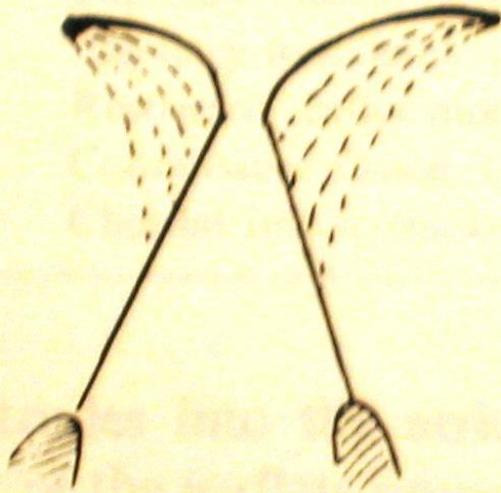


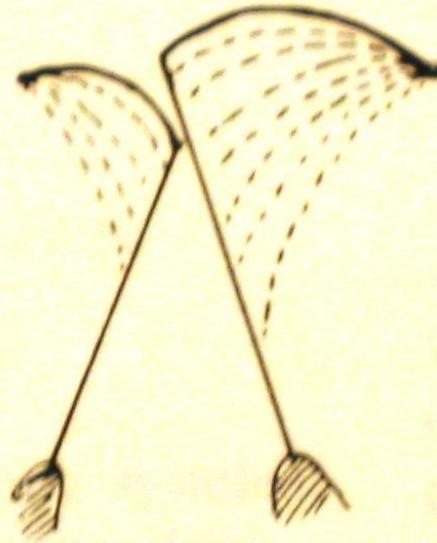
PATOLOGIA MITRAL SINDROME DE SHONE

José I. Aramendi
Hospital de Cruces

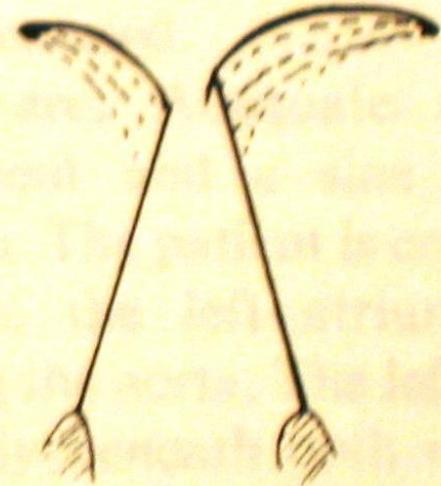
CLASIFICACION INSUFICIENCIA MITRAL CARPENTIER 1983



TYPE I
NORMAL LEAFLET MOTION



TYPE II
LEAFLET PROLAPSE



TYPE III
RESTRICTED LEAFLET MOTION

Mitral valve stenosis

Carpentier

Type A (normal papillary muscle) Supravalvular ring

Leaflet fusion

Type B (Abnormal papillary muscle)

Parachute mitral valve

Hammock valve

Papillary muscle abnormality

RECONSTRUCTIVE SURGERY IN CONGENITAL MITRAL VALVE INSUFFICIENCY (CARPENTIER'S TECHNIQUES)

<i>Type</i>	<i>n</i>	<i>% of 145</i>
Type I: Normal leaflet motion	31	21.4
1. Annular dilatation	7	4.8
2. Cleft leaflet	18	1.2
3. Leaflet defect	6	4.1
Type II: Leaflet prolapse	79	54.5
1. Chordal elongation	46	31.7
2. Papillary muscle elongation	24	16.5
3. Absence of chordae	9	6.2
Type III: Restricted leaflet motion	35	24.1
A: Normal papillaary muscles	15	
1. Commissure papillary fusion	8	5.5
2. Short chordae	7	4.8
B: Abnormal papillary muscles	20	
1. Parachute mitral valve	5	3.4
2. Hammock mitral valve	12	8.2
3. Papillary muscle hypoplasia	3	2

<i>Procedures</i>	<i>n</i>	<i>% of 138</i>
Annuloplasty		
Carpentier prosthetic ring	70	48
Plication of the commissures	6	4
Plication of the anulus	5	3
Semicircular suture	5	3
Cleft suture without patch	14	10
Pericardial patch enlargement		
Anterior leaflet	14	10
Posterior leaflet	8	5
Chordal shortening	46	33
Papillary muscle shortening	24	17
Chordal transposition	12	8
Commissurotomy	15	10
Splitting of papillary muscle	13	9
Fenestration of papillary muscle	9	6

Repair of congenital malformations of the mitral valve
Edvin Prifti N=94

Mitral valve stenosis 21

Type A (normal papillary muscles) 8

Papillary muscles commissure fusion 6

Supravalvular ring 2

Type B (abnormal papillary muscles) 13

Parachute mitral valve 4

Hammock mitral valve 9

Repair of congenital malformations of the mitral valve

Edvin Prifti,

25 pt. < 1year of age

Variables	Failed (n = 8)	Successful (n = 86)	<i>p</i> Value
• Age < 12 months	6/2	19/67	0.004
• Mitral valve stenosis/ • mitral valve insufficiency	5/3	16/70	0.013
• Hammock valve	5/8	4/82	0.006
• Congestive heart failure	3/5	2/84	0.004
• pulmonary artery pressure	46 ± 12	27.6 ± 8	0.001

MORTALITY

OVERALL	8/94	8%
MV Stenosis	5/21	25% all hammock v.

MITRALVALVE REPLACEMENT

N= 54

Congenital 43

Mitral stenosis 14

Mitral regurgitation 12

Complete atrioventricular septal defect 12

Partial atrioventricular septal defect 5

Endocarditis 9

Bacterial 7

Viral 2

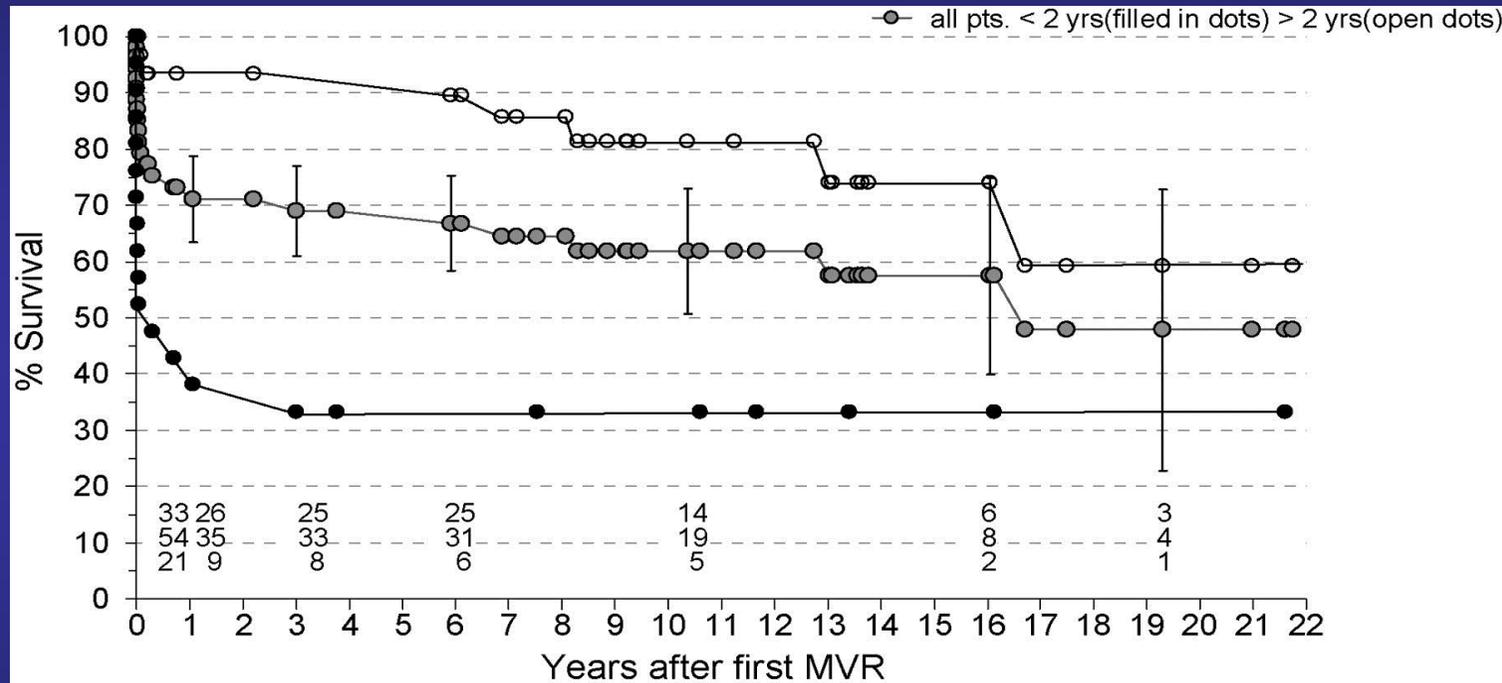
(normal mitral valve 5, previous repair of atrioventricular septal defect 2, previous rheumatic fever 1, marfan syndrome 1)

Rheumatic 2

Thirty-day mortality was 42% in patients < 2-year-old, and 6% in older patients.

Break-point: Prosthetic valve size <23

Actuarial freedom from death (including 30-day death) following mitral valve replacement for the entire cohort, and for subgroups of patients 2-year-old



Beierlein W. et al.; Eur J Cardiothorac Surg 2007;31:860-865

SHONE'S ANOMALY

N= 27

MITRAL ANATOMY

Parachute mitral valve	10
Typical mitral stenosis	14
Supravalvular ring	11
normal valve	3

Brown J. W. et al.; Ann Thorac Surg 2005;79:1358-1365

Surgical Procedures on the Left Ventricular Outflow Tract Performed in 27 Patients

<u>Procedure</u>	<u>No. of Procedures</u>
Coarctation or recoarctation repair	17
Resection of subaortic stenosis	9
Myotomy/myectomy	6
Open aortic valvotomy	4
Balloon aortic valvotomy	4
Patch aortoplasty	3
Balloon angioplasty of coarctation	2
Closed aortic valvotomy	1
Ross-Konno procedure	1
Ross procedure	1
Total	48

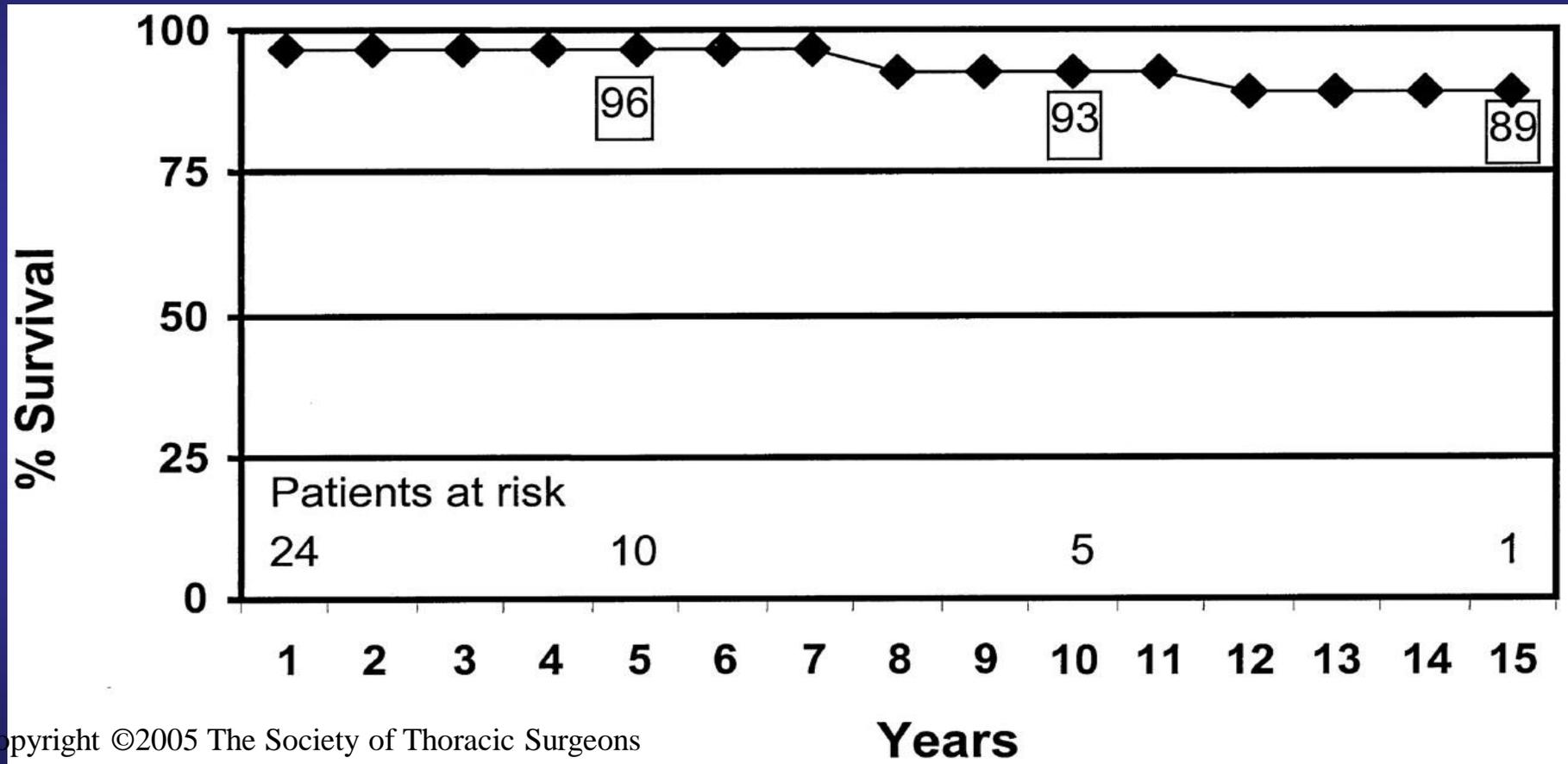
Surgical Procedures on the Left Ventricle Inflow Performed in 17 Patients

<u>Procedure</u>	<u>No. of Procedures</u>
Resection of supramitral ring	9
Mitral valve replacement	7 (1 initial)
Mitral valve dilatation	4
Mitral valvuloplasty	1
Mitral commissurotomy	1
Total	22

no mortality at initial operation

3 late deaths at reoperation

Kaplan-Meier estimate of survival in patients with Shone's anomaly



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Brown J. W. et al.; Ann Thorac Surg 2005;79:1358-1365

MITRAL VALVE REPAIR IN CHILDREN

STELLIN, G. PADOVA N=93

- REGURGITATION 48
- STENOSIS 45
- ASSOCIATED LESIONS 72%
- Mean age 4.5yr
- Hammock valve 6. parachute 12
- Hospital mortality 7.5% . Late mortality 8%
- Freedom fom replacement:
 - 77% at 20yr

Surgical repair of congenital mitral valve malformations in infancy and childhood: A single-center 36-year experience.

Stellin, Giovanni; Padalino, Massimo; MD, PhD; Vida, Vladimiro; MD, PhD; Boccuzzo, Giovanna; Orru, Emanuele; Biffanti, Roberta; Milanese, Ornella; Mazzucco, Alessandro

Journal of Thoracic & Cardiovascular Surgery. 140(6):1238-1244, December 2010.

MV malformations	No. of patients
MV regurgitation (or prevalent)	48
Type I (normal leaflet motion)	32
Annular dilatation	12
Cleft anterior leaflet	18
Leaflet defect	2
Type II (leaflet prolapse)	9
Elongated chordae	9
Type III (restricted leaflet motion)	7
Type A (normal papillary muscles)	
Papillary muscles commissure fusion	1
Short chordae	2
Type B (abnormal papillary muscles)	
Parachute MV	2
Hammock MV	2
MV stenosis (or prevalent)	45
Type A (normal papillary muscles)	24
Papillary muscle commissure fusion	14
Supravalvular membrane	7
Double-orifice MV	3
Type B (abnormal papillary muscles)	21
Parachute MV	10
Hammock MV	4
Shone complex	5
Arcade MV	2
Total	93

CMV, Congenital mitral valve; MV, mitral valve.

Technique	No. of patients
Papillary muscle splitting	35
Cleft closure	21
Annuloplasty (Kaye, Paneth)	20
Supravalvular ring excision	16
Commissurotomy	15
Chordal shortening	9
Ring annuloplasty	6
Commissuroplasty	4
Accessory orifice closure	2

Outcomes	Variables				
	Complex MV (OR)	MV stenosis (OR)	Parachute MV (OR)	Before 1984 (OR)	Age, y (OR)
Any adverse outcome	3.173	<i>2.520</i>	-	-	-
Early death	-	-	7.122	18.40	0.636
Late death	-	<i>6.973</i>	-	-	-
Preoperative medical therapy	-	-	4.889	-	-
Reoperation on MV	-	-	6.817	-	-
Operation on MV before 1 y of age	-	7.219	-	-	-
Heart failure at follow-up	-	-	6.100	-	-
Arrhythmias	-	-	6.377	4.744	-

Variables significant at a level of .05 < P < .1 are shown in italics. MV, Mitral valve; OR, odd ratio.

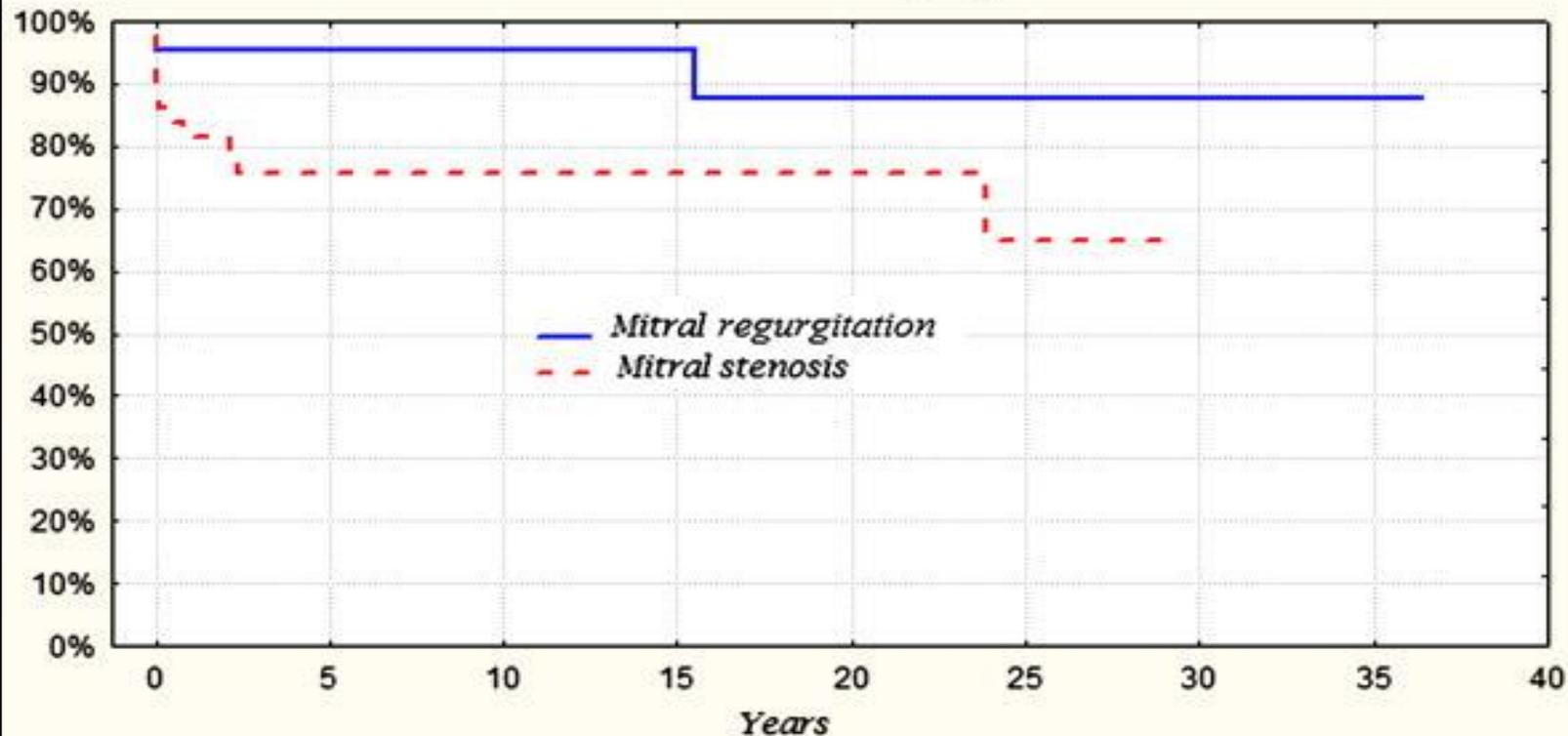
Surgical repair of congenital mitral valve malformations in infancy and childhood: A single-center 36-year experience.

Stellin, Giovanni; Padalino, Massimo; MD, PhD; Vida, Vladimiro; MD, PhD; Boccuzzo, Giovanna; Orru, Emanuele; Biffanti, Roberta; Milanesi, Ornella; Mazzucco, Alessandro

Journal of Thoracic & Cardiovascular Surgery. 140(6):1238-1244, December 2010.

DOI: 10.1016/j.jtcvs.2010.05.016.x

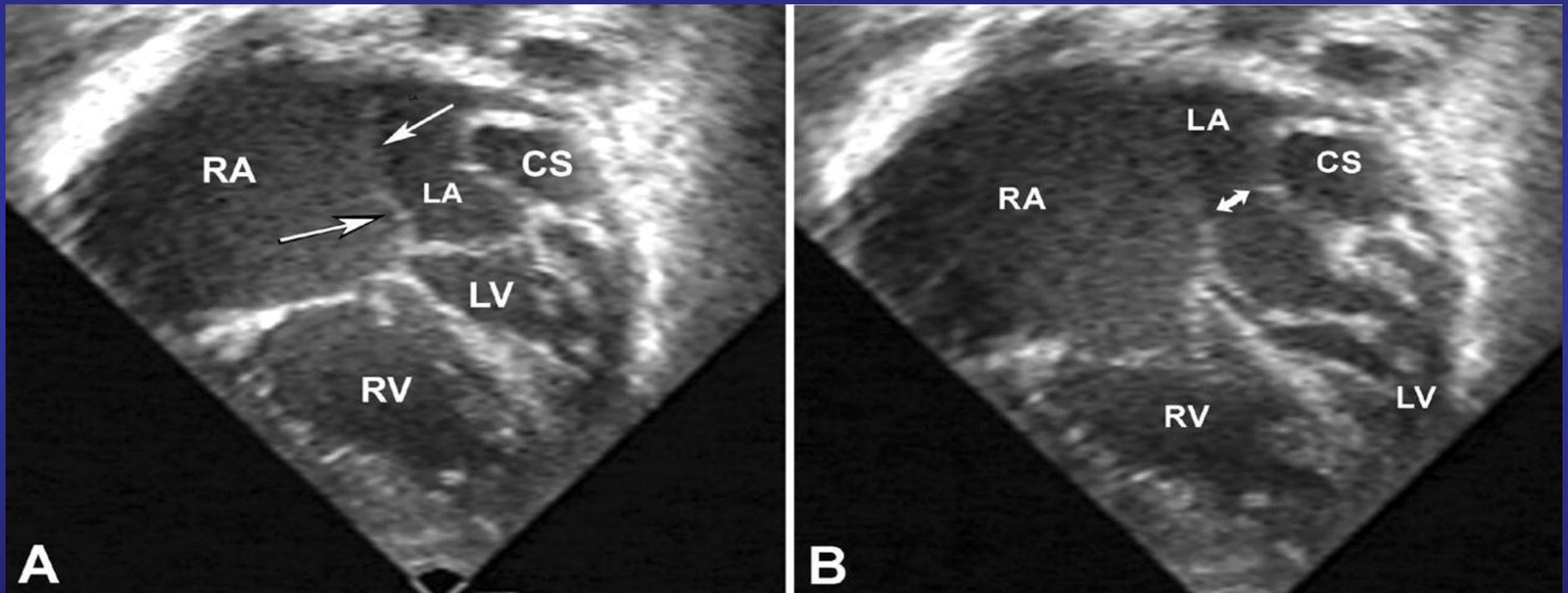
Cumulative survival in mitral regurgitation and mitral stenosis



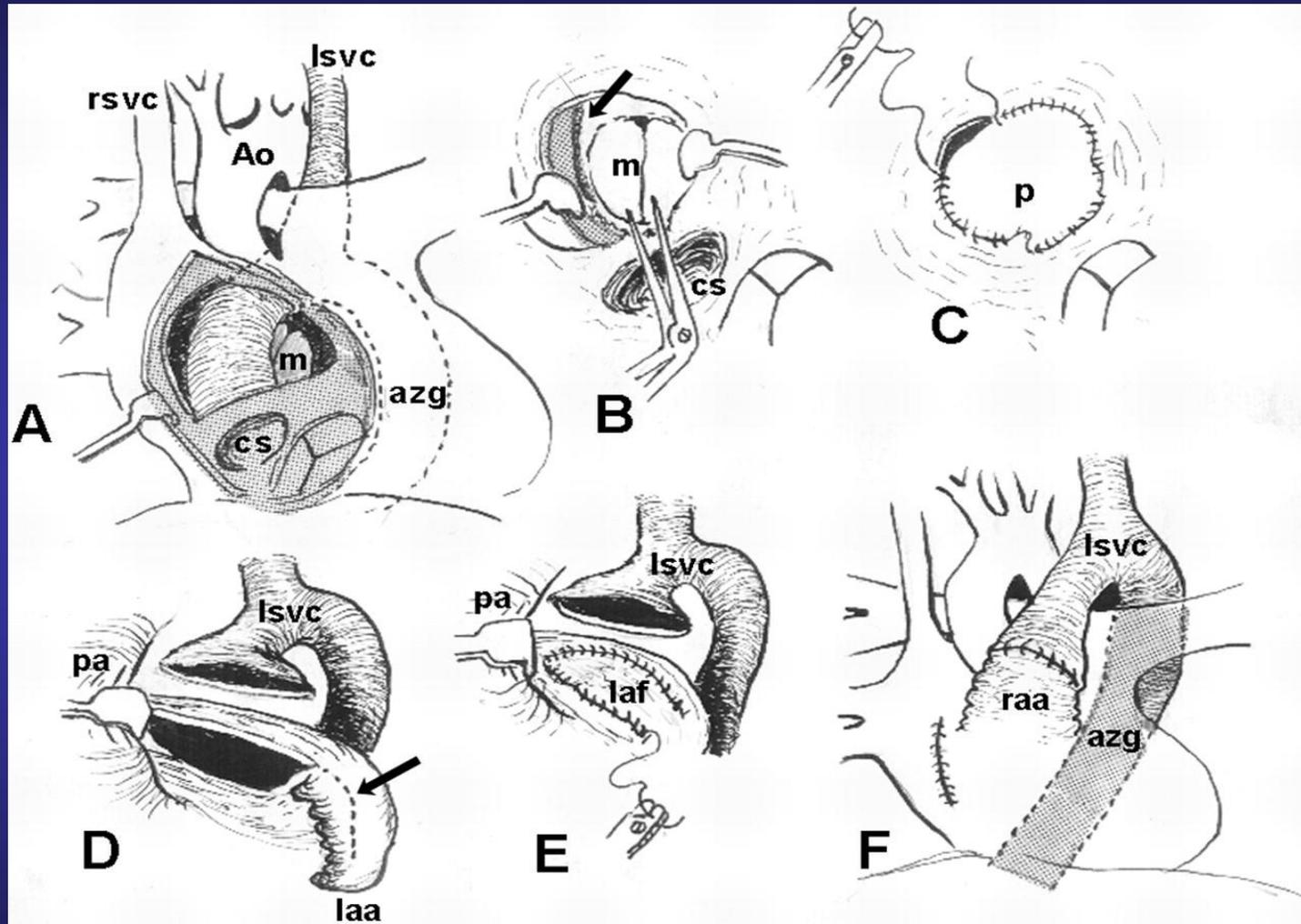
OBSTRUCCION TRACTO DE ENTRADA VI POR VENA CAVA SUPERIOR IZDA

- Ausencia de vena innominada
- Dilatación del seno coronario
- Hipertensión pulmonar severa
- Imita cor triatriatum
- Anillo mitral menor que el tricuspídeo

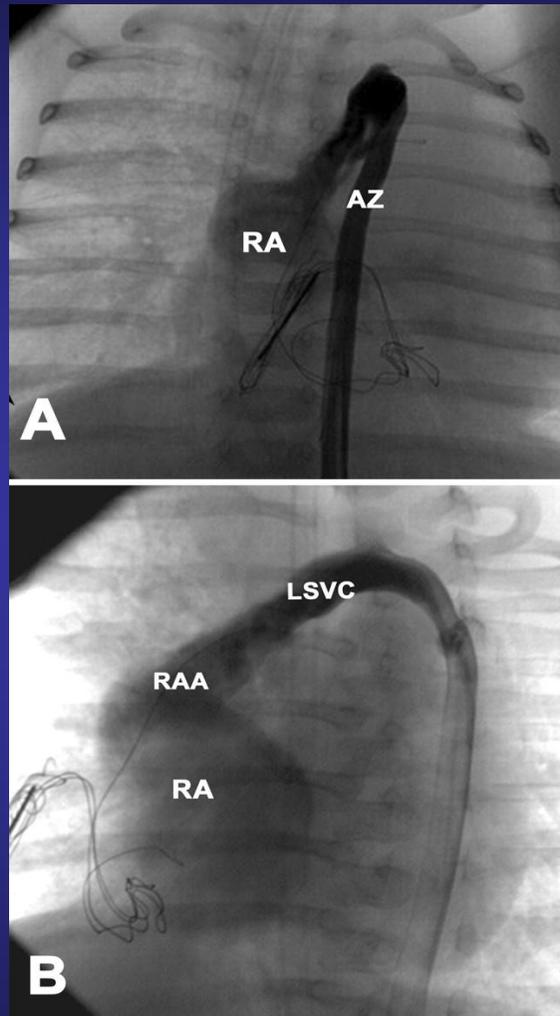
OBSTRUCCION TRACTO DE ENTRADA VI POR VENA CAVA SUPERIOR IZDA



Vargas F. J. et al.; Ann Thorac Surg 2006;82:191-196



Postoperative catheterization



Vargas F. J. et al.; Ann Thorac Surg 2006;82:191-196

HOSPITAL DE CRUCES (2000-2015)

PATOLOGIA MITRAL N=44

• Cleft aislado	5
• IM post canal AV	17
• M. Hipertrófica obstructiva	3
• S de Shone	9
– V. Hamaca	3
– Paracaídas	5
– Prolapso	1
• Obstrucción entrada VCSI	2
• V. hamaca parcial	3
• V. hamaca	2
• Pseudoparacaídas	2
• Estenosis protésica	1

HOSPITAL DE CRUCES

PATOLOGIA MITRAL N=44

• Sustitución mitral	15
• Reparación mitral	23
– Cierre cleft	2
– Ampliación parche velo post	9
– Anillo Carpentier	3
– Anillo reabsorbible Kalangos	5
– Comisuras, m. papilares	3
– Comisurotomía + neocuerda Goretex	1
• Reoperación (1repar, 4sust)	5

HOSPITAL DE CRUCES

PATOLOGIA MITRAL N=44

- Edad media 82m (5d-520m)
- 12p. <1 año (27%)
- Mortalidad 6/44 13.6%
 - Sustitución 3/19 15,7%
 - Reparación 3/23 13%

Síndrome de Shone

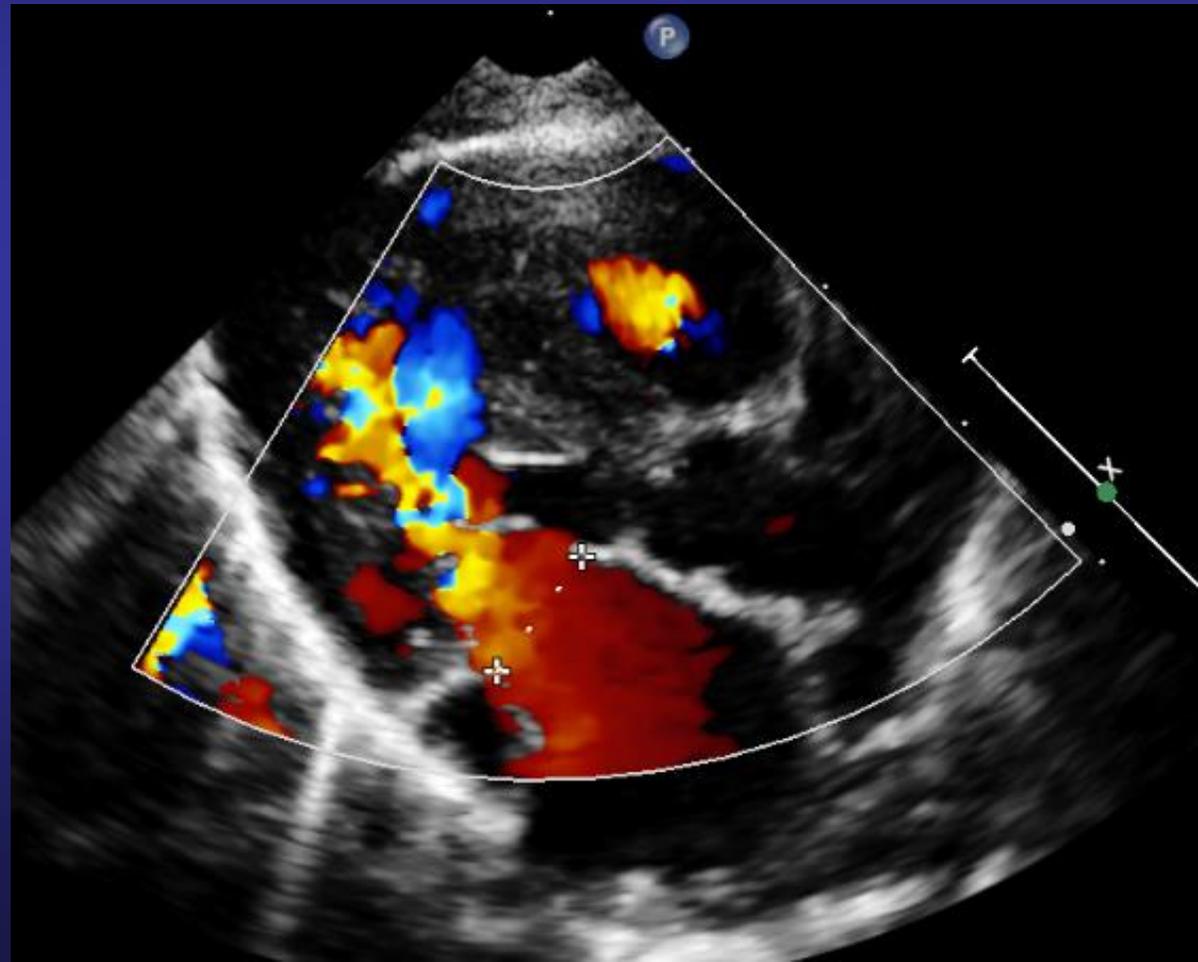
9 días:

ampliación de arco

3 meses:

estenosis aórtica +

Doble lesión mitral



Síndrome de Shone

v. Aórtica monocuspide

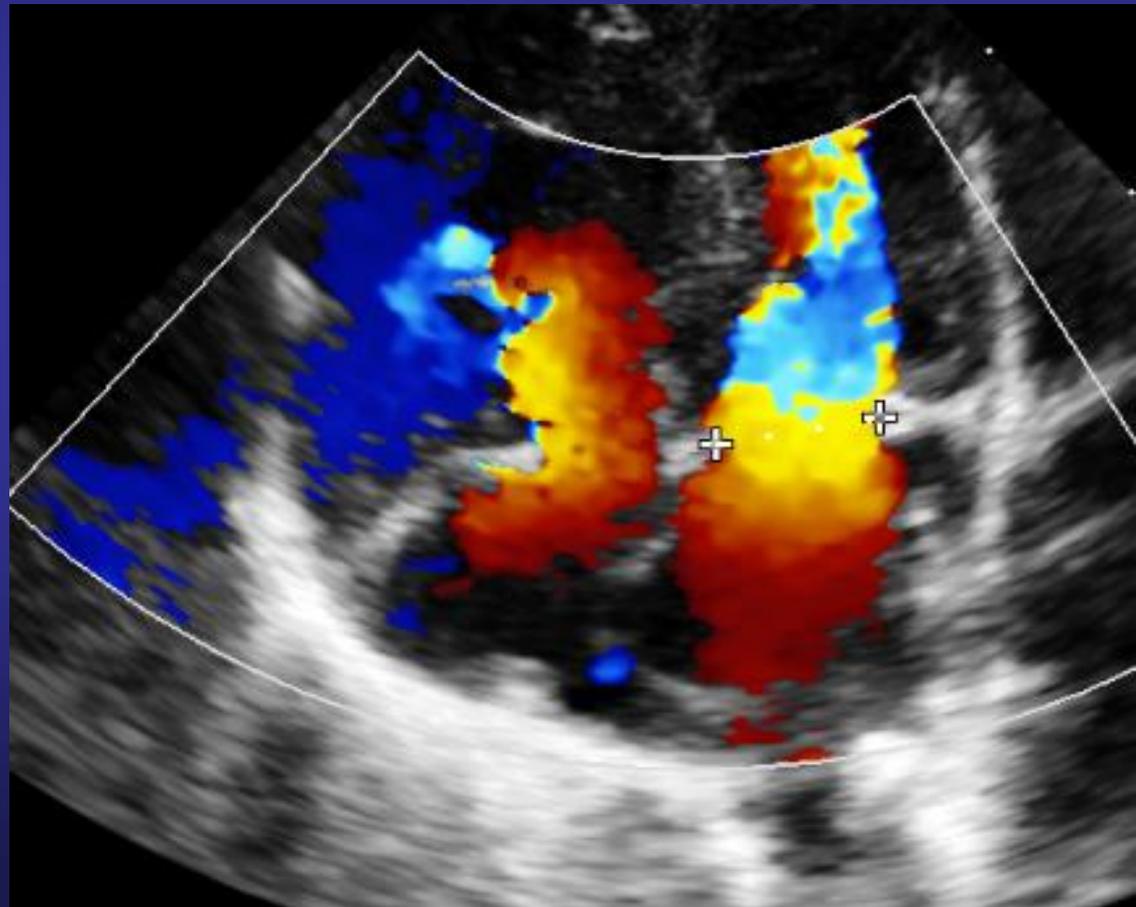
Comisurotomía

9mm diámetro Z=1

v. Mitral en hamaca

Comisurotomía +

Neo cuerda Goretex



PHILIPS

KILIAN

25/02/2015

10:24:12

T1s1.2 MI 1.2

JPEG

21/10/2014 2004326

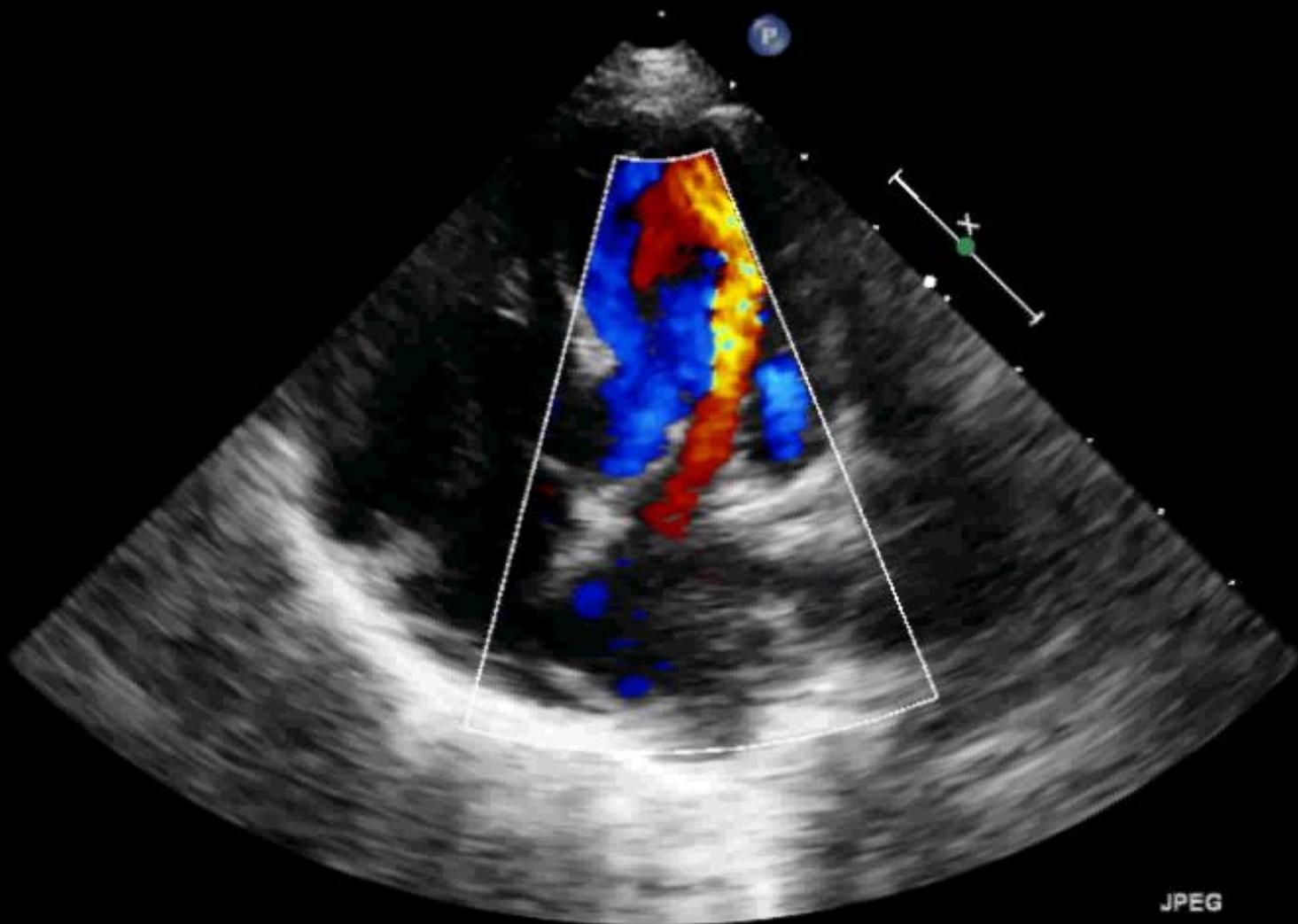
S8-3/CRUCES

FA 20Hz
9.0cm

2D
73%
C 50
P Des.
Gral.

FC
77%
3.0MHz
FP Alt.
Med.

M3 M4
+77.0
-77.0
cm/s



JPEG

*** lpm

PHILIPS

KILIAN

25/02/2015

10:23:54

TIs1.6

MI 1.0

JPEG

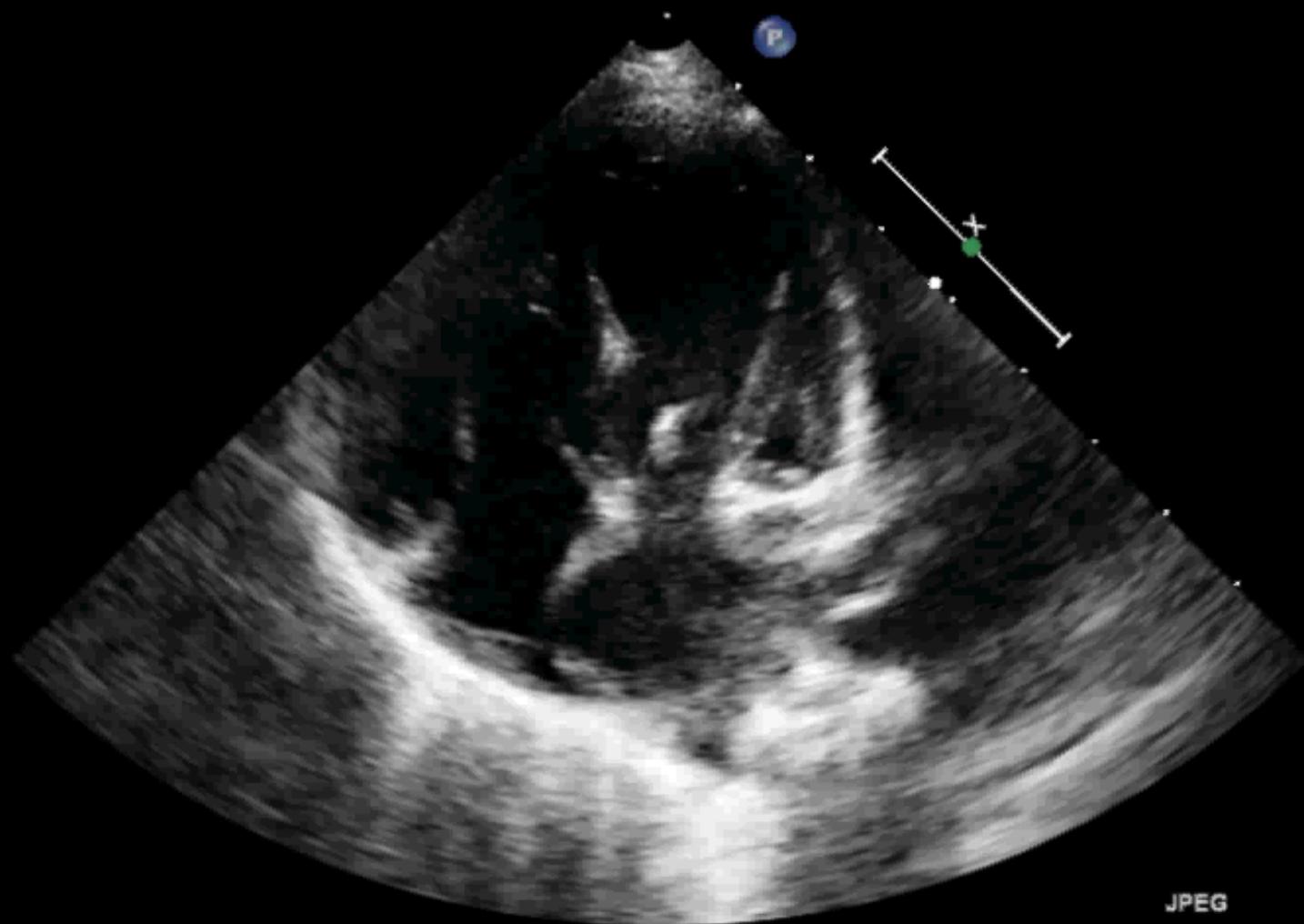
21/10/2014 2004326

S8-3/CRUCES

FA 61Hz
9.0cm

M3

2D
67%
C 50
P Des.
Gral.



JPEG

*** lpm

Síndrome de Shone

Hipoplasia de arco
2 CIV muscular y
Perimembranosa
Patología mitral:
Dudoso doble orificio
Prolapso
Reparación de arco
Banding AP



Síndrome de Shone

6 meses después
CIV flujo bidireccional
Gradiente 100 mmHg
Doble orificio mitral
Gradiente máximo
18 mmHg
Prolapso
Flujo aórtico normal

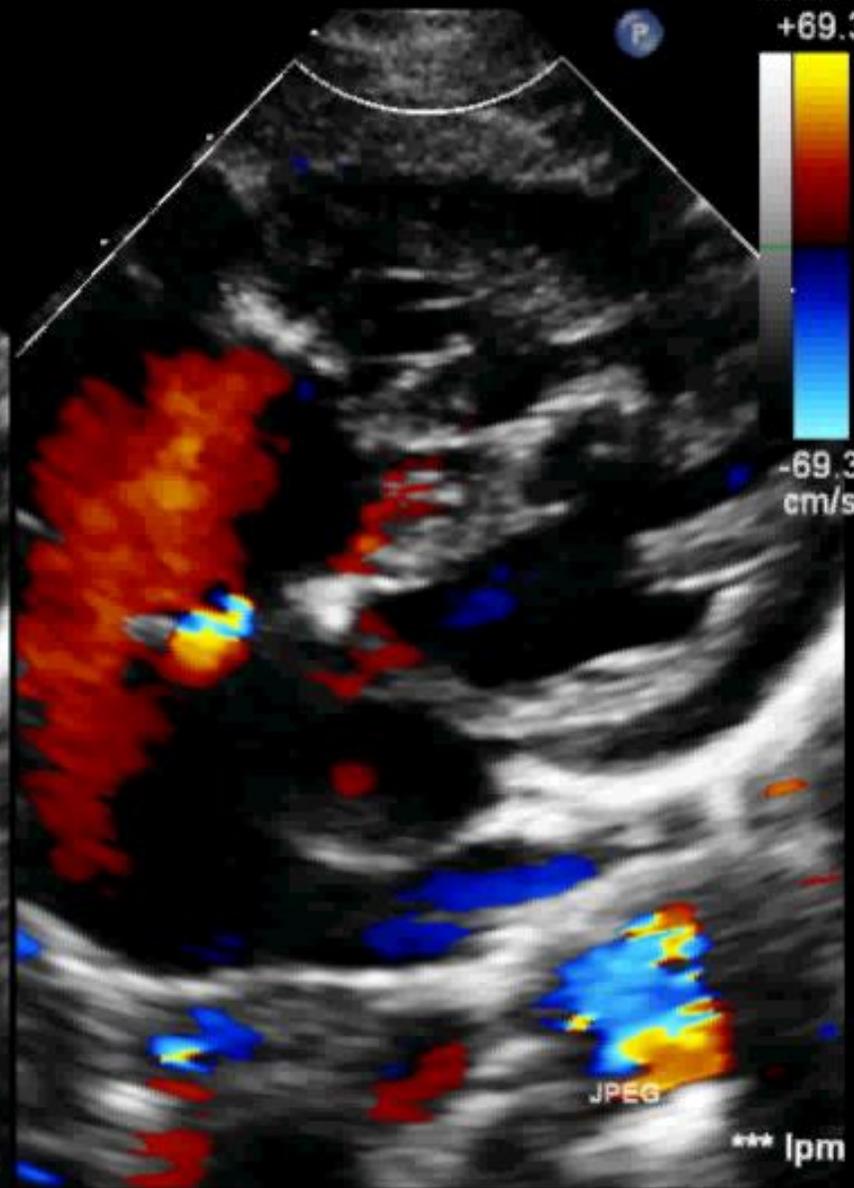


2016178

S8-3/CRUCES

FA 9Hz
9.0cm

2D
71%
C 50
P Des.
Gral.
FC
77%
3.0MHz
FP Alt.
Med.



CONCLUSIONES

En el síndrome de Shone 1º actuar sobre el tracto de salida

La estenosis mitral en el 1º año de vida es difícil de reparar

La sustitución valvular mitral por debajo de los 2 años conlleva una alta mortalidad

La insuficiencia mitral tiene un comportamiento más benigno y es posible repararla en la mayoría de los casos y a cualquier edad con resultados variables. Ganar tiempo.

El anillo mitral reabsorbible amplía las posibilidades de reparación en niños pequeños al permitir el crecimiento

La válvula mitral en hamaca tiene el peor pronóstico

VALVULA HAMACA PARCIAL

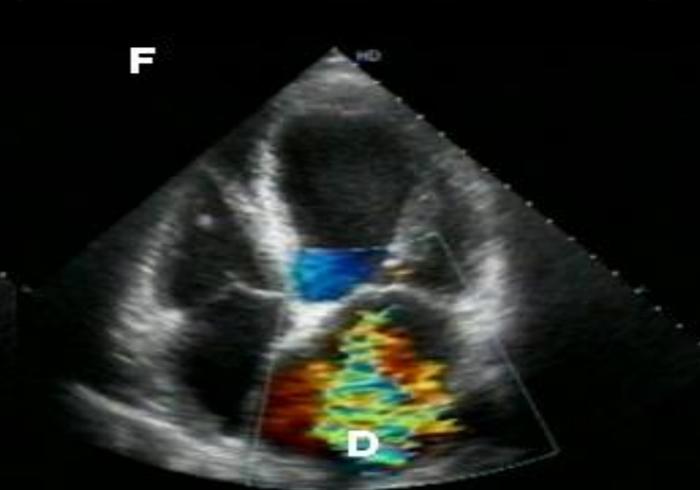
DEFINICION

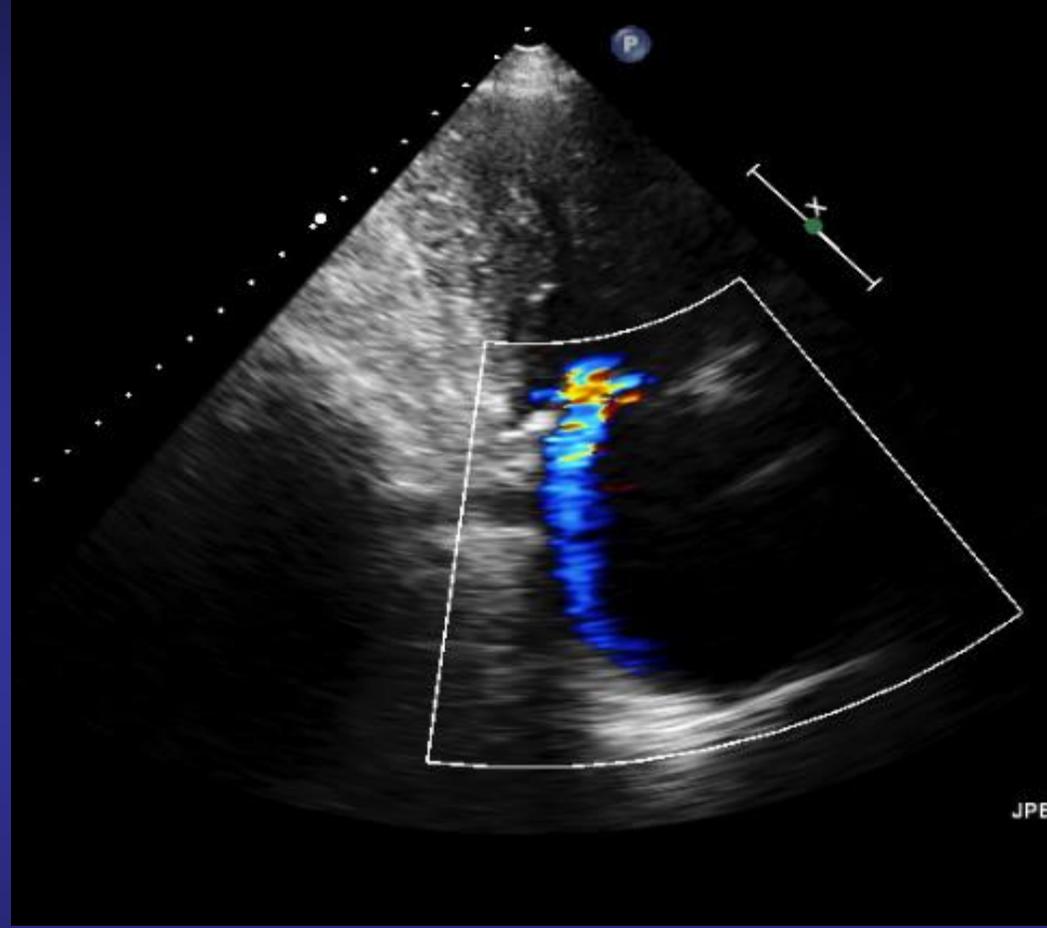
- Afecta solamente al velo posterior
- Presencia de m. papilares accesorios (3 ó 4 fusionados en empalizada)
- Produce insuficiencia mitral severa
- Mecanismo: restricción en sístole de P2 tipo III b de Carpentier

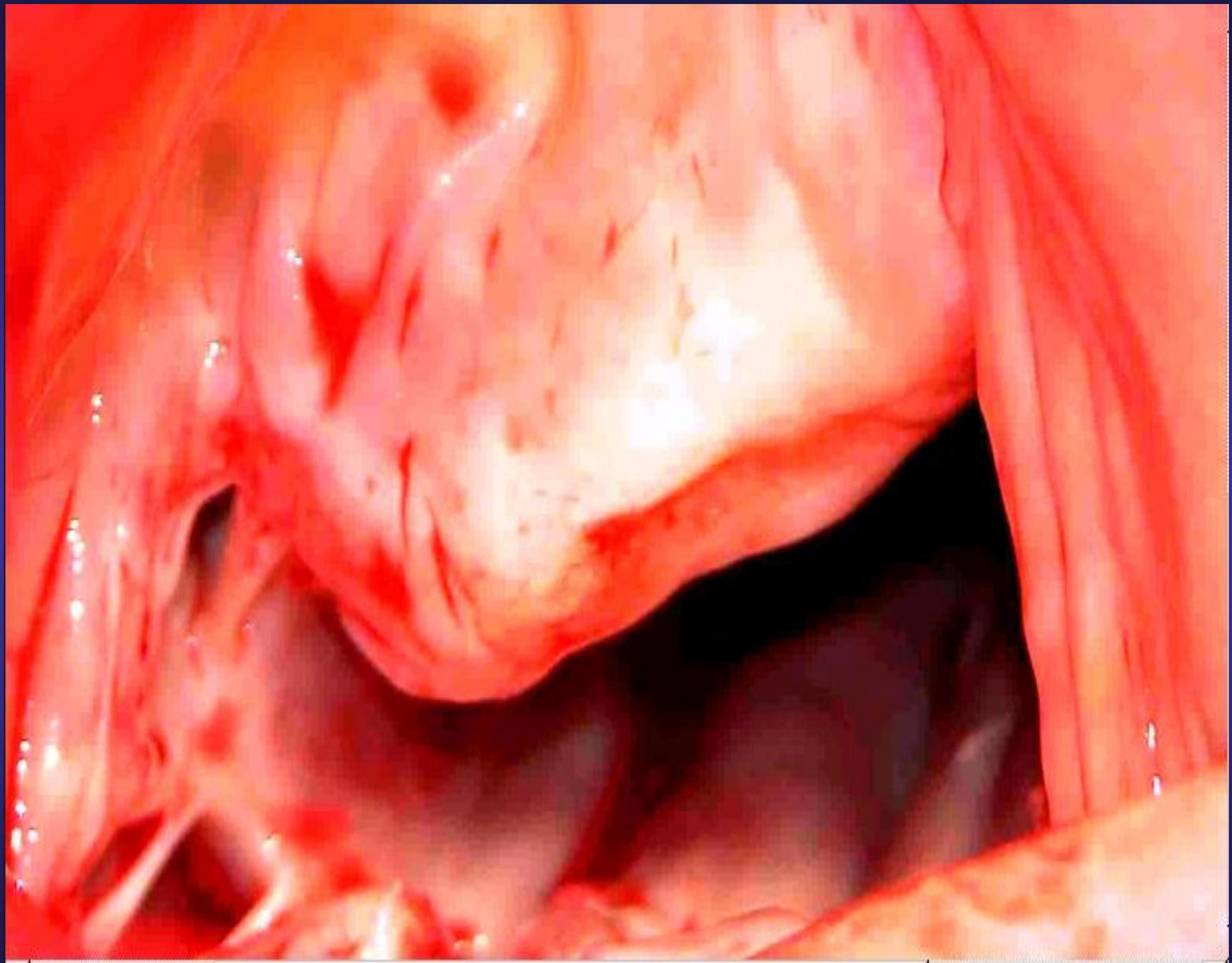
Partial Hammock Valve: Surgical Repair in Adulthood

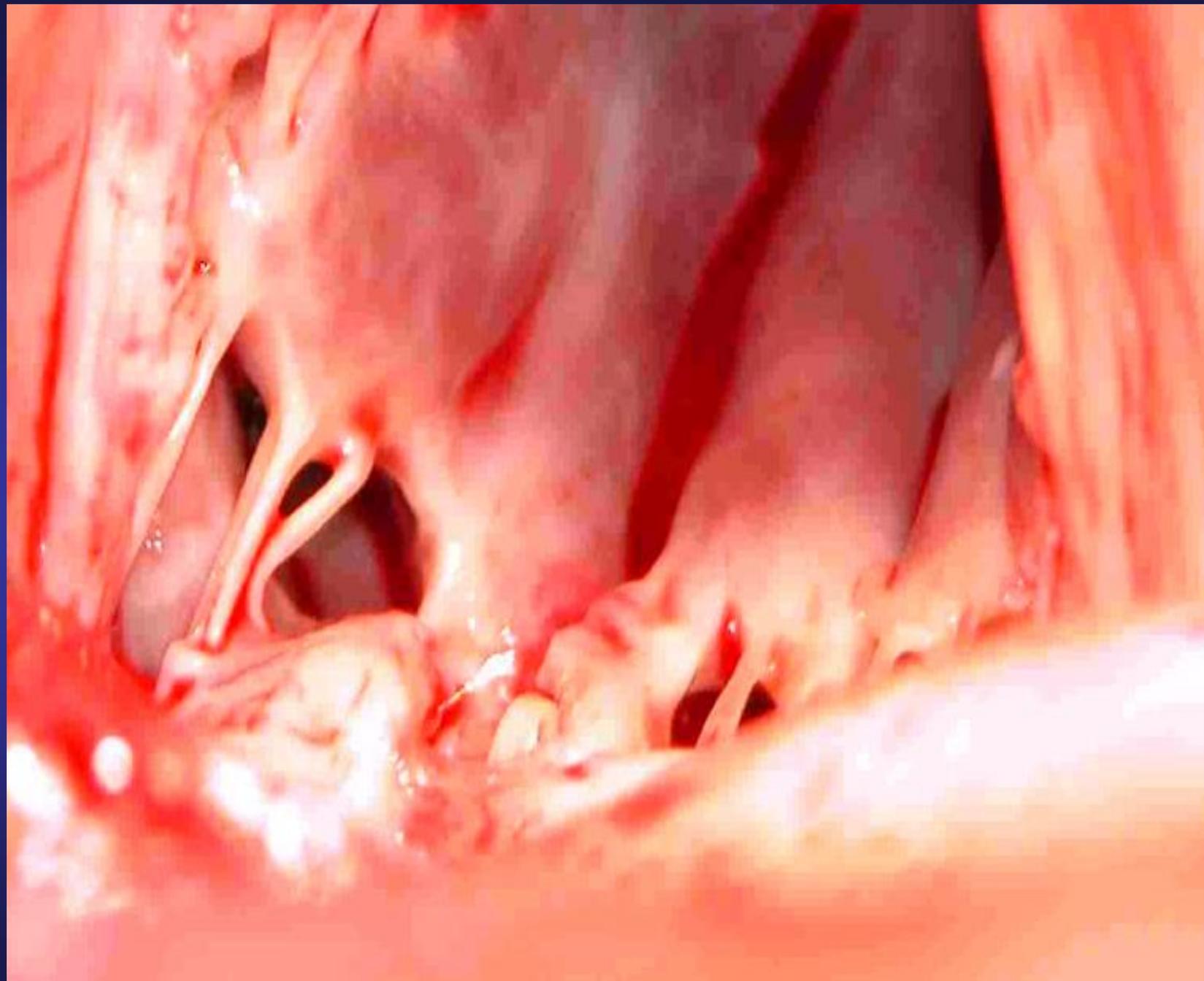
José I. Aramendi, MD, Miguel A. Rodríguez, MD,
Roberto Voces, MD, Pedro Pérez, MD, and
David Rodrigo, MD

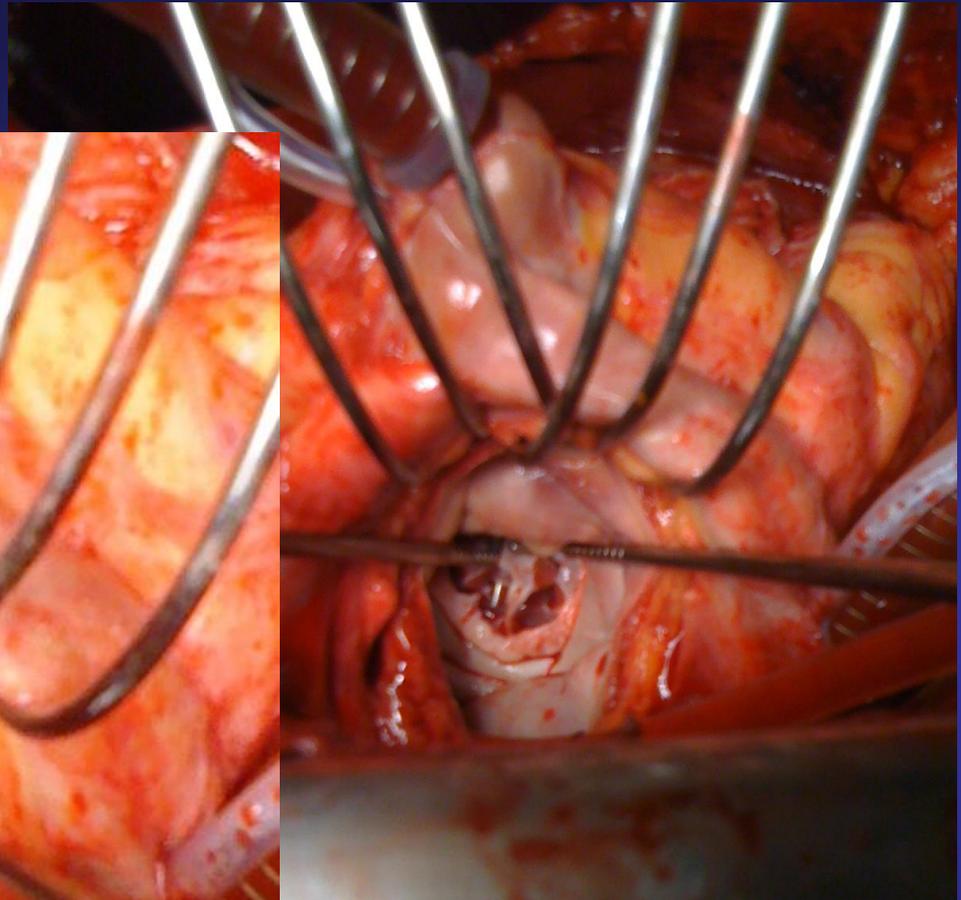
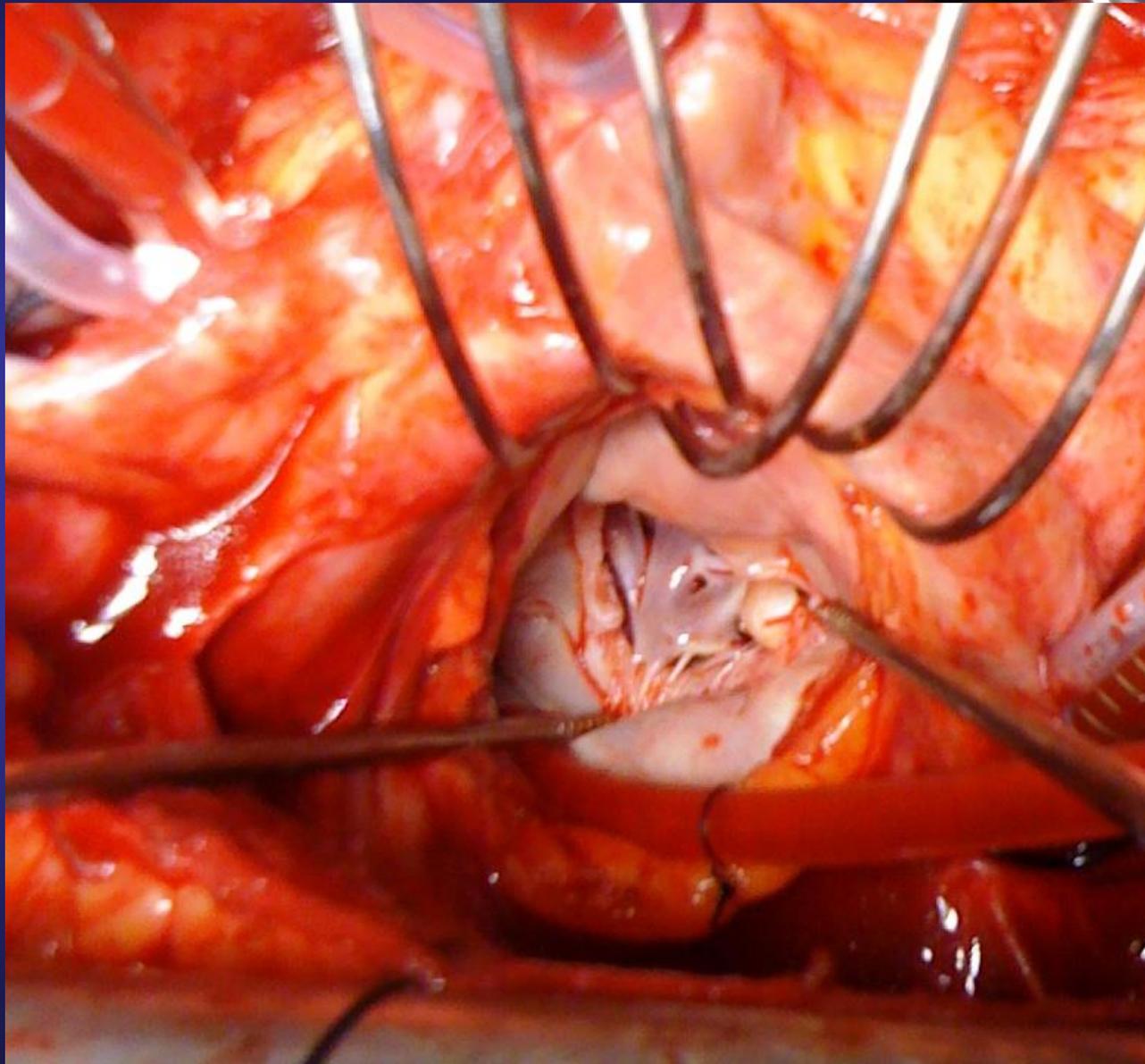
Ann Thorac Surg 2006;82:1103-1106

A**B****C****D****E****F**

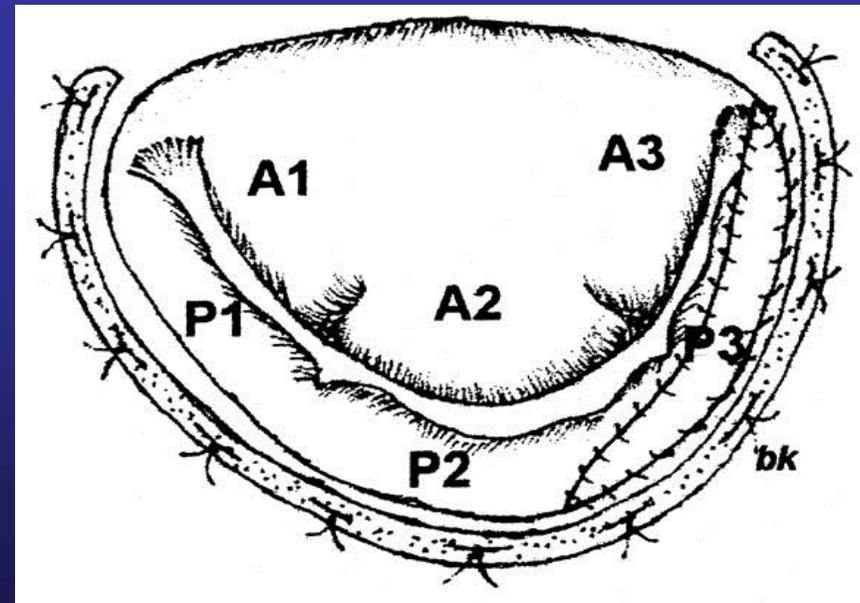
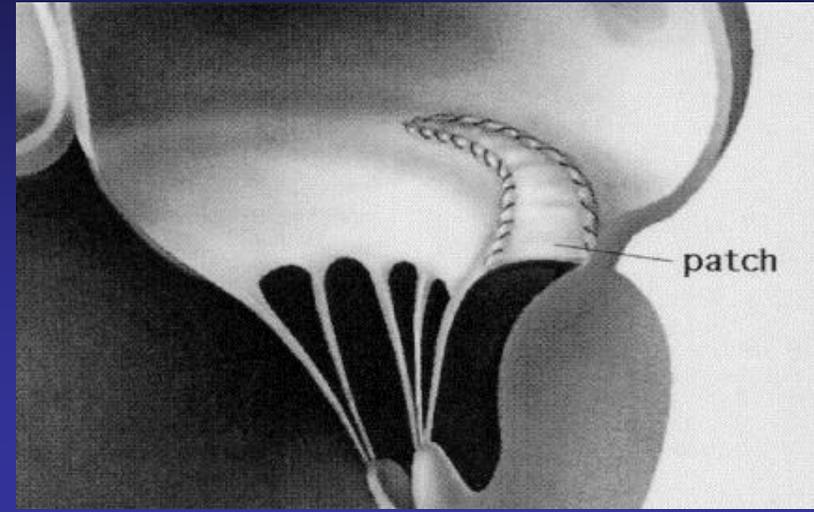
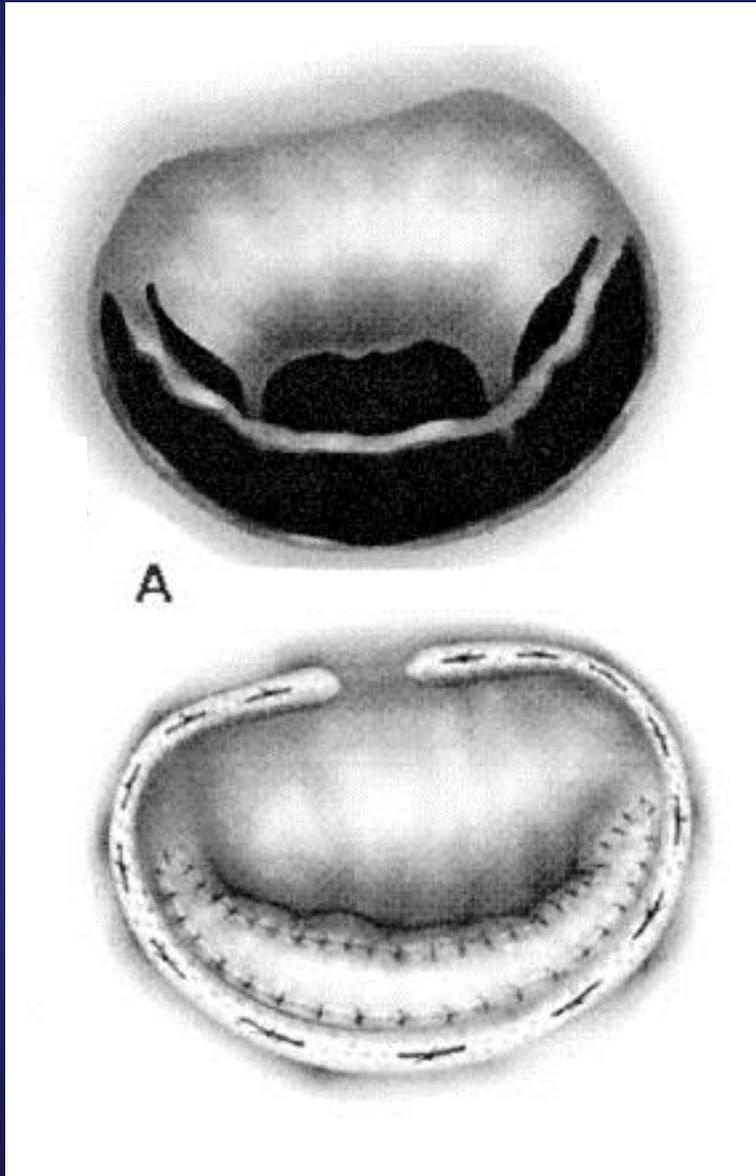








TECNICA QUIRURGICA



DOBRE

RESULTADOS

1995-2011

- 15 pacientes 9M/6H
- Edad media 57 a. (5d – 80a) NYHA III FE VI 52%
- Tipo de reparación:
 - Ampliación de velo posterior 12 80%
 - División m. papilares 2 13%
 - Punto de Alfieri 1 6%
- Anillo mitral 13/15 86%
 - Anillo Carpentier 10 66%
 - Anillo Kalangos 3 20%
- T CEC 127' T ISQ 95'
- No mortalidad.
- 14/15 r. sinusal antiagregación Disgren

PARTIAL HAMMOCK VALVE

FOLLOW-UP

- Mean 42,8 meses (1m-16a) 52,5 pa.año
- Mortalidad tardía 4 no cardiaca
- Insuficiencia mitral
 - 0 4 pa.
 - I 9 pa.
 - II 2 pa.

CONCLUSIONES

- La válvula mitral en hamaca parcial es una nueva lesión congénita que se manifiesta en edad pediátrica y adulta
- Lesión infrecuente aunque pasa desapercibida, a menudo etiquetada de “fibrosis de velo posterior” de causa incierta
- El diagnóstico es intraoperatorio. El ECO 3D puede ser útil para identificar los m. papilares
- Sospecharla si hay retracción en sístole de P2 en ausencia de cardiopatía isquémica o reumática
- Puede ser reparada con resultados fiables y reproducibles
- Cardiopatías congénitas del adulto