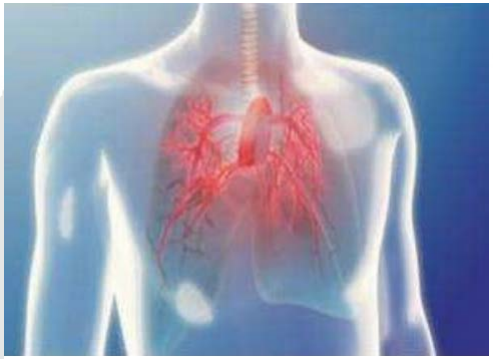
La comunicació interauricular en la gent gran: una cardiopatia no tan simple



Sílvia Montserrat

2 desembre 2019

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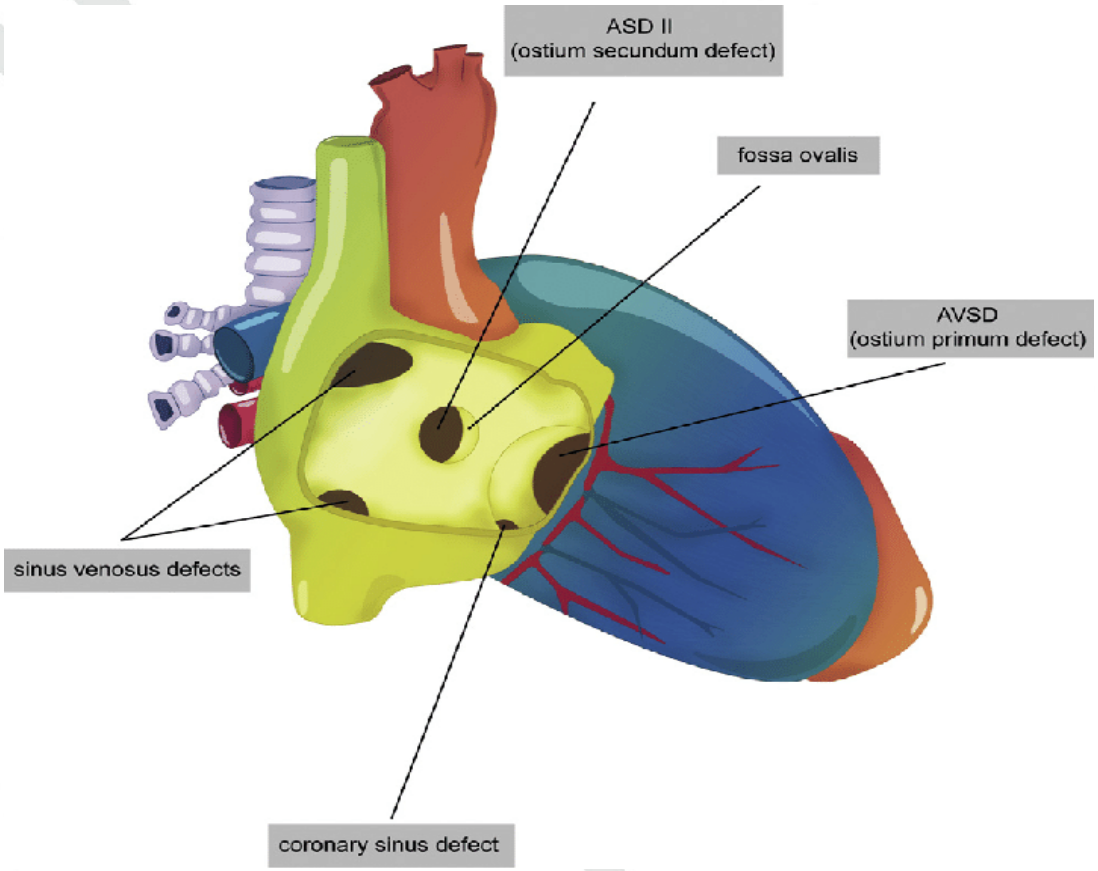
Tipus de CIA



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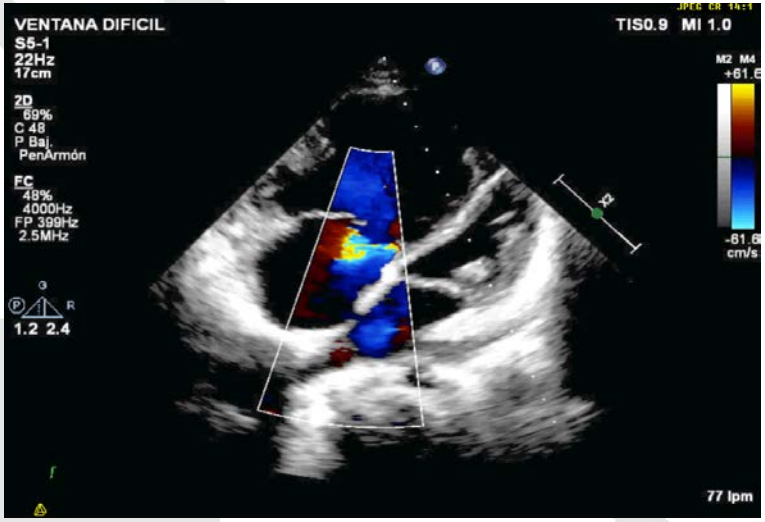
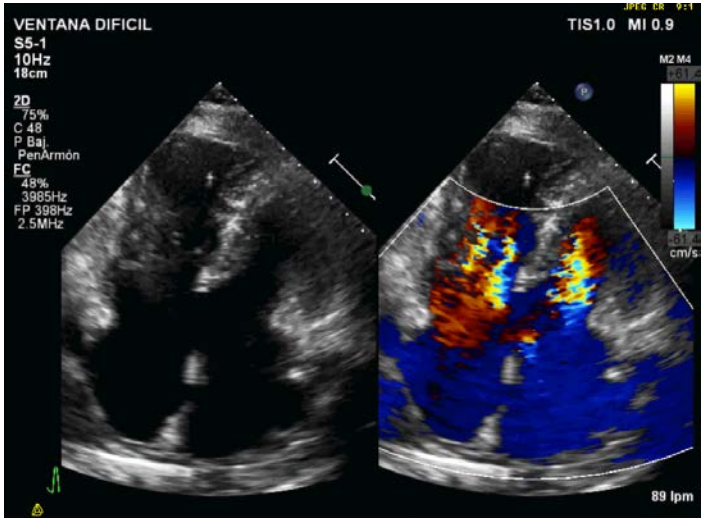
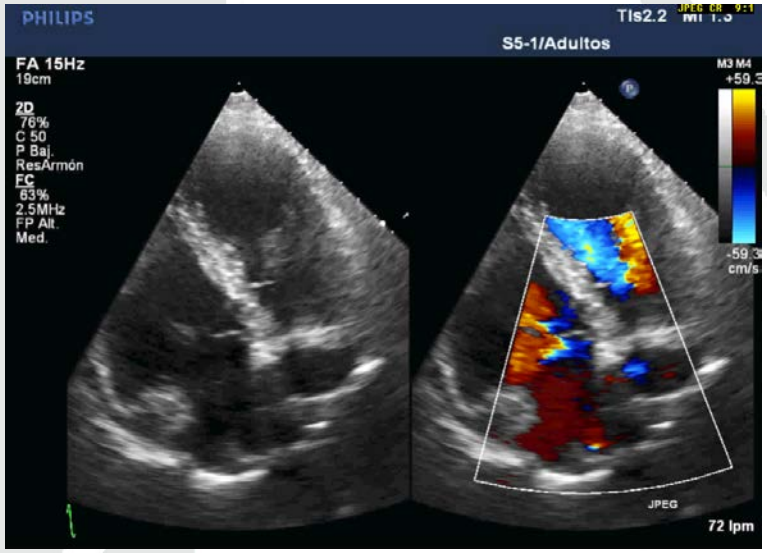
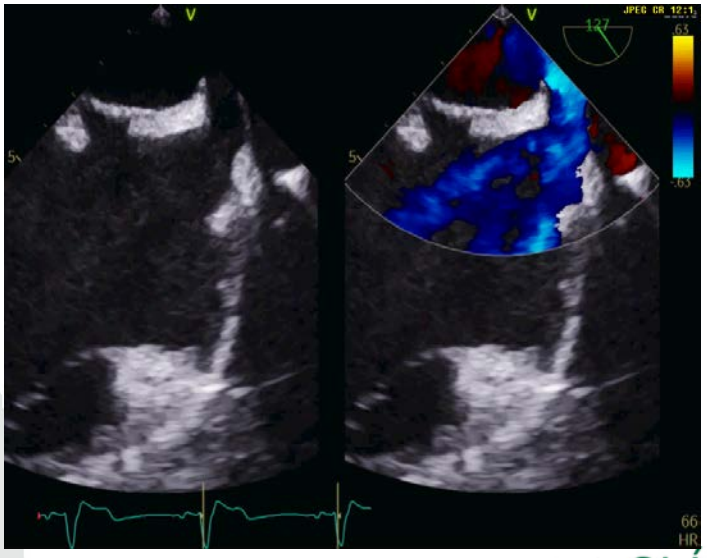
Tipus de CIA









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CIA en la gent gran

La CIA és la tercera cardiopatia congènita més comuna.

Supervivència edat adulta de CC

- 98% lleus: CIA OS
- 90% moderades: CIA sí venós, Canal AV parcial

La CIA en gent gran:

- Edat i comorbilitats:

  benefici /risc del tractament: quirúrgic i percutani.
- FA/ Insuficiència tricuspidea massiva
- Valoració de la Hipertensió pulmonar.
- Sdr. Eisenmenger. Tractament
- Altres causes de hipertensió pulmonar

CIA en gent gran: tractament quirúrgic

- Es detecta en un 25-30% dels pacients en edat adulta[1].
- Estudis restrospectius [2] i de series [3-4] quirúrgiques en pacients de > 60 anys, avalen una milloria dels símptomes i suggereixen una milloria de la supervivència

1. *Lindsey JB, Hillis LD.Lancet. 2007;369:1244–1246*
2. *Konstantinides S, et al. N Engl J Med. 1995;333:469–473.*
3. *Sutton MG et al. Circulation. 1981;64:402–409.*
4. *Nasrallah AT, et al. Long-term results. Circulation. 1976;53:329–331.*

Risc-benefici del tractament quirúrgic > 60 anys

23. Oyer PE, Stinson EB, Griepp RB, Shumway NE: Valve replacement with the Starr-Edwards and Hancock prostheses: comparative analysis of late morbidity and mortality. Ann Surg **186:** 301, 1977
24. Cohn LH, Sanders JH, Collins JJ: Actuarial comparison of Hancock porcine and prosthetic disc valves for isolated mitral valve replacement. Circulation **54** (suppl III): III-60, 1976
25. Cohn LH, Koster JK, Mee RBB, Collins JJ: Long-term follow-up of the Hancock bioprosthesis heart valve: a 6-year review. Circulation **60** (suppl I): I-87, 1979

Atrial Septal Defect in Patients Ages 60 Years or Older: Operative Results and Long-term Postoperative Follow-up

MARTIN G. ST. JOHN SUTTON, M.B., ABDUL J. TAJIK, M.D., AND DWIGHT C. MCGOON, M.D.

SUMMARY **Between 1955 and 1977, 66 patients ages 60 years or older underwent operative closure of secundum atrial septal defect. Of these, 56 (85%) were catheterized preoperatively. The 56 patients were divided into three groups to assess the effects of pulmonary hypertension on operative mortality, symptoms and longevity. The 17 group 1 patients had peak systolic pulmonary artery pressures (PAPs) of less than 40 mm Hg; the 21 group 2 patients had PAPs of 40–60 mm Hg; and the 18 group 3 patients had PAPs of more than 60 mm Hg. Among the three groups, there was no significant difference in Qp/Qs, right or left atrial pressures, right or left ventricular end-diastolic pressures and Qs, although pulmonary vascular resistance was significantly higher ($p < 0.01$) in group 3 than in group 1. Four patients died, yielding an operative mortality of 6%. All four patients had undergone additional operative procedures. Operative mortality was unrelated to preoperative symptom class, PAP or pulmonary vascular resistance. Forty-seven patients were followed up for 2–20 years (mean 6.6 years), and of these, 41 (87%) improved by at least one functional class. Symptomatic benefit occurred in all groups, regardless of preoperative PAP, pulmonary vascular resistance or functional class. Actuarial survival curves showed that longevity at 5 and 10 years postoperatively was significantly increased ($p < 0.01$) for patients with atrial septal defect treated surgically compared with that predicted for age-matched patients treated medically.**

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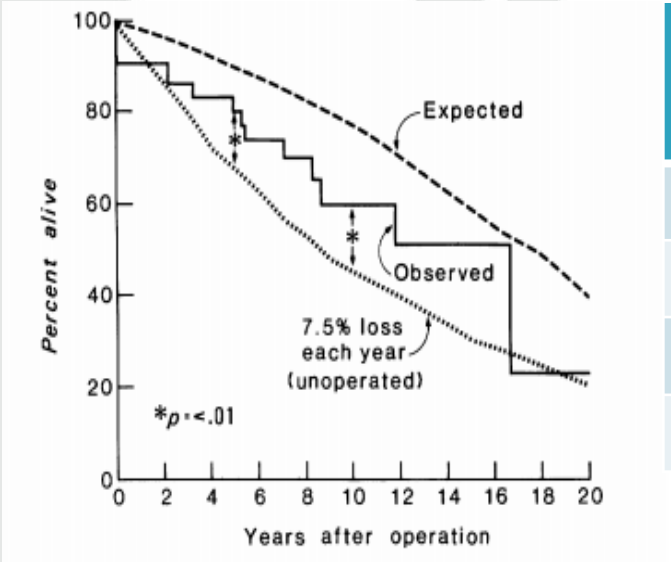
Criteris d'inclusió
> 60 anys (major 83 anys)
CIA OS 64 i sí venós sinus 8.
RVP< 15 U/ m ^{2}
Gener 1955 al Desembre 1977



PAPs (n=56)	40 (n=17)	40-60 (n=21)	>60 (n=18)
CF III-IV pre	38%	69%	76%
FA pre	20%	67%	67%
Exitus	2	4	6
CF I- II post	75%	85%	83%

6% mortilitat 30 dies. (4 exitus: 14 U/m^2 +bypass, 2 anuloplàsties T, 1 PTS T)

Sutton MG et al. Circulation. 1981;64:402–409.

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Our data, which represent the first long-term follow-up study of a large number of patients 60 years of age or older who underwent operative closure of atrial septal defect, indicate that (1) atrial septal defect in the elderly is not rare and must be considered in a patient with congestive heart failure and associated atrioventricular valve regurgitation; (2) most patients (67%) have disabling symptoms (functional classes III and IV); (3) the severity of symptoms increases with increase in PAP and right ventricular work; (4) most patients are symptomatically improved on a long-term basis by operation; (5) surgical closure can be performed, with a low risk of mortality; (6) operative mortality is not increased by the presence of congestive heart failure or pulmonary hypertension; (7) operation significantly increases longevity and restores the patients' subsequent survival to normal; and (8) operative closure of atrial septal defect can be recommended for all patients 60 years old or older, except for those with the most severe pulmonary vascular obstructive disease and those with associated debilitating medical illnesses that result in shortened survival.

Sutton MG et al. Circulation. 1981;64:402–409

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Risc-benefici del tractament quirúrgic > 40 anys

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PII S0735-1097(01)01635-7

Adult Congenital Heart Disease

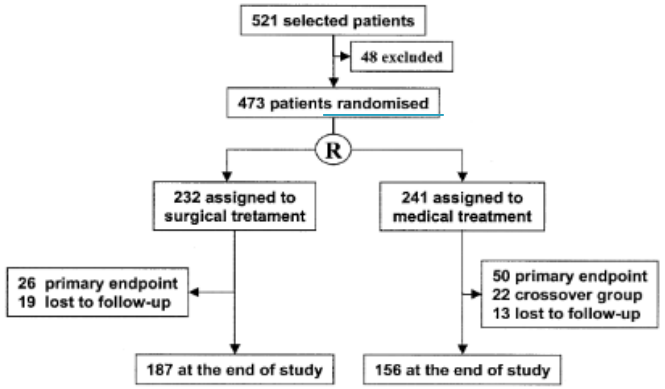
Surgical Treatment for Secundum Atrial Septal Defects in Patients >40 Years Old

A Randomized Clinical Trial

Fause Attie, MD, FACC, Martín Rosas, MD, PHD, Nuria Granados, MD, Carlos Zabal, MD, Alfonso Buendía, MD, Juan Calderón, MD
México City, México

AMOR·SCIENTIA·QVE·INSERVIANT·CORDI
MEXICO
INSTITVTO·N·DE CARDIOLOGIA IGNACIO·CHAVEZ

JACC Vol. 38, No. 7, 2001
December 2001:2035–42



Attie et al. J Am Coll Cardiol 2001;38(7):2035-42.

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Risc-benefici del tractament quirúrgic > 40 anys

Criteris d'inclusió

- > 40 anys
- CIA OS 88.2% o sí venós sinus 11.8%.
- Qp/ Qs ≥ 1,7
- PAPs < 70 mm Hg
- Sense comorbilitats
- **CF I i II**
- November 1985 and August 1998

< 50 anys:

palpitacions, vèrtigen i/o buf casual

> 50 anys:

díspnea, palpitacions, FA o flutter A o síncope

Attie et al. J Am Coll Cardiol 2001;38(7):2035-42

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Endpoint	Medical (n =241) >60 a (n=66)	Surgical (n=232) >60 a (n=45)	p value
Primari			
Total events	50 (20,7%)	26 (11,1%)	0,0046
IC	9 (3,7%)	5 (2,1%)	0,3982
Embòlia pulmonar	4 (1,6%)	5 (2,1%)	0,8266
Embòlia perifèrica	1(0,4%)	3 (1,3%)	0,4058
Ictus	4 (1,6%)	6 (2,6%)	0,3727
Neumònia recurrent	24 (10%)	6 (2,6%)	0,0012
Mort sobtada	7 (2,9%)	2 (0,9%)	0,0837
Secundari			
Mortalitat total	14 (5,8%)	10 (4,3%)	0,1934

Seguiment mig 7,3 (2-13 anys)

Attie et al. J Am Coll Cardiol 2001;38(7):2035-42

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	Medical	Surgical
Milloria CF *	0 %	57 %
Empitjorament CF *	6,2 %	0 %
IT massiva	10 %	12 %
FA /Flutter A	8,7 % (7 anys)	7,4 % (2 anys)

Onset of AF or flutter was related to long-term mortality in the medical group, but not in the surgical group

Temps sense event major CV: 13.7±0.2 anys (95% IC 12.9-14.9 anys).

Supervivència sense events:

99% a 1 any, 89% a 5 anys, 79% a 10 anys i 58% a 15 anys

Attie et al. J Am Coll Cardiol 2001;38(7):2035-42

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JACC Vol. 38, No. 7, 2001
December 2001:2035–42

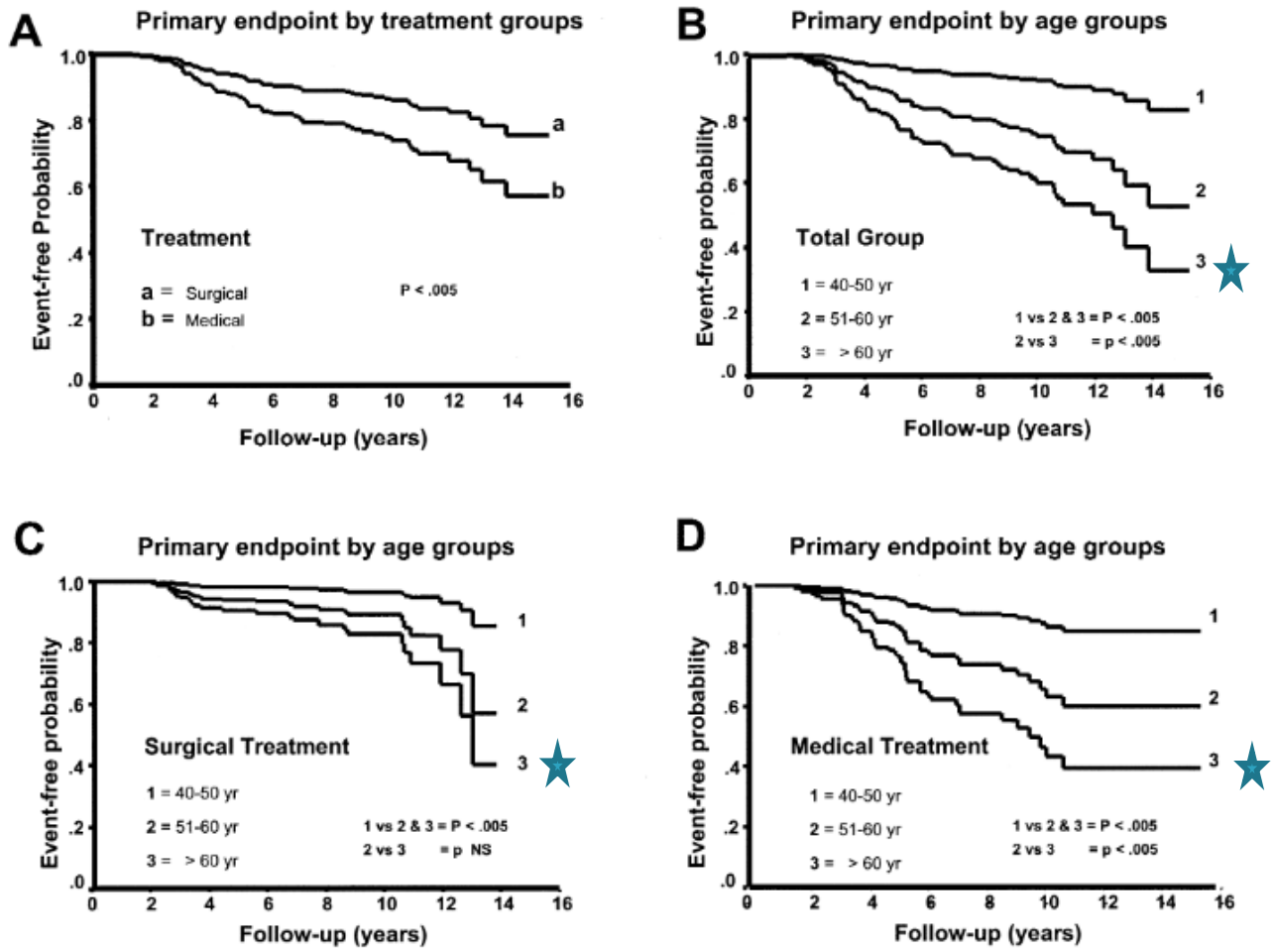

Figure 2. Event-free probability for primary end point by type of treatment and age at entry.

Attie et al. J Am Coll Cardiol 2001;38(7):2035-42

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Table 4. Forward Multivariate Cox Regression Analysis for Secondary End Point (Death)

Variables	B	SE	Wald's Statistic	*df*	Sig*	R	HR	95% CI Lower	95% CI Upper
Medical treatment	1.4088	0.5438	6.7122	1	0.0096	0.1357	4.0911	1.4092	11.8768
CI <3.5 l/m^2	1.1329	0.4819	5.5259	1	0.0187	0.1173	3.1045	1.2072	7.9836
Age at entry (years)			10.2493	2	0.0059	0.1562			
50–59	1.1198	0.5975	3.5119	1	0.0609	0.0768	3.0643	0.9499	9.8846
≥60	1.9799	0.6199	10.2021	1	0.0014	0.1790	7.2418	2.1489	24.4046
Previous AF or flutter	1.2263	0.4856	6.3782	1	0.0116	0.1308	3.4087	1.3160	8.8292
mPAP (>35 mm Hg)	1.6636	0.4937	11.3531	1	0.0008	0.1911	5.2783	2.0055	13.8918

*Model significance: overall score (6*df*) = 49.13, −2LLR (6*df*) = 207.105, p < 0.0001.
AF = atrial fibrillation; B = regression coefficient; *df* = degree of freedom; R = R-adjusted value; SE = standard error of B; other abbreviations as in Tables 1 and 2.

Attie et al. J Am Coll Cardiol 2001;38(7):2035-42

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Conclusions. We conclude that surgical treatment of ASDs was superior to medical treatment in improving both the composite of major cardiovascular events and overall mortality in patients >40 years old. Superiority was related to the cardiac index, mPAP and age at diagnosis. Because of the higher risk of morbidity and mortality, we believe that anatomic closure should always be performed as the initial treatment for ASD in adults >40 years old, with a Qp/Qs ratio ≥1.7 and pulmonary artery systolic pressure <70 mm Hg. The operation must be performed as soon as possible, even when the symptoms and hemodynamic impact seem to be minimal.

Attie et al. J Am Coll Cardiol 2001;38(7):2035-42

Tractament quirúrgic : Cirurgia mini-invasiva +/- MAZE +/- anell tricuspidi

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Tancament quirúrgic CIA + FA posterior

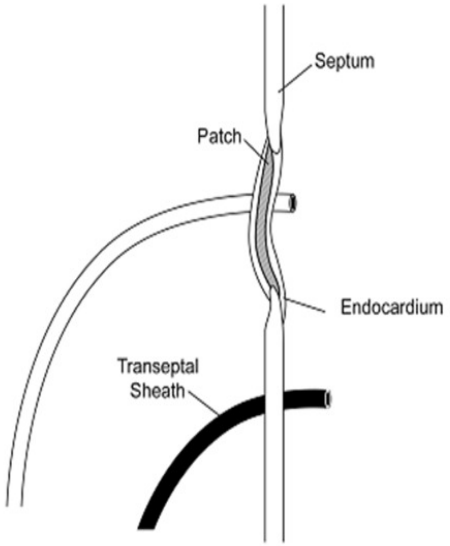
Table 1

Clinical features of patients with complete follow up

	At presentation (n=89)		At last follow up (n=86)	
	Group I (35–50 years)	Group II (51–72 years)	Group I (35–50 years)	Group II (51–72 years)
Number	51	38	51	35
NYHA class (%)				
I	4 (8)	3 (7)	40 (78)*	15 (43)*
II	18 (35)	13 (34)	4 (8)*	10 (28.5)*
III–IV	29 (57)	22 (56)	7 (14)*	10 (28.5)*
Mean (range) PAP (mm Hg)	37.8 (21–49)	40.3 (26–51)	21.3 (12–38)*	35.9 (23–49)
In AF (%)	12 (23.5)	17 (44)*	4 (8)*	12 (34)
Taking diuretics (%)	13 (25.5)	10 (26.3)	11 (21.5)	16 (46)
Taking warfarin (%)	11 (21.5)	17 (44)*	4 (8)*	12 (34.2)

*p<0.05 versus corresponding group at presentation.

AF, atrial fibrillation; NYHA, New York Heart Association; PAP, pulmonary artery pressure.



Gosh et al. Heart. 2002 Nov; 88(5): 485–487.

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Risc-benefici del tancament percutani

European Heart Journal (2011) **32**, 553–560
doi:10.1093/eurheartj/ehq352

CLINICAL RESEARCH
Congenital heart disease

Benefit of atrial septal defect closure in adults: impact of age

Michael Humenberger[1†], Raphael Rosenhek[1†], Harald Gabriel[1], Florian Rader[1], Maria Heger[1], Ursula Klaar[1], Thomas Binder[1], Peter Probst[1], Georg Heinze[2], Gerald Maurer[1], and Helmut Baumgartner[1,3*]

[1]Department of Cardiology, Medical University of Vienna, Vienna, Austria; [2]Center for Medical Statistics, Informatics and Intelligent Systems, Medical University of Vienna, Vienna, Austria; and [3]Adult Congenital and Valvular Heart Disease Center, Department of Cardiology and Angiology, University of Muenster, Albert Schweitzer Str. 33, 48149 Muenster,

Criteris d'inclusió
> 40 anys
CIA OS Anatomia favorable
RVP< 5 U/ m ^{2} després de test NO o tractament mèdic

Amplatzer: AAS 6 mesos

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Risc-benefici del tancament percutani

N=233

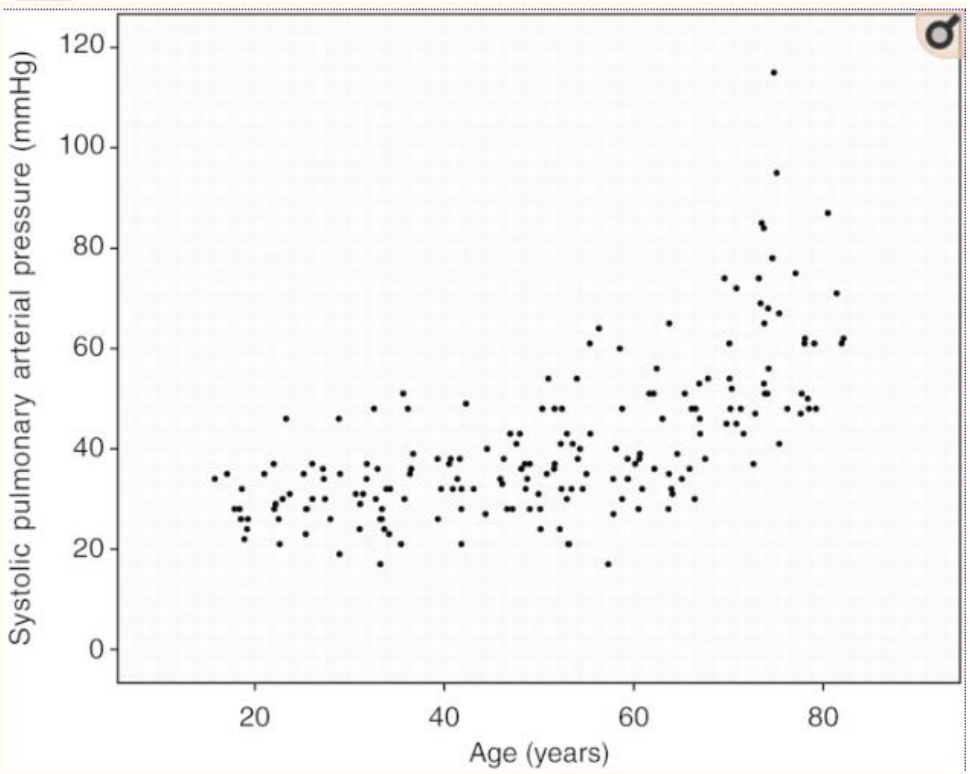

Figure 1

Correlation between systolic pulmonary artery pressure and age ($r = 0.65$, $P < 0.0001$).

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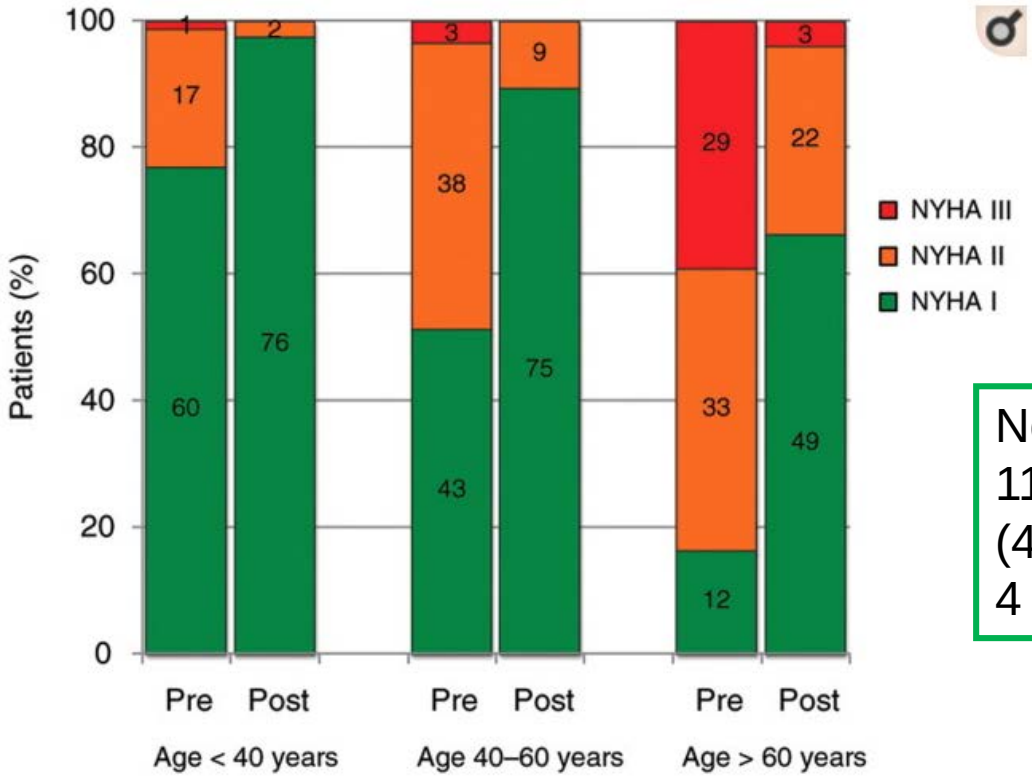
Risc-benefici del tancament percutani: CF i FA

N=233

N=78 N=84 N=74



Nova FA:
11 FA 1ª setm post
(4 RS, 4 CVE, 3 CVF)
4 FA entre 3-6 mesos

13 % a 3% 49 a 11% 83 a 34%

FA 0% FA parox 9,5% FA parox 18,9%
FA per 2,4% FA per 32,4%

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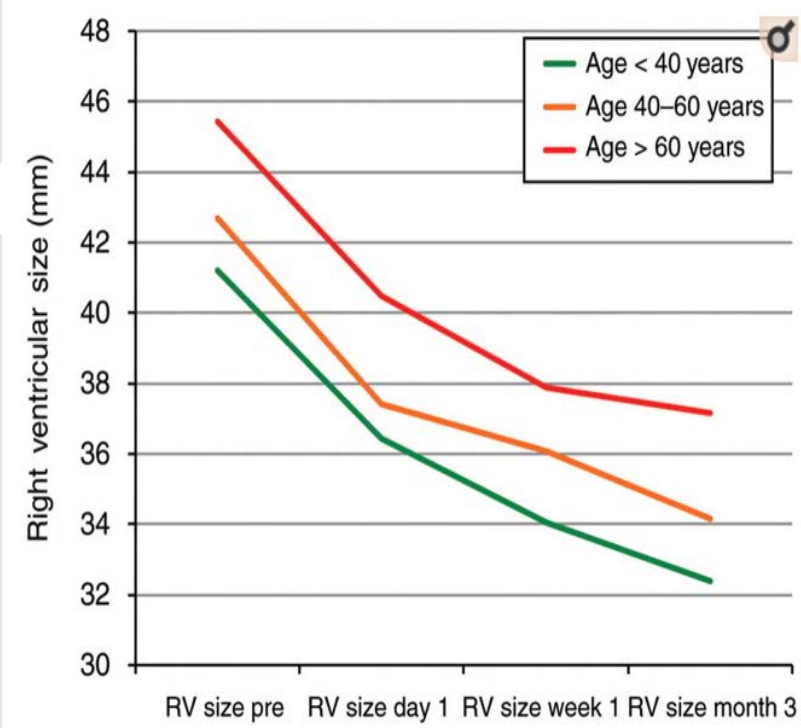
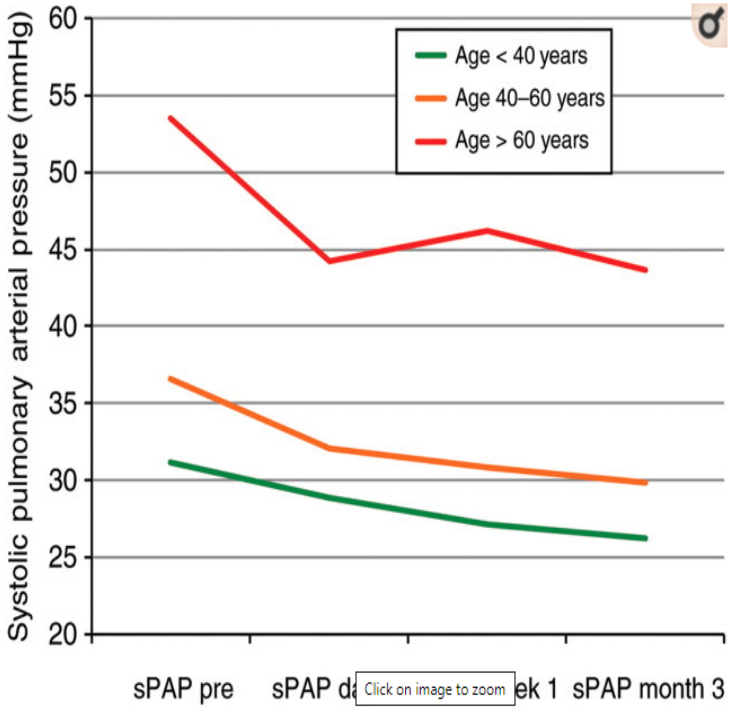
Risc-benefici del tancament percutani: VD, PAP, IT





De 41 ± 7, 43 ± 7, i 45 ± 6 mm
A 32 ± 5, 34 ± 5, i 37 ± 5 mm

De 31 ± 7, 37 ± 10, i 53 ± 17 mmHg
a 26 ± 5, 30 ± 6, i 43 ± 14 mmHg

IT moderada 9,7 % (78% > 60 anys) (de 23p pre a 17 p post)
IT severa 3% (7p pre a 2p post)

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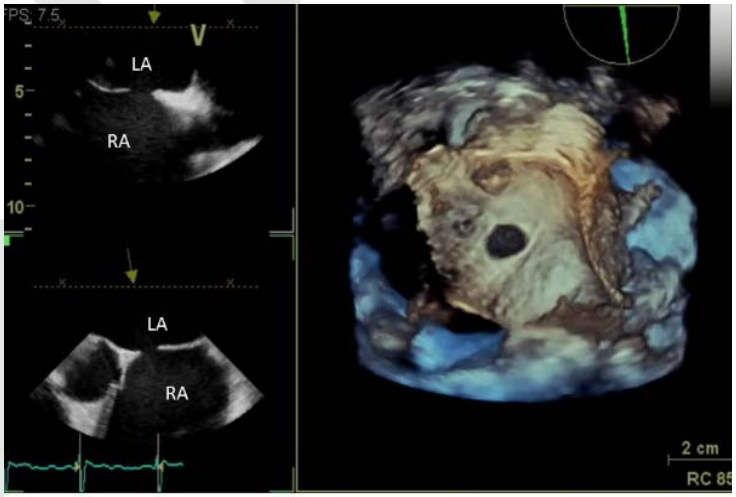
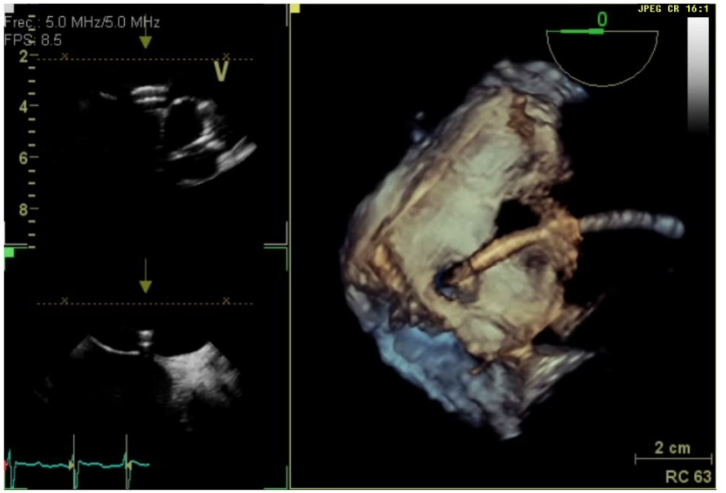
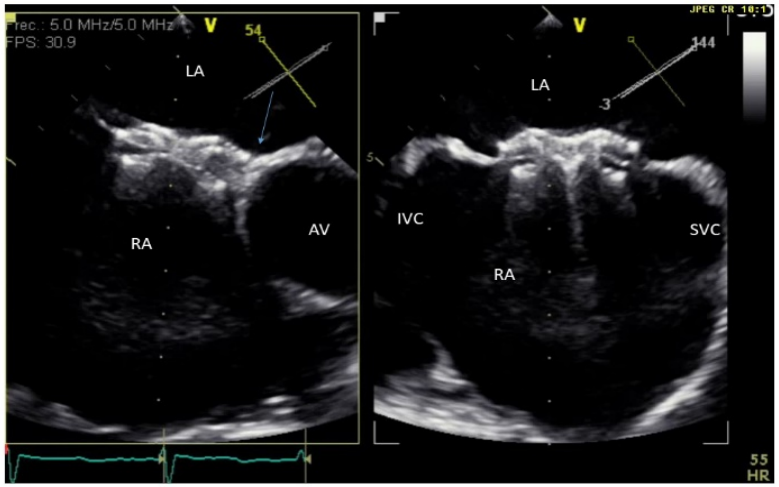
Conclusions

Transcatheter ASD closure can be safely and successfully performed in adults at any age. Regression of RV size and PAP as well as symptomatic improvement can be expected across all age groups. However, the best outcome is achieved in patients with less functional impairment and less elevated PAP. Considering the continuous increase in symptoms, RV remodelling, and PAP with increasing age, ASD closure must be recommended irrespective of symptoms early after diagnosis even in adults of advanced age.

Tancament percutani de la CIA







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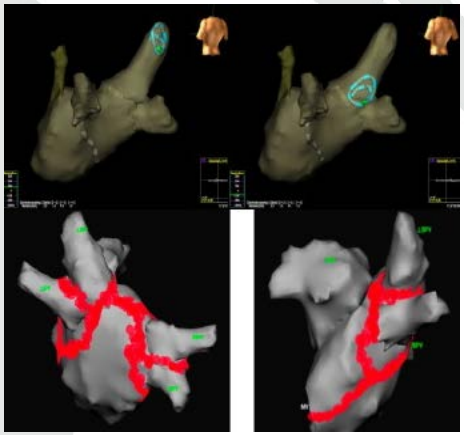
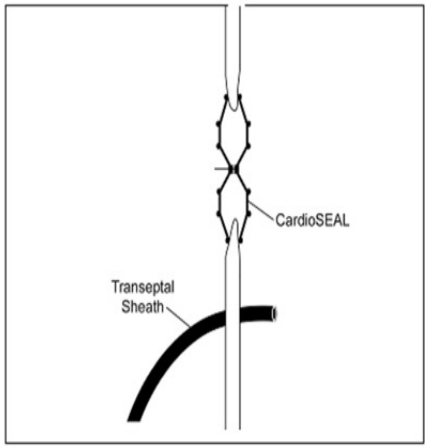
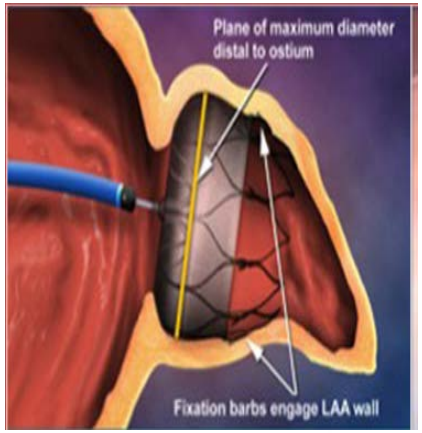
Complicacions del tancament CIA edat avançada

- Erosió: 0.04-0.3% :bora superior petita, defectes grans respecte la mide del septum, dispositius oversized device
- FA: anticoagulació (Ictus), ablació, casos seleccionats tancament orelleta.







Conduction abnormality and arrhythmia after transcatheter closure of atrial septal defect. *Komar M, et al Circ J. 2014; 78(10):2415-21.*

- Evolució IT massiva + ICD : Tricuclip?
- Vasculopatia pulmonar severa: Augmenta mortalitat!!!

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Prevalença de CIA+ HTP

CIA: 1,6 de cada 1000 nounats
CC: 97 % sobreviuen a l'edat adulta

CIA OS Diagnòstic 25-30% a l'edat adulta

Comunicació interauricular	Definició HTP	Nº de pacients	% HTP
Clínica Mayo [1]	RVP > 7U/m2	702	6%
EuroHeart Survey [2]	PAPs > 40 m Hg	896	23%
CONCOR registry [3]	PAPs > 40 mm Hg	437	8.2%

1.Circulation 1987;76:1037-42
2.Heart. 2007;93:682-7.
3.Int J Cardiol. 2014;174:299-305.

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Prevalença de CIA+ HTP

Table 1. Risk of PVD in Differing Lesions Associated With CHD and Increased Pulmonary Blood Flow.[24-27] These data are adapted from reference 98.

CHD WITH INCREASED PULMONARY BLOOD FLOW AND/OR PRESSURE		
DEFECT	**RISK OF PVD**	**AGE OF OCCURRENCE**
Truncus Arteriosus	~100%	<2 years
A-V Septal Defect	~100%	~2 years
Transportation of Great Arteries + VSD	~70%-100%	1-2 years
Patent Ductus Arteriosus	~15%-20%	>2 years
Ventricular Septal Defect	~15%-20%	>2 years
Atrial Septal Defect	~20%	>20 years

Defects in bold and italics represent high-flow/direct high-pressure lesions; defects in italics represent high-flow/variable direct high-pressure lesions; ASD is a high-flow lesion without a direct pressure stimulus from the systemic ventricle.

Advances in P DOI:10.21693/1933-088X-18.1.19
Pulmonary Hypertension *Volume 18, Number 1; 2019*

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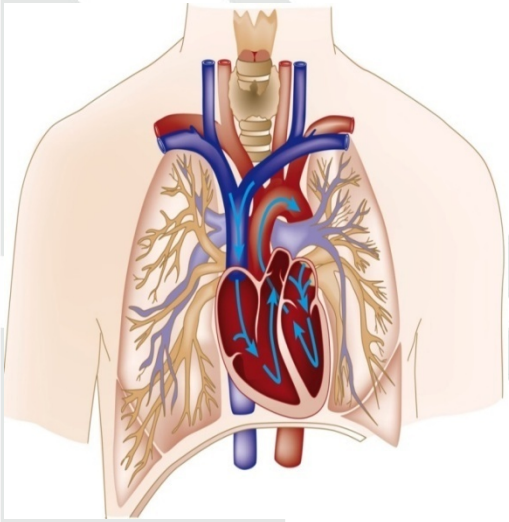
Nova classificació



1. HAP

1.1 HAP idiopática (HAPi)

1.2 HAP hereditaria (HAPh)

1.3 Inducida por fármacos y tóxicos*

1.4 Asociada (HAPA):

1.4.1 Conectivopatía

1.4.2 Infección por HIV

1.4.3 Hipertensión portal (HPoP)

1.4.4 Cardiopatía congénita

1.4.5 Esquistosomiasis

1.5 Respondedores a largo plazo de los bloqueadores de los canales de calcio

1.6 Enfermedad pulmonar veno-oclusiva y/o hemangiomatosis pulmonar capilar

1.7 HP persistente del recién nacido (PPHN)

2. HP por enfermedad del corazón izquierdo

3. HP por enfermedad pulmonar y/o hipoxia

4. HPTEC (Hipertensión Pulmonar Tromboembólica crónica) y otras obstrucciones de la arteria pulmonar

5. HP poco clara y/o causada por mecanismos multifactoriales

5.1 Alteraciones hematológicas

5.2 Alteraciones sistémicas

5.3 Alteraciones metabólicas

5.4 Otras

Clasificación actualizada de fármacos y tóxicos asociados con HAP[2]

Confirmado	Posible
Aminorex Fenfluramina Dexflenfuramina Benfluorex **Metanfetaminas*** **Dasatinib*** Aceite de colza ~~Inhibidores selectivos de la recaptación de serotonina~~	Cocaina Fenilpropanolamina **L-Triptófano**# Hierba de San Juan **Anfetaminas**# Interferón α y β Agentes alquilantes Bosutinib **Antivirales directos hepatitis C**& **Leflunomida**& **Indirubin** (hierba china Qing-Dai)&

[1]Galiè N, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J.* 2016;37(1):67-119

[2]Simonneau et al. Haemodynamic definitions and updated Clinical classification of pulmonary hypertension. *Eur Respir J* 2019;53(1)

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Nova definició

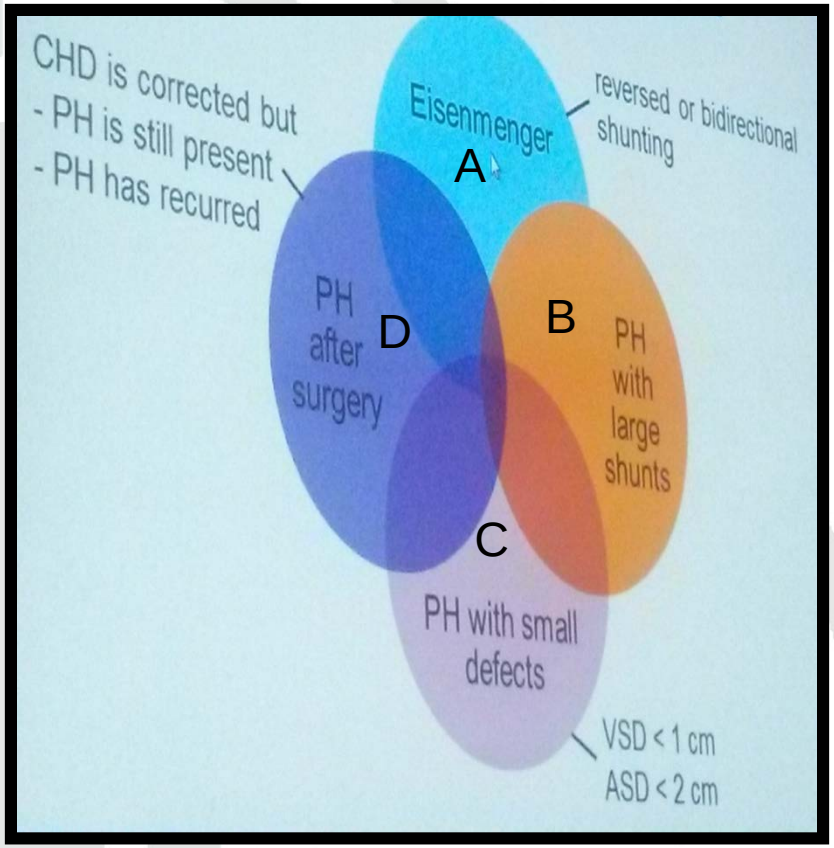
ABANS (Galié et al. EHJ 2015)[1]		Grup
HP	PAPm ≥ 25 mmHg	Tots
HP precapilar	PAPm ≥ 25 mmHg PEP ≤ 15 mmHg	1,3,4 y 5
HP poscapilar	PAPm ≥ 25 mmHg PEP > 15 mmHg	2 y 5
HP poscapilar aislada	GTPd < 7 mmHg RVP ≤ 3 UW	
HP combinada (pre y poscapilar)	GTPd ≥ 7 mmHg RVP > 3 UW	

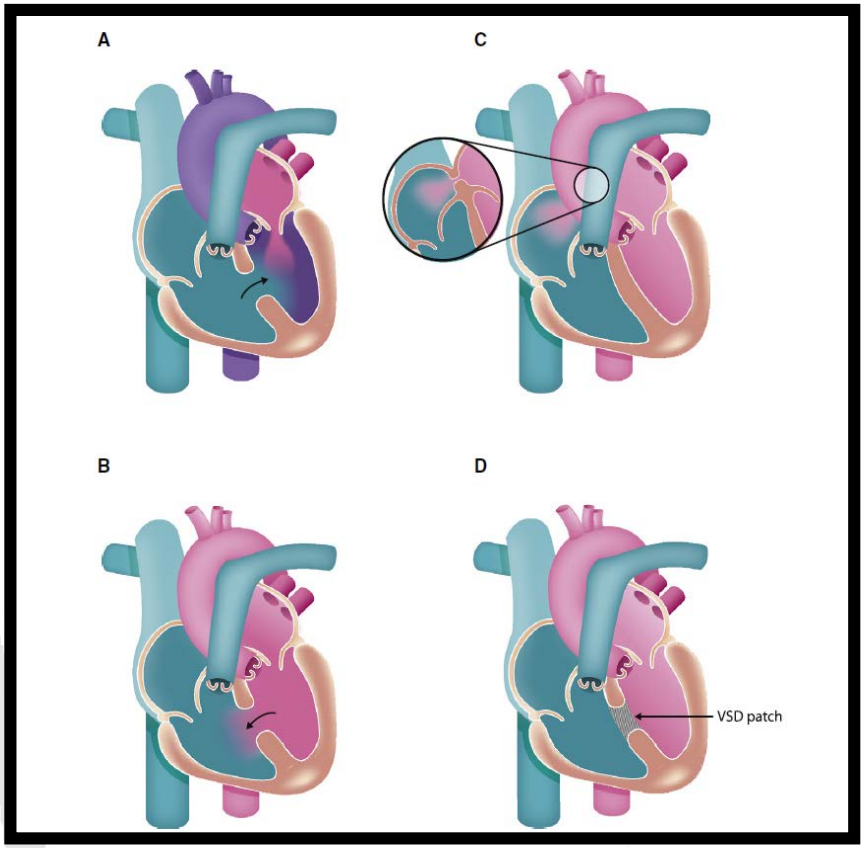
ARA (propuesta NIZA 2018)[2]			Grup
HP	**HP pre-capilar**	PAPm > 20 mmHg PEP ≤ 15 mmHg RVP ≥ 3 UW	1, 3, 4 y 5
	HP post-capilar aislada	PAPm > 20 mmHg PEP > 15 mmHg RVP < 3 UW	2 y 5
	HP combinada (pre y post capilar)	PAPm > 20 mmHg PEP > 15 mmHg RVP ≥ 3 UW	2 y 5

[1]Galiè N, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J.* 2016;37(1):67-119

[2]Simonneau et al. Haemodynamic definitions and updated Clinical classification of pulmonary hypertension. *Eur Respir J* 2019;53(1)

Classificació clínica dels shunts i HTP





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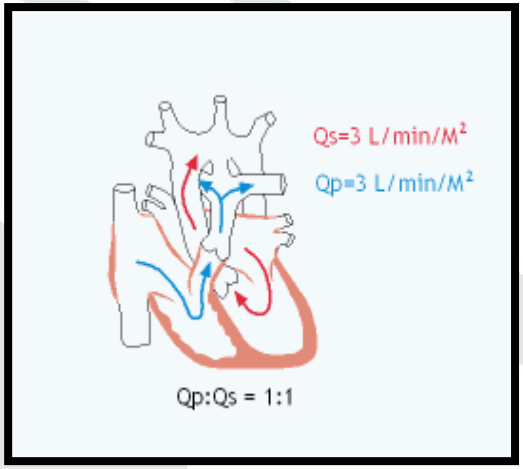
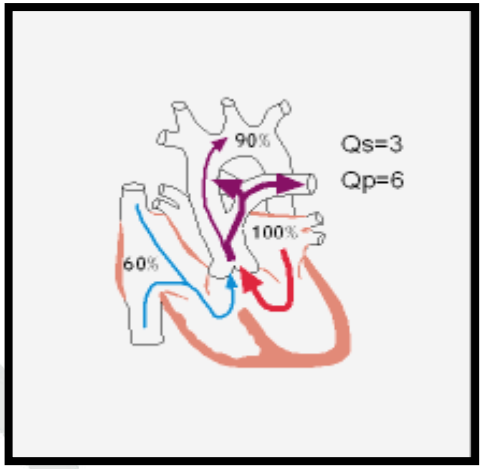
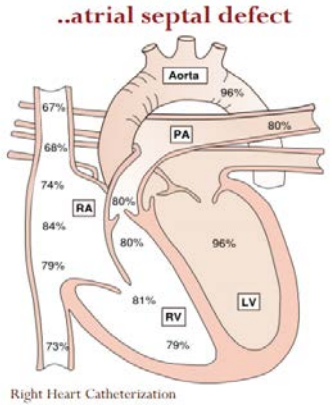
ESTUDIS HEMODINÀMICS: Shunts. oximetries







Salt oximètric > 7%

- En condicions normals el CC p = CC s, Qp/ Qs =1.
- Si hi ha un shunt E-D (CIA): CC p = CC s + shunt.
- Si hi ha un shunt D-E (Eisenmenger): CC s = CC p + shunt.

Qp/ Qs major de 1.5, s'ha de tancar.

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ESTUDIS HEMODINÀMICS:
Shunts. oximetries

- Hipertensió pulmonar ≠ malaltia vascular pulmonar

- Llei d' Ohm

$$PAP\ m = RVP * Qp + PAE$$

PAPm: Pressió artèria pulmonar mitja
PAE m: pressió aurícula esquerra

Qp: flux pulmonar
Qp index: flux pulmonar indexat

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ESTUDIS HEMODINÀMICS:
Shunts. oximetries

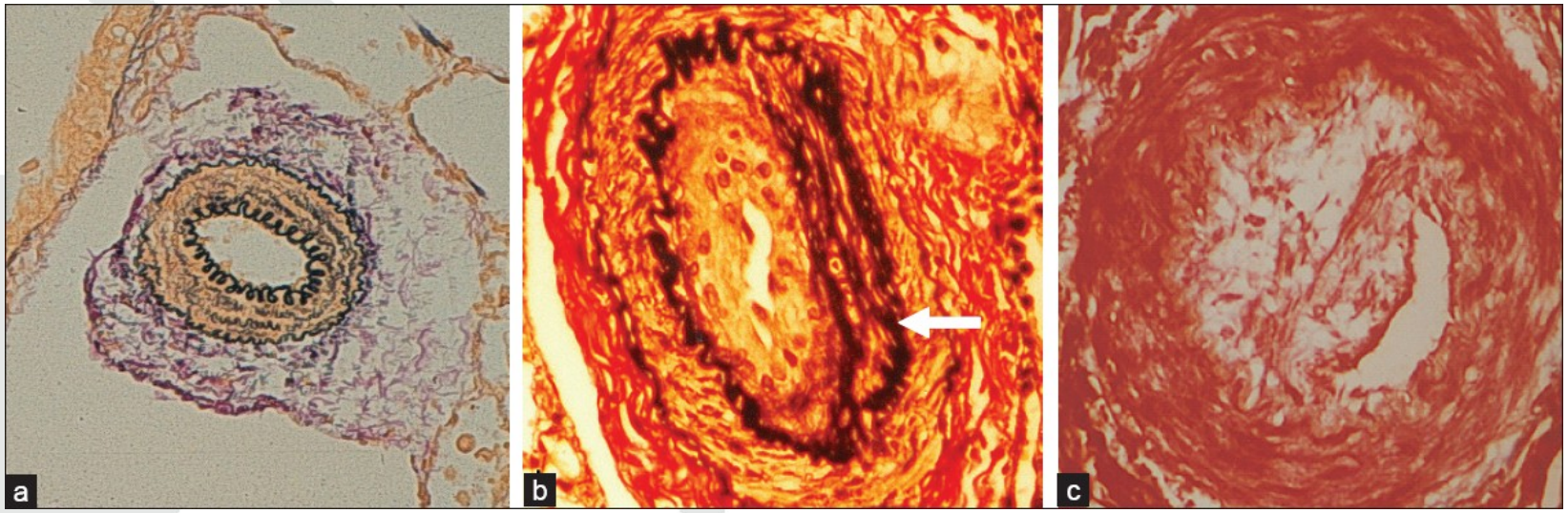
RVP = (PAPm - PAE m) / Q p (WU)

RVPi = (PAPm - PAE m) / Q p index (L / m2) WU * m2

PAPm: Pressió artèria pulmonar mitja
PAE m: pressió aurícula esquerra

Qp: flux pulmonar
Qp index: flux pulmonar indexat

Sdr. Eisenmenger



Disfunció de l'endoteli vascular i lliberació de substàncies vasoactives (endotelina 1 i tromboxà A2) i un dèficit de NO i prostaciclines

Fins lesions plexiformes, que són irreversibles.

Tractament de la CIA amb HTP

PAPm <2/3 PASm
RVP <4 UW * m2
RVP: RVS <0.3
Qp / Qs> 1.5

SÍ

TANCAMENT PERCUTANI O QUIRÚRGIC

NO

TEST VASODILATADOR
TEST OCLUSIÓ
DESATURACIÓ EXERCICI
BOSENTAN 6 MESOS

TRACTAMENT MÈDIC

Table 1. Recommendations for Closure of Simple Shunt Defects in Adults With PAH

Recommendation for Closure of ASD	ESC/ERS 2015[13]	ACC 2018[14]
YES	PVRi <4 WU m²	Systolic PA pressure <one-half systemic PVR / SVR <0.3
INDIVIDUALIZE	PVRi 4 to 8 WU m²	Systolic PA pressure one-half to two-thirds systemic PVR / SVR 0.3 to 0.66
NO	PVRi >8 WU m²	Systolic PAP >two-thirds systemic PVR / SVR >0.66 and Qp/Qs <1.0

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Tractament sdr. Eisenmenger

Recommendations	Class[a]	Level[b]
Bosentan is recommended in WHO-FC III patients with Eisenmenger syndrome	I	B
Other ERAs, PDE-5is and prostanoids should be considered in patients with Eisenmenger syndrome	IIa	C
Combination drug therapy may be considered in patients with Eisenmenger syndrome	IIb	C

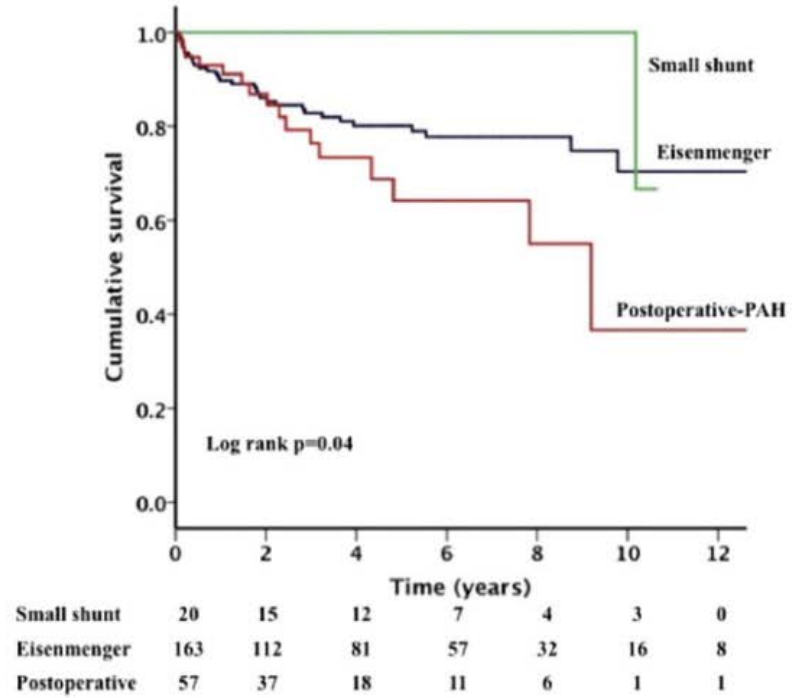
ESC/ERS GUIDELINES

Recommendations for Eisenmenger Syndrome		
COR	LOE	Recommendations
Therapeutic		
I	A	Bosentan is beneficial in symptomatic adults with Eisenmenger syndrome with ASD or VSD.
IIa	B-R	In symptomatic adults with Eisenmenger syndrome, bosentan and PDE-5 inhibitors are reasonable in combination if symptomatic improvement does not occur with either medication alone.

2018 ACHD GUIDELINES

Tractament de la CIA amb HTP

REHAP
Subanálisis en pacientes con HAP asociada a cardiopatías congénitas



Alonso et al. International Journal of Cardiology 184 (2015):717-723

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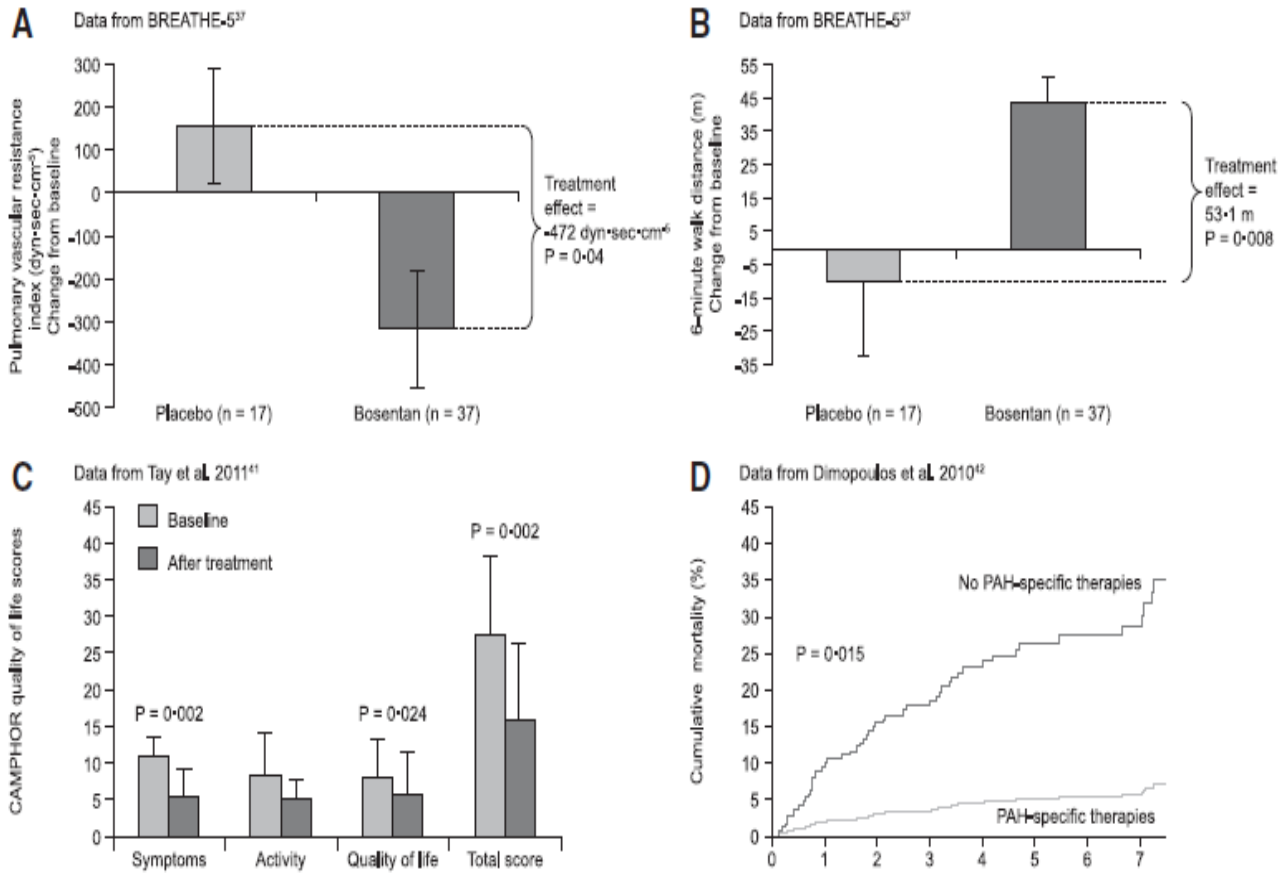
Tractament de la CIA amb HTP



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Tractament de la CIA amb HTP

- No hi ha guies basades en la evidència, pero sí estudis de seguiment de centres concrets que indueixen aquest maneig. No tancar defectes en el Sdr. Eisenmenger
- No tancar defectes petits amb fisiopatologia de HTP idiopàtica
- En pacients amb edat avançada, descartar altres causes de hipertensió pulmonar (Insuficiència mitral, disfunció diastòlica, patologia pulmonar, ...)

Int J Cardiol 2013; 168: 3897 a 3801

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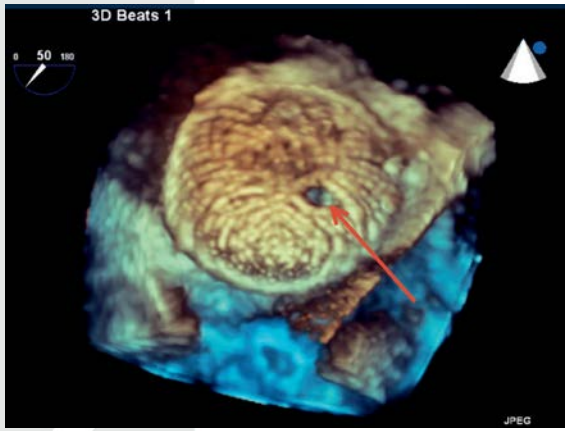
Tractament de la CIA amb HTP

Atrial Septal Defect–Associated Pulmonary Hypertension: Outcomes of Closure With a Fenestrated Device



42 pacients. Complets 25

Table 3. Change in Parameters Following FASD Implantation

Parameters	Baseline	Short-term follow-up	Long-term follow-up	P value
O_2 saturation at rest (%)	93±4 (n=25)	96±3 (n=22)	97±2 (n=12)	0.0066 (n=12)
O_2 saturation after exercise (%)	88±7 (n=7)	84±31 (n=9)	75±41 (n=5)	†
6MWT distance (m)	228±183 (n=14)	351±179 (n=12)	457±83 (n=5)	0.0081 (n=12)
NYHA functional class	n=27	n=23	n=12	
I	0 (0%)	7 (30.43%)	8 (66.7%)	
II	8 (32.0%)	13 (56.5%)	3 (25.0%)	
III	17 (68.0%)	3 (13.0%)	1 (8.3%)	
IV	0 (0%)	0 (0%)	0 (0%)	
RA pressure (mm Hg)	12±3 (n=24)	10±3 (n=17)‡	8±4 (n=5)	0.0502 (n=17)
LA pressure (mm Hg)	13±4 (n=23)	14±4 (n=16)‡	11±1 (n=4)	0.9252 (n=16)
MPAP (mm Hg)	46±19 (n=24)	42±17 (n=17)‡	42±22 (n=5)	0.0004 (n=17)

†Due to low n and high standard deviations for short- and long-term, only descriptive statistics are given. ‡Represents cardiac catheterization data from the immediate post-deployment period.

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Tractament de la CIA amb HTP

	Determinants of prognosis	Estimated 1-year mortality		
		Low risk < 5%	Intermediate risk 5-10%	High risk > 10%
Evaluación clínica	Clinical signs of right heart failure	Absent	Absent	Present
	Progression of symptoms	No	Slow	Rapid
	Syncope	No	Occasional syncope	Repeated syncope
	FC	I, II	III	IV
Pruebas de esfuerzo	6MWD	> 440 m	165 - 440 m	< 165 m
	CPET	Peak VO_2 > 15 ml/min/kg (> 65% pred.) VE/VCO_2 slope < 36	Peak VO_2 11 - 15 ml/min/kg (35-65% pred.) VE/VCO_2 slope 36 - 44.9	Peak VO_2 < 11ml/min/kg (< 35% pred.) VE/VCO_2 slope ≥ 45
Marcadores bioquímicos	NT-proBNP plasma levels	BNP < 50 ng/l NT-proBNP < 300 ng/l	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP > 300 ng/l NT-proBNP > 1400 ng/l
Evaluación ecocardiográfica	Imaging (echo, CMR)	RA area < 18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal pericardial effusion	RA area > 26 cm^2 Pericardial effusion
Evalaución hemodinámica	Haemodynamics	RAP < 8 mmHg CI ≥ 2.5 l/min/m^2 SvO_2 > 65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m^2 SvO_2 60–65%	RAP > 14 mmHg CI < 2.0 l/min/m^2 SvO_2 < 60%

Objetivo terapéutico: bajo riesgo

Alt risc: epoprostenol sc o ev

Galiè N, et al. Eur Heart J 2016; 37:67-119.

Risc intermig: Afegir a ARE+PDE5i el Selexipag vo
CIA reparada: inici combinacions: Macitentan+sildenafil/ bosentan+riociguat/ Selexipag+ AREi/o PDE5i

Galiè N et al. Risk stratification and medical therapy of pulmonary arterial hypertension. *Eur Respir J.* 2019;53(1)

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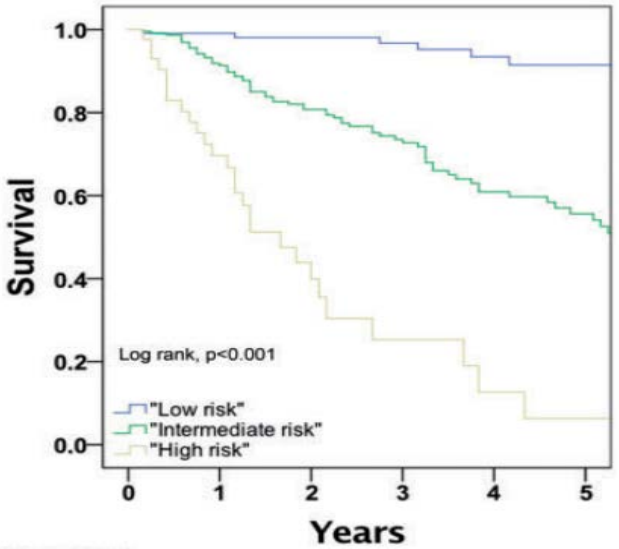
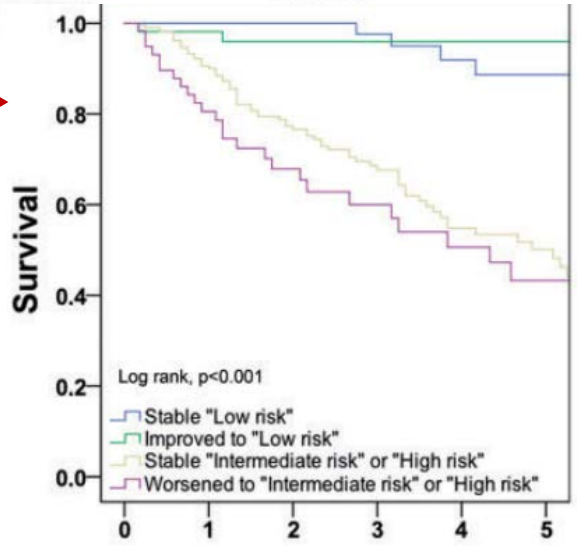
Tractament de la HTP

Recommendations	Class[a]	Level[b]
It is recommended to evaluate the severity of PAH patients with a panel of data derived from clinical assessment, exercise tests, biochemical markers and echocardiographic and haemodynamic evaluations (*Tables 13* and *14*)	I	C
It is recommended to perform regular follow-up assessments every 3–6 months in stable patients (*Table 14*)	I	C
Achievement/maintenance of a low-risk profile (*Table 13*) is recommended as an adequate treatment response for patients with PAH	I	C
Achievement/maintenance of an intermediate-risk profile (*Table 13*) should be considered an inadequate treatment response for most patients with PAH	IIa	C





Galiè N, *et al. Eur Heart J* 2016; 37:67-119.

Kylhammar D, EurHeart J 2018; 39:4175-418

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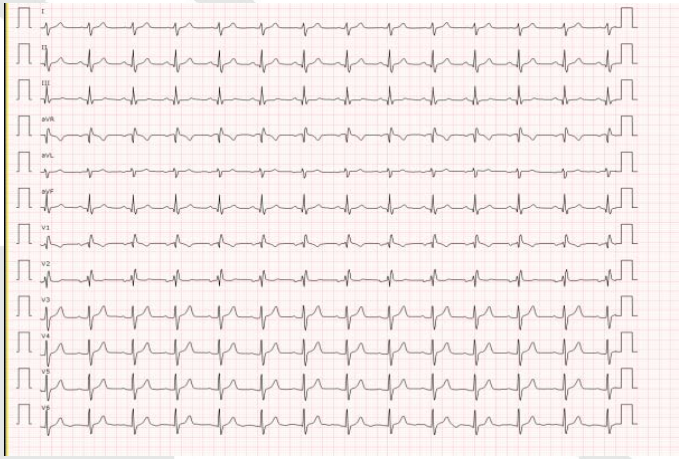
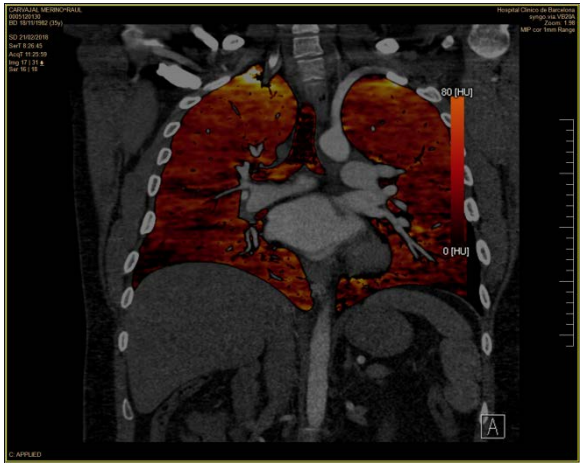
Altres causes d'HTP i patologies associades:

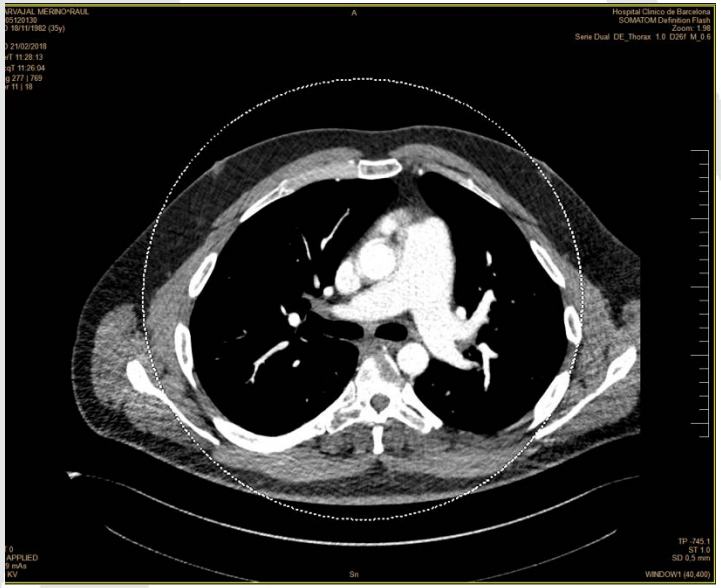
- TEP
- MPOC
- Valvulopatia mitral
- Disfunció diastòlica…

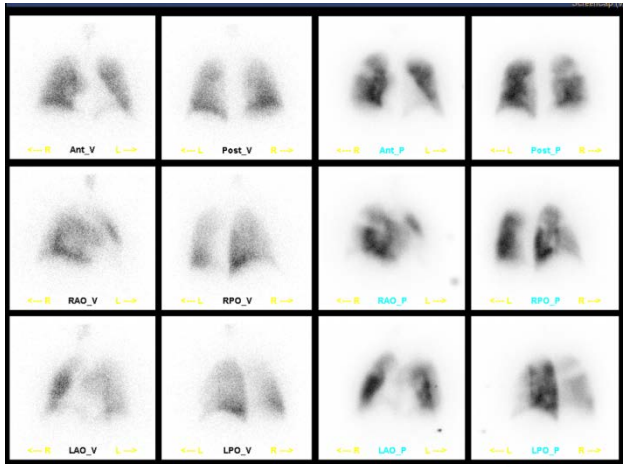
- Estenosi aòrtica.

Altres causes d'HTP: TEP + CIA









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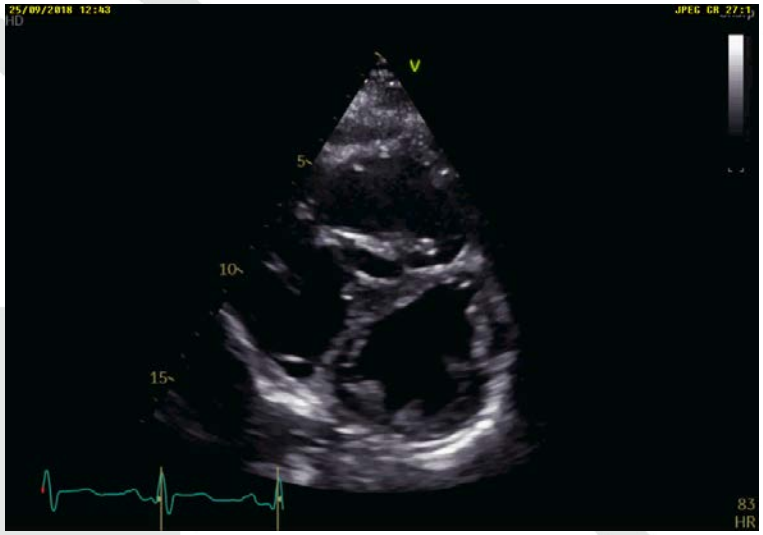
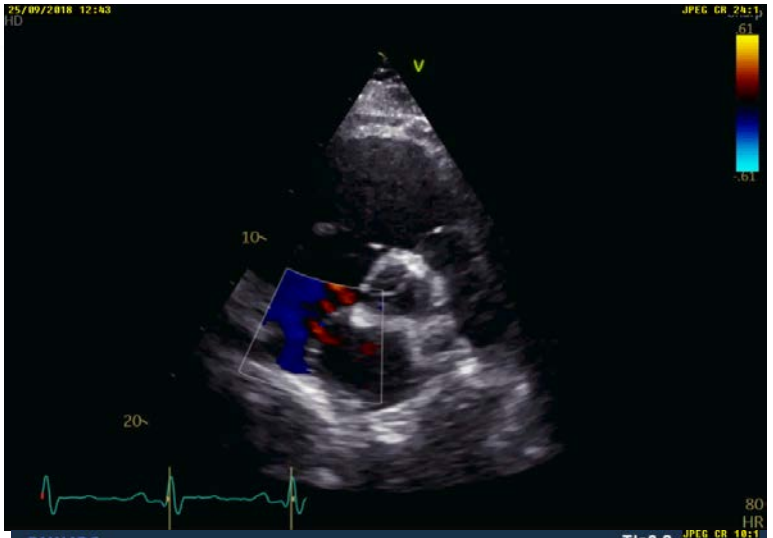
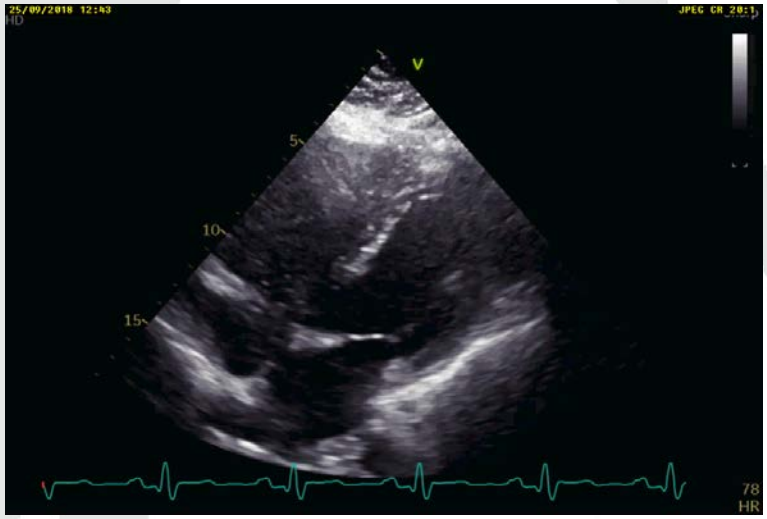
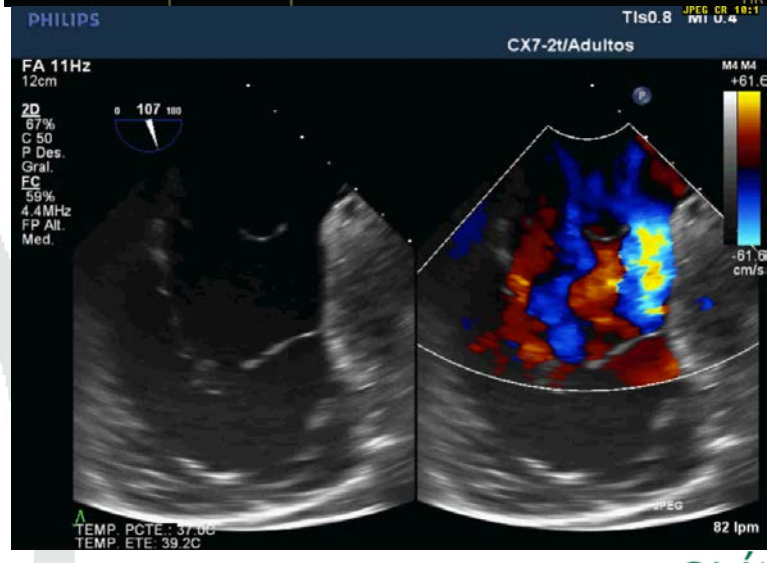
Altres causes d'HTP: TEP + CIA









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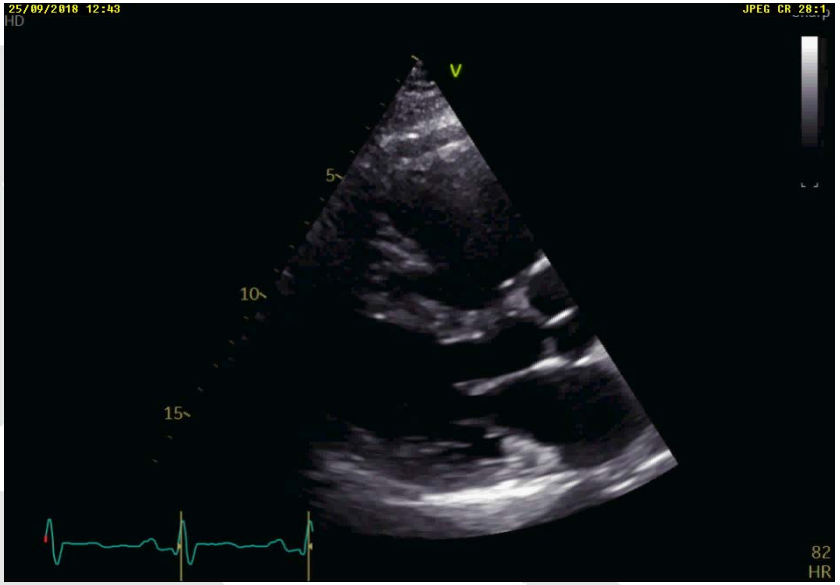
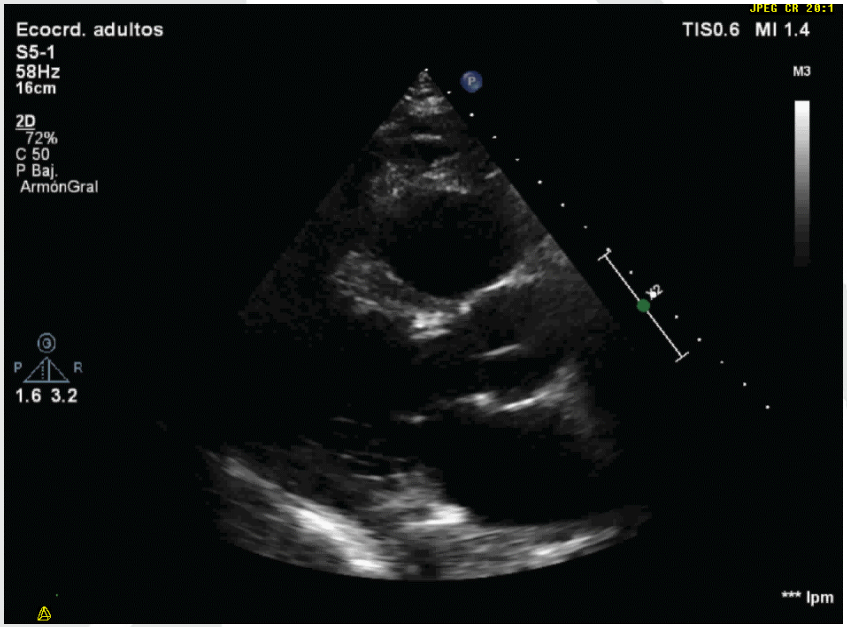
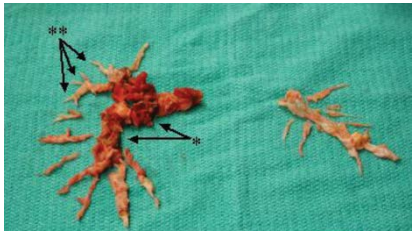


	Basal	(*)
QT (l/min)	8,02	16,50
IC (l/min/m2)	3,56	7,33
FC (lat./min)	79,00	125,00
PAP (S/D/M) (mmHg)	49,00/12/29	99,00/35/60
PAOP (mmHg)	5,00	19,00
PAD (mmHg)	02	10
TAS (S/D/M) (mmHg)	118,00/ 75,00/ 84,00	144,00/ 101,00/ 106,00
RVP (Wood)	2,99	2,48
RVP (din·s·cm-5)	238,90	198,15
RVS (Wood)	10,22	5,82
RVS (din·s·cm-5)	816,58	465,02
PvO2 (mmHg)	49	30
SvO2 (%)	82	52





	Basal
QT (l/min)	5,15
IC (l/min/m2)	2,23
FC (lat./min)	70,00
PAP (S/D/M) (mmHg)	33,00/16/22
PAOP (mmHg)	8,00
PAD (mmHg)	05
TAS (S/D/M) (mmHg)	116,00/ 69,00/ 79,00
RVP (Wood)	2,72
RVP (din·s·cm-5)	217,33
RVS (Wood)	14,37
RVS (din·s·cm-5)	1148,16
PvO2 (mmHg)	41
SvO2 (%)	70

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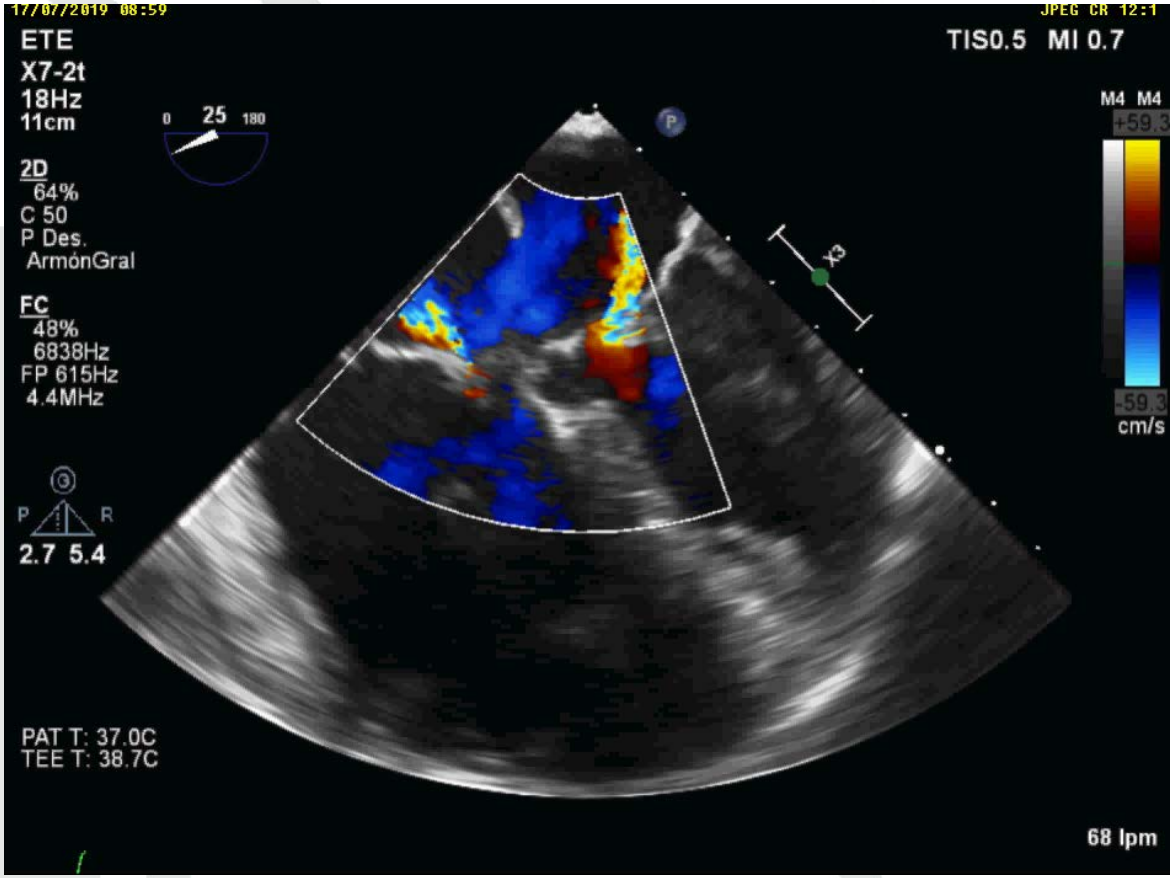
Altres causes d'HTP: IM+ CIA



- HTA
- Disfunció diastòlica

PAP m = RVP * Qp + PAE

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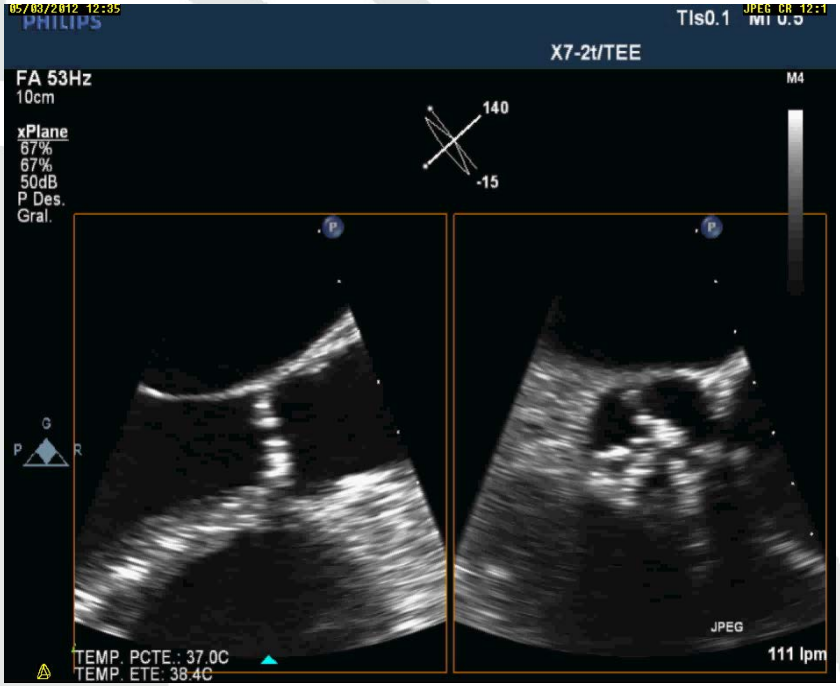
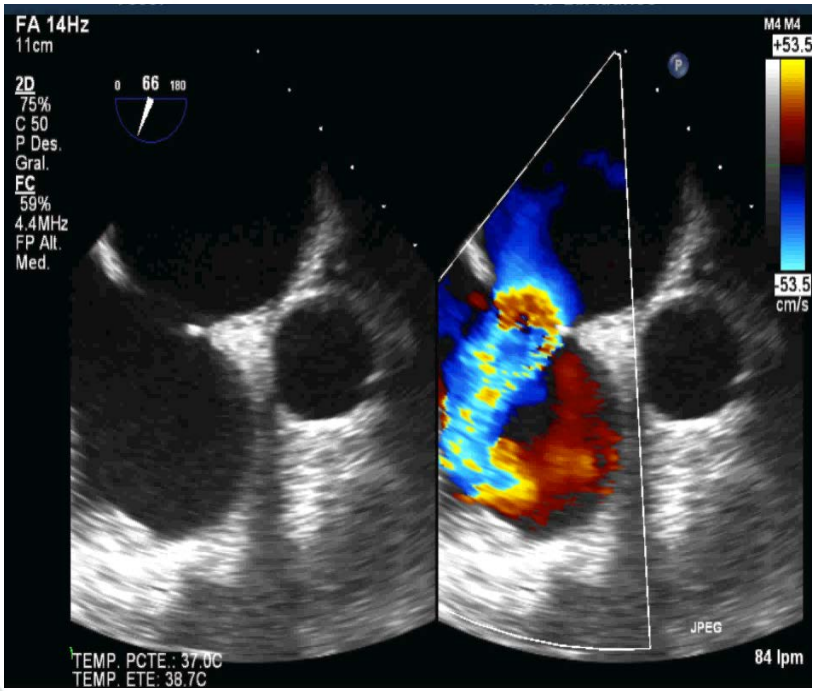
Patologies associades: EAo + CIA





PTS Ao + tancament CIA quirúrgic
TAVI + tancament CIA percutani

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CONCLUSIONS

- La CIA en el adult es detecta en un 25-30% dels pacients, i no és un fet estrany en gent gran.
- El tractament quirúrgic de la CIA en majors de 60 anys amb CF III-IV millora la simptomatologia i sembla millorar la supervivència, i en pacients en CF I- II millora la morbi-mortalitat, sobre tot en els pacients amb alteracions del IC, PAPm i major edat. El tancament s'ha de fer precoçment.
- El tractament percutani de la CIA és segur i efectiu, millora la CF, la mide del ventricle dret i la hipertensió pulmonar.
- Falta evidència sobre el tractament de la insuficiència tricuspidea en edat avançada, en cas de moderada-severa amb dilatació de l'anell T indicar cirurgia Mini invasiva versus tancament percutani +/- tricuclip
- Tractament de la FA: Anticoagulació. Ablació FA. Tancament orelleta en casos seleccionats

CONCLUSIONS

- La hipertensió pulmonar en les cardiopaties congènites es classifica al grup 1.
- L'evolució a sdr. Eisenmenger contraindica el tancament dels shunts i indica tractament farmacològic en pacients en CF II-III.
- Les cardiopaties congènites reparades amb hipertensió pulmonar són les que tenen pitjor pronòstic. L'ús de nous fàrmacs pot millorar les hospitalitzacions i l'evolució de la malaltia. Mantenir en baix risc (verd).
- En pacients d'edat avançada s'ha de valorar altres causes d'augment de la PAP (TEP, Valvulopatia mitral, disfunció diastòlica, MPOC…). Tromboendarterectomia, cirurgia valvular…
- En cas d'estenosi aòrtica individualitzar.

Moltes gràcies

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