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CONGRESSO NACIONAL
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25 — 27 OUT 2017
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Alfândega do Porto



Coarctação da Aorta

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Jorge Moreira

Sem conflitos de interesse

Coarctação da Aorta

- 5-8% das Cardiopatias Congénitas
- Prevalência - 3-4:10.000/ nados-vivos
- 25-35 novos casos/ano em Portugal
- Maior incidência no sexo masculino (1,5:1)
- Associação a certos síndromes:
 - S. Turner (35%), S. Noonan, S. DiGeorge, S. VCF

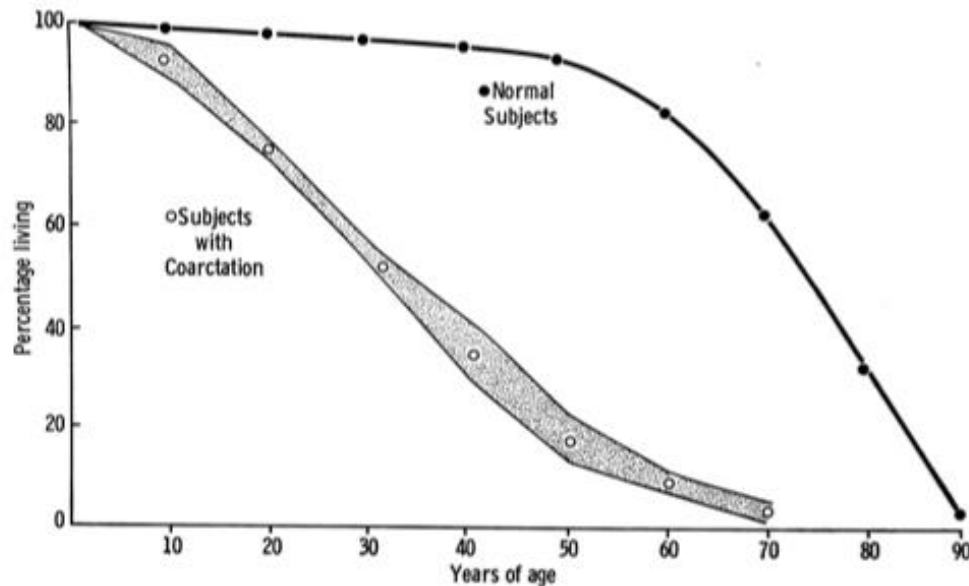
Coarctação da Aorta

British Heart Journal, 1970, 32, 633-640.

Natural history of coarctation of the aorta

Maurice Campbell

From the Cardiac Department, Guy's Hospital, London S.E.1, and the Institute of Cardiology, London W.1



- Prognóstico mau
- Sobrevida mediana 31A
- 75% óbitos < 46A
- Causas de mortalidade:
 - ICC (26%)
 - ruptura aórtica (21%)
 - endocardite (18%)
 - hemorragia intracraniana (12%)

Coarctação da Aorta



1760 Descrição
(Morgagni)



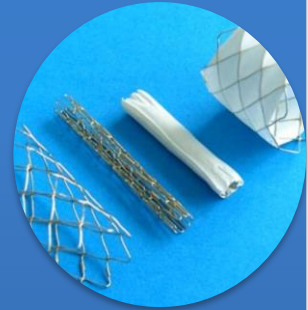
1944
Cirurgia
(Craaford
&Nylon)



1970s
Estabilização
PGE1



1980s
Angioplastia
balão

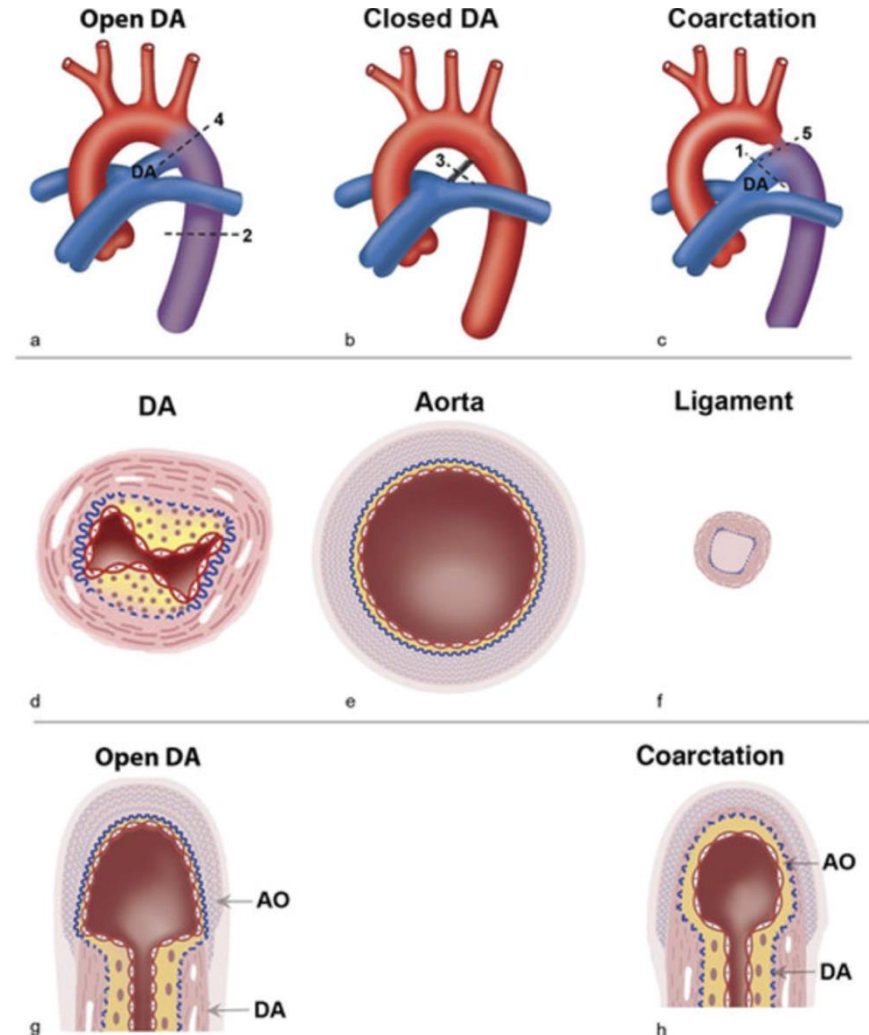


1990s
Stents
intravasculares



Coarctação da Aorta

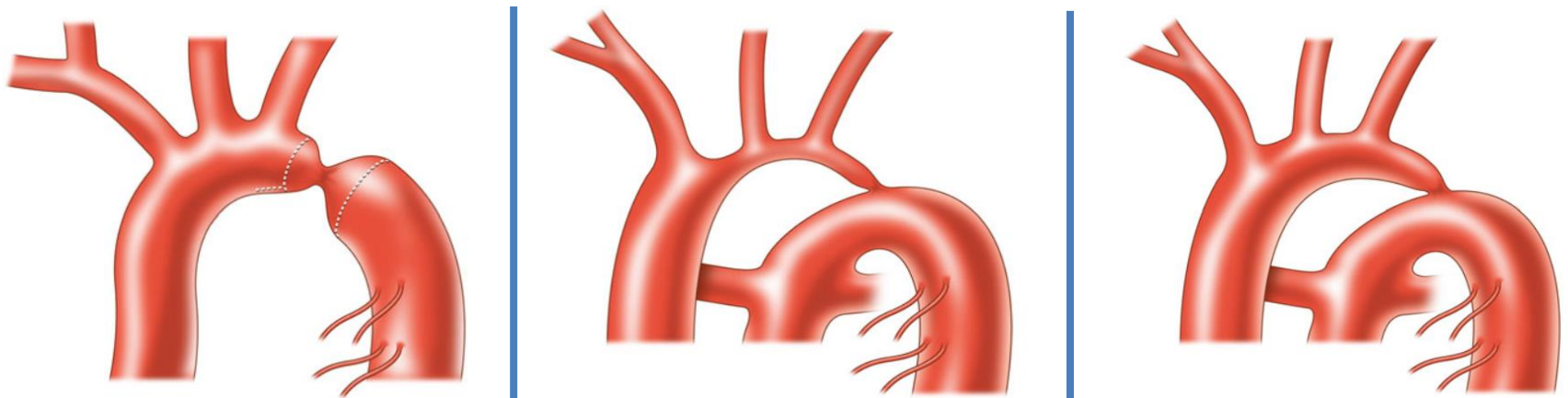
- Estreitamento do lumen aórtico no Istmo
 - Camada média espessada
 - Hiperplasia da íntima
- Arteriopatia generalizada
 - Necrose cística da média
- Teorias fisiopatológicas
 - Hemodinâmica
 - Tecido ductal ectópico



Coarctação da Aorta

Espectro morfológico

- Ampla diversidade de lesões
 - Forma isolada/ hipoplasia major do arco/ hipoplasia do istmo
- Critérios de definição da hipoplasia
 - **Moulaert *et al*** (regra 60-50-40); **T. Karl** (Peso + 1 mm); **Z-score** (<-2)



Coarctação da Aorta

Anomalias associadas

- Válvula aórtica bicúspide → 50-75%
- PDA (período neonatal)
- CIV → 40-50%
- Anomalias da válvula mitral, regiões sub e supra valvulares
- Aneurismas cerebrais → 2,5-10%
- Outras cardiopatias complexas

Coarctação da Aorta

Padrões clínicos

Table 1 Recognising CoA in patients of all ages

Age	Symptoms
Foetal	Ventricular disproportion (RV>LV) Great vessel disproportion Continuous diastolic flow at isthmus level Associated congenital heart disease (BAV, arch hypoplasia, subaortic stenosis, mitral valve abnormalities, VSD, ASD, PDA) Syndromic phenotypes (Turner, PHACE, DiGeorge, Noonan, velocardiofacial)
Neonatal and infantile	→ Collapse, acidosis → Heart failure → Systolic/ continuous murmur → Weak or absent femoral pulses, radio-femoral pulse delay Supine arm-leg blood pressure gradient Necrotizing enterocolitis Renal failure Cardiomyopathy (rare)

BAV, bicuspid aortic valve; LV, left ventricle; RV, right ventricle; VSD, ventricular septal defect; ASD, atrial septal defect; PDA, patent ductus arteriosus.

Coarctação da Aorta

Padrões clínicos

Table 1 Recognising CoA in patients of all ages

Age	Symptoms
Childhood, adolescent, adult	- Systolic/ continuous murmur → Weak or absent femoral pulses, radio-femoral delay - Supine arm-leg blood pressure gradient → Hypertension (headache, epistaxis, retinopathy) - Reduced exercise capacity, exercise induced hypertension - Leg fatigue and claudication - Cold feet - Left ventricular hypertrophy, arrhythmia and heart failure - Infective endocarditis - Aortic dissection, rupture - Intracranial haemorrhage

BAV, bicuspid aortic valve; LV, left ventricle; RV, right ventricle; VSD, ventricular septal defect; ASD, atrial septal defect; PDA, patent ductus arteriosus.

Coarctação da Aorta

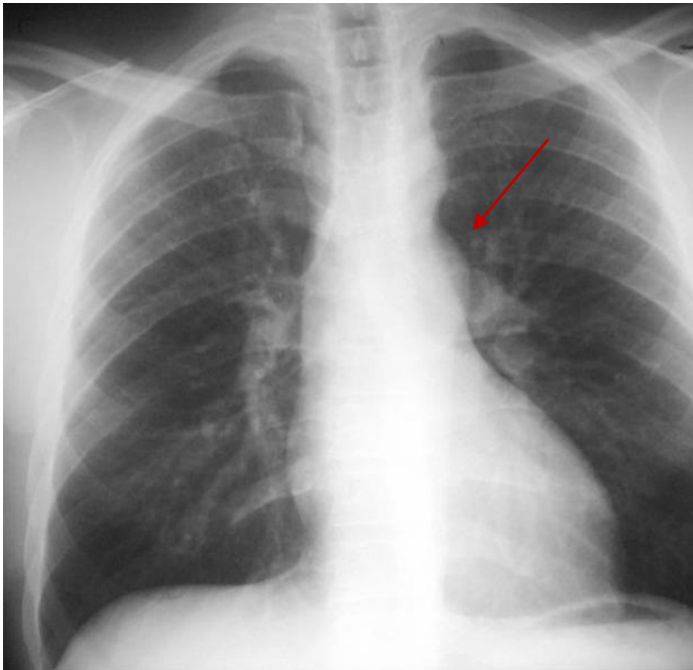
Diagnóstico

- Aspectos clínicos
- **Ecocardiografia TT**
- Imagiologia avançada (RMC e TC) → adolescentes e adultos
- Cateterismo cardíaco
 - Realizado, habitualmente, com intenção terapêutica

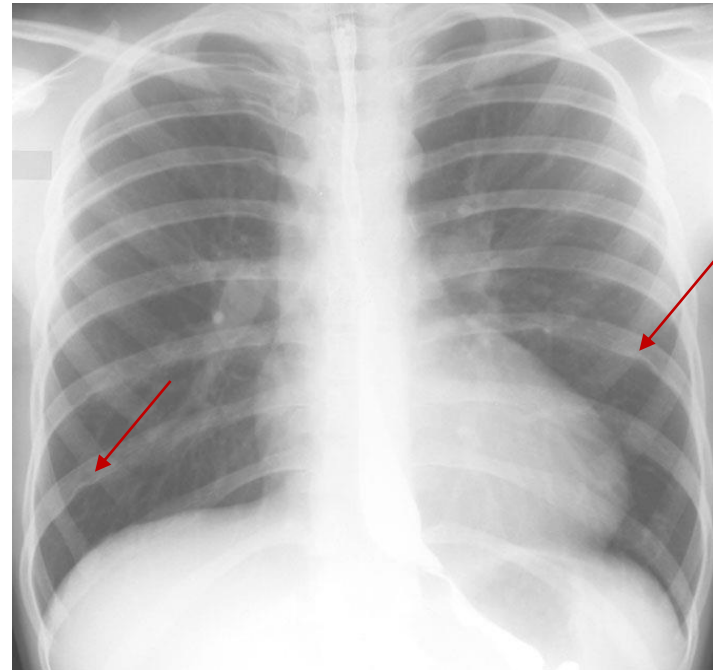
Coarctação da Aorta

Diagnóstico

- Radiografia Torácica



Sinal do “3”

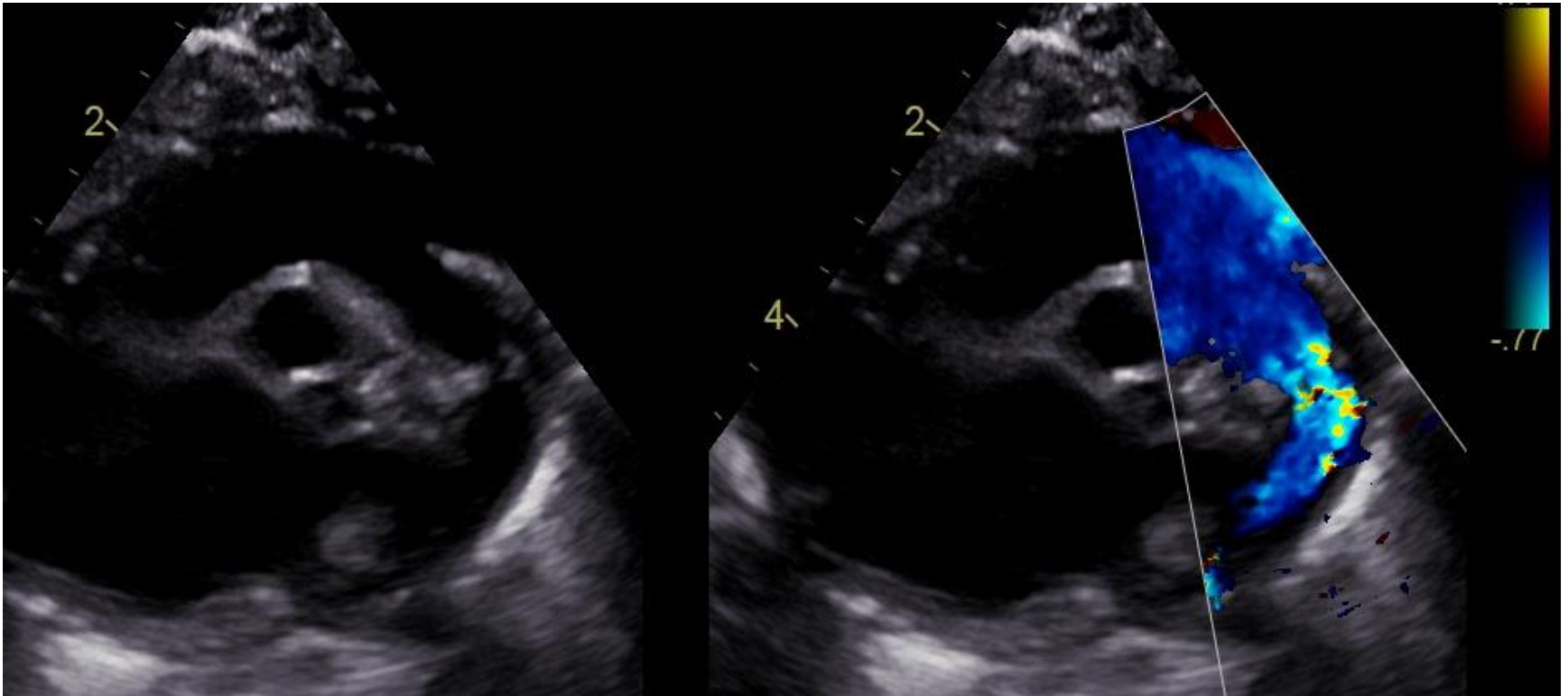


Erosão das costelas

Coarctação da Aorta

Diagnóstico

- **Ecocardiograma** – meio de diagnóstico mais útil

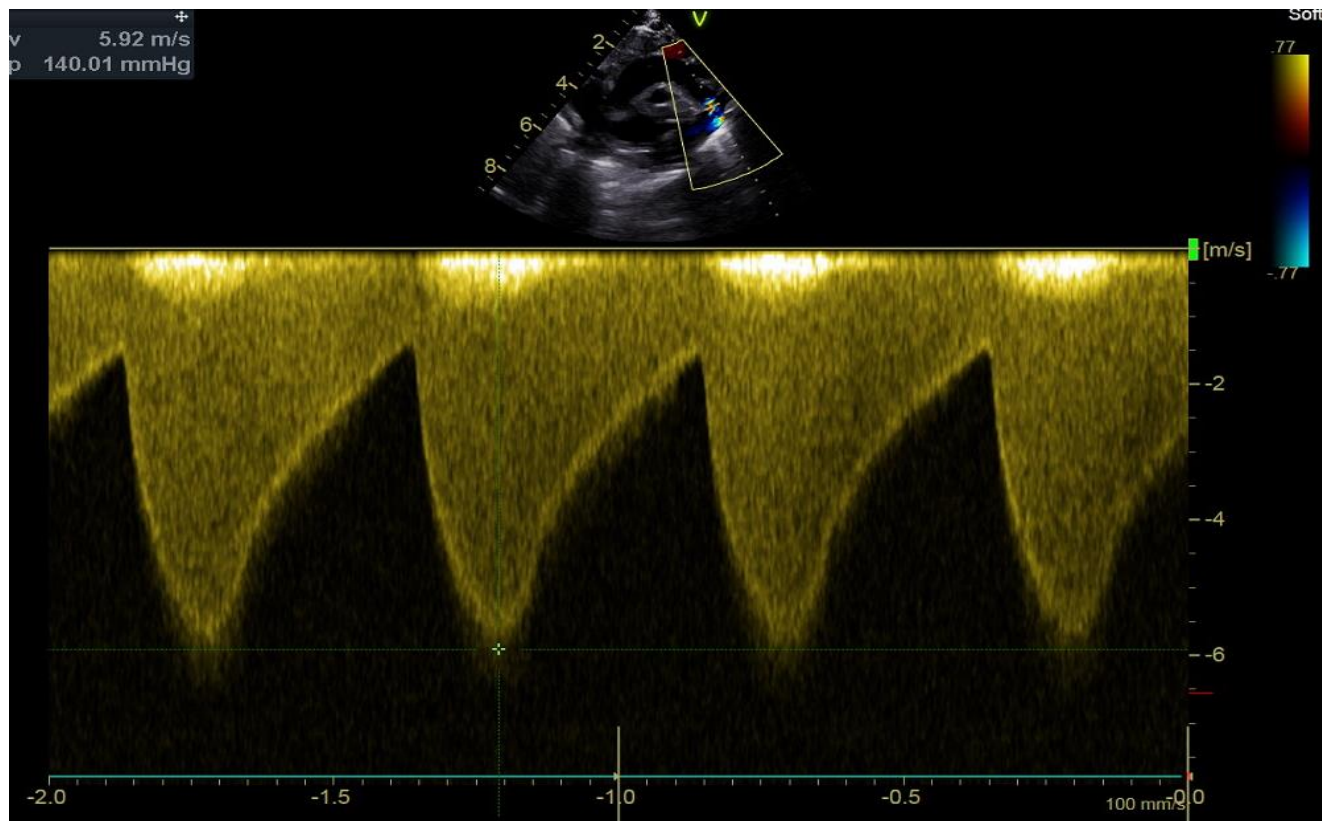


TL, 6meses

Coarctação da Aorta

Diagnóstico

- **Ecocardiograma** – prolongamento diastólico do fluxo na Ao D



TL, 6meses

Coarctação da Aorta

Diagnóstico

- PF, 12 anos (52 Kg) – **CoAo nativa**



Angio TC



Reconstrução 3D

Coarctação da Aorta

Diagnóstico

- RN FARN, 21 dias (3,2Kg) – Suspeita pré-natal de CoAo



Angio TC

Tronco bovino +
hipoplasia do AAo



Reconstrução 3D

Coarctação da Aorta

Diagnóstico

- VHM, 17 anos → Coarctação Aorta Nativa + Circulação colateral abundante



Angio-RMC

Coarctação da Aorta

Tratamento – Indicações

Box 1 Indication for treatment of native coarctation of the aorta (CoA) and re-CoA

Indications for treatment:

- ▶ Supine non-invasive pressure gradient >20 mm Hg between upper and lower limbs
- ▶ Peak-to-peak coarctation gradient ≥ 20 mm Hg
- ▶ Peak to peak coarctation gradient <20 mm Hg with radiological evidence of significant coarctation with significant collateral flow
- ▶ Pathological blood pressure response during exercise
- ▶ Significant left ventricular hypertrophy
- ▶ Hypertension with $\geq 50\%$ aortic narrowing relative to the aortic diameter at the level of the diaphragm
- ▶ Upper limb hypertension

Coarctação da Aorta

Tratamento – Indicações

Indications	Class ^a	Level ^b
All patients with a non-invasive pressure difference >20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (>140/90 mmHg in adults), pathological blood pressure response during exercise, or significant LVH should have intervention	I	C
Independent of the pressure gradient, hypertensive patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) should be considered for intervention	IIa	C
Independent of the pressure gradient and presence of hypertension, patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) may be considered for intervention	IIb	C

Coarctação da Aorta

Comparison of Surgical, Stent, and Balloon Angioplasty Treatment of Native Coarctation of the Aorta

An Observational Study by the CCISC
(Congenital Cardiovascular Interventional Study Consortium)

Objectives	The purpose of this study was to compare the safety and efficacy of surgical, stent, and balloon angioplasty (BA) treatment of native coarctation acutely and at follow-up.
Background	Controversy surrounds the optimal treatment for native coarctation of the aorta. This is the first multicenter study evaluating acute and follow-up outcomes of these 3 treatment options in children weighing >10 kg.
Methods	This is a multicenter observational study. Baseline, acute, short-term (3 to 18 months), and intermediate (>18 months) follow-up hemodynamic, imaging data, and complications were recorded.
Results	Between June 2002 and July 2009, 350 patients from 36 institutions were enrolled: 217 underwent stent, 61 underwent BA, and 72 underwent surgery. <u>All 3 arms showed significant improvement acutely and at follow-up in resting systolic blood pressure and upper to lower extremity systolic blood pressure gradient (ULG). Stent was superior to BA in achieving lower ULG acutely. Surgery and stent were superior to BA at short-term follow-up in achieving lower ULG. Stent patients had shorter hospitalization than surgical patients (2.4 vs. 6.4 days; $p < 0.001$) and fewer complications than surgical and BA patients (2.3%, 8.1%, and 9.8%; $p < 0.001$). The BA patients were more likely to encounter aortic wall injury, both acutely and at follow-up ($p < 0.001$).</u>
Conclusions	Stent patients had significantly lower acute complications compared with surgery patients or BA patients, although they were more likely to require a planned reintervention. At short-term and intermediate follow-up, stent and surgical patients achieved superior hemodynamic and integrated aortic arch imaging outcomes compared with BA patients. <u>Because of the nonrandomized nature of this study</u> , these results should be interpreted with caution. (J Am Coll Cardiol 2011;58:2664–74) © 2011 by the American College of Cardiology Foundation

Coarctação da Aorta

Table 7

Intermediate Follow-up Outcomes by Integrated Imaging

	Surgery (n = 16)	Balloon (n = 16)	Stent (n = 56)	p Value (2-Sided)
Any complications*	25.0%	43.8%	12.5%	0.020‡
Aortic wall injury	12.5%	43.8%	7.1%	0.003‡
Dissection/intimal tear	0.0%	6.3%	1.8%	0.598
Aneurysm	12.5%	43.8%	5.4%	<0.001
Coarct:Dao ratio, mean	0.98	0.79	0.80	0.011‡
Coarct:Dao ratio ≥0.6	88%	93%	89%	1.000
Any reobstruction	18.8%	18.8%	14.3%	0.923
Mild†	6.3%	18.8%	12.5%	
Moderate	6.3%	0%	1.8%	
Severe	6.3%	0%	0%	

*Defined as any moderate to severe reobstruction, aortic wall injury (aneurysm, dissection, intimal tear) or stent fracture. †Mild reobstruction was not considered as a complication in our analysis. ‡p < 0.05.

Coarct:Dao = narrowest coarctation dimension (mm)/the dimension of the descending aorta at the level of the diaphragm (mm).

Table 8

Reintervention

	Surgery (n = 72)	Balloon (n = 61)	Stent (n = 217)
Patients with reintervention	4	6	44
Patients with planned procedures	0	2	35
Patients with unplanned procedures	4	4	9
Time to first planned reintervention, yrs	NA	1.43 ± 1.70	1.14 ± 1.15
Time to first unplanned reintervention, yrs	2.24 ± 2.23	1.28 ± 1.43	2.84 ± 1.43

Forbes *et al*

JACC Vol. 58, No. 25, 2011

December 13/20, 2011:2664–74

Coarctação da Aorta

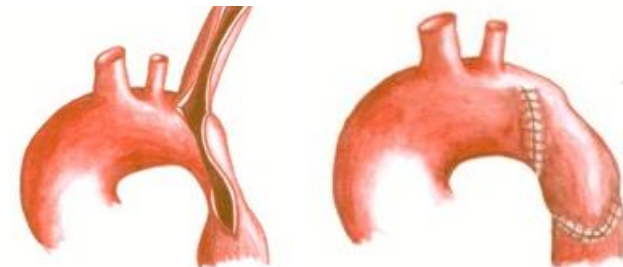
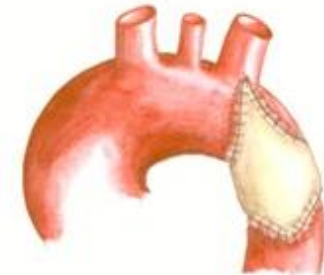
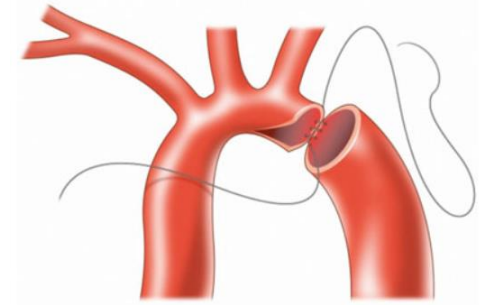
Tratamento Cirúrgico

- Método de eleição → RN, lactentes e crianças mais jovens
- Diversidade de técnicas cirúrgicas
- Abordagem dependente da idade e anatomia do arco
- Mortalidade geralmente baixa (1-4%)
- Morbidade
 - HTA paradoxal, lesão do nervo laríngeo recorrente, S. Roubo da Subclávia, Paraplegia

Coarctação da Aorta

Técnicas cirúrgicas

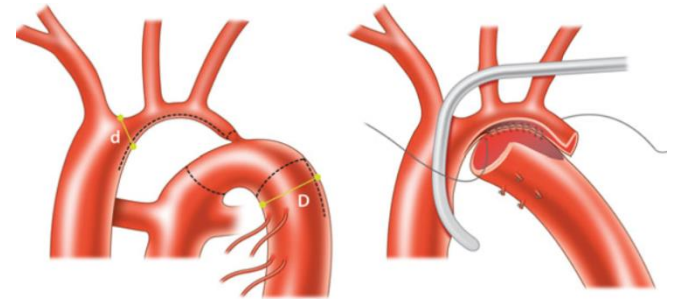
- Anastomose topo-a-topo
- Aortoplastia com *patch* (1957)
 - 7% desenvolvem aneurisma
- Aortoplastia com *flap* de Subclávia (1966)



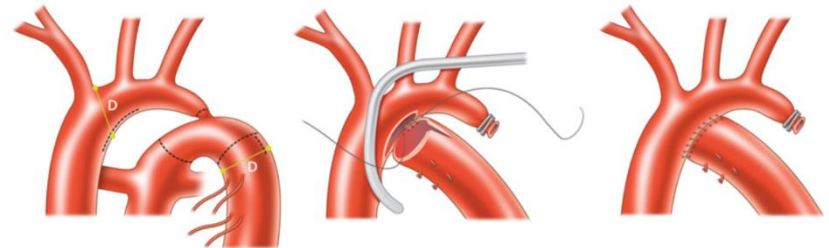
Coarctação da Aorta

Técnicas cirúrgicas

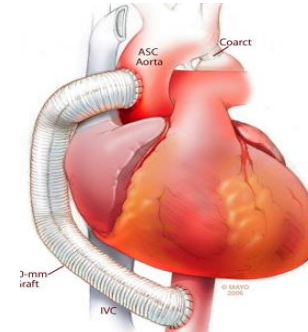
- Anastomose topo-a-topo alargada (1971)



- Anastomose termino-lateral
(por esternotomia ou toracotomia)



- Bypass extra-anatômico



Coarctação da Aorta

Timing cirúrgico

- RN com lesão crítica
 - 1-2 dias após estabilização clínica
- 6-12 meses (idealmente)
 - < Risco de HTA tardia
- Diagnóstico mais tardio
 - Intervenção logo que possível (< 6 meses)

Coarctação da Aorta

Tratamento - Intervenção percutânea

- ***Stent***

- Crianças (>25Kg), adolescente e adulto
- Lesão focal
- Stent bare/covered
 - Stents biodegradáveis indisponíveis
- Necessidade eventual de re-dilatação
- Complicações: migração/embolização, fracturas
- Menor risco de complicações vasculares (re-estenose, aneurisma)



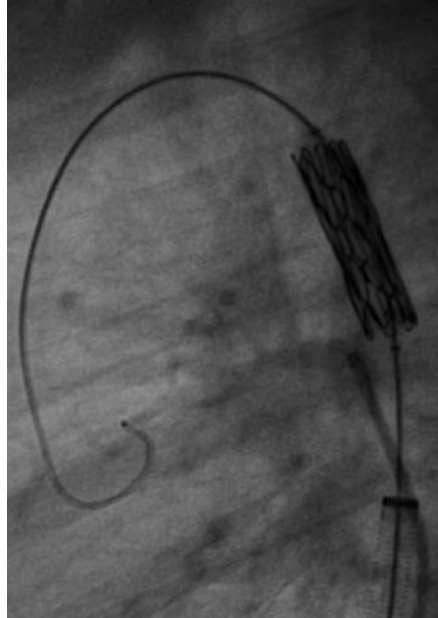
Coarctação da Aorta

Tratamento - Intervenção percutânea

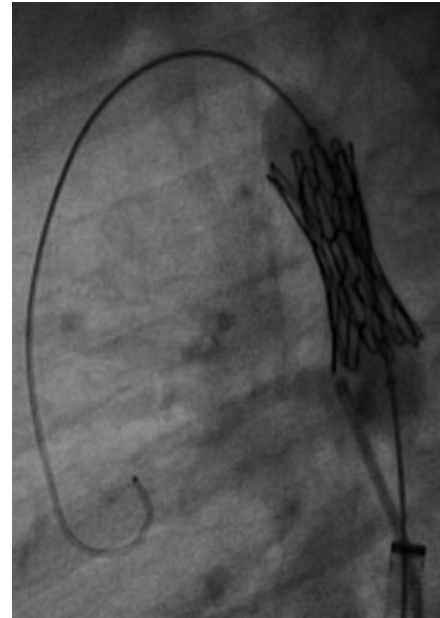
- FP, 9 anos (34 Kg) + HTA → CoAo nativa



Aortografia
Projecção lateral



Inner BiB
12x35 mm



Outer BiB 12x35
mm (5 atm)



Angiografia
pós-implantação

Coarctação da Aorta

Complicações imediatas

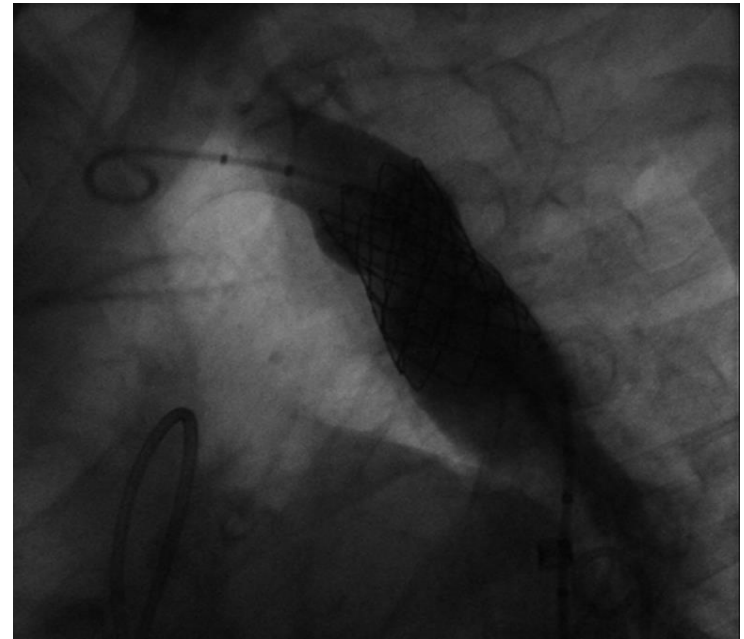
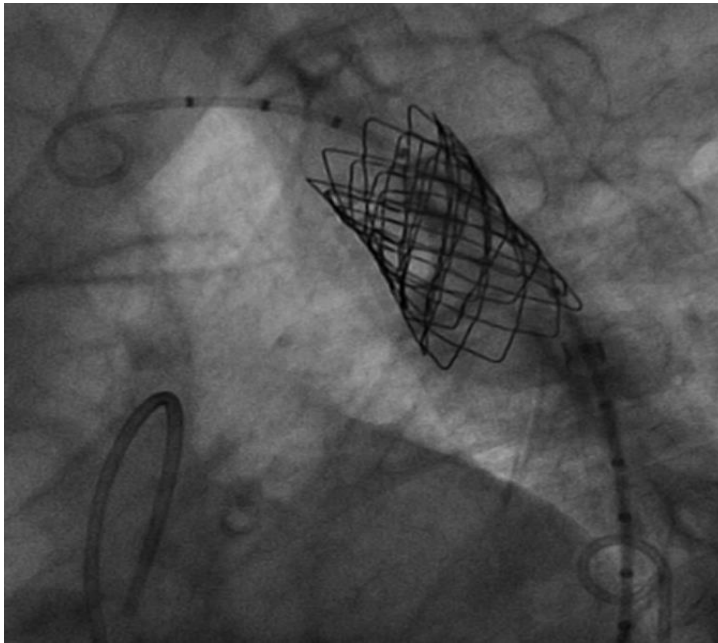
- Sp Implantação de CP Stent (CoAo Nativa)
 - Oclusão de vaso femoral → Sistema Proglide®



Coarctação da Aorta

Tratamento - Intervenção percutânea

- MM, 18 anos → Recoarctação (SP op de Correção CoAo com *flap* de subclávia em 1990)



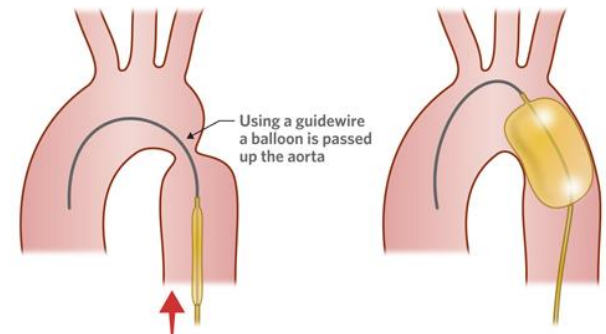
2 CP *Stents* cobertos

Coarctação da Aorta

Tratamento - Intervenção percutânea

- **Angioplastia com balão**

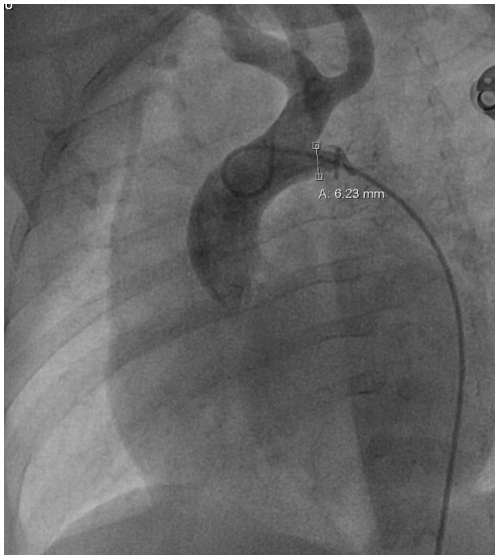
- Método implica lesão na íntima/média com risco dissecção/ruptura
- Paliativa em RN de alto risco
- Recoarctação em lactentes e crianças mais pequenas
- Complicações mais frequentes a longo prazo:
 - Re-estenose (< 40%)
 - Aneurismas (20-40%)



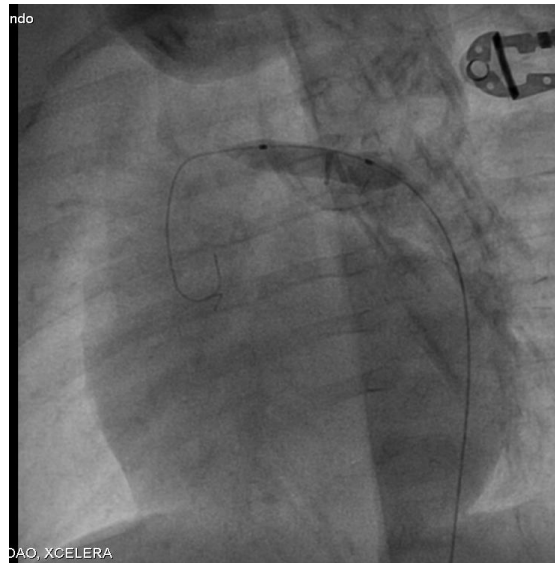
Coarctação da Aorta

Tratamento - Intervenção percutânea

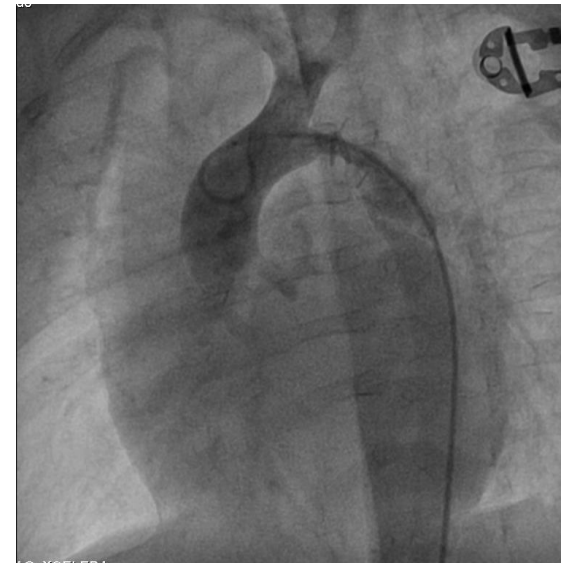
- **Angioplastia com balão** → GR, 5 meses (6Kg): Re-coartação



Obstrução crítica
PigTail 4F



Balão SABER®
6x20 mm



Aortografia
pós-dilatação

Coarctação da Aorta

Tratamento percutâneo – CH S. João/Porto

- **Jul 2008 – Set 2017**

- **n= 25 (≤ 18 anos)**

- **CoAo nativa – 14**

- » *Stent* – 14

- » Balão - 0

- **Re-CoAo – 11**

- » *Stent* – 9 (1 doente com 2 *stents*)

- » Balão – 3 (todos < 12 meses)

Coarctação da Aorta

AHA Scientific Statement

Indications for Cardiac Catheterization and Intervention in Pediatric Cardiac Disease

A Scientific Statement From the American Heart Association

Endorsed by the American Academy of Pediatrics and Society for Cardiovascular Angiography and Intervention

Timothy F. Feltes, MD, FAHA, Chair; Emile Bacha, MD; Robert H. Beekman III, MD, FAHA; John P. Cheatham, MD; Jeffrey A. Feinstein, MD, MPH; Antoinette S. Gomes, MD, FAHA; Ziyad M. Hijazi, MD, MPH, FAHA; Frank F. Ing, MD; Michael de Moor, MBBCh; W. Robert Morrow, MD; Charles E. Mullins, MD, FAHA; Kathryn A. Taubert, PhD, FAHA; Evan M. Zahn, MD; on behalf of the American Heart Association Congenital Cardiac Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Clinical Cardiology, and Council on Cardiovascular Radiology and Intervention

6.1. Native Coarctation and Recoarctation2621- 2623

Coarctação da Aorta

Complicações a longo prazo

Table 2 Long-term complications of CoA

Long-term complications

Local	Recoarctation, aneurysm, dissection, rupture, fistulae, endocarditis
Ascending aorta	Aneurysm, dissection, rupture, sinus of Valsalva fistula
Aortic valve	(Bicuspid valve), stenosis, regurgitation
Left ventricle	Hypertrophy, dilation, systolic dysfunction and/or diastolic dysfunction, heart failure, sudden cardiac death
Coronaries	Premature atherosclerosis, ischaemic heart disease
Cerebral	Berry aneurysms, intracranial bleeds, atherosclerosis, stroke
Systemic	Hypertension, reduced exercise capacity

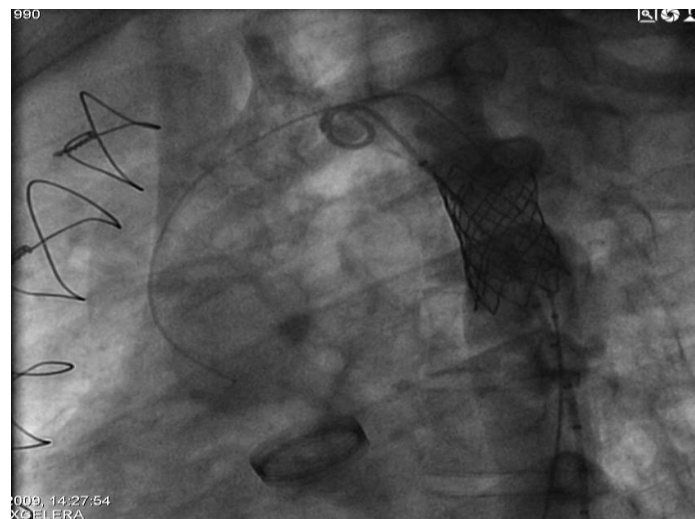
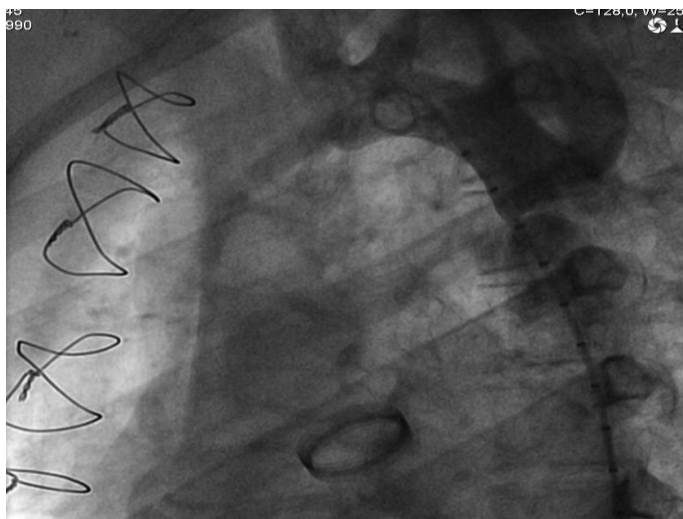
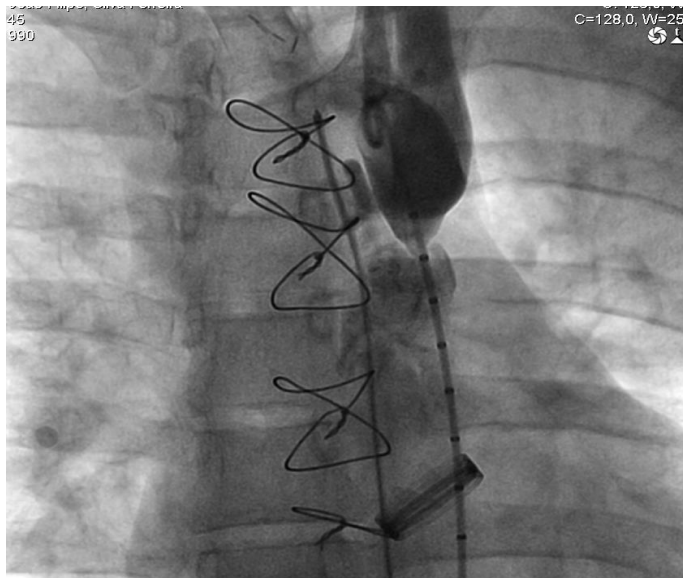
Coarctação da Aorta

Complicações a longo prazo

- JF, 17 anos – Status pós-Bentall (dissecção da aorta ascendente)
 - Coarctação da Aorta



Coarctação da Aorta



CP Stent 39mm, em BIB 18mm

Coarctação da Aorta

Lesão residual / Re-estenose

- Definição gradiente > 20 mmHg
- Mais comum após técnicas clássicas convencionais
- Período neonatal (até 30%)
- Hipoplasia do arco aórtico (~33%)

Necessidade de reavaliação periódica

- Exame clínico, ecodoppler, RM cardíaca combinados

Coarctação da Aorta

Factores de risco para HTA

- Tratamento cirúrgico tardio
 - 7% em < 1 ano; 33% em > 14 anos (Cohen, *et al*; 1989)
- Técnica cirúrgica utilizada
- Arco aórtico gótico (“não-românico”)
- Anomalia intrínseca da parede vascular



Coarctação da Aorta

Seguimento

- Follow-up vitalício necessário
- MAPA 24h anual >10 anos
- Prova de esforço periódica
- Imagiologia avançada (eco, RMC e TC)
- Re-intervenção frequente (25%)
 - Re-coarctação 4-14%
 - Disfunção válvula aórtica
 - Outras malformações ardiovasculares

Coarctação da Aorta

Participação em Desporto (competição)

Coarctation of the Aorta: Untreated

Recommendations

1. Athletes with coarctation and without significant ascending aortic dilation (z score ≤ 3.0 ; a score of 3.0 equals 3 standard deviations from the mean for patient size) with a normal exercise test and a resting systolic blood pressure gradient < 20 mm Hg between the upper and lower limbs and a peak systolic blood pressure not exceeding the 95th percentile of predicted with exercise can participate in all competitive sports (Class I; Level of Evidence C).
2. Athletes with a systolic blood pressure arm/leg gradient > 20 mm Hg or exercise-induced hypertension (a peak systolic blood pressure exceeding the 95th percentile of predicted with exercise) or with significant ascending aortic dilation (z score > 3.0) may be considered for participation only in low-intensity class IA sports (Class IIb; Level of Evidence C).

AHA/ACC SCIENTIFIC STATEMENT:

Eligibility and Disqualification,
Recommendations for Competitive

Athletes
JACC, 2015

Coarctação da Aorta

Participação em Desporto (competição)

Coarctation of the Aorta: Treated by Surgery or Balloon and Stent

Recommendations

1. Athletes who are >3 months past surgical repair or stent placement with <20 mm Hg arm/leg blood pressure gradient at rest, as well as (1) a normal exercise test with no significant dilation of the ascending aorta (z score <3.0), (2) no aneurysm at the site of coarctation intervention, and (3) no significant concomitant aortic valve disease, may be considered for participation in competitive sports, but with the exception of high-intensity static exercise (classes IIIA, IIIB, and IIIC), as well as sports that pose a danger of bodily collision (*Class IIB; Level of Evidence C*).
2. Athletes with evidence of significant aortic dilation or aneurysm formation (not yet at a size to need surgical repair) may be considered for participation only in low-intensity (classes IA and IB) sports (*Class IIB; Level of Evidence C*).

AHA/ACC SCIENTIFIC STATEMENT:

Eligibility and Disqualification,
Recommendations for Competitive
Athletes
JACC, 2015



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Coarctação da Aorta

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