Hipertensão Pulmonar nas Cardiopatias Congénitas

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Guidelines for the diagnosis and treatment of pulmonary hypertension

2009

Table 4 Updated clinical classification of pulmonary hypertension (Dana Point, 2008[1])

1 Pulmonary arterial hypertension (PAH)
- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2
  - 1.2.2 ALK1, endoglin (with or without hereditary haemorrhagic telangiectasia)
  - 1.2.3 Unknown
- 1.3 Drugs and toxins induced
- 1.4 Associated with (APAH)
  - 1.4.1 Connective tissue diseases
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis
  - 1.4.6 Chronic haemolytic anaemia
- 1.5 Persistent pulmonary hypertension of the newborn

1′ Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomatosis

2 Pulmonary hypertension due to left heart disease
- 2.1 Systolic dysfunction
- 2.2 Diastolic dysfunction
- 2.3 Valvular disease

3 Pulmonary hypertension due to lung diseases and/or hypoxaemia
- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental abnormalities

4 Chronic thromboembolic pulmonary hypertension

5 PH with unclear and/or multifactorial mechanisms
- 5.1 Haematological disorders: myeloproliferative disorders, splenectomy.
- 5.2 Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neurofibromatosis, vasculitis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: tumoural obstruction, fibrosing mediastinitis, chronic renal failure on dialysis

ALK-1 = activin receptor-like kinase 1 gene; APAH = associated pulmonary arterial hypertension; BMPR2 = bone morphogenetic protein receptor, type 2; HIV = human immunodeficiency virus; PAH = pulmonary arterial hypertension.

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European Heart Journal
doi:10.1093/eurheartj/ehp297

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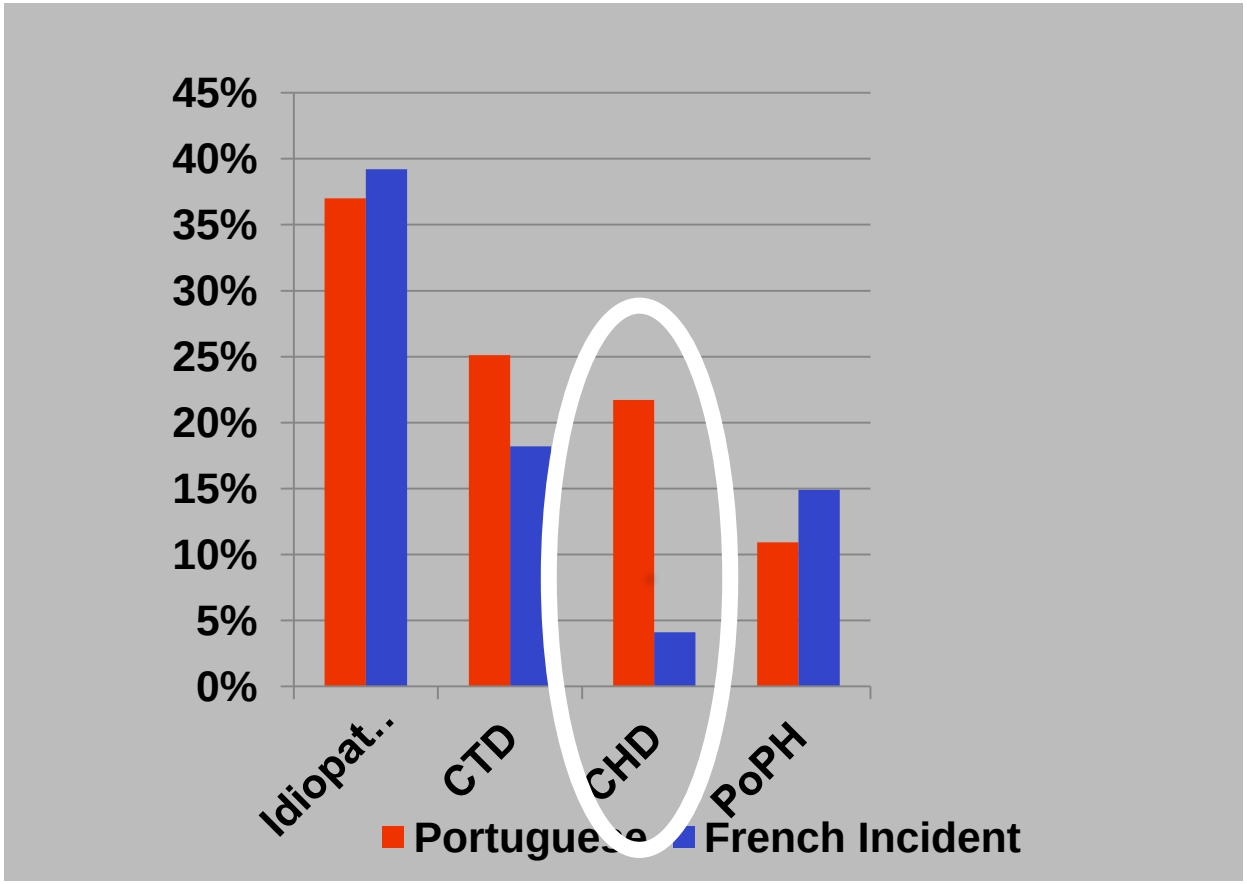
Pulmonary Hypertension in Portugal: first data from a nationwide registry

- 5 centros
- dts ≥ 18 anos
- admitidos entre Jan 2008 e Dez 2010

- HAP confirmada por cateterismo *(PAPm ≥ 25mmHg e PCPm e/ou teled VE≤15mmHg)*

Disease subtype, group 1 PAH



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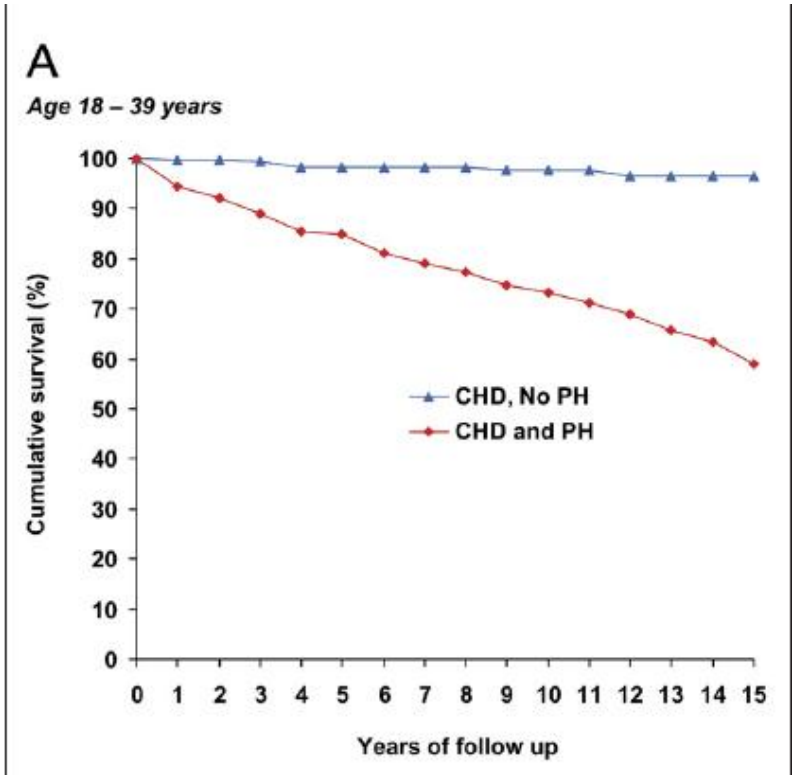
Baptista R, BioMed Research International, 2013
Humbert M et al; AJRCCM 2006; 173:1023-1030

Diagnosis of Pulmonary Hypertension in the Congenital Heart Disease Adult Population

Impact on Outcomes

Lowe BS JACC 2011; 58:538-46



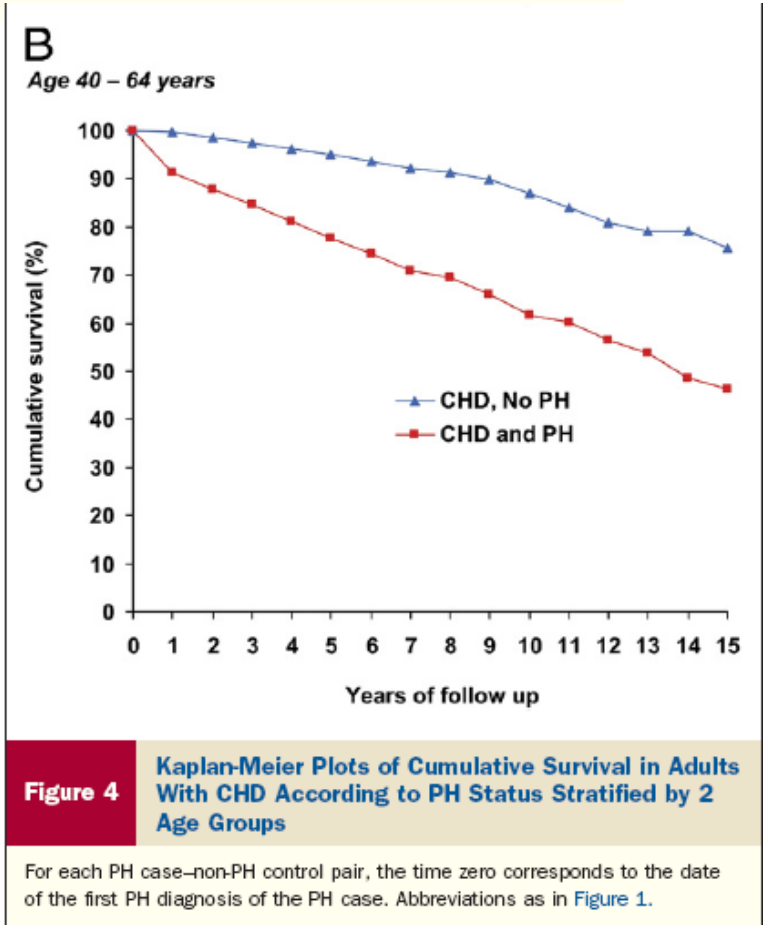
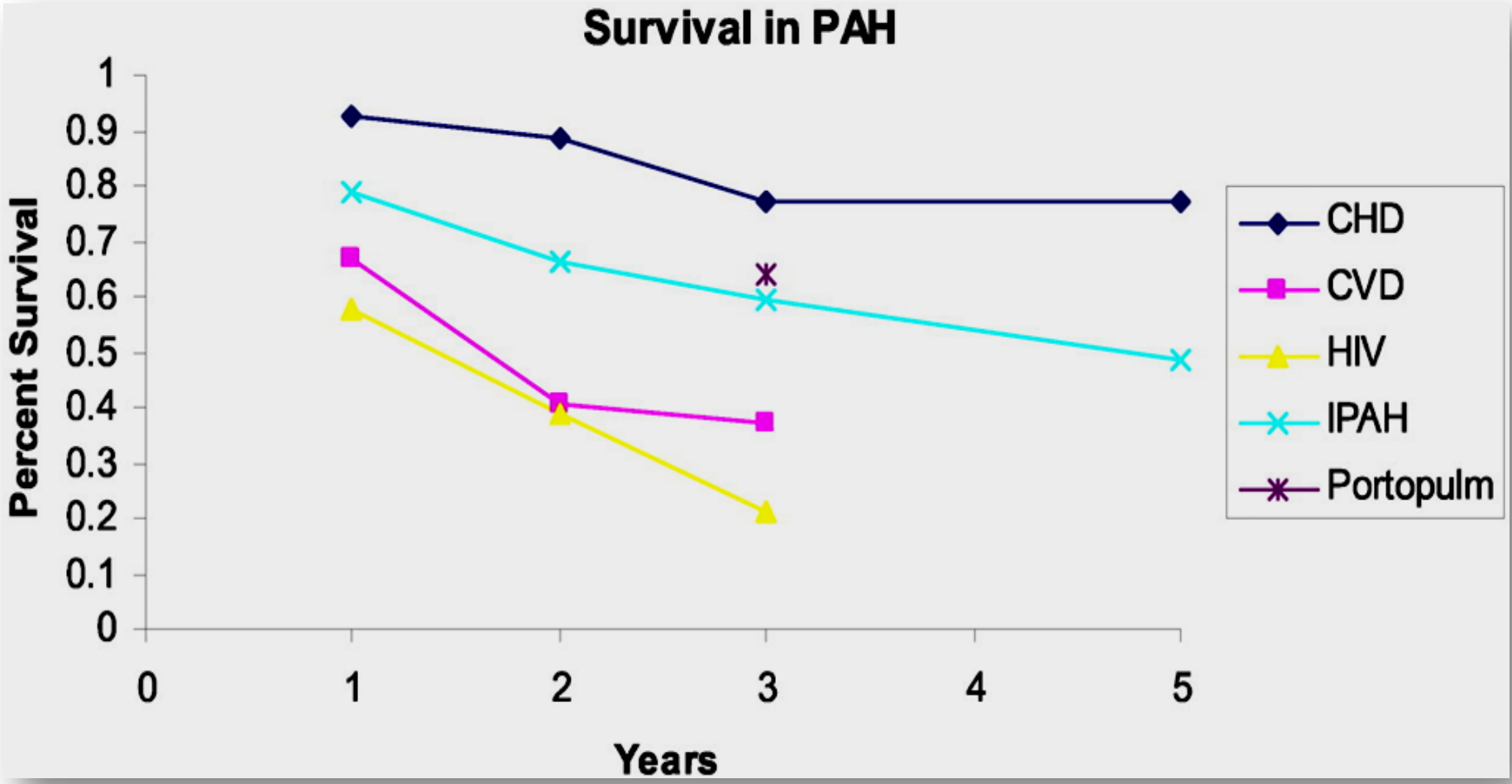


Figure 4 Kaplan-Meier Plots of Cumulative Survival in Adults With CHD According to PH Status Stratified by 2 Age Groups

For each PH case–non-PH control pair, the time zero corresponds to the date of the first PH diagnosis of the PH case. Abbreviations as in Figure 1.

A diagnosis of PH in adults with CHD is associated with a more than 2-fold higher risk for all-cause mortality and 3-fold higher rates of HSU, reflecting high morbidity. (J Am Coll Cardiol 2011;58:538–46) © 2011 by the



Mean survival of patients with PAH based on etiology

McLaughlin, V. V. et al. Chest 2004;126:78S-92S

Am J Cardiol. 2014 ;113(1):147-55.

Four- and Seven-Year Outcomes of Patients With Congenital Heart Disease-Associated Pulmonary Arterial Hypertension (from the REVEAL Registry).

Barst RJ[1], Ivy DD[2], Foreman AJ[3], McGoon MD[4], Rosenzweig EB[5].

For the overall CHD-associated PAH cohort, longer 6-minute walk distance, lower mean right atrial pressure, brain natriuretic peptide level <50 pg/ml, and the presence of acute vasoreactivity were predictors of survival at 4 years from enrollment; younger age and lower mean right atrial pressure were predictors of survival at 7 years from diagnosis. In conclusion, these observations **support predicted physiologic differences** (e.g., hemodynamics) between patients with IPAH or HPAH and patients with CHD-associated PAH, with or without a systemic-pulmonary shunt. **These differences, however, did not translate into significantly improved 4- and 7-year survival rates in patients with ES versus IPAH or HPAH and CHD-associated PAH.**

Hipertensão Pulmonar nas Cardiopatias Congénitas

Prevalência

	Adultos com CC	
		shunts **sistémico-pulmonares** ***shunts*** **cirúrgicos**
Euro Heart Survey (PsAP>40mmHg)	**5 -10%**	**HP - 28%** **SE - 12%** *Engelfriet PM, Heart 2007; 93:682-687*
Dutch CONCOR registry (PsAP>40mmHg)	**HP - 4,2%** **SE – 1,1%**	**HP – 6,1%** (SE -58%) **SE- 3,5%** **Pós encerramento – 3%** *Duffels MG, Int J Cardiol 2007;120:198- 204*
Estudo Canadiano	**5,8%**	*Lowe BS, JACC 2011; 58 (5): 538-546*

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A probabilidade de desenvolver HP/SE depende do tipo e dimensão do defeito

CIV	**11%**
DSAV	**41%**
CIA *ostium secundum*	**8%**
Janela aorto-pulmonar **Truncus**	**100%**

Incidência de acordo com o tipo de defeito*(> 2 A, não operado)*

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Duffels MG, Int J Cardiol 2007;120:198- 204

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- **Apesar da precocidade no diagnóstico e dos avanços na cirurgia a HP associada a CC continua a ser um problema importante e preocupante**

The frequency of Eisenmenger syndrome has declined from 8% in the 1950s to 4% among contemporary adult congenital heart patients under follow-up at tertiary centers in London, Toronto, and Zurich

Oechlin EN. AHJ 2000; 86:1111-1116

- Dados mais recentes (> 2002) do Royal Brompton Hospital (Londres) revelam ↑ do nº D com SE (±9%/ano)

HAP em situações mais complexas (Fontan, cardiopatias complexas, pós–cirurgia)

Marelli AJ Circulation 2007; 115(2):163-172
Mulder B Eur Respir Rev 2010;19(118):308-313

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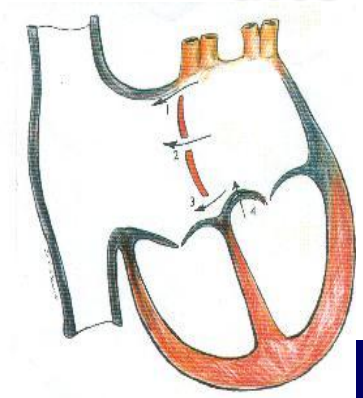
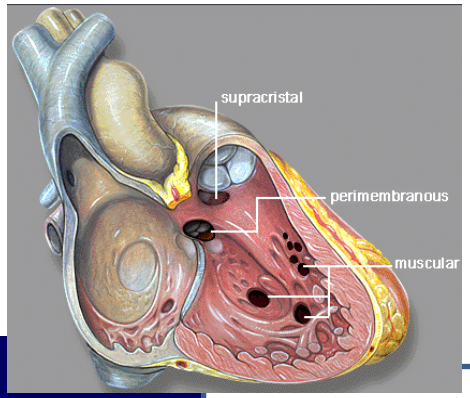
HAP nas CC - Fisiopatologia

Cardiopatias com *shunt* sistémico-pulmonar e alto débito pulmonar inicial

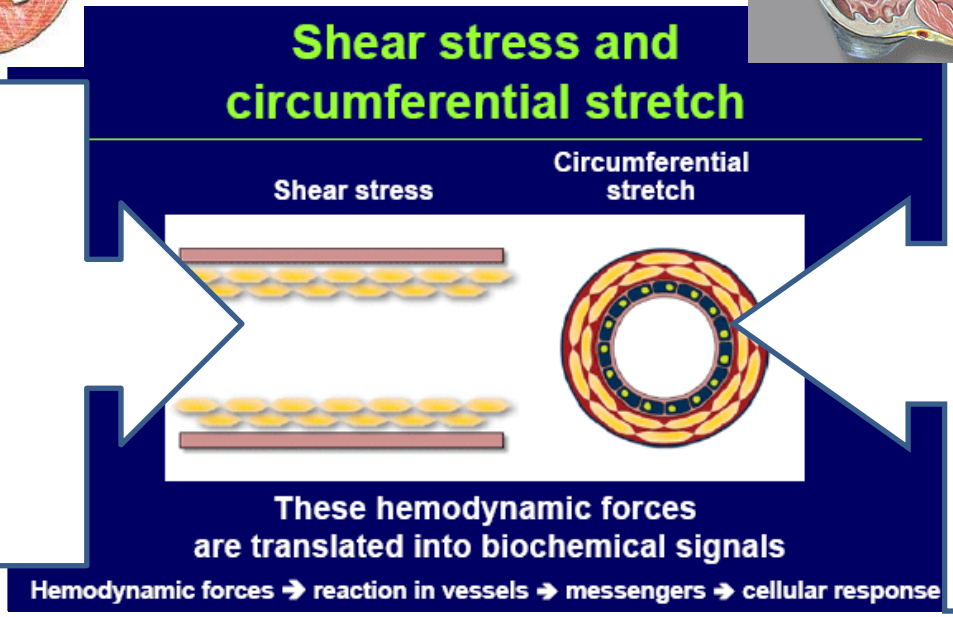




***Shunts* pré-tricúspides**

CIA, RVPA

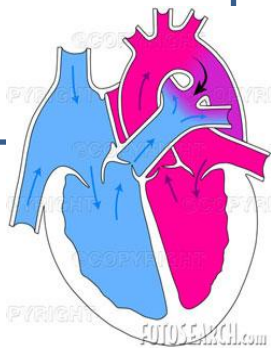
- ↑*volume*
- **HP tardia**
- ***PAP* ↑**



***Shunts* pós-tricúspides**

CIV, PCA, DCSAV, Truncus

- **↑pressão e volume**
- **HP precoce**
- **PAP ↑ ↑**



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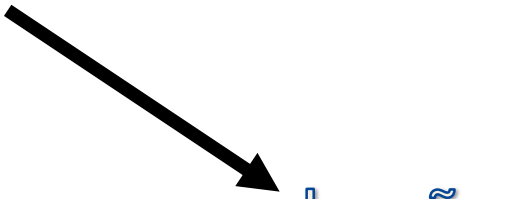
HIPERTENSÃO ARTERIAL PULMONAR
Fisiopatologia

Factores de risco:

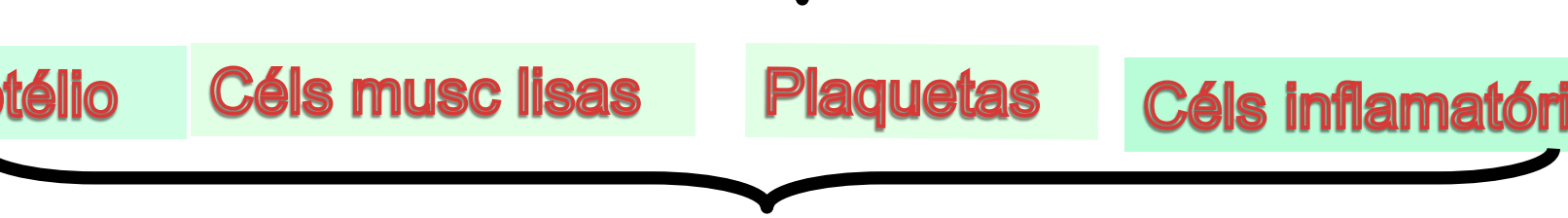
Anorexígenos, azeite tóxico
Conectivites, CC («shunts»),
HIV, HTPortal

Predisposição genética

Mutações-BMPR2, ALK1
Polimorfismos – 5HTT,
e-NOS, CPS



Lesão vascular



Endotélio | Céls musc lisas | Plaquetas | Céls inflamatórias

Vasoconstrição | **Proliferação** | **Trombose** | **Inflamação**

SINDROMA DE EISENMENGER

«... resulting from lesions that have an unrestrictive comunication with exposure of pulmonary bed to arterial pressure» **Paul Wood, BMJ 1958**

Left-to-right shunt

↓

Increased pulmonary blood flow
(shear stress/circumferential stretch)

↓

Endothelial dysfunction and vascular remodeling
Smooth muscle cell proliferation, increase in extracellular matrix, intravascular thrombosis

↓

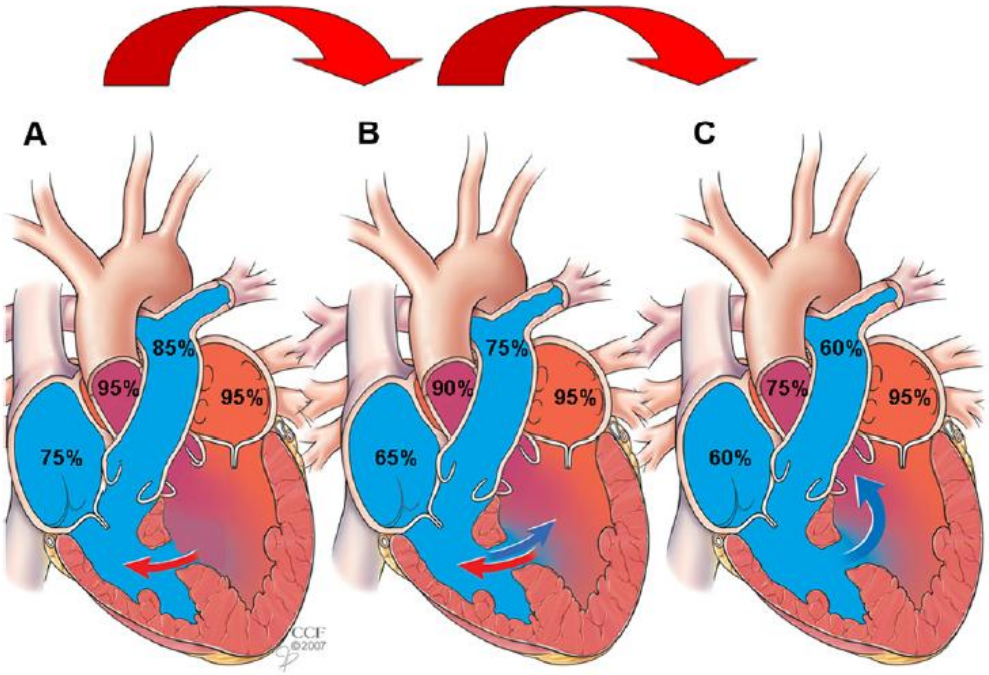
Increase in PVR

↓

Inverted shunt: right-to-left

↓

Cyanosis (Eisenmenger syndrome)



Vongpatanasin. Ann Inter Med 1998; 128:745-55

Formas de HAP nas CC

A - Sindroma de Eisenmenger

B – HAP ainda sem inversão do *shunt* (sem cianose)

C- HAP com defeitos pequenos (semelhante HAPi)

D- HAP após cirurgia

Clinical classification of congenital, systemic-to-pulmonary shunts associated with pulmonary arterial hypertension

A. Eisenmenger's syndrome

Eisenmenger's syndrome includes all systemic-to-pulmonary shunts due to large defects leading to a severe increase in PVR and resulting in a reversed (pulmonary-to-systemic) or bidirectional shunt. Cyanosis, erythrocytosis, and multiple organ involvement are present.

B. Pulmonary arterial hypertension associated with systemic-to-pulmonary shunts

In these patients with moderate to large defects, the increase in PVR is mild to moderate, systemic-to-pulmonary shunt is still largely present, and no cyanosis is present at rest.

C. Pulmonary arterial hypertension with small[a] defects

In cases with small defects (usually ventricular septal defects <1 cm and atrial septal defects <2 cm of effective diameter assessed by echocardiography) the clinical picture is very similar to idiopathic PAH.

D. Pulmonary arterial hypertension after corrective cardiac surgery

In these cases, congenital heart disease has been corrected but PAH is either still present immediately after surgery or has recurred several months or years after surgery in the absence of significant post-operative residual congenital lesions or defects that originate as a sequela to previous surgery.

[a]The size applies to adult patients.
PAH = pulmonary arterial hypertension; PVR = pulmonary vascular resistance.

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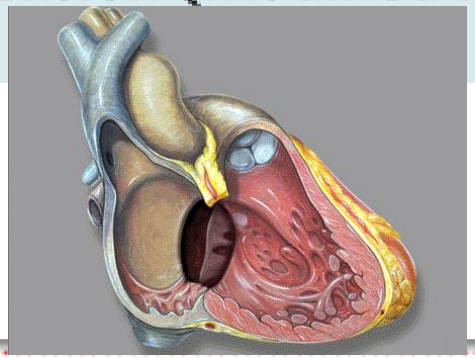
Anatomical-pathophysiological classification of congenital systemic-to-pulmonary shunts associated with pulmonary arterial hypertension (modified from Venice 2003)



Type

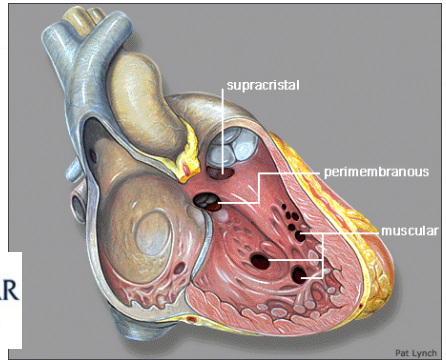
1.1 Simple pre-tricuspid shunts
- 1.1.1 Atrial septal defect (ASD)
  - 1.1.1.1 Ostium secundum
  - 1.1.1.2 Sinus venosus
  - 1.1.1.3 Ostium primum
- 1.1.2 Total or partial unobstructed anomalous pulmo venous return

1.2 Simple post-tricuspid shunts
- 1.2.1 Ventricular septal defect (VSD)
- 1.2.2 Patent ductus arteriosus

1.3 Combined shunts

Describe combination and define predominant defect

1.4 Complex congenital heart disease
- 1.4.1 Complete atrioventricular septal defect
- 1.4.2 Truncus arteriosus
- 1.4.3 Single ventricle physiology with unobstructed pulmonary blood flow
- 1.4.4 Transposition of the great arteries with VSD (without pulmonary stenosis) and/or patent du arteriosus
- 1.4.5 Other



Dimension (specify for each defect if more than one congenital heart defect exists)

2.1 Haemodynamic (specify Qp/Qs)[a]
- 2.1.1 Restrictive (pressure gradient across the defect)
- 2.1.2 Non-restrictive

2.2 Anatomic[b]
- 2.2.1 Small to moderate (ASD ≤2.0 cm and VSD ≤1.0 cm)
- 2.2.2 Large (ASD >2.0 cm and VSD >1.0 cm)

Direction of shunt
- 3.1 Predominantly systemic-to-pulmonary
- 3.2 Predominantly pulmonary-to-systemic
- 3.3 Bidirectional

Associated cardiac and extracardiac abnormalities

Repair status
- 5.1 Unoperated
- 5.2 Palliated [specify type of operation(s), age at surgery]
- 5.3 Repaired [specify type of operation(s), age at surgery]

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HAP ≠ Doença vascular pulmonar

SE

Shunt E-D	*Shunt* E-D	*Shunt* E-D predominante	*Shunt* D-E
PAP normal	↑ PAP HP hipercinética	↑ PAP	↑ ↑ PAP
RVP normal	RVP normal	↑ RVP	↑ ↑ RVP
Operabilidade			
Sim	*Sim*	?	*Não*

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HIPERTENSÃO ARTERIAL PULMONAR

DEFINIÇÃO HEMODINÂMICA

Dana Point 2008

PAPm ≥ 25mmHg

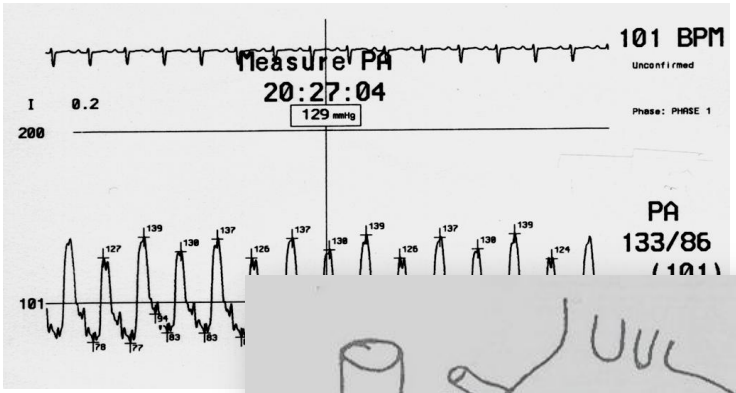
pressão capilar normal (<15 mmHg)

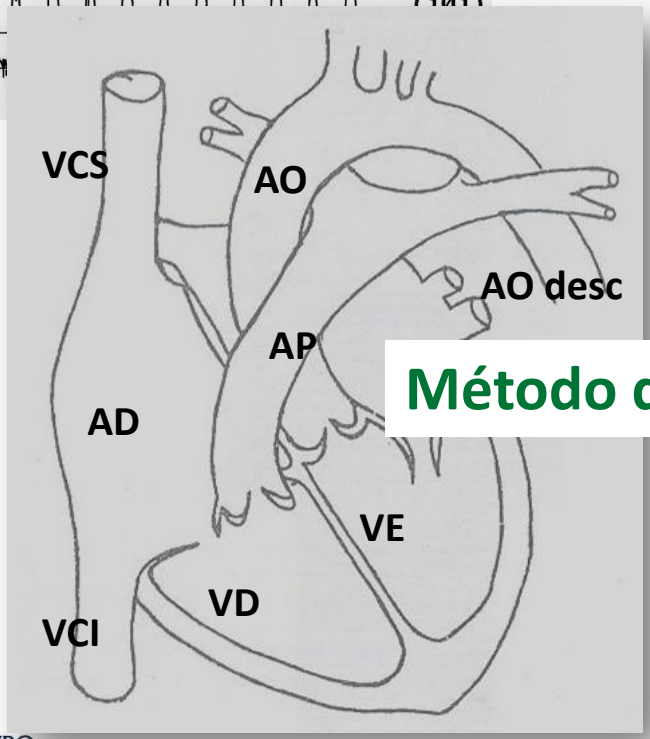
DC normal ou ↓

Resistência Vascular Pulmonar (RVP) > 3 UW

RVP= PAPm/QP

A determinação correcta das pressões, débitos e resistências pulmonares não dispensa o estudo hemodinâmico





Método de Fick

- ✓ PAPm
- ✓ Débito pulmonar (QP)
- ✓ Débito sistémico (QS)
- ✓ QP:QS
- ✓ Resistência vascular pulmonar (RVP/RVPi)
- ✓ RVP/RVS

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Guidelines for the diagnosis and treatment of pulmonary hypertension 2009

The Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT)

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric Cardiology (AEPC)

HAPi vs sindroma de Eisenmenger

	HTAPi	Sindroma de Eisenmenger
Resposta do VD * Dimensão * Função	Dilatação Deterioração rápida	Hipertrofia (lesões pós-tricuspide) Preservada – muito estável
Débito cardíaco	Reduzido	Mantido pelo shunt D-E
Prognóstico	Mau	Menos grave
Cianose	Raramente severa em repouso	Severa em repouso em dts estáveis Eritrocitose frequente
Complicações sistémicas	Pouco frequentes	Muito frequentes
Sínd cromossómicos associados	Não	Frequente (S. Down)
Percepção da limitação	Percepção adequada da limitação	Subestima limitação (longa evolução desde infância)
Coexistência de alt. estruturais cardíacas	Rara	Frequente (DCSAV, coração univentricular)

Dimopoulos K. Curr Opin Cardiol 2008; 23:545-554

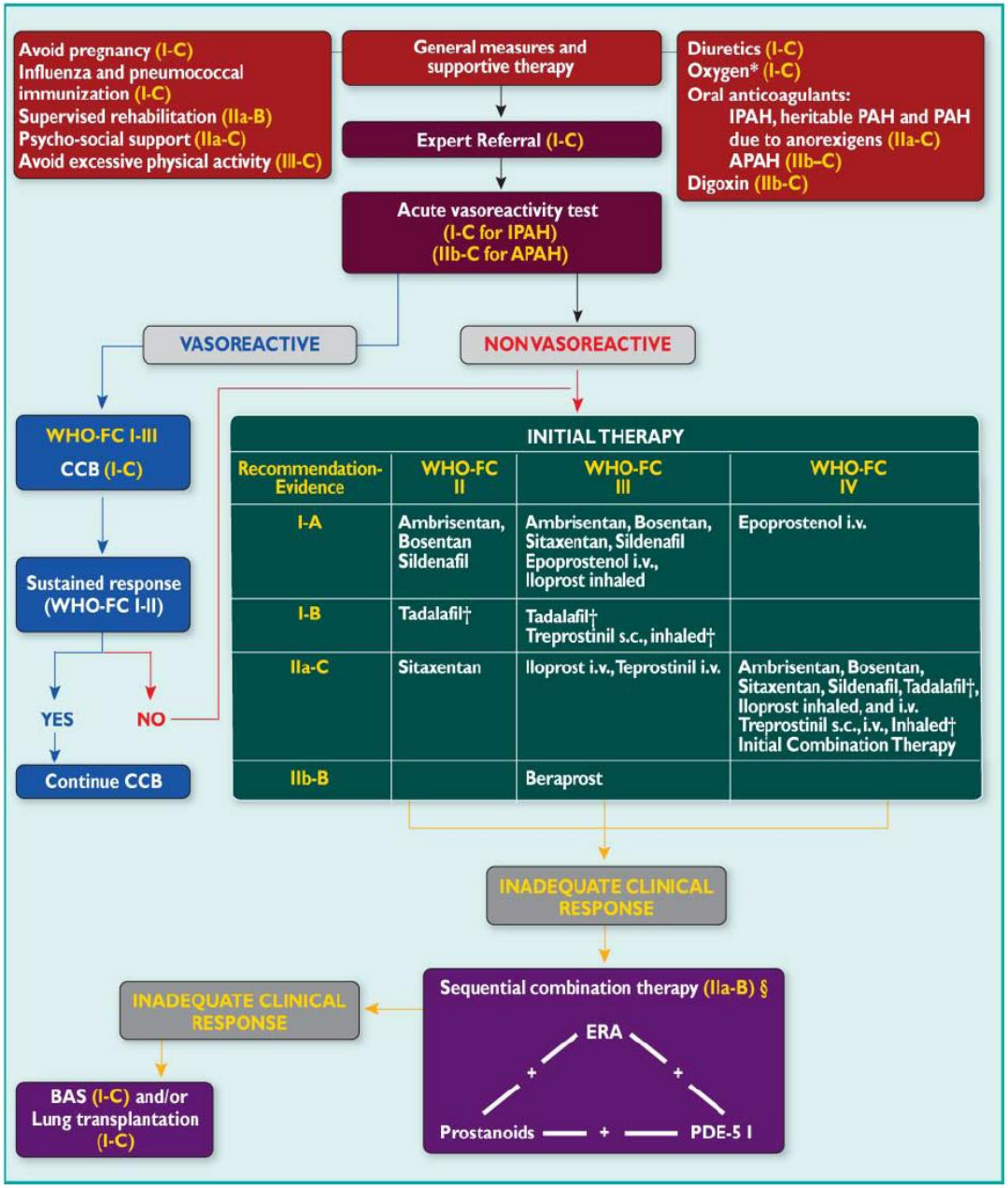

Figure 2 Evidence-based treatment algorithm for pulmonary arterial hypertension patients (for group 1 patients only). *To maintain arterial blood O_2 pressure ≥8 kPa (60 mmHg). †Under regulatory review in the European Union. §IIa-C for WHO-FC II. APAH = associated pulmonary arterial hypertension; BAS = balloon atrial septostomy; CCB = calcium channel blocker; ERA = endothelin receptor antagonist; IPAH = idiopathic pulmonary arterial hypertension; PDE5 I = phosphodiesterase type-5 inhibitor; WHO-FC = World Health Organization functional class.

General measures and supportive therapy

Avoid pregnancy (I-C)
Influenza and pneumococcal immunization (I-C)
Supervised rehabilitation (IIa-B)
Psycho-social support (IIa-C)
Avoid excessive physical activity (III-C)

- Gravidez contraindicada (30-50% mortalidade materna)
- Evitar: desidratação, hipovolémia
- Cuidados especiais com cirurgia não cardíaca e manejo anestésico
- Tentar manter/restaurar RS

Eritrocitose secundária

- **Flebotomia por rotina está contraindicada**
- **Indicada se sintomas de hiperviscosidade *e* hematócrito ≥ 65%**
- **Excluir/tratar deficiência de ferro**

Replacement therapy for iron deficiency improves exercise capacity and quality of life in patients with cyanotic congenital heart disease and/or the Eisenmenger syndrome

Edgar L.W. Tay [a,*], Ana Peset [a], Maria Papaphylactou [a], Ryo Inuzuka [a], Rafael Alonso-Gonzalez [a], Georgios Giannakoulas [a], Aphrodite Tzifa [a], Sara Goletto [a], Craig Broberg [a], Konstantinos Dimopoulos [a,b], Michael A. Gatzoulis [a,b]

- **Reposição isovolémica**

If symptoms of hyperviscosity are present, phlebotomy with isovolumic replacement should be considered usually when the haematocrit is > 65%

IIa C

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Prophylactic measures are the mainstay of care to avoid complications. The following exposures/activities should be avoided:
• Pregnancy
• Iron deficiency and anaemia (no routine, inappropriate phlebotomies to maintain a pre-determined haemoglobin)
• Dehydration
• Infectious disease: annual influenza vaccination, pneumovax

- **Aconselhamento adequado e medidas gerais são importantes para reduzir a morbilidade e mortalidade**

Other risk reduction strategies include:
• Use of an air filter in an intravenous line to prevent air embolism
• Consultation of a GUCH cardiologist before administration of any agent and performance of any surgical/interventional procedure
• Prompt therapy of upper respiratory tract infections
• Cautious use or avoidance of agents that impair renal function
• Contraceptive advice

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O2

General measures and supportive therapy

- Diuretics (I-C)
- Oxygen* (I-C)
- Oral anticoagulants:
  - IPAH, heritable PAH and PAH due to anorexigens (IIa-C)
  - APAH (IIb–C)
- Digoxin (IIb-C)

- **Pode ser útil**

  Bowyer, BHJ 1986;55:385-390

- **Ausência de benefício de O2 nocturno (sem ↓PA ou ↑sobrevida)**

  Sandoval, AJRCC 2001; 164:1682- 1687

The use of supplemental O_2 therapy should be considered in cases in which it produces a consistent increase in arterial oxygen saturation and reduces symptoms	IIa	C

Anticoagulação

- Controversa
- Alteração hemostase

In the absence of significant haemoptysis, oral anticoagulant treatment should be considered in patients with PA thrombosis or signs of heart failure	IIa	C

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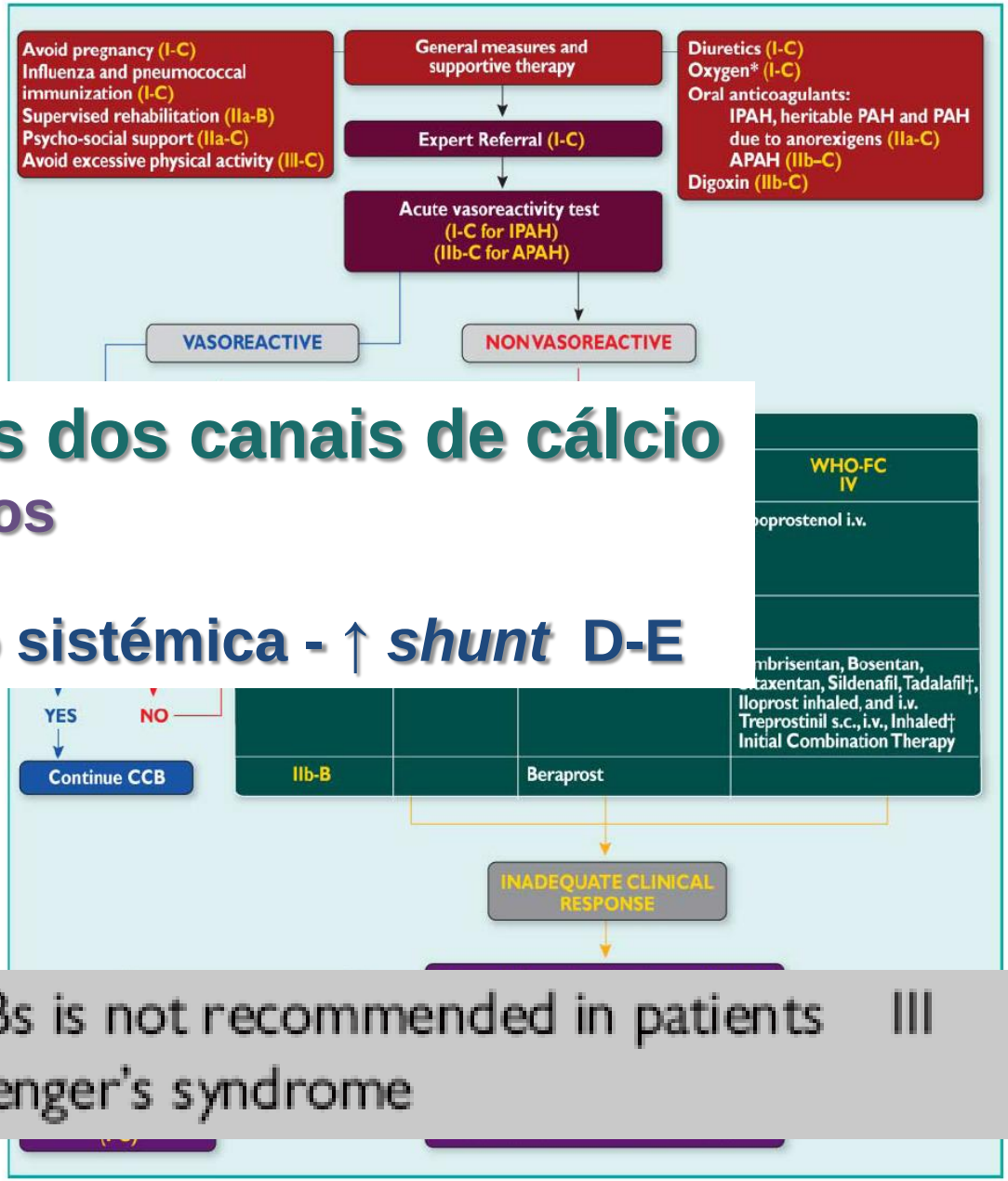
Antagonistas dos canais de cálcio

Contraindicados

-vasodilatação sistémica - ↑ *shunt* D-E



The use of CCBs is not recommended in patients with Eisenmenger's syndrome — III — C

Figure 2 Evidence-based treatment algorithm for pulmonary arterial hypertension patients (for group 1 patients only). *To maintain arterial blood O_2 pressure ≥8 kPa (60 mmHg). †Under regulatory review in the European Union. §IIa-C for WHO-FC II. APAH = associated pulmonary arterial hypertension; BAS = balloon atrial septostomy; CCB = calcium channel blocker; ERA = endothelin receptor antagonist; IPAH = idiopathic pulmonary arterial hypertension; PDE5 I = phosphodiesterase type-5 inhibitor; WHO-FC = World Health Organization functional class.

Guidelines for the diagnosis and treatment of pulmonary hypertension

No SE a terapêutica específica está indicada na classe ≥ III NYHA

Table 25 Recommendations for PAH associated with congenital cardiac shunts

Statement	Class[a]	Level[b]
The ERA bosentan is indicated in WHO-FC III patients with Eisenmenger's syndrome	I	B
Other ERAs, phosphodiesterase type-5 inhibitors, and prostanoids should be considered in patients with Eisenmenger's syndrome	IIa	C
In the absence of significant haemoptysis, oral anticoagulant treatment should be considered in patients with PA thrombosis or signs of heart failure	IIa	C
The use of supplemental O_2 therapy should be considered in cases in which it produces a consistent increase in arterial oxygen saturation and reduces symptoms	IIa	C
If symptoms of hyperviscosity are present, phlebotomy with isovolumic replacement should be considered usually when the haematocrit is > 65%	IIa	C
Combination therapy may be considered in patients with Eisenmenger's syndrome	IIb	C
The use of CCBs is not recommended in patients with Eisenmenger's syndrome	III	C

European Heart Journal
doi:10.1093/eurheartj/ehp297

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Recommendations	Class[a]	Level[b]
Targeted PAH therapy in CHD should only be performed in specialized centres	I	C
The ERA bosentan should be initiated in WHO-FC III[c] patients with Eisenmenger syndrome	I	B
Other ERAs, phosphodiesterase type-5 inhibitors, and prostanoids should be considered in WHO-FC III[c] patients with Eisenmenger syndrome	IIa	C
Combination therapy may be considered in WHO-FC III[c] patients with Eisenmenger syndrome	IIb	C
The use of calcium channel blockers should be avoided in patients with Eisenmenger syndrome	III	C

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Guidelines for the diagnosis and treatment of pulmonary hypertension

The treatment strategy for patients with PAH associated with CHD and in particular for subjects with Eisenmenger's syndrome is mainly based on clinical experience of experts rather than formally evidence based

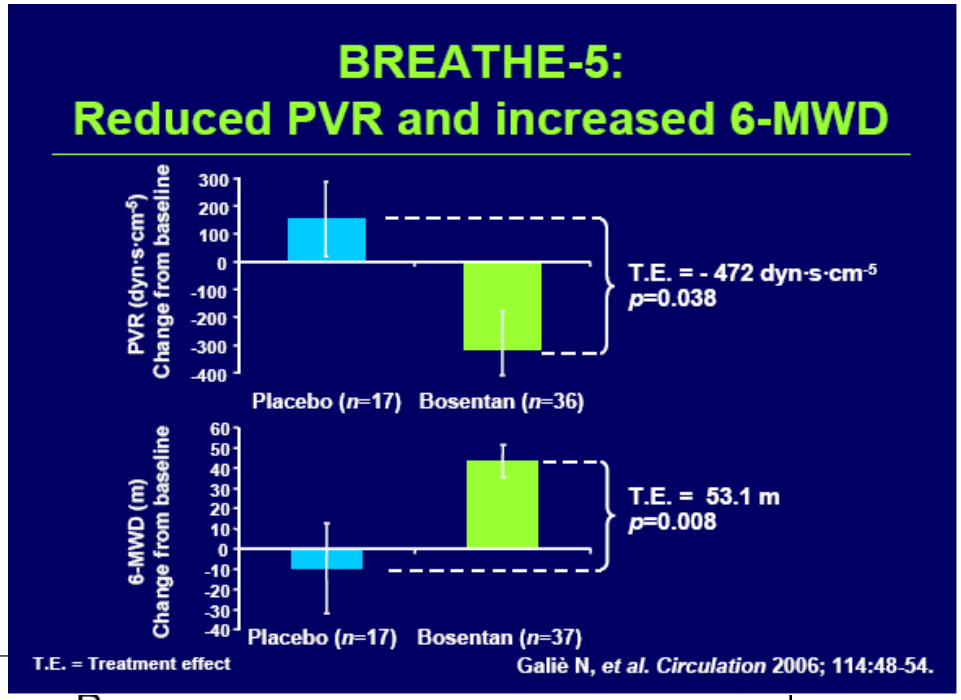
EUROPEAN SOCIETY OF CARDIOLOGY®

European Heart Journal
doi:10.1093/eurheartj/ehp297

Terapêutica - resultados

- Melhoria clínica (FC, 6MWT)
- Melhoria hemodinâmica
- ↓ PVR
- *Sem baixar a saturação O2*

Breath 5 Galiè. Circulation 2006;114:48-54



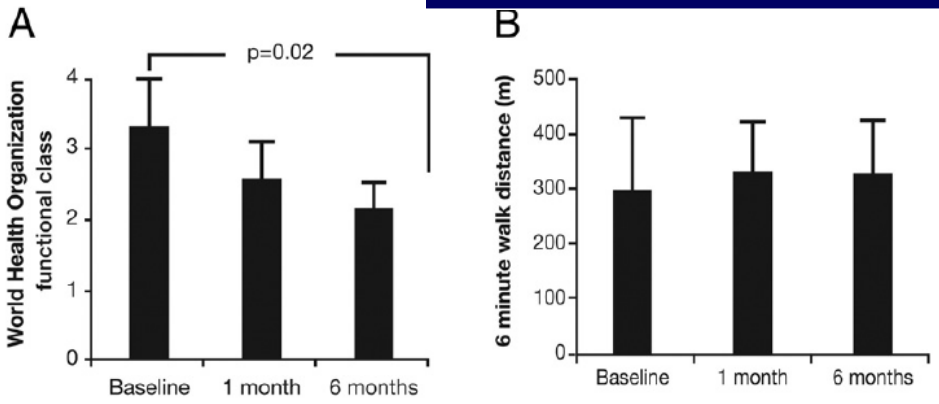


Figure 5 Effects of Sildenafil on World Health Organization Functional Class and 6-Min Walk Distance in Patients With ES

Improvements in World Health Organization functional class and 6-min walk distance after treatment with sildenafil in patients with Eisenmenger syndrome (ES). Reproduced, with permission, from Chau et al. (41).

Terapêutica - resultados

- ↑ Tempo até agravamento clínico

Pulmonary arterial hypertension associated with a congenital heart defect: advanced medium-term medical treatment stabilizes clinical condition.
Duffels M, van Loon L, Berger R, Boonstra A, Vonk-Noordegraaf A, Mulder B.
Congenit Heart Dis. 2007; 2(4):242-9

Advanced therapy may delay the need for transplantation in patients with the Eisenmenger syndrome

Tom Adriaenssens[1], Marion Delcroix[2], Kristien Van Deyk[1], and Werner Budts[1]*

European Heart Journal (2006) 27, 1472–1477

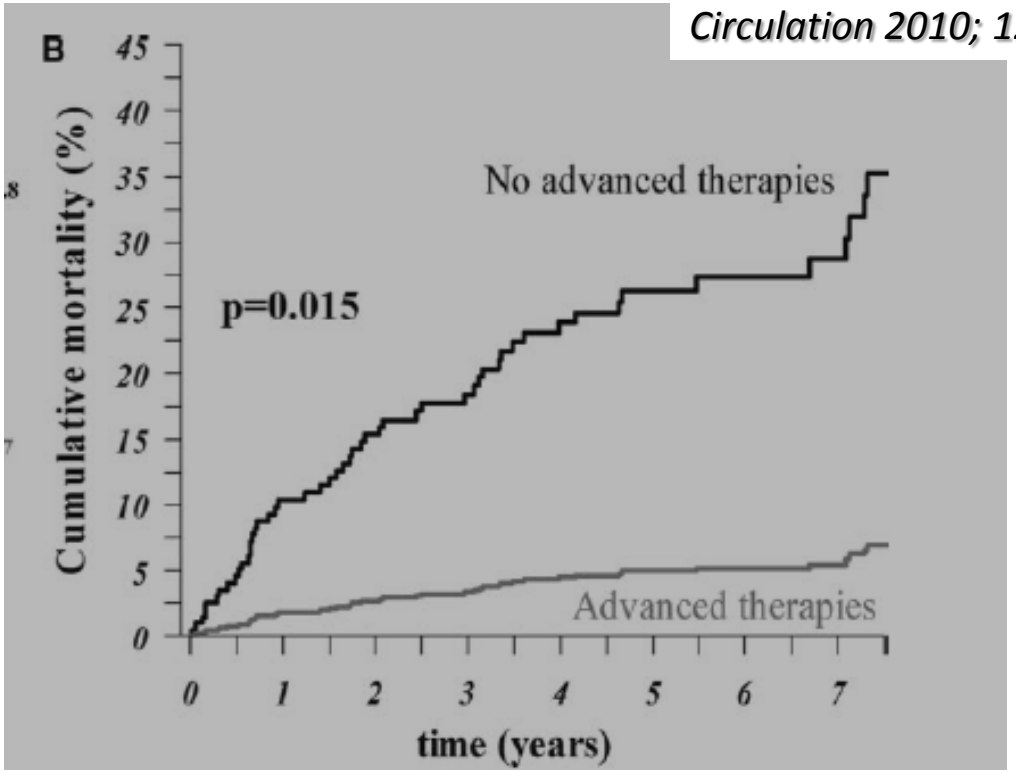
Terapêutica - resultados

- Melhoria na sobrevida

Improved Survival Among Patients With Eisenmenger Syndrome Receiving Advanced Therapy for Pulmonary Arterial Hypertension

Konstantinos Dimopoulos, Ryo Inuzuka, Sara Goletto, Georgios Giannakoulas, Lorna Swan, Stephen J. Wort and Michael A. Gatzoulis

Circulation 2010; 121:20-25



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Terapêutica

- **Resultados a longo prazo**

Disease targeting therapies in patients with Eisenmenger syndrome: Response to treatment and long-term efficiency ☆

Gerhard-Paul Diller [a,b,1], Rafael Alonso-Gonzalez [a,1], Konstantinos Dimopoulos [a,b], Maria Alvarez-Barredo [a], Chiehyang Koo [a], Aleksander Kempny [a], Carl Harries [a], Lisa Parfitt [a], Anselm S. Uebing [a], Lorna Swan [a], Philip S. Marino [a,b], Stephen J. Wort [a,b], Michael A. Gatzoulis [a,b,*]

Int J Cardiol 2012

Prolonged beneficial effect of bosentan treatment and 4-year survival rates in adult patients with pulmonary arterial hypertension associated with congenital heart disease

Jeroen C. Vis[a, b], Marielle G. Duffels[a], Pepijn Mulder[a], Rianne H.A.C.M. de Bruin-Bon[a], Berto J. Bouma[a], Rolf M.F. Berger[c], Elke S. Hoendermis[c], Arie P.J. van Dijk[d], Barbara J.M. Mulder[a, b],

Int J Cardiol 2011

…the mortality rate of 20% of patients after 4 years of follow-up remains high

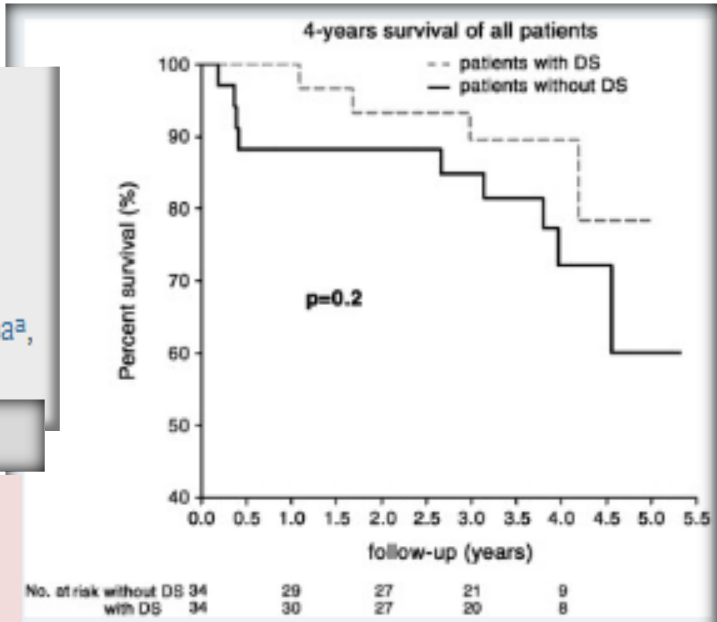

Fig. 1. 4-years survival of all patients. DS; Down syndrome.

Treat-and-repair??

Com as novas terapêuticas será possível tornar um defeito «inoperável» numa situação com solução cirúrgica ??

Evaluating operability in adults with congenital heart disease and the role of pretreatment with targeted pulmonary arterial hypertension therapy

Konstantinos Dimopoulos *, Ana Peset, Michael A. Gatzoulis

Int J Cardiol 2008; 129:163-171

Can "Inoperable" Congenital Heart Defects Become Operable in Patients with Pulmonary Arterial Hypertension? Dream or Reality?

Beghetti M. Cong Heart Dis. 2012

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Treat-and-repair??

A FAVOR	CONTRA
▪Anular shunt D-E ↓ eventos cerebrovasculares ↓ abcessos cerebrais	▪ Reduções «modestas» da RVP
Evitar a hipoxémia /cianose ↑ Tolerancia ao esforço	▪ Risco cirúrgico elevado
▪Prevenir as complicações da eritrocitose ↓ alterações hemostáticas ↓ outras complicações sistémicas	▪ Converter a fisiologia de Eisenmenger numa forma de HAP idiopática (pior prognóstico)
▪Proteger a circulação pulmonar	▪ Experiência limitada: ▪ só os casos bem sucedidos são relatados ▪ sem dados a longo prazo

Treat-and-repair??

A FAVOR	CONTRA
▪A ↓ ↓	P
Evi ↑	
▪P eri ↓ alterações hemostáticas ↓ outras complicações sistémicas	HAP idiopática (pior prognóstico)
▪Proteger a circulação pulmonar	▪ Experiência limitada: ▪ só os casos bem sucedidos são relatados ▪ sem dados a longo prazo

- eventualmente em casos *border-line*, cuidadosamente seleccionados
- no sindroma [illegible]enmenger

Conclusões

- Em Portugal a HAP associada a CC é um dos principais tipos de HAP
- A HAP associada a CC é uma situação com gravidade clínica e que influencia negativamente o prognóstico
- Semelhanças com outras formas de HAP
- Aspectos específicos (diagnóstico, clínica, prognóstico e tratamento)
- A existência de Hipertensão Pulmonar associada a CC (*shunts*) implica a sua correcta caracterização e avaliação hemodinâmica

Conclusões

- As medidas gerais e o aconselhamento adequado continuam a ter importância na redução da morbilidade e mortalidade
- Os fármacos específicos para HAP abriram novas perspectivas, com melhoria sintomática, funcional e da sobrevida
- Não existe cura para a HAP

- Redução da incidência ← correcção precoce da CC

- Formas de HAP em cardiopatias mais complexas

Long-term effect of bosentan in pulmonary hypertension associated with complex congenital heart disease

Rui Baptista[a,b,*], Graça Castro[a], António Marinho da Silva[a], Pedro Monteiro[a,b], Luís Augusto Providência[a,b]

Rev Port Cardiol. 2013;32(2):123–129

Conclusions: Bosentan was safe and was associated with improved exercise capacity in patients with PAH and complex CHD. This improvement was sustained for up to four years and the safety profile was similar to simple CHD patients.

Teste de vasoreactividade

HAP i

- Orientação terapêutica (antag Ca)
- Prognóstico

- ↓PmAP ≥10mmHg e PmAP ≤40mmHg
- DC igual ou ↑

CC – shunts

Operabilidade?

RVPi < 6U UW/m2

RVP/RS < 0.3

Encerramento do *shunt*

Teste não indicado

RVPi 6-9UW/m2 ↓20% e <6

RVP/RS 0.3 a 0.5 ↓20% e <0.3

Teste

Potenciais candidatos a intervenção

Decisão individual

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Lopes and O´Leary, Cardiol Young. 2009; 19:431-45

SINDROMA DE EISENMENGER

Melhor prognóstico - porquê ?

- Manutenção da função VD
- Manutenção da HVD fetal
- Melhor enchimento VE (shunt D-E)
- Menor compressão VE pelo VD

Severe Pulmonary Hypertension Without Right Ventricular Failure: The Unique Hearts of Patients With Eisenmenger Syndrome

William E. Hopkins, MD, and Alan D. Waggoner, MHS, RDCS

(Am J Cardiol 2002;89:34–38)

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Survival in different subgroups of pts with PAD-CHD

	PAH-CHD ES	PAH -CHD shunt S-P	PAH-CHD small defects	PAH – CHD corrective surgery
1 y	96%	100%	80%	98%
5 y	88%	95%	80%	82%
10 y	85%	88%	80%	71%
P vs PAH – CHD corrective surgery	0,02	0,002	0,57	

Previous cardiac corrective surgery may represent a risk factor in pts with PAH-CHD

Galiè, Am J Respir Crit Care Med 2011