



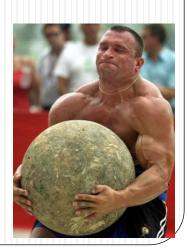
# Ventrículo derecho sistémico

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Unitat Integrada de Cardiopaties Congènites de l'Adolescent i l'Adult Vall d'Hebrón-Sant Pau

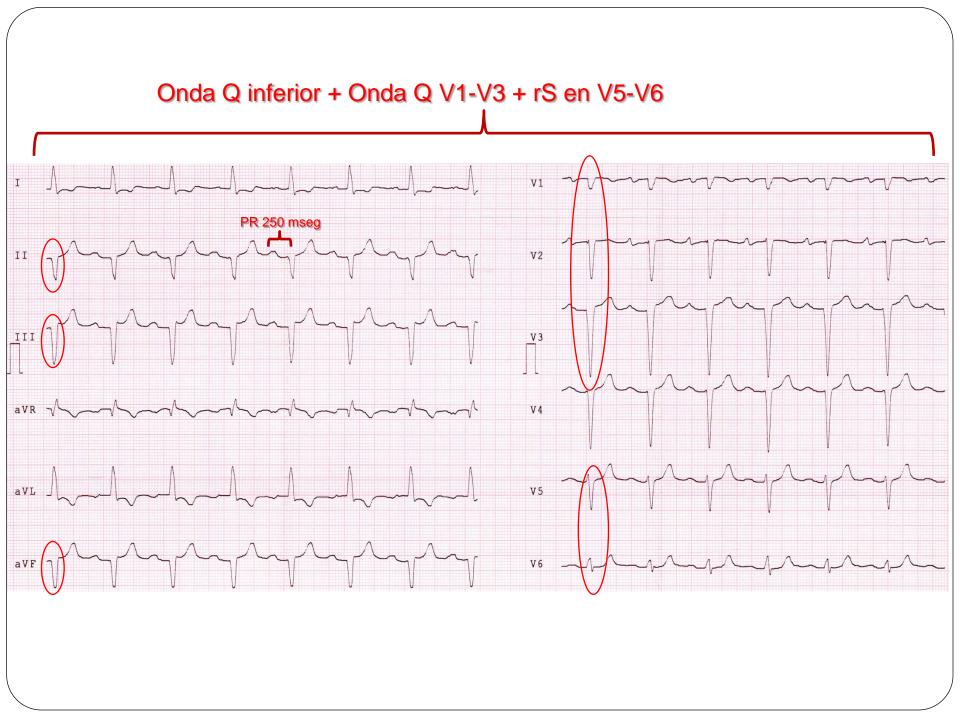
3 Novembre 2014



Mujer con fecha nacimiento: 18-ag-1965

### \*Visita UCCA 13/10/2011 > 46 años

- Dislipemia. HTA (tto Enalapril 5 mg/12 h)
- Enfermedad de Graves (tto con Tirodril)
- ✓ Asintomática cardiovascular
- ✓ Exploración: Peso: 66,5 Kgs. Talla: 176 cm. TA 105/70 mmHg. Sat O2 98%. Buen aspecto. Tolera decúbito. AC: RCR. Soplo pansistólico en ápex y mesocardio, llegando hasta foco pulmonar, en donde adquiere carácter eyectivo. No signos de ICC. Pulsos pedios presentes y simétricos
- ✓ Análisis: sin nada a destacar



# ¿Cuál es el diagnóstico?

A. Necrosis septal e inferior + BAV de primer grado, en una mujer de 46 años con FRCV: HTA y DL

(esta doctora se ha equivocado con la charla...)

B. Malposición de electrodos

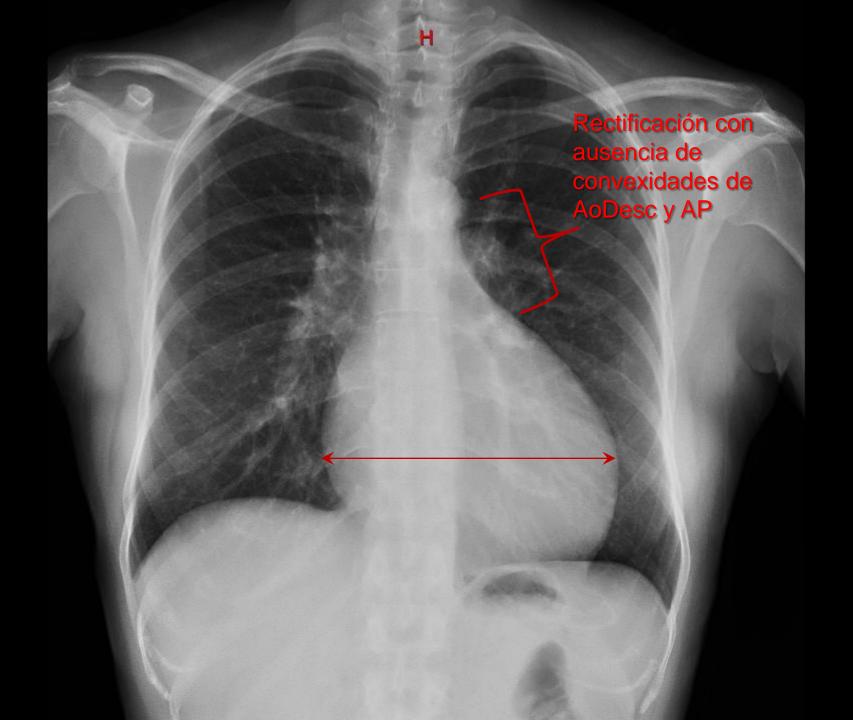
(la enfermera se ha equivocado con los electrodos...)

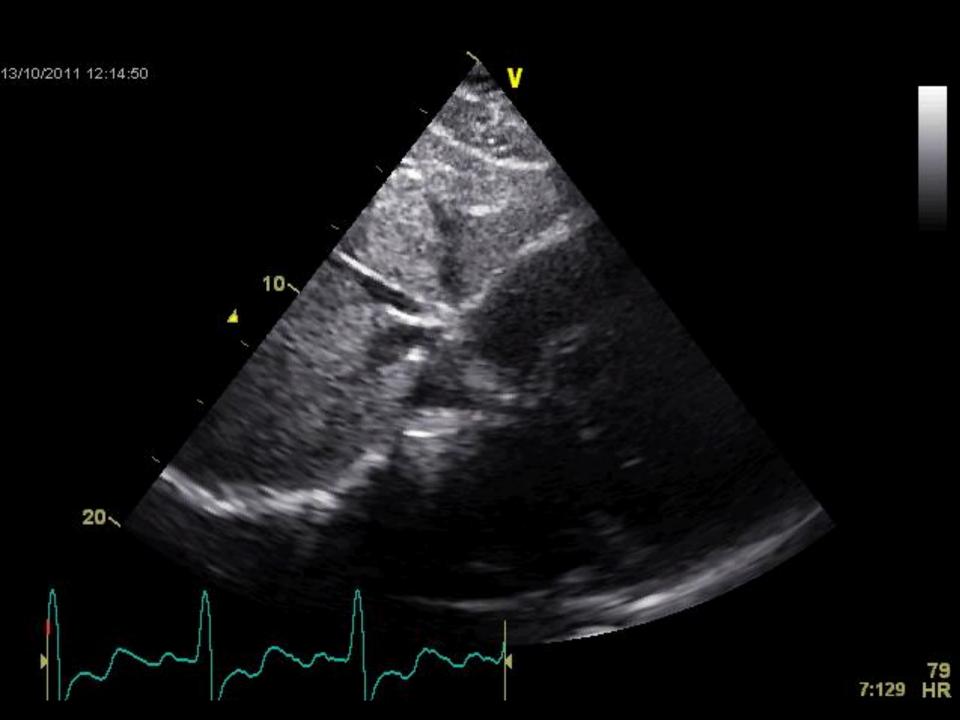
C. El ECG es de otro paciente

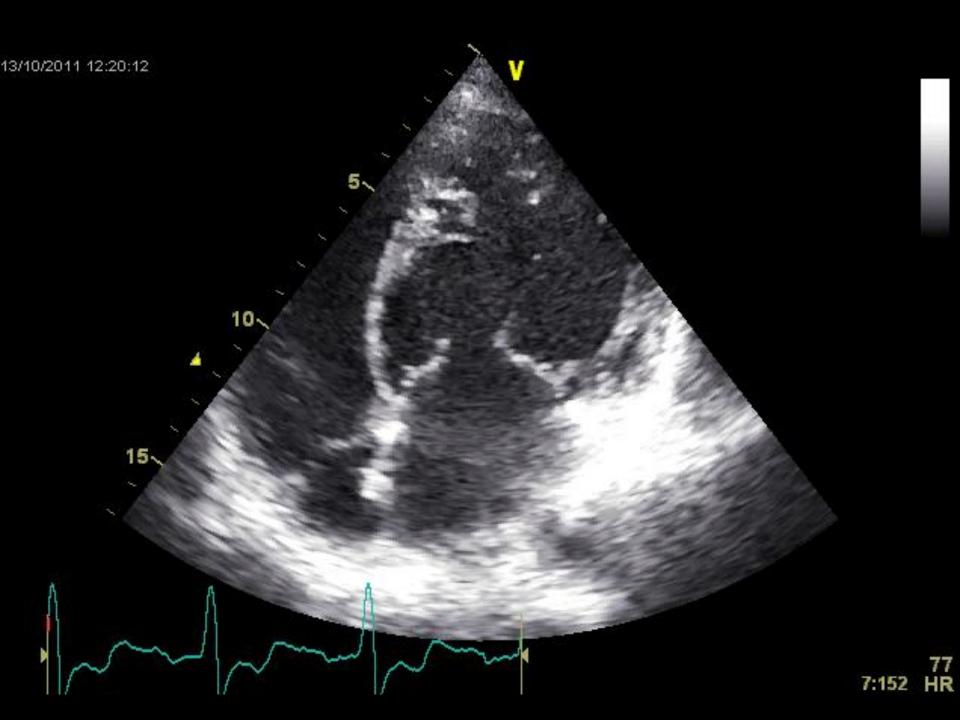
(la enfermera se ha equivocado con los nombres...)
esta enfermera no da una...
y la doctora tampoco...

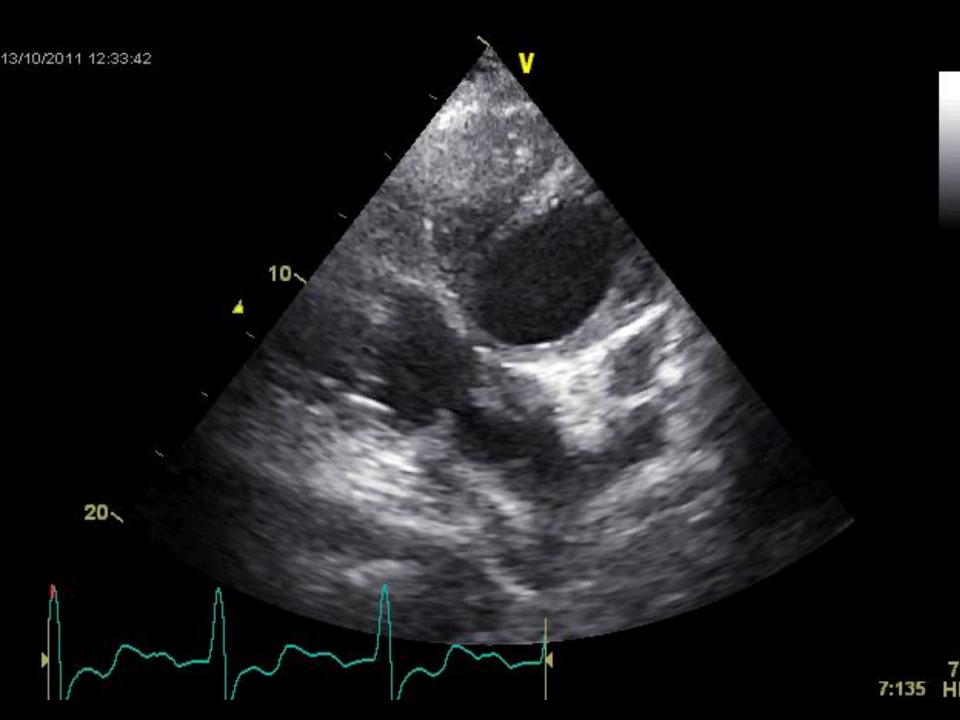


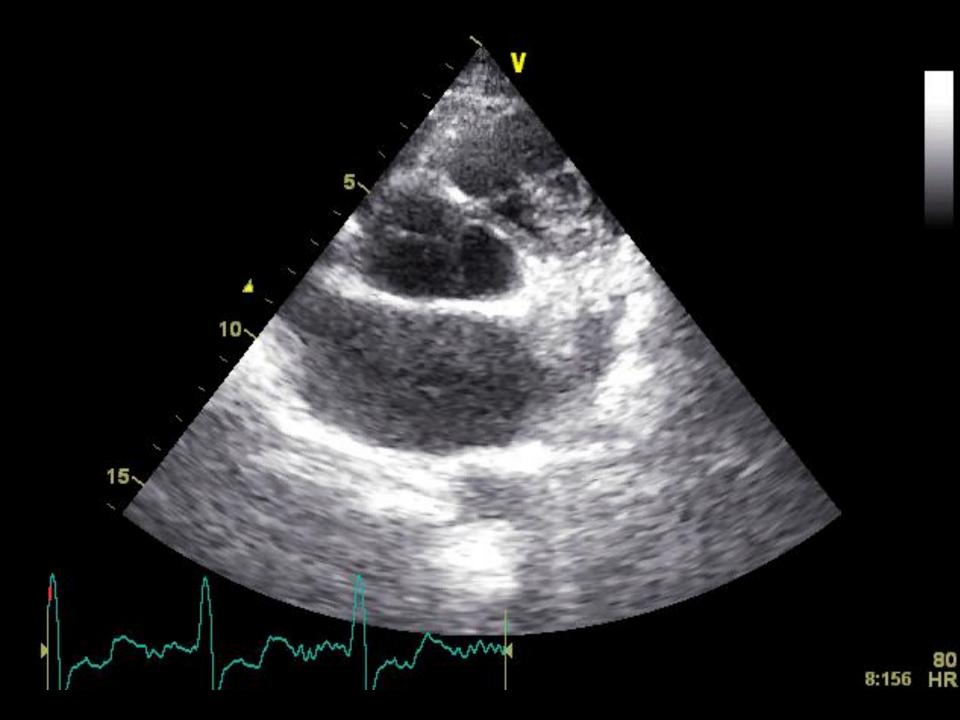
D. ¡No tengo ni idea! Necesito más datos..

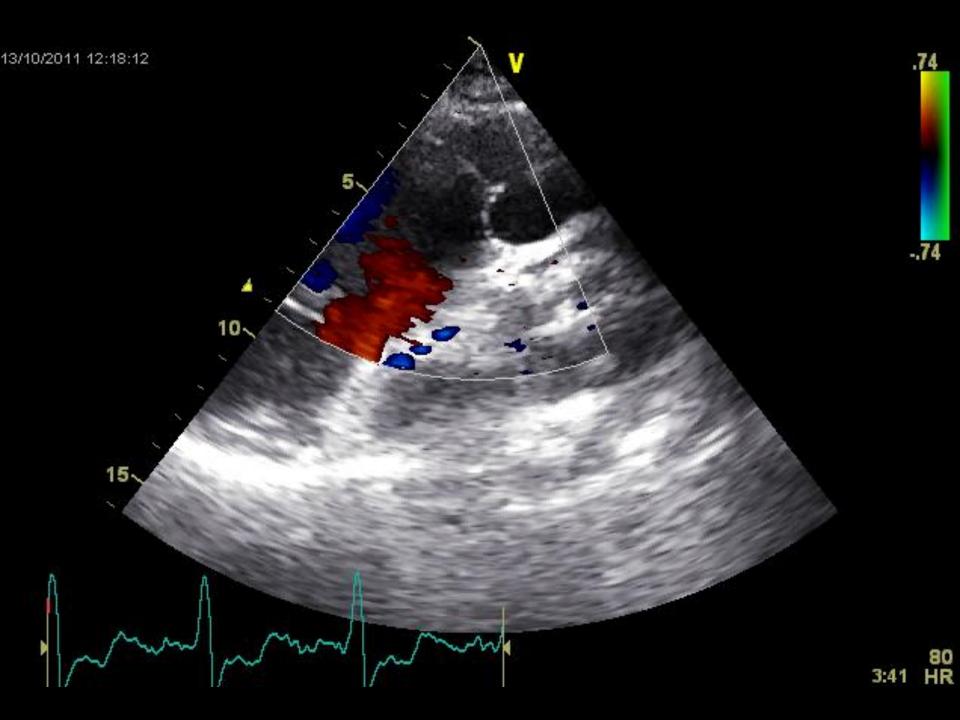


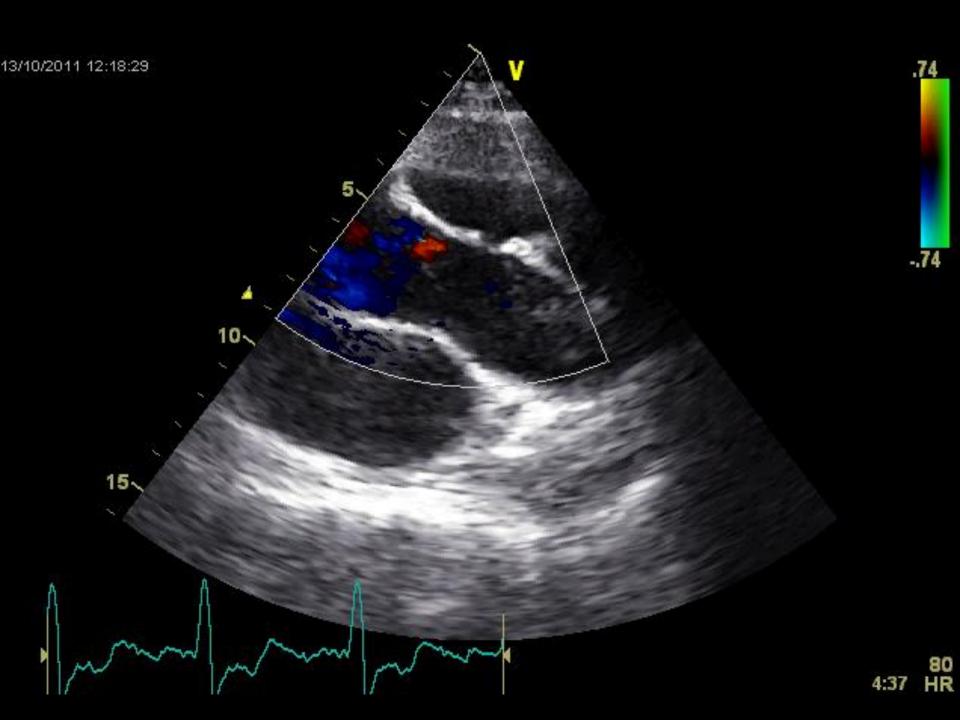






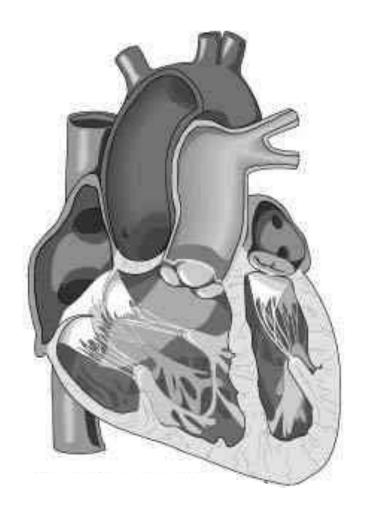


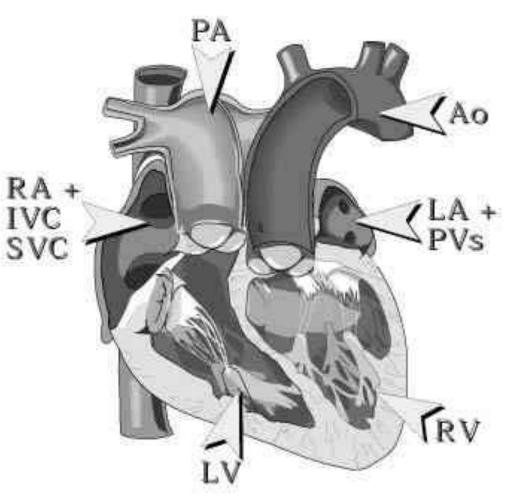






# **TGAcc**





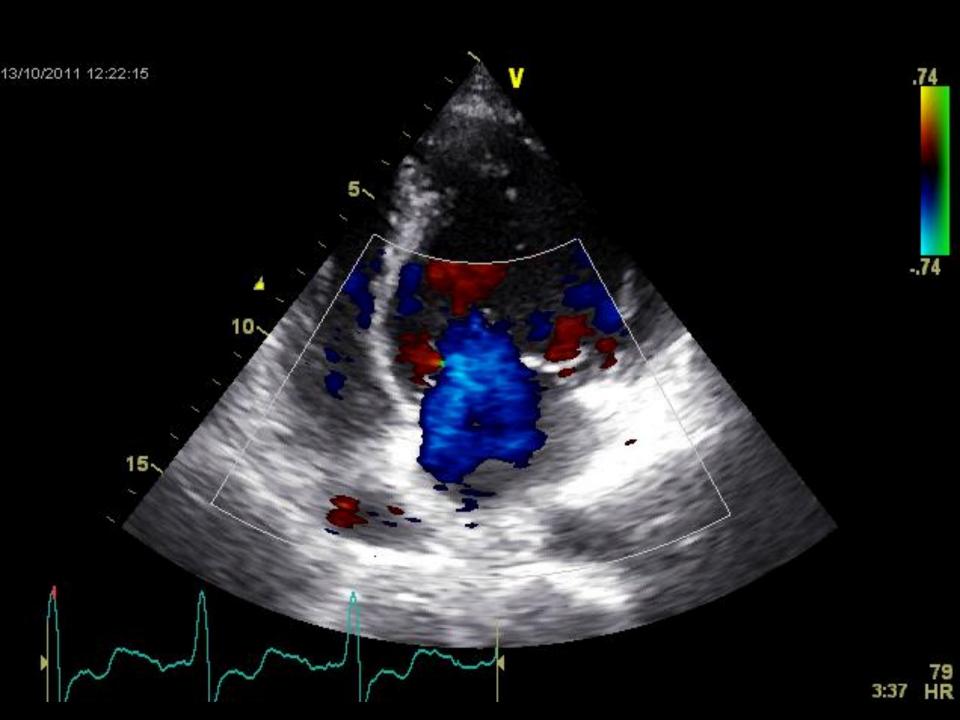
# Nuestra paciente...

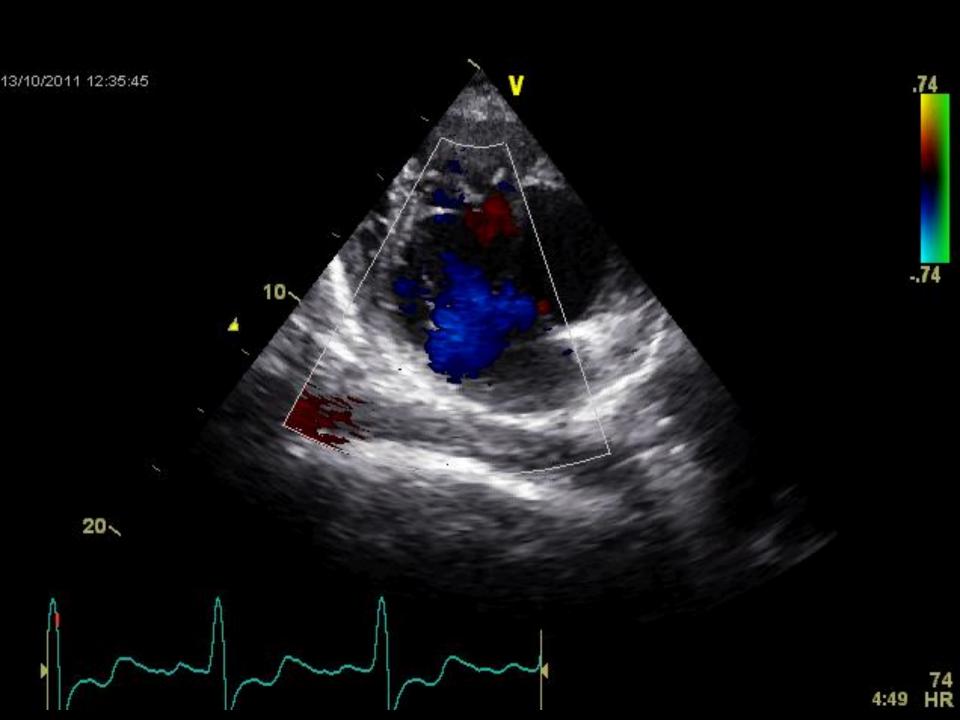
- Diagnóstico de CC acianótica e ICC a los 3 meses de edad,
   coincidiendo con infección respiratoria digoxina
- A los 29 meses de edad: Se orientó como TGV congénitamente corregida, con CIV asociada.
  - -No se detectaron signos de ICC. Sat. arterial de O2 normal.
- -El diagnóstico se confirmó por cateterismo cardiaco a los 6 años de edad (23/6/1972), constatando, además:
  - Existencia de un FOP.
  - CIV con cortocircuito izquierda-derecha ligero-moderado (Qp/Qs de 1.48)

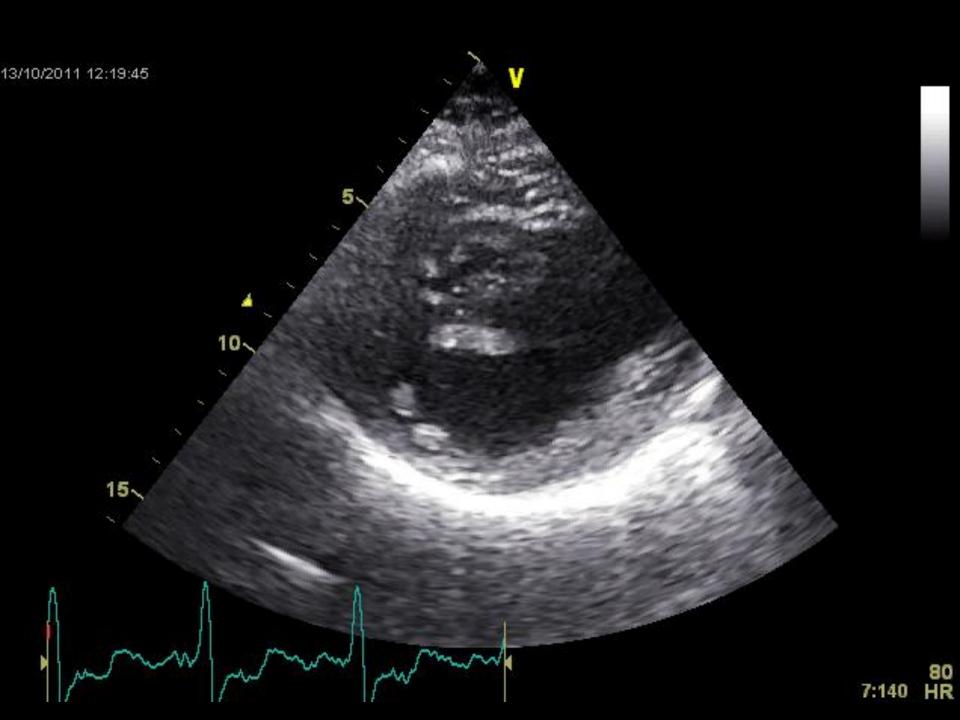
- Inicia con crisis de palpitaciones desde el 1997
  \*\*Holter: ESV frecuentes, generalmente aisladas, o en parejas, una salva regular de 4 latidos a 155 Ipm, y una salva de 28 latidos entre 125-155 Ipm muy sugestiva de ACXFA paroxística o TA multifocal.
  →Atenolol
- Persistencia de palpitaciones
   \*\*Holter (1999,2003) con registro de episodios de FA paroxística y TPSV por TA ectópica
- → Atenolol → Sotalol → Amiodarona (en el 2003 hipertiroidismo secundario) → Flecainida + Atenolol sin palpitaciones ni paroxismos de FA

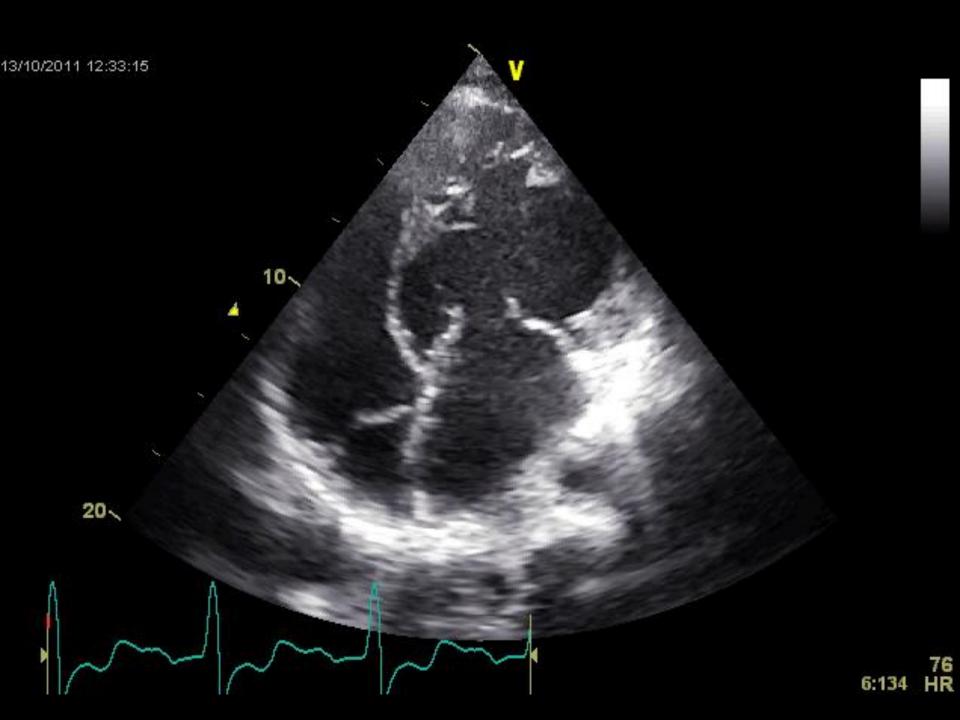
• Desde el 2/7/1984 se detecta una regurgitación de la válvula A-V sistémica, que se cataloga de severa en los últimos años.

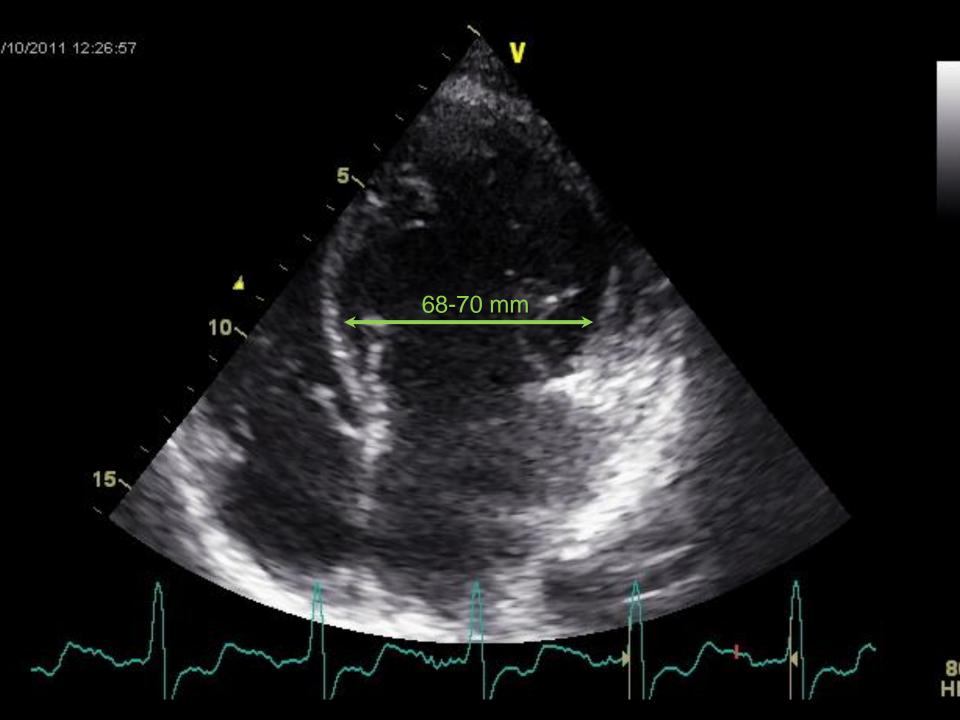
• Desde el punto de vista clínico la paciente se mantiene en buena clase funcional, refrendada por ergometrías con consumo de gases.

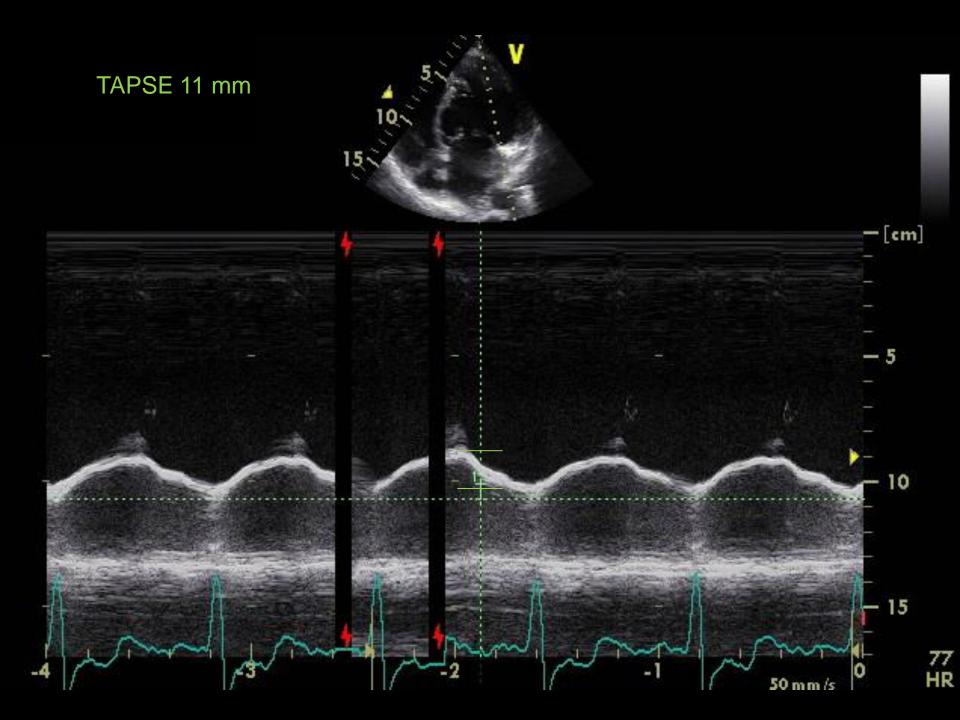


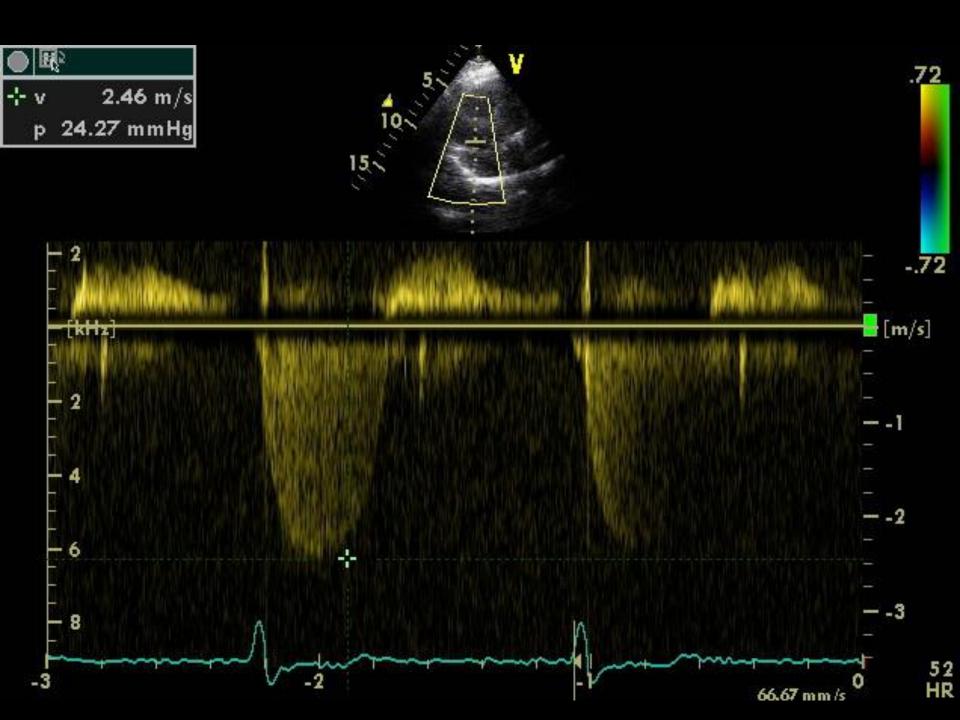


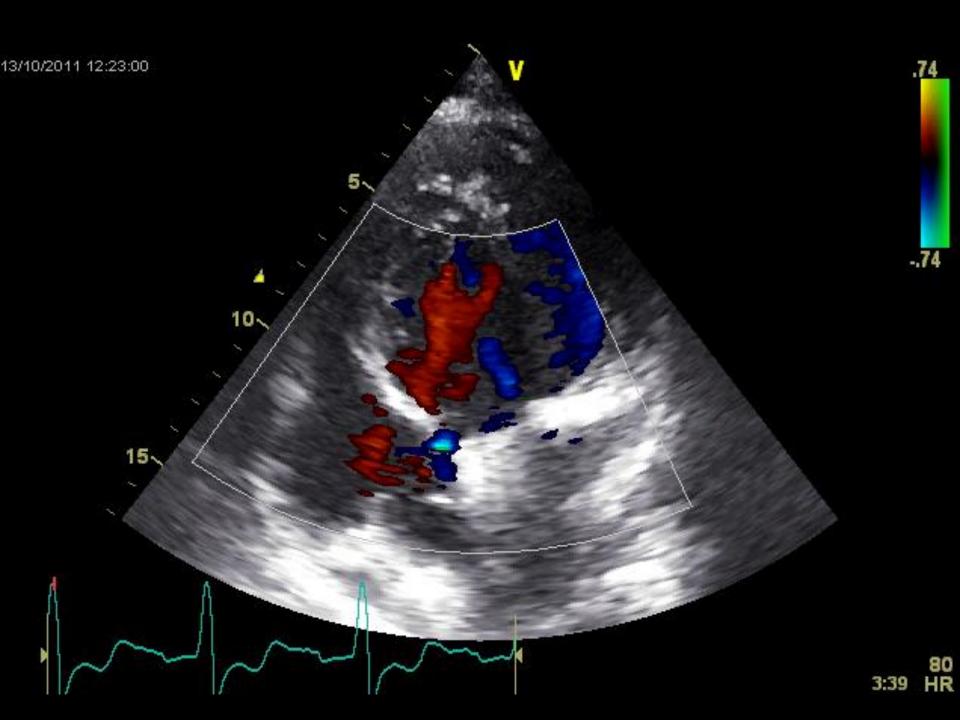


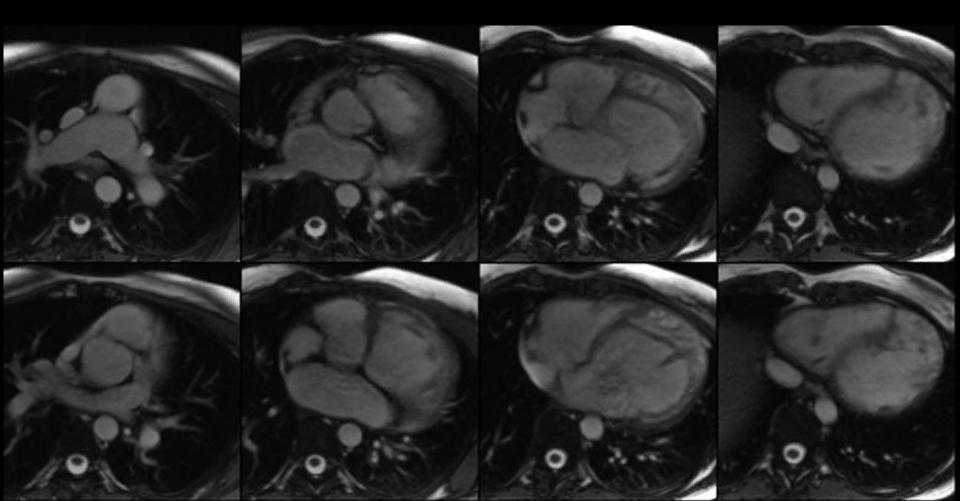


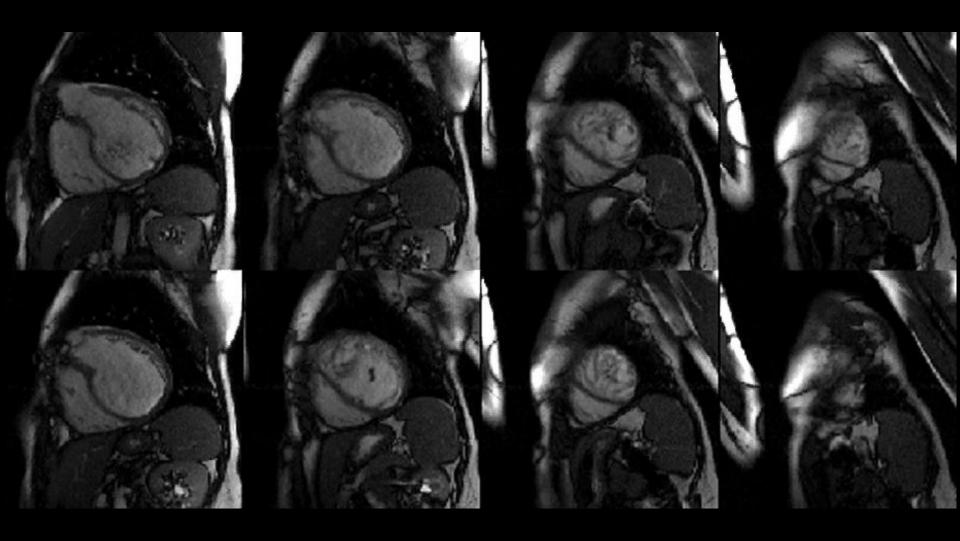












	VTD VD	FEVD	Qp/Qs	FRg AV
2003	308 ml	50%	1.88	26%
2007	262 ml (154 ml/m2)	57%	1.6	48%
2009	293 ml (162 ml(m2)	57%	1.7	45%
2011	310 ml (151 ml/m2)	52%	1.83	60%
2012	188 ml/m2	41%	1.4	41%

- ✓ Ventrículo derecho sistémico: FE 41% (VTD: 188 ml/m2 y VTS: 110 ml/m2).
- ✓ Ventrículo izquierdo subpulmonar FE 55% (VTD: 66 ml/m2 y VTS: 29 ml/m2).

Los estudios evolutivos de función del ventrículo derecho sistémico por RMN han mostrado una estabilidad de la fracción de eyección entorno a 50-55% pero el control de 2012 mostró ya un deterioro de la función sistólica hasta FE 41%

# Y, ¿ahora qué hacemos?...

A. Reparación fisiológica:

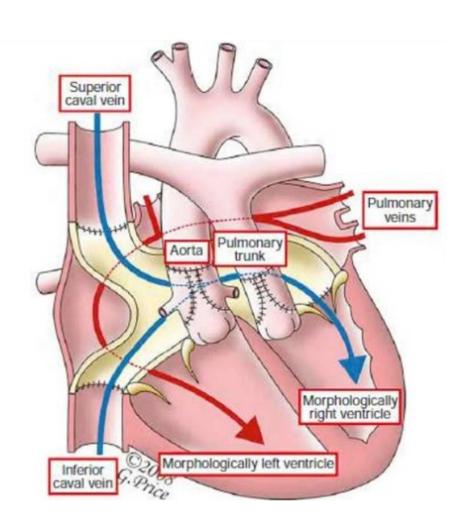
Cirugía sobra la válvula AV sistémica (VT) + IQ CIV

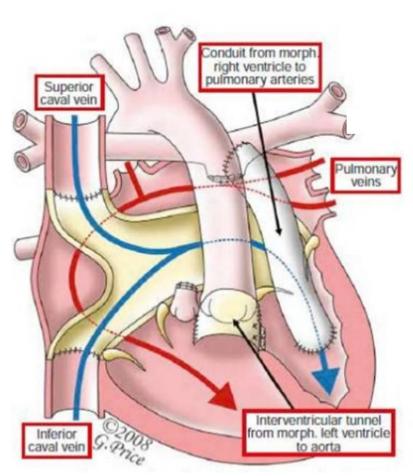


- B. Reparación anatómica
  - 1) Doble recambio: Switch auricular (senning/mustard)+ switch arterial (Jatene)
  - Switch auricular + Rastelli+/- IQ CIV... +/-banding pulmonar... +/-Glenn...
- C. TRC +/- DAI
- D. Tratamiento médico y rezar.. Pero rezar mucho...

# Switch auricular + Jatene

# Switch auricular + Rastelli





### **Doble Switch**



EUROPEAN JOURNAL OF CARDIO-THORACIC SURGERY

European Journal of Cardio-thoracic Surgery 35 (2009) 879-884

www.elsevier.com/locate/ejcts

# Results of the double switch operation for congenitally corrected transposition of the great arteries\*

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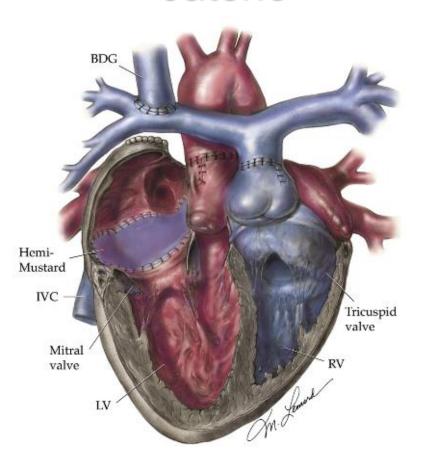
Received 3 September 2008; received in revised form 1 January 2009; accepted 3 January 2009

#### Abstract

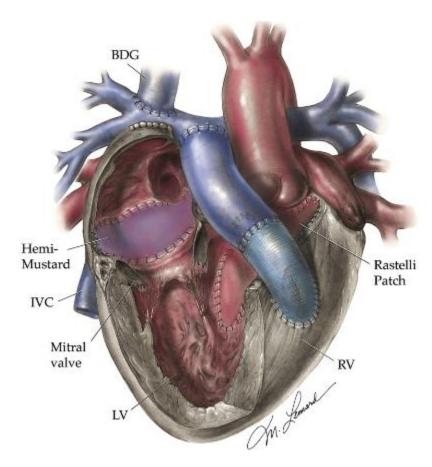
Background: Congenitally corrected TGA (CC-TGA) is characterized by discordant atrioventricular and ventricula arterial connections. In absence of right ventricular outflow tract obstruction (RVOTO), repair by atrial and arterial switches remains a challenging procedure for which long term follow-up is uncertain. Methods: From 1995 to 2007, 20 patients (median age: 26 months) with CC-TGA had double switch procedure. Segmental anatomy was {SLL} in all patients, dextrocardia in two patients, mesocardia in two patients. Ventricular septal defect was present in 17 patients, aortic coarctation in 2 patients and interrupted aortic arch (IAoA) in 1 patient. Five patients had tricuspid valve regurgitation. Six patients had AV blocks, 4 patients had pacemaker implantation prior to repair. Pulmonary artery banding was performed in 17 patients, for congestive heart failure (14 patients) or left ventricular retraining (3 patients). Three patients, including one patient with IAoA had primary repair. After LV retraining, repair was performed when indexed LV mass to LV volume ratio was above 1.5. A median follow-up of 60 months was achieved in all. Results: There were no deaths. Postoperative pacemaker implantation was required in four patients. Reoperation for Senning obstruction was necessary in one patient, and pacemaker battery replacement in another patient. One patient had mild neoaortic insufficiency, two had mild tricuspid regurgitation and two had mild mitral regurgitation. All were in NYHA I—II. Actuarial survival at 10 years was 100% and freedom from reoperation at 5 and 10 years were 93% and 77.4%, respectively. Conclusion: Double switch for CC-TGA without RVOTO can be performed with no mortality and low morbidity. Since these results seem to last for several years, it should be considered as the optimal procedure.

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# Hemi-Mustard + Glenn bidireccional + Jatene



# Hemi-Mustard + Glenn bidireccional + Rastelli



### Hemi-Mustard-Glenn bidireccional-Jatene/Rastelli

# The hemi-Mustard/bidirectional Glenn atrial switch procedure in the double-switch operation for congenitally corrected transposition of the great arteries: Rationale and midterm results

Sunil P. Malhotra, MD,<sup>a</sup> V. Mohan Reddy, MD,<sup>b</sup> Mary Qiu, BS,<sup>b</sup> Timothy J. Pirolli, MD,<sup>b</sup> Laura Barboza, BS,<sup>b</sup> Olaf Reinhartz, MD,<sup>b</sup> and Frank L. Hanley, MD<sup>b</sup>

Objective: This study was undertaken to assess the risks and benefits of the double-switch operation using a hemi-Mustard atrial switch procedure and the bidirectional Glenn operation for congenitally corrected transposition of the great arteries. To avoid complications associated with the complete Senning and Mustard procedures and to assist right-heart hemodynamics, we favor a modified atrial switch procedure, consisting of a hemi-Mustard procedure to baffle inferior vena caval return to the tricuspid valve in conjunction with a bidirectional Glenn operation.

Methods: Between January 1994 and September 2009, anatomic repair was achieved in 48 patients. The Rastelli-atrial switch procedure was performed in 25 patients with pulmonary atresia and the arterial-atrial switch procedure was performed in 23 patients. A hemi-Mustard procedure was the atrial switch procedure for 70% (33/48) of anatomic repairs.

Results: There was 1 in-hospital death after anatomic repair. There were no late deaths or transplantation. At a median follow-up of 59.2 months, 43 of 47 survivors are in New York Heart Association class I. Bidirectional Glenn operation complications were uncommon (2/33), limited to the perioperative period, and seen in patients less than 4 months of age. Atrial baffle–related reoperations or sinus node dysfunction have not been observed. Tricuspid regurgitation decreased from a mean grade of 2.3 to 1.2 after repair (P = .00002). Right ventricle–pulmonary artery conduit longevity is significantly improved.

Conclusions: We describe a 15-year experience with the double-switch operation using a modified atrial switch procedure with favorable midterm results. The risks of the hemi-mustard and bidirectional Glenn operation are minimal and are limited to a well-defined patient subset. The benefits include prolonged conduit life, reduced baffle- and sinus node-related complications, and technical simplicity. (J Thorac Cardiovasc Surg 2011;141:162-70)

Pacing and Cardiac Resynchronization Therapy



### Effects of cardiac resynchronization therapy on echocardiographic indices, functional capacity, and clinical outcomes of patients with a systemic right ventricle

Gaël Jauvert<sup>1,2\*</sup>, Juliette Rousseau-Paziaud<sup>3</sup>, Elisabeth Villain<sup>2</sup>, Laurence Iserin<sup>2</sup>, Françoise Hidden-Lucet<sup>4</sup>, Magalie Ladouceur<sup>2</sup>, and Daniel Sidi<sup>2</sup>

<sup>1</sup>Clinique Bizet, Paris, France; <sup>2</sup>Hôpital Necker, Paris, France; <sup>3</sup>Centre Cardiologique du Nord, Saint Denis, France; and <sup>4</sup>Hôpital de la Pitié Salpétrière, Paris, France

Received 22 September 2008; accepted after revision 29 October 2008; online publish-ahead-of-print 26 November 2008

#### Aims

Surgically (SC) or congenitally corrected (CC) transposition of the great arteries (TGA), associated with a systemic right ventricle (RV), is often complicated by heart failure. This retrospective study assessed the functional and mechanical effects of cardiac resynchronization therapy (CRT) in patients presenting with TGA.

### Methods and results

Seven patients with SC (n=5) or CC (n=2) TGA (mean age 24.6  $\pm$  12 years), a failing systemic RV, and intraventricular dyssynchrony, underwent implantation of a CRT-P. Permanent pacemakers were previously implanted in five patients. The leads were implanted by a combined transvenous and epicardial approach in the five patients with SC TGA. Echocardiography, including tissue Doppler imaging and cardiopulmonary exercise testing were performed before and during CRT. Since, in all patients, ventricular dyssynchrony was due to delayed septal wall contraction, the interventricular septum and RV free wall were stimulated synchronously, with a view to resynchronize a maximum amount of myocardium. After  $19.4 \pm 8.1$  months of CRT, mean QRS duration decreased from  $160 \pm 31$  to  $120 \pm 28$  ms (P=0.03), intraventricular delay from  $104 \pm 27$  to  $14 \pm 15$  ms (P=0.01), New York Heart Association functional class from 3.0 to 1.57 (P=0.01), and peak oxygen consumption increased from  $13.8 \pm 2.5$  to  $22.8 \pm 6.7$  mL/kg/min (P=0.03). One patient died suddenly at 23 months of follow-up.

#### Conclusions

CRT was technically feasible and associated with improvements in cardiac mechanical function and clinical status in patients with TGA, failing systemic RV, and intraventricular dyssynchrony.

# ESC Guidelines for the management of grown-up congenital heart disease. 2010

# **Table 18** Indications for intervention in congenitally corrected transposition of the great arteries

Indications	Classª	Level
Systemic AV valve (tricuspid valve) surgery for severe regurgitation should be considered before systemic (subaortic) ventricular function deteriorates (before RVEF <45%)	lla	С
Anatomic repair (atrial switch + arterial switch or Rastelli when feasible in case of non-restrictive VSD) may be considered when LV is functioning at systemic pressure	IIb	С

## IQ sobre la Válvula AV sistémica (VT)

### Cardiovascular Surgery

# Tricuspid Valve Surgery in Adults With a Dysfunctional Systemic Right Ventricle Repair or Replace?

Roderick W.C. Scherptong, MD; Hubert W. Vliegen, MD, PhD; Michiel M. Winter, MD; Eduard R. Holman, MD, PhD; Barbara J.M. Mulder, MD, PhD; Ernst E. van der Wall, MD, PhD; Mark G. Hazekamp, MD, PhD

Background—In patients with a right ventricle (RV) in the systemic position, tricuspid valve surgery for regurgitation beyond adolescence is a subject of debate. The aim of the present study was to evaluate the complications, survival, and benefit of tricuspid surgery in adult patients with an atrium-level correction for transposition of the great arteries or congenitally corrected transposition of the great arteries.

Methods and Results—All adult patients (n=16; 7 men, 9 women; age 35±11 years) who underwent tricuspid valvuloplasty (n=8) or replacement (n=8) in the period 1999 to 2008 were included. Complications and survival were analyzed, and postoperative changes in RV function and functional class were evaluated. Tricuspid regurgitation was graded 1 to 4 according to its severity, RV dysfunction was graded as 1 to 4 (1=no dysfunction to 4=severe dysfunction), and functional status was determined according to New York Heart Association class. Although complications occurred in 11 patients, all could be managed adequately. Three patients died 109, 180, and 659 days after surgery, respectively, the first patient after tricuspid valve replacement and the latter 2 after tricuspid valvuloplasty. Overall, tricuspid valve function improved (from grade 3.1±0.8 to 0.9±1.0; P=0.001) and functional class improved (from 2.7±0.6 to 2.1±0.8; P=0.007), whereas RV function remained unchanged. After tricuspid valvuloplasty, however, recurrent moderate tricuspid valve regurgitation was observed frequently (n=3; 37%).

Conclusions—Mortality is rather low after tricuspid surgery in adult patients with mild to moderate RV dysfunction. In general, tricuspid valve function and functional class improve significantly after surgery, and systemic RV function is preserved. Tricuspid valvuloplasty, however, is associated with a high rate of recurrence of regurgitation. (Circulation. 2009;119:1467-1472.)

## IQ sobre la Válvula AV sistémica (VT)

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#### **Congenital Heart Disease**

# Congenitally Corrected Transposition of the Great Arteries

Ventricular Function at the Time of Systemic Atrioventricular Valve Replacement Predicts Long-Term Ventricular Function

François-Pierre Mongeon, MD,\* Heidi M. Connolly, MD,\* Joseph A. Dearani, MD,† Zhuo Li, MS,‡ Carole A. Warnes, MD\*

Rochester, Minnesota

0				

The objective was to evaluate the systemic ventricular ejection fraction (SVEF) at the time of systemic atrioventricular valve (SAVV) replacement as a predictor of SVEF ≥1 year after surgery in patients with congenitally corrected transposition of the great arteries (CCTGA).

#### Background

Progressive SAVV regurgitation causes systemic ventricular failure in CCTGA patients, who are commonly referred late for intervention. Survival after surgery is poor when the pre-operative SVEF is <44%.

#### Methods

We retrospectively reviewed 46 patients (pre-operative SVEF  $\geq$ 40% in 27 patients and  $\leq$ 40% in 19 patients) with 2 good-sized ventricles, a morphologically right systemic ventricle, and SAVV regurgitation requiring surgery. Median follow-up was not different in patients with a pre-operative SVEF  $\geq$ 40% (8.8 years) or  $\leq$ 40% (7.7 years, p = 0.36).

#### Results

Pre-operative SVEF was the only independent predictor of ≥1-year post-operative SVEF (p < 0.0001). The late SVEF was preserved (defined as ≥40%) in 63% of patients who underwent surgery with an SVEF ≥40% compared with 10.5% of patients who underwent surgery with an SVEF <40%. Pre-operative variables associated with late mortality were an SVEF ≤40%, a subpulmonary ventricular systolic pressure ≥50 mm Hg, atrial fibrillation, and New York Heart Association functional class III to IV.

#### Conclusions

Post-operative systemic ventricular function after SAVV replacement can be predicted from the pre-operative SVEF. For best results, operation should be considered at an earlier stage, before the SVEF falls below 40% and the subpulmonary ventricular systolic pressure rises above 50 mm Hg. (J Am Coll Cardiol 2011;57:2008–17) © 2011 by the American College of Cardiology Foundation

- > 26/07/2012: Sustitución de válvula AV sistémica por prótesis mecánica Sorin Bicarbon 29 mm y cierre de CIV subpulmonar
  - Hallazgos: CIV subpulmonar de 1 cm de diámetro. Dilatación del anillo AV sistémico severa.
  - Implante en posición intraanular de prótesis mecánica de 29mm mediante puntos sueltos en U de Ticron apoyados en teflon de 7mm, respetando los velos de la válvula nativa. Cierre del septo y atriotomía.

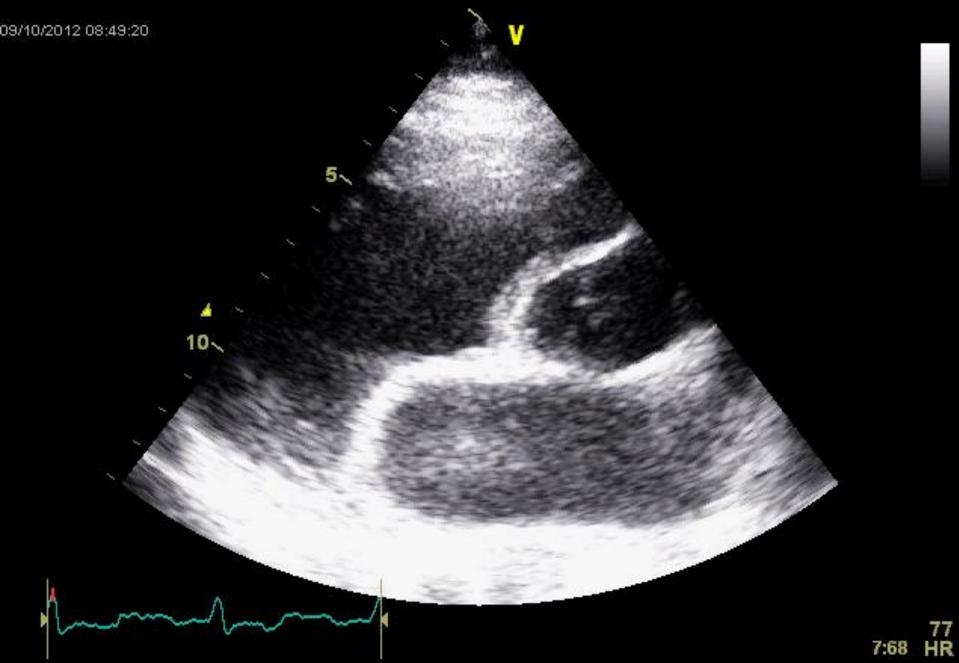
#### Incidencias:

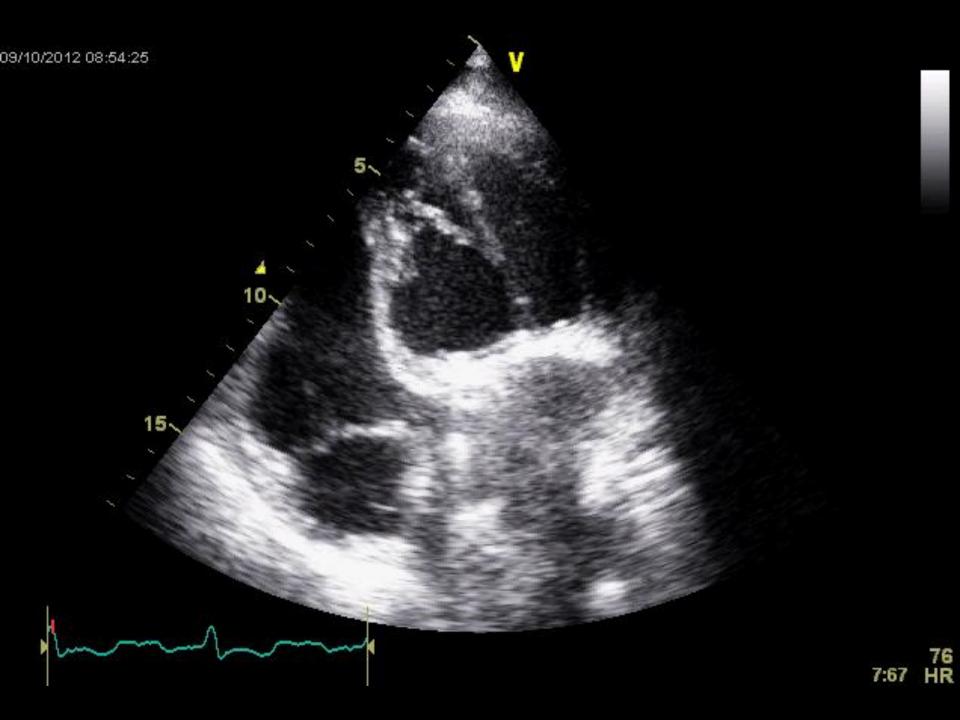
- FAR revertida con flecainida
- pericarditis postpericardiotomía buena respuesta a ibuprofeno.

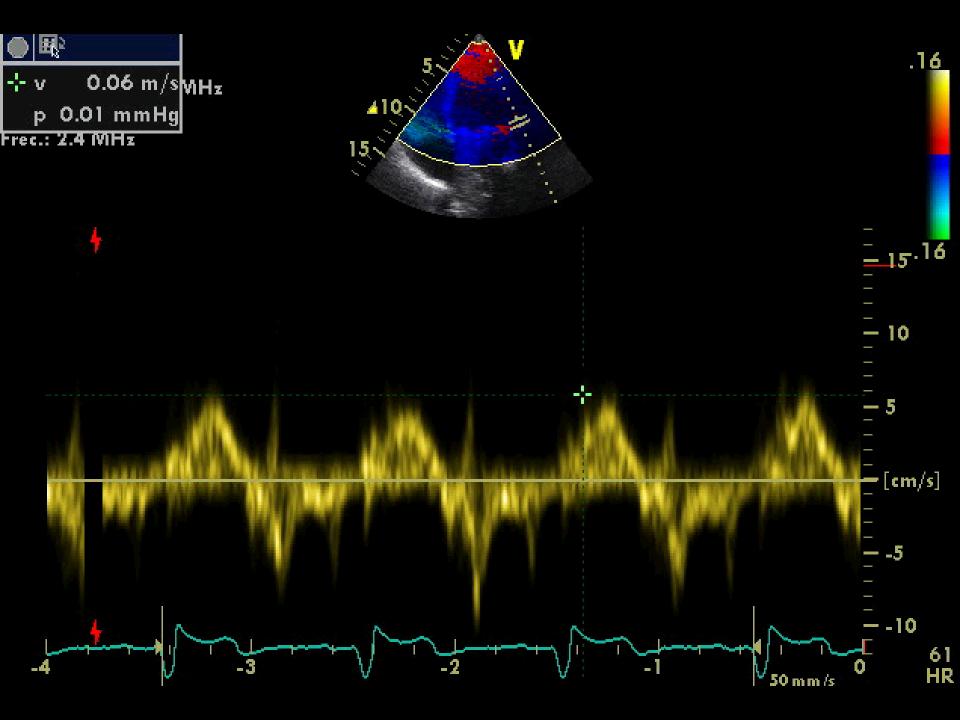
### • ETT prealta:

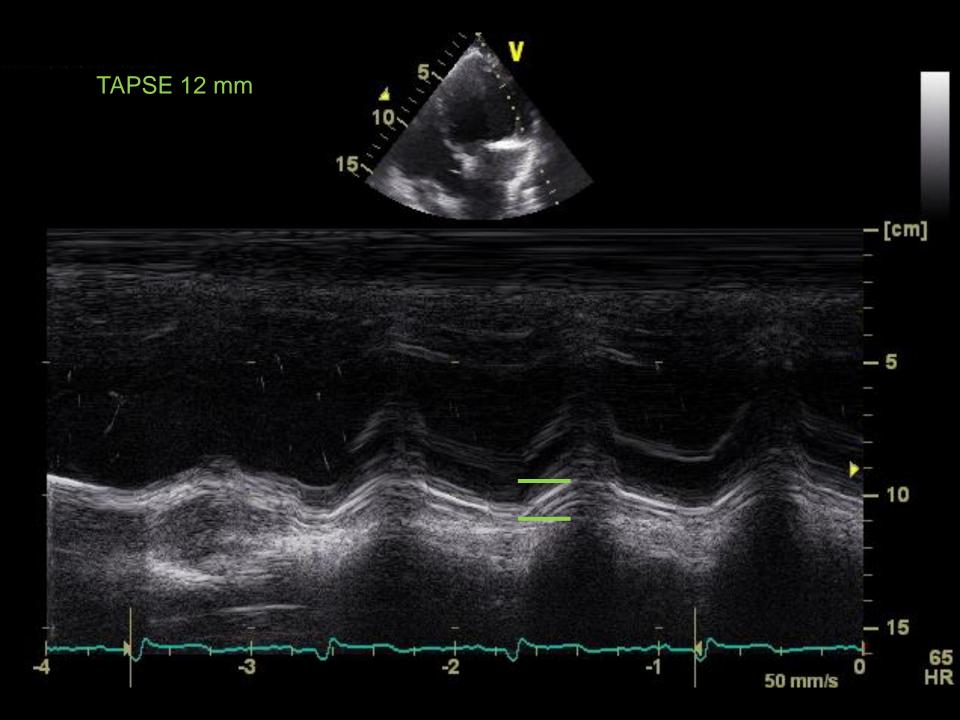
- Prótesis mecánica normofuncionante
- Ausencia de derrame pericárdico
- Ausencia de CIV residual
- FEVD sistémico moderadamente deprimida.
- Tratamiento al alta: Acenocumarol, Flecainida 100mg/12h, Atenolol 25mg/24h, Ibuprofeno pauta descendente, Omeprazol 20mg/24h

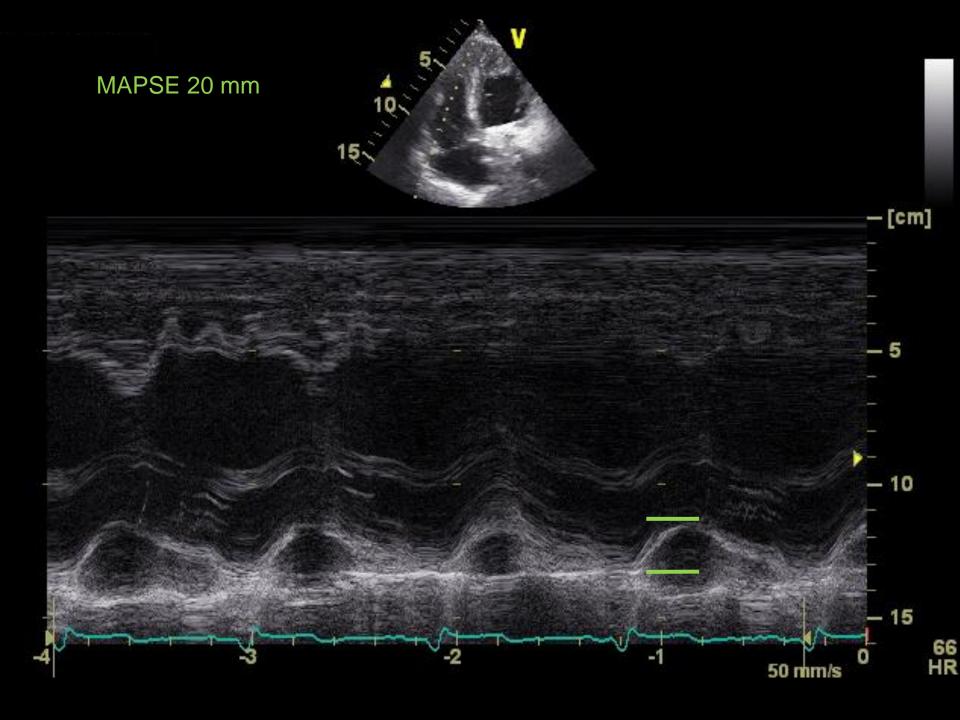


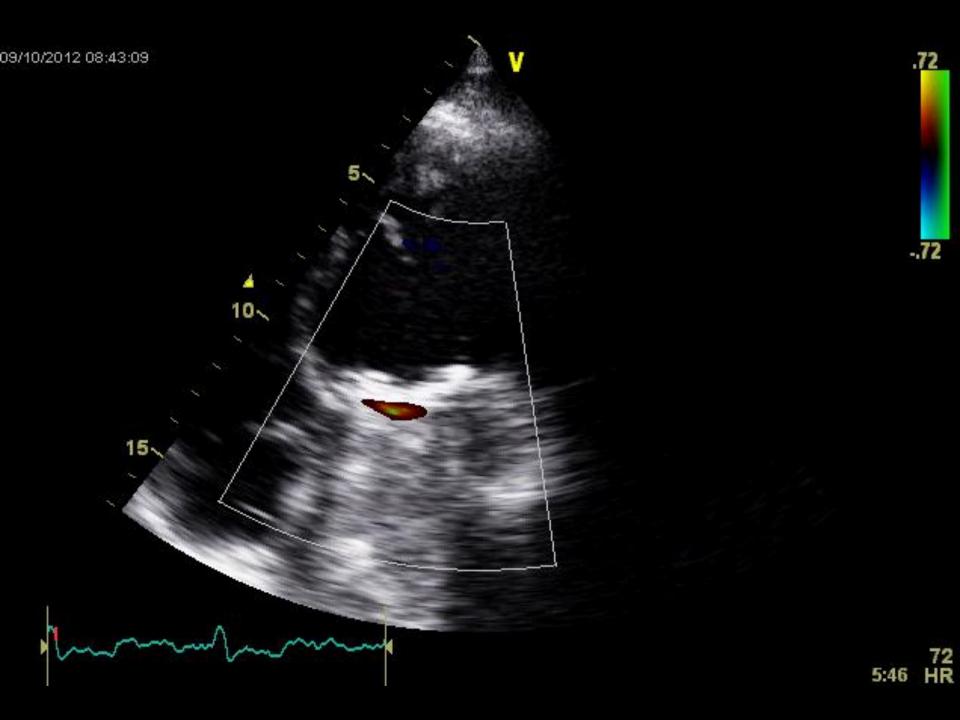


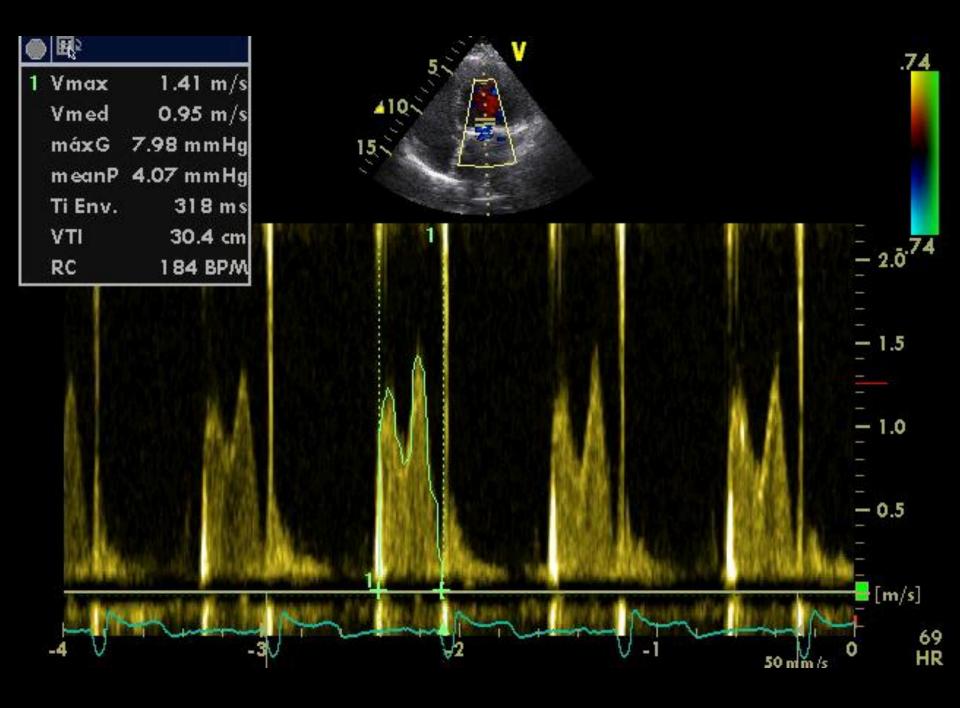


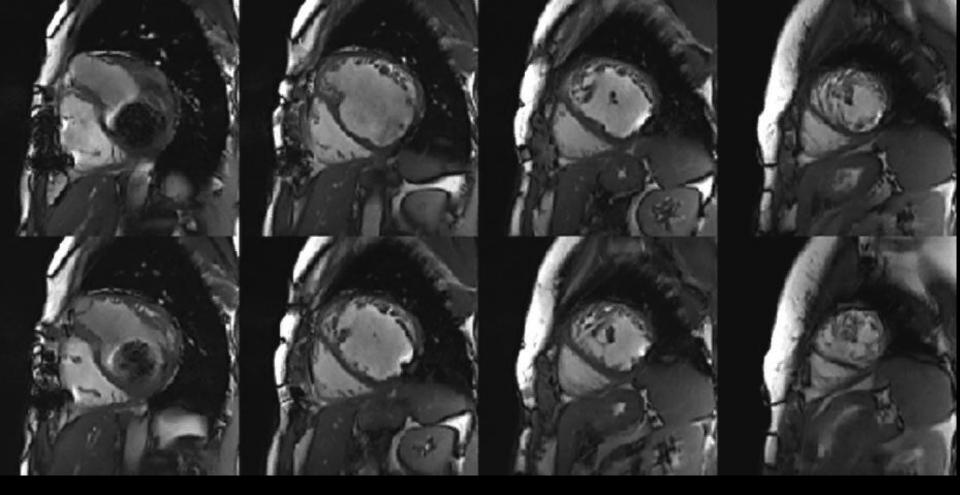








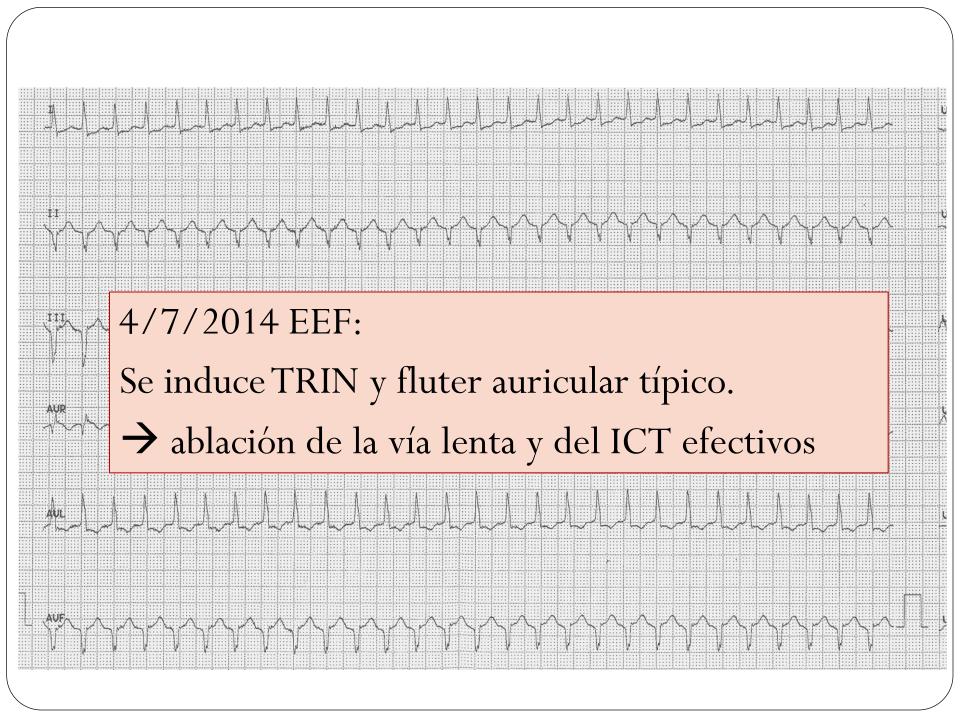




	VTD VD	VTSVD	FEVD	VTDVI	VTSVI	FEVI
2012 preoperatoria	188 ml/m2	110 ml/m2	41%	66 ml/m2	29 ml/m2	60 %
2012 postoperatoria	123 ml/m2	65 ml/m2	47 %	47 ml/m2	16 ml/m2	66 %

### Seguimiento

- Sin síntomas ni signos de IC
- Tratamiento: Enalapril 5 mg/12 h, Flecainida 100 mg/12 h, Atenolol 25 mg/12 horas, Acenocumarol
- ECG: sin cambios.
- Holter sin bloqueo AV ni pausas significativas. ESV y EV poco frecuentes



# ccTGA = discordancia AV y VA = L-TGA = doble discordancia = aislada inversión ventricular

- <1% del total de las cardiopatías congénitas
- Los ventrículos están invertidos en comparación con la situación normal, la Ao surge anterior del VD (en el lado izquierdo) y la AP nace posterior, desde el VI (en el lado derecho).
- Puede existir isomerismo auricular y la orientación cardiaca puede incluir levocardia, dextrocardia, o mesocardia.

- Lesiones asociadas frecuentes (80-90%):
  - CIV (70%) y EP (40%. Valvular y frecuentemente subvalvular).
  - Anomalías de la válvula tricúspide sistémica ("Ebstein like")
  - Trastornos de la conducción (congénito —adquirido: incidencia BAVc 2%/año. Más frecuente tras IQ CIV- IQ VAV)
- > Historia natural: en dependencia de las lesiones asociadas...
  - CIV  $\rightarrow$  ICC
  - CIV + EP  $\rightarrow$  cianosis
  - Aislada -> frecuentemente asintomáticos hasta la edad adulta
  - Disfunción V. sistémico (VD) Insuficiencia válvula AV sistémica (VT) (4<sup>a</sup>-5<sup>a</sup> década)
  - Arritmias supraventriculares (5ª-6ª década)
  - Arritmias ventriculares

Br Heart 7 1995;74:57-59

# **TGAcc**

### Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up

Patrizia Presbitero, Jane Somerville, Filippo Rabajoli, Susan Stone, Maria Rosa Conte

### Abstract

Objective-To assess the clinical course of adult patients with corrected transposition of the great arteries without associated anomalies.

Design-All patients with corrected transposition of the great arteries without associated anomalies were reviewed with complete clinical and echocardiographic assessment. The complications were evaluated in each decade.

Setting-Tertiary centre with a specific unit dealing with "grown-up" adolescent and adult congenital heart disease, designated as a quaternary centre and a general hospital with a referral centre for "grown-up" congenital heart disease.

Patients—18 patients (nine male and nine female) aged 16-61 years followed for 1-30 years (mean 10 years).

Results-There were no deaths. Six patients had a worsening ability index during follow up. Complications were: (a) complete heart block in seven, three of whom required pacemaker insertion; (b) significant left atrioventricular valve regurgitation in 50%, appearing only in the third decade (12%), with increasing frequency thereafter. Infective endocarditis was responsible for increasing left atrioventricular valve regurgitation in only one patient; (c) supraventricular arrhythmia appeared in the fifth decade, and occurred in all patients over the age of 60 years. One patient aged 61 had recurrent sustained ventricular tachycardia; and (d) congestive heart failure developed only after 50 years in 66%. One patient had severe left atrioventricular valve regurgitation; the function of the systemic ventricle was only moderately reduced in the other three. Three of the nine women had seven uneventful preg-

Conclusions-Patients with corrected transposition of the great arteries without associated defects may remain undiagnosed until adult life. Symptoms occur rarely before the fourth and fifth decades, when rhythm disturbance, left atrioventricular valve regurgitation, and moderately impaired systemic ventricular function cause congestive cardiac failure. The role of pacemaker insertion or surgery for left atrioventricular valve regurgitation needs further assessment.

Keywords: corrected transposition of the great arteries without associated defects; natural history of corrected transposition of the great arteries; "grown-up" congenital heart disease.

Corrected transposition of the great arteries (atrioventricular discordance and ventriculoarterial discordance) as a lone anomaly is rare. Patients with this condition reach adult life without earlier diagnosis or recognition of having heart disease. Left atrioventricular valve regurgitation, complete heart block, and systemic ventricular dysfunction develop as the years advance. The incidence of such complications and the age at which they occur have not been completely documented. The aim of this survey was to illustrate the natural history of a group of patients with corrected transposition of the great arteries without associated defects.

### Patients and methods

Data were collected from two centres specialising in the care of patients with "grown up" congenital heart disease, namely, the Royal Brompton National Heart and Lung Hospital (RBH), London, United Kingdom, and the Hospital Giovanni Bosco (HGB), Turin, Italy. From this population data of patients with corrected transposition of the great arteries followed in adulthood were reviewed retro-

Selection criteria were: (a) age of patients 16 years or over at first presentation in the centre; (b) follow up of more than 1 year; and (c) diagnosis of corrected transposition of the great arteries without major associated defects confirmed by echocardiography or angiography, or both.

There were 18 patients (10 from the RBH and eight from the HGB). The mean (range) age at presentation was 35 (16-61) years and mean (range) follow up of the 18 patients was 10 (1-30) years. All patients were reviewed with complete clinical and echocardiographic assessment. The ability index (AI)1 was used for grading symptoms. The complications, evaluated decade by decade, were incidence of arrhythmia, complete heart block, cardiac failure, left atrioventricular valve regurgitation, and bacterial endocarditis.

The reasons for the initial referral of these patients to a cardiologist were (a) abnormal

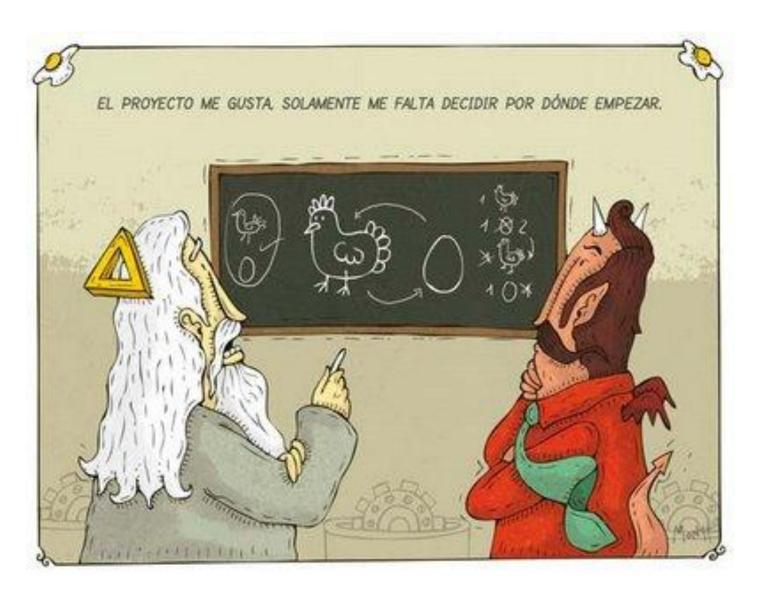
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¡Gracias!

