La conferma della diagnosi: l'ecocardiografia e il cateterismo cardiaco destro. Dalle linee guida alla pratica clinica

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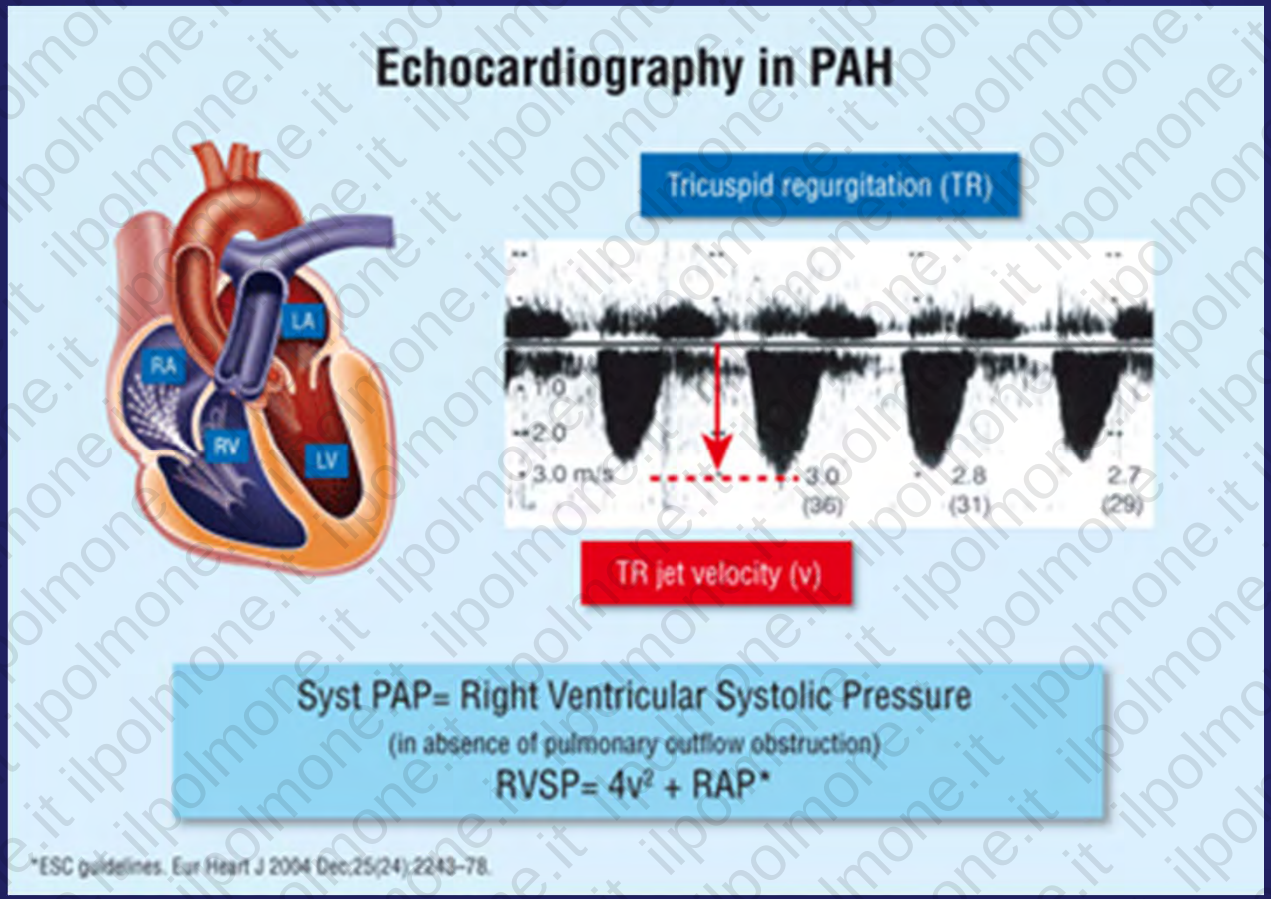
ERS guidelines 2009

ECOCARDIOGRAMMA



DOPPLER ECHOCARDIOGRAPHY

ACCP evidence-based clinical practice guidelines

In patients with a clinical suspicion of PAH, Doppler echocardiography should be performed to evaluate the level of RVSP, and to assess the presence of associated anatomic abnormalities such as right atrial enlargement, right ventricular enlargement, and pericardial effusion, left ventricular systolic and diastolic dysfunction, left-sided chamber enlargement, or valvular heart disease. Doppler echocardiography with contrast should be obtained to look for evidence of intracardiac shunting

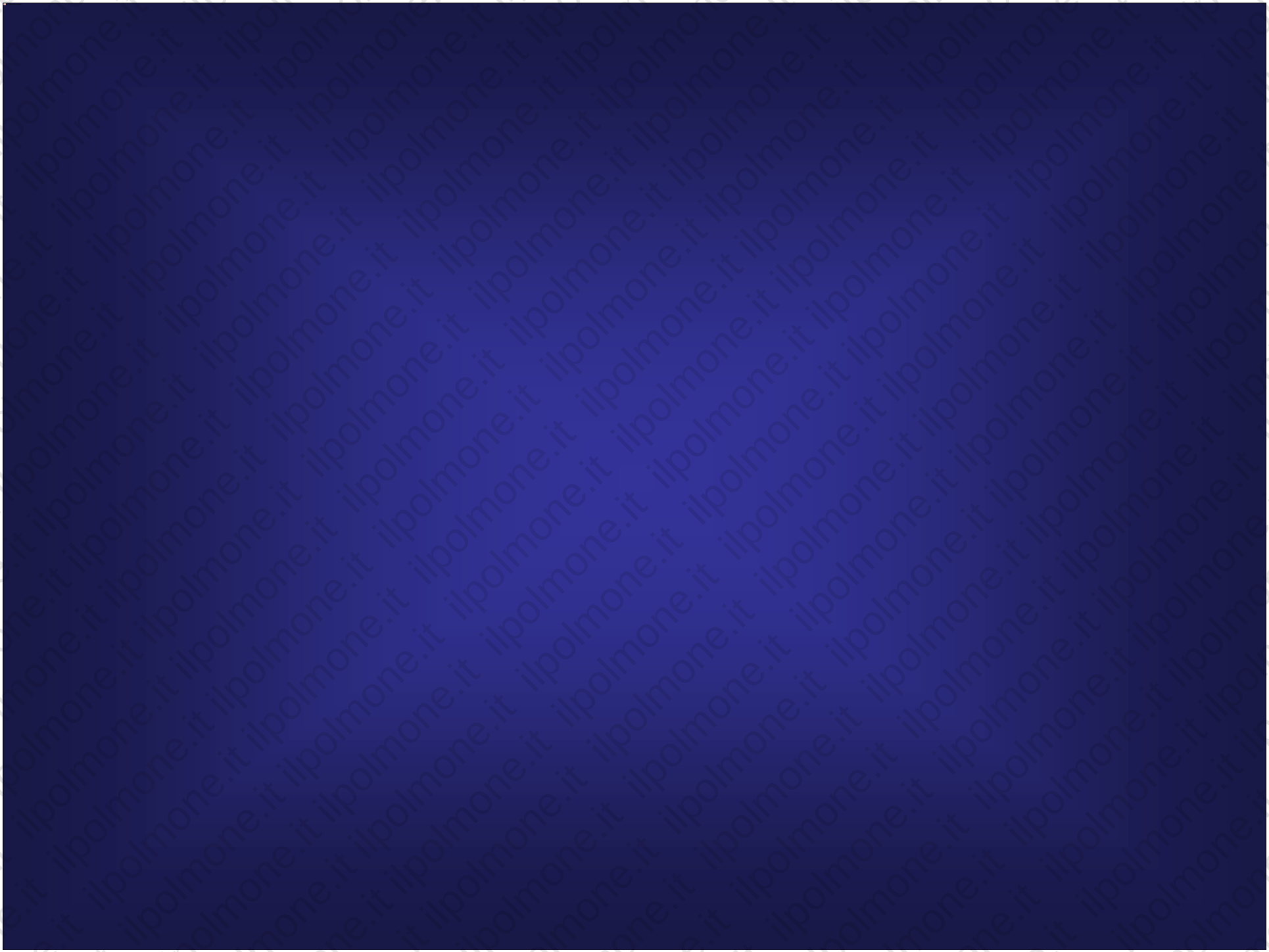
Fisher MR. Arthritis Rheum 2006;54:3043-50

DETECTION OF PULMONARY HYPERTENSION

ECHOCARDIOGRAPHY

Marked right heart dilatation

Tricuspid regurgitation



Doppler Echocardiography

- ✓ Right atrial size
- ✓ Pericardial effusion
- ✓ Left ventricular eccentricity index (D2/D1 =1 normal; D2/D1 >1 PAH)
- ✓ RV Tei index
- ✓ TAPSE (tricuspid annul plane systolic excursion) 3,0 cm normal; < 1,8 poor prognosis
- ✓ TDI
- ✓ IVC diameter
- ✓ Q by the sub-aortic VTI
- ✓ Assesment of LV filling pressures

Tei Index

Tei et al, *J Am Soc Echocardiogr* 1996 9:838-47

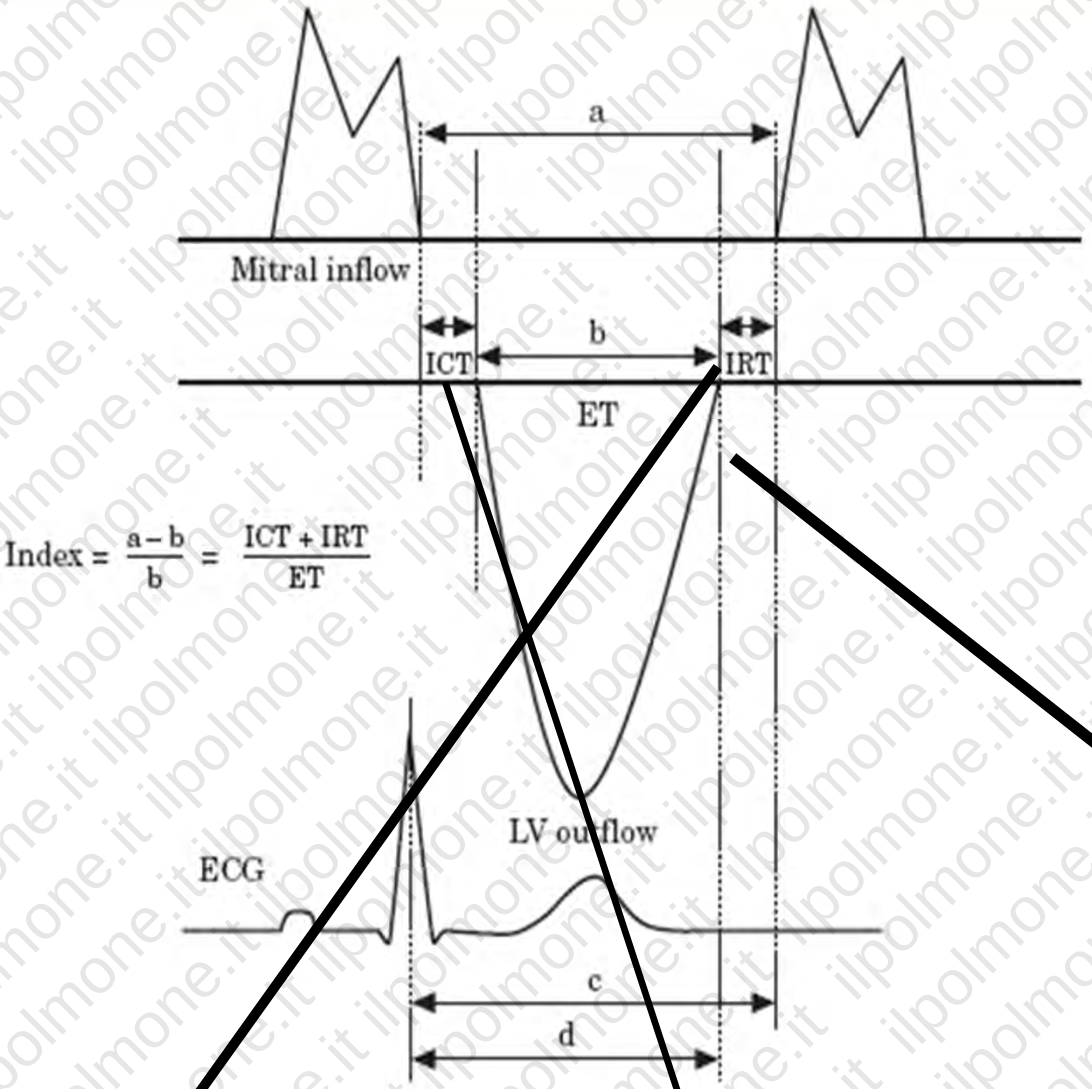
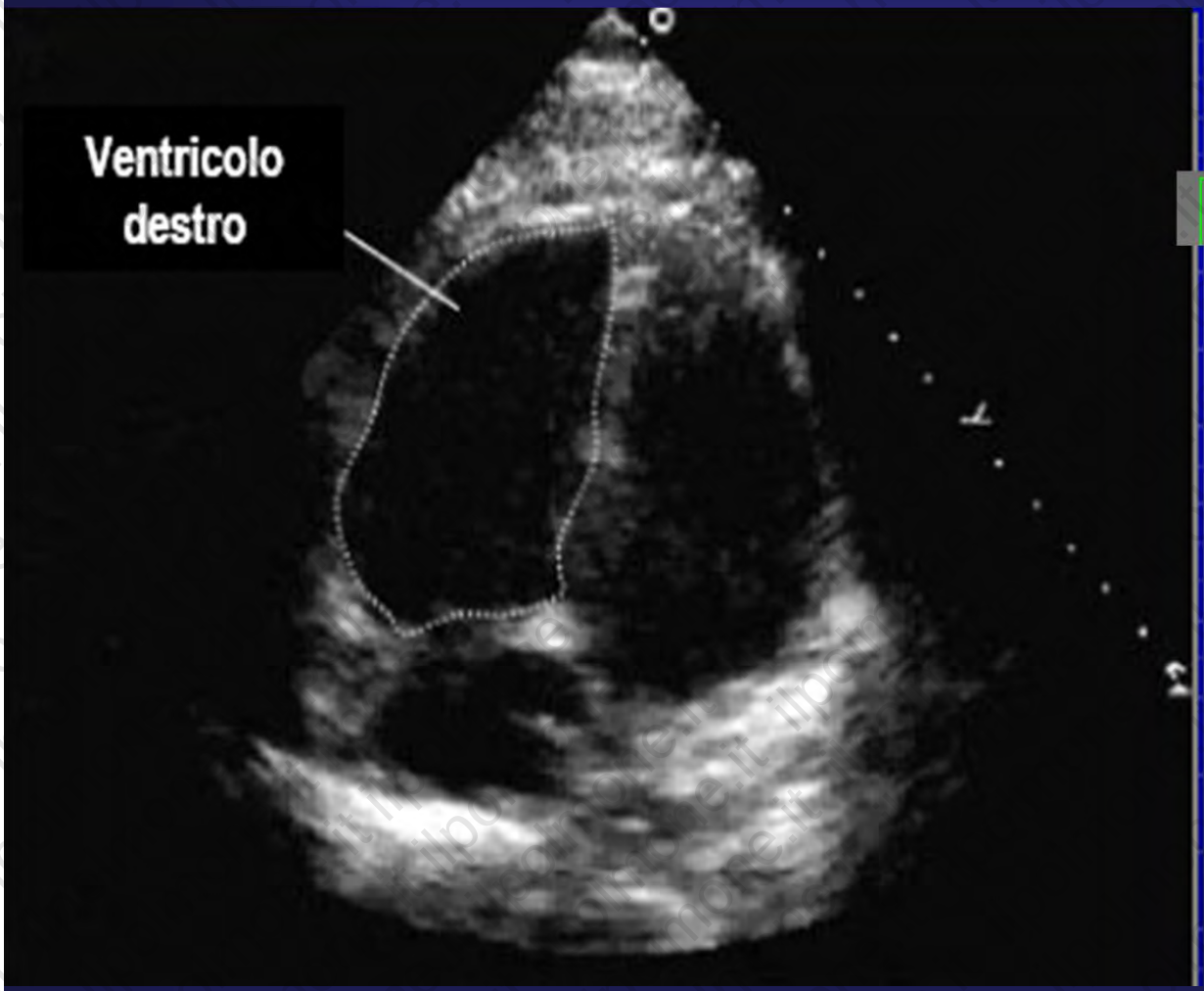


Figura 4. *Misure Doppler standard sistoliche (efflusso del ventricolo destro-VD) e diastoliche (afflusso transtricuspidale) del VD. A sinistra schema di misurazione dei parametri Doppler sistolici e diastolici del VD.* A destra in alto *pattern Doppler dell'efflusso sistolico del VD,* in basso *pattern Doppler dell'afflusso transtricuspidale del VD. A = velocità di picco atriale; AT = tempo di accelerazione; E = velocità di picco protodiastolica; ET = tempo di eiezione; IVRT = tempo di rilasciamento isovolumetrico; PEP = periodo preespulsivo; QS_2 = sistole elettromeccanica (dall'inizio del complesso QRS alla fine dell'eiezione sistolica del VD).*

TDI (Doppler Tissue imaging)



Myocardial Performance Index

DOPPLER ECHOCARDIOGRAPHY

ACCP evidence-based clinical practice guidelines

TAPSE

Tricuspid annular plane systolic excursion is a simple measure of RV ejection fraction

The evaluation of right ventricular systolic function is important for its clinical and prognostic value in pulmonary hypertension

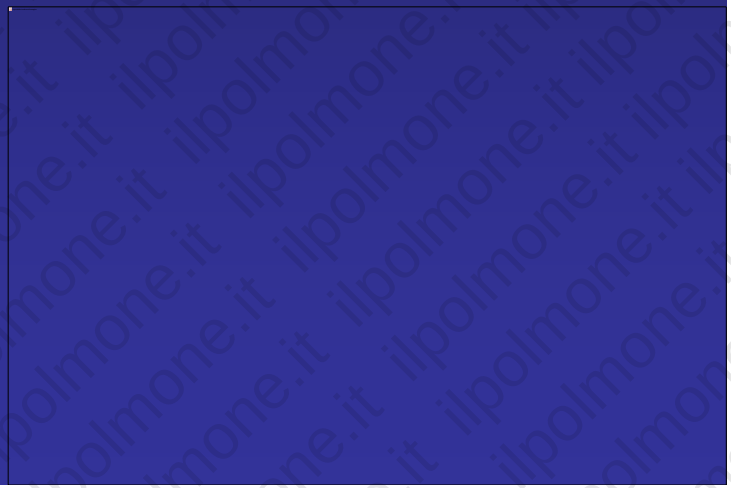
It may be affected by co-existing chronic obstructive pulmonary disease

Values of TAPSE < 20 mm is correlated negatively to the pulmonary pressure

A TAPSE less than 19.6 mm indicate a RVEF less than 40%.

Values <18 mm is a negative prognostic survival factor

Kjaergaard J. Eur J Heart Fail 2007;9:610-6
Tamborini G. Int J Cardiol 2007;115:86-9
Forfia PR. Am J Respir Crit Care Med 2006; 174:1034-41
Lee CY. Echocardiography 2007;24:118-25



Doppler Echocardiography

Limitations

RVSP increases with age and BMI

Range 15-57 mmHg

If we consider positive a low RSVP, the number of false positive cases increases

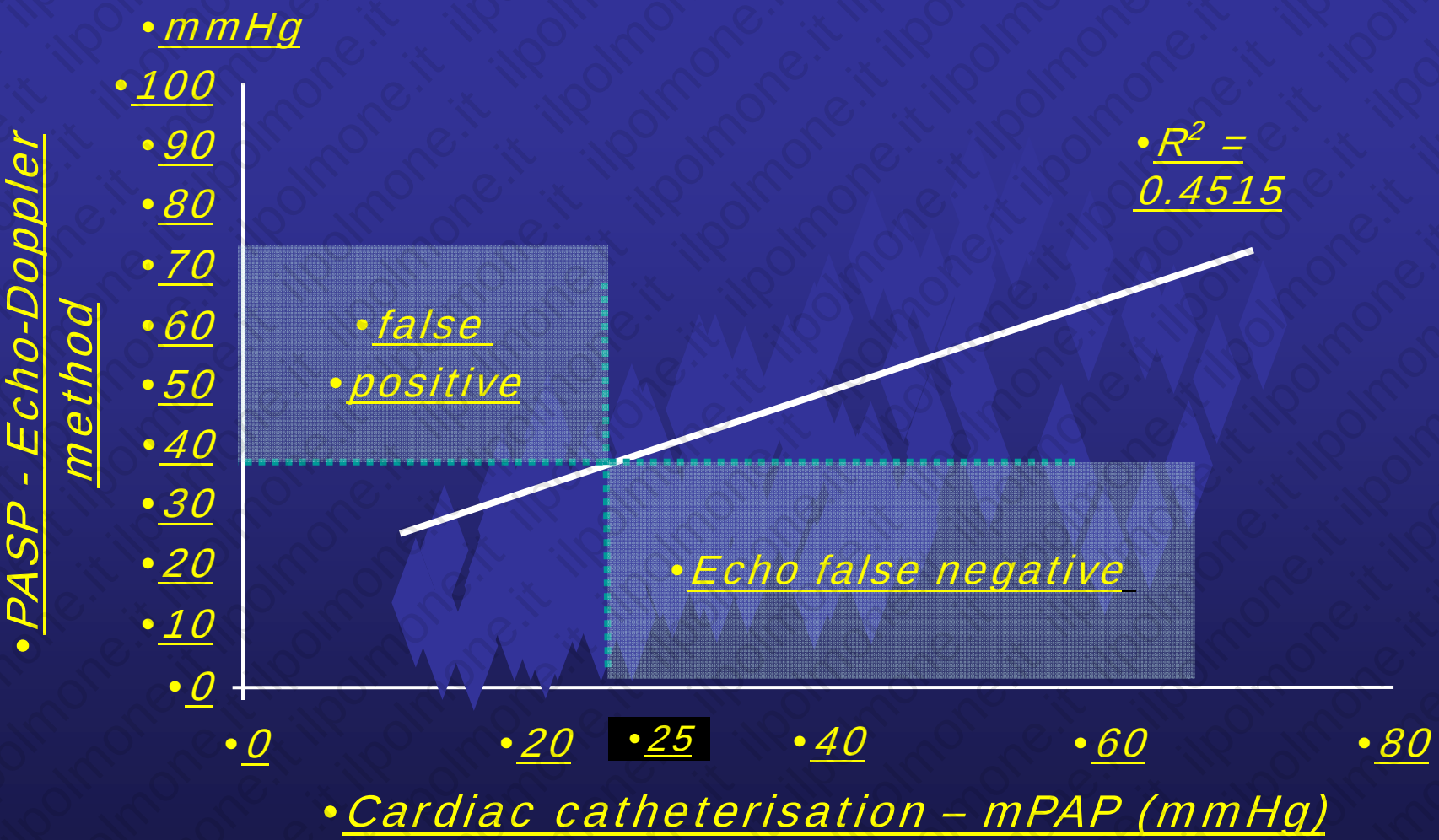
DOPPLER ECHOCARDIOGRAPHY

EcoDoppler is not useful for CI and PVR

Often RVSP is lower in patient with severe PAH

Often RVSP is higher in patient with normal PAH

RHC confirmation is absolutely mandatory



- Mukerjee D, et al. Rheumatology 2004; 43:461-6

Screening for PAH (echocardiography)

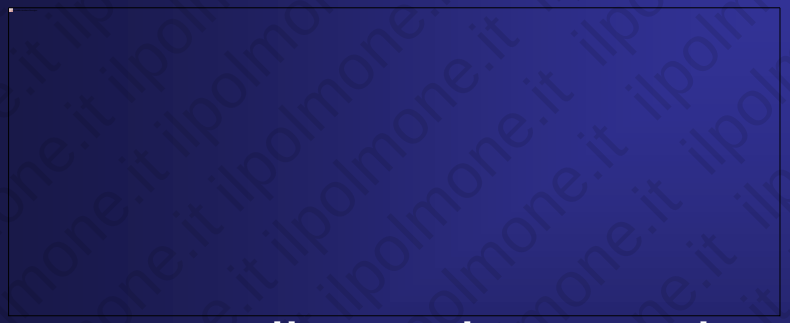
- ✓HIV infection: 0,1%- only if symptoms
- ✓Portal hypertension: 1-2%- only if symptoms
- ✓Systemic sclerosis:10-15%- once a year
- ✓Congenital heart disease: 10-15%- once a year
- ✓Anorexigens: 0,01% - if symptoms

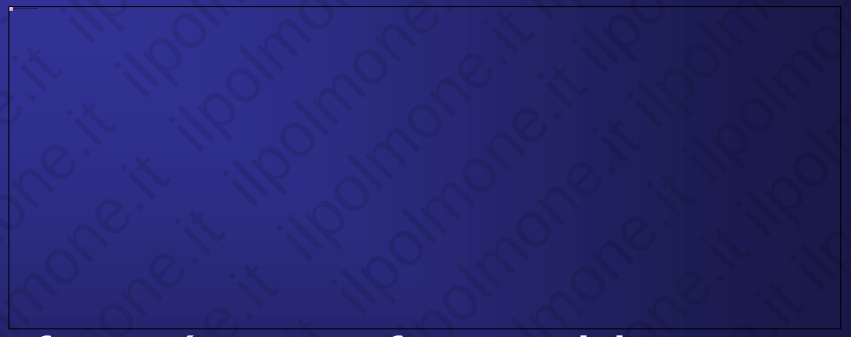
ESC Guidelines- Galiè et al et al. *Eur Heart J* 2004 25:2243-78

ECHOCARDIOGRAPHY AND PULMONARY FUNCTION AS SCREENING TEST FOR PULMONARY ARTERIAL HYPERTENSION IN SYSTEMIC SCLEROSIS

137 Pts (52 with and 85 without fibrosis)

RHC
- 99 Pts with PAH (54 without fibrosis)
- 38 Pts without PAH (31 without fibrosis)





ECOcardiography: good specifity of TG (97% of Pts with TG>45 mmHg had PAH at RHC)

Mukerjee D et al. Rheumatology (Oxford) ,2004 Apr;43(4):461-6

Early Detection of Pulmonary Arterial Hypertension in Systemic Sclerosis

19 female patients with SSc in NYHA class I e II were enrolled and submitted to all diagnostic tests

Right heart catheterisation detected PAH in in 15/19 patients

Serra W. Cardiovasc Ultrasound 2010; 8:25

Early Detection of Pulmonary Arterial Hypertension in Systemic Sclerosis

Serra W. Cardiovasc Ultrasound 2010; 8:25

Early Detection of Pulmonary Arterial Hypertension in Systemic Sclerosis

Conclusion:

The alterations in cardiorespiratory function describe an initial impairment in patient with systemic sclerosis.

Echocardiographic findings non-invasively reveal the presence of increased pulmonary pressure and PVR.

Serra W. Cardiovasc Ultrasound 2010; 8:25

Early Detection of Pulmonary Arterial Hypertension in Systemic Sclerosis

599 Pts

33 with PAH at echocardiography

RHC
- 18 Pts with PAH
- 3 Pts with left ventricular dysfunction
- 12 Pts without PAH

Diagnosis: moderate PAH

Hachulla E, et al. Arthritis Rheum ,2005 Dec;52(12):3792-800

Assessment of PAH in Patients with Systemic Sclerosis: Comparison of Noninvasive Tests with Results of Right-Heart Catheterization

Forty-nine patients with SSc were evaluated for suspected PH based on clinical findings, progressive dyspnea, and pulmonary function tests (PFT).

RHC identified 24/49 (49%) PAH patients

Echo classified 38 subjects correctly (14/24 with and 24/25 without PH; sensitivity 58%, specificity 96%)

CONCLUSION: In evaluation of SSc with suspected PH, echo appeared to be the most useful among the noninvasive tests, mainly due to the high specificity, high positive predictive value, and highest AUC.

However, due to the low sensitivity of noninvasive testing, RHC should remain the gold standard

Hsu VM., et al. J Rheumatol ,2008 Mar;35(3):458-65

Echocardiography as an outcome measure in scleroderma-related pulmonary arterial hypertension: a systematic literature analysis by the EPOSS group

Echo was considered partially validated with respect to criteria on validity based on significant correlations between echo measures and right-heart catheterization in patients with SSc at risk of PAH/PH.

However, echo was found to lack specificity (lack of content validity), since measurements of echo pulmonary pressure may be influenced by left-heart disease and interstitial lung disease

Structured literature review on full-text English articles was performed using the PubMed and Cochrane databases

J Rheumatol. 2010 Jan;37(1):105-15. Epub 2009 Dec 1.

Right heart catheterization
History

400 a.c. Utilizzo dei cateteri su cadavere per esplorare la funzione valvolare cardiaca.

1711: Hales effettua cateterismo cardiaco su cavallo usando tubi di vetro e trachea di oca.

1844: Il francese Bernard conia il termine “cateterismo cardiaco ed effettua rilevazioni pressorie su animali.

1929: Forssmann esegue su se stesso il primo cateterismo cardiaco sull’uomo.

1940: Cournand e Richards (riprendono esperimento di Forssmann)

1956: Nobel a Forssmann, Cournand e Richards

1970: Schwan e Ganz (catetere a palloncino)

Objectives of right heart catheterization

1. To confirm Eco-Doppler diagnosis and to have a marker of disease's gravity
2. To valuate vasoreactivity
3. To valuate possible shunts
4. For prognosis
5. To valuate efficacy of specific drugs in follow-up

Rare Lung Diseases

Ospedale San Giuseppe Experience (RHC 2002- october2010)

284 RHC in 225 pts

PH - Pulmonary Hypertension -

PH has been defined as an increase in **mean pulmonary arterial pressure (PAP) ≥25 mmHg at rest** as assessed by right heart catheterization (RHC)

The significance of a mean PAP between 21 and 24 mmHg is unclear.
Patients resenting with PAP in this range need further evaluation in epidemiological studies.

Recent re-evaluation of available data has shown that **the** normal mean PAP at rest is 14 ±3 mmHg, with an upper limit of normal of~20 mmHg.

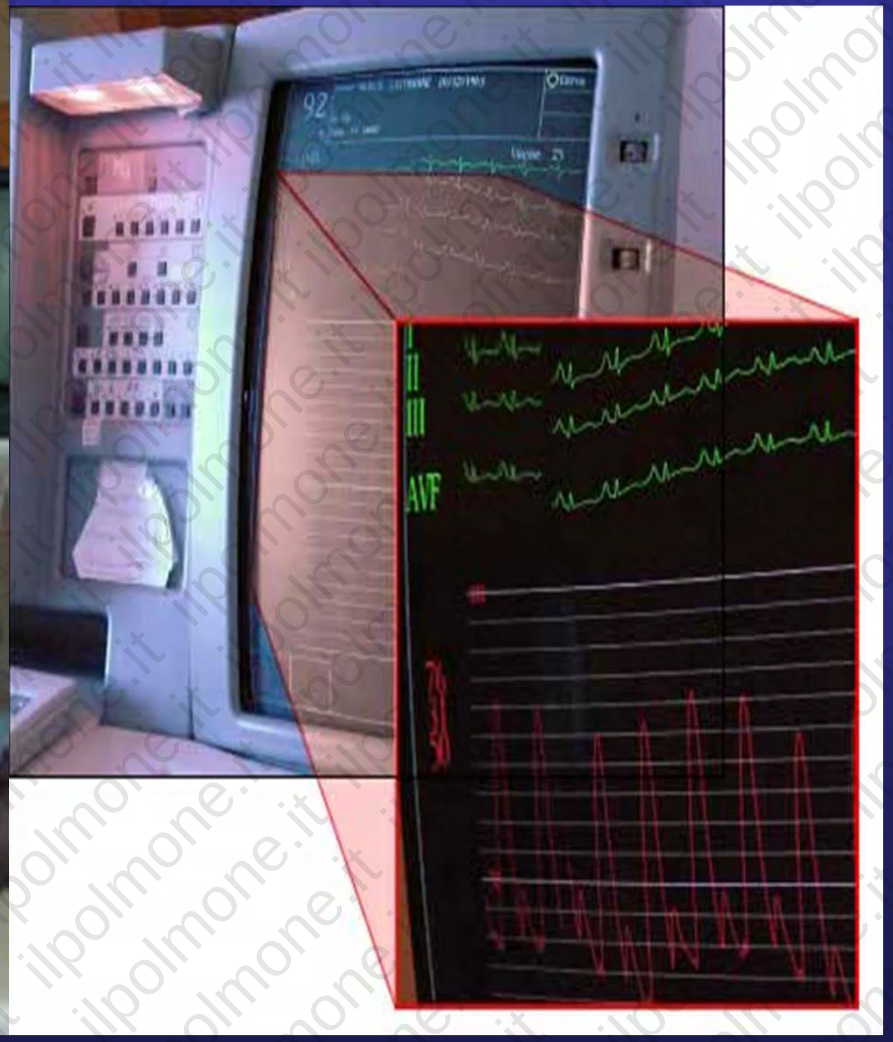
* assessed by right heart catheterization

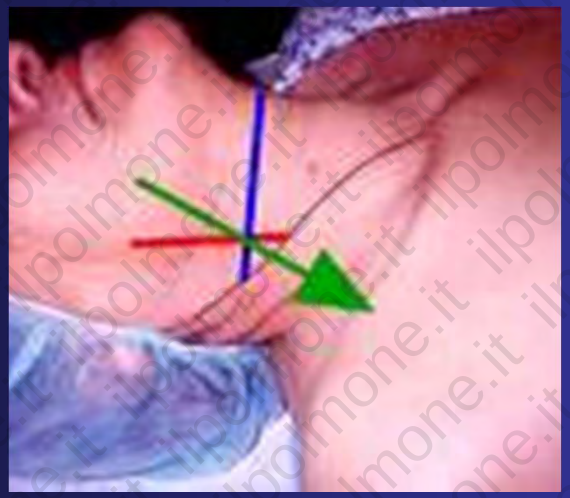
1. Galiè N, Hoeper MM, Humbert M et. al. Guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Heart J. 2009 Oct;30(20):2493-537

Right-heart catheterisation

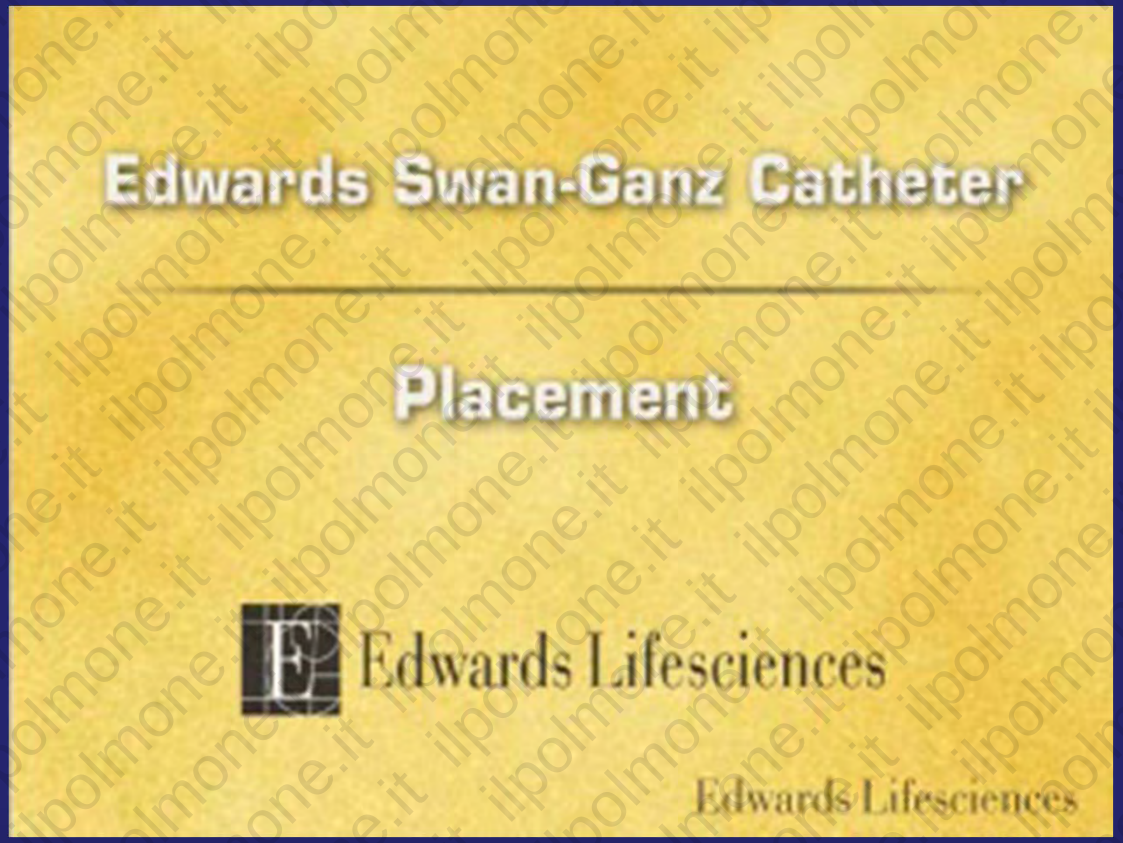
- Involves threading a catheter through a vein until it reaches the pulmonary artery
- Allows direct measurement of pressure inside:
  - Right atrium
  - Right ventricle
  - Pulmonary arteries
- Right-heart catheterisation is used to:
  - Confirm the diagnosis of PAH
  - Assess haemodynamic impairment
  - Test vasoreactivity

Galiè N *et al Eur Heart J* 2004; 25: 2243–2278


I
II
III
AVF




Positioning of the
catheter


Edwards Swan-Ganz Catheter
Placement
Edwards Lifesciences
Edwards Lifesciences

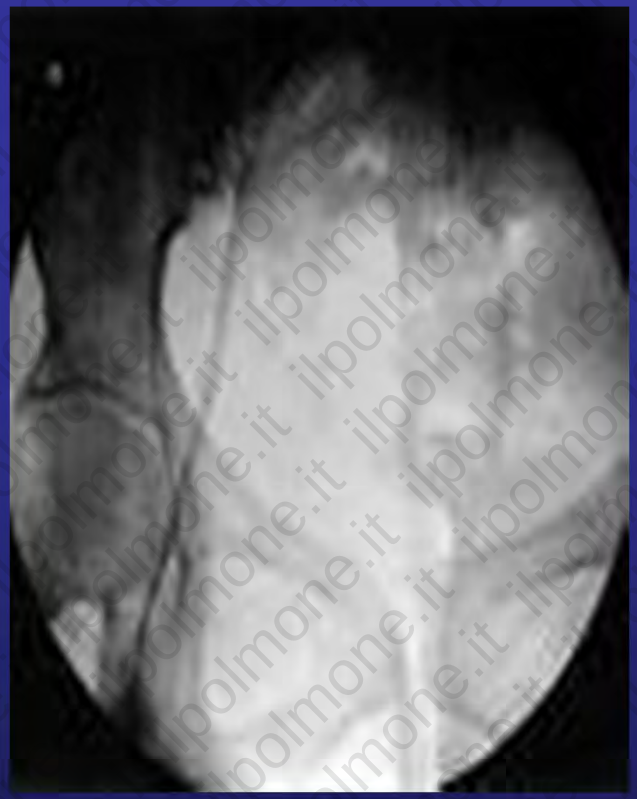
SCHWAN GANZ

lungo 110 cm con diametro esterno di 2,3 mm (7 french).

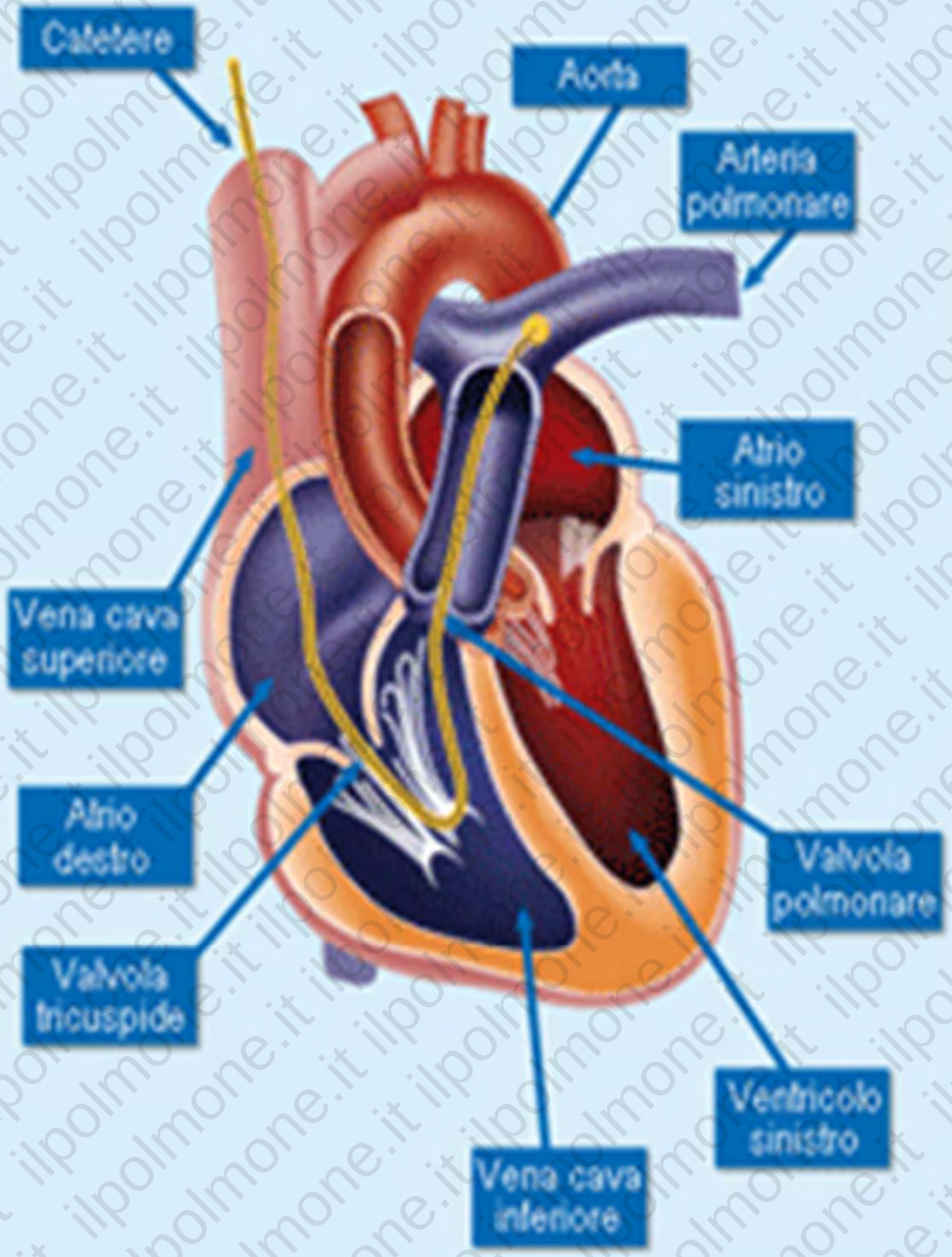
All' interno vi sono 2 canali: il primo percorre l'intera lunghezza del catetere per andare ad aprirsi alla sua estremità (apertura distale), mentre il secondo, più corto, termina in un'apertura situata a 30 cm dall'estremità del catetere (apertura prossimale).

L'estremità del catetere è provvista di un palloncino della capacità di 1,5 ml. Il palloncino completamente gonfiato crea una protezione per l'estremità del catetere.

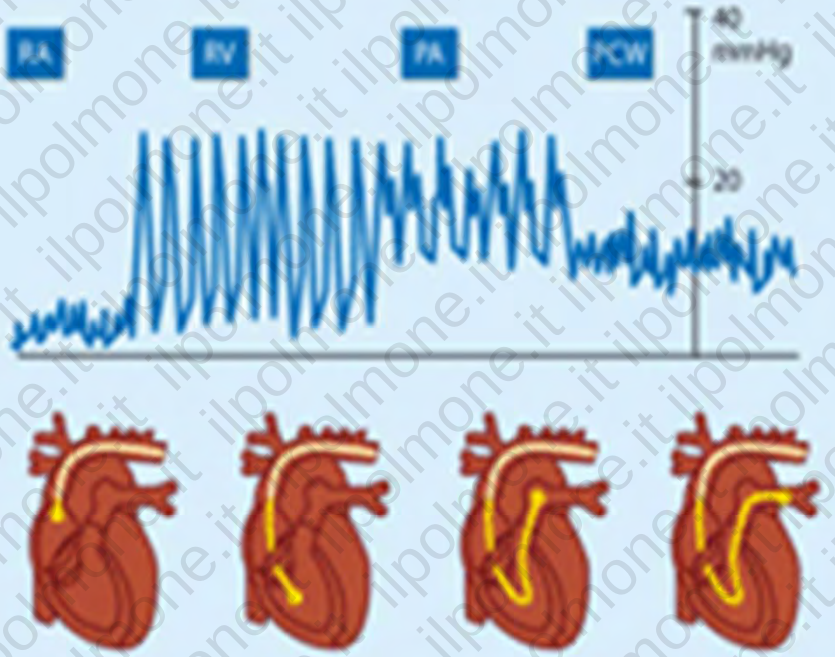
Situato sulla superficie esterna del catetere, a 4 cm dall'estremità, vi è un termistore.



Cateterismo del cuore destro



Caratteristiche onde di pressione intracardiaca durante il passaggio attraverso il cuore



Tipo di pressione	Valore medio (mmHg)	Intervallo di normalità (mmHg)
Atrio Dx	3	0-8
Ventricolo Dx		
Picco sistolico	25	15-30
Telediastolica	4	0-8
Arteria polmonare		
Media	15	9-16
Picco sistolico	25	15-30
Telediastolica	9	4-14
Pressione arteria polmonare a catetere bloccato (wedge)		
Media	9	2-12

Normal Values for Cardiac Index and Related Measurements

Measurement	Normal Value	SD
O_2 uptake	143 mL/min/m^{2*}	14.3
Arteriovenous O_2 difference	4.1 dL	0.6
Cardiac index	3.5 L/min/m^2	0.7
Stroke index	46 mL/beat/m^2	8.1
Total systemic resistance	1130 dynes-sec-cm^{-5}	178
Total pulmonary resistance	205 dynes-sec-cm^{-5}	51
Pulmonary arteriolar resistance	67 dynes-sec-cm^{-5}	23

PAP = PVR *CO + Wedge

PVR ↑

- PAH Idiopatica
- PAH Familiare
- PAH porto-polmnare
- Malattie del collagene
- Esposizioni a tossici/farmaci
- Malattie polmoanari
- Tromboembolia cronica

- Emangiomatosi capillare polmonare
- Compressione vascolare ab estrinseco

CO ↑

- Shunt sistemico-polmonari
- Patologia congenita
- Tireotossicosi
- Fistole AV
- PAH porto-polmonare

Wedge ↑

- Malattie cuore sx
- Patologie valvolari sx
- Malattia veno-occlusiva polmonare

Haemodynamic definitions of Pulmonary Hypertension [a]

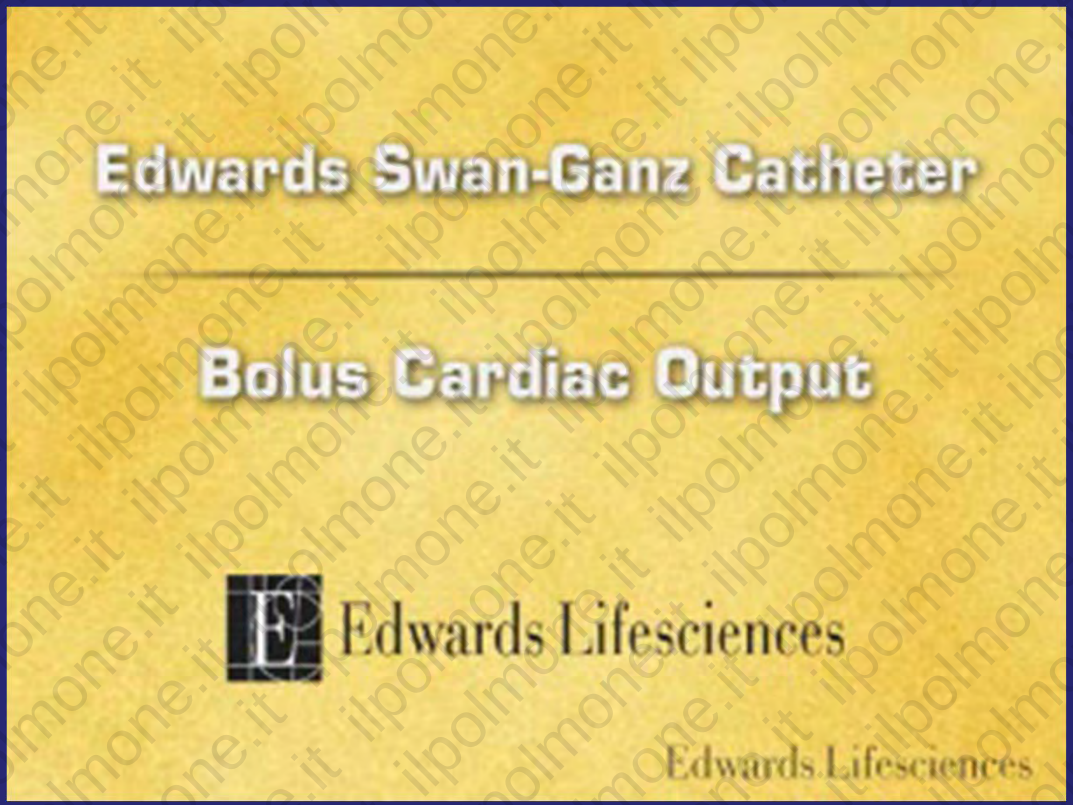
Definition	Characteristics	Clinical group(s)
Pulmonary hypertension (PH)	Mean PAP ≥ 25 mmHg	All
Pre-capillary PH	Mean PAP ≥ 25 mmHg PWP ≤ 15 mmHg CO normal or reduced	1. Pulmonary arterial hypertension 3. PH due to lung diseases 4. Chronic thromboembolic PH 5. PH with unclear and/or multifactorial mechanisms
Post-capillary PH . Passive Reactive (out of proportion)	Mean PAP ≥ 25 mmHg PWP >15 mmHg CO normal or reduced [c] TPG ≤ 12 mmHg TPG >12 mmHg	2. PH due to left heart disease

a All values measured at rest.

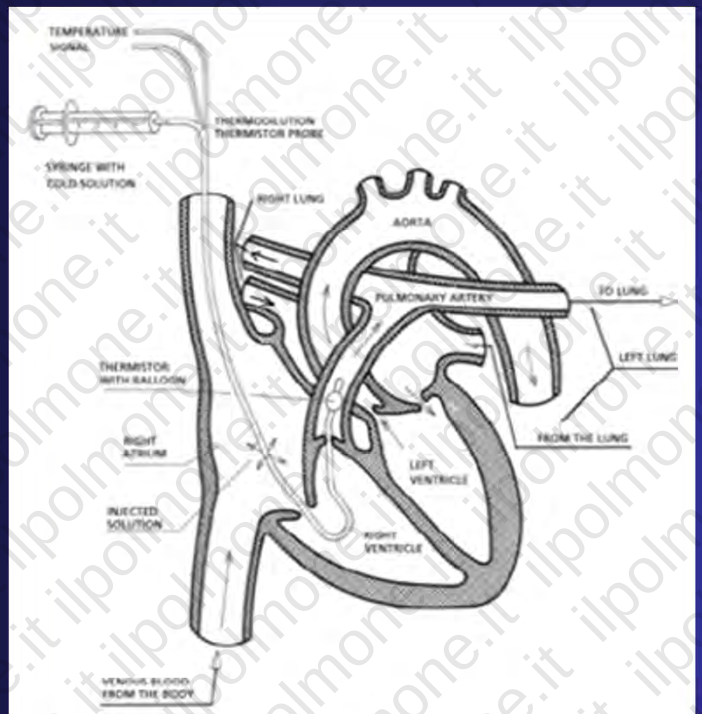
c High CO can be present in cases of hyperkinetic conditions such as systemic-to-pulmonary shunts (only in the pulmonary circulation), anaemia, hyperthyroidism, etc.

CO = cardiac output; PAP = pulmonary arterial pressure; PH = pulmonary hypertension; PWP = pulmonary wedge pressure; TPG = transpulmonary pressure gradient (mean PAP – mean PWP).

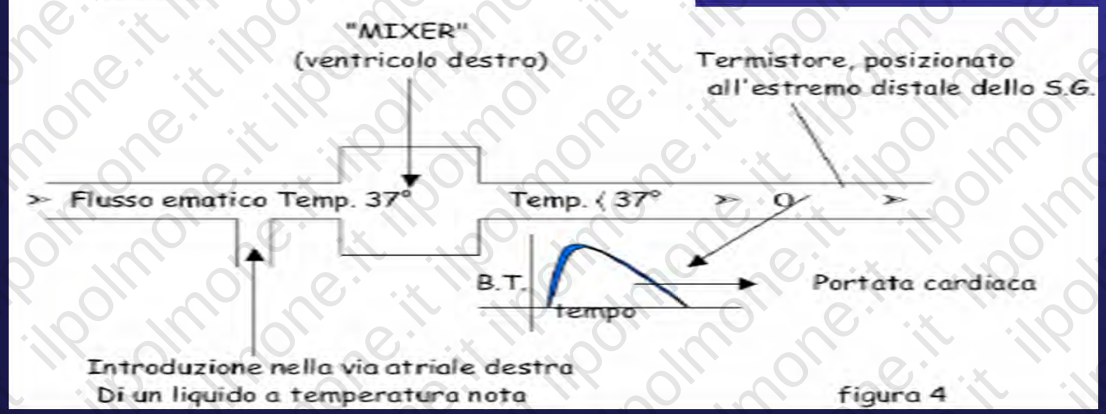
1. Galiè N, Hoeper MM, Humbert M et. al. Guidelines for the diagnosis and treatment of pulmonary hypertension. Eur Heart J. 2009 Oct;30(20):2493-537


Edwards Swan-Ganz Catheter
Bolus Cardiac Output
Edwards Lifesciences
Edwards Lifesciences

CARDIAC OUTPUT



“ Fegler (1954) e poi Ganz, postularono che le variazioni di temperatura del sangue, in seguito all’iniezione di una soluzione fredda a temperatura nota, sono direttamente proporzionali alla portata cardiaca.”



Vasoreactive test

Table 10 Route of administration, half-life, dose ranges, increments, and duration of administration of the most commonly used agents for pulmonary vasoreactivity tests

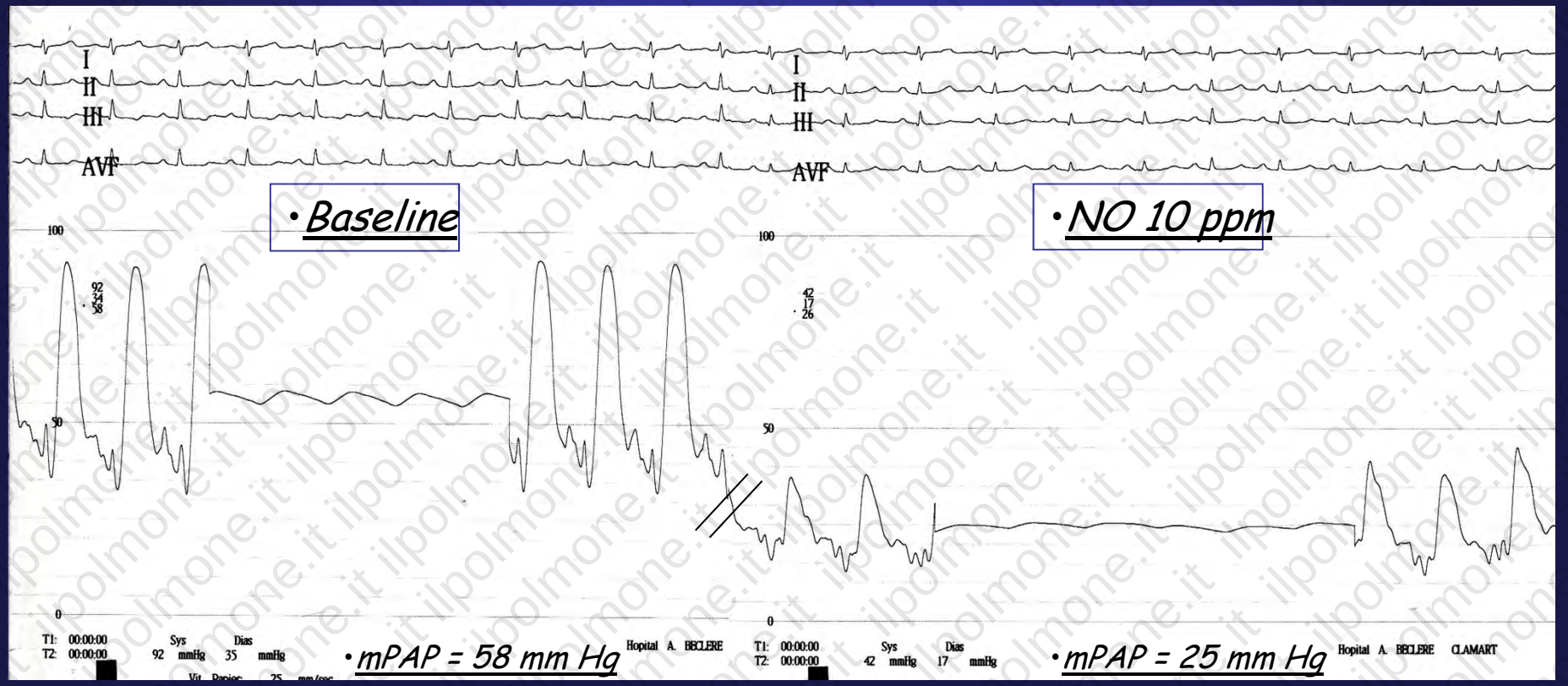
Drug	Route	Half-life	Dose range[a]	Increments[b]	Duration[c]
Epoprostenol	Intravenous	3 min	2–12 ng/kg/min	2 ng/kg/min	10 min
Adenosine	Intravenous	5–10 s	50-350 μg/kg/min	50 μg/kg/min	2 min
Nitric oxide	Inhaled	15–30 s	10–20 p.p.m	–	5 min[d]

[a]Initial dose and maximal tolerated dose suggested (maximal dose limited by side effects such as hypotension, headache, flushing, etc.).
[b]Increments of dose by each step.
[c]Duration of administration on each step.
[d]For NO, a single step within the dose range is suggested.

Responders (only in 10- 15% of patients in acute and 7% in chronic): ↓ mPAP ≥ 10 mmHg
and mPAP ≤ 40 mmHg
CO ↑ o ↔

ASSESSMENT OF PAH SEVERITY
ACUTE VASOREACTIVITY TESTING

- The goal of acute vasoreactivity testing is to select safely PAH patients who could respond favorably to long-term oral vasodilator therapy.
- This test identifies patients with a better long-term prognosis



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CASO CLINICO

ECHOCARDIO

DATE	PAPs
14/12/09	88 mmHg
10/11/10	75 mmHg

RHC

PAPs	PAP m	CO	CI	PVR	WEDGE
119	73	4,99	2,77	12.4	11
95	55	4,33	2,43	18	7

PLEASE, PAY MORE ATTENTION!

GRAZIE