ACHD Symposium 2017

THE QUEENSTOWN MEETING

30 Nov - 2 Dec, Queenstown, New Zealand

Tips for Evaluation of Pulmonary Hypertension in the Adult with Congenital Heart Disease

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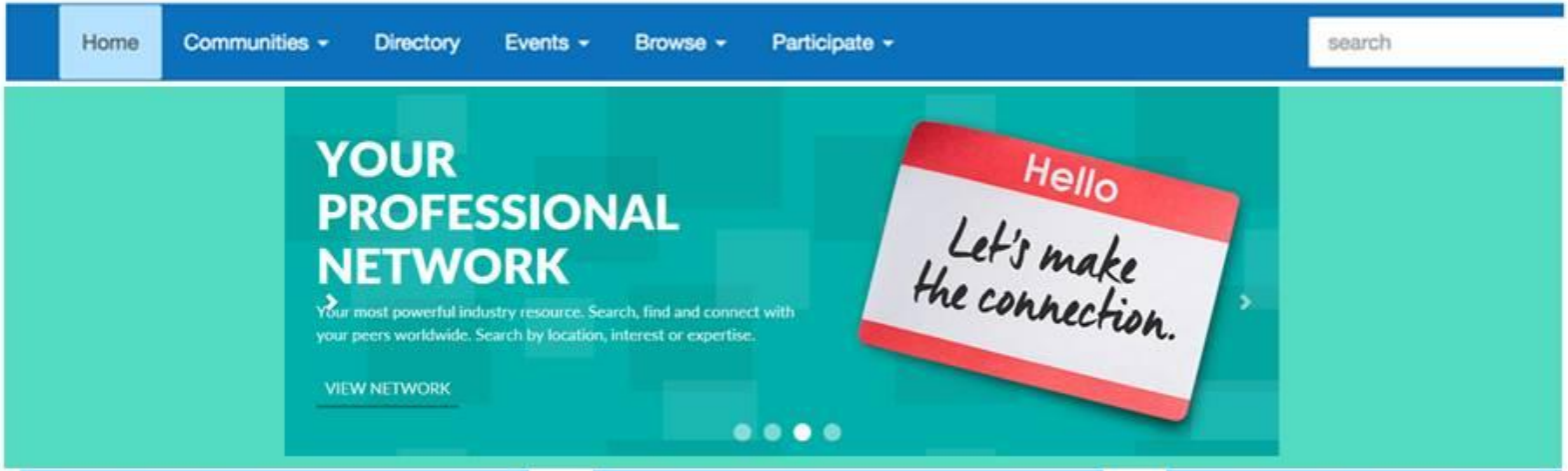
International Society for Adult Congenital Heart Disease



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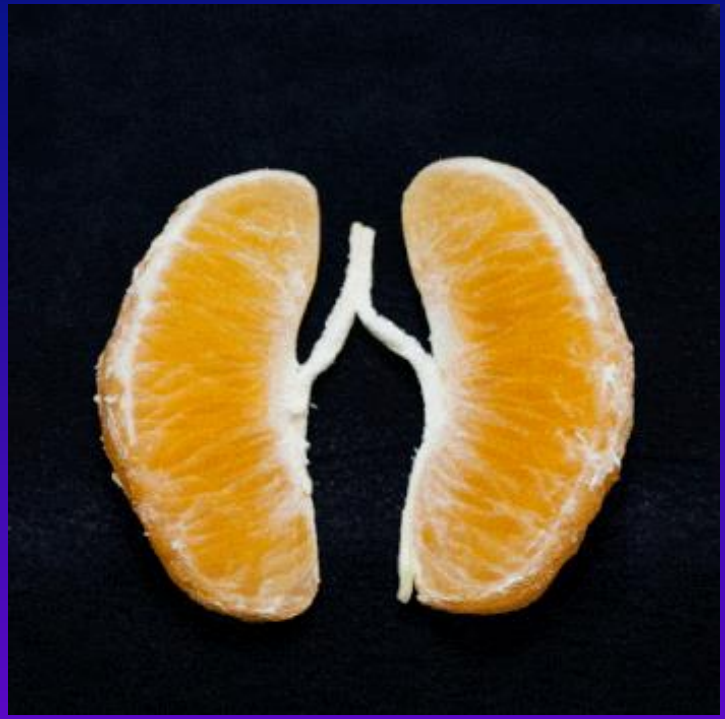
HIGHER LOGIC
ALL TOGETHER



Questions

Functional status

Severity

Reversibility

Prognosis

Treatment and Operability

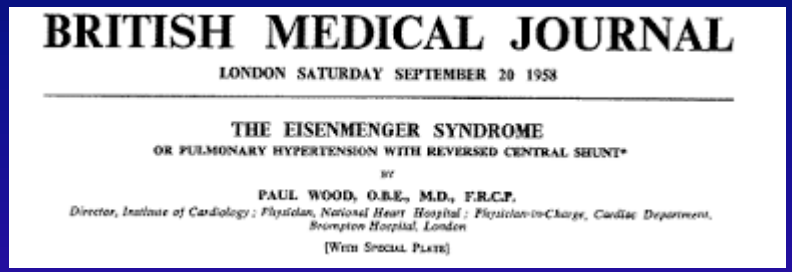
Eisenmenger syndrome

32 yo man

Non restrictive VSD

Cyanosis

Dyspnea



BRITISH MEDICAL JOURNAL

LONDON SATURDAY SEPTEMBER 20 1958

THE EISENMENGER SYNDROME

OR PULMONARY HYPERTENSION WITH REVERSED CENTRAL SHUNT*

BY

PAUL WOOD, O.B.E., M.D., F.R.C.P.

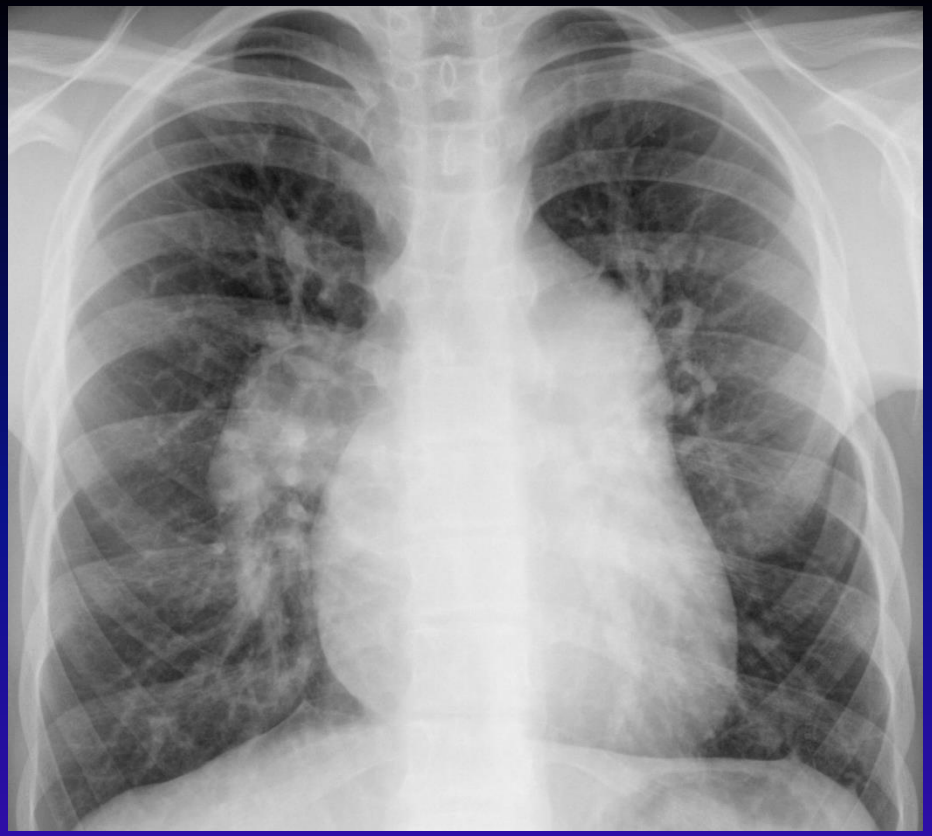
Director, Institute of Cardiology; Physician, National Heart Hospital; Physician-in-Charge, Cardiac Department, Brompton Hospital, London

[WITH SPECIAL PLATE]

Reasonably active until 3 years before his death

Developed progressive CHF, died of hemoptysis

Eisenmenger V. Z Klin Med. 1897; 32



Nice 2013 PAH/CHD

1. Eisenmenger syndrome
2. Left to right shunts

   Correctable

   Noncorrectable
3. PAH with coincidental CHD
4. Post operative PAH

JACC Vol 62, No.25, Suppl D, 2013

Tests – traditional and evolving

Anatomy – Echo (TTE, TOE), CT

Hemodynamics

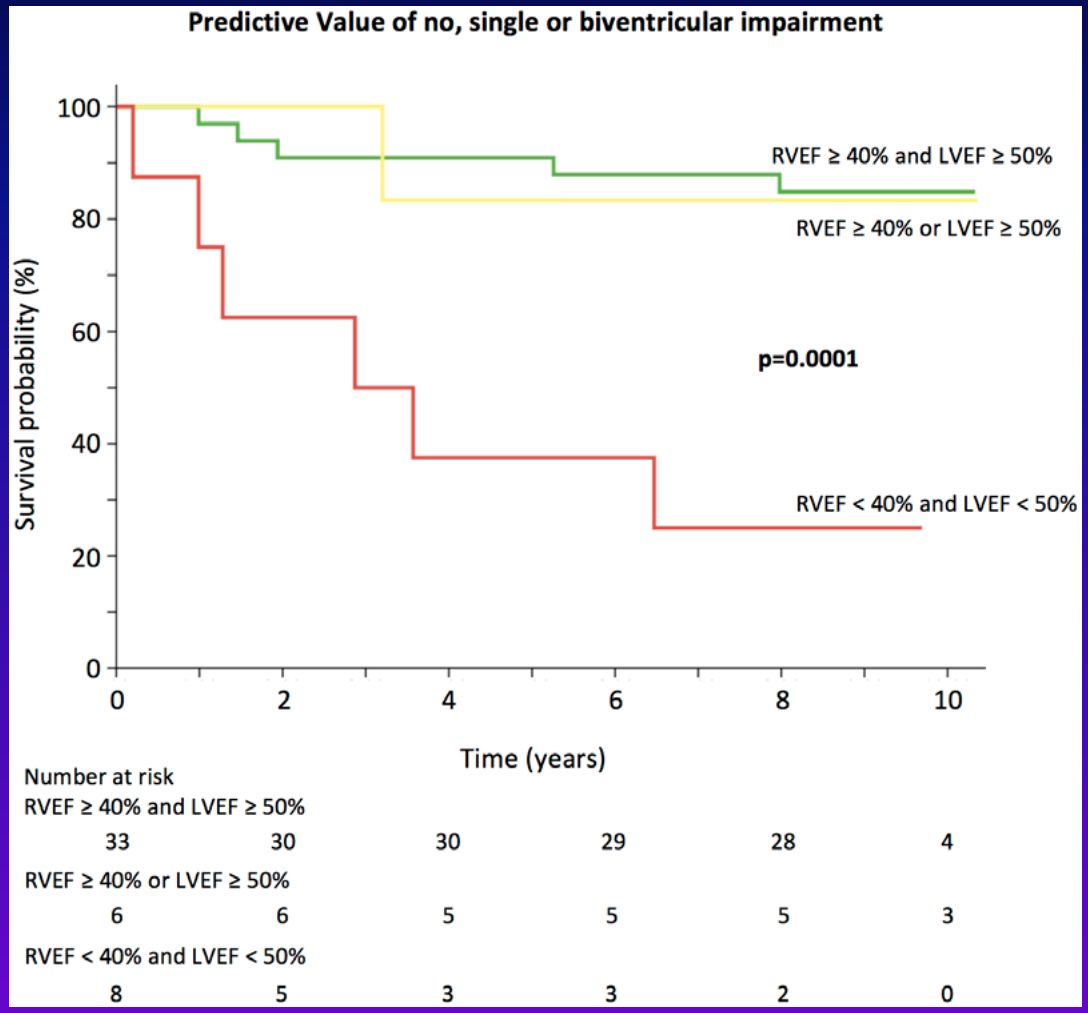
Genetic testing

Echo parameters of RV function

TAPSE, RV strain, Fractional area change

MRI

Impaired Right, Left, or Biventricular Function and Resting Oxygen Saturation Are Associated With Mortality in Eisenmenger Syndrome
A Clinical and Cardiovascular Magnetic Resonance Study



Jensen et al, Circulation CV imaging 2015

Functional status

NYHA/Ability Index

QoL scores - Camphor and others

Exercise testing

- 6 Minute walk
- CP Exercise testing

Other parameters

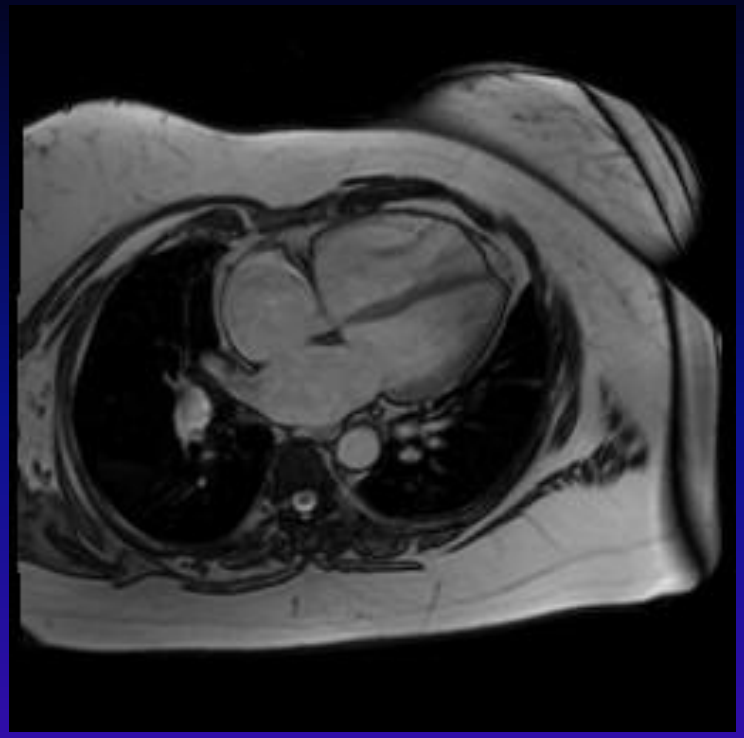
Eisenmenger group – predictors of death

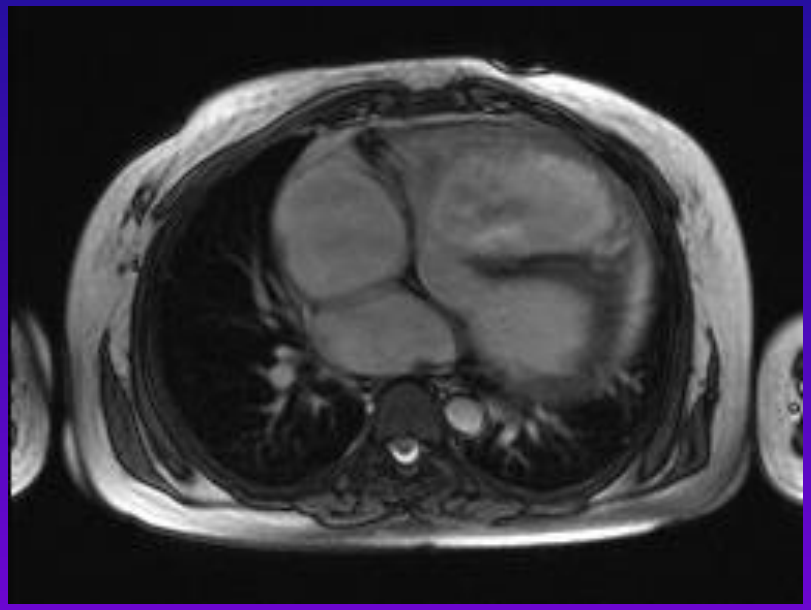
- Age
- Pretricuspid shunt
- Oxygen saturation at rest
- Presence of sinus rhythm
- Presence of pericardial effusion

Kempney et al Circulation April 2017

Cardiac Cath – what do we want to know?

- Understand anatomy
- Venous occlusion
- Phobias
- Developmental issues





Need to clearly understand the anatomy and be systematic !

Pulmonary venous desaturation

Systemic desaturation

?

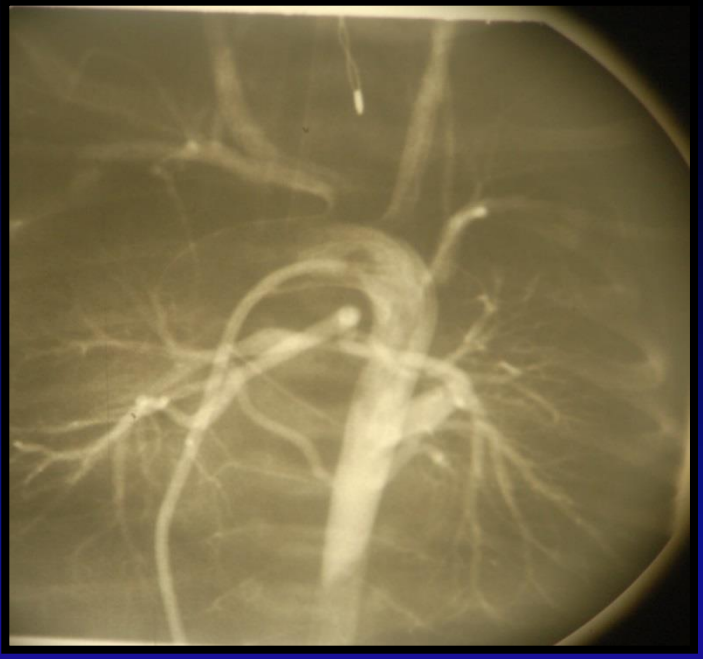
CI
PVR(I)
Qp:Qs
Rp:Rs

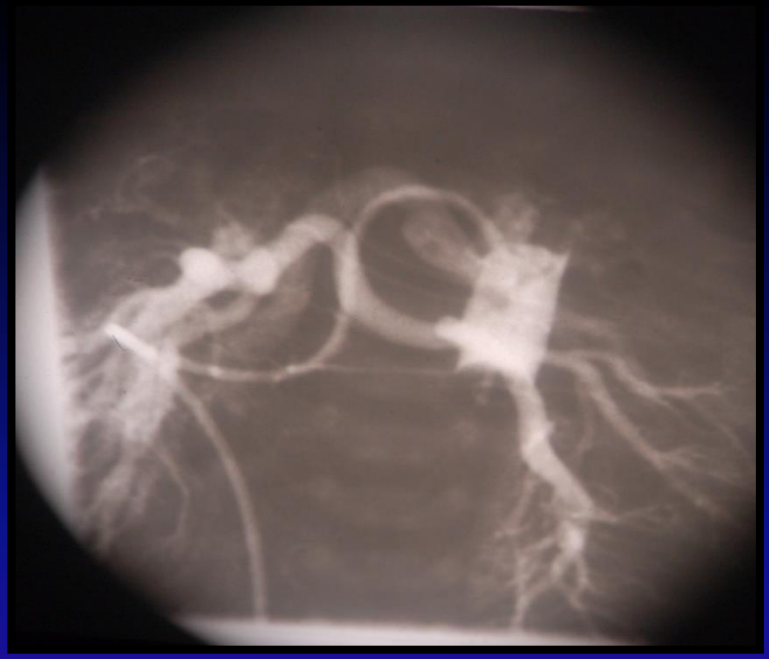
2 (or more) sources of Pulmonary blood flow ?

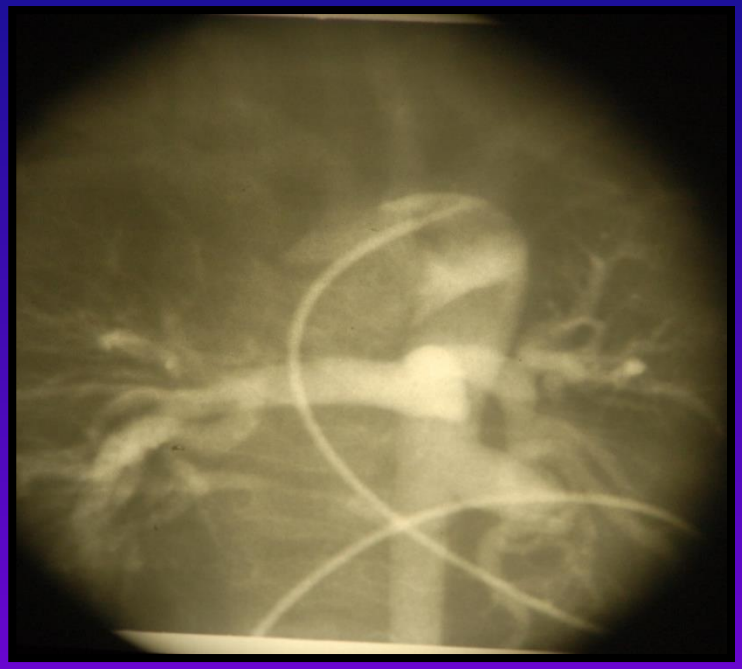
Low Cardiac output

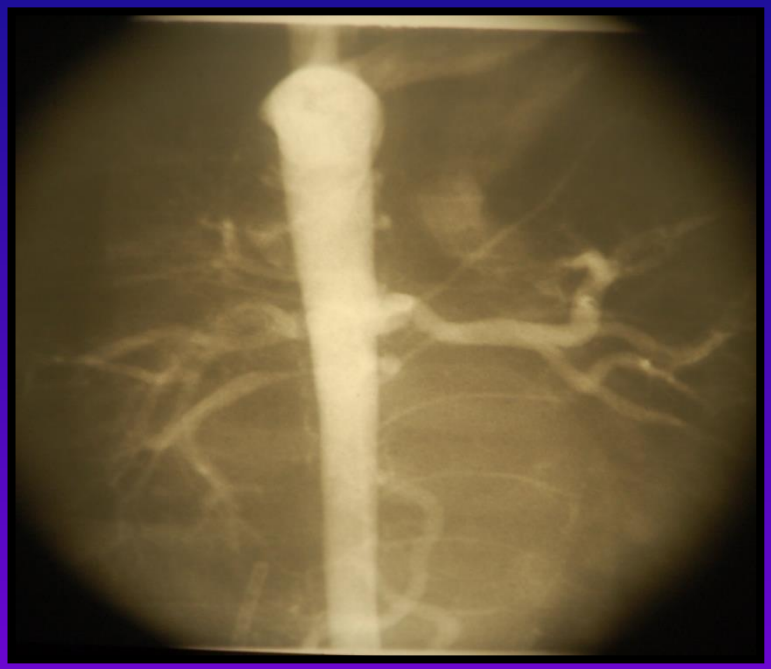
Bilateral SVCs?
IVC interruption
Anomalous pulmonary venous drainage?

Complex Pulmonary Supply and Drainage









opies Dr J Andrew, Alnwick Street, Warkworth
Dr R.N.Easthorpe, Cardiologist, Wellington Hospital
Brian Barratt-Boyes Mr Monro, Cardiology

C2212 Age 4 Ward 7 Code C37F

19.10.71 TRANSPOSITION GREAT VESSELS WITH HIGH PVR (PREVIOUS BLALOCK/HANLON PROCEDURE) ATRIAL BAFFLE REPAIR UNDER BYPASS

BRIAN BARRATT-BOYES
MR MONRO
Gen.DR SEELYE

Bypass No 2428

Findings: This child had a Blalock/Hanlon procedure in infancy, performed at Wellington Hospital. Subsequent follow-up has shown a progressive increase in pulmonary vascular resistance, despite the absence of any other communication. Study was recently repeated here and this showed the pulmonary vascular resistance to be virtually equal to systemic and pulmonary flow perhaps slightly less than systemic. While these resistance levels would have contraindicated operation in the absence of transposition it was felt that in the present circumstances a baffle repair should improve the situation by abolishing cyanosis and therefore decreasing blood viscosity.

At operation the pericardial space was completely obliterated, the adhesions being particularly dense over the lateral RA wall. The ventricles were not fully exposed but the heart was clearly diffusely enlarged. The anatomy was consistent with transposition with the aorta arising anteriorly from a morphological RV and the pulmonary artery posteriorly and to the left from LV. When RA was opened there was a circular large ASD occupying the lower part of the atrial septum and with a diameter of about 1.5cm. The pulmonary veins drained correctly to LA and the two AV valves looked normal.

After placement of the baffle the SVC pressure was excessively high - about 40mm.Hg mean - and we therefore went back on so bypass and inserted a further pericardial patch into the baffle. After this SVC pressure was probably about twice IVC and although I was disappointed that there was still some apparent SVC obstruction, we decided to accept this pressure. (Pressures at this level have been encountered occasionally before and have not led to postoperative difficulties.) In addition, pulmonary artery pressure had fallen significantly and was now only just over half that in the aorta.

Pressures were as follows:

after:	11.50a.m.	SVCR 35	1.10 p.m.	RPA 34/21	+ m 28	Ao 64/32	+ m 44	Anneedle 53/33
	1.00p.m	22						

Personal Health Passport

A publication of the Adult Congenital Heart Association

Sponsored by the American College of Cardiology Foundation

Fontan circulation

Non pulsatile flow

Loading conditions eg dehydration

Sedation/Ventilation

End expiratory pressures

Isolated atrial septal defect with pulmonary vascular obstructive disease — long-term follow-up and prediction of outcome after surgical correction

PETER M. STEELE, M.B.B.S. (HONS), VALENTIN FUSTER, M.D., MARC COHEN, M.D., DONALD G. RITTER, M.D., AND DWIGHT C. MCGOON, M.D.

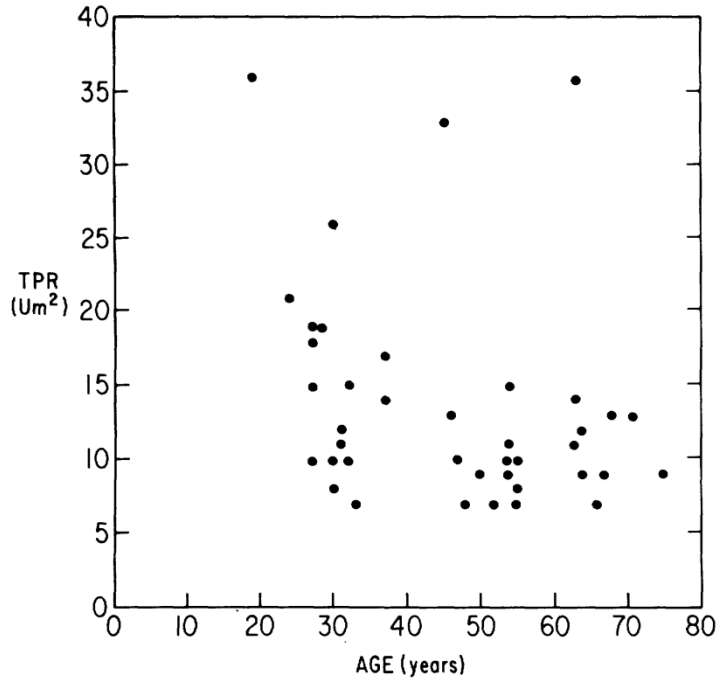


FIGURE 1. Relationship between the age of the patient and the total pulmonary resistance (TPR), in Wood units, at diagnostic catheterization.

'In conclusion

(1) Atrial septal defect with high total pulmonary resistance is uncommon and predominates in adult female patients.

(2) Total pulmonary resistance (or pulmonary arteriolar resistance) is the best predictor of surgical outcome. In patients with total pulmonary resistance less than 15 U/m2, surgical treatment is advised.

(3) In patients with borderline total pulmonary resistance, the systemic arterial oxygen saturation provides a good prediction of surgical outcome.'

Circulation 76, No. 5, 1037-1042, 1987.

Suitable for closure ?

Table 4 **Criteria for Closing Cardiac Shunts in PAH Patients Associated With Congenital Heart Defects***

PVRi, Wood units/m^2	PVR, Wood units	Correctable†
<4	<2.3	Yes
>8	>4.6	No
4-8	2.3–4.6	Individual patient evaluation in tertiary centers

*Criteria: the long-term impact of defect closure in the presence of pulmonary arterial hypertension (PAH) with increased PVR is largely unknown. There are a lack of data in this controversial area, and caution must be exercised. †Correctable with surgery or intravascular nonsurgical procedure.
PVR = pulmonary vascular resistance; PVRi = pulmonary vascular resistance index.

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December 24, 2013:D34–41

Pt No.	Date of Procedure	Sex	Age	Qp/Qs	M	PVRI	Vasodilator	Lowest PVRI	Device Size (mm)	OUTCOME
1	24/07/2003	F	31	1.2		12.7	yes	11.9	32	alive on vasodilators
2	04/02/2003	F	26	1.2		18	yes	11.7	32	died 2.5 years post ASD
3	27/9/2001	F	46	2.1		5.5	no	5.5	30	died 5.5 years post ASD
4	23/10/2001	F	28	1.1**		13.9	yes	6.5**	32	alive and well
5	08/02/2000	F	66	6.2		2.1	no	2.1	22	died after CABG surgery
6	25/02/2005	F	69	0.8		8.3	yes	3.9	28	alive TR 3.3m/s
7	26/05/2003	M	58	1.9		Data lost	-	-	22	alive and well
8	18/09/2001	F	45	2.4		3.4	no	3.4	28	alive and well. No TR
9	22/02/2000	F	57	2.4		4.3	no	4.3	20	alive and well, TR 2.2m/s
10	17/10/2000	M	31	1.7		4.7	no	4.7	20	alive and well
11	22/08/2000	F	30	2.1		4.2	no	4.2	30	alive and well. No TR
12	03/06/2003	F	10	1.3		4.2	no	4.2	26	alive and well. No TR
13	29/04/2003	F	5	1.9		3.0	no	3.0	22	alive and well

Progressive Pulmonary Hypertension Post Atrial Septal Defect Device Closure—Early Symptomatic Improvement may not Predict Outcome

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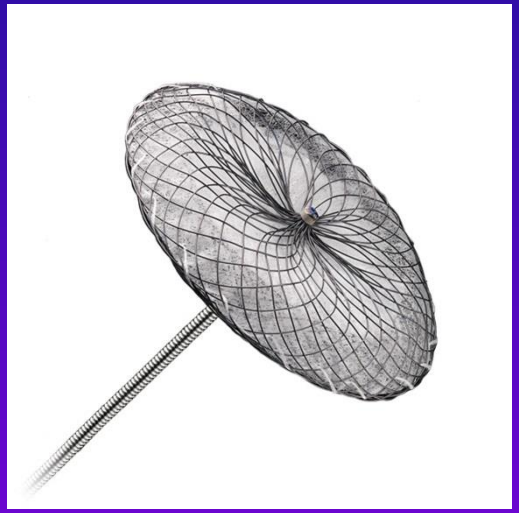
Heart, Lung and Circulation
2010;19:713–716



Other contributors

Obesity/OSA

Diastolic dysfunction

Abnormal pulmonary function

Thrombus

Genetics

Importance of Follow up

Always refer back
to the heart.
It is where
the world
began.

Jenny Bornholdt

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