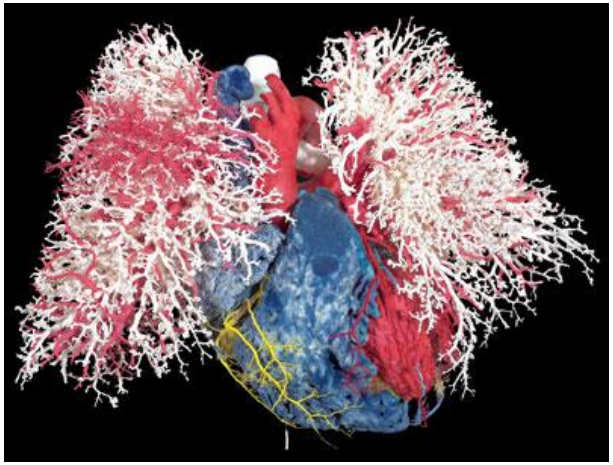
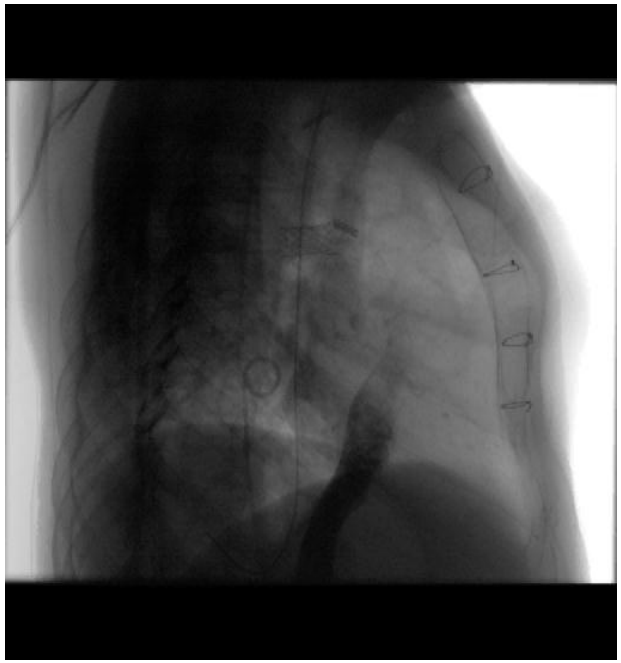
La Circulación de Fontan y la Presión Pulmonar.





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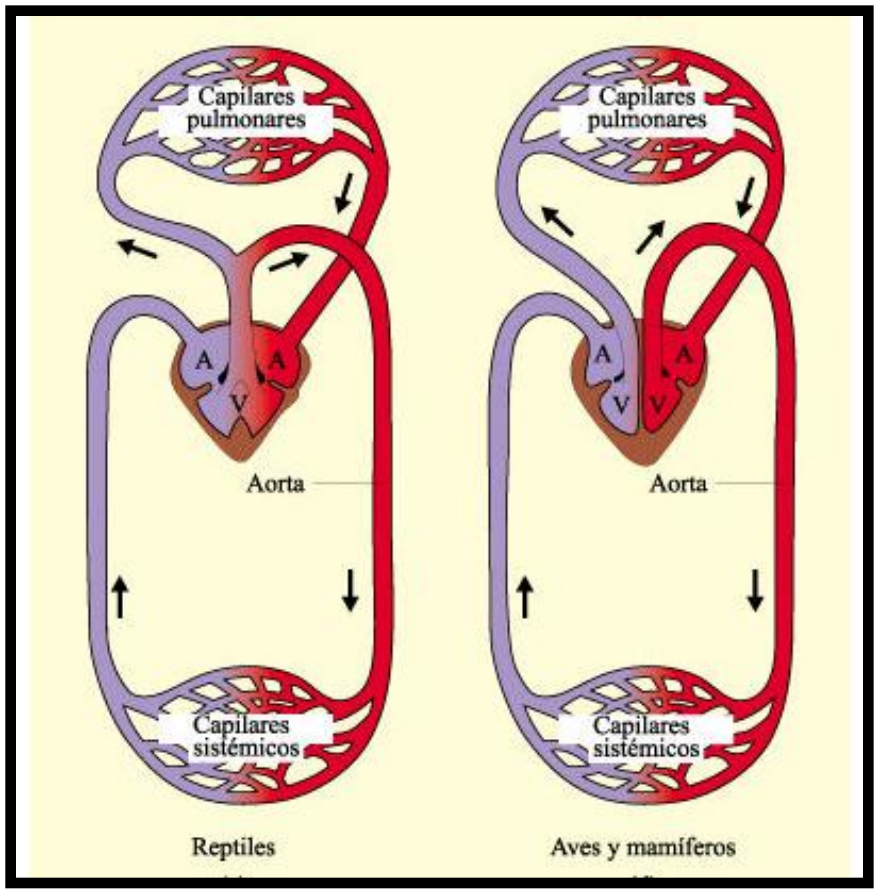
Hospital Universitario La Paz
SaludMadrid
Comunidad de Madrid

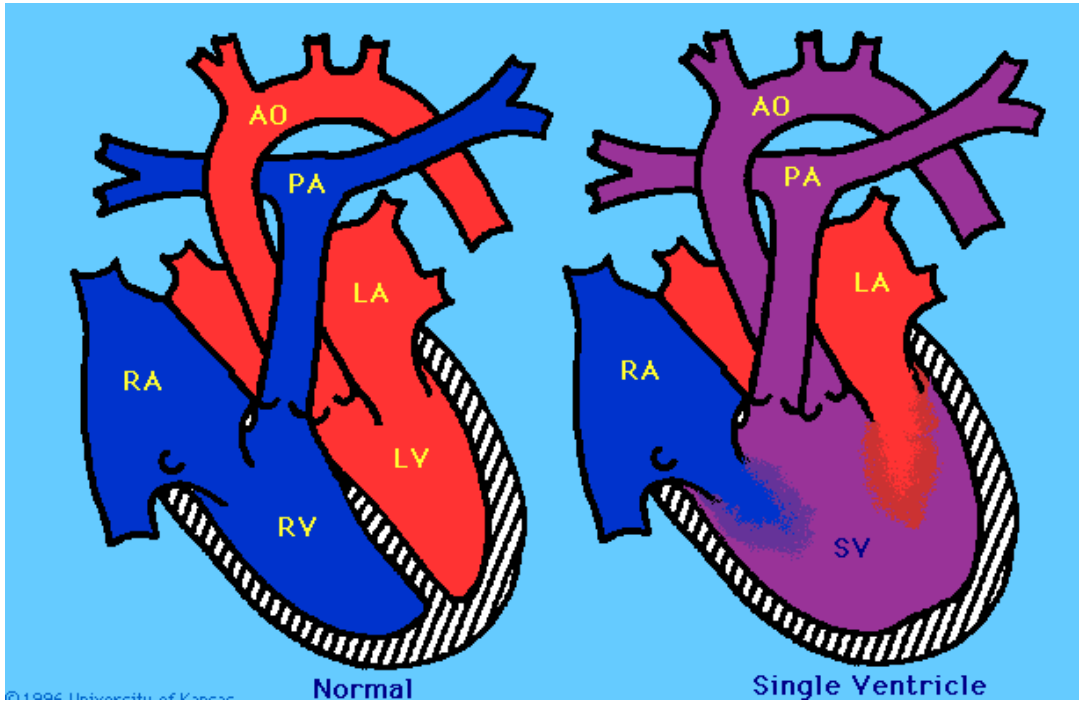
INTRODUCCIÓN

- Corazón univentricular: Circulaciones en paralelo
- Corazón biventricular: Circulaciones en serie



REPARACIÓN UNIVENTRICULAR: OBJETIVO

- Ante una situación funcional de Ventrículo Único (VU):
- Las técnicas quirúrgicas de reparación univentricular, hoy en día **neonatales en su 1er tiempo,** persiguen:
  - La supervivencia del paciente
  - Convertirlo en buen candidato al Fontan



- **FONTAN**→Tratamiento definitivo del VU que maneja el gasto sistémico; Separación completa de las circulaciones sistémica y pulmonar: el drenaje de las v. cavas va directamente al pulmón sin pasar por el corazón → Imprescindible PP baja

TÉCNICAS QUIRÚRGICAS I

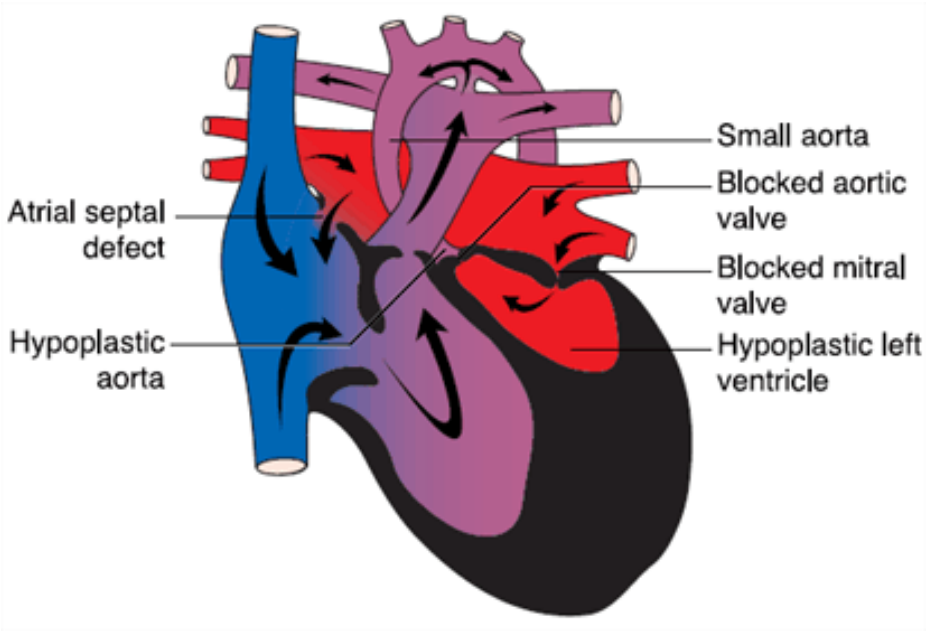
- La cirugía busca **minimizar los factores de riesgo** que acechan al paciente a lo largo de su trayectoria hasta alcanzar el Fontan:
  - Isquemia pulmonar→Fístulas sistémico pulmonares
  - CIA restrictiva → Apertura de CIA, Blalok-Hanlon
  - Sobrecarga de volumen→"Banding" del tronco de la arteria pulmonar

1945 1950 1952

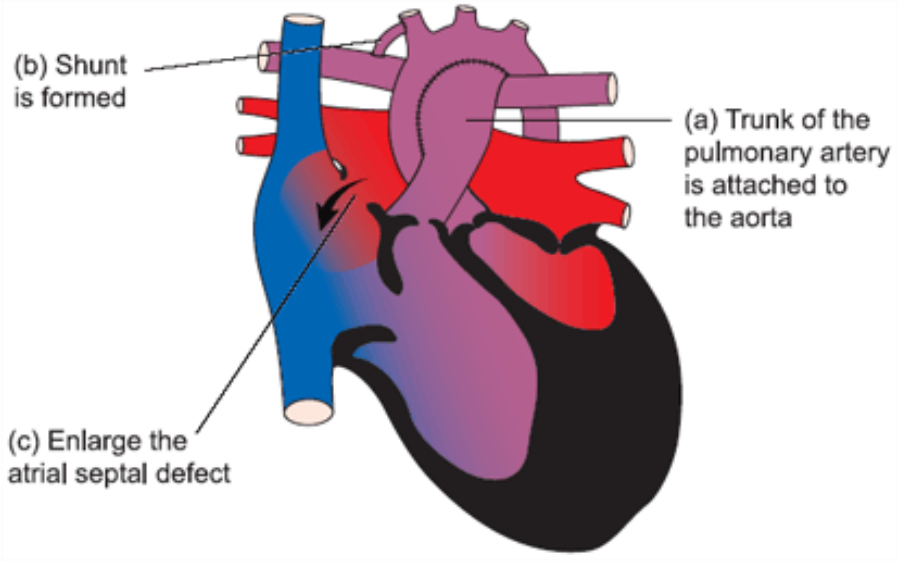
- En la situación de **palliación inicial** el objetivo será mantener ambas circulaciones en equilibrio: **Qp/Qs =1**

TÉCNICAS QUIRÚRGICAS II

- **Minimizar factores de riesgo** que acechan al paciente a lo largo de su vida hasta el Fontan:
  - Sobrecarga de presión por obstrucción a la vía de salida izquierda→ Stansel o resección de foramen bulbo ventricular restrictivo, reconstrucciones de arco aórtico, Norwood estadio I

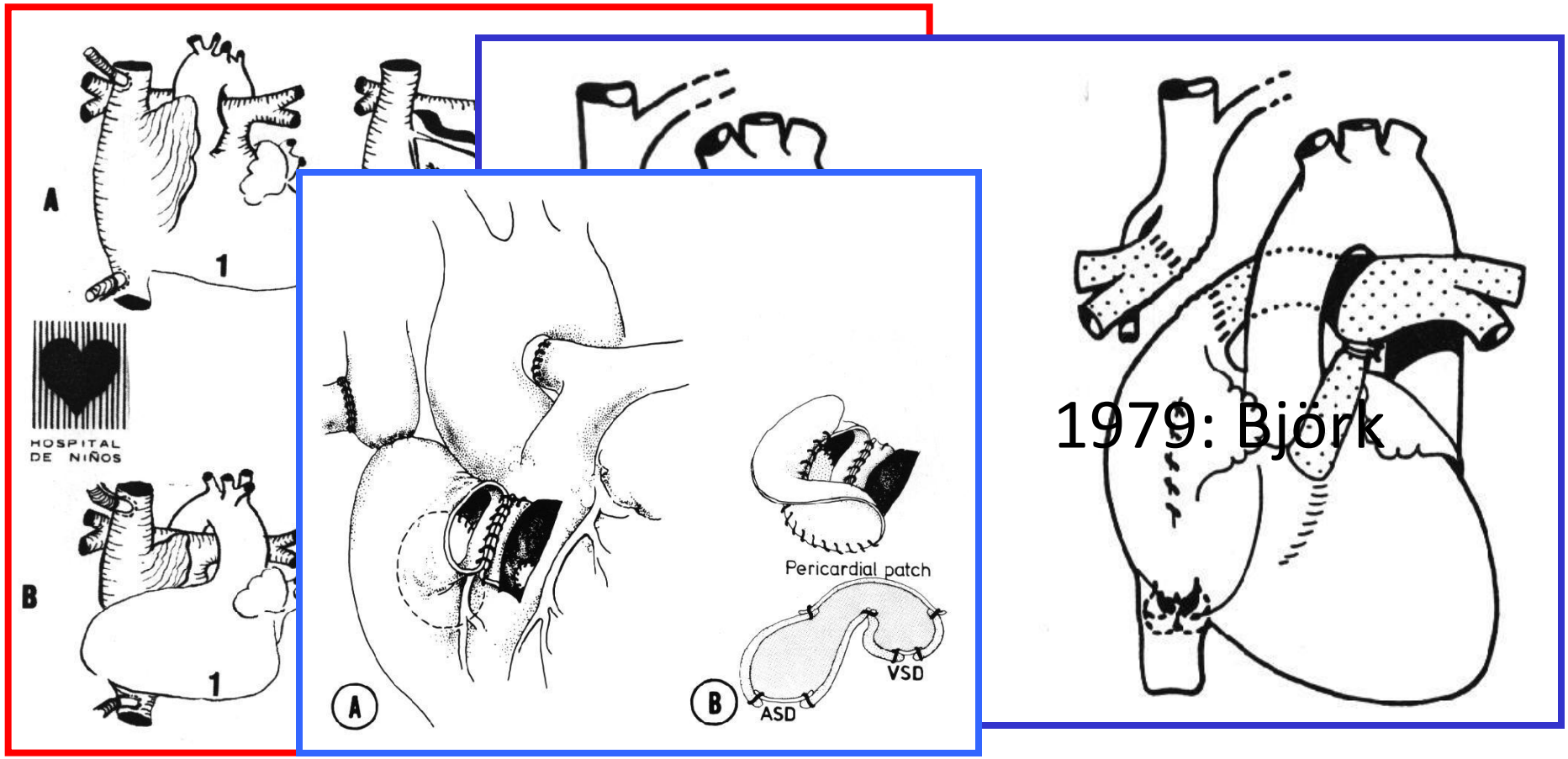


1981



TÉCNICAS QUIRÚRGICAS III

Fontan atriopulmonar: Conexión de AD con TP para q sangre de cavas vaya directa al pulmón + septación de la aurícula con parche para que sangre de VVPP drene hacia el VU



The Fontan/Kreutzer Procedure At 40: An Operation for the Correction of Tricuspid Atresia

Guillermo O. Kreutzer, MD,[a] Andrés J. Schlichter, MD,[b] and Christian Kreutzer, MD[c]

Pediatric Cardiac Surgery Annual

" Certainly, it is one of the triumphs of cardiac surgery in congenital heart disease".

It is clearly a matter of perspective
– is the glass partially empty or partially full?

Forty Years of The Fontan Operation: A Failed Strategy

Jack Rychik

"The Fontan/Kreutzer operation (results in profound physiological disturbances with very serious consequences. Pervasive abnormalities
of multiple organ systems are affected as time goes on"...

"odds are 1 out 4 that a child after Fontan operation will be dead by the time he or she reaches their late 20s…"

Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2010;13:84-90

Fallo del Fontan : ¿Que pasa con el tiempo?

- AD gigante → Arritmias (extrasístoles A, reentrada A, FA), pérdida energética, trombos-embolias, ACV
- Deterioro de clase funcional
- Obstrucción del Fontan: alt drenaje de V cavas o de VVPP, Ascitis, Enteropatía pierdeproteínas.
- Cianosis por fístulas AV
- Disfunción ventricular: x arritmias, cianosis, VU derecho
- Muerte súbita
- Disfunción de VAV

TÉCNICAS QUIRÚRGICAS IV

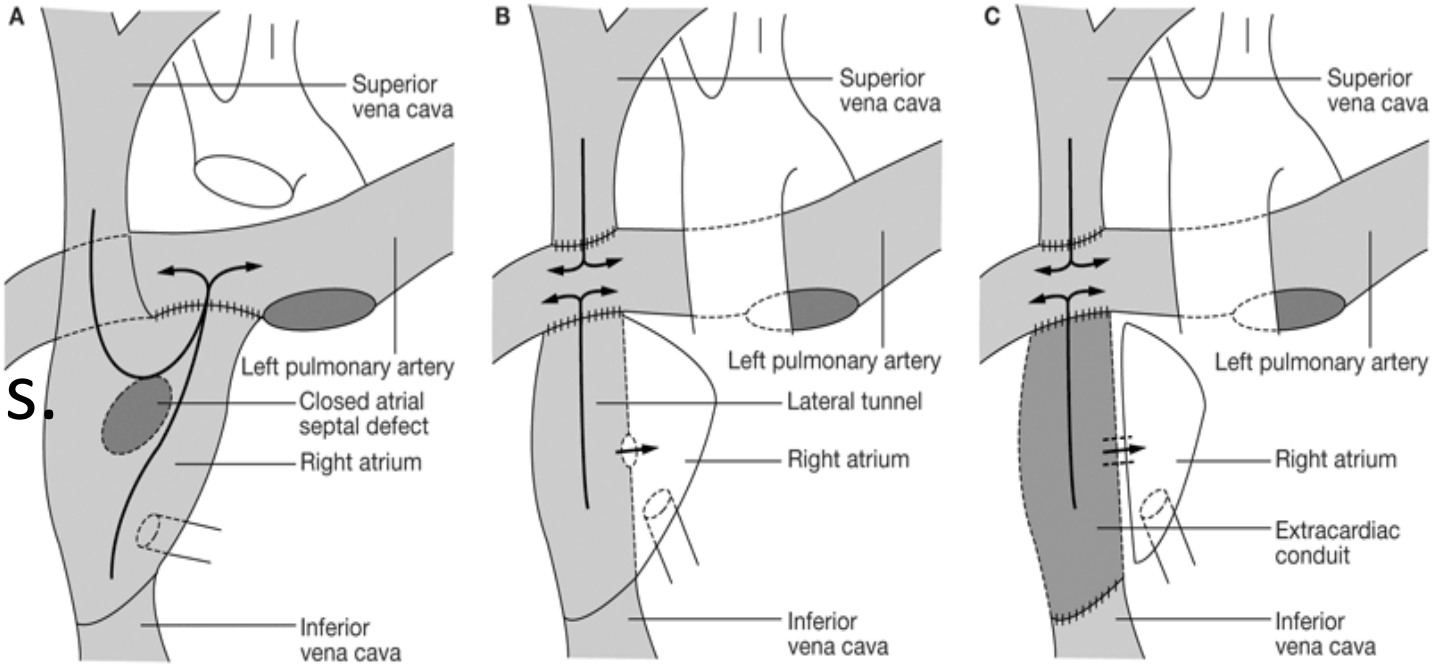
Fontan cavopulmonar: Conexión directa de VCS con RPD (Glenn) y la VCI con las RRPP para q sangre de cavas vaya directa al pulmón

1988: M. de Leval (GOSH): túnel lateral. Hacer Fontan por etapas (actualm Glenn 4-6 meses, Fontán hacia los 3 años)

1990: C. Marcelletti: conducto extracardíaco. Si pac >15 kg, se puede poner conducto de tamaño adulto

1999:

C. Mavroudis. Conversión de Fontan

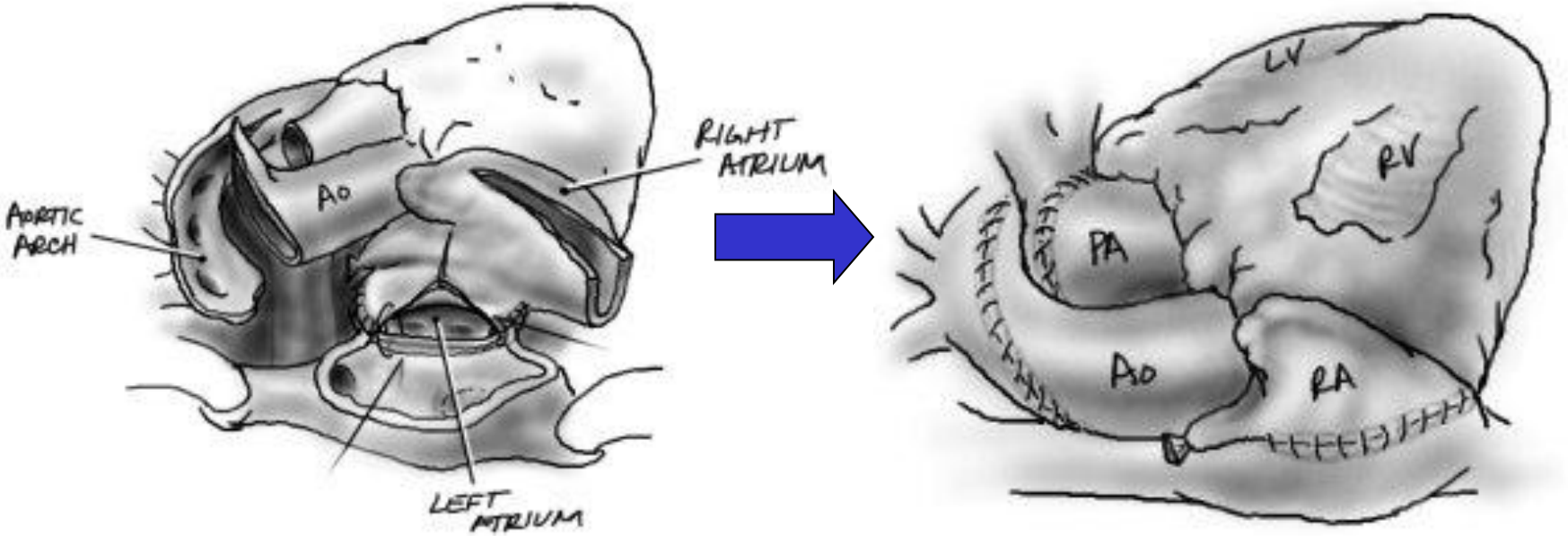


TÉCNICAS QUIRÚRGICAS V

1986

Bailey: Transplante cardíaco

- 1 sola cirugía y obtenemos un ♥ biventricular, se puede realizar en cualquier momento, pero no está exenta de problemas…
- escasez de donantes, sobre todo en niños pequeños
- si cirugías previas mayor riesgo de sangrado y mayor MH
- Efectos secundarios de los inmunosupresores
- imprescindible PP y RVP bajas (< 6 unidades Wood)

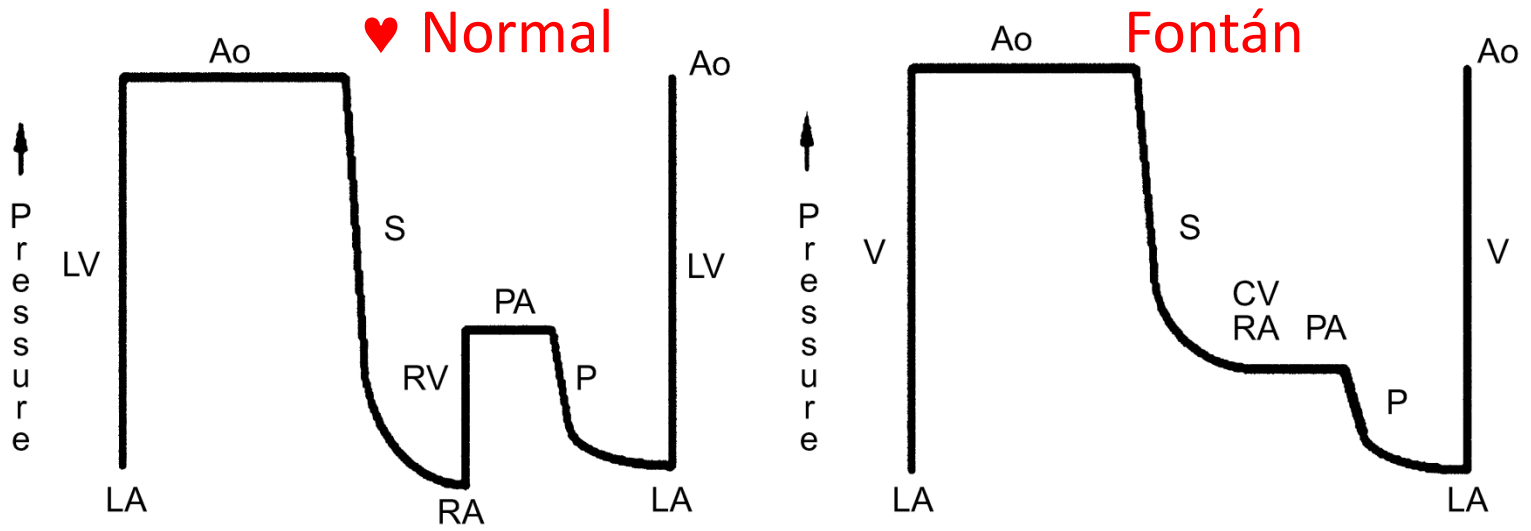


Resultados de cirugía del V. Único

Cirugía	Supervivencia	Complicaciones
Fístula SP	50% a 20 años	Fallo VU, arritmias
Glenn bidireccional	50% a 20 años	Cianosis, fístulas AV pulmonares
Fontan	80% a 10 años 50% a 5 años si epp	Arritmias, fallo VU, enteropatía pp
Trasplante ♥	50% a 10 años	Inmunosupresión

ACC/AHA 2008 Guidelines for adults with congenital heart disease.
Warnes et al. DOI:10.1161/CIRCULATION aha.108.190690

FISIOLOGÍA DEL BUEN FONTAN…



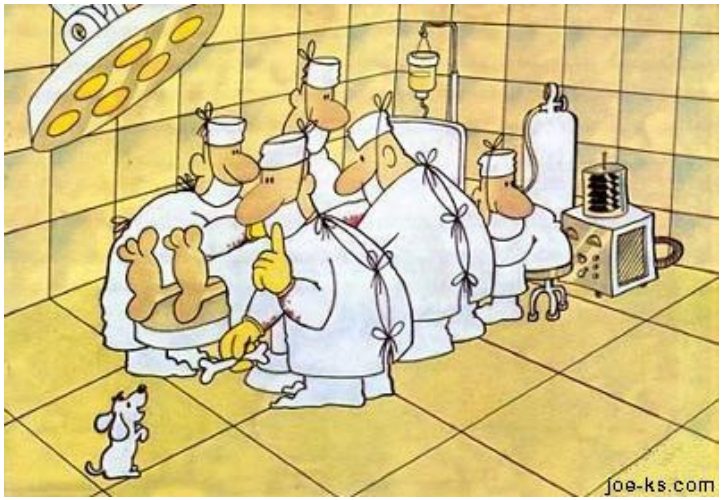
- **Fontán**: Circulaciones en serie, sin ventrículo antes del pulmón→ estado de bajo GC contínuo con poca reserva de precarga, que causa disfunción diastólica del VU
- Imprescindible asegurar buen árbol pulmonar (Diám, desarrollo RRPP)
- El buen Fontán requiere ↓ **RVP** (RVP es el > determinante del GC en el Fontán)
- El resto de los factores no influyen demasiado en el GC
    - Contractilidad VU: Sólo si disfunción severa ↓GC
    - FC: Importante sincronía AV. Sólo taqui/bradi severas ↓GC
    - Postcarga habitualmente ↑ como consecuencia del ↓GC, no causa

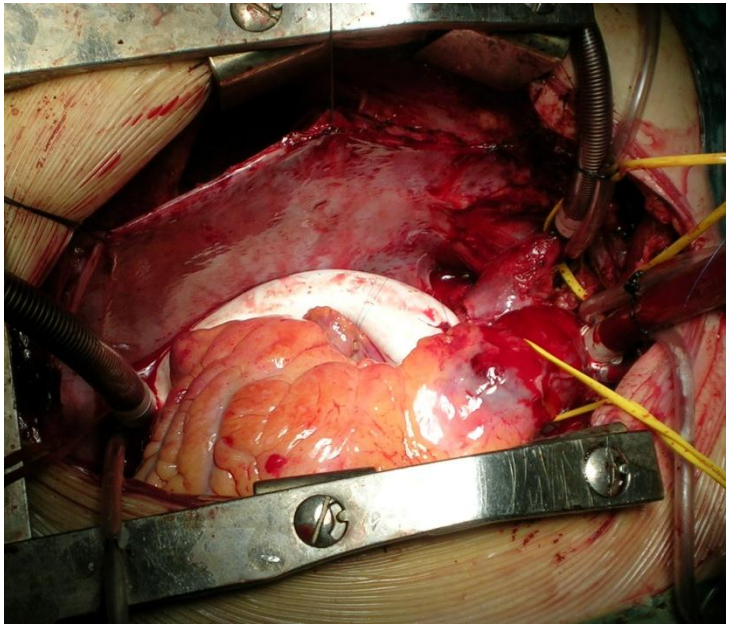
The Fontan Circulation: who controls cardiac output?. M. Gewillig et Al. ICVTS 10 (2010):428-433

Los 10 mandamientos y la operación de Fontan

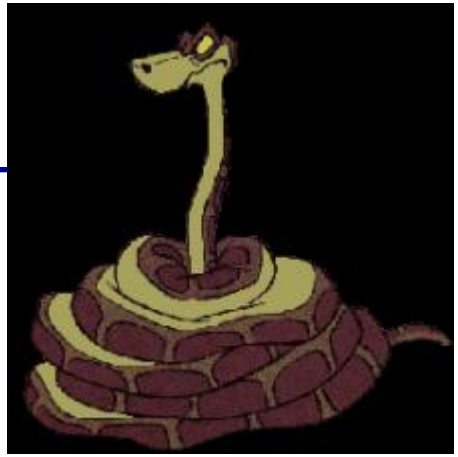
1978: Choussat y Fontan

- Edad > 4 años
- *Ritmo Sinusal*
- Drenaje normal de cavas
- Tamaño normal de AD (AD pequeña contraindica túnel lateral)
- PP media < 15 mmHg
- Resistencias P < 4 U Wood
- Ratio diam AP-Ao > 0,75 (RRPP de buen calibre). Buen árbol pulmonar
- Buena función del VU (FE > 60%, PTDVU < 12 mmHg)
- *Buena función VAV*
- *No shunts que interfieran en la hemodinámica (eliminar previamente colaterales veno-venosas y arterio-venosas)*



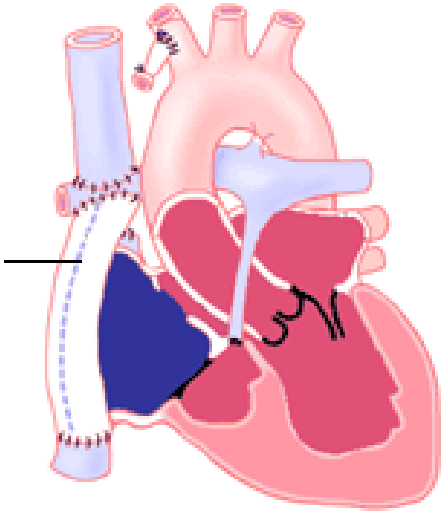


FONTAN FALLA POR:



Aumento progresivo de la RVP

- Atresia o estenosis Mi con FOP restrictivo
- Down
- Fístula SP de larga duración, o con distorsión de RRPP
- Vivir a alta altura respecto el nivel del mar
- AMD crónica (fibrosis P)
- Microtrombos crónicos en RRPP
- Edema linfático
- Flujo P no pulsátil
- Respuesta alterada del endotelio P

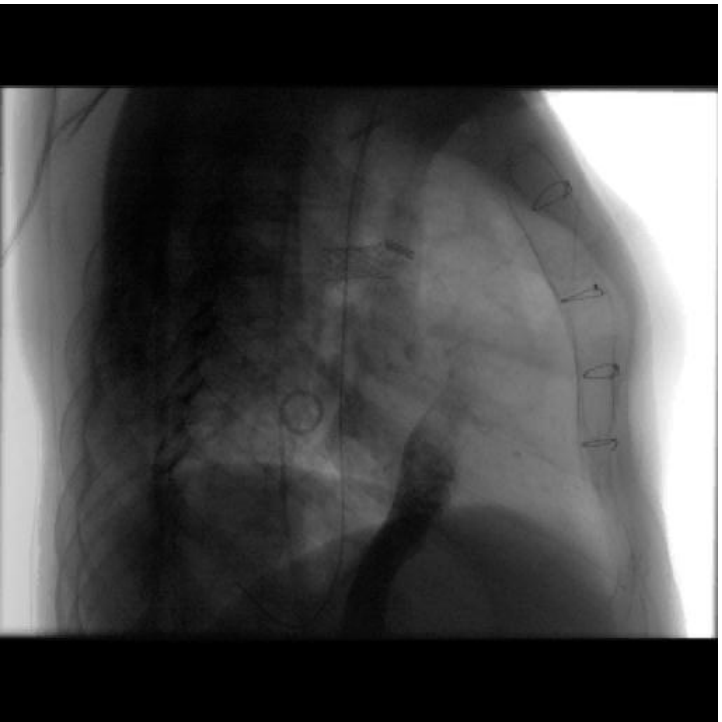


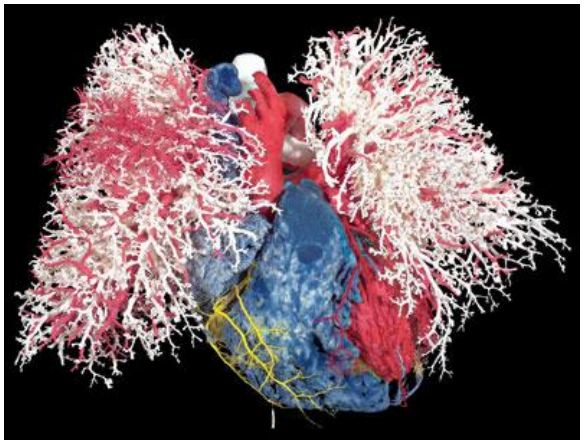
Disf VU con PTD >12 mmHg

- aumenta precarga x sobrecarga de volumen
- VU derecho, miocardiopatía, mala protección miocárdica, arritmia
- Insuf VAV
- bajo GC crónico con disfunción secundaria renal y hepática

La circulación de Fontan y la presión pulmonar

¿Podemos hablar de hipertensión pulmonar?
¿Cuáles son sus consecuencias?
¿Como la tratamos?
y ¿Cuando falla?





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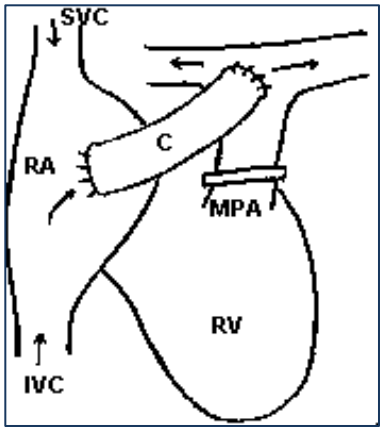
Pediatrica Cardiologia
Hospital “La Paz”

Pulmonary Circulation & Univentricular Physiology

- What did we know about the Pulmonary circulation in 1978?....



1978 Choussat and Fontan

"THE TEN COMMANDMENTS OF THE FONTAN OPERATION"

which should be fulfilled to achieve a successful outcome in Fontan operation....

Age above 4 years
No distortion of lung arteries
Normal venous drainage
Normal ventricular function
Adequate pulmonary artery size
No atrio-ventricular valve leak
Low pulmonary artery pressure (below 15 mmHg)
Low lung blood vessel resistance
Normal heart rhythm
Normal right atrial size

passive lung blood flow....
Lung vascular bed as a "tubing system" with mechanical role

Pulmonary Circulation & Univentricular Physiology

- What do we know in 2012?...

PULMONARY VASCULAR DISEASE **BEFORE** GLENN OR FONTAN

- **Periods of excessive lung blood flow**
  - hyperfunctioning shunt,
  - non-protective bandings
- **INTRAUTERINE PVD**
  (HLHS with Intact or Restrictive IAS)
- **LUNG HYPOPLASIA/UNDERDEVELOPMENT**
  in univentricular heart babies
- **EFFECT OF COMORBIDITIES**
  - Chromosomopathies or MCA syndroms
  - Gastroesophageal reflux
  - Repeated lung infections
  - Skeletal abnormalities (scoliosis, vertebral..)

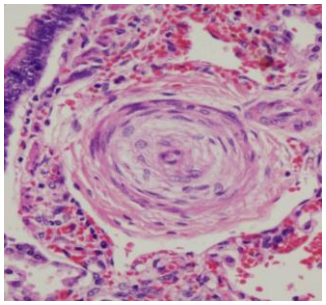
PULMONARY VASCULAR DISEASE DEVELOPING **AFTER** THE FONTAN

- Non pulsatile lung flow on the endothelium
- Consecuences of increased CVP from the physiological 5-8 mmHg….to the 10-13-18 mmHg
  - on the lymphatic system
  - on the liver, kidney…
- Consecuences of Chronic LUNG thromboembolisms
- Systolic and diastolic Ventricular dysfuntion (70%)
- Restrictive neumopathy

Dana Point 2008

1. HIPERTENSION ARTERIAL PULMONAR

- Idiopathic PAH
- Heritable (BMPR2, ALK1, Endoglin with or without HHT, Unknown)
- Drugs and toxins induced
- Associated with: Connective tissue diseases, HIV infection, Portal Hypertension, *Shunt Sistémico-pulmonar*, Schistosomiasis, chronic hemolytic anemia
- PPHN (HTP PERSISTENTE DEL RN)



2. HTP por enfermedad CORAZÓN IZDO

- Disfunción Sistólica
- Disfunción Diastolica
- Valvulopatías

3. HTP POR ENFERMEDAD PULMONAR/HIPOXIA

4. HTP TROMBOEMBÓLICA CRÓNICA (CTEPH)

5. PH WITH UNCLEAR OR MULTIFACTORIAL MECHANISMS

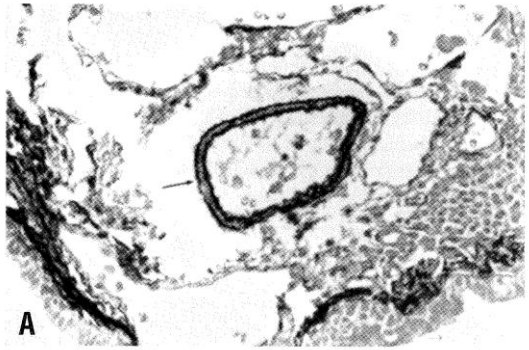
PODEMOS HABLAR DE Hipertensión PULMONAR EN EL FONTAN?

- **NO, si la definición es de PMAP > 25 mmHG…**
  (estarian todos muertos)

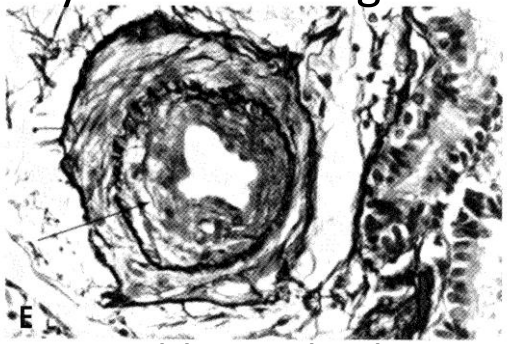
PERO SI ……DE ENFERMEDAD VASCULAR PULMONAR

PVD BEFORE THE FONTAN…

Histomorphometric analysis of pulmonary vessels in single ventricle for better selection of patients for the Fontan operation Levy M, et al. J Thorac Cardiovasc Surg 2002



Normal terminal bronchial artery: thin and regular wall



Terminal bronchial artery increased wall thickness and intimal fibrosis

Lung biopsy in 40 patients with TA or single ventricle at the time of Glenn/Fontan surgery

mPAP mmHg	Number of Patients	Abnormal Histology
< 18 mmHg	35	18 (51%)
>18 mmHg	5	5 (100%)

Extension of muscle in the walls in the distal intraacinar arteries

IN ALL CASES OF OF FONTAN FAILURE!!

Guidelines and Consensus

A consensus approach to the classification of pediatric pulmonary hypertensive vascular disease: Report from the PVRI Pediatric Taskforce, Panama 2011

Volume 1, Number 2 (April-June 2011)
Pulmonary Circulation
ISSN: 2045-8932
E-ISSN: 2045-8940

1. Prenatal/developmental pulmonary vascular disease
2. Perinatal Pulmonary Vascular Maladaptation (PPHN)
3. Heart Disease
4. Bronchopulmonary dysplasia
5. Isolated Pulmonary Arterial Hypertension
6. Multifactorial in congenital malformation syndromes
7. Lung Diseases
8. Thromboembolic Disease
9. Hypobaric hypoxic exposure
10. Associated with other Disorders

A consensus approach to the classification of pediatric pulmonary hypertensive vascular disease: Report from the PVRI Pediatric Taskforce, Panama 2011

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3.3. Pulmonary vascular disease following staged palliation for single ventricle physiology

3.3.1. After stage 1

- *PA banding*
- *modified Norwood*
- *Hybrid procedure*
- *aortopulmonary or ventricular pulmonary shunt*
- *stenting PDA,…*

3.3.2. After SVC to PA anastomosis (Glenn).

RVP > 3 UW.m2

GTP > 6 mmHg

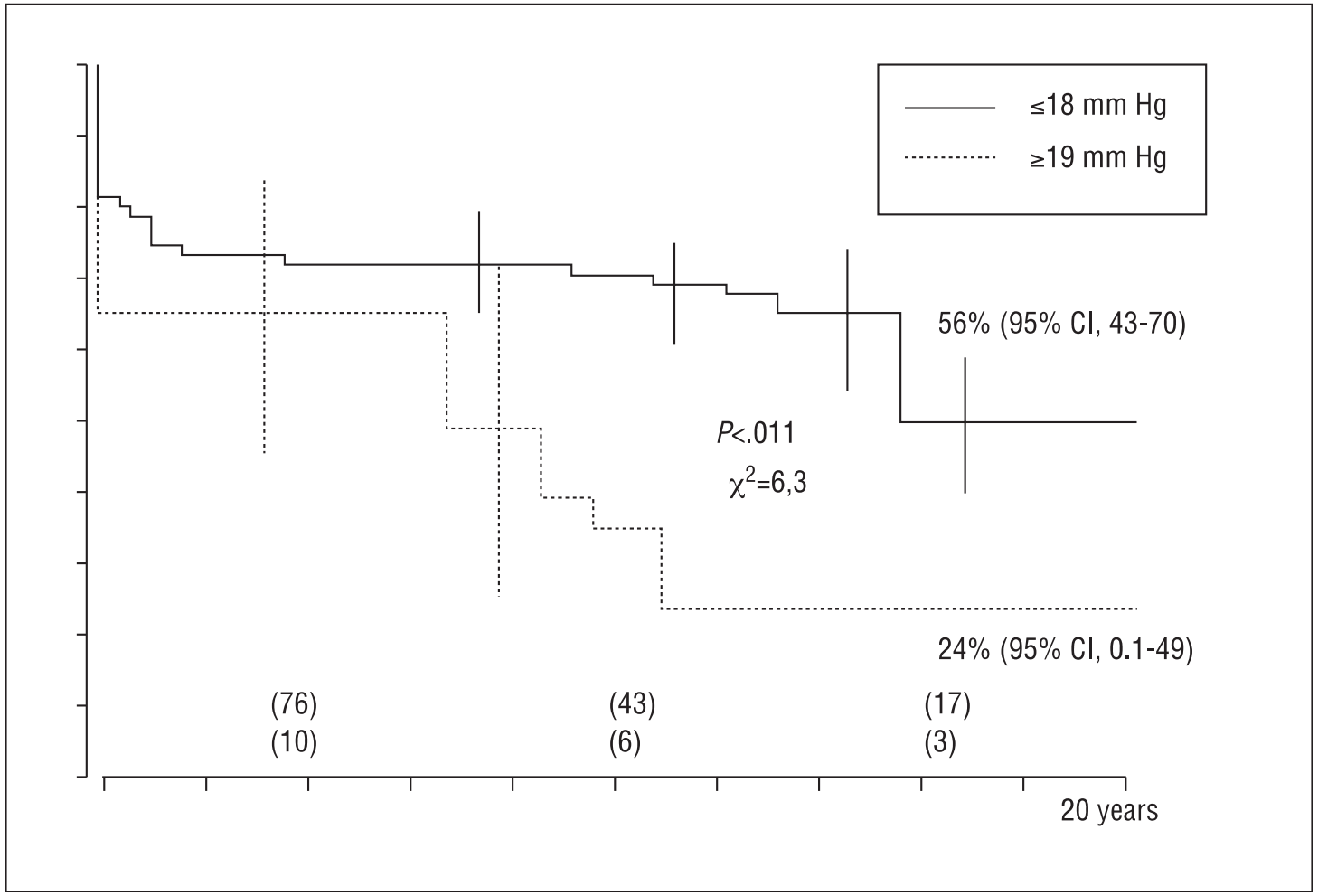
3.3.3. After total cavopulmonary anastomosis (Fontan).

- *These patients may have a PA pressure less than 25 mmHg but an increased PVRI and Pulmonary Vascular Disease…*

Cuales son las consecuencias de la Enfermedad Vascular Pulmonar en el Fontan?

Single-Stage Fontan Procedure: Early and Late Outcome in 124 Patients

Mario Cazzaniga, Luis Fernández Pineda, Fernando Villagrá*, Julio Pérez de León*, Ricardo Gómez*, Pedro Sánchez* and José Díez Balda**



Single-Stage Fontan Procedure: Early and Late Outcome in 124 Patients

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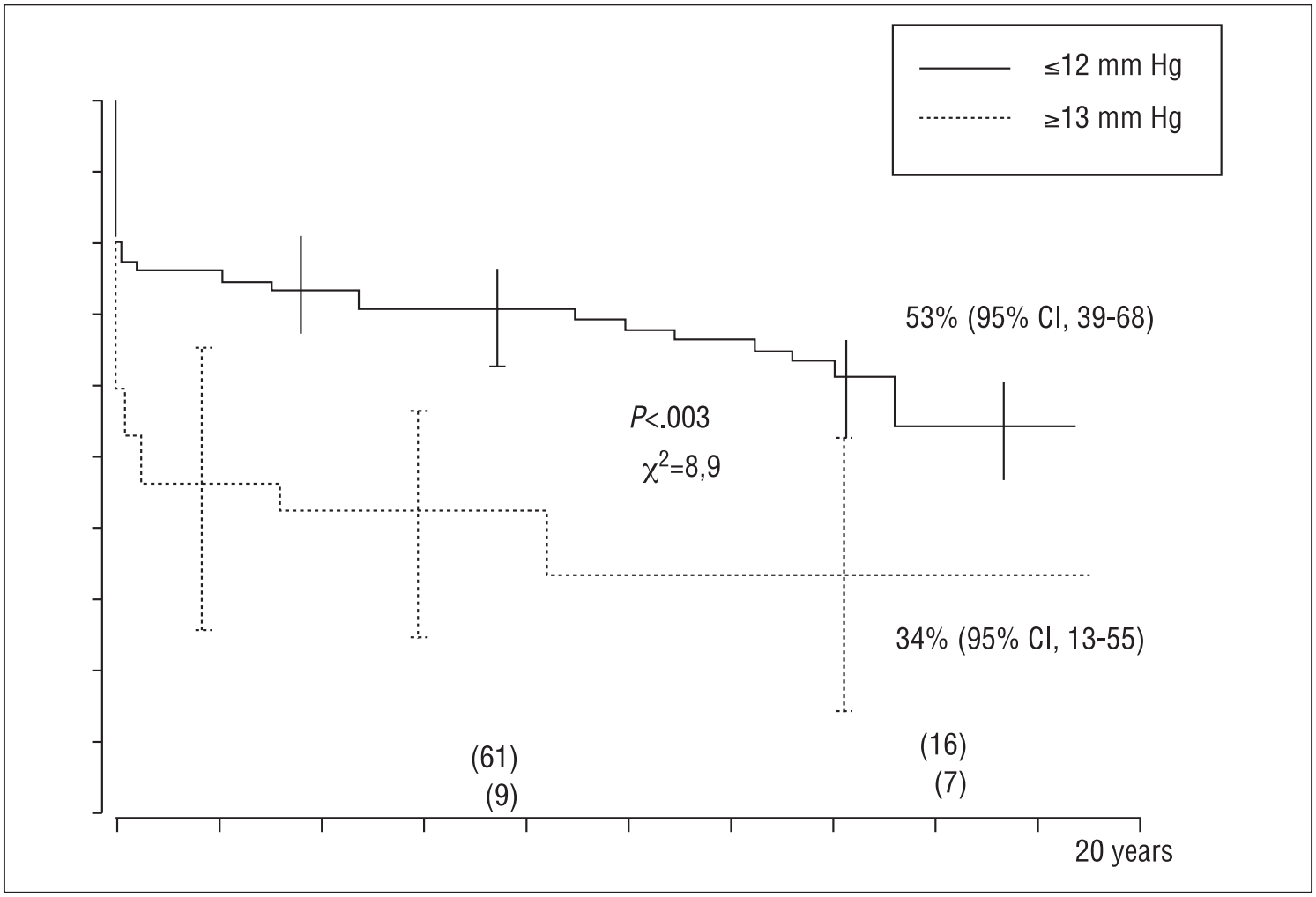
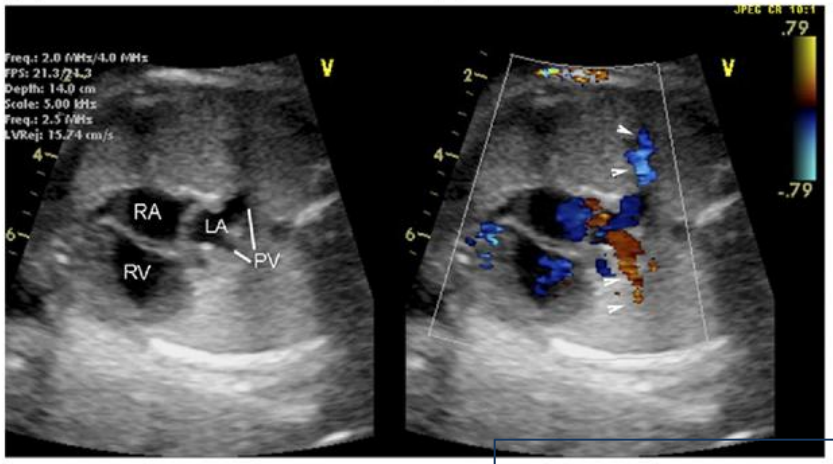


PULMONARY VASCULAR DISEASE BEFORE THE FONTAN

PVD BEFORE THE FONTAN…

INTRAUTERINE PULMONARY VASCULAR DISEASE

IN HLHS WITH RESTRICTIVE OR INTACT INTERATRIAL SEPTUM



IMAGES IN CARDIOLOGY

Hypoplastic Left Heart Syndrome With Intact Atrial Septum

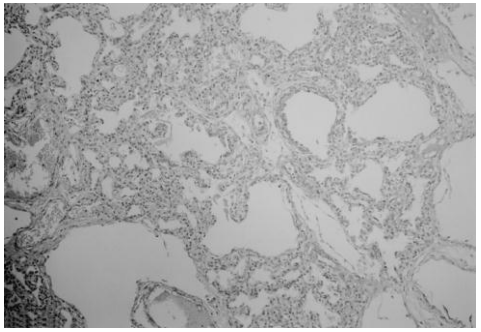
Sequelae of Left Atrial Hypertension In Utero

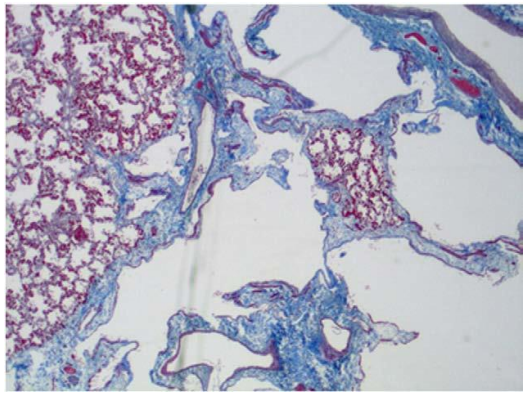
JACC 2011 Vol. 57, No. 20, 2011

Hypoplastic Left Heart Syndrome with Restrictive Atrial Septal Defect and Congenital Pulmonary Lymphangiectasis

Cardiovasc Pathol 1999;8:49–51

Muscularization of left atrial wall and pulmonary veins wall



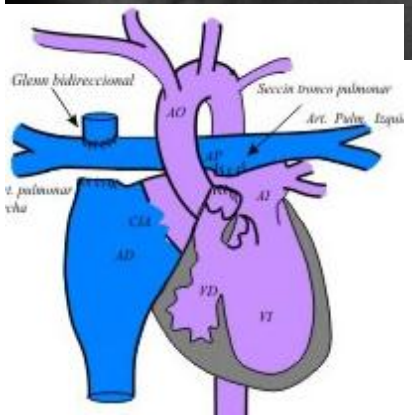


PULMONARY LINFANGIECTASIA

PVD BEFORE THE FONTAN…

Down syndrome patients
High PVR

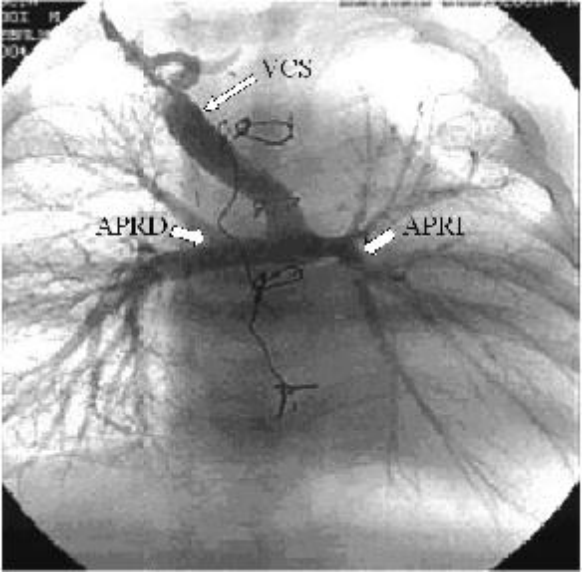
- HYPOPLASIA VASCULAR BED
- LUNG HYPOPLASIA
- SAOS
- REPEATED LUNG INFECTIONS



STAGE I:
systemic-pulmonary shunt

Severe Hiposaturation
In spite of functioning
BT shunt

FAILURE
UNIVENTRICULAR CIRCULATION
(Glenn o Fontan)



STAGE II (Glenn):

- Severe Hiposaturation
- Glenn "take-down" needed due to high PVR

ORIGINAL ARTICLE

Gen Thorac cardiovasc surg **2008**

Single ventricle repair in children with Down syndrome

STAGE III (Fontan):

Glenn en 6 S. Down

FONTAN COMPLETED ONLY in 16%

n"
stop.

3 didn´t meet

2. Fontan

ESTRICT SELECTION PATIENTS
Lung Biopsy at Glenn

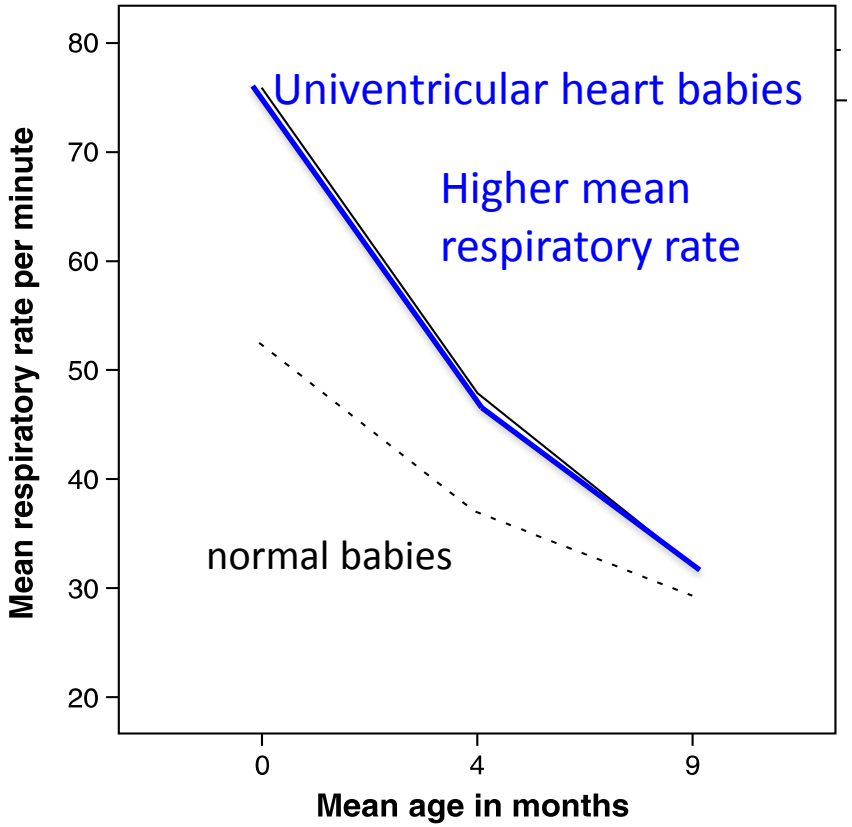
postoperative **Success**
PH Crisis tracheostomy

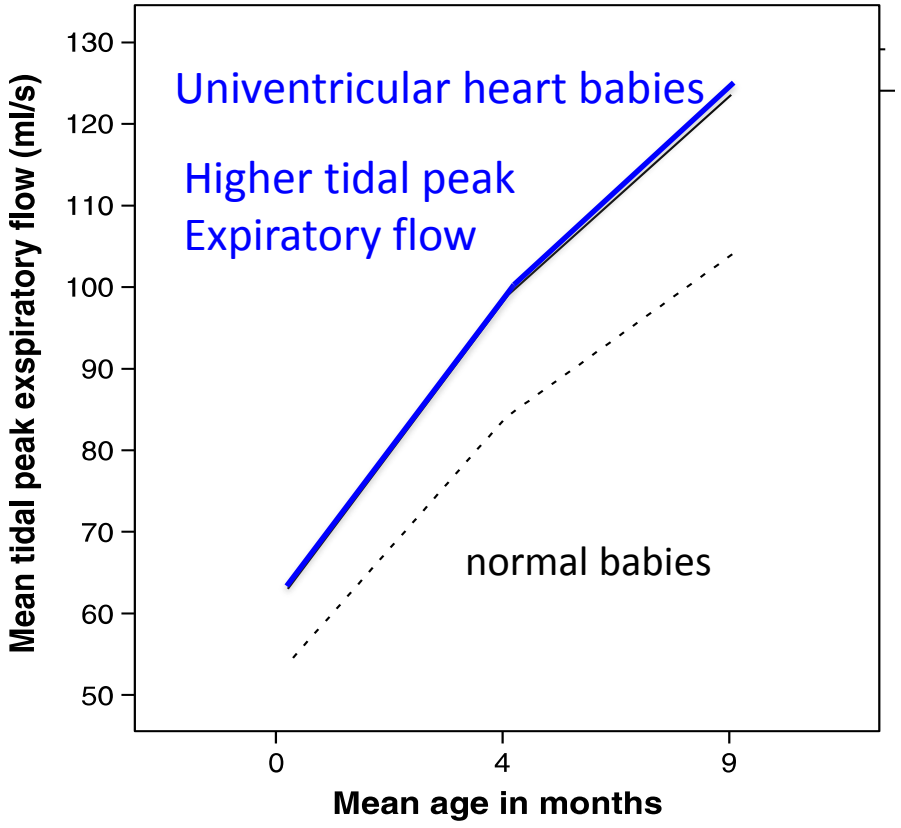
PVD in DOWN

PVD BEFORE THE FONTAN…

Differing lung function development in infants with univentricular hearts compared with healthy infants Matthews IL, Acta Pediatrica 2008

Conclusion: The pattern of lung function development is different in the patients with univentricular hearts compared to healthy controls. Lung function measured at birth is predictive of later lung function.



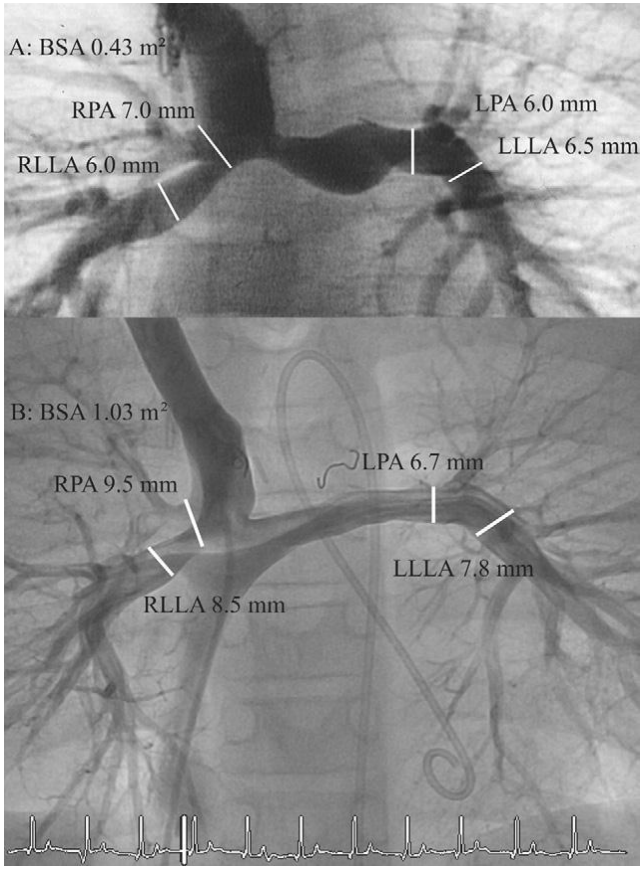


PULMONARY VASCULAR DISEASE
AFTER THE FONTAN

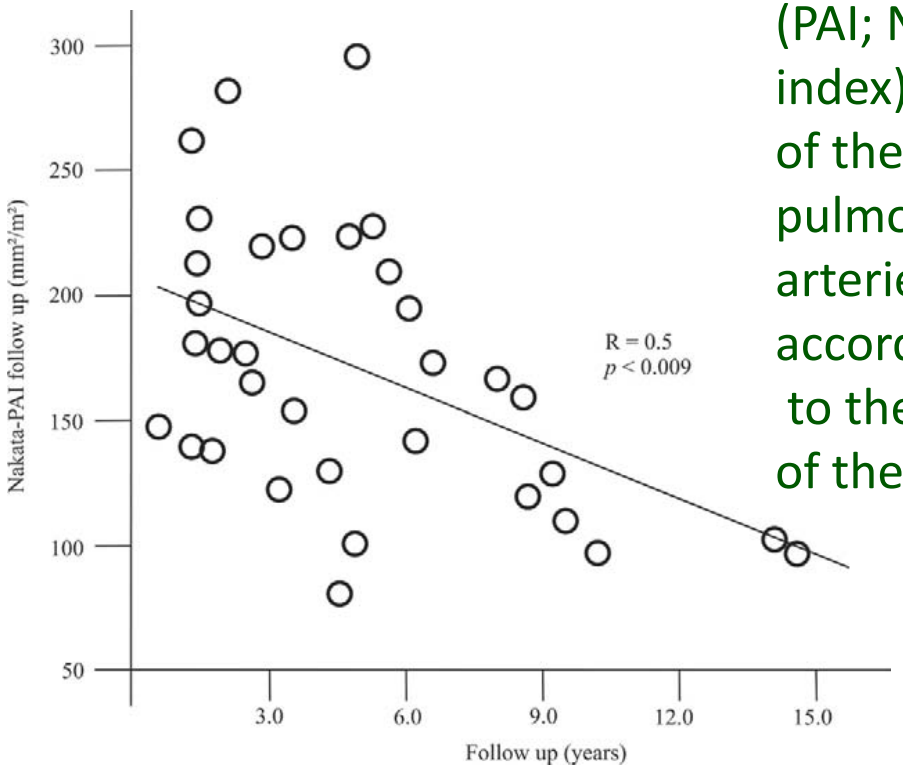
PVD developing AFTER THE FONTAN…

Absence of Pulmonary Artery Growth After Fontan Operation and Its Possible Impact on Late Outcome

Ovroustsky S. *Ann Thorac Surg* 2009;87:826-831



same patient 9 years after Fontan operation



Significant decrease in the pulmonary artery index (PAI; Nakata index) of the central pulmonary arteries according to the duration of the follow-up.

PVD developing AFTER THE FONTAN…

Pulmonary Thromboembolism in Fontan

- Anatomic Risk factors:

  low flow state, stasis in venous pathways, right to left shunts, blind cul de sacs, prosthetic materials, and/or atrial arrhythmia

- Prothrombotic tendency in Fontan patients
  - A hypercoagulable state: deficiencies in proteins C and S,…
  - Endothelial dysfunction *before* and *after* the Fontan

    (hypoxia, hiperviscosity,…increased Von Villebrand factor levels

Coagulation abnormalities in patients with single- ventricle physiology precede the Fontan procedure

J Thorac Cradiovasc Surg 20028;123 (3):459.

Altered endothelial function following the Fontan procedure

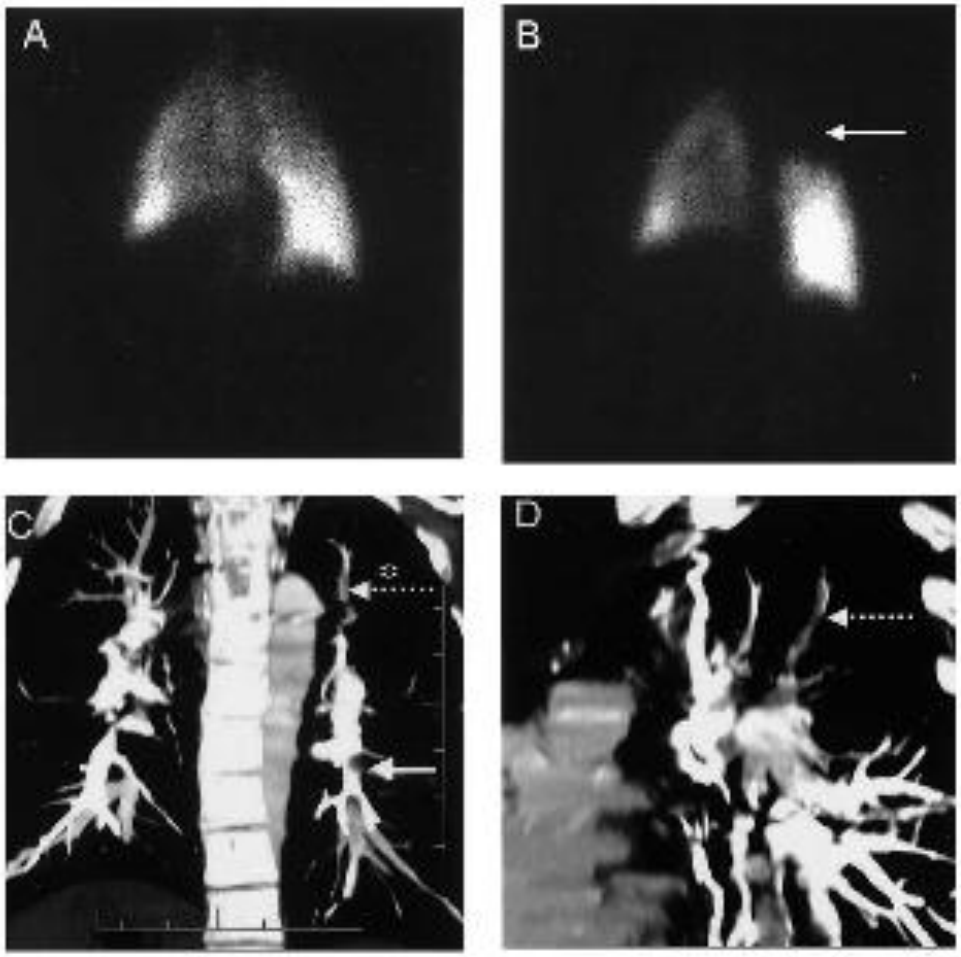
Cardiol Young 2008;18(1): 70-4

PVD developing AFTER THE FONTAN…

Prevalence of "Silent" Pulmonary Emboli in Adults After the Fontan Operation

JACC 2003

Chetan Varma, MBBS, MD, Matthew R. Warr, BSc, Aaron L. Hendler, MD, Narinder S. Paul, MD, Gary D. Webb, MD, FACC, Judith Therrien, MD
Toronto, Canada



30 patients (mean age 26 ± 7 years

18 Had an Atriopulmonary connection

(17%) adult Fontan patients had an intermediate or high probability for PE on VQ scan, confirmed on CT pulmonary angiography.

No patient had a thrombophilia tendency.

Late age at time of Fontan operation and ***type of Fontan*** anatomy ($p < 0.001$) associated with increased risk of silent PE.

no patients on coumadin (n =9) were diagnosed with an asymptomatic PE,

prevalence of silent PE in nonanticoagulated

Fontan patients to 24%

A Multicenter, Randomized Trial Comparing Heparin/Warfarin and Acetylsalicylic Acid as Primary Thromboprophylaxis for 2 Years After the Fontan Procedure in Children

(J Am Coll Cardiol 2011;58:645–51)

111 eligible patients were randomized (57 to ASA, 54 to heparin/warfarin)

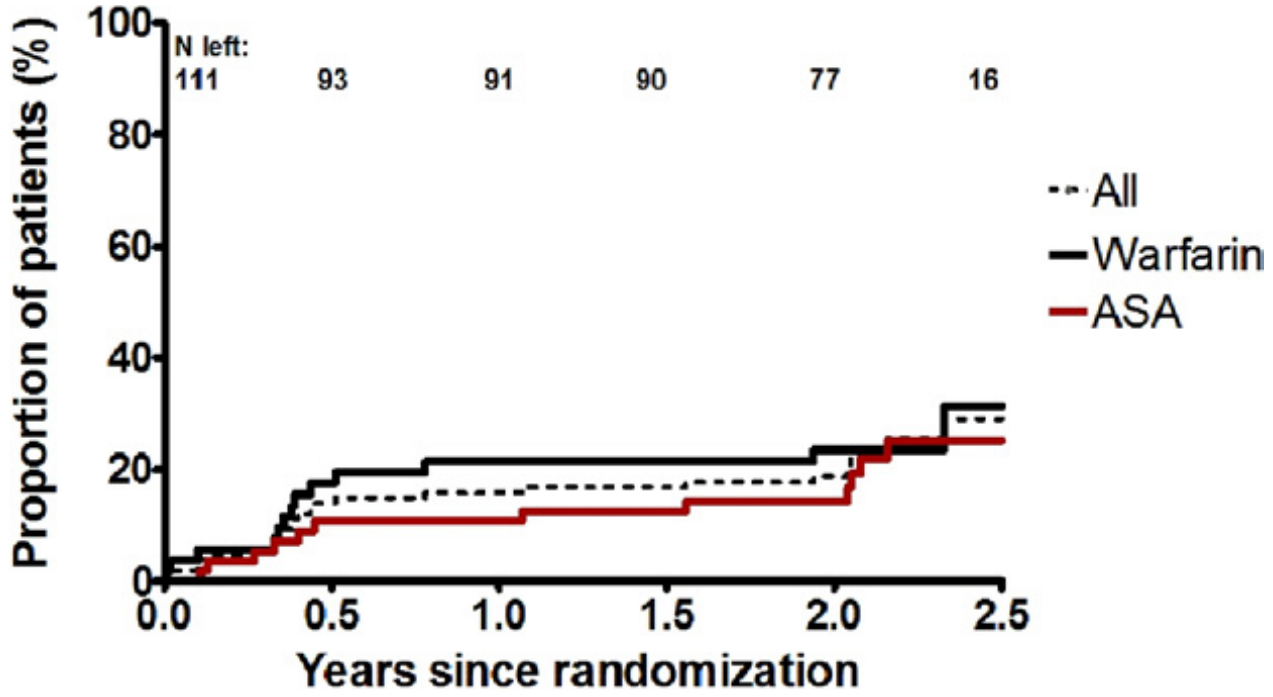
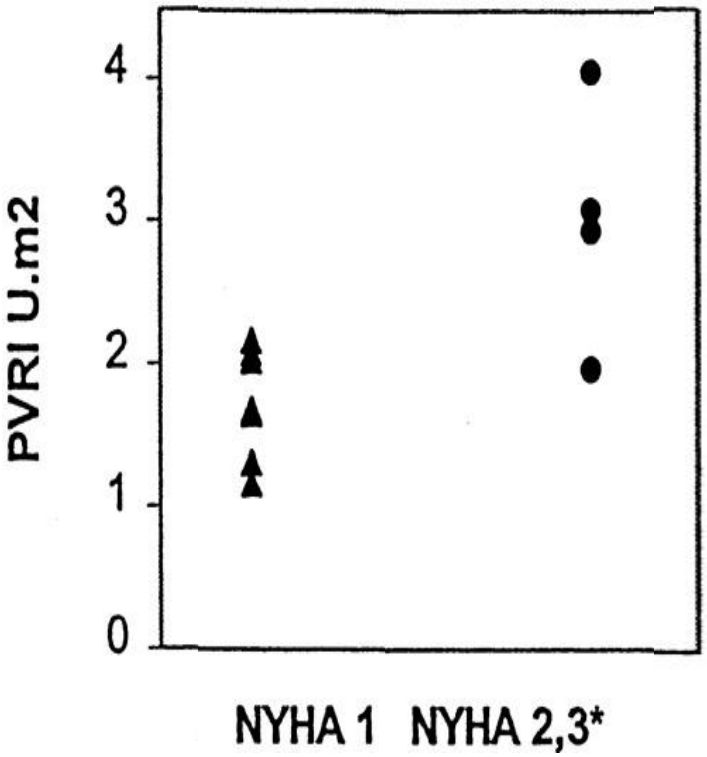


Figure 3 Thrombosis After Randomization

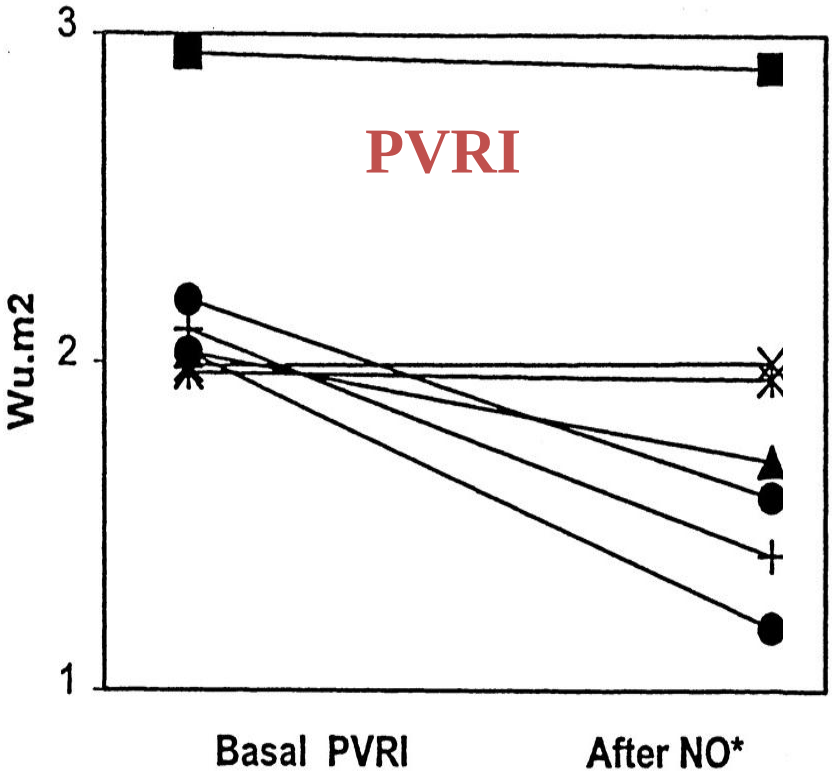
PVD developing AFTER THE FONTAN…

Basal Pulmonary Vascular Resistance and Nitric Oxide Responsiveness Late After Fontan-Type Operation

Heart Association
Learn and Live



Patients with higher PVRI were in NYHA classes 2 and 3.



Effect of exogenous NO on PVRI of patients with nonpulsatile flow in the pulmonary arteries. Significant drop of mean PVRI (*P<0.05).

Khambadkone S et al. Circulation 2003;107:3204-3208

Evidence of pulmonary vascular disease after heart transplantation for Fontan circulation failure

- 15 failed Fontan patients undergoing heart transplantation.
- Compared PVRI and TPG before and after Heart transplant
- in a failing Fontan a *normal PVR does not exclude PVD*

TPG (TRANSPULMONARY GRADIENT): mean PAP-LAP

Variable	n	Before Tx		After Tx
All patients				
PAP (mm Hg)	13	17.0 ± 3.7		19.7 ± 3.3
TPG (mm Hg)	12	5.3 ± 2.3		12.0 ± 2.1
PVR (Wood units · m^2)	6	1.8 ± 1.1	P<0.0001	2.7 ± 1.0

Mitchel et al. J Thorac Cardiovasc Surg 2004;128: 693-702

Evidence of pulmonary vascular disease after heart transplantation for Fontan circulation failure

Underlying PVD became apparent
after heart Tx in patients with late-failing Fontan

	Early Fontan		Late Fontan		All patients	
	n	Mean ± SD	n	Mean ± SD	n	Mean ± SD
Mean PAP (mm Hg)						
Pretransplantation	4	17.0 ± 5.0	11	16.7 ± 3.3	15	16.8 ± 3.6
Posttransplantation	4	17.8 ± 4.3	10	20.0 ± 2.4	14	19.9 ± 3.3
Mean TPG (mm Hg)						
Pretransplantation	4	6.0 ± 2.0	10	4.5 ± 2.8	14	4.9 ± 2.8
Posttransplantation	4	10.5 ± 1.7	10	12.8 ± 2.0	14	12.1 ± 1.4
Mean PVR (Wood units · m^2)						
Pretransplantation	4	2.1 ± 1.4	5	1.5 ± 0.7	8	1.7 ± 0.6
Posttransplantation	4	2.0 ± 0.4	10	3.8 ± 0.8	14	3.3 ± 1.7

The Fontan circulation itself induces pulmonary vascular disease??

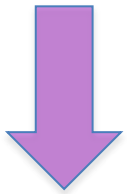
J Thorac Cardiovasc Surg 2004;128: 693-702

PVD developing AFTER THE FONTAN…

Pulsatile flow is thought to lower pulmonary vascular resistance by passive recruitment of capillaries and by active vasodilation.

Shear-Stress-induced EDRF release from the small arteries

Effect of Non pulsatile lung flow on the endothelium

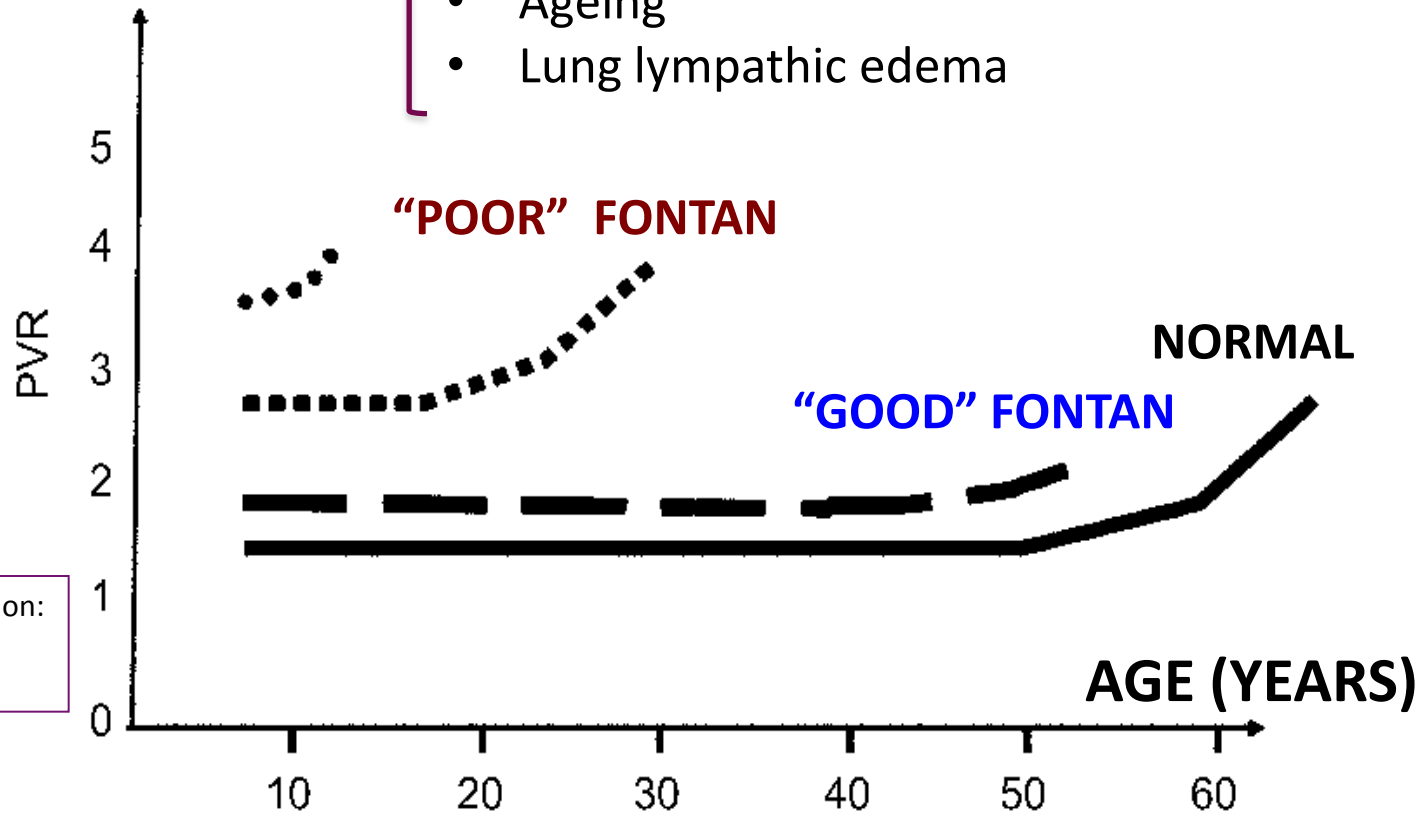


ENDOTHELIAL DYSFUCTION

PVD developing AFTER THE FONTAN…

Progressive increase in PVR after the Fontan surgery:

- Suboptimal PA growth after Glenn / Fontan
- Absence of pulsatile pulmonary flow
- Functional loss of lung segments
- Ageing
- Lung lympathic edema



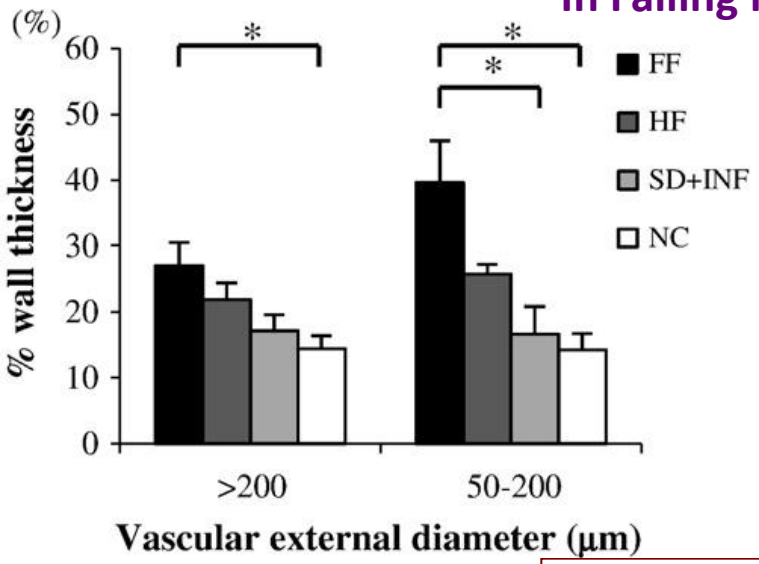
The Fontan Circulation: Who controls the cardiac output?

Gewillig M

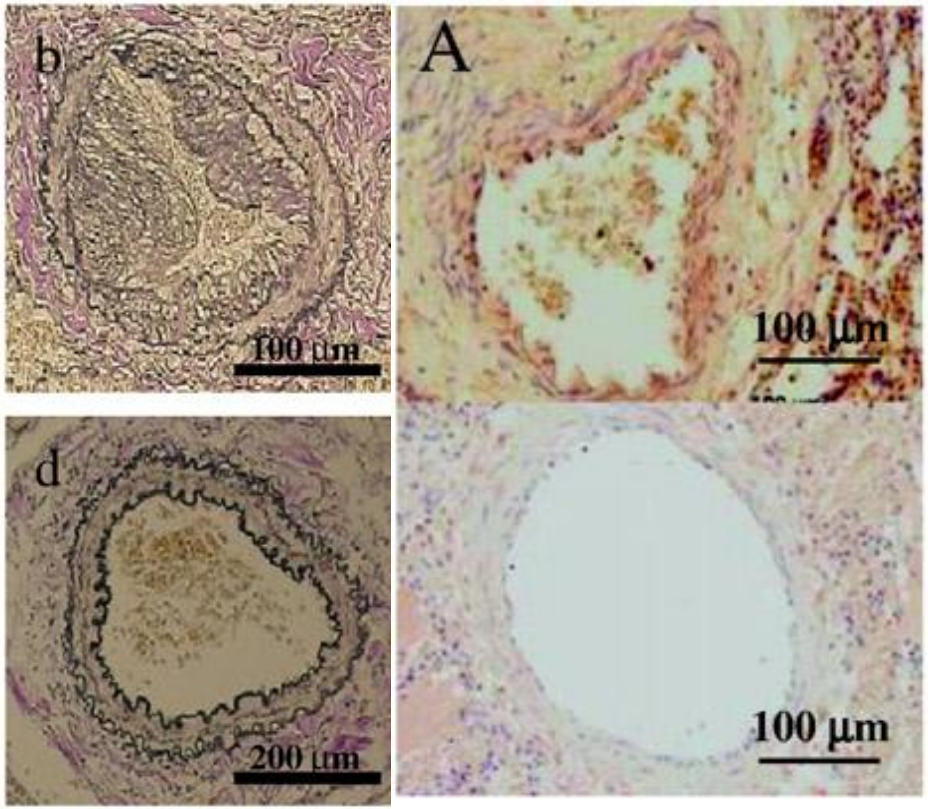
Interactive CardioVascular and Thoracic Surgery 10 (2010) 428-433

Clinical correlation and prognostic predictive value of neurohumoral factors in patients late after the Fontan operation.
Am Heart J 2005;150(3):588–94.

INCREASED serum LEVELS OF ENDOTHELIN-1 IN FONTAN PATIENTS



Increassed wall thickness: medial and intimal Hypertrophy In Failing fontan



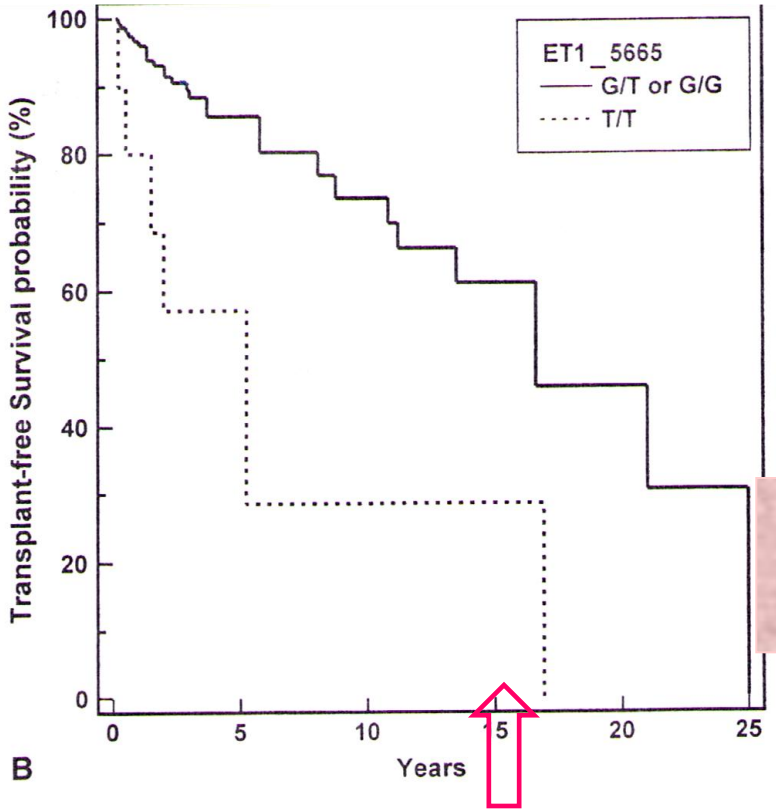
Overexpression of endothelin-1 and endothelin receptors in the pulmonary arteries of failed Fontan patients. Int J Cardiol. 2011 Feb 26.

The endothelin-1 G5665T polymorphism impacts transplant-free survival for single ventricle patients

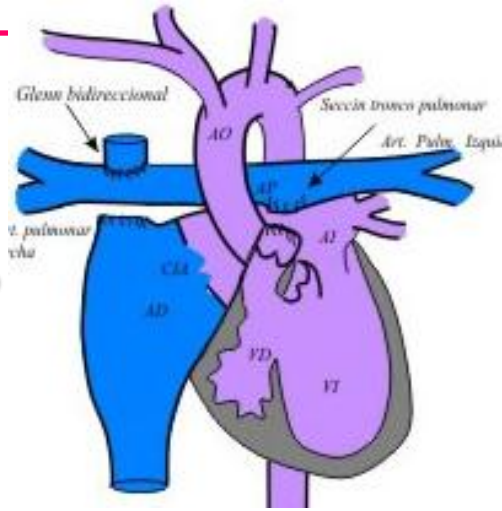
J Thorac Cardiovasc Surg 2008

165 patients

UNIVENTRICULAR heart and Fontan Surgery

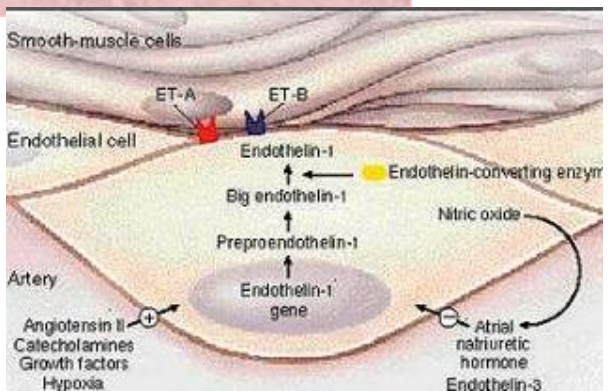


Homocigotes variant TT
Worse prognosis



RELATIONSHIP
SURVIVAL
free of heart Tx
Fontan failure

POLIMORFISMS GEN
PRE-PROENDOTHELIN-1



¿Cómo tratamos la Enfermedad Vascular Pulmonar en el Fontan?

Bosentan Induces Clinical, Exercise and Hemodynamic Improvement in a Pre-Transplant Patient With Plastic Bronchitis After Fontan Operation

Sotiria C. Apostolopoulou, MD, PhD, John Papagiannis, MD, and Spyridon Rammos, MD, PhD

J Heart Lung Transplant 2005;24:1174 – 6

D-TGA+ VSD + PS
At age 5: Glenn
At age 7: lateral tunnel Fontan.

8 months later

PLASTIC BRONCHITIS

Mean PAP 20 mmHg,
end-diast. VP 14 mm
TPG 5 mmHg

Fontan
FENESTRATION

NO resolution of PLASTIC BRONCHITIS

At age 14 **BOSENTAN**

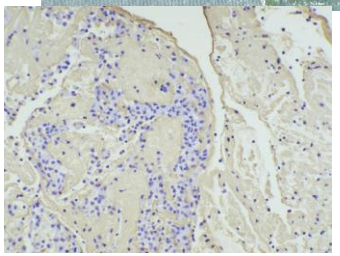
16 WEEKS

Symptomatic improvement

mPAP	**18**
TPG	**8**
PVR	**192**
Qp/Qs	**0.7**
WHO	**III**
Sat O2	**90%**

mPAP	**15**
TPG	**5**
PVR	**111**
Qp/Qs	**0.9**
WHO	**II**
Sat O2	**94%**

Pulmonary vasodilation therapy with sildenafil citrate in a patient with plastic bronchitis after the Fontan procedure for hypoplastic left heart syndrome



J Thorac Cardiovasc Surg 2006;132:1232-1233

3 y.o child HLHS, neonatal Norwood
Glenn at 3 months
Fontan at 3 years
PLASTIC BRONCHITIS had started 2 months after the Fontan

Mean PAP 16 mm Hg
PVRI 8.1 W.U.m2.
C.I. 2.8 L/min/m2

epoprostenol therapy

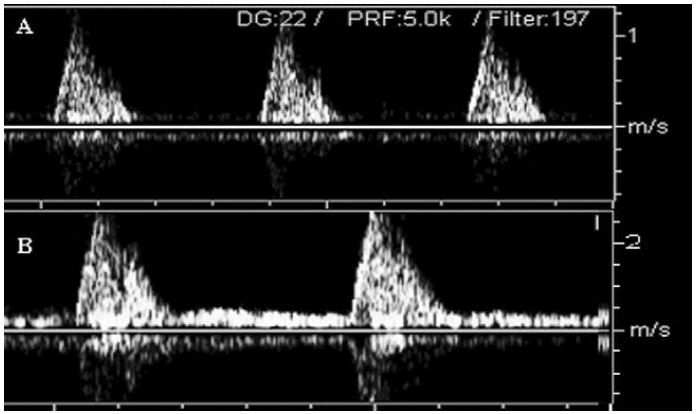
Mean PAP 12 mm Hg
PVRI 4.5 Wood U.m2.

Unresolved plastic bronchitis

SILDENAFIL added

Resolution of plastic bronchitis and epoprostenol Withdrawal
6 months F.U.

Resolution of Protein-Losing Enteropathy and Normalization of Mesenteric Doppler Flow With Sildenafil After Fontan Ann Thorac Surg 2006



HLHS newborn, Norwood , Glenn

At age 4; mean PAP of 10 mm Hg
TPG of 3 mm Hg.
Nonfenestrated Fontan

AT AGE 6: PROTEIN LOSING ENTEROPATHY
mPAP 14 mmHg, ***restrictive ASD***
Atrioseptostomy and
Fontan fenestration
Resolution of the PLE

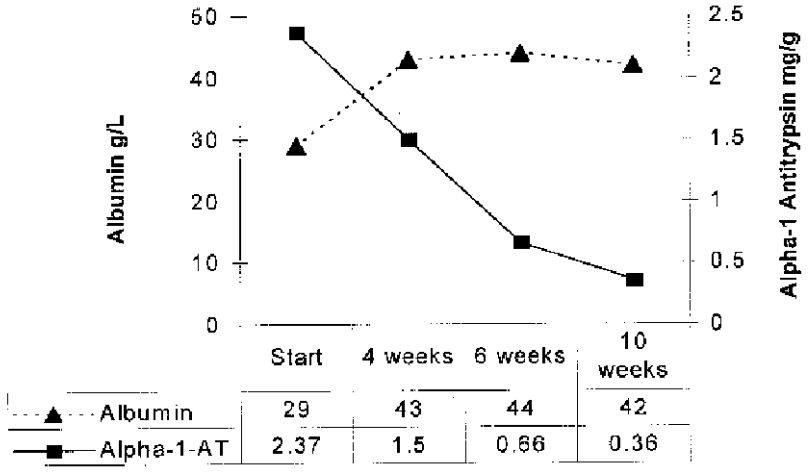


Fig 2 Albumin and alpha-1 antitrypsin (AT) levels over time on sildenafil treatment.

AT AGE 8: Sat O2 78%
basal mPAP 13 mm Hg
occlusion 14 mm Hg
PARTIAL FENESTRATION CLOSURE

3 mo.

PROTEIN LOSING ENTEROPATHY

High gradient through the fenestration

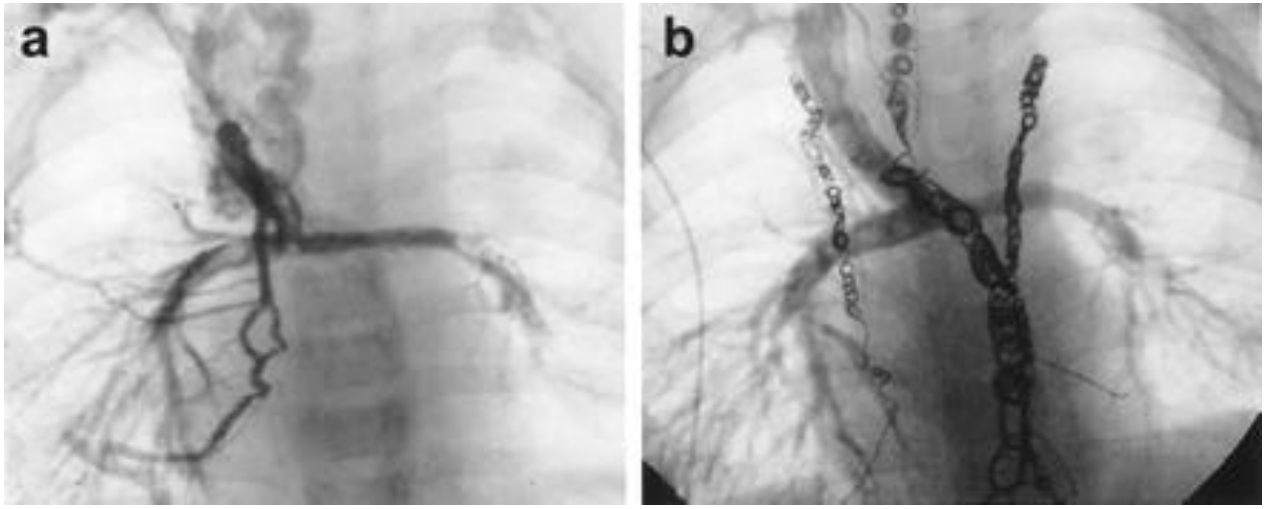
6 weeks

SILDENAFIL

PLE RESOLUTION

Pulmonary artery growth after Norwood and bidirectional Glenn procedure

Interact CardioVasc Thorac Surg 2008;7:328-330



- **HLHS with Restrictive IAS**
  Hybrid procedure.
- Glenn+ enlargement ASD at 6 moths

9 months old

Nakata index 73 mm^2/m^2

mPAP 18 mmHg

Sat O2: 75%
After closure of venous collaterals

SILDENAFIL

4 years old

Nakata index: 117 mm^2/m^2

mPAP 13 mmHg

Sat O2: 83%

Successful

FONTAN

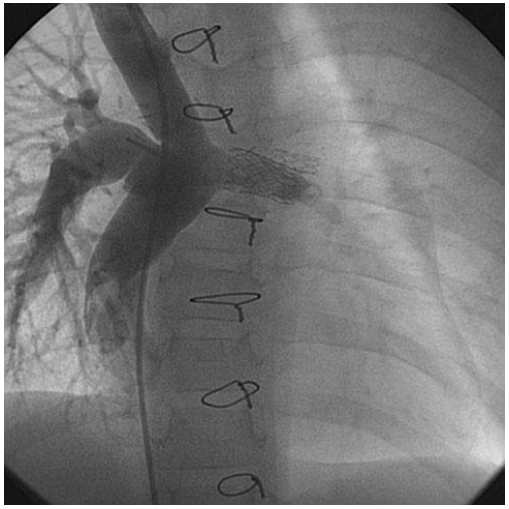
mPAP 15 mmHg

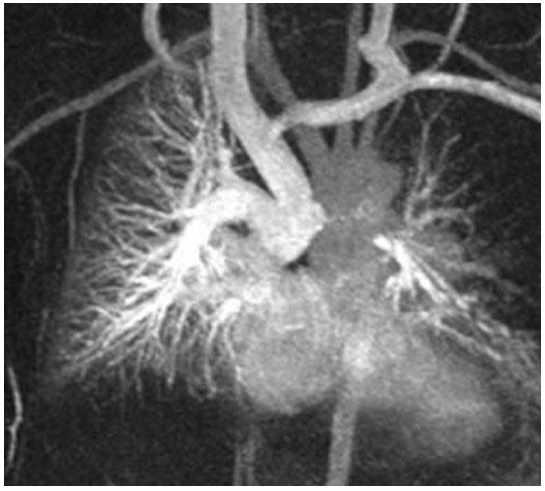
Sat O2: 98%

1 year follow-up !!

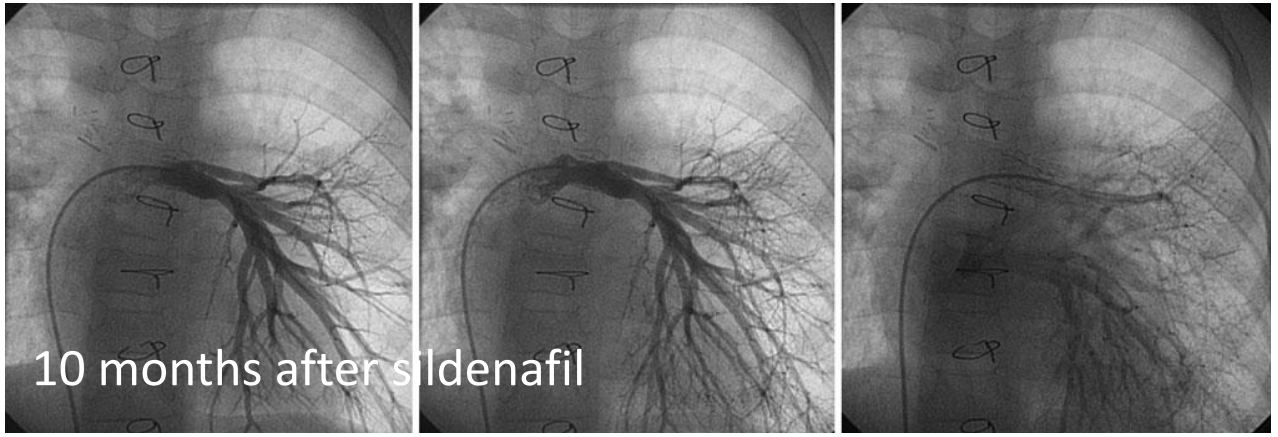
Lost Unilateral Capillary Perfusion During Nonpulsatile Pulmonary Circulation: Successful Recovery by Oral Sildenafil

Pediatr Cardiol 2011





Arterial phase Capillary phase Venous phase.



Hospital Universitario La Paz

SaludMadrid

Comunidad de Madrid

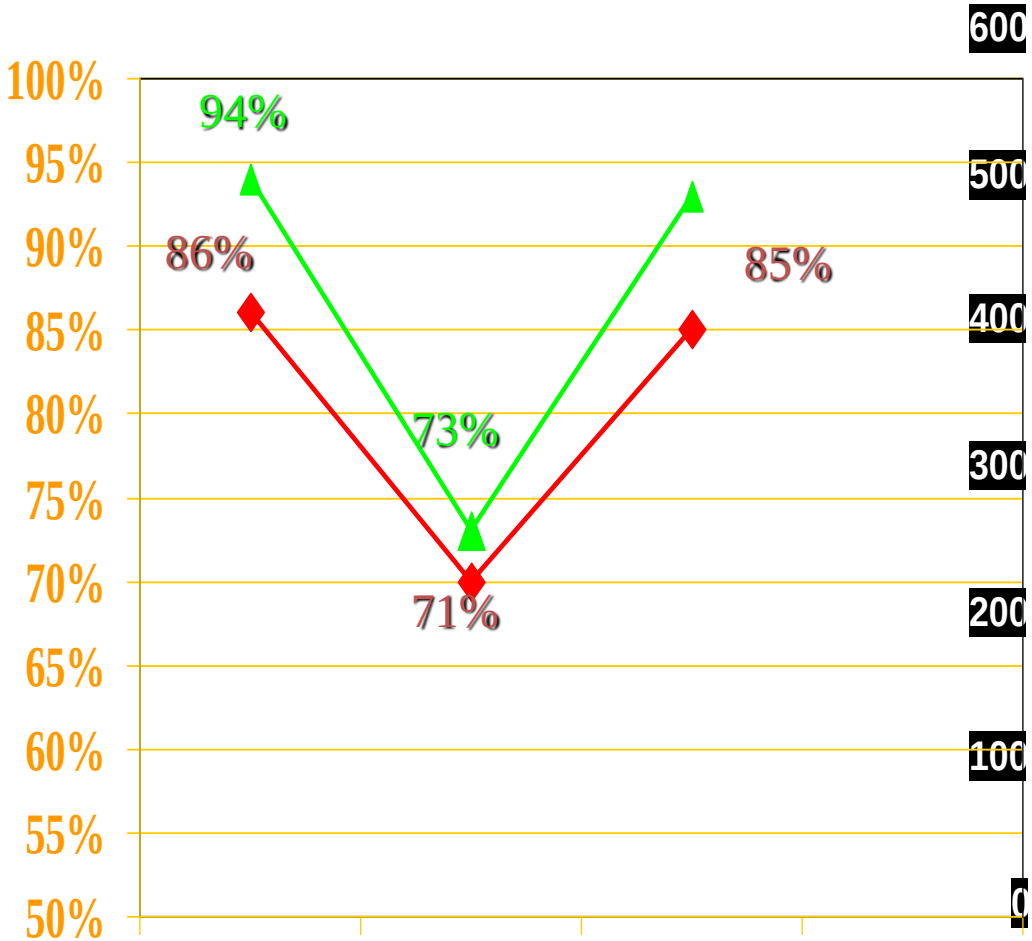
11 FONTAN PATIENTS TREATED WITH SILDENAFIL AND/OR BOSENTAN

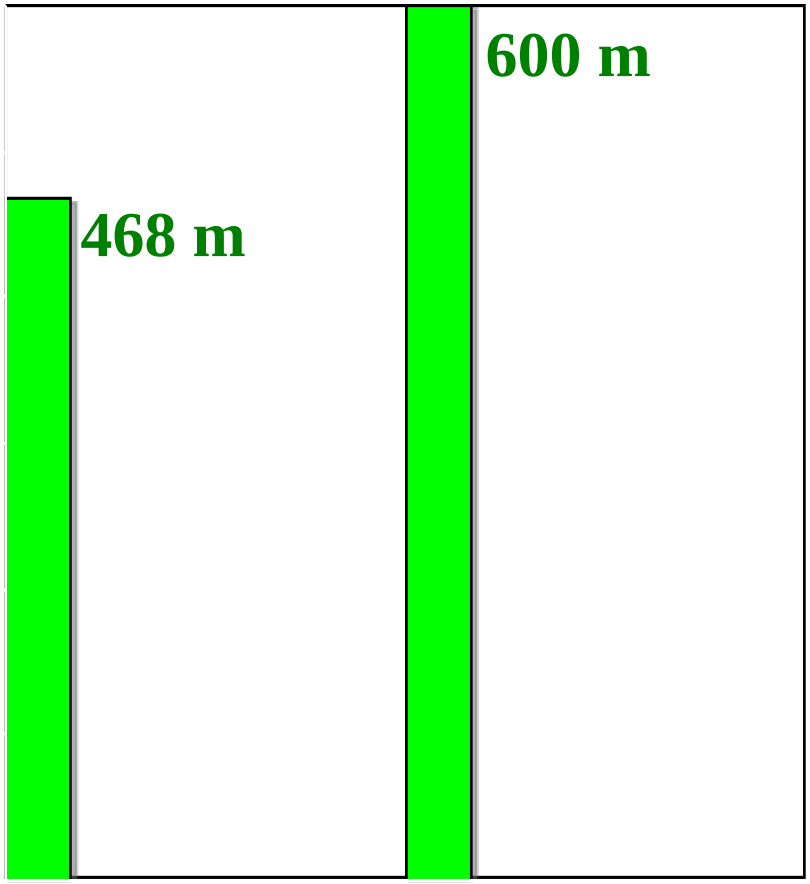
EXPERIENCE
PEDIATRIC CARDIOLOGY
" LA PAZ"
CHILDREN´S HOSPITAL
2006-2011

	2 PLASTIC BRONCHITIS	4 PLE	1 ASCITIS	3 SINGLE LUNG FENESTRATED FONTAN	1 BEFORE PARTIAL FENESTRATION CLOSURE
❖ **Resolution**	0/2	0/4	1/1	↑ O2 sat 6MWDT	↓ RVP and mPAP
❖ **NYHA Improvement**	0/2	2/4	1/1	2/3	1/1
❖ **Follow-up and outcome**	9 mo.	3.5 y. S+B NYHA III → II 3.5 y. S+B NYHA II → I	6 mo. IV → I	3 y. II → I 2 y. III → I 2 y. II → II	3 y. B II → I
	2/2 Dead In waiting list or at heart tx	**1/4 Dead** 6 mo. (S)	0/3 dead	0/3 dead	0/3 dead

9 years
HLHS+NORWOOD.
FENESTRATED FONTAN TO RPA
Disconnected LPA small left Bt Shunt

SILDENAFIL
1 mg/kg/8 horas





SEPT. 2008

MARCH 2009

Hospital Universitario La Paz

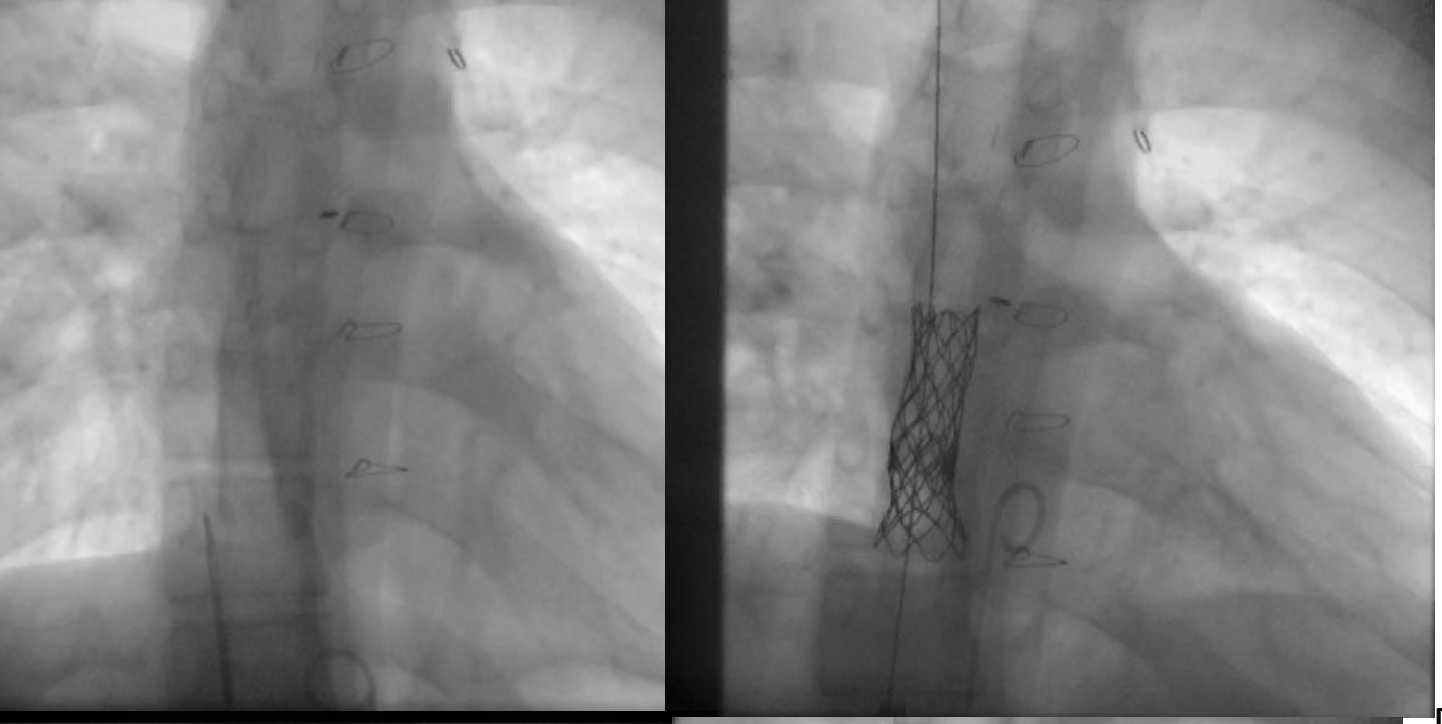
SaludMadrid

Comunidad de Madrid

PEDIATRIC CARDIOLOGY
" LA PAZ"
CHILDREN´S HOSPITAL

FONTAN PATIENTS
BEFORE TREATMENT WITH SILDENAFIL AND/OR BOSENTAN…

- ❑ FUNCTIONAL CLASS: ANAMNESIS, PHYSICAL EXAM, QUALITY OF LIFE
- ❑ CAREFUL READING OF THE CLINICAL RECORDS…
  (PREVIOUS CATHs, SURGERIES, INTENSIVE CARE, periods of excessive blood flow, Restrictive ASD if MA, …
- ❑ REVIEWING OLD CATHS movies…
- ❑ COMPLETE BLOOD TESTING: SEROLOGIES, thyroid function, HEPATIC FUNCTION, microalbuminuria,…
- ❑ EXERCISE TESTING OR 6MWDT
- ❑ MRI: cardiac output, ventricular function, VALVAR REGURGITATION, venous or arterial collaterals, conduit STATUS, residual shunts,…
- ❑ CARDIAC CATHETERIZATION:
  DG: MPAP, VENTRICULAR DIASTOLIC PRESSURE, TPG AND PVRI
  INTERVENTIONS: collateral embolization, conduit stenting, etc,…



17 años
Fontan
extracardiaco

Cuadro ascitis

Implante stents
En el conducto
Diametro de 8 a 15 mm

2m despues
Persiste ascitis, edemas

SILDENAFILO

4m: resolucion

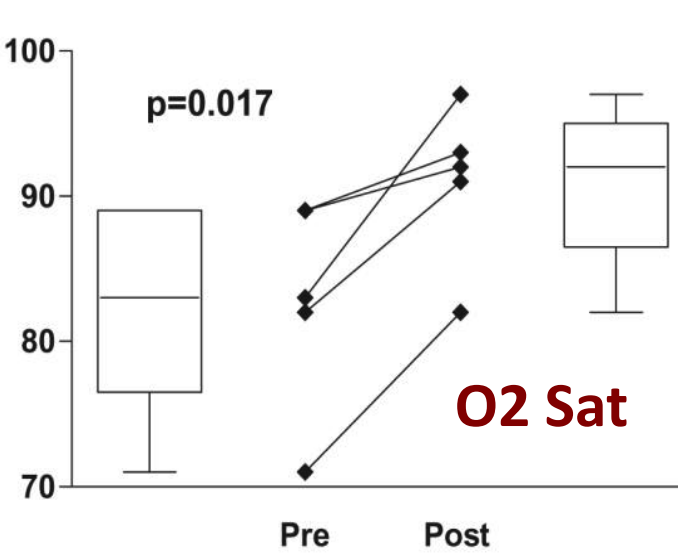
Sildenafil Increases Systemic Saturation and Reduces Pulmonary Artery Pressure in Patients with Failing Fontan Physiology

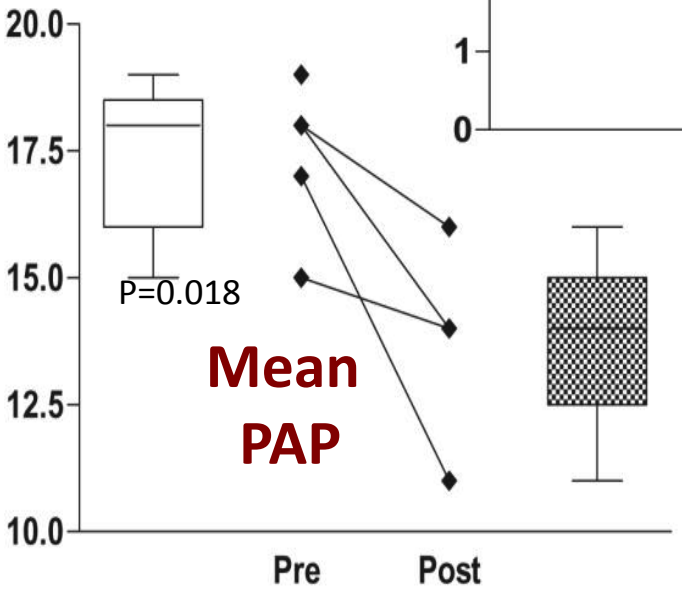
Retrospective review

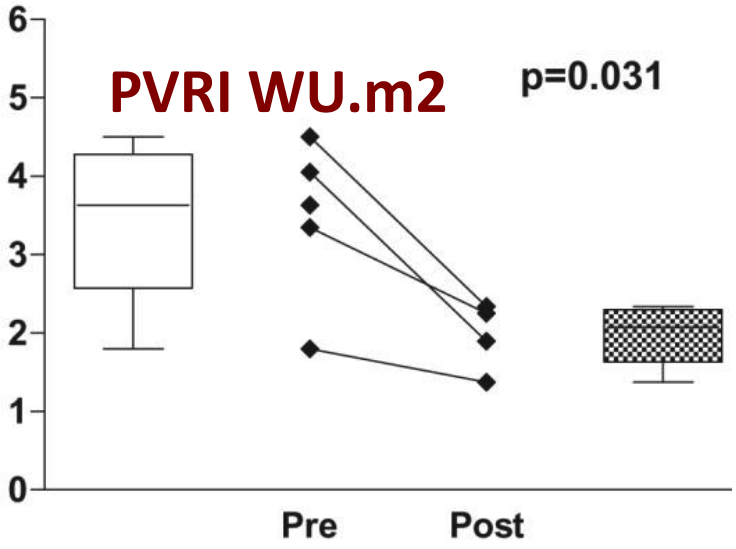
Congenit Heart Dis. 2009 April ; 4(2): 107–111

6 Fontan patients cyanosis /pleural effusion (EARLY failures)

who had been treated with sildenafil 1 mg/kg t.i.d for 6 -51 months

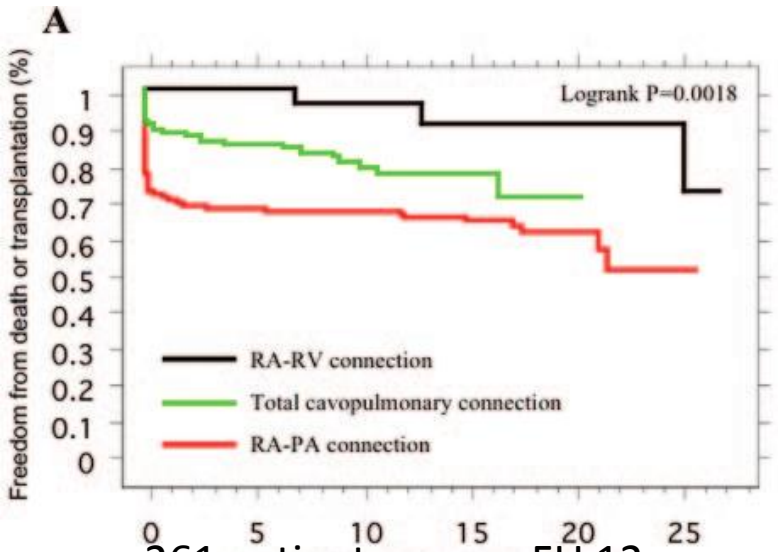




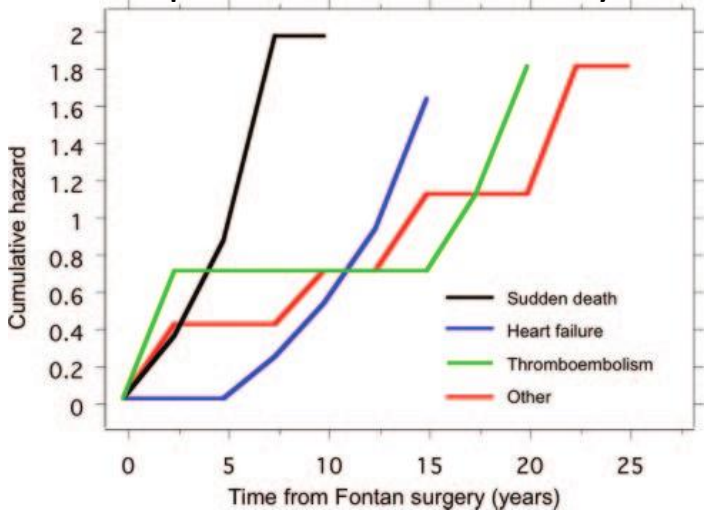


Pulmonary Vasodilators in the Failing Fontan...

What do we mean by “failing Fontan”?



261 patients, mean FU 12 years



- Protein-losing enteropathy
- Plastic Bronchitis.

- Recurrent arrythmias
- Heart failure
- NYHA III or IV
- Sudden death
- Hepatic disease
- Recurrent thromboembolic events
- Increasing cyanosis
- Deceased tolerance to exercise

Long-Term Survival, Modes of Death, and Predictors of Mortality in Patients With Fontan Surgery

Circulation 2008, Children´s Boston

The effect of bosentan in patients with a failing Fontan circulation

Multicentric open label, non-controlled study.

10 patients with a "Failing Fontan" circulation (7 fenestrated)

Baseline,
During
After 16 w. Bos.

- ❖Saturations of oxygen at rest and during exercise (6MWDT)
- ❖Questionaire quality of life

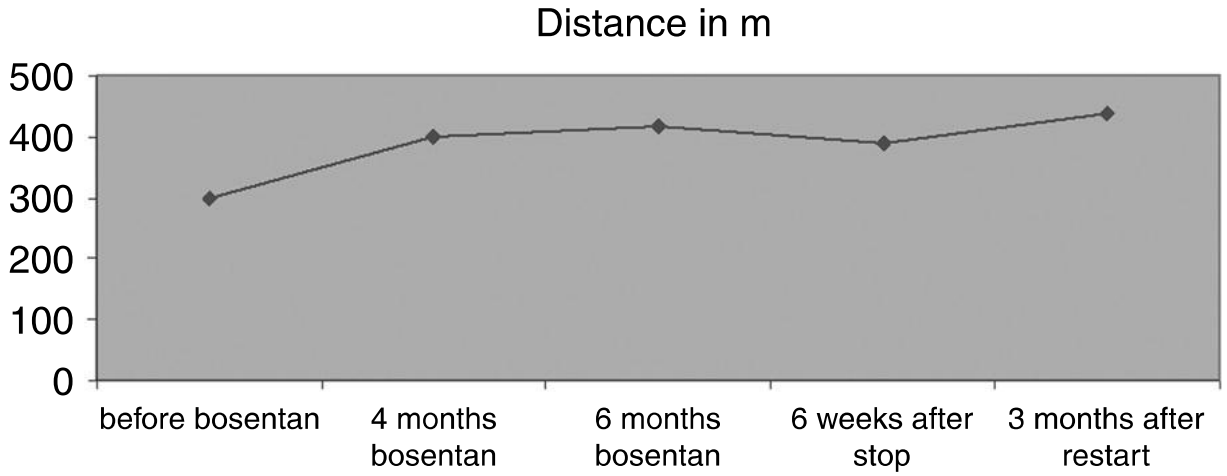
INCLUSION CRITERIA

- Sat O2 < 90% at rest or exercise
- Chronic low C.O. with ⇧ SVP
  (hepatomegaly /high YVP
  pleural effusions / ascites)
- Protein-losing enteropathy,
- Exercise intolerance (WHO II-IV)

The effect of bosentan in patients with a failing Fontan circulation

- The changes in saturations of oxygen, exercise performance, **did not reach statistical significance *for the whole group*.**

- Saturations and/or exercise capacity *improved in 5/10 patients,* which deteriorated again when the drug was discontinued.

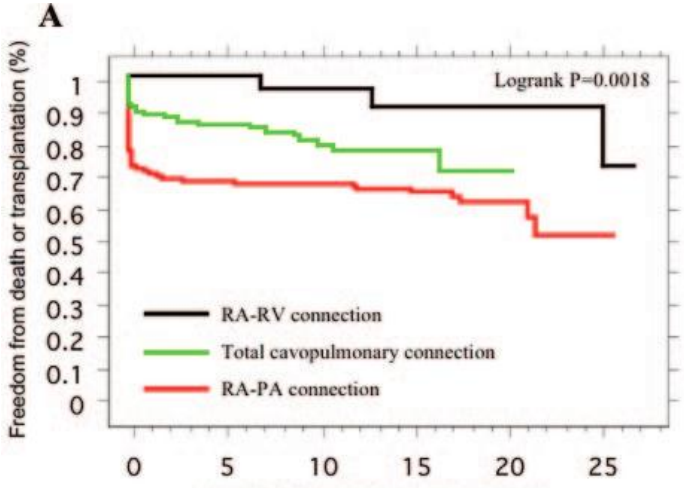


¿Cual sería el momento

de comenzar a tratar la
Enfermedad Vascular Pulmonar
en el Fontan?

Pulmonary Vasodilators in the Failing Fontan...

What do we mean by “failing Fontan”?



261 patients, mean FU 12 years

IF PULMONARY VASODILATORS COULD HAVE A BENEFFICIAL EFFECT…
WHY SHOULD WE WAIT UNTIL PLE OR PLASTIC BRONCHITIS?...

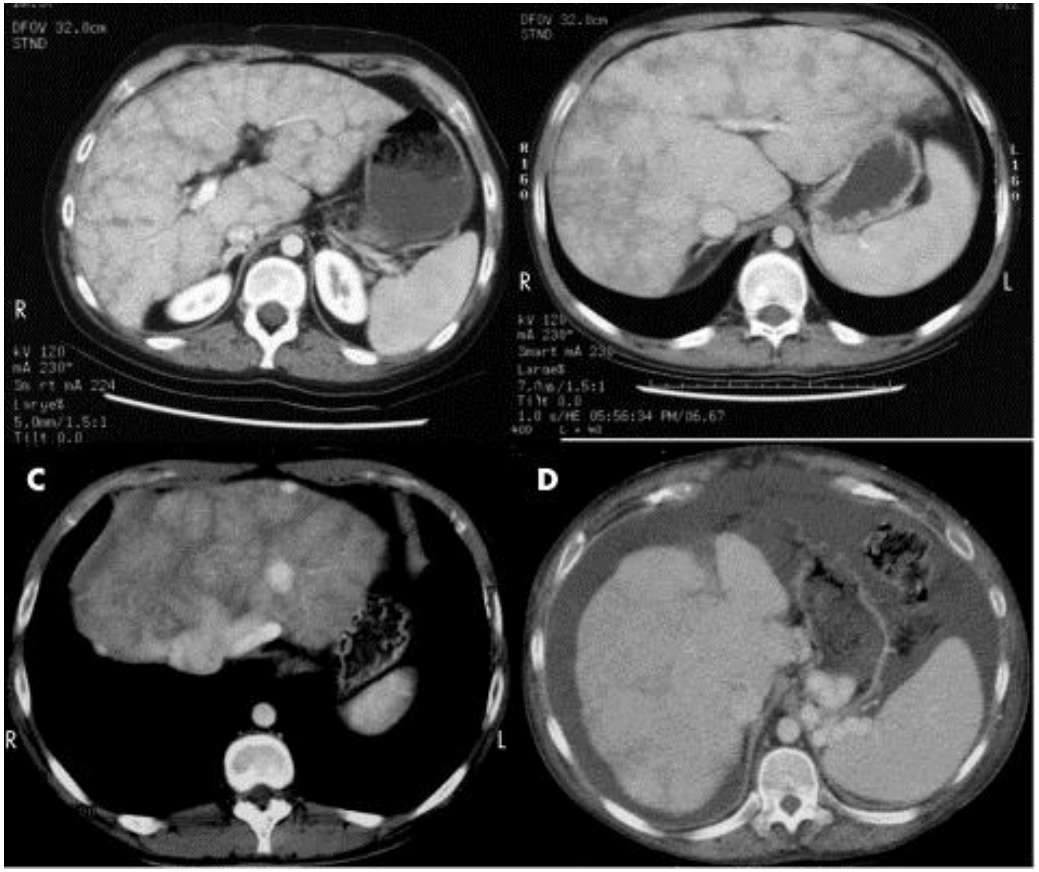
- Protein-losing enteropathy
- Plastic Bronchitis.

- Recurrent arrythmias
- Heart failure
- NYHA III or IV
- Sudden death
- Hepatic disease
- Recurrent thromboembolic events
- Increasing cyanosis
- Deceased tolerance to exercise

Long-Term Survival, Modes of Death, and Predictors of Mortality in Patients With Fontan Surgery

Circulation 2008,

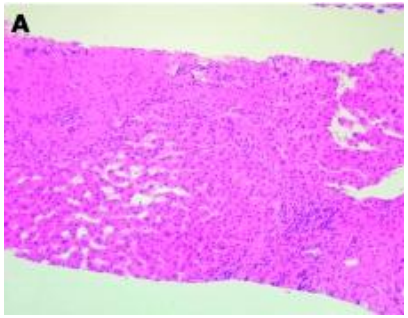
Children´s Boston



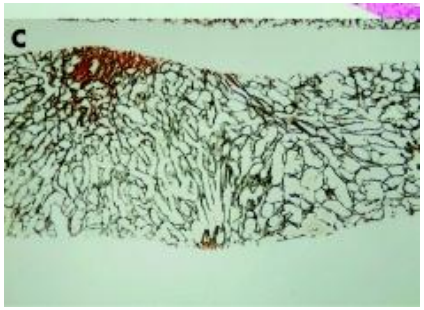
Hepatic changes in the failing Fontan circulation

Heart. 2007 May; 93(5): 579–584

12 patients atrio-pulmonary Fontan referred for conversion, due to:
PLE
Persistent Arrythmia
Exercise intolerance
Peripheral edemas



All: Parenchymal atrophy and sinusoidal dilatation

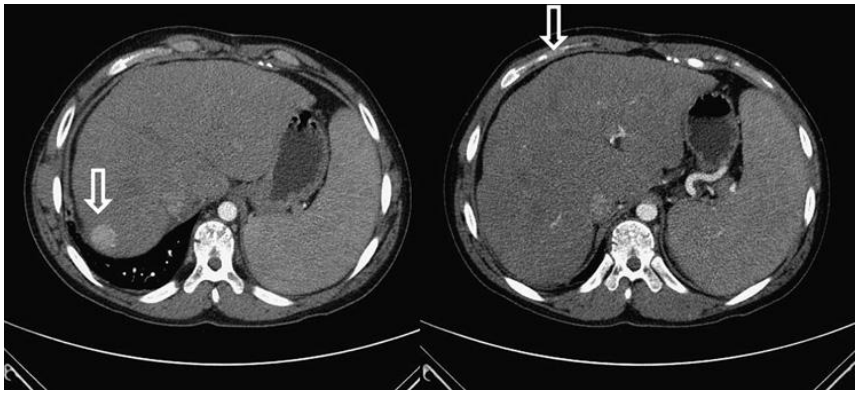


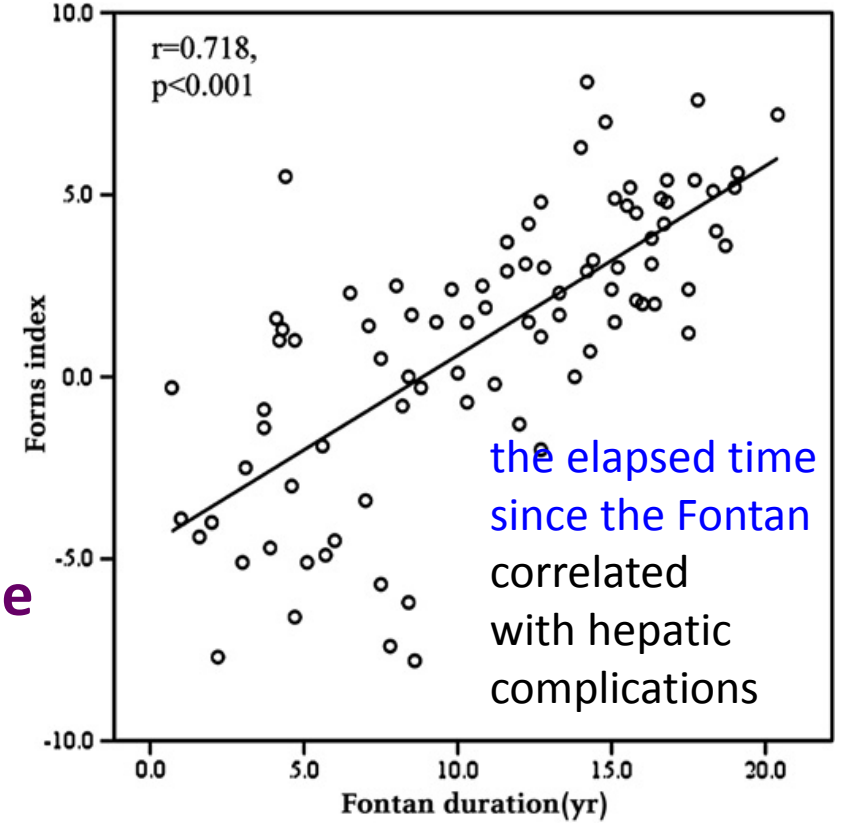
7/12 patients, cardiac cirrhosis

The mean (SD) duration since the initial Fontan procedure was 14.1 (5.0) (range 6.9–26.4) years.

Late hepatic complications after Fontan operation; non-invasive markers of hepatic fibrosis and risk factors

Heart 2010;**96**:1750—1755.





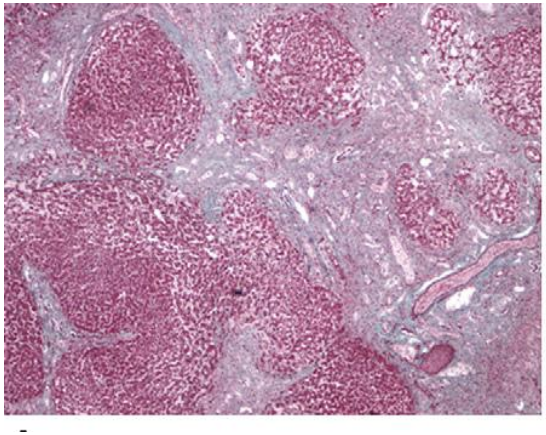
the elapsed time since the Fontan correlated with hepatic complications

30-60% of Fontan patients may have hepatic complications by the time they become teenagers….

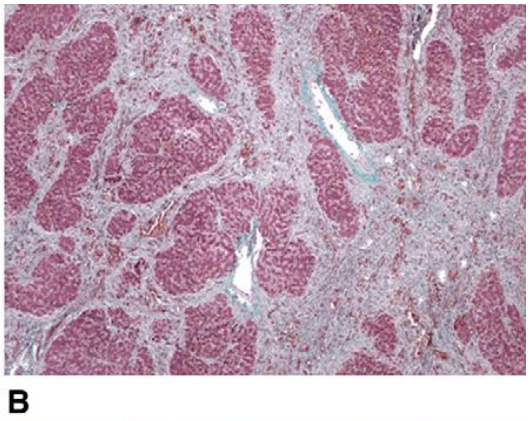
Hepatomegaly, splenomegaly, abnormal transaminases, elevated GGT, elevated bilirubin and coagulopathy, thrombocytopenia

platelet count, GGT, age and cholesterol AST/ALT ratio prothrombin time

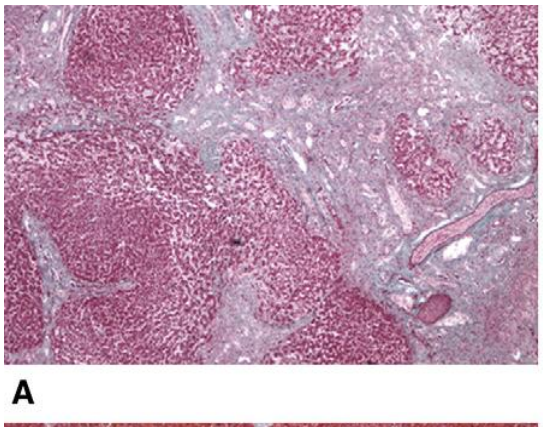
Hepatic pathology may develop before the Fontan operation in children with functional single ventricle: An autopsy study



portal fibrosis only 1 day after the Fontan operation



Portal-based cirrhosis 4 years after the Fontan procedure



Significant portal fibrosis in a long-term survivor after the Fontan (16 years)

Nephropathy in patients after Fontan palliation.

- Int J Cardiol. 2009 Feb 20;132(2):244-7.

- Nine patients **(43%) had a pathologic MICROALBUMINURIA** (>20 microg/mg).

- **Strong positive correlations were found between the MCR and pre- and post-Fontan PVR** (r=0.51, p<0.05 and r=0.61, p=0.02 respectively).

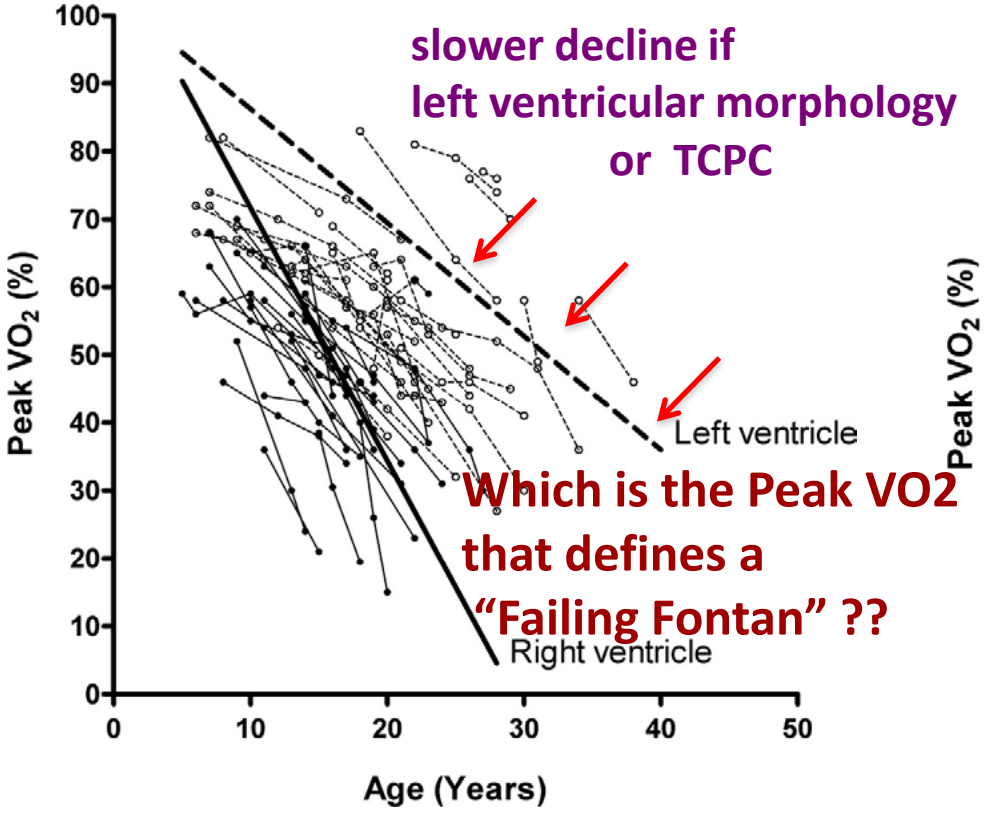
- No significant differences between the MCR-normal and MCR-abnormal groups in age, gender, type of single ventricle, type of the procedure, age at or time since Fontan operation
- Moreover, no patient receiving a lisinopril-equivalent dose greater than 0.4 mg/kg/day had pathologic microalbuminuria

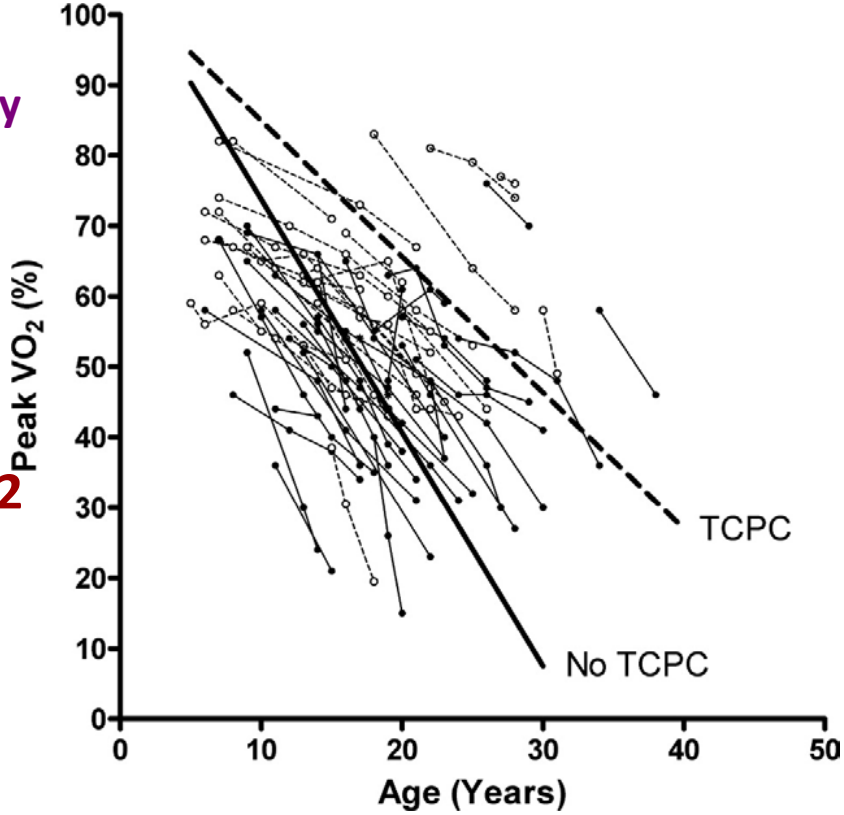
Natural History of Exercise Capacity After the Fontan Operation: A Longitudinal Study

Giardini et al (Ann Thorac Surg 2008;85:818–22)

Longitudinal study of 53 Fontan patients with cardiopulmonary exercise tests

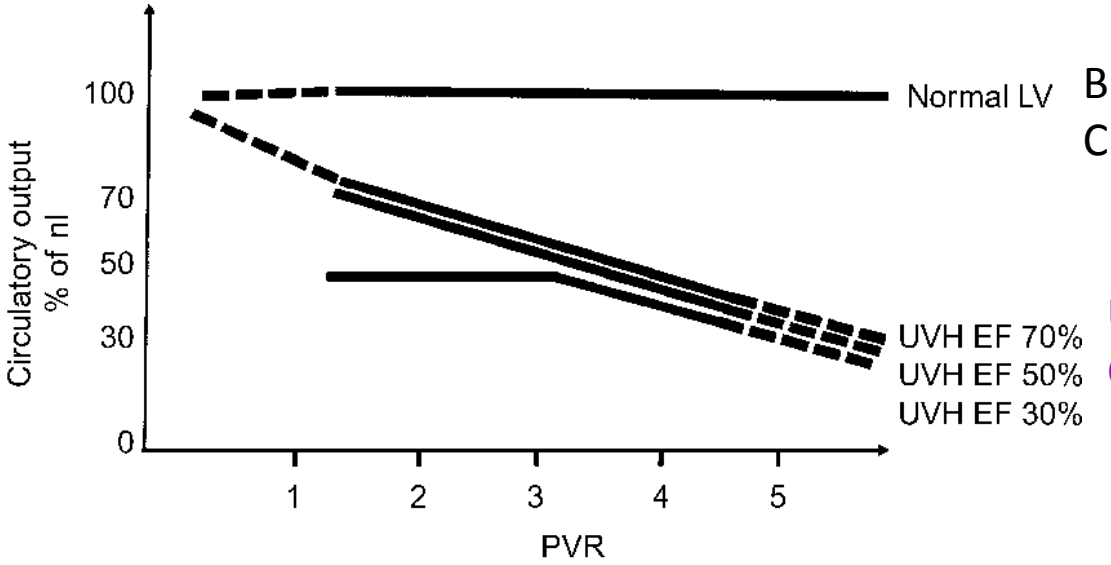
Exercise capacity progressively declines….





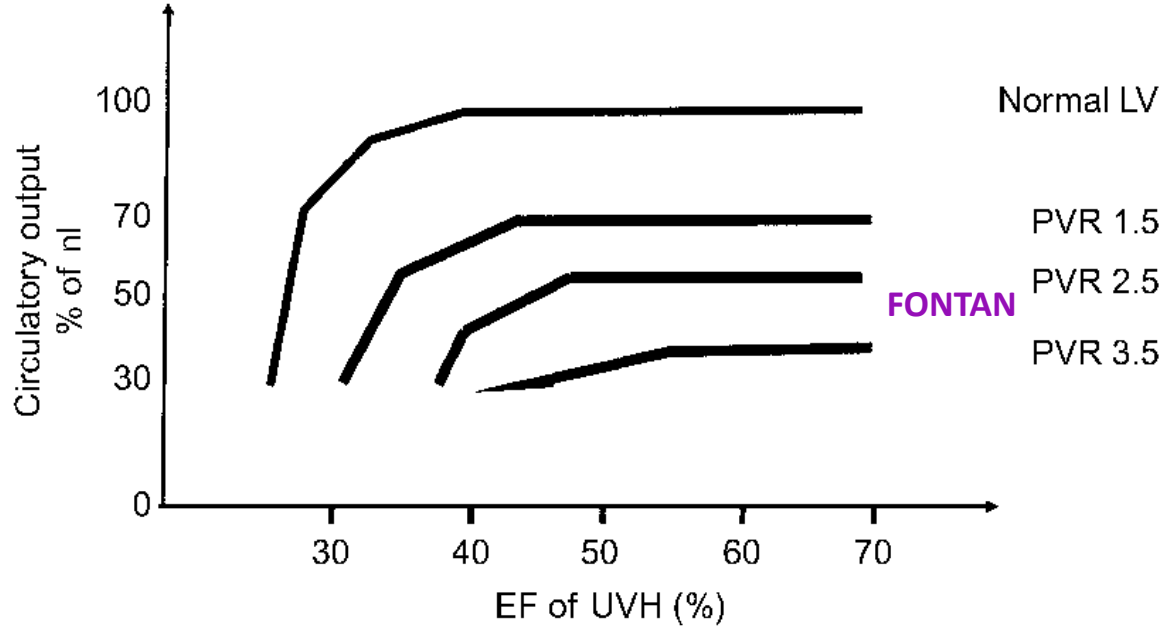
The Fontan circulation: who controls cardiac output?

M. Gewillig et al. / Interactive CardioVascular and Thoracic Surgery 10 (2010) 428-433



BIVENTRICULAR CIRCULATION

UNIVENTRICULAR CIRCULATION

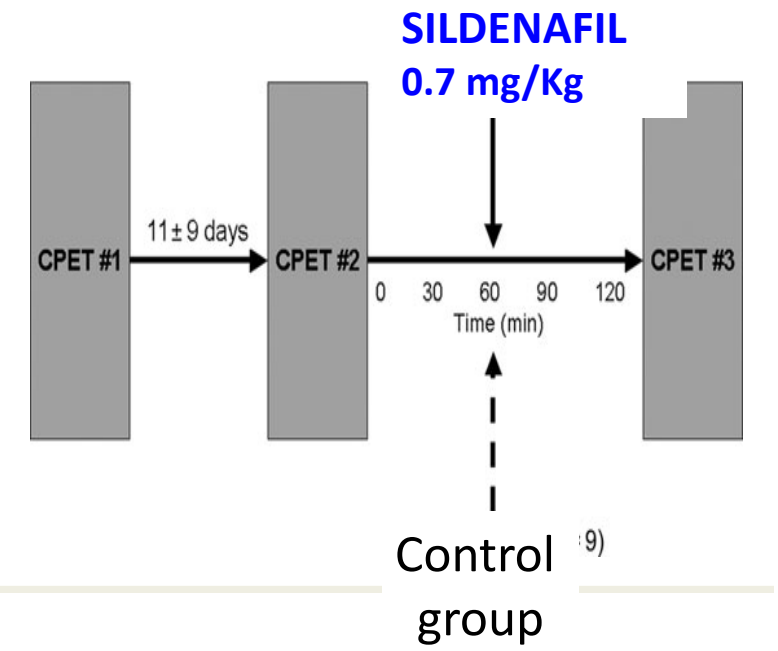


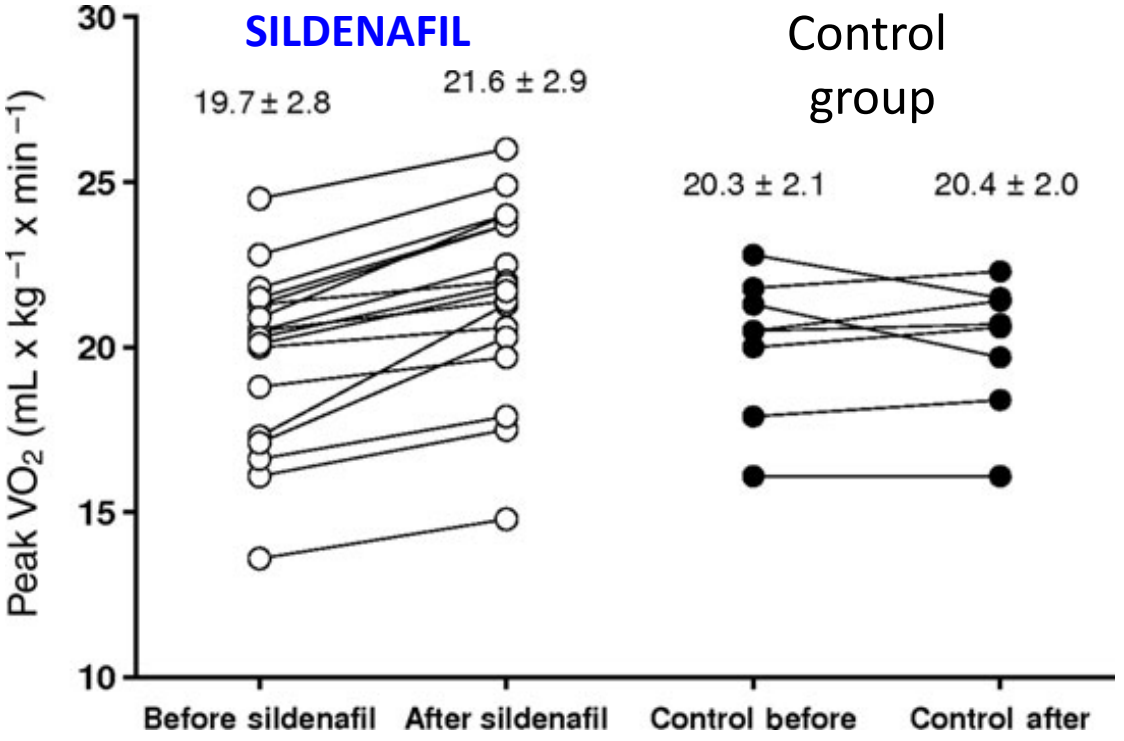
Effect of sildenafil on haemodynamic response to exercise and exercise capacity in Fontan patients

Giardini A, et al European Heart Journal (2008) 29, 1681–1687

- Prospective study 27 Fontan adult patients (age 22.8 + 4.9 years)

- **Acute effect** of a single dose of sildenafil in CPET **(peak VO2)**



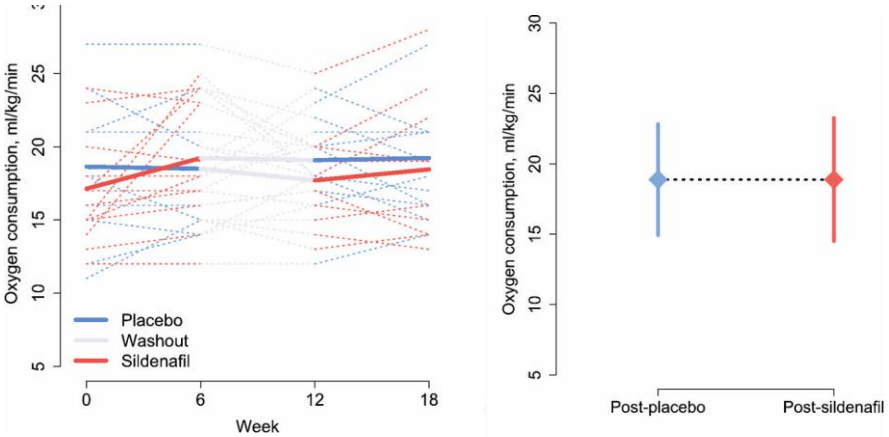


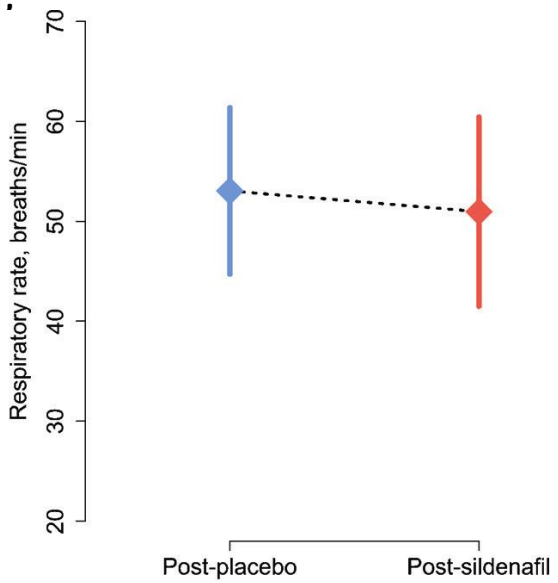
Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

Impact of Oral Sildenafil on Exercise Performance in Children and Young Adults After the Fontan Operation: A Randomized, Double-Blind, Placebo-Controlled, Crossover Trial

Circulation. 2011;123:1185-1193

- no significant effect of sildenafil on cardiopulmonary measures at rest
- **no improvement in VO2 max <u>after a 6-week sildenafil</u>.**
- sildenafil improved ventilatory efficiency and exercise performance at the anaerobic threshold but not VO2max

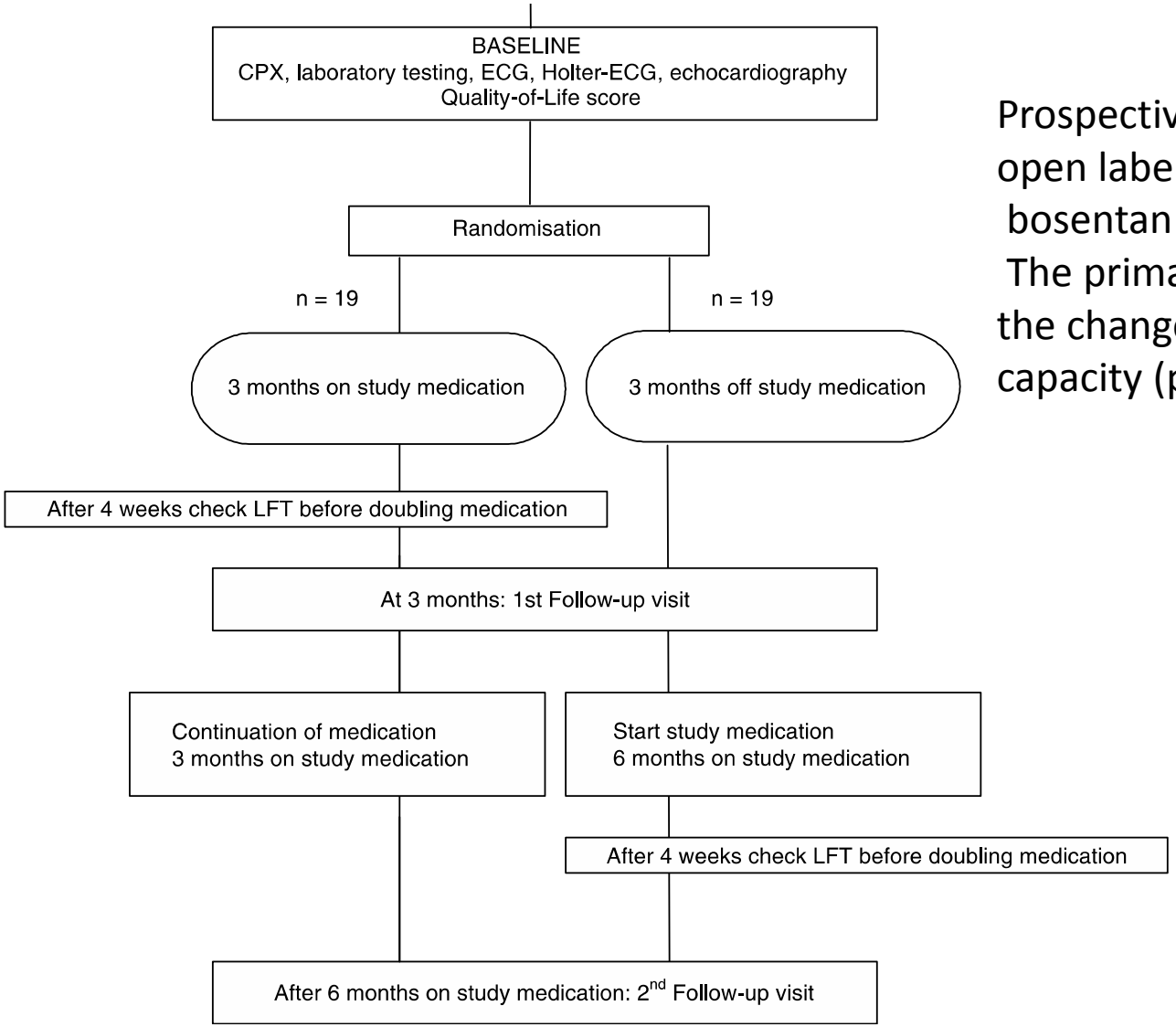




- **Improvement in oxygen consumption at the anaerobic threshold** and the ***<u>statistically significant improvement in</u>*** 2 subgroups (those with single left ***<u>ventricular or mixed ventricular morphology and those with BNP > 100 pg/mL</u>***

Rationale and design of a trial on the role of bosentan in Fontan patients: Improvement of exercise capacity?

M.J. Schuuring et al. / Contemporary Clinical Trials 32 (2011) 586–591

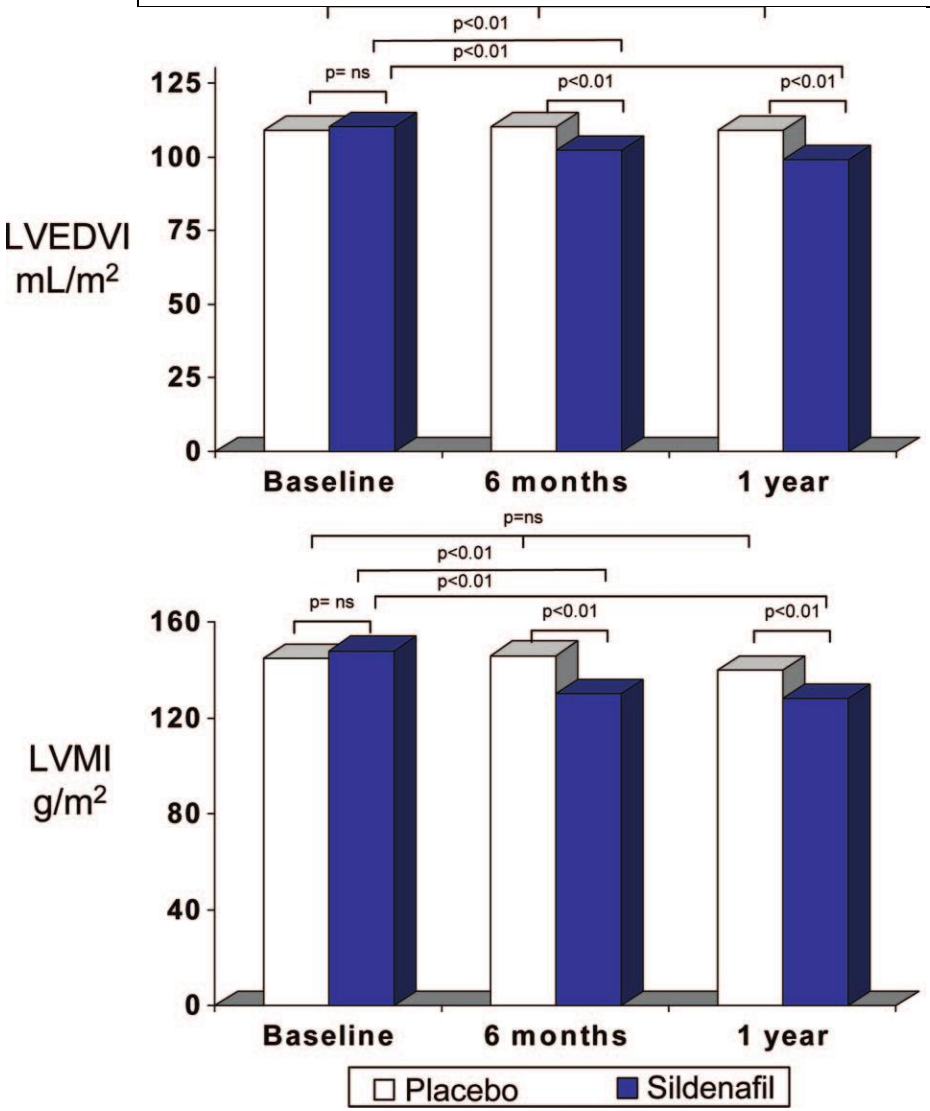


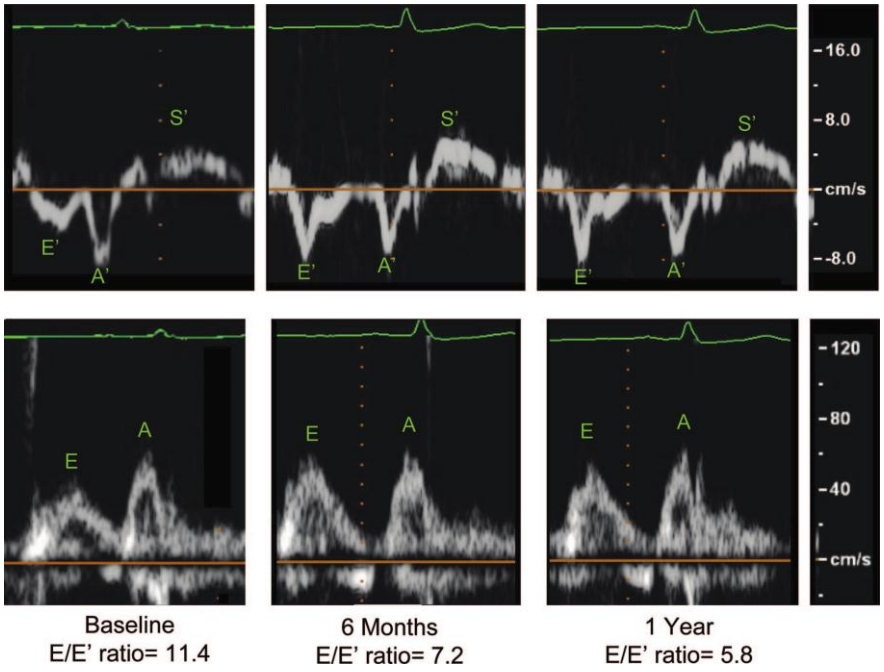
Prospective, multicenter, randomized open label trial to study the effect of bosentan inadult Fontan patients. The primary endpoint will be the change in maximum exercise capacity (peak V'O2).

PDE5 Inhibition With Sildenafil Improves Left Ventricular Diastolic Function, Cardiac Geometry, and Clinical Status in Patients With Stable Systolic Heart Failure : Results of a 1-Year, Prospective, Randomized, Placebo-Controlled Study

Guazzi M

Circ Heart Fail. 2011;4:8-17.





Baseline
E/E' ratio= 11.4

6 Months
E/E' ratio= 7.2

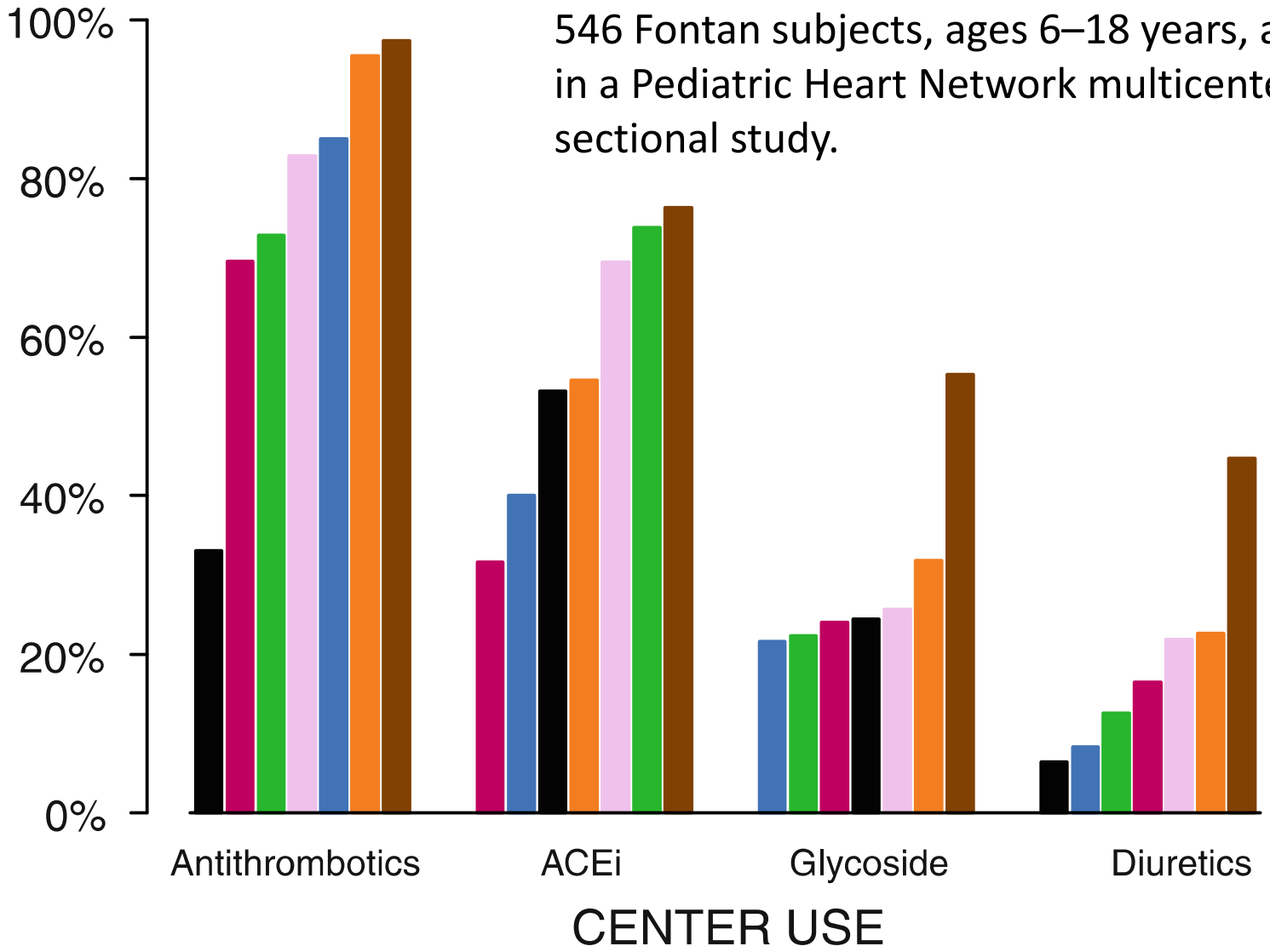
1 Year
E/E' ratio= 5.8

Pediatr Cardiol (2010) 31:1219–1228
DOI 10.1007/s00246-010-9807-5

ORIGINAL ARTICLE

The Fontan Patient: Inconsistencies in Medication Therapy Across Seven Pediatric Heart Network Centers

546 Fontan subjects, ages 6–18 years, assessed in a Pediatric Heart Network multicenter cross-sectional study.



Enalapril Does Not Enhance Exercise Capacity in Patients After Fontan Procedure

Circulation. 1997;96:1507-1512

Randomized, placebo trial 18 Fontan Pts (6-19 years after Fontan)

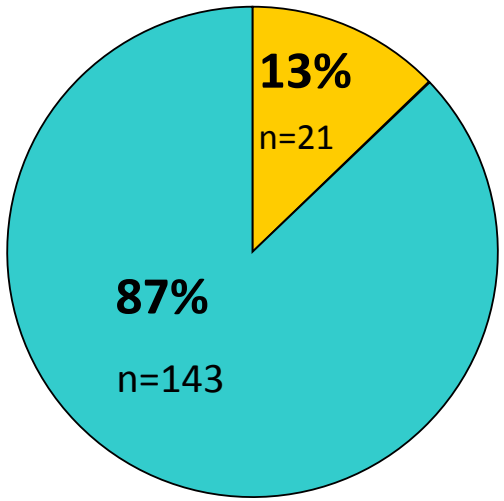
10 weeks of enalapril /placebo

	Enalapril	Placebo
Heart rate at rest, bpm	88.9±13.8	86.9±12.6
Maximum heart rate, bpm	156±22	156 ±21
Oxygen saturation at rest	97±4	96±5
Maximum oxygen saturation	95±5	94±4
Cardiac output at rest, L/min	2.3±0.7	2.3±0.6
Maximum cardiac output, L/min	4.9±1.6	5.3±1.6
Cardiac index at rest, L · min-1 · m2	1.7±0.3	1.7±0.3
Maximum cardiac index, L · min-1 · m2	3.5±0.9	3.8±0.9
Minute ventilation (E) at rest, L/min	13.5±4	13.0±5
Maximum minute ventilation (E), L/min	57.5±17	55.4±19
Oxygen consumption (VO2) at rest, mL · kg-1 · min-1	5.3±1	5.2±1
Maximum oxygen consumption (VO2), mL · kg-1 · min-1	18.3±9	20.5±7
E/VO2 at rest	62.2±13	62.5±17
Exercise time, min	6.4±2.6	6.7±2.6

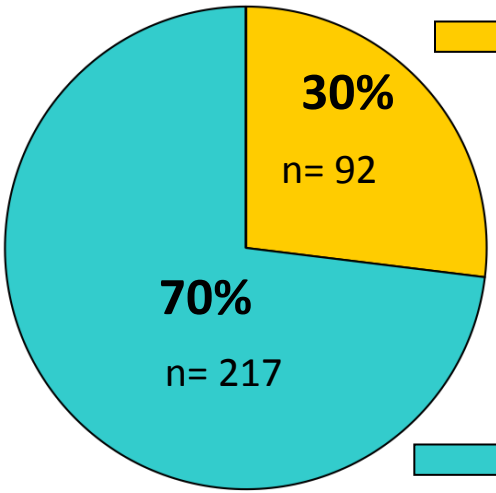
REHIPED

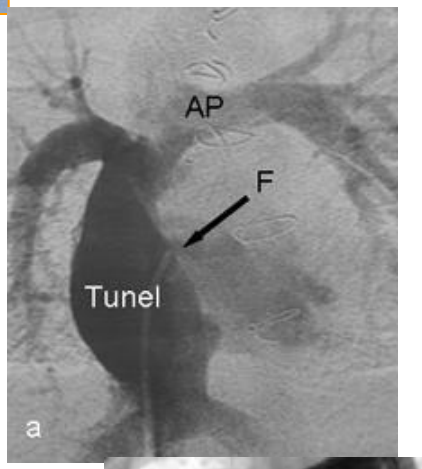
REgistro de pacientes con HIpertensión Pulmonar PEDiátrica

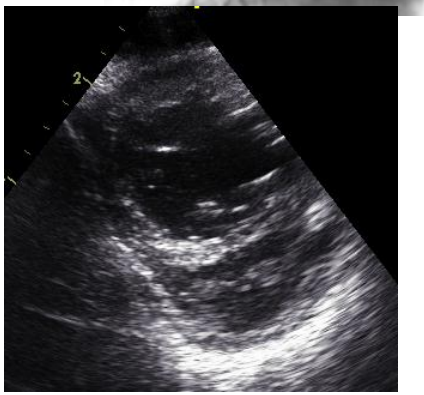
April 2011
164 pts



April 2012
310 pts





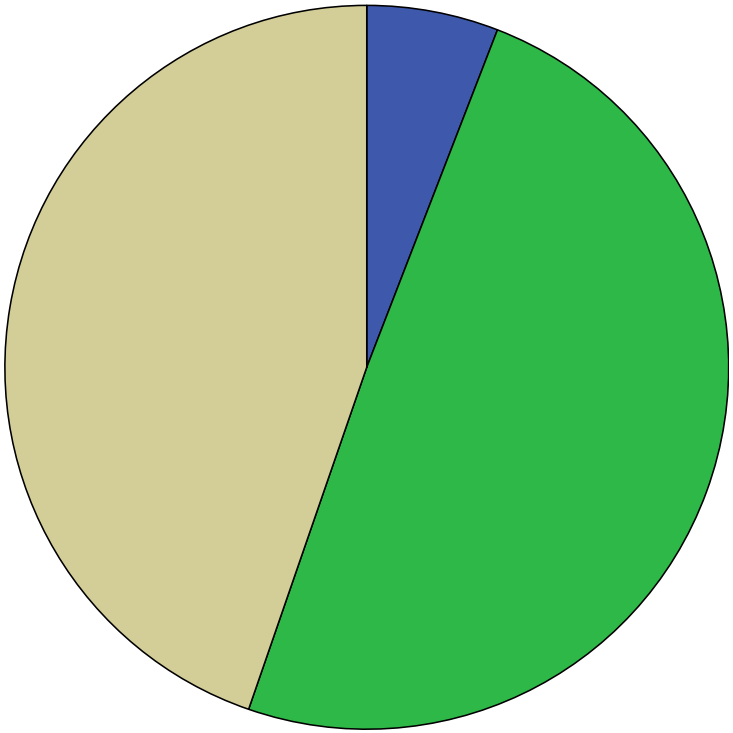


REHIPED

REgistro de pacientes con HIpertensión Pulmonar PEDiátrica

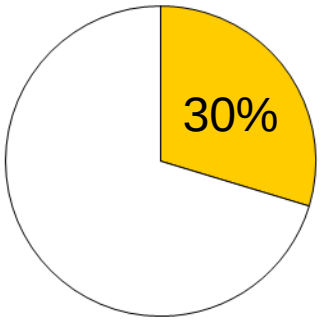
AGE AT INCLUSION

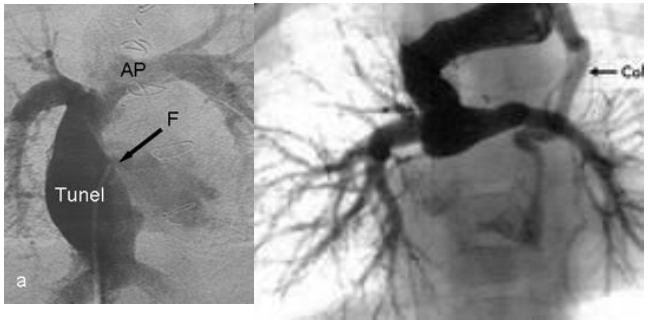
- 0-2 years
- 2-8 years
- 8-18 years

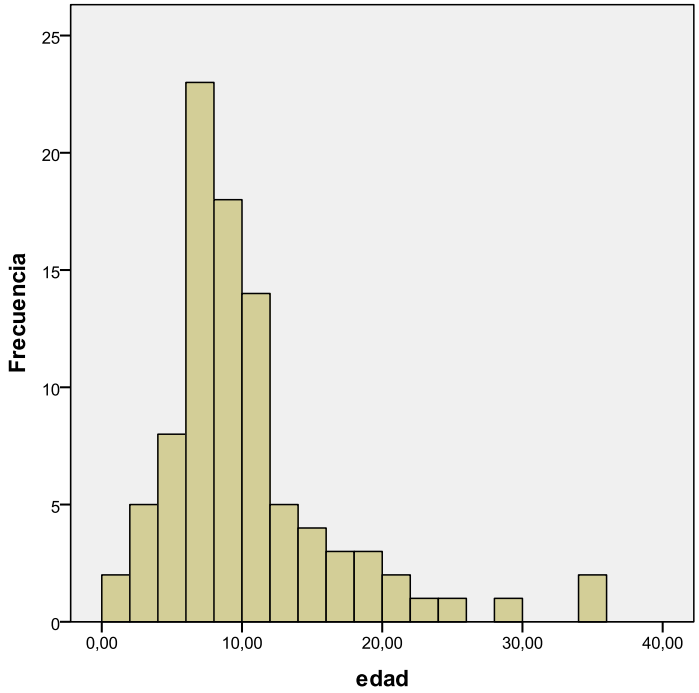


Single ventricle physiology mPAP < 25 mmHg

(92/311)







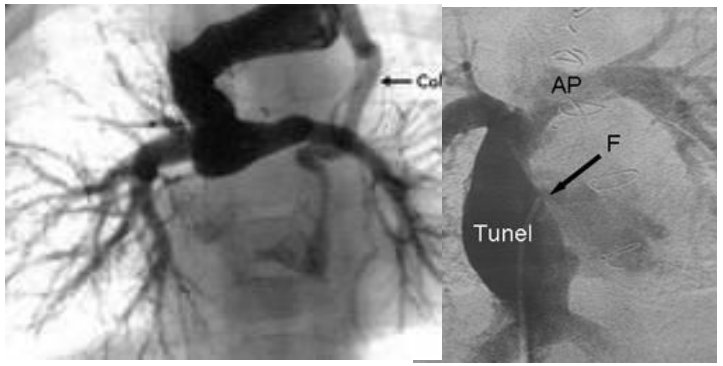
REHIPED
REgistro de pacientes con HIpertensión Pulmonar PEDiátrica

HEMODYNAMIC DATA

86 GLENN/FONTAN: 98% cath data (84/86)

catheterized

	mean ± SD
Systolic Pulmonary pressure / Systolic Aortic pressure (%)	**23 ± 16**
Mean Pulmonary pressure (MPAP) mmHg	15 ± 7
CARDIAC INDEX (l/min/m^2)	4 ± 1.9
PVRI (WU.m^2)	1.4 ± 1
PVRI /SVRI	0.14 ± 0,13
GRAD. TRANSPULMONAR	**5.5 ± 7.5**
PWP	**10.4 ± 3.2**



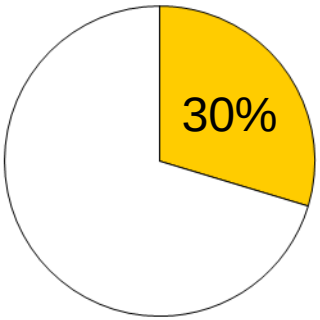
REHIPED

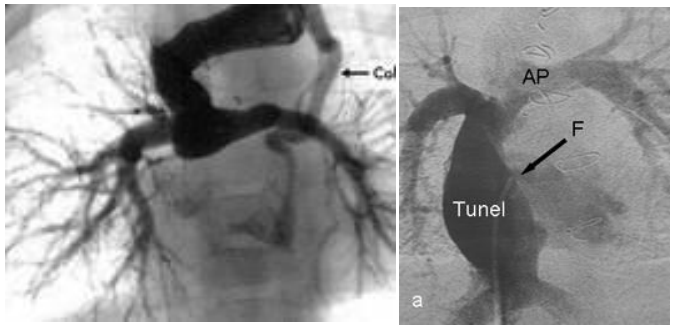
REgistro de pacientes con HIpertensión Pulmonar PEDiátrica

REASON TO START TREATMENT

Single ventricle physiology
mPAP < 25 mmHg

(92/311)





Protein-losing enteropathy or plastic bronchitis	**16.5**
Improve O2 sat. In single lung /fenestrated fontan	**13**
Decrease pvri before fontan / glenn surgery	**35**
Decrease pvri before fenestration closure fontan	**19**
Improve 02 Sat in Glenn	**23**
Improve Functional Class Fontan	**24**
Growth of PA before Glenn/Fontan surgery	**16.5**

%

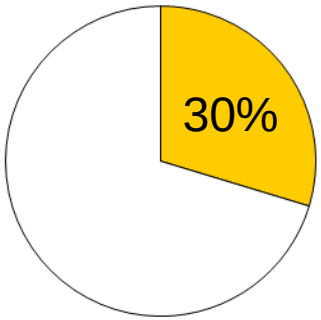
REHIPED

REgistro de pacientes con HIpertensión Pulmonar PEDiátrica

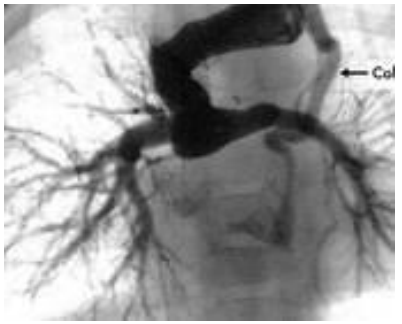
Specific TREATMENT

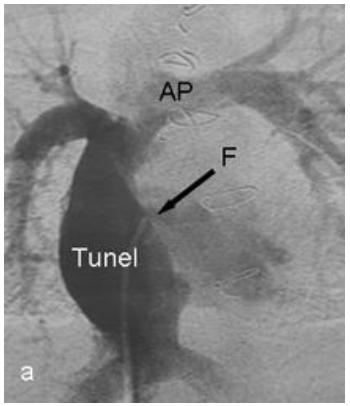
Single ventricle physiology
mPAP < 25 mmHg

(92/311)



DRUG	n
SILDENAFIL	71
BOSENTAN	28
PROSTANOIDS	2





QUESTIONS FOR THE FUTURE:

- WHEN SHOULD WE START vasodilator therapy in Fontan patients?

NYHA III OR IV?
PLE?
PB?
STABLISHED Hepatic disease?
Significant exercise intolerance?

OR BEFORE COMPLICATIONS APPEAR?

EVEN BEFORE THE FONTAN SURGERY ??

- WE NEED:
  -- RANSOMIZED TRIALS to determine possible role of sildenafil /bosentan in the management of fontan patients

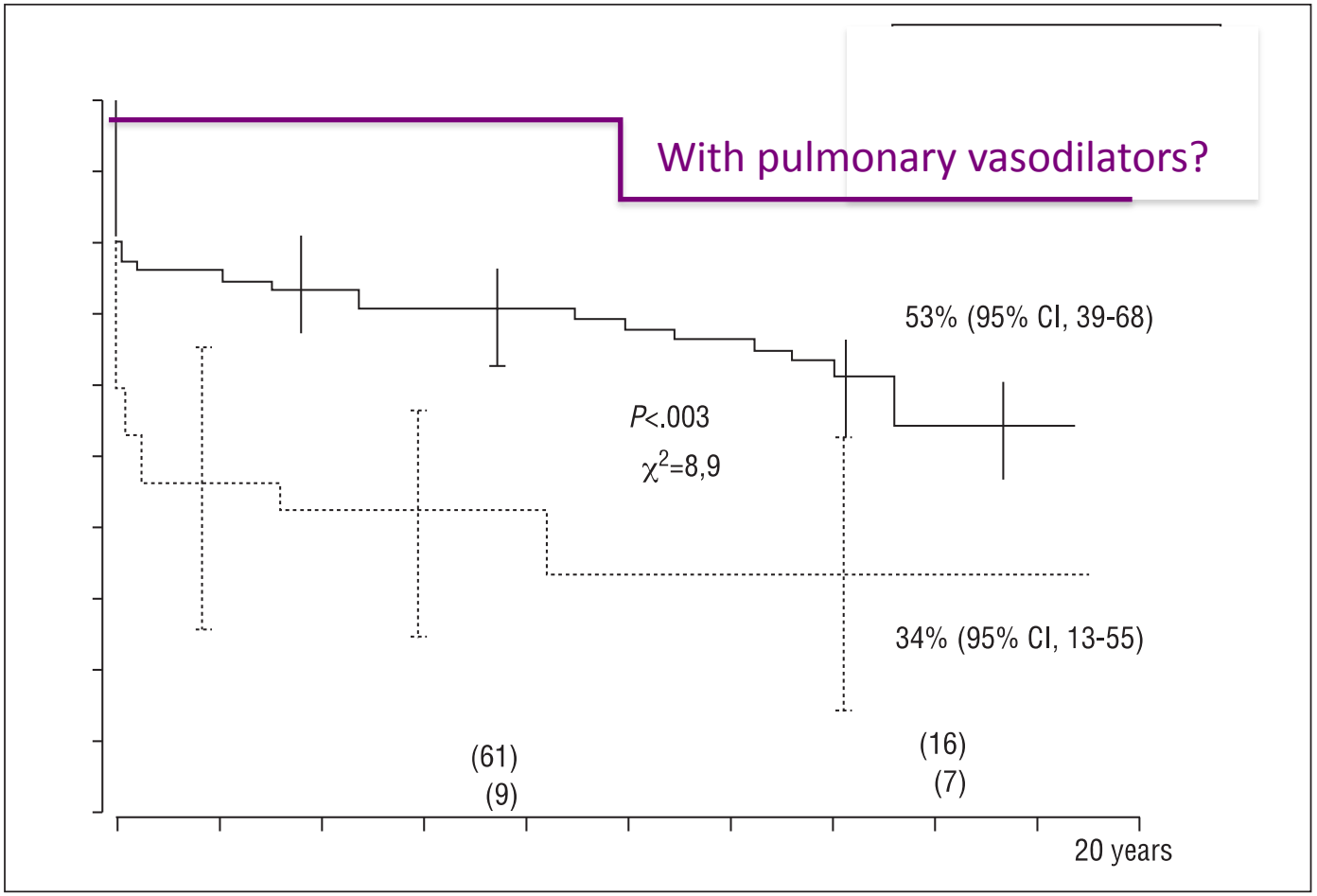
  -- LONGER FOLLOW –UP OF THE REPORTED PATIENTS…

QUESTIONS FOR THE FUTURE:

Freedom From Death or TX

Freedom From hepatic disease?

Freedom From NHYA III?



¿Y Cuando todo falla?

- ❑ RECONVERSIÓN ……

en el Fontan atriopulmonar PRIMERA OPCIÓN
(ANTES DE VASODILATORES PULMONARES)

- ❑ Trasplante cardiaco…..

-- mayor mortalidad inmediata,
-- igual supervivencia tardía
-- PLE predictor riesgo, pero se resuelve
en el 100% de los que sobreviven al trasplante

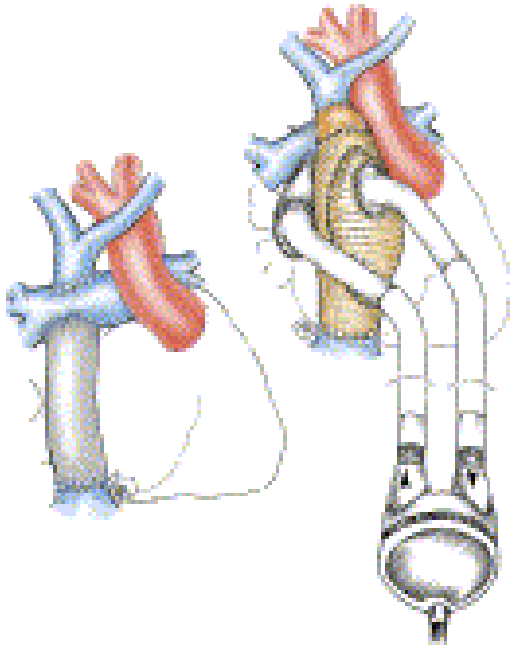
- ❑ TX Hepatocardiaco?

(considerar BIOPSIA HEPÁTICA antes de indicar el TX cardiaco)

- ❑ Asistencias Ventriculares???

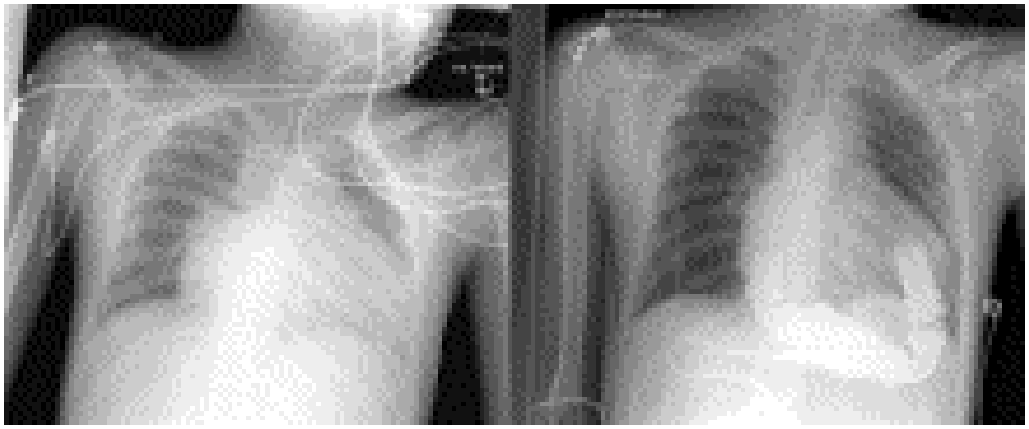
Case report
Right-Sided Univentricular Cardiac Assistance in a Failing Fontan Circulation

The Annals of Thoracic Surgery
Volume 86, Issue 3, September 2008: 1018–1020



A new era: Use of an intracorporeal systemic ventricular assist device to support a patient with a failing Fontan circulation

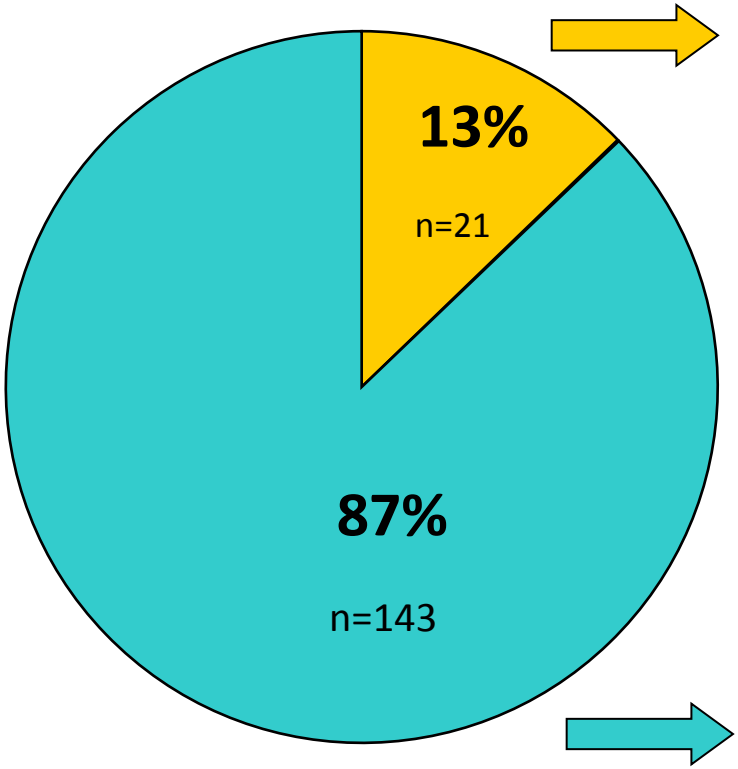
The Journal of Thoracic and Cardiovascular Surgery
Volume 142, Issue 3, September 2011, Pages e138–e140



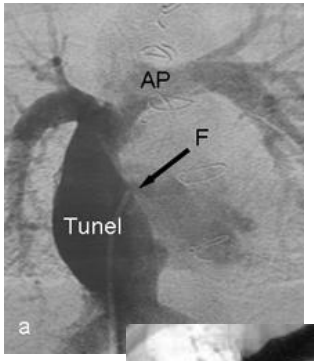
GRACIAS POR SU ATENCIÓN…

REHIPED

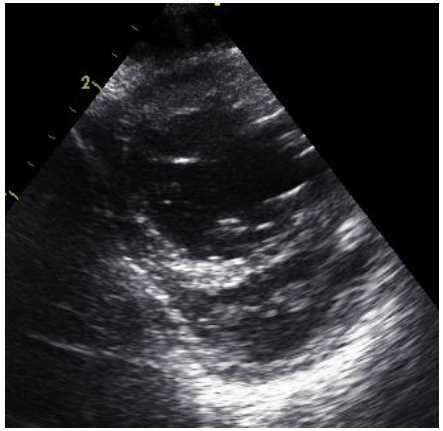
REgistro de pacientes con HIpertensión Pulmonar PEDiátrica



13% patients
receiving PH drugs
SINGLE VENTRICLE PHYSIOLOGY
mPAP < 25 mmHg



87% patients
PULMONARY HYPERTENSION
mPAP > 25 mmHg
And PVRI > 3 WU.m^2



Patients=164

Guidelines and Consensus

A consensus approach to the classification of pediatric pulmonary hypertensive vascular disease: Report from the PVRI Pediatric Taskforce, Panama 2011

Volume 1, Number 2 (April-June 2011)
Pulmonary Circulation
ISSN: 2045-8932
E-ISSN: 2045-8940

1. Prenatal/developmental pulmonary vascular disease
2. Perinatal Pulmonary Vascular Maladaptation (PPHN)
3. Heart Disease
4. Bronchopulmonary dysplasia
5. Isolated Pulmonary Arterial Hypertension
6. Multifactorial in congenital malformation syndromes
7. Lung Diseases
8. Thromboembolic Disease
9. Hypobaric hypoxic exposure
10. Associated with other Disorders

A consensus approach to the classification of pediatric pulmonary hypertensive vascular disease: Report from the PVRI Pediatric Taskforce, Panama 2011

Volume 1, Number 2 (April-June 2011)
Pulmonary Circulation
ISSN: 2045-8932
E-ISSN: 2045-8940

3.3. Pulmonary vascular disease following staged palliation for single ventricle physiology

3.3.1. After stage 1

- *PA banding*
- *modified Norwood*
- *Hybrid procedure*
- *aortopulmonary or ventricular pulmonary shunt*
- *stenting PDA,…*

3.3.2. After SVC to PA anastomosis (Glenn).

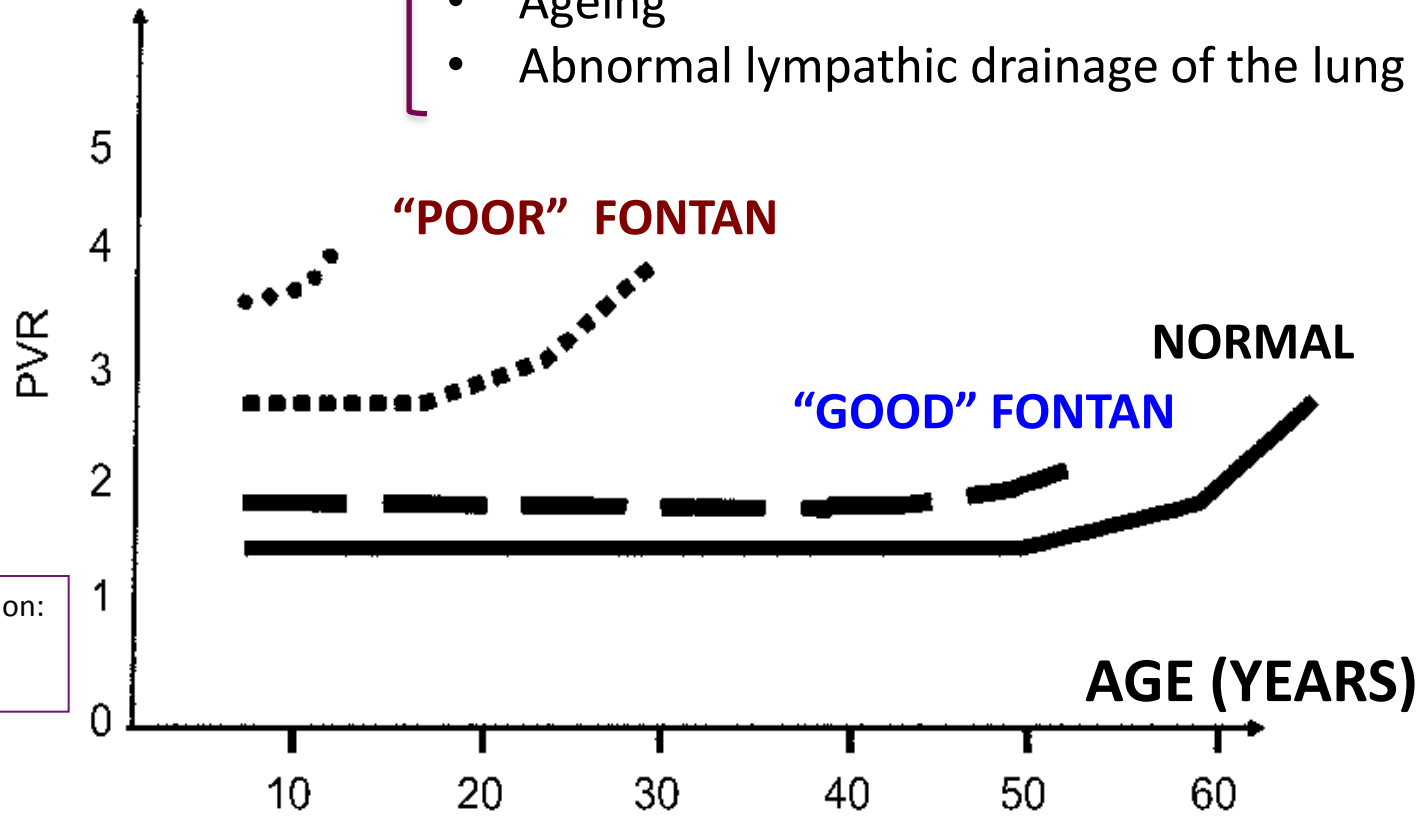
3.3.3. After total cavopulmonary anastomosis (Fontan).

- *These patients may have a PA pressure less than 25 mmHg but an increased PVRI and Pulmonary Vascular Disease…*

PVD developing AFTER THE FONTAN…

Progressive increase in PVR after the Fontan surgery:

- Suboptimal PA growth after Glenn / Fontan
- Absence of pulsatile pulmonary flow
- Functional loss of lung segments
- Ageing
- Abnormal lympathic drainage of the lung



The Fontan Circulation: Who controls the cardiac output?

Gewillig M

Interactive CardioVascular and Thoracic Surgery 10 (2010) 428-433

Evidence of pulmonary vascular disease after heart transplantation for Fontan circulation failure

- 15 failed Fontan patients undergoing heart transplantation.
- Compared PVRI and TPG before and after Heart transplant
- in a failing Fontan a *normal PVR does not exclude PVD*

Variable	n	Before	After	*P* value
All patients				
PAP (mm Hg)	13	17.0 ± 3.7	19.7 ± 3.3	.064
TPG (mm Hg)	12	5.3 ± 2.3	12.0 ± 2.1	<.0001
PVR (Wood units · m^2)	6	1.8 ± 1.1	2.7 ± 1.0	.296
Late Fontan failures				
PAP (mm Hg)	9	17.0 ± 3.4	20.6 ± 2.5	.057
TPG (mm Hg)	8	4.9 ± 2.5	12.8 ± 2.0	.0004
PVR (Wood units · m^2)	3	1.5 ± 0.9	3.5 ± 0.7	.155

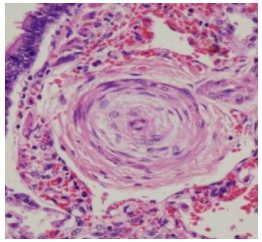
Mitchel et al. J Thorac Cardiovasc Surg 2004;128: 693-702

1. PULMONARY ARTERIAL HIPERTENSION

Dana Point 2008

- Idiopathic PAH
- Heritable (BMPR2, ALK1, Endoglin with or without HHT, Unknown)
- Drugs and toxins induced
- Associated with: Connective tissue diseases, HIV infection, Portal Hypertension,

Shunt Sistémico-pulmonar,
Schistosomiasis, chronic hemolytic anemia



- PPHN (HTP PERSISTENTE DEL RN)

1´. Pulmonary Veno Occlusive Disease (Pvo) And/Or Pulmonary Capillary Hemangiomatosis (Pch)

2. PH DUE TO LEFT HEART DISEASE

- Systólic dysfunction
- Diastolic dysfunction
- Valvulopathies

3. PH DUE TO HYPOXIA OR LUNG DISEASE

4. CHRONIC TROMBOEMBOLIC PH (CTEPH)

5. PH WITH UNCLEAR OR MULTIFACTORIAL MECHANISMS

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INTERACTIVE CARDIOVASCULAR AND THORACIC SURGERY

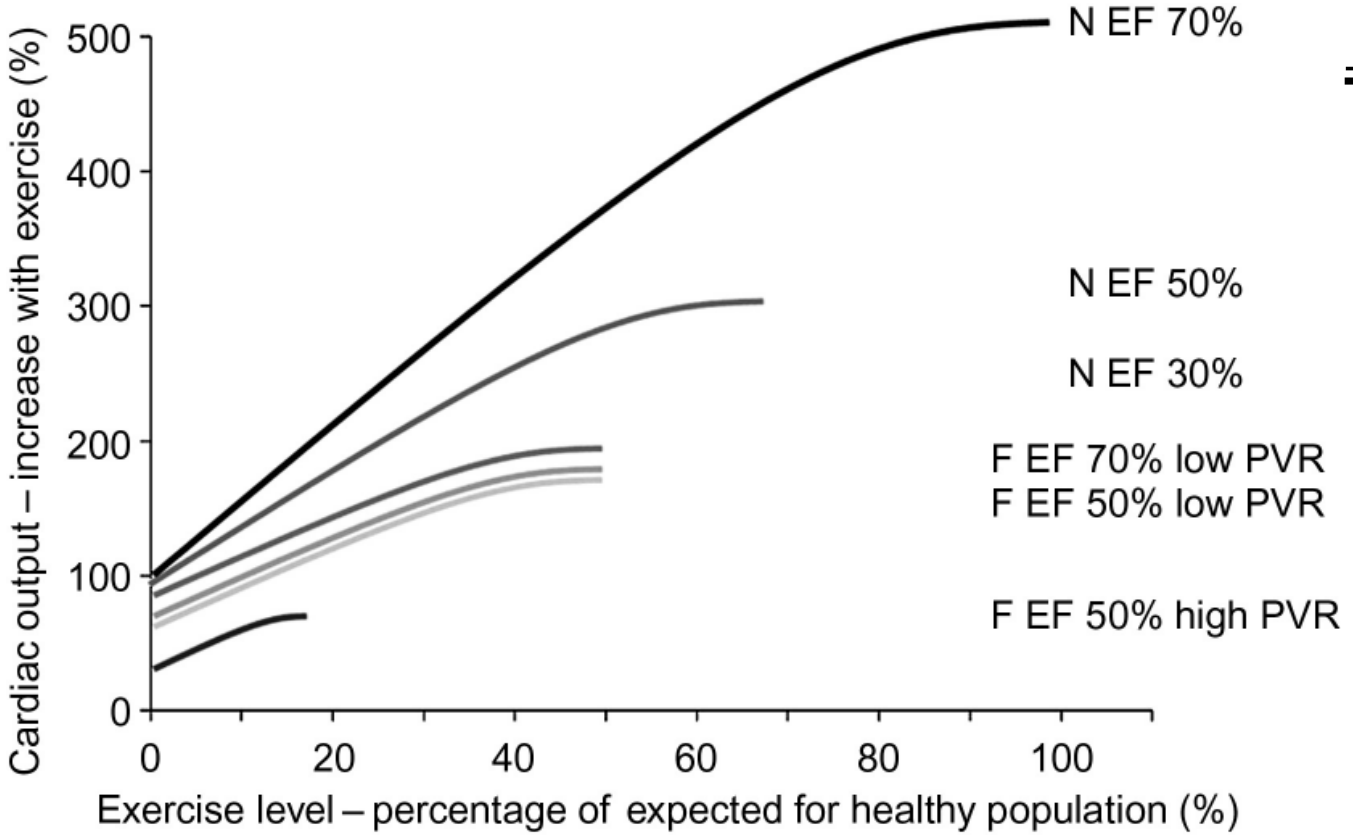


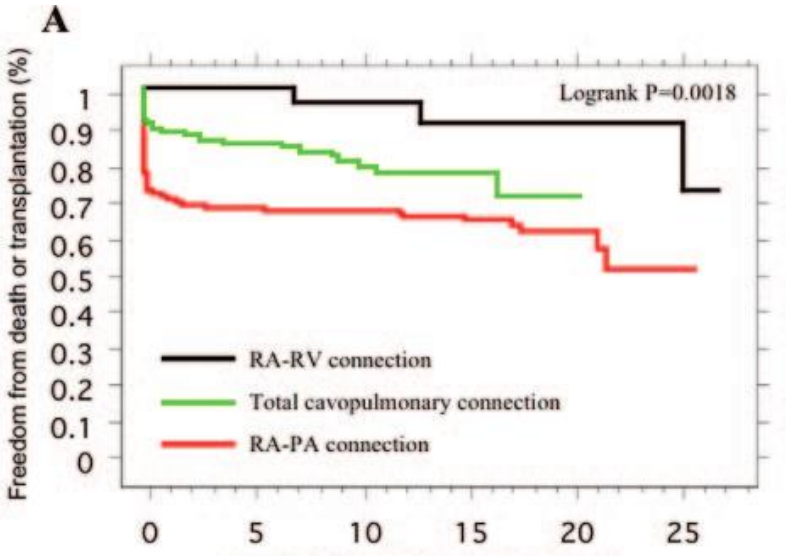
Fig. 4. Relationship of output during exercise, pulmonary vascular resistance (PVR) and ventricular function. A normal (N) subject with a biventricular circuit can increase his output by a factor of 5; if ventricular function is impaired, this will first result in decreased maximal output and subsequently in reduced output at low-level of exercise. In Fontan patients (F) output is

The effect of bosentan in patients with a failing Fontan circulation

Patient	Sat before	Sat end	Sat after stop (FU)	Sat after restart	6MWT before	6MWT end	6MWT stop	6MWT restart	REMARKS
patient 1	86	87	87	NA	NA	NA	NA	NA	VO_2max did not change on medication
patient 2	94	95	94	94	300	400	389	439	effect on 6MWT, restarted on bosentan
patient 3	95	94	Continued	Continued	445	434	Continued	Continued	effect on PLE, weaned from steroids, continued on Bosentan
patient 4	84	86	87	NA	520	500	NA	NA	no effect
patient 5	87	89	88	NA	370	350	NA	NA	no effect, suboptimal LV function
patient 6	84	88	79	NA	520	430	NA	NA	effect on saturation at rest and exercise and VO_2max, bosentan stopped after study
patient 7	91	91	92	NA	170	285	NA	NA	effect on 6MWT, bosentan stopped after study
patient 8	85	93	NA	NA	474	384	NA	NA	No effect on PLE
patient 9	66	88	NA	NA	3 minutes	8 minutes	NA	NA	effect of saturations and 6MWT, continued on sildenafil

Pulmonary Vasodilators in the Failing Fontan...

What do we mean by “failing Fontan”?



- Protein-losing enteropathy
- Plastic Bronchitis.

- Recurrent arrythmias
- Heart failure
- NYHA III or IV
- Sudden death
- Hepatic disease
- Recurrent thromboembolic events
- Increasing cyanosis
- Deceased tolerance to exercise

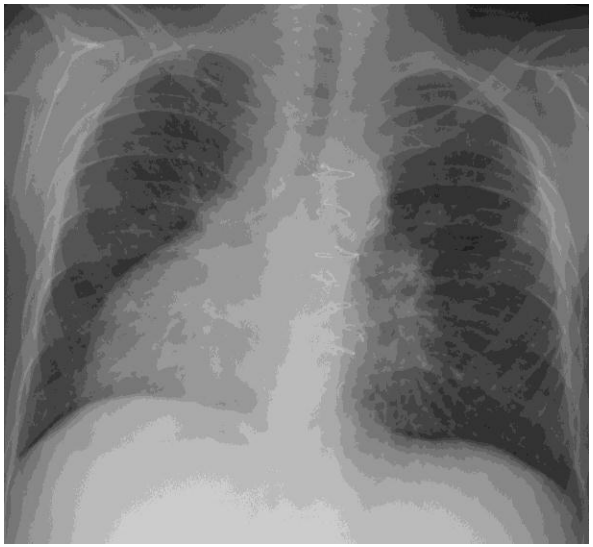
Long-Term Survival, Modes of Death, and Predictors of Mortality in Patients With Fontan Surgery

Circulation 2008,

Children´s Boston

REHIPED

REgistro de pacientes con HIpertensión Pulmonar PEDiátrica



CIRCULACIÓN UNIVENTRICULAR

12 años
Cardiopatia compleja en dextrocardia
4 cirugías cardiacas
Enfermedad pulmonar crónica
Derivación cardiopulmonar (Fontan)

ENTEROPATIA PIERDE PROTEINAS
Edemas, desnutrición, inmuodeficiencia…

PAP 20/14/18
PTD VI 12
PCP 13
RPt: 9 UW/m2
Rpa 2.5 UW/m2

Única alternativa:
Tx cardiaco
con mortalidad
operatoria del 40-50%

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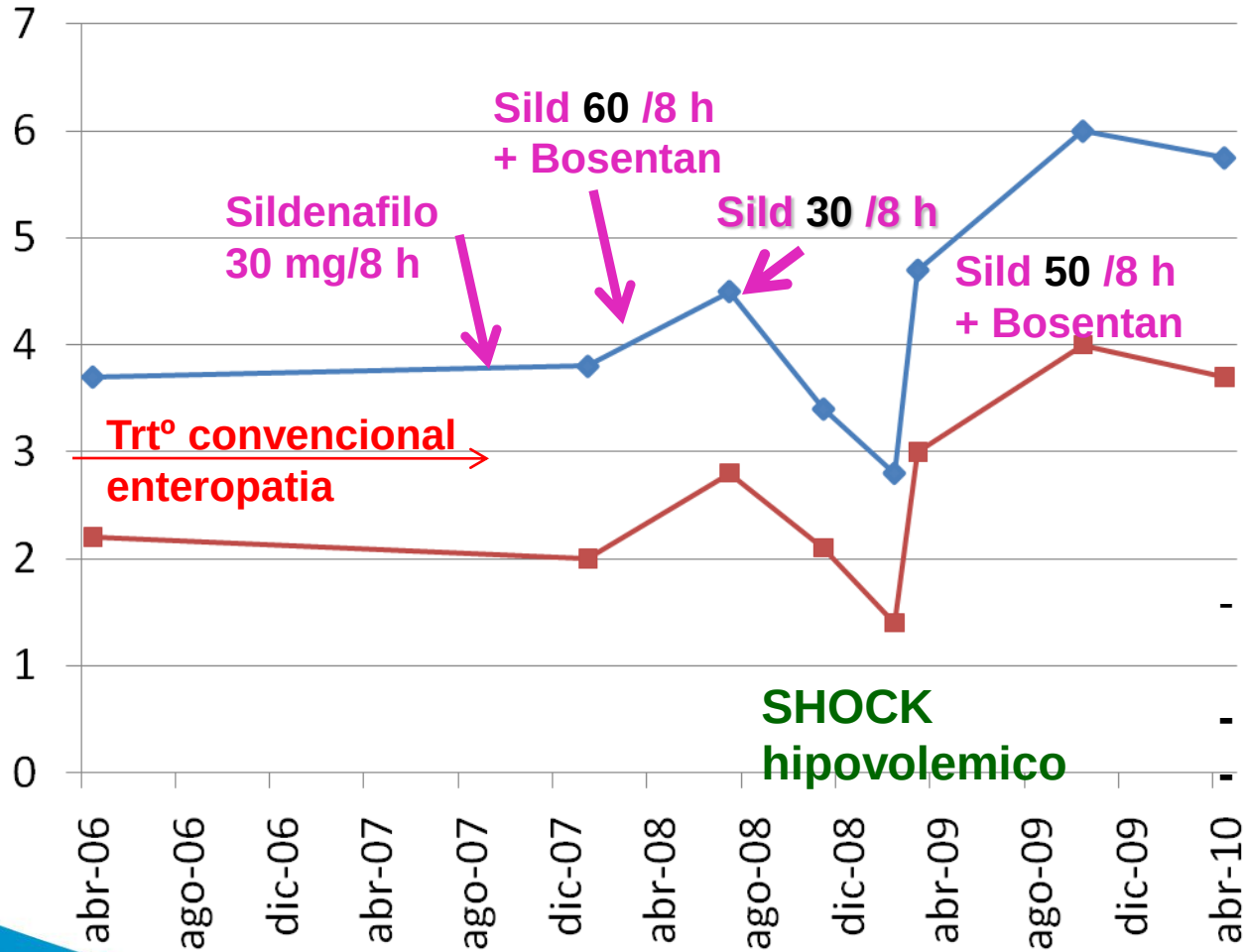

- Disminucion diureticos y corticoides
- Grado funcional IV a II
- Escolarizacion

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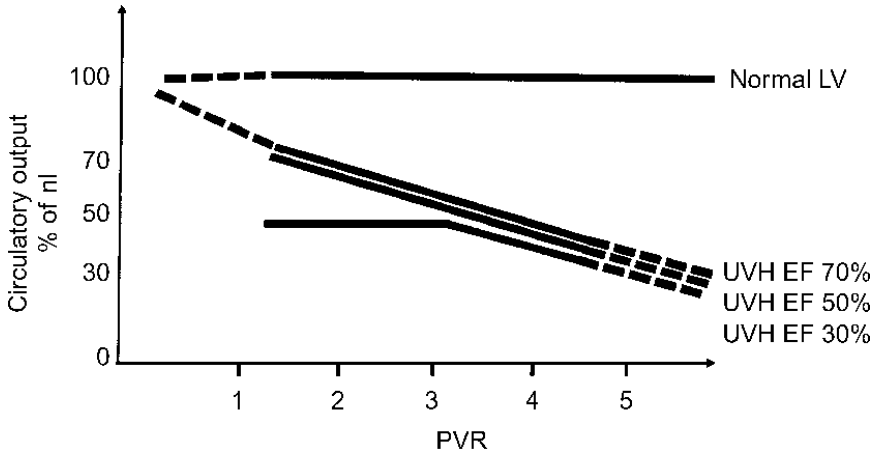

Fig. 3. Relationship of output at rest and pulmonary vascular resistance (PVR) modulated by ventricular function. In a normal subject (solid black line), cardiac output is not influenced by a mild increase of PVR up to 5 Woods Units. An increased PVR is invariably associated with decreased cardiac output at rest in all Fontan patients. If PVR is low, a good output is achieved in patients with normal or moderately depressed ventricular function; however, severely depressed ventricular function invariably results in low output. EF, ejection fraction; F, Fontan; UVH, univentricular heart; LV, left ventricle.

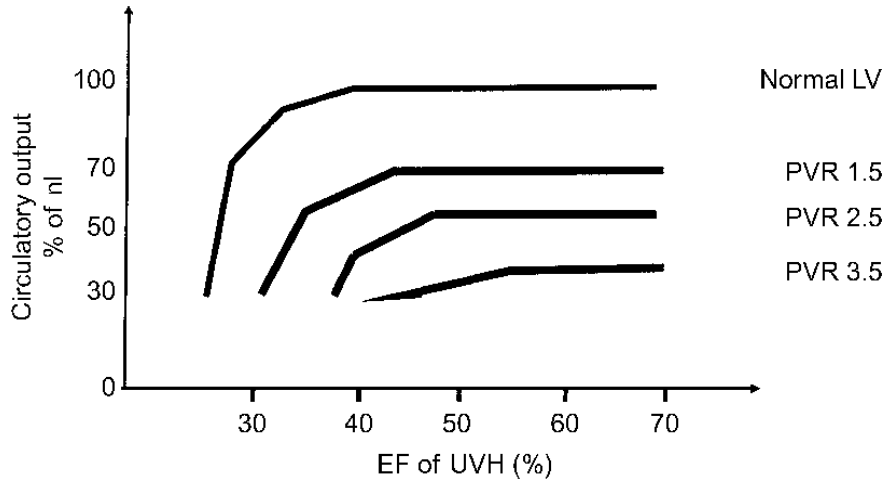

Fig. 2. Relationship of output at rest and ventricular function modulated by PVR. In a normal subject, cardiac output at rest is minimally influenced by ventricular function, except when severely depressed. In Fontan patients, minor changes of PVR result in marked changes of output; only when severely impaired, the ventricle will influence output. EF, ejection fraction; PVR, pulmonary vascular resistance; UVH, univentricular heart; LV, left ventricle.

M. Gewillig et al. / Interactive CardioVascular and Thoracic Surgery 10 (2010) 428–433

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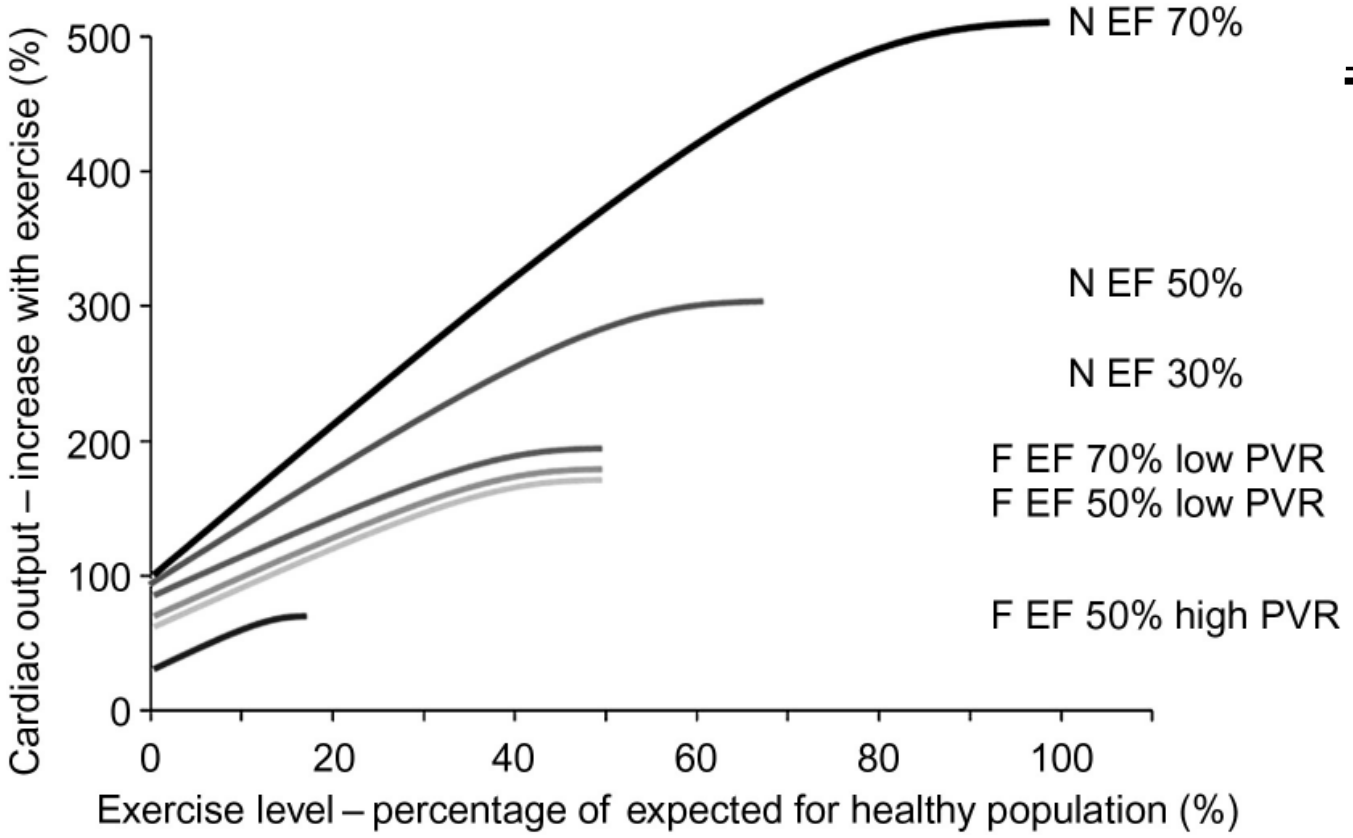

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Experiencia la paz en faling fontan

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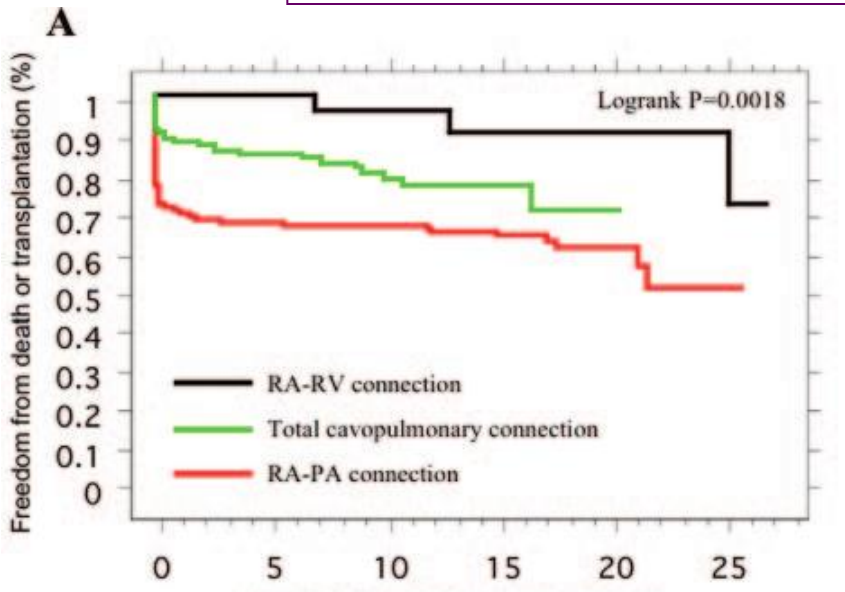
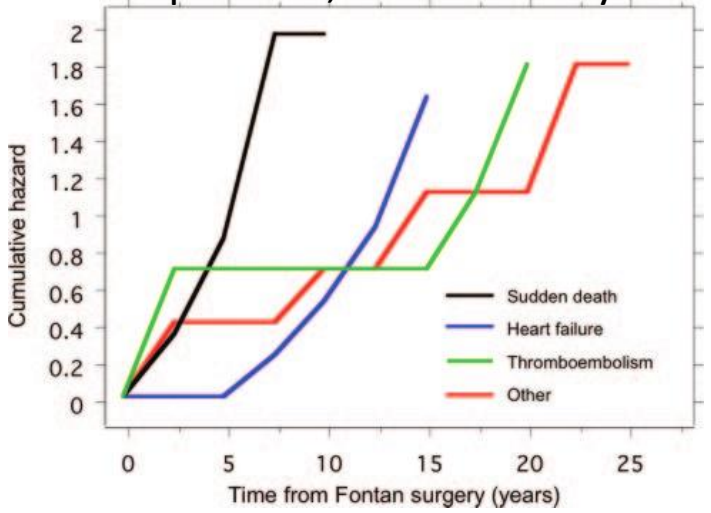


261 patients, mean FU 12 years



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Circulation 2008, Children´s Boston

Gen Thorac Cardiovasc Surg (2008) 56:104–108
DOI 10.1007/s11748-007-0201-z

ORIGINAL ARTICLE

Single ventricle repair in children with Down's syndrome

Naoki Wada, MD · Yukihiro Takahashi, MD
Makoto Ando, MD · In-Sam Park, MD
Takashi Sasaki, MD

Mortality After Total Cavopulmonary Connection in Children With the Down Syndrome

Monesha Gupta-Malhotra, MBBS[a,*], Virgil E.V. Larson, PhD[b], Ronald M. Rosengart, MD[d], Hongfei Guo, PhD[c], and James H. Moller, MD[b]

(Am J Cardiol 2010;105:865–868)

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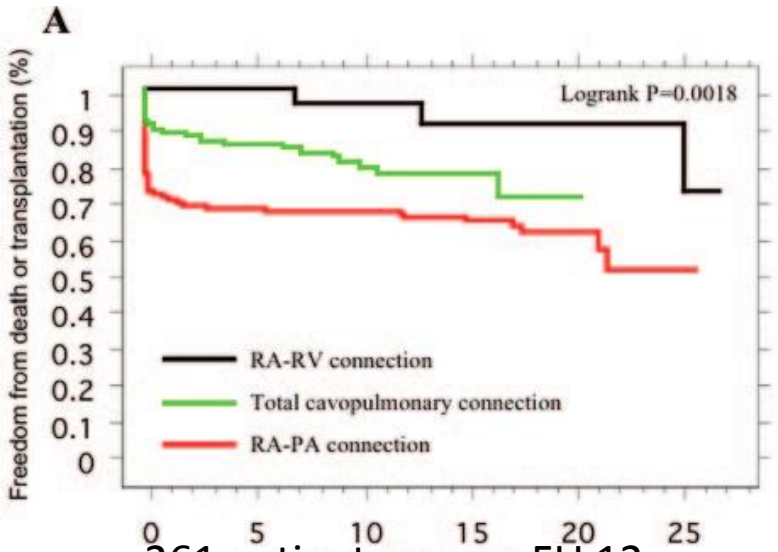

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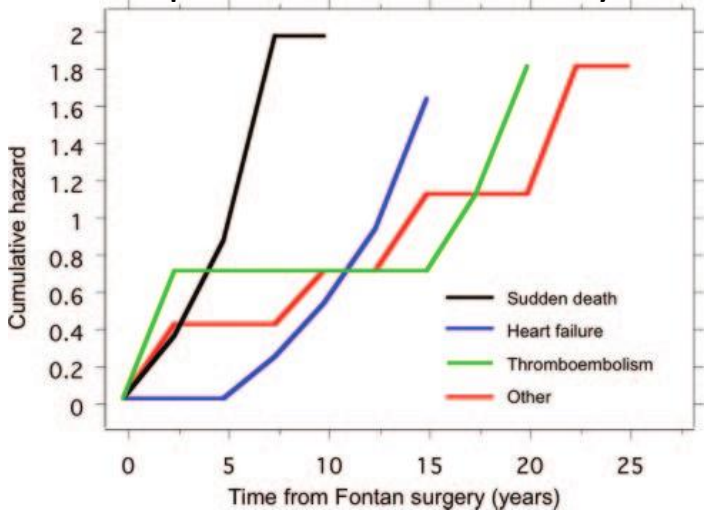

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