

# **International Symposium THE RIGHT HEART THE NEW FRONTIER Highlights**

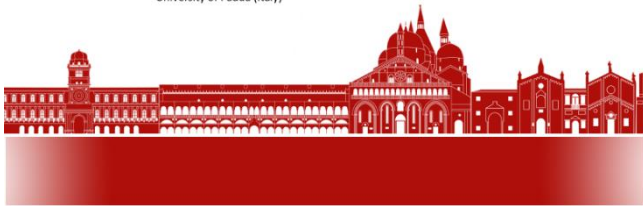
**Padua (Italy), March 01-03, 2017**

## **Introduction**



International Symposium on:  
**THE RIGHT HEART.  
THE NEW FRONTIER**

Padua, March 1<sup>st</sup> - 3<sup>rd</sup>, 2017  
March 1<sup>st</sup> at Palazzo del Bo - March 2<sup>nd</sup> - 3<sup>rd</sup> at Botanical Garden  
University of Padua (Italy)



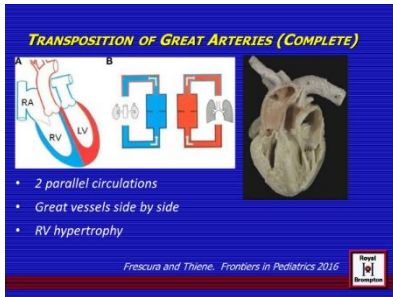
Prof. Iliceto, chairman of the symposium, opened the congress, by highlighting the high scientific level of the University of Padua in full compliance with the scientific level of this congress focused on the physiology, physiopathology, diagnosis, prognosis and treatment of the right heart diseases. Many top researchers in cardiology focused on right heart diseases, coming from all the world attended this symposium

together with young physicians and cardiologists. This congress represented a very unique occasion for a full update on the Right Heart from physiology to pathology and the related pharmacological and surgical treatments.

To follow the presentations of this congress, click on the link below:

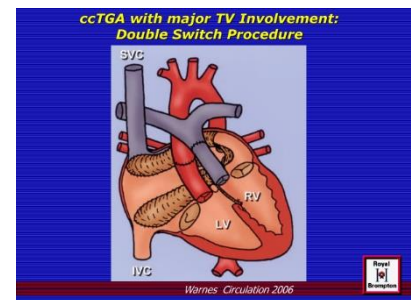
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# The spectrum of congenital heart diseases with systemic right ventricle

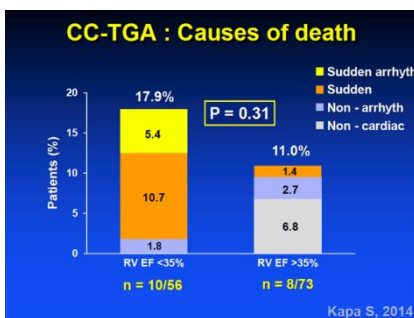


Prof. Gatzoulis from London (UK), spoke about the spectrum of congenital heart diseases with systemic right ventricle. The speaker went deeper in his talk, by presenting very interesting data on the transposition of the great arteries (TGA), the congenitally corrected TGA and on the so called “single ventricle” (RV). More in particular Prof. Gatzoulis spoke about specific procedures for the TGA correction, like the Mustard procedure and the congenitally corrected TGA and about

specific pathological pictures like the hypoplastic left heart syndrome. In the main part of his lecture, the speaker discussed the main abnormalities which characterize these syndromes, by highlighting the role played by the myofibril arrangements leading to the major contractile alterations and the related ventricular dysfunctions. Speaking about the determinants of the systemic RV function, Prof. Gatzoulis highlighted the role played by the myocardial fibrosis in the development of the RV hypertrophy. In the second part of his lecture, the speaker presented very interesting data given by clinical studies on patients affected by these



congenital diseases. Speaking about therapy, Prof. Gatzoulis presented very interesting data on the poor effect of ACE inhibitors and on the cardiac resynchronization therapy. In the last part of his lecture, the speaker talked about the congenitally corrected TGA, the related symptomatology and outcomes. In conclusion, Prof. Gatzoulis pointed out that despite the peripheral-central interactions and the exercise training have a clear potential, the premature RV failure is inevitable in many patients.



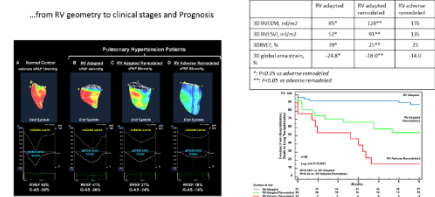
- What's about the tricuspid valve replacement based on the data presented by the speaker?
- What's about PA banding for a failing systemic RV?
- What's about the neurohormonal activation in ACHD?
- What is the effect of the resynchronization therapy on the RV patients?
- What is the effect of the exercise training on the systemic RV?
- What is the correlation between ventilator efficiency /aerobic capacity and the event-free survival in adults with atrial repair TGA?

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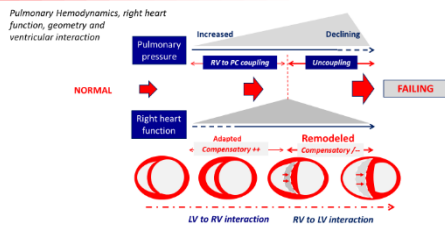
# Right heart structure and function

Characterization of Right Ventricular Remodeling in PAH by 3-Dimensional Wall Motion Tracking Echocardiography  
 Keiko Ryo et al. Circ Cardiovasc Imaging. 2015.



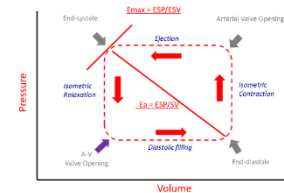
a particular attention to the coupling methodologies. In the main part of his speech, Prof. Grassi talked about very interesting and innovative techniques like the “REDUCE-LAP” that is a transcatheter intracardiac shunt developed for patients with heart failure with preserved ejection fraction, by presenting the preliminary results

Phenotyping the Right Heart Function and Structure Over Time



The structure and function of the right heart was the topic Prof. Guazzi spoke about in his lecture. The speaker coming from Milan (IT), started his talk, by presenting very interesting data on the history of the right ventricle starting from Leonardo! Going deeper in his lecture the speaker talked about anatomy, physiology and right ventricle functions with a

Ventricular-Arterial Coupling Defined by Emax/Ea Ratio



of a multicenter phase 1 trial. Finally, the speaker talked about another very interesting technique like the right heart function and structure phenotyping over time. In conclusion, Prof. Grassi pointed out that the interdependence of the left and the right ventricles, is one of the main important and also forgotten reasons leading to the RV dysfunction and a potentially early marker of an unfavourable clinical course.

- Why does the RV fail?
- What is the Berrheim effect and its reversal, based on the data presented by the speaker?
- How does the right heart work and how its work is better defined?
- Why does a loss in right heart function translate in a worse outcome?

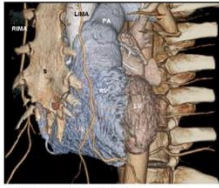
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# Echocardiographic assessment of the right ventricle

## Echo Assessment of the Right Ventricle

### Challenges for Tomographic Imaging of RV



- operator-dependent evaluation
- thin-walled chamber behind the sternum
- separate inflow and outflow portions
- asymmetrical, crescentic shape, wrapped around LV
- variations of shape with loading conditions
- heavily trabeculated

CT 3D Image courtesy of Dr F. Falga

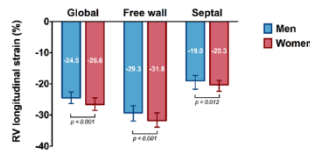
Badano LP et al. Eur J Echocardiogr 2010

The echocardiographic assessment of the right ventricle was the topic discussed by Prof. Muraru. At the beginning of her lecture the speaker, coming from Padua (IT) presented very interesting data on the measurements performed with the 2D echo modality, by highlighting that different probe positions can lead to different linear measurements. Going deeper in her lecture, Prof. Muraru spoke about the parameters to be quantified for the evaluation

of the right ventricle systolic function and highlighted the role played by the 3D echo modality. Prof. Muraru spoke also about automated algorithms developed for the 3D echo quantification and presented very interesting unpublished data on the all-cause mortality prediction in patients with different cardiac diseases thanks to the 3D echo modality measurements of the RV volumes. In the last part of her lecture, the speaker presented a huge amount of data on advanced echo and CMR imaging and spoke about other parameters like

## Echo Assessment of the Right Ventricle

### How to Interpret RV Strain Measurements for Clinical Purposes?



Take into account:

- Method (3- or 6-segments)
- Gender
- Vendor



Muraru D et al. Circ Cardiovasc Imaging 2016

the longitudinal vs radial RV shortening and on the RV longitudinal strain as an index of true ventricular deformation. In Conclusion, Prof. Muraru pointed out that the 3D echocardiography modality is superior to the conventional 2D echo technique for the RV size and systolic function evaluation, in terms of accuracy, reproducibility and outcome prediction.

## Echo Assessment of the Right Ventricle

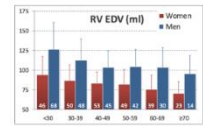
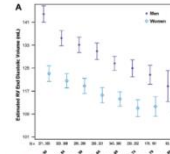
### Reference Values for RV Volumes Depend on Age, Gender and Body Size



n=441 healthy subjects



n=507 healthy subjects



Kawut SM et al. Circulation 2011

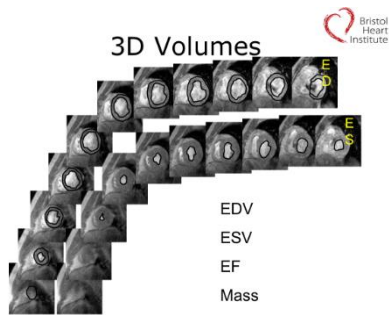
Maffessanti F, Muraru D et al. Circ Cv Img 2013

- Is the 3D echo accurate?
- How to interpret RV strain measurements for clinical purposes?
- What's about the automated software algorithms for 3D echo quantification based on the data presented by the speaker?
- What are the limitations of the RV diameters from the speaker point of view?
- What's about the advanced echo and the CMR imaging from the speaker point of view?
- What is the only true global measure of the RV function from the speaker point of view?

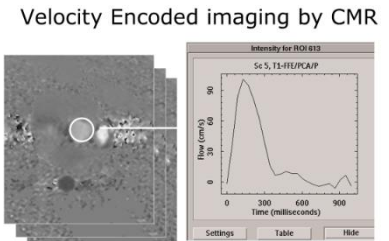
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# CMR assessment of the right ventricle

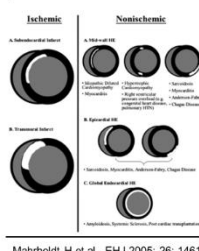


The CMR assessment of the right ventricle was the topic of the lecture discussed by Prof. Bucciarelli Ducci. The speaker, coming from Bristol (UK), introduced her talk by presenting very interesting data on the right ventricle function detected by CMR. Going deeper in her lecture, the speaker talked about the CMR flow detection and the velocity encoded imaging, by presenting very interesting data on the HASTE sequence, the SSFP cine, the aortic and the pulmonary



flow and on the pulmonary and tricuspid Valve diseases. In the main part of her lecture, the speaker talked about the CMR application in the scarring/fibrosis and ischemia detection, by presenting very interesting imaging data given by clinical cases. More in particular Prof. Bucciarelli Ducci spoke about the RV

involvement in the LV



cardiomyopathies and about the differential diagnosis thanks to the CMR application. Finally, the speaker talked about Pulmonary hypertension and other topics like the ECV mapping and the 4D flow applications. In conclusion, Prof. Bucciarelli Ducci, pointed out that CMR is a very affordable technique for the detection of the main physiological and pathological images of the RV.

- What 's about the CMR functional detection of the RV?
- What's about the measurements of the right ventricular volumes from the speaker point of view?
- What are the additional RV focused images presented by the speaker?
- What's about CMR detection of RV scarring/fibrosis?
- What are the main characteristics of MRI images in ARVC?

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# Why and how to assess the right atrium?

**RA Size**

**RA Linear Dimensions**

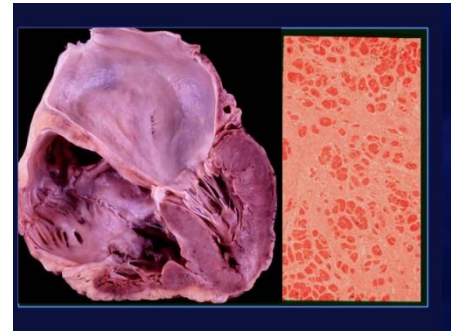
The minor axis of the right atrium should be measured in the apical 4-chamber view as the distance between the lateral right atrial wall and inter-atrial septum, at the mid-atrial level defined by half of right atrial long axis.

**RA Volume**

2D volumetric measurements are usually based on tracings of the blood-tissue interface on the apical 4-chamber view. At the tricuspid valve level, the contour is closed by connecting the two opposite sections of the tricuspid ring with a straight line. Volumes can be computed by using either the single plane area-length or the disc summation technique.

	Women	Men
RA minor axis dimension (cm/m <sup>2</sup> )	1.9 ± 0.3	1.9 ± 0.3
RA major axis dimension (cm/m <sup>2</sup> )	2.5 ± 0.3	2.4 ± 0.3
2DE right atrial volume (ml/m <sup>2</sup> )	21 ± 6	25 ± 7

Prof. Khandheria spoke about the assessment of the right atrium and more in particular on the rationale and the time to perform it. The speaker, coming from Milwaukee (USA), started his lecture, by presenting data on the main abnormalities of the RA, the remodelling in atrial fibrillation, cardioversion, the pulmonary hypertension and on the



congenital heart disease. Going deeper in his lecture Prof. Khandheria spoke about the methodologies for the right atrium investigation like echocardiography, cardiac CT and cardiac MRI and highlighted that in the ASA guidelines only few lines of more than 60

pages have been dedicated to right atrium. In the main part of his presentation, the speaker talked about RA physiology, by presenting very interesting data on the reservoir, conduit and contractile phases. In conclusion, Prof. Khandheria pointed out, that the Right Atrium is a forgotten chamber but starting from now the studies on right atrium are to be growing in the view of a global interaction between right atrium and right ventricle.

**How common is Right atrial appendage thrombus in Atrial fibrillation ?**

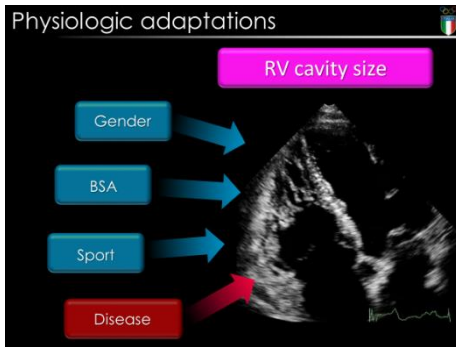
Real time 3D echo of RAA: Image source Francesco F et al. J Am Coll Cardiol. 2010;3(9):966-975

- Why is heart atrium important from the speaker point of view?
- What is the main limitation of the present techniques for the detection of the RA images?
- How common is the right atrial appendage thrombus in atrial fibrillation?
- What's about the recommendations for cardiac chamber quantification by echocardiography in adults from the speaker point of view?
- What's about the right atrium physiology based on the data presented by the speaker?

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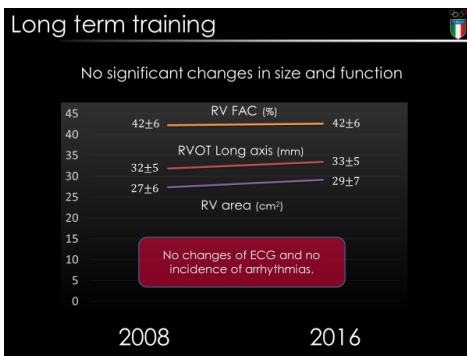
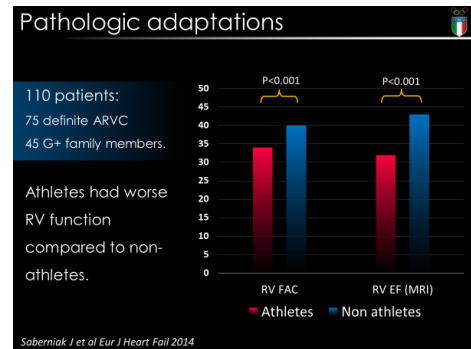
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# Right ventricular adaptation and maladaptation to exercise



The mechanisms of adaptation and maladaptation to exercise of the right ventricle was the topic of Prof. Caselli presentation. The speaker, coming from Rome (IT), talked about the physiological adaptations of the right ventricle starting from the wall thickness and the cavity size. Going deeper in his lecture, Prof. Caselli presented very interesting data on a study running in 1011 Olympic athletes, designed for the

detection of the RV adaptation from an anatomical and functional point of view. In the main part of his presentation the speaker presented very interesting data on arrhythmogenic cardiomyopathy in patient with positive mutations for cardiomyopathy and in subject without any mutation and discussed the data given by



mouse experimental models, where the animals exposed to an intensive exercise developed RV fibrosis and arrhythmias. Finally, Prof. Caselli talked about the limits of the maladaptation hypothesis and presented very interesting data given by a study performed on Italian Olympic athletes subjected to very strong exercise activities along the years. In conclusion, the speaker pointed out that currently there are no sufficient evidence for a pure exercise-induced cardiomyopathy.

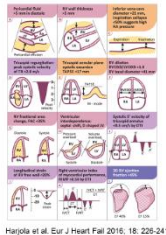
- What's about the gender impact on the right ventricle adaptation to exercise?
- Is too much sport exercise harmful for the heart?
- What are the main limitations of the maladaptation hypothesis?
- What's about the data on the Italian Olympic athletes presented by the speaker?

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# Epidemiology and clinical significance

Echocardiographic parameters in the assessment of right ventricular failure

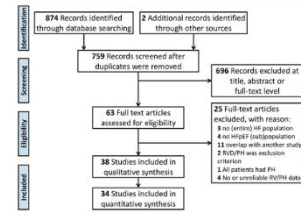


Hajjola et al. Eur J Heart Fail 2016; 18: 228-241

Prof. Metra, coming from Brescia (IT) spoke about the epidemiology and the clinical significance of the right ventricular failure, by presenting very interesting data given by the specific guidelines. Going deeper in his lecture the speaker talked about causes, epidemiology and prognostic features. More in particular Prof. Metra presented data on the main mechanisms of RV failure, by highlighting the importance of ischemia,

infarction, cardiomyopathy and RV valve diseases as the main causes of the right ventricular dysfunction. Speaking about epidemiology, Prof. Metra presented very interesting data on predictors of the RV function in patients with HF with preserved and reduced EF. In the last part of his lecture, the speaker talked about the prognostic significance of the right ventricular dysfunction in patients affected by pulmonary hypertension and HF with preserved EF, by presenting very interesting data on the

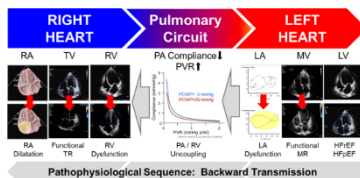
RV dysfunction in heart failure with preserved ejection fraction: a systematic review and meta-analysis



Gorter et al. Eur J Heart Fail 2016; 18: 1472-1487

pathophysiological mechanisms shared in patients with right ventricular dysfunction and pulmonary hypertension. Finally, Prof. Metra talked about new parameters for a better detection of the prognostic significance of the presence of the right ventricular dysfunction, by presenting data on pulmonary arterial capacitance. In conclusion, Prof. Metra pointed out that from the prognostic point of view an integrated assessment of the RV-arterial coupling yields the best prognostic prediction.

Sequence of pathophysiological factors contributing to pulmonary hypertension in left ventricular heart failure



Rosenkranz et al. Eur Heart J. 2015;37(12):942-954. doi:10.1093/eurheartj/ehv512

- What are the main mechanisms leading to RV failure?
- What are the main practical rules for women with bleeding during CHC presented by the speaker?
- What's about the cardiopulmonary interaction and pathobiology of pulmonary hypertension?
- What are the determinants of RV dysfunction?
- What's about the changes of the right ventricular function from a prognostic point of view?

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# Acute right ventricular failure

## Ventricular interdependence

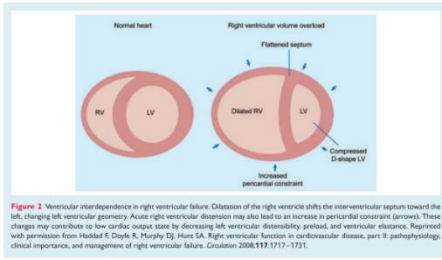


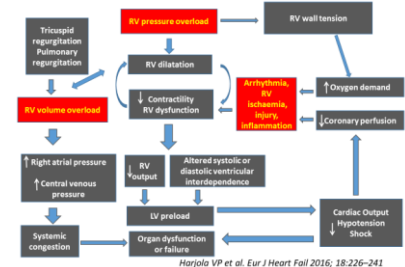
Figure 2. Ventricular interdependence in right ventricular failure. Dilatation of the right ventricle shifts the interventricular septum toward the left, changing left ventricular geometry. Acute right ventricular dilation may also lead to an increase in pericardial pressure (arrows). These changes may contribute to low cardiac output state by decreasing left ventricular distensibility, preload, and ventricular elastance. Reprinted with permission from Haddad F, Doshi K, Murphy DJ, Hane SA. Right ventricular function in cardiovascular disease, part II: pathophysiological importance, and management of right ventricular failure. *Circulation* 2008;117:1717-1731.

Haddad F et al. *Circulation* 2008;117:1717-1731.

data on the differences between right and left ventricular response to the increasing afterload and preload from a ventricular interdependence point of view. In the main part of his presentation Dr. Harjola spoke about the causes and the differential diagnosis of acute RV failure, by presenting very interesting data on acute pulmonary embolism, acute RV

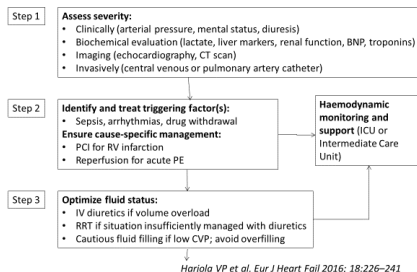
The acute right ventricular failure was the topic at the core of Dr. Harjola presentation. The speaker, coming from Helsinki (FI), at the beginning of his talk, addressed the audience by highlighting that this one is a very complicated topic from the basic physiology and the intensive care point of view. Going deeper in his lecture, Dr, Harjola spoke about pathophysiology, by highlighting that acute RV is a syndrome with multiple aetiologies and presented experimental

## Pathophysiology of acute RV failure



Harjola VP et al. *Eur J Heart Fail* 2016; 18:226-241

## Management of acute RV failure



Harjola VP et al. *Eur J Heart Fail* 2016; 18:226-241

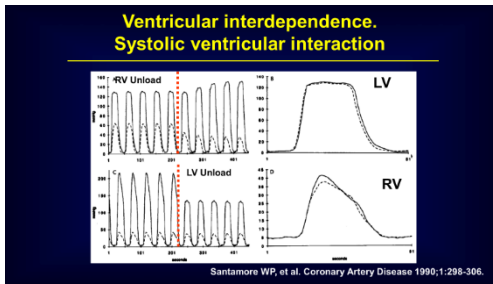
infarction, pulmonary hypertension and on acute RV failure in intensive care patients. In the last part of his lecture the speaker talked about the management of the acute RV failure, by discussing on six steps, characterized by the assessment of severity, the identification and the treatment of the triggering factor, the optimization of the fluid status, the maintenance of the arterial pressure, the use of inotropies for reducing the cardiac filling pressure and finally the use of further measures for the afterload reduction.

- What's about the management of acute RV failure from the speaker point of view?
- What are the main steps of the protective ventilation strategy?
- What's about the acute RV failure in intensive care patients?
- What's about the acute RV failure infarction?
- What are the main steps of the risk-adjusted management of patients affected by acute pulmonary embolism?

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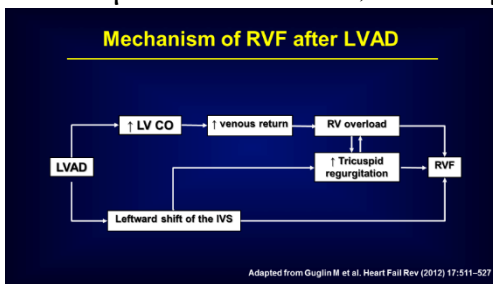
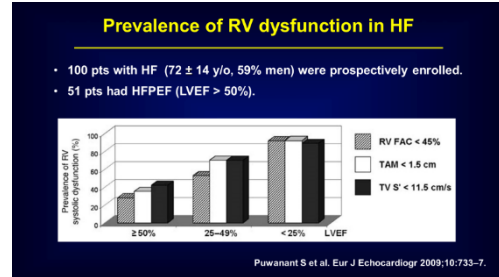
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# The right ventricle failure in left heart failure patients



The right ventricle failure in left heart failure patients was the topic discussed by Prof. Popescu. The speaker, coming from Bucharest (RO), presented very interesting data about the ventricular interdependence and the systolic ventricular interaction. Going deeper in his talk Prof. Popescu presented data on the significant LV contribution to RV systolic

function and on the contributions given by the motion of the intraventricular septum. In the main part of his presentation, the speaker talked about the prevalence of RV dysfunction in HF and its prognostic value, by presenting very interesting data given by clinical trials on HF patients with right ventricular dysfunction. In the last part of his lecture, Prof. Popescu presented very interesting data on the treatment of



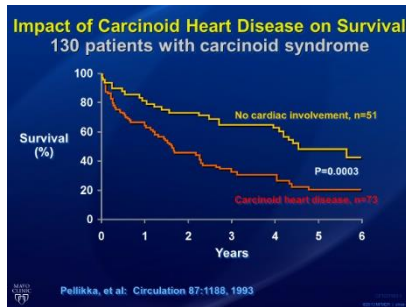
patients affected by right ventricular dysfunction and HF with preserved and reduced EF. Finally, the speaker spoke about the risk of right ventricular dysfunctions in patients undergoing LVAD. In conclusion, Prof. Popescu pointed out that right ventricular dysfunction is a very important aspect in patients with HF, that significantly affects their outcome.

- What is the relationship between the prevalence of RV dysfunction and the prevalence of HF?
- What's about the treatment of the right ventricular dysfunction patients?
- What is the mechanism leading to the right ventricular dysfunction in patients undergoing LVAD?

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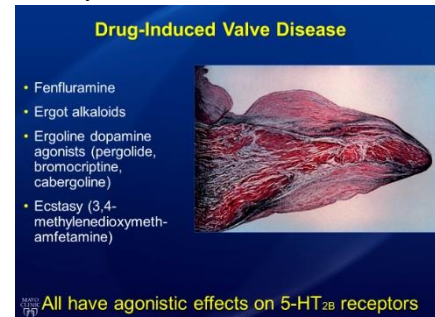
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# Carcinoid Heart Disease: the Right Ventricle and Valves

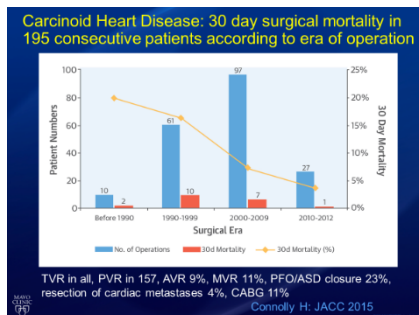


Prof. Pellikka, talked about the carcinoid heart disease and the involvement of the right ventricle and the valves. The speaker coming from Rochester (USA), started her speech by highlighting the impact of carcinoid heart disease on survival. Going deeper in her lecture the speaker talked about the carcinoid syndrome, its initial presentation, the main symptomatology from fatigue to edema and ascites and about echocardiography

as the main procedure for a very effective diagnosis. In the main part of her lecture, Prof. Pellikka presented very interesting data on the similarity of the mechanism of action between carcinoid and drugs in inducing valve disease and



spoke about the progression of the disease, by highlighting the role of the right ventricle from a prognostic point of view. In the last part of her talk, Prof. Pellikka spoke about management and presented data on the mortality due to surgery. In conclusion, the speaker pointed out that a multidisciplinary team composed by physicians with different specializations is mandatory for a better management of the patients.

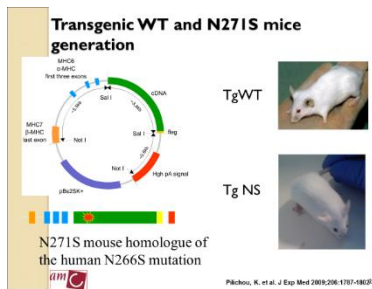


- What are the main examinations to be performed for a correct carcinoid diagnosis?
- What are the main agents useful for the management of Carcinoid HD patients?
- What are the main professionals to be involved in the multidisciplinary team indicated by the speaker?
- What is the effect of the valve replacement in patients with severe carcinoid valvular heart disease?

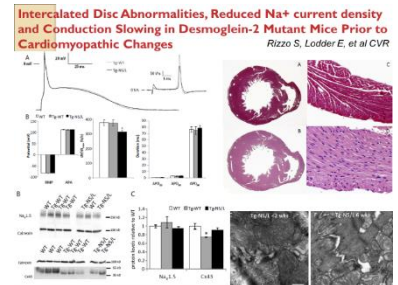
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# ARVC advances at the University of Padua



Prof. Basso from Padua (IT), spoke about the ARVC advances at the University of Padua, by presenting very interesting data on the arrhythmogenic right ventricular cardiomyopathy, that is a congenital disease studied in this University. More in particular, the speaker talked about the history of this disease, the genetic origin of ARVC, the recapitulation of this disease in transgenic mice, the advances in diagnosis and finally on the risk stratification and the prevention of SD. Going deeper in her lecture, Prof. Basso presented very interesting data on the genes involved in the development of this disease, by highlighting that ARVC is a desmosomal disease. Speaking about the recapitulation of ARVC in mice, the speaker presented very interesting data given by the pre-clinical studies running in these animals and highlighted that in humans there are the same cardiac lesions seen in the mice. From the diagnostic point of view, Prof. Basso presented very impressive data given by clinical cases managed in Padua and finally, spoke about an ongoing research project, by presenting data on DPS zebrafish.



**AC:**  
**Ongoing Research Projects**

- Biomarkers (miRNA)
- iPSCs
- Knock in DSP mouse
- DSP zebrafish

- What are the main genes involved in the development of ARVC?
- What's about ARVC as a progressive myocardial dystrophy based on the data presented by the speaker?
- What is necessary to do when a patient with a desmo mutation is discovered?
- What's about DSP zebrafish based on the data presented by the speaker?
- What's about the IETA studies discussed by the speaker?

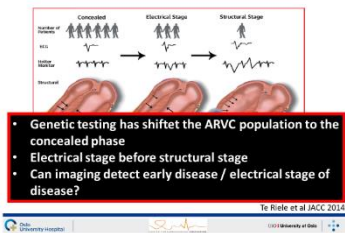
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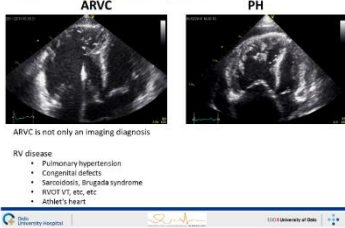
# Guidelines about cardiac imaging assessment

Disease progression in ARVC

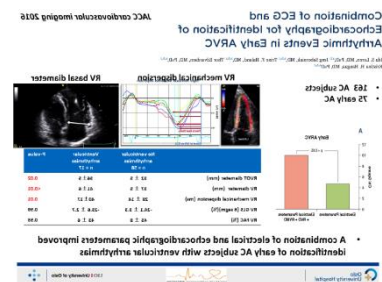


measurements can help in the detection of the reduced RV function.

RV disease, differential diagnoses



The guidelines about the cardiac imaging assessment was the topic discussed by Prof. Haugaa in her lecture. The speaker coming from Oslo (N), talked about the main criteria for the diagnosis of ARVC. Going deeper in her lecture, Prof. Haugaa presented imaging data on the stratification of these patients, by highlighting that in specific cases the strain measurements can help in the detection of the reduced RV function. In the main part of her talk, the speaker presented many clinical cases of ARVC patients assessed on the basis of the 2010 task force criteria and spoke about the best follow-up procedures for these patients and about the differential diagnosis with other cardiac diseases. In conclusion, the speaker pointed out that imaging by echo and CMR are important tools for a correct TFC ARVC diagnosis.



- What's about the ICD treatment based on the data presented by the speaker?
- What's about the diagnostic performance of imaging in ARVC using the 2010 Task Force criteria?
- How often and what to look for the follow-up of ARVC patients?
- What's about the differential diagnosis of the RV disease?
- What is the correct age for the first CMR in positive genetic babies from the speaker point of view?

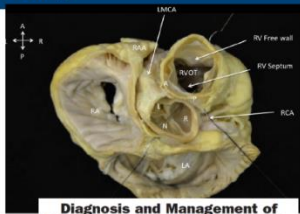
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# Idiopathic RVOT tachycardia

## Anatomy



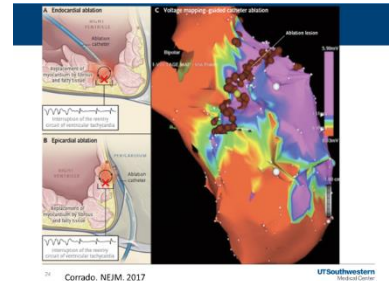
Diagnosis and Management of Idiopathic Ventricular Tachycardia

Kurt S. Hoffmayer, PharmD, MD, and Edward P. Gerstenfeld, MD

(Curr Probl Cardiol 2013;38:131-158.)

UT Southwestern Medical Center

The main topic at the core of Prof. Link presentation, was the idiopathic RVOT tachycardia. The speaker, coming from Dallas (USA), presented very interesting data on RVOT, LVOT, ARVC ventricular tachycardia and on athletes with epicardial ventricular tachycardia. Speaking about RVOT Prof. Link, highlighted that this is the most common idiopathic ventricular tachycardia in women and that it may be the



Corrado, NEJM, 2017

UT Southwestern Medical Center

first sign of an underlying cardiomyopathy. In the main part of his lecture, the speaker presented data on the clinical and electrophysiological spectrum of the LVOT tachycardia, on

ARVC and on the ECG

comparison between RVOT and ARVC leading to the development of a score model for a correct diagnosis. Prof. Link presented also data on the 3D electroanatomical voltage mapping compared to CMR, showing that there is a very good correlation between these two techniques. Finally, the speaker, talked about athletes affected by ventricular arrhythmias and presented very interesting data on the relationship between VT and the endurance athletes.

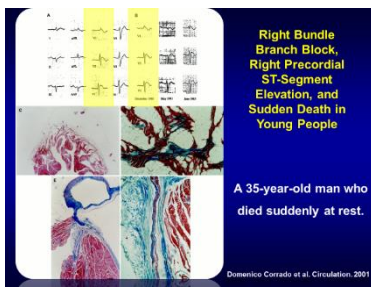
TABLE 1 Baseline Characteristics				
	All Patients (N = 473)	Group A (n = 449)	Group B (n = 24)	P Value*
Age, yrs	48 ± 16	49 ± 16	42 ± 15	0.152
Male	47 (10)	48 (11)	9 (38)	0.851
ICD (before ablation)	31 (6.6)	27 (6.0)	4 (16.7)	0.382
White/black/Asian	54/217	43/217	11/10/0	0.685
NYcturbulent patient	146 (30.9)	140 (30.9)	6 (25.0)	0.201
First presentation				
DCHA	6 (1.3)	6 (1.3)	0	0.205
Pre-syncope	18 (3.8)	17 (3.8)	1 (4.2)	0.668
Palpitations	26 (5.5)	23 (5.1)	3 (12.5)	0.174
Other	7 (1.5)	5 (1.1)	2 (8.3)	0.507
Total	28 (5.9)	27 (6.0)	11 (45.8)	0.001
ECG characteristics				
First documented VA	52 (11.0)	41 (9.1)	11 (45.8)	0.212
VT	278 ± 37	283 ± 39	297 ± 22	0.043
VT cycle length, ms				
Ventricular tachycardia				
Superior axis	12 (2.5)	12 (2.6)	0	0.205
Inferior axis	18 (3.8)	17 (3.8)	1 (4.2)	0.668
Both axes	26 (5.5)	23 (5.1)	3 (12.5)	0.174
Total	27 (5.7)	26 (5.8)	11 (45.8)	0.001
Endurance athletes	15 (3.2)	14 (3.1)	1 (4.2)	0.209
Endurance athletes	15 (3.2)	14 (3.1)	1 (4.2)	0.209
Training, yrs	15 (6.2)	14 (6.2)	1 (10.0)	0.209
MET-hrs	2,813	2,342	5,405	0.001
MET-hrs (IQR)	(888, 5,171)	(607, 3,847)	(6,270, 12,840)	0.001
Family history of ARVC	14 (2.9)	14 (3.1)	0	0.205
ARVC associated	14 (2.9)	14 (3.1)	0	0.205
Non-associated	23 (4.9)	23 (5.1)	0	0.002
Total	28 (5.9)	28 (6.0)	0	0.001
Any pathologic	29 (6.1)	29 (6.4)	0	0.001

- What does happen to the ejection fraction when PVCs are corrected with the ablation?
- What's about the ablation outcomes for RVOT VT?
- What are the main electrophysiological characteristics of ARVC?
- What are the main imaging technique applied in the diagnosis of these patients?

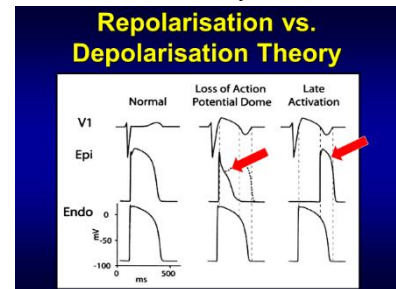
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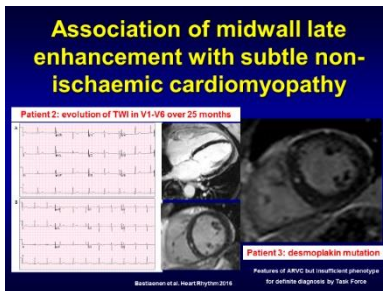
# ARVC and Brugada syndrome: what is new?



The main topics at the core of Prof. Behr presentation, were the novelties about ARVC and the Brugada syndrome. The speaker, coming from London (UK), presented very interesting data, starting from a new consensus document on the J-wave syndromes. Going deeper in his lecture, Prof. Behr spoke about the main ECG patterns indicative for Brugada syndrome, starting from the history of the



development of the disease mechanism of action. In the main part of his lecture, the speaker talked about the procedures of repolarization through the application of RV canine models and about the identification of the mutations affecting the  $I_{T_o}$  KCNE5/KCND3. Prof. Behr spoke also about RVOT, embryology, the connexin43 protein and the related hypothesis on the development of the Brugada syndrome. More in particular Prof. Behr presented very interesting data given by a clinical study running in his center on Brugada syndrome patients and their relatives. Finally, the speaker talked about connexin43 and the open epicardial ablation and the changes in the ECG pattern after ablation.



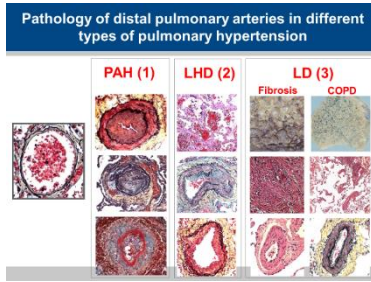
- What's about the hypothesis on the onset of the Brugada syndrome due to fibrosis and altered gap junction expression in RVOT?
- What's about the morphometric collagen analysis?
- What are the main left ventricular late enhancement patterns presented by the speaker?
- What is the association of the midwall late enhancement with subtle non-ischaemic cardiomyopathy?

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# Pulmonary arterial hypertension in 2017: classification and prognostication

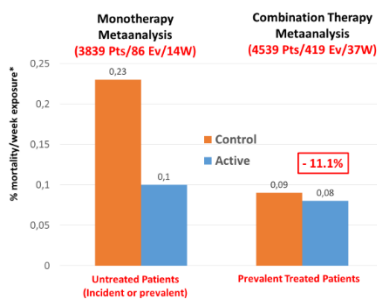


Prof. Galie', coming from Bologna (IT), spoke about the classification and the prognostication of the pulmonary arterial hypertension in 2017, by highlighting at the beginning of his presentation that PH is not a disease but a pathophysiological condition and can be found in more than 50 diseases. Going deeper in his speech, Prof. Galie' talked about the PH clinical classification and presented very interesting data on the pathology of the distal pulmonarias arteries. In the main part of his lecture, the speaker presented data on the survival incidence dived into three clinical classification groups and discussed about an algorithm for the PH diagnosis. Prof. Galie' presented very interesting data on the pulmonary arterial hypertension, its treatment algorithm and on its risk assessment. Finally, talking about survival the speaker presented very impressive data on the prognostic value of the RV remodelling models based on echo parameters given by clinical studies performed in his clinical center.

**Management of pulmonary hypertension in left heart disease**

Recommendations	Class <sup>a</sup>	Level <sup>b</sup>
Optimization of the treatment of the underlying condition recommended before considering assessment of PH-LHD (i.e. treat structural heart disease)	I	C
It is recommended to identify other causes of PH (i.e. COPD, SAS, PE, CTEPH) and to treat them when appropriate before considering assessment of PH-LHD	I	C
It is recommended to perform invasive assessment of PH in patients on optimized volume status.	I	C
Patients with PH-LHD and a severe pre-capillary component as indicated by a high DPG and/or high PVR should be referred to an expert PH center for a complete diagnostic work-up and an individual treatment decision.	IIa	C
The importance and role of vasoreactivity testing is not established in PH-LHD, except in patients who are candidates for heart transplantation and/or LV assist device implantation.	III	C
The use of PAH approved therapies is not recommended in PH-LHD	III	C

www.escardio.org Galie' H. et al. Eur Heart J 2015; 36: 3092-3103



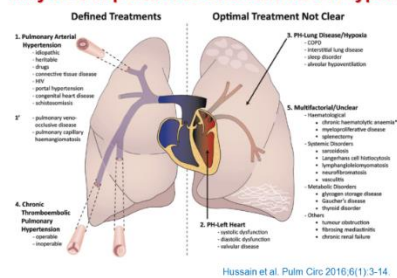
- What's about the clinical classification of PH from the speaker point of view?
- What is the survival incidence of the PH patients according to the three clinical groups?
- What's about the PAH prognosis based on the data presented by the speaker?
- What's about the RV remodelling and the Prognosis among and within PAH types?

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# Is cardiac catheterization really needed to differentiate Group I from Group II patients?

## Why is it important to differentiate PH type?



Hussain et al. *Pulm Circ* 2016;6(1):3-14.

The question about the need to perform the cardiac catheterization with the aim to differentiate group I from group II patients, was the topic at the core of Prof. Linhart presentation. The speaker coming from Prague (CZ), at the beginning of his presentation talked about the haemodynamic definitions of pulmonary hypertension and on the importance of a definitive PH

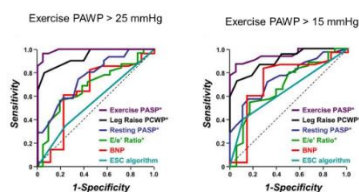
differentiation from the phenotype point of view. Going deeper in his lecture, the speaker presented very interesting imaging data

## Echocardiographic probability of pulmonary hypertension in symptomatic patients with a suspicion of pulmonary hypertension

Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs'	Echocardiographic probability of pulmonary hypertension
≤ 2.8 or not measurable	No	Low
≤ 2.8 or not measurable	Yes	Intermediate
2.9 - 3.4	No	Intermediate
2.9 - 3.4	Yes	High
>3.4	Not required	High

Galè N et al. *Eur Heart J* 2016; Jan 1;37(1):67-119

## Exercise induced LVEDP increase for unmasking HF-pEF



Borlaug BA et al. *Circulation: Heart Failure*. 2010;3:588-595

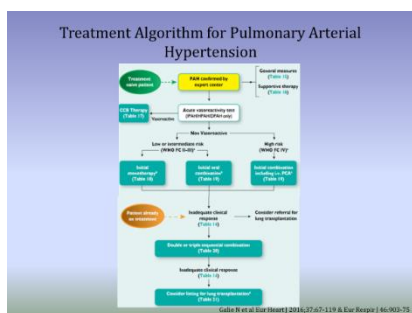
on PH patients with the RV and the LV phenotype and spoke about a very effective scoring system for the prediction of the pulmonary vascular disease. In the main part of his presentation, the speaker talked about another phenotype, the so called left heart failure with CPC-PH and pronounced RV overload and presented very interesting imaging data on the handgrip influence of the LV filling. Finally, Prof. Linhart spoke about those patients who may benefit from RHC.

- Why is it important to differentiate the PH types?
- What's about the echocardiographic probability of the PH with RV phenotype?
- What are the typical characteristics of the RV phenotype in these patients?
- Is there an opposite phenotype-PAH with left heart disease?
- Can loading conditions change the "label"?
- Who may benefit from RHC?

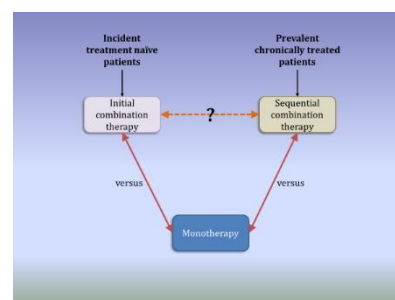
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# Evolving treatment paradigm in pulmonary hypertension: aggressive upfront therapy or sequential addition of drugs

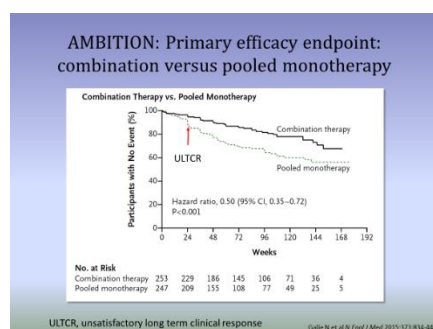


The Evolving treatment paradigm in pulmonary hypertension: aggressive upfront therapy or sequential addition of drugs, was the topic at the core of Prof. Gibbs presentation. The speaker coming from London (UK), presented very interesting data on treatment, starting from an algorithm of the pulmonary arterial hypertension therapy. Going deeper in his presentation, the speaker



talked about the initial combination strategies versus the sequential combination therapy both compared to monotherapy, by presenting very interesting data given by a meta-analysis on 16 trials of sequential combination therapy and 2 trials on initial combination therapy. In the main part of his lecture, Prof. Gibbs spoke about the Ambition trial comparing monotherapy vs initial combination therapy and its combined endpoint including

also an unsatisfactory long-term response after at least 6 months of randomized therapy, by highlighting that the initial combination therapy is better than the monotherapy in all the studied outcomes. Finally, the speaker presented data on the initial triple combination therapy. In conclusion, Prof. Gibbs pointed out that compared to monotherapy, the combination therapy reduces the incidence of the combined clinical worsening endpoints without any effect on mortality.



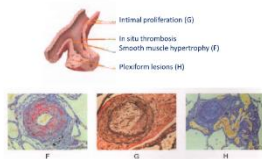
- What are the main drugs recommended in the treatment algorithm presented by the speaker?
- What is the initial combination therapy for PAH?
- What is the effect of the combination therapy on the mortality for all causes, based on the data presented by the speaker?
- What's about the initial triple combination therapy from the speaker point of view?

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# Pathophysiology and treatment of chronic thromboembolic pulmonary hypertension

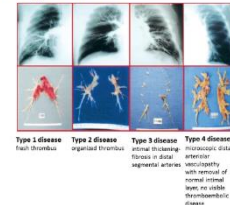
## 'Open Vessel' theory



The Pathophysiology and the treatment of the chronic thromboembolic pulmonary hypertension was the topic Prof. Pengo talked about. The speaker coming from Padua (IT), at the beginning of his lecture talked about CTEPH, its classification and its pathophysiology based on many hypotheses like the

antiphospholipid antibodies and the open vessel theory proposed by Moser and Braunwald in 1973. Going deeper in his lecture, Prof. Pengo spoke about epidemiology and the independent risk factors for CTEPH. In the main part of his presentation, the speaker talked about treatment, by presenting data on the pulmonary

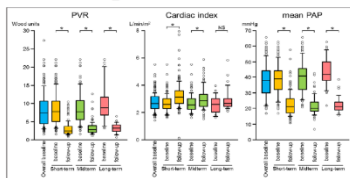
## CTEPH: Surgical Classification



Fransoletto M, et al. Journal of Internal Medicine. 2012

thromboendarterectomy as the treatment of choice and other procedures like medical treatment, balloon pulmonary angioplasty and lung transplantation. Speaking about drugs Prof. Pengo highlighted that with the exception of Riociguat, the medical therapy does not produce any improvement in the exercise capacity. In conclusion, the speaker pointed out that CTEPH is a potentially treatable disorder and PEA is the treatment of choice.

## Long-Term Outcomes



Circulation. 2016;134:2030-2032.

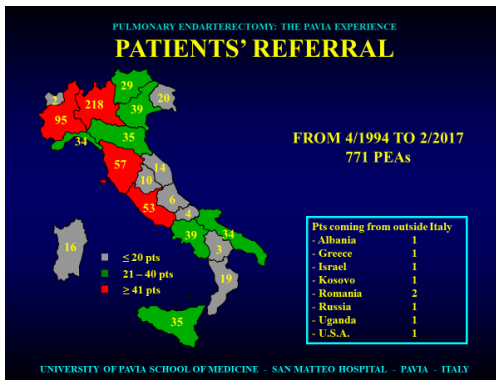
- What is CTEPH from the speaker point of view?
- What are the main pathophysiological hypothesis of CTEPH presented by the speaker?
- What are the main risk factors for CTEPH presented by the speaker?
- What is the main CTEPH treatment procedure presented by the speaker?
- What's about the medical treatment of CTEPH?
- What's about balloon pulmonary angiography based on the data presented by the speaker?

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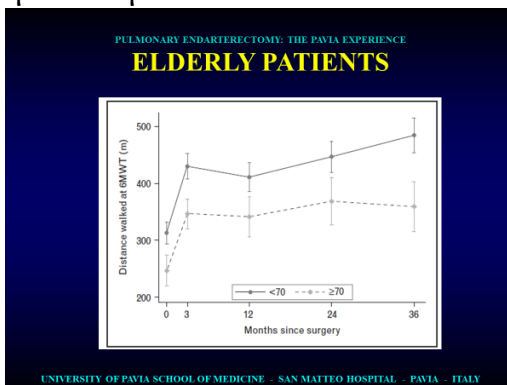
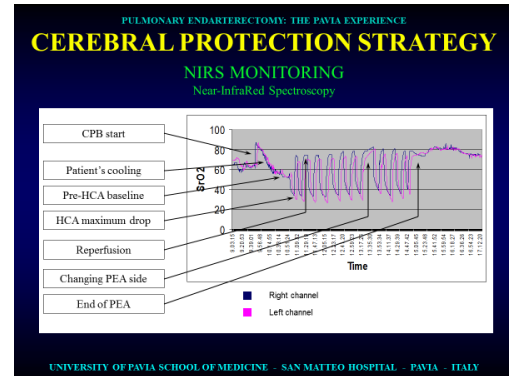
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# Technique and outcomes of pulmonary endarterectomy surgery. How to select the right patient?



and the related DLTx procedure. Speaking about indications for surgery, Dr. D'Armini presented very interesting data on clinic, hemodynamic and anatomy that are the parameters of reference for the surgical treatment and for the choice of the right type of surgery. Talking about the surgical treatment, the speaker pointed out that in his center they have



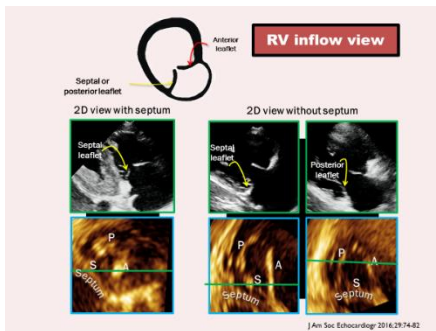
changed the original San Diego protocol in a less invasive surgical procedure, the so-called Pavia protocol, more effective for the cerebral protection and characterized by the use of minimally-invasive surgical instruments. Dr. D'Armini pointed out that thanks to the development of this technique, the distal pulmonary lesions are now operable. Finally, the speaker presented data on the results in elderly patients, by highlighting that the good results obtained in these patients depend on their correct channel selection.

- What's about transplants and conservative surgery from the speaker point of view?
- What are the main differences about the San Diego and the Pavia protocols?
- What's about the surgical trips and tricks presented by the speaker?
- What's about the operability of the distal lesions?
- What are the main topics of the Pavia CTEPH program presented by the speaker?

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# The normal tricuspid valve



Prof. Addetia talked about the normal tricuspid valve. The speaker coming from Chicago (USA), presented very interesting data on the Imaging features of the 3D echocardiography applied in the diagnosis of the tricuspid valve disease. At the beginning of her lecture, Prof. Addetia spoke about the differences between 2D and 3D echocardiography for the detection of the

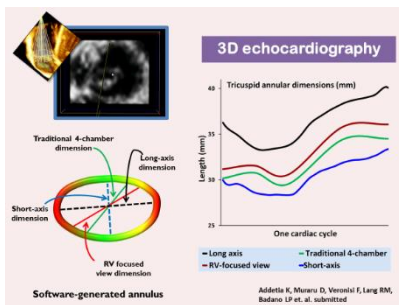
real tricuspid valve structure. Going deeper in her presentation the speaker talked about the main features obtained with the 2D imaging and the advantages of the 3D echocardiography for the correct detection of all the leaflets and the other structures of the tricuspid valve. In the main part of her lecture, Prof. Addetia presented very interesting imaging and anatomic data on

**THE TRICUSPID VALVE COMPLEX**

1. Three leaflets
  - Anterior
  - Septal
  - Posterior
2. Fibrous annulus
3. Chordae tendinae
4. Papillary muscles
5. RA myocardium
6. RV myocardium

Courtesy Dr. Stephen P. Sanders, Professor of Pediatrics (Cardiology), Harvard Medical School

the real structure composing the tricuspid valve. In the last part of her talk, the speaker presented very interesting data on the tricuspid valve in pulmonary hypertension patients. Finally, Prof. Addetia presented data on the correlation between 2D, 3D and intra-operative measurements of the tricuspid valve annulus. In conclusion, Prof. Addetia pointed out that the 3D echocardiography has opened the way for a better detection of the real tricuspid valve



structure and composition.

- How many leaflets does the tricuspid valve have?
- What are the structures that compose the tricuspid valve?
- What's about 3D for the tricuspid valve measurements?
- What's about the correlation between the annulus measurements with 2D, 3D echocardiography and the intra-operative measurements?

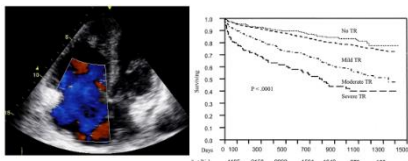
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# Pathophysiology of functional tricuspid regurgitation

## PATHOPHYSIOLOGY OF FUNCTIONAL TR

Tricuspid regurgitation and survival



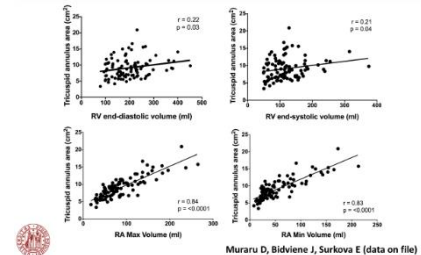
Nath, J. et al. J Am Coll Cardiol 2004  
Shiran A et al, J Am Coll Cardiol 2009

Prof. Badano talked about the pathophysiology of functional tricuspid regurgitation. The speaker coming from Padua (IT), at the beginning of his presentation addressed the audience with the observation that people dyes for the tricuspid regurgitation, but despite this, the tricuspid valve is the Cinderella valve from a research point of view. Going deeper in his lecture, Prof. Badano presented very interesting data on new considerations about the pathophysiology

of the tricuspid valve through the 3D imaging technique. In the main part of his talk, Prof. Badano spoke about the tricuspid annulus in functional regurgitation, by presenting very interesting data given by a computer reconstruction imaging study. Prof. Badano presented also interesting data on the measurements of the tricuspid annulus by 2D and on its changes in shape in the functional regurgitation.

## PATHOPHYSIOLOGY OF FUNCTIONAL TR

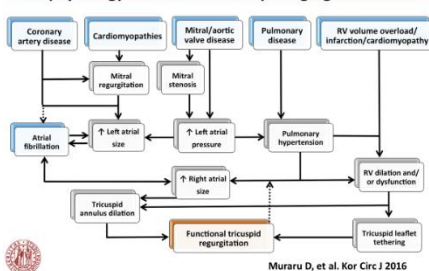
Tricuspid annulus in functional tricuspid regurgitation



In the last part of his lecture, the speaker presented very innovative imaging data on the tricuspid annulus in the functional regurgitation state, given by a clinical study running in his center with the aim to understand the main mechanisms of the functional tricuspid regurgitation. Finally, Prof. Badano presented very interesting data on the valve tenting in functional regurgitation. In conclusion, the speaker pointed out that the 3D echocardiography offers unique opportunities to quantitate the valve geometry and the relative importance of its components in the development of the functional regurgitation.

## PATHOPHYSIOLOGY OF FUNCTIONAL TR

Pathophysiology of functional tricuspid regurgitation in 2016



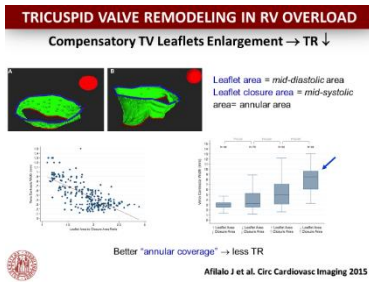
Muraru D, et al. Kor Circ J 2016

- What's about the 3D assessment of the TR functional state?
- What's about the TR apparatus assessment through 3D Imaging?
- What are the main mechanisms of the papillary muscles movements in the functional regurgitation?
- How to measure the tricuspid annulus with 2D technology?
- What's about the dedicated software for TV analysis by transthoracic 3D?
- What are the main mechanisms of functional tricuspid regurgitation?
- What's about the valve tenting in functional regurgitation, based on the data presented by the speaker?

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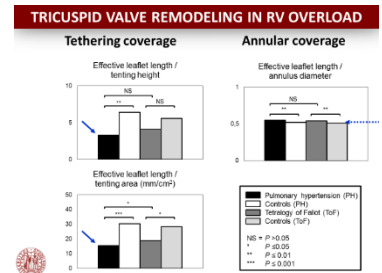
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# Tricuspid valve remodeling in right ventricular overload



Tricuspid valve remodeling in right ventricular overload was the topic Prof. Badano talked about. The speaker coming from Padua (IT), presented very interesting data on the variability of the TR severity in pulmonary hypertensive patients, based on the contraction and the planimetry of the leaflets covering the annulus of the tricuspid valve. Going deeper in

his lecture, the speaker presented very interesting data given by a 3D imaging study on patients affected by pulmonary hypertension and tricuspid regurgitation with the aim to assess the multiple components of the TV



remodeling in various forms of RV overload. The speaker presented and discussed a huge amount of data with the intention to explain the main correlations between the tricuspid changes in patients with various forms of RV overload. In conclusion, Prof. Badano pointed out that the TR is more severe in patients with RV pressure overload, than in patients with

volume overload.

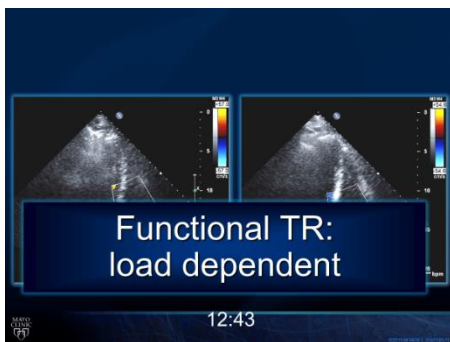
- What's about the 3D echocardiographic measurements in patients with tricuspid valve remodeling in RV overload presented by the speaker?
- What's about the coaptation length in pulmonary hypertension patients compared to controls?
- What's about the determinants of the multivariate analysis presented by the speaker?

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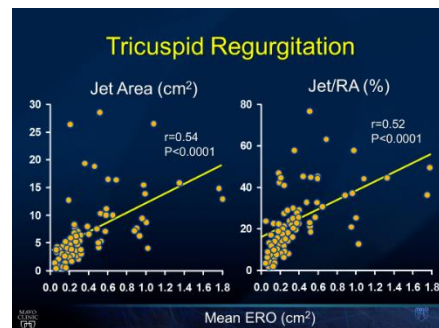
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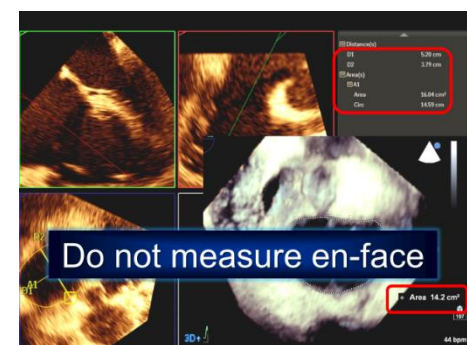
# Assessment of tricuspid regurgitation severity



The Assessment of tricuspid regurgitation severity was the topic of Prof. Pislaru presentation. The speaker coming from Rochester (USA), presented very interesting data on the so called forgotten valve as the tricuspid valve is. Going deeper in his presentation Prof. Pislaru spoke about tricuspid regurgitation and its evaluation depending on high quality imaging. In the



main part of his presentation, the speaker talked about the TR assessment, by presenting very interesting data on the mechanisms, the severity and the haemodynamic impact of the regurgitation. More in particular Prof. Pislaru highlighted the main differences between the functional TR that is load dependent and the organic TR that depends on abnormal leaflets or support apparatus abnormalities. From the severity point of view, the speaker talked about all the



tools applicable for a qualitative and quantitative TR severity assessment, by highlighting the role played by P.I.S.A. for the physiological assessment of the TR severity. In the last part of his lecture, Prof. Pislaru presented very interesting data on the haemodynamic impact of the Tricuspid regurgitation. In conclusion, the speaker pointed out that the quantification of the TR is possible and reliable and this is important for the prediction of the clinical outcome of patients affected by

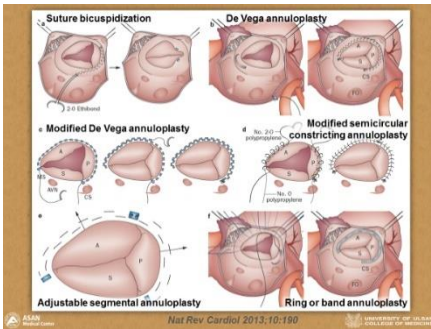
severe TR.

- What are the main TR mechanisms?
- What's about the TR severity and the TR haemodynamic impact?
- What's about the PISA parameters for the quantitative assessment of the Tricuspid regurgitation?
- What's about Vena Contracta as a new method for the assessment of the TR severity?
- What's about the assessment of TR regurgitation by the detection of physiological data?
- What's about the survival of patients affected by idiopathic TR?

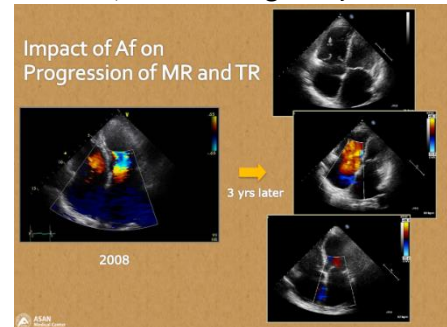
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# Which parameter will predict successful and durable tricuspid annuloplasty?

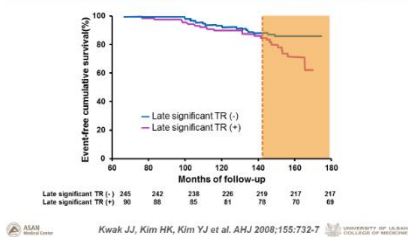


Prof. Song, coming from Seoul (Seoul, ROK) spoke about the parameters to be chosen for a well-established prediction of a successful and durable tricuspid annuloplasty. The speaker at the beginning of his talk addressed the audience with this question: how to predict a successful result of the tricuspid annuloplasty in patients affected by TR? Going deeper in his lecture, the speaker presented very interesting imaging data



in order to try to solve the problem. More in particular Prof. Song spoke about the haemodynamic TR assessment, by presenting data on the impact of the atrial fibrillation and the volume status on the tricuspid regurgitation. Speaking about prognostications in functional TR, the speaker pointed out that the maintenance of the late significant TR after any intervention,

Event-free survival depending on significant TR



like left-sided valve surgery worsens the outcome. In the last part of his lecture, Prof. Song spoke about surgical interventions and the independent variables associated with the clinical outcomes. More in particular the speaker presented very interesting data on the surgical methods to be applied and on how to predict the surgical outcomes. In conclusion, Prof. song pointed out that further investigations are needed for establishing reasonable therapeutic options and optimal timing of intervention.

- how to predict a successful result of the tricuspid annuloplasty in patients affected by TR?
- What's about the haemodynamic assessment?
- What is the correlation between atrial fibrillation and Tricuspid regurgitation?
- What's about the volume status in patients with TR?
- Which are the main surgical methods to be applied in TR patients?
- How to predict the surgical outcomes?

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# More than valve repair: effect of cone reconstruction on right ventricular geometry and function in patients with Ebstein anomaly

**Ebstein Anomaly**  
Carpenter definition

In 1988 Carpenter et al described four grades of Ebstein's anomaly:

- **TYPE A:** adequate RV volume
- **TYPE B:** large atrialized RV component, with free movement of the anterior leaflet
- **TYPE C:** severely restricted anterior leaflet movement (potential RVOT obstruction)
- **TYPE D:** almost complete RV atrialization (very small infundibular component)



Carpenter A, Chavand S, Mace L, et al. A new reconstructive operation for Ebstein's anomaly of the tricuspid valve. *J Thorac Cardiovasc Surg* 1988; 96:62-70.

performed by Wilhelm Ebstein in 1866. Going deeper in his lecture Prof. Stellin spoke about the principal anatomical features of the Ebstein anomaly and its 4 Types. In the main part of his presentation, the speaker talked about the indications for surgery and about the surgical techniques

**Ebstein Anomaly**  
Cone operation: the Mayo Clinic experience (2013)

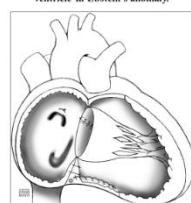
- **Early results**
  - Early mortality (1%).
  - Early reoperation for recurrent TR (13%)
    - (Re-repair 50%, replacement 50%).

Sheward JAJ, Nair NM, O'Leary PP, et al. Anatomic repair of Ebstein's malformation: Issues learned with cone reconstruction. *Ann Thorac Surg* 2012; 93(1):220-6.

More than valve repair: effect of cone reconstruction on right ventricular geometry and function in patients with Ebstein anomaly was the topic Prof. Stellin talked about. The speaker coming from Padua (IT), presented very interesting data on the Ebstein Anomaly, as a disease of the right ventricle, starting from the historical notes on its the description

**Ebstein Anomaly**  
Cone operation: da Silva (1989)

Preoperative demonstration of the displaced tricuspid valve and atrialized right ventricle in Ebstein's anomaly.



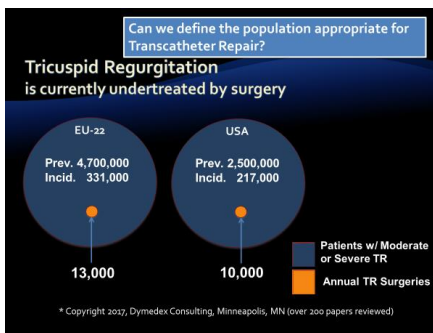
applied for the anomaly repair, by presenting very interesting data on the traditional repair, the valve different techniques replacement, the Mayo clinic new repair technique and finally on the Cone operation its early postoperative managing and the related Mayo clinic experience. In conclusion, Prof. Stellin pointed out that the cone da Silva Ebstein operation is the most anatomically natural type of TV repair.

- What are the main characteristics of the original Con operation developed by Da Silva in 1989?
- What are the main issues of the Cone operation in the early postoperative management?
- What's about the Mayo clinic experience in the Cone operation development?
- What's about the Cone operation modifications presented by the speaker?
- What's about the early mortality for Cone operation based on the Mayo clinic experience?

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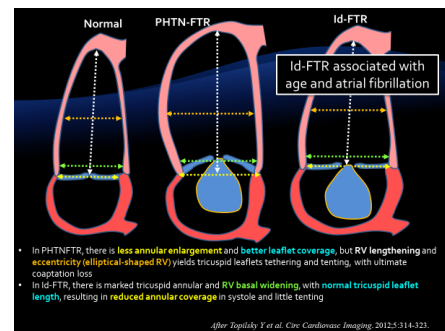
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# Transcatheter repair of the tricuspid valve: is it suitable for my patient?

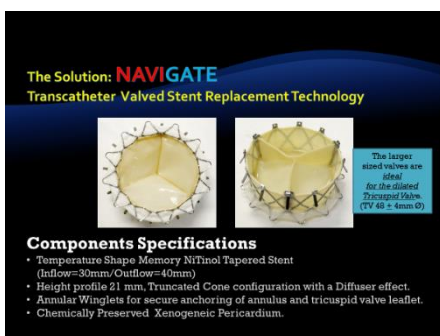


Transcatheter repair of the tricuspid valve: is it suitable for my patient? was the topic Prof. Hahn talked about. The speaker coming from New York (USA), presented very interesting data on the TR and its surgery. At the beginning of her lecture, Prof. Han pointed out that TR is undertreated in a time when left heart surgery is so developed. Going deeper in her talk, the speaker presented very interesting data on the rationale for the interventional

treatment of the TR, by highlighting that in 2010 the mortality rate for TR was about 8%, 3 to 4 time higher when compared to other single valve open procedures. In the main part of her presentation, the speaker talked about the pathophysiology of TR and its outcomes based on the echocardiographic measurements, by highlighting that the idiopathic TR is associated with the atrial fibrillation and that this phenomenon is age-dependent. In the second part of her lecture, Prof. Hahn presented data on the diagnostic methodology, speaking about 2D imaging and quantitative doppler measurements. Finally, the speaker talked about the percutaneous approaches for TR repair, by presenting very impressive data on the transcatheter solutions, their approaches and the



related anatomic targets. Prof. Hahn spoke also about the 4TECH TriCinch concept, by presenting the early clinical outcomes from this new technique given by the FORMA and the SPACER trials. The speaker at the end of her lecture presented also very innovative data on the future of the TR surgical procedure, characterized by the tricuspid valve replacement, through the application of NAVIGATE that is a new valved stent via a transcatheter replacement technology already in human experimental phase I trial.



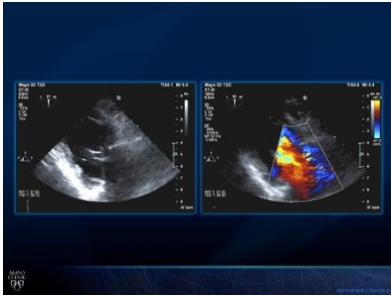
- Why is TR undertreated at the time of the left heart surgery?
- Can we define the appropriate population for Transcatheter repair?
- What's about tenting and papillary muscle displacement?
- How to accurately assess the severity of the TR?
- What are the main anatomic targets of the transcatheter tricuspid surgery?
- What's about the first percutaneous tricuspid valve annuloplasty repair performed in 2016?
- What about FORMA and SPACER presented by the speaker?
- What's about NAVIGATE valved stent and its experimental human trial, presented by the speaker?

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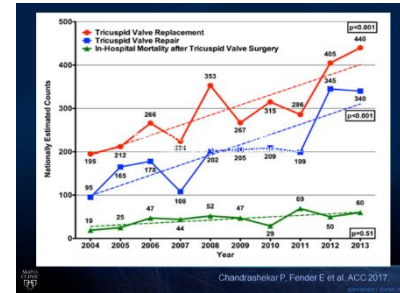


# When it is too late?



When it is too late? was the topic Prof. Pislaru talked about. The speaker coming from Rochester (USA), presented very interesting data based on real clinical cases and their solutions, characterized by two patients with severe TR but with different co-morbidities and with possible different outcomes depending on the decision of the physicians.

Going deeper in his lecture, Prof. Pislaru spoke about the treatments options for the more complicated patient based on guidelines, calculators, national



experience and finally on personal experience. The speaker

presented very interesting and impressive data on the decision to be taken in such a patient. Finally, he talked about his decisions on that specific patient and pointed out that the approach to TR is profoundly changed thanks to the improvement in surgical outcomes and in this context an aggressive surgical approach to TR, even isolated, is in order.

**How do we improve TR management?**

- Accept that severe TR is not a benign condition
- +
- Consider early(er) intervention
- +
- Percutaneous interventions – game changer?

- What's about the surgical option for a patient with TR and severe co-morbidities?
- Will the patient benefit prolong the life or improve symptoms?
- Is the surgical risk too high based guidelines, calculators and experience?
- When should you not operate?
- How do we improve TR management?

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International Symposium on:  
**THE RIGHT HEART.  
THE NEW FRONTIER**

Padua, March 1<sup>st</sup> - 3<sup>rd</sup>, 2017  
March 1<sup>st</sup> at Palazzo del Bo - March 2<sup>nd</sup> - 3<sup>rd</sup> at Botanical Garden  
University of Padua (Italy)



These are only some of the topics addressed in the congress's sections

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