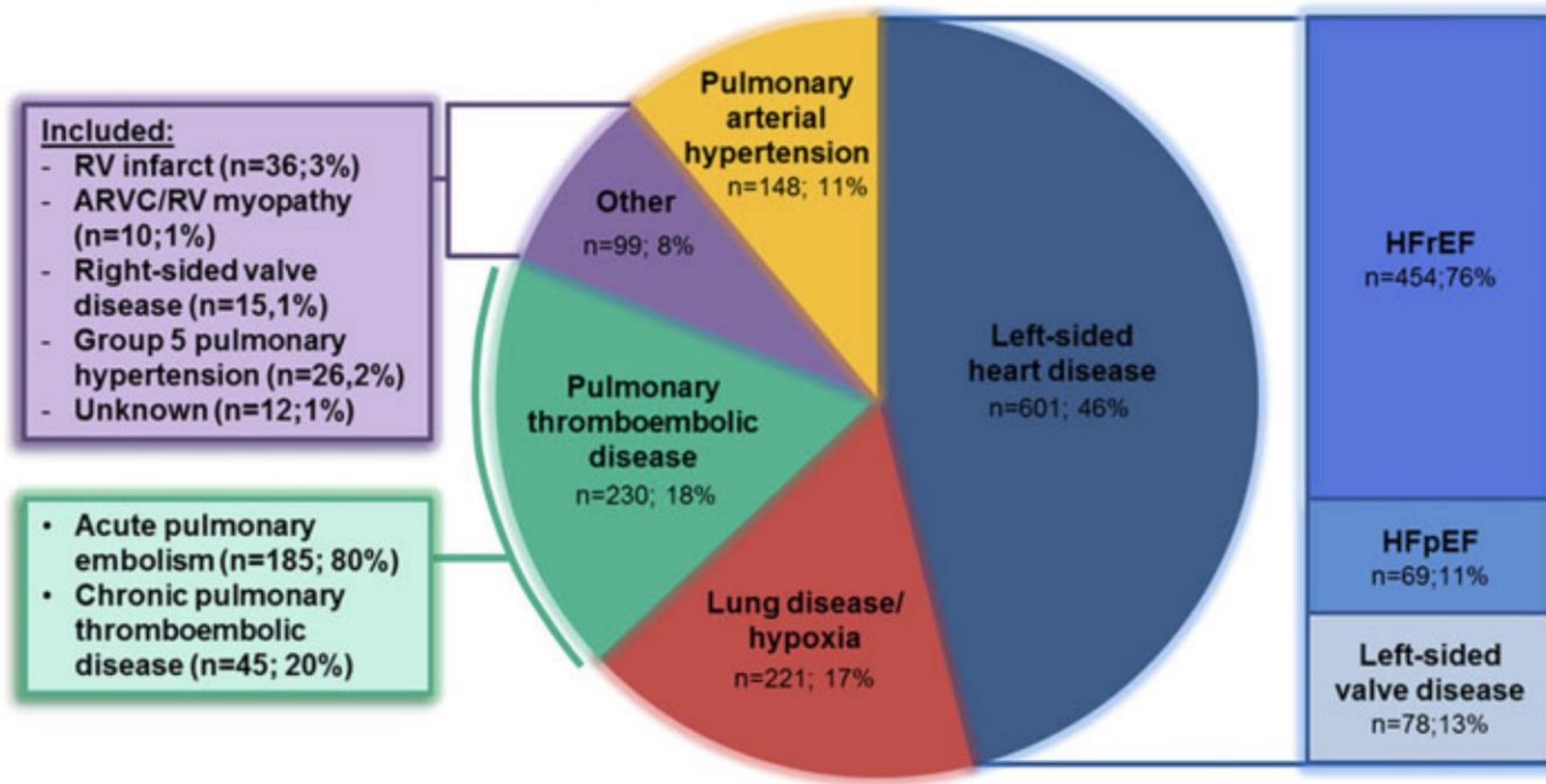
Links-rechts shunt
Tricuspidalisklepinsufficiëntie

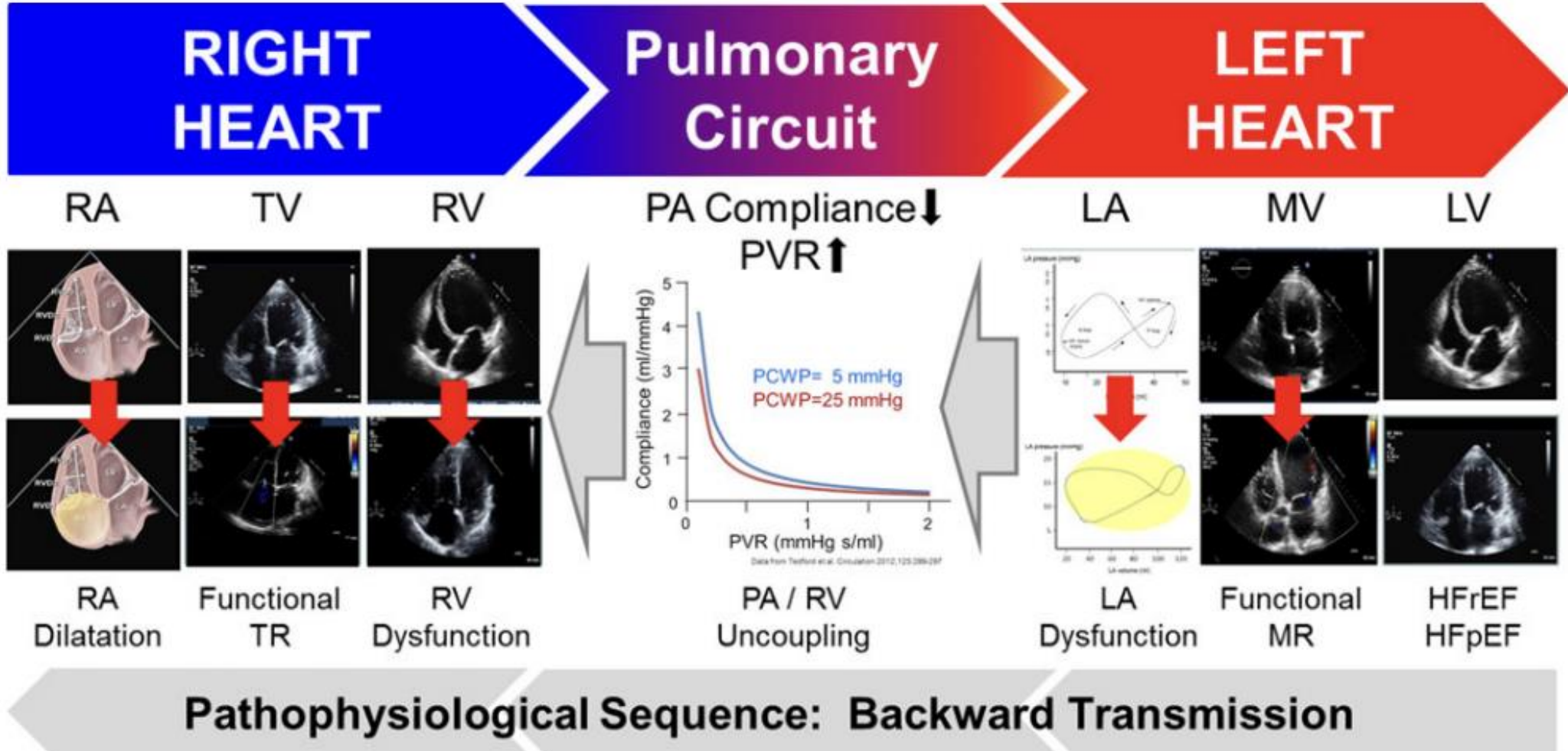
Afname contractiliteit

Cardiomyopathie (oa ARVC)
Post-pericardiotomie (/LVAD)
Ischemie



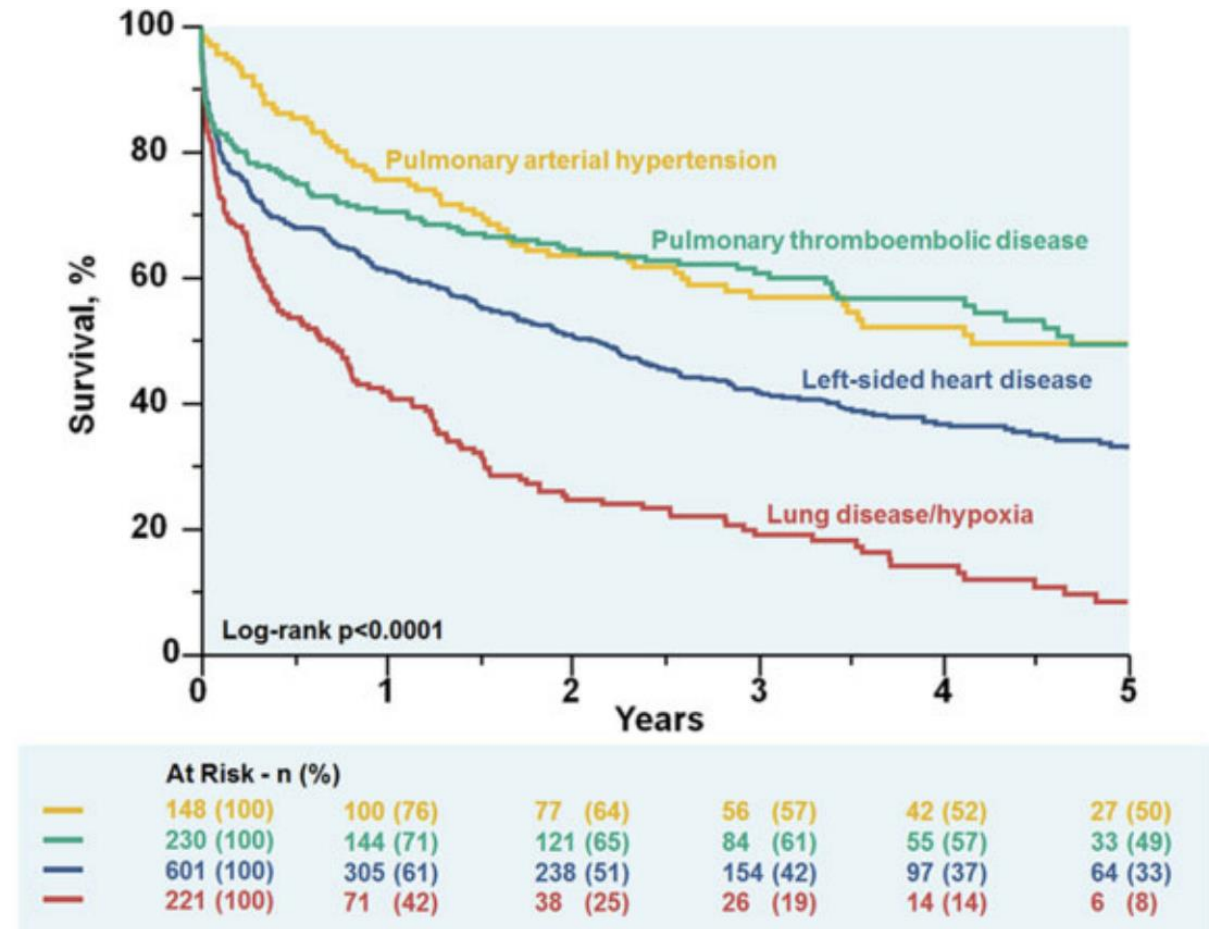
Oorzaken van ernstige RV dysfunctie (4% van n=65,000)





RV falen voorspelt klinisch beloop van hartfalen

- 1299 pts met ernstige RV dysfunctie¹
 - 1jr overleving 61% (range 58-64%)
 - 5jr overleving 33% (range 28-37%)
- Pooled analyse²
 - per 5mm ↓TAPSE = 26% toename
 - per 5% ↓FAC = 16% toenamein mortaliteit in 5 jaar



1. Padang et al. Eur Heart J 2020;41:1273-82
2. Gorter et al. Eur J Heart Fail 2016;18:1472-87

Beoordeling RV falen bij de hartfalenpatient

Klinisch

Imaging

Hemodynamisch

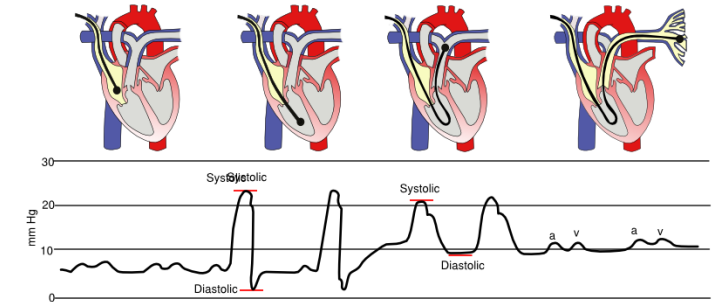
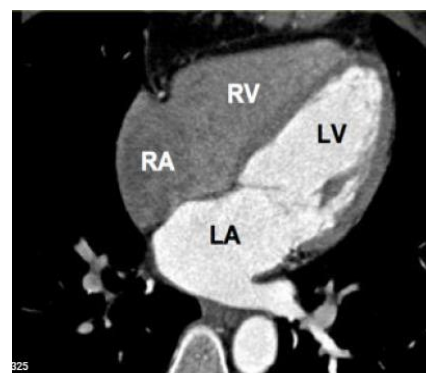
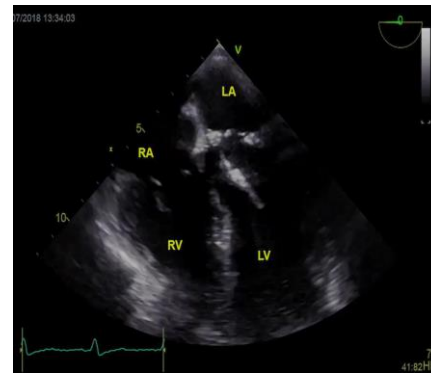
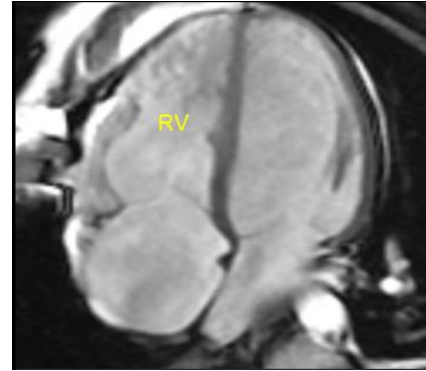
RIGHT SIDED FAILURE

(Cor Pulmonale)

- Fatigue
- ↑ Peripheral Venous Pressure
- Ascites
- Enlarged Liver & Spleen
- May be secondary to chronic pulmonary problems
- Distended Jugular Veins
- Anorexia & Complaints of GI Distress
- Weight Gain
- Dependent Edema

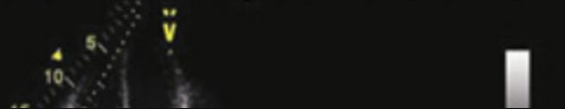


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Tricuspid Annular Plane Systolic Excursion



(TAPSE)

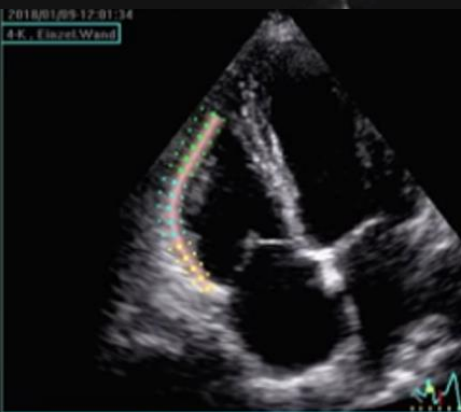


Tissue Velocity Imaging (TVI)



Fractional area change (FAC)

- RVIT ———
- RV SAX ·······
- RV LAX - - - -
- End diastolic area 
- End systolic area 



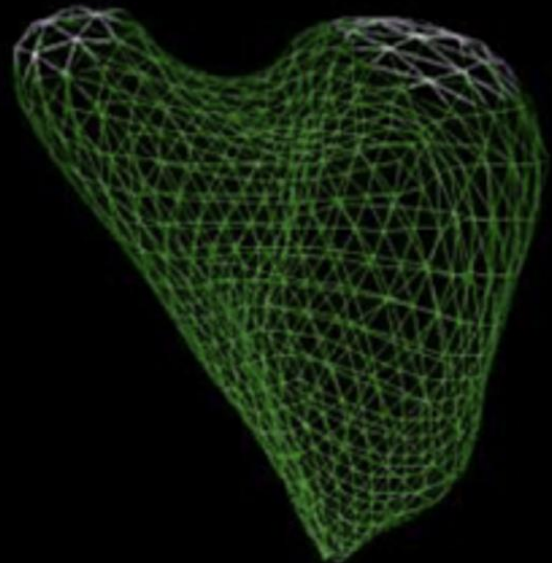
Peak Systolic Strain



Besteed aandacht aan goede beoordeling van RV!

Overige parameters:

- TAPSE/sPAP
- PAPI = (PASP – PADP) / RAP
- RVSWI = (mean PAP – mean RAP) * CI/HR * 0.0136



Beoordeling RV is belangrijk voor ELKE hartfalenpatient

Verwijzing advanced HF polikliniek i.v.m. verminderde LV functie eci bij jonge patient (38 jr oud):

Verslag - 25-05-2022 13:37:00:

Klinische gegevens/Vraagstelling:

ventriculaire tachycardie vermoedelijk uit RVOT; aanwijzingen voor ischemie? cardiomyopathie?

Verslag:

MRI hart volgens cardiomyopathie protocol

Beoordeling:

[REDACTED]

Kwaliteit:

Goed

Functie:

Matige linkerventrikelfunctie met ejectiefractie van 38%.

Morfologie:

Normale cardiale morfologie. Verwijd linkerventrikel met een geïndexeerd einddiastolisch volume van 106 ml/m².

Ook het geïndexeerde systolische volume is verhoogd namelijk 65 ml/m².

Geïndexeerde linkerventrikel massa binnen normale grenzen.

Wandbeweging stoornissen:

Geen regionale wandbewegingsstoornissen.

Late Enhancement:

Geen

Perfusie afwijkingen in rust:

Geen

Perfusie afwijkingen met adenosine (stress):

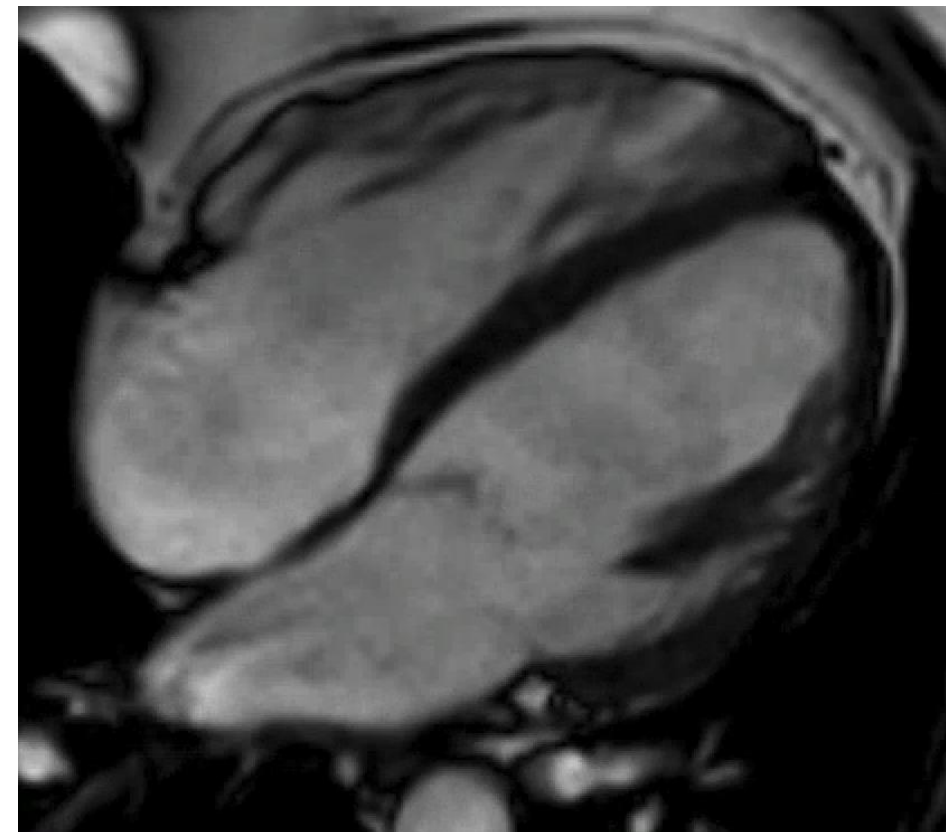
Geen

Overige afwijkingen:

Geen. Geen afwijkingen RVOT. Geen intramyocardiaal oedeem.

Conclusie:

Geen (infarct) littekens. Geen ischemie. Matige functie (38%) en wijd linkerventrikel



Diagnose ARVC gesteld




Behandeling RV falen

- Algemeen:
 - Adequate volume status (“loading conditions”)
 - Onderliggend ziektebeeld behandelen
 - Geen indicatie voor PH-specifieke therapie
- Weinig goede studies naar reguliere hartfalenmedicatie
 - Verschillende studies ongoing naar HFpEF en PHT
 - Lokale ervaring:
 - ACE-remmer
 - ARB
 - ... SGLT2-remmer?
 - sacubitril/valsartan?
 - (betablokker)

Congenital heart disease

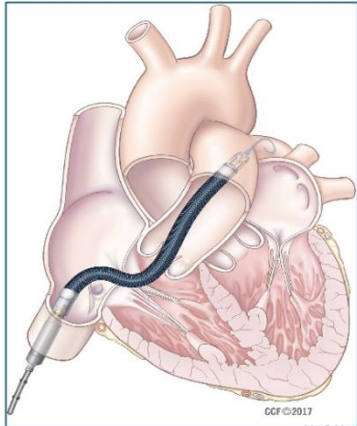
Original research

Sacubitril/valsartan in the treatment of systemic right ventricular failure

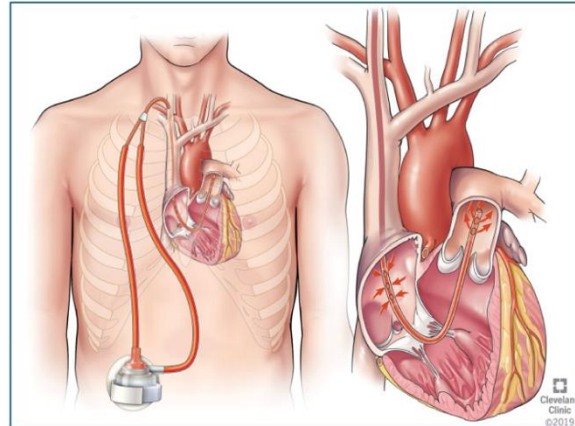
Tjitske E Zandstra ¹, Marieke Nederend,¹ Monique R M Jongbloed ^{1,2},
Philippine Kiès,¹ Hubert W Vliegen,¹ Berto J Bouma,³ Laurens F Tops,⁴ Martin J Schalij,⁴
Anastasia D Egorova ¹

Direct RV Bypass (RA → PA)

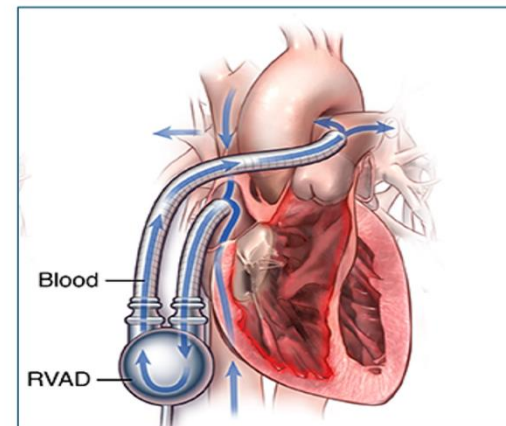
Indirect RV Bypass (RA → AO)



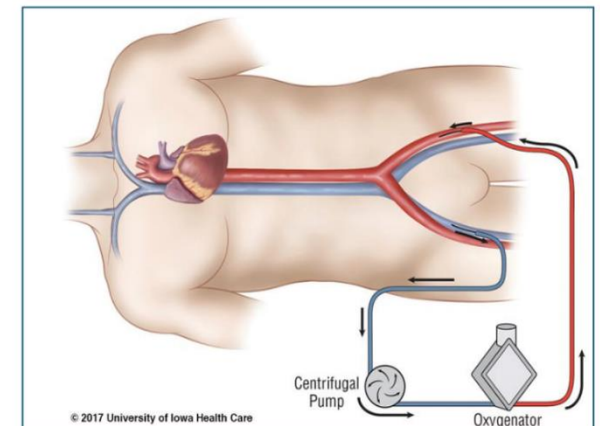
Impella RP



Protek Duo



(Tandem) RVAD



VA-ECLS



Harttransplantatie

Conclusie

- De rechter ventrikel is geen “linker ventrikel aan de rechterzijde”
- Besteed altijd aandacht aan beoordeling van RV functie
 - Belang voor juiste diagnose
 - Belang voor prognose
 - Evt. therapeutische mogelijkheden hangen er vanaf
- Behandeling toegespitst op specifieke patient
 - Onderliggend ziektebeeld behandelen
 - Cornerstone = optimaliseren preload / afterload
 - HF medicatie insluipen als mogelijk

LOOK RIGHT

A black and white photograph of a zebra crossing on a road. The crossing consists of several wide, parallel white stripes. The words "LOOK RIGHT" are painted in large, bold, white capital letters across the bottom of the crossing. In the background, a person is walking on the sidewalk, and a bicycle is parked near a building entrance. A dark metal fence runs along the sides of the road.

Diagnose ARVC

TABLE 1 Revised 2010 TFC for ARVD/C*

1 Global or Regional Dysfunction and Structural Alterations

Major

2D echocardiographic criteria

Regional RV akinesia, dyskinesia, or aneurysm and 1 of the following measured at end diastole:

- PLAX RVOT ≥ 32 mm (PLAX/BSA ≥ 19 mm/m²),
- PSAX RVOT ≥ 36 mm (PSAX/BSA ≥ 21 mm/m²), or
- FAC $\leq 33\%$

CMR criteria

Regional RV akinesia or dyskinesia and 1 of the following:

- RV EDV/BSA ≥ 110 ml/m²
- RV ejection fraction $\leq 40\%$

RV angiography criteria

Regional RV akinesia, dyskinesia, or aneurysm and 1 of the following:

Minor

2D echocardiographic criteria

Regional RV akinesia or dyskinesia and 1 of the following measured at end diastole:

- PLAX RVOT ≥ 29 to < 32 mm (PLAX/BSA ≥ 16 - < 19 mm/m²), or
- PSAX RVOT ≥ 32 to < 36 mm (PSAX/BSA ≥ 18 - < 21 mm/m²), or
- FAC $> 33\%$ $\leq 40\%$

CMR criteria

Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and 1 of the following:

- RV EDV/BSA ≥ 100 - < 110 ml/m² (male) or ≥ 90 - < 100 ml/m² (female)
- RV ejection fraction > 40 - $\leq 45\%$

2 Tissue Characterization of Wall

Major

Residual myocytes $< 60\%$ by morphometric analysis (or $< 50\%$ if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

Minor

Residual myocytes 60%-75% by morphometric analysis (or 50%-65% if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample with or without fatty replacement of tissue on endomyocardial biopsy

3 Repolarization Abnormalities

Major

Inverted T waves in right precordial leads (V₁, V₂, and V₃) or beyond in individuals > 14 yrs of age (in the absence of complete RBBB)

Minor

Inverted T waves in V₁ and V₂ in individuals > 14 yrs of age (in the absence of complete RBBB) or in V₄, V₅, and V₆

Inverted T waves in leads V₁, V₂, V₃, and V₄ in individuals > 14 yrs of age in the presence of a complete RBBB

4 Depolarization/Conduction Abnormalities

Major

Non-sustained or sustained VT of LBBB morphology with superior axis

Minor

Non-sustained or sustained VT of RVOT configuration, LBBB morphology with inferior axis or with unknown axis

> 500 PVCs per 24 h on Holter monitoring

6 Family History

Major

ARVD/C in first-degree relative who meets current TFC

ARVD/C confirmed pathologically at autopsy or surgery in a first-degree relative

Identification of a pathogenic mutation categorized as associated or probably associated with ARVD/C in the patient under evaluation

Minor

History of ARVD/C in first-degree relative in whom it is not possible to determine whether the family member meets current TFC

Premature sudden death (< 35 yrs of age) due to suspected ARVD/C in a first-degree relative

ARVD/C confirmed pathologically or by current TFC in second-degree relative

Complexe set van diagnostische criteria:
4 criteria (2 major; 1 major+2 minor; of 4 minor) voor definitieve diagnose
Diagnose "per exclusionem": andere oorzaken redelijkerwijs uitgesloten





European Journal of Heart Failure (2018) 20, 16–37
doi:10.1002/ejhf.1029

HFA POSITION PAPER

Right heart dysfunction and failure in heart failure with preserved ejection fraction: mechanisms and management. Position statement on behalf of the Heart Failure Association of the European Society of Cardiology

Thomas M. Gorter¹, Dirk J. van Veldhuisen¹, Johann Bauersachs², Barry A. Borlaug³, Jelena Celutkiene⁴, Andrew J.S. Coats^{5,6}, Marisa G. Crespo-Leiro⁷, Marco Guazzi⁸, Veli-Pekka Harjola⁹, Stephane Heymans¹⁰, Loreena Hill¹¹, Mitja Lainscak¹², Carolyn S.P. Lam¹³, Lars H. Lund¹⁴, Alexander R. Lyon¹⁵, Alexandre Mebazaa¹⁶, Christian Mueller¹⁷, Walter J. Paulus¹⁸, Burkert Pieske¹⁹, Massimo F. Piepoli²⁰, Frank Ruschitzka²¹, Frans H. Rutten²², Petar M. Seferovic²³, Scott D. Solomon²⁴, Sanjiv J. Shah²⁵, Filippos Triposkiadis²⁶, Rolf Wachter²⁷, Carsten Tschöpe¹⁹, and Rudolf A. de Boer^{1*}



European Journal of Heart Failure (2016) 18, 226–241
doi:10.1002/ejhf.478

Contemporary management of acute right ventricular failure: a statement from the Heart Failure Association and the Working Group on Pulmonary Circulation and Right Ventricular Function of the European Society of Cardiology

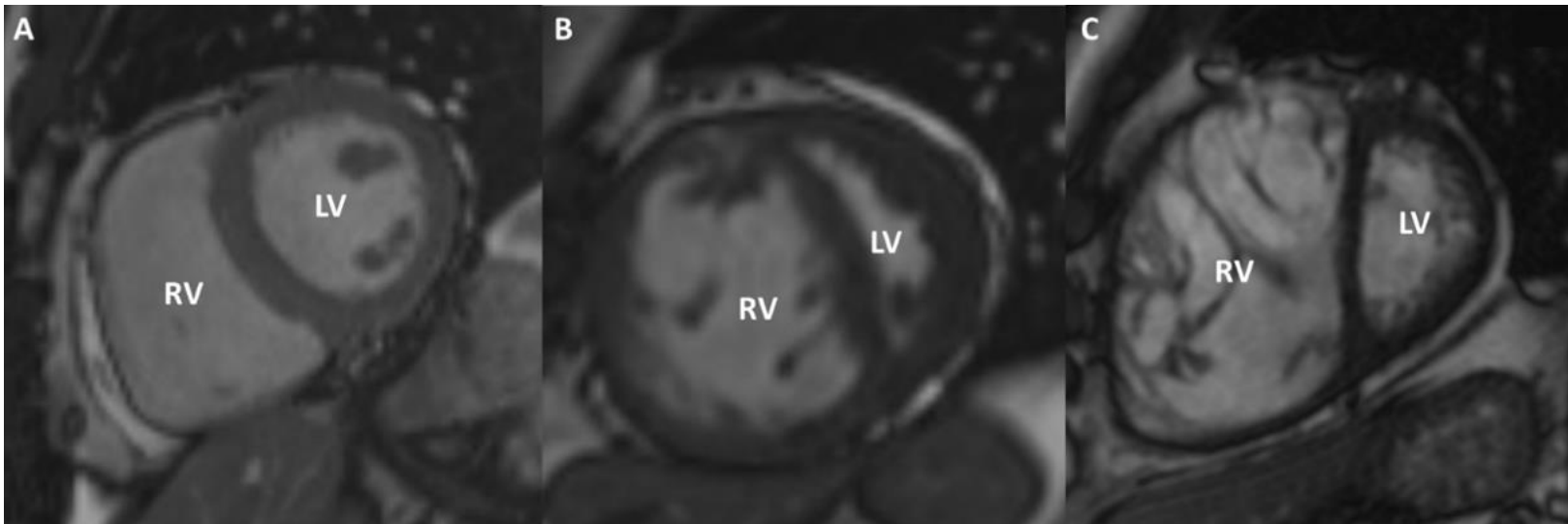
Veli-Pekka Harjola^{1*}, Alexandre Mebazaa^{2,3,4}, Jelena Čelutkienė⁵, Dominique Bettex⁶, Hector Bueno^{7,8,9}, Ovidiu Chioncel¹⁰, Maria G. Crespo-Leiro¹¹, Volkmar Falk¹², Gerasimos Filippatos¹³, Simon Gibbs¹⁴, Adelino Leite-Moreira¹⁵, Johan Lassus¹⁶, Josep Masip¹⁷, Christian Mueller¹⁸, Wilfried Mullens¹⁹, Robert Naeije²⁰, Anton Vonk Nordegraaf²¹, John Parissis²², Jillian P. Riley¹⁴, Arsen Ristic²³, Giuseppe Rosano^{24,25}, Alain Rudiger²⁶, Frank Ruschitzka²⁷, Petar Seferovic²⁸, Benjamin Sztrymf²⁹, Antoine Vieillard-Baron³⁰, Mehmet Birhan Yilmaz³¹, and Stavros Konstantinides^{32,33}



UMC Utrecht

Beoordeling rechter ventrikel morfologie / functie

- RV heeft complexe geometrie en vorm
 - Positie van het septum
 - Trabecularisatie
 - Moderatorband
- “Normale RV” komt in vele vormen: accurate beoordeling wandbeweging vereist ervaring!



Normaal

PAH

Tetralogie van Fallot



Pectus excavatum

