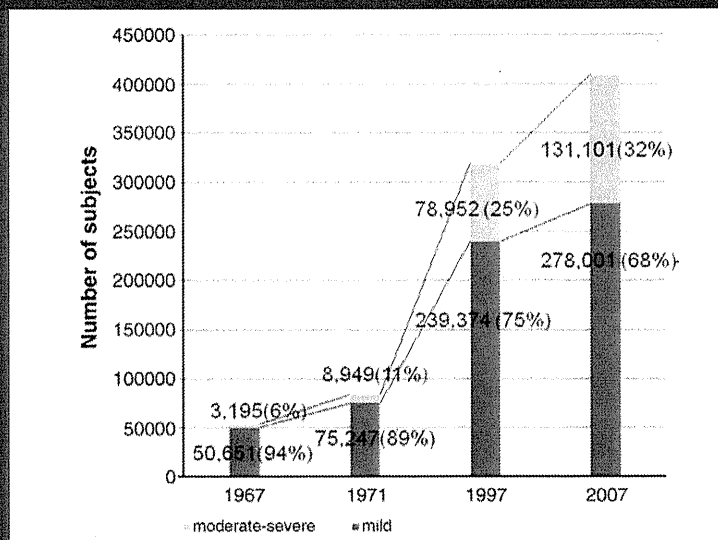


Prevalence of adult patients with CHD in Japan

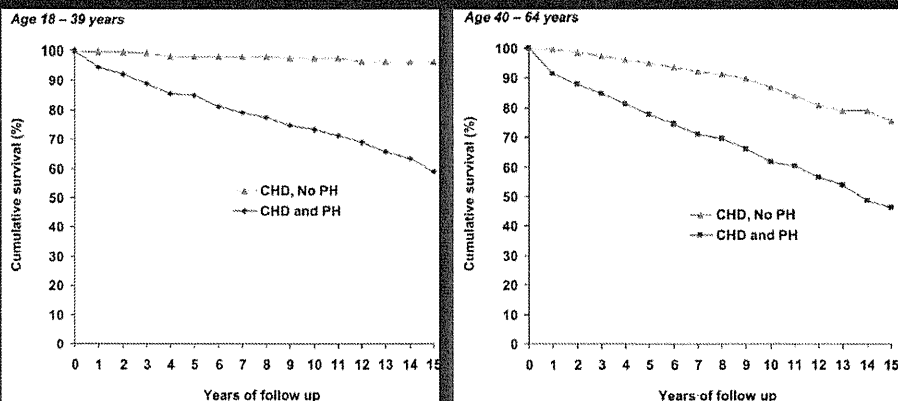


Shiina Y, et al. Inter J Cardiol 2009

Diagnosis of Pulmonary Hypertension in the Congenital Heart Disease Adult Population

Impact on Outcomes

Boris S. Lowe, MB, ChB,*† Judith Therrien, MD,*† Raluca Ionescu-Ittu, PhD,*‡
 Louise Pilote, MD, MPH, PhD,‡§ Giuseppe Martucci, MD,* Ariane J. Marelli, MD, MPH*



J Am Coll Cardiol 2011;58:538-46

Various Conditions of ACHD with PH

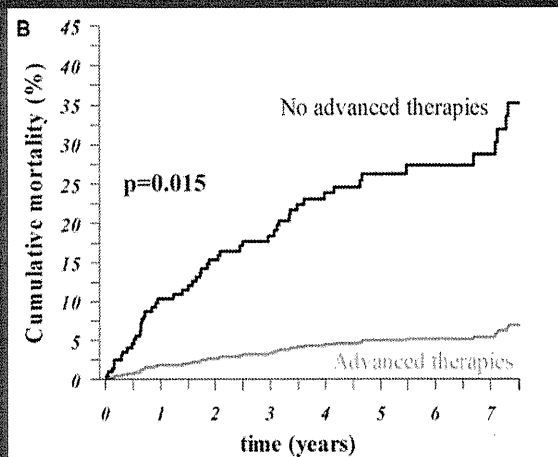
- Eisenmenger Syndrome
- Left to Right Shunt disease with PH
- Post operative PH (without shunt)

Various Conditions of ACHD with PH

- Eisenmenger Syndrome
- Left to Right Shunt disease with PH
- Post operative PH (without shunt)

Improved Survival Among Patients With Eisenmenger Syndrome Receiving Advanced Therapy for Pulmonary Arterial Hypertension

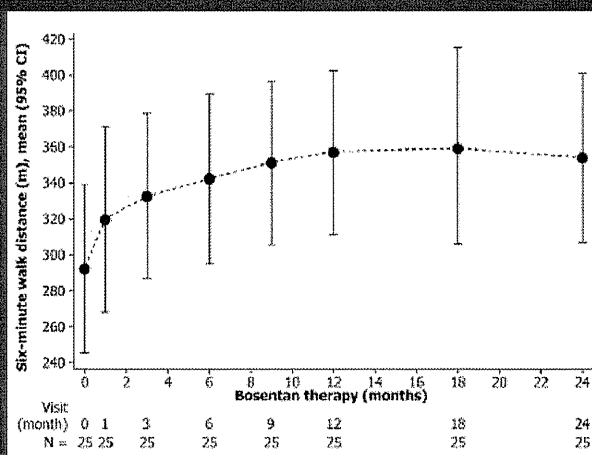
Konstantinos Dimopoulos, MD, MSc, PhD, FESC^{1,2}; Ryo Inuzuka, MD³; Sara Goletto, MD;
 Georgios Giannakoulas, MD, PhD, FESC¹; Lorna Swan, MD, MRCP;
 Stephen J. Wort, BA, MBBCh, MRCP, PhD¹; Michael A. Gatzoulis, MD, PhD, FESC



Circulation. 2010;121:20-25.

Efficacy and Safety of *Bosentan* for Pulmonary Arterial Hypertension in Adults With Congenital Heart Disease

Oliver Monfredi, MBChB, MRCP^a; Linda Griffiths, RGN, RSCN^b; Bernard Clarke, MD^{a,b}, and
 Vaikom S. Mahadevan, MD^{a,b,c}



Am J Cardiol 2011

What is the goal of management in ACHD patients with PH

- To improve clinical symptoms
- To reach to the complete repair
(Lung transplantation)

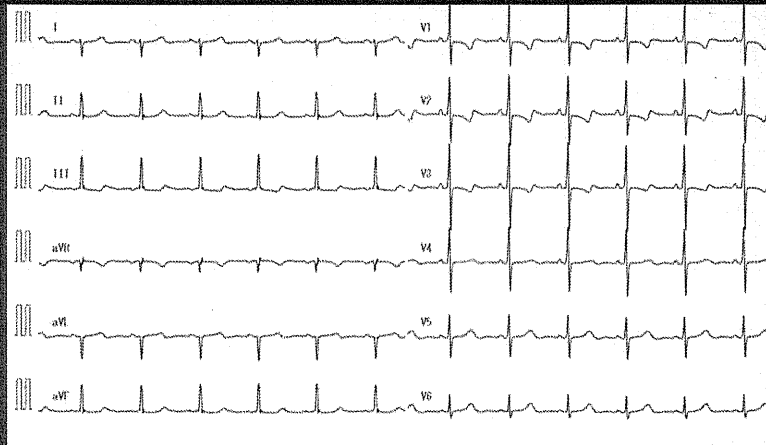
Various Conditions of ACHD with PH

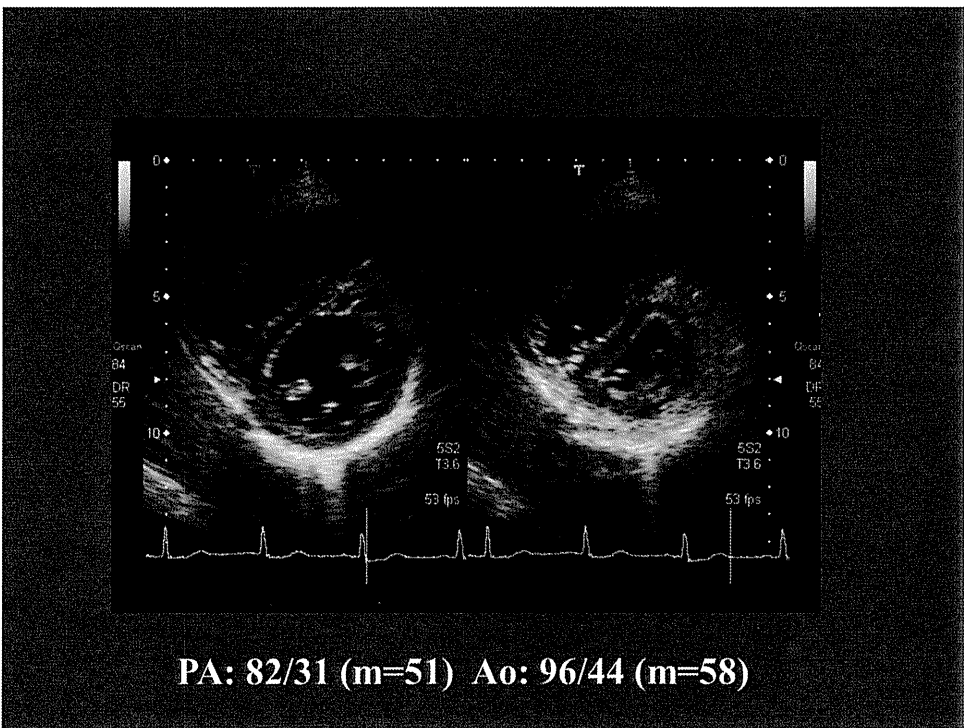
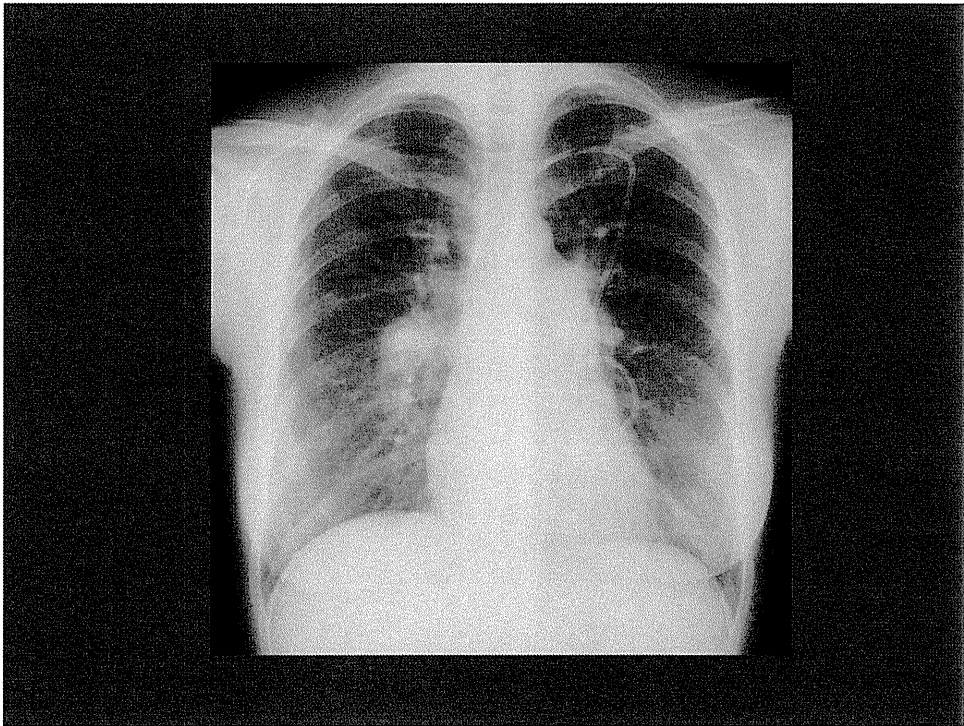
- **Eisenmenger Syndrome**
- **Left to Right Shunt disease with PH**
- **Post operative PH (without shunt)**

What is the goal of management in ACHD patients with PH

- To improve clinical symptoms
- To reach to the complete repair surgery or catheter intervention

32 years female





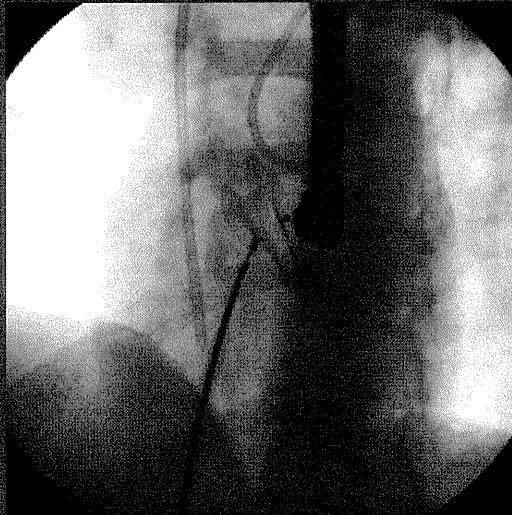
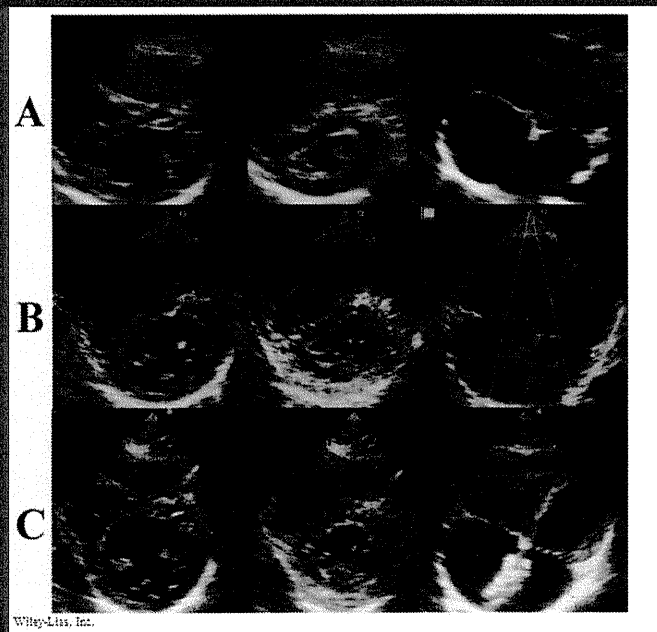


Table I. Sequential Hemodynamic Data by Cardiac Catheterization

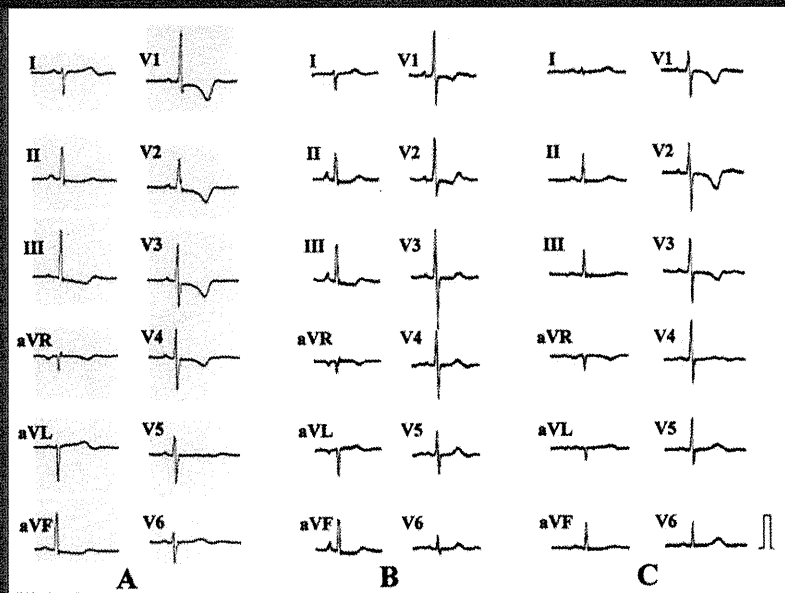
	Before epoprostenol therapy	Before ASD occlusion	Soon after ASD occlusion	1 year after ASD occlusion
PAP (mm Hg)	106/32 (58)	82/31 (51)	53/22 (36)	57/23 (39)
PVR (dyne sec/cm ⁻⁵)	824	471	N/A	256
Qp/Qs (L/min/m ²)	3.7/2.4	6.8/3.4	N/A	5.5/5.3
Qp/Qs	1.5	2.0	N/A	1.0

Hirabayashi A, Akagi T, *Catheter Cardiovasc Interv* 2009



Wiley-Liss, Inc.

Hirabayashi A, Akagi T, Catheter Cardiovasc Interv 2009



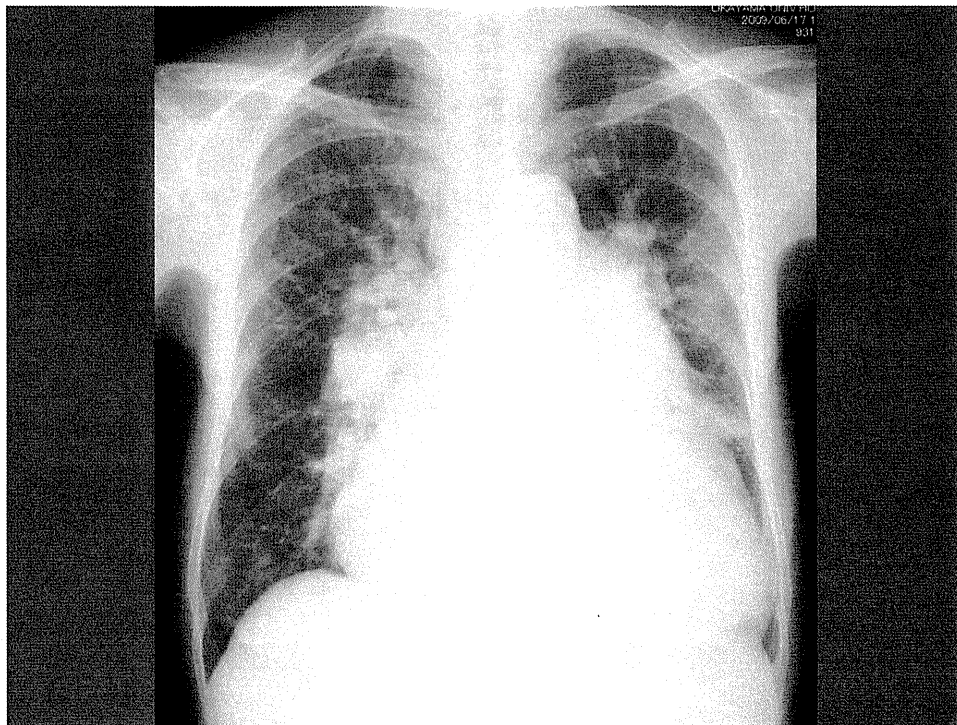
Wiley-Liss, Inc.

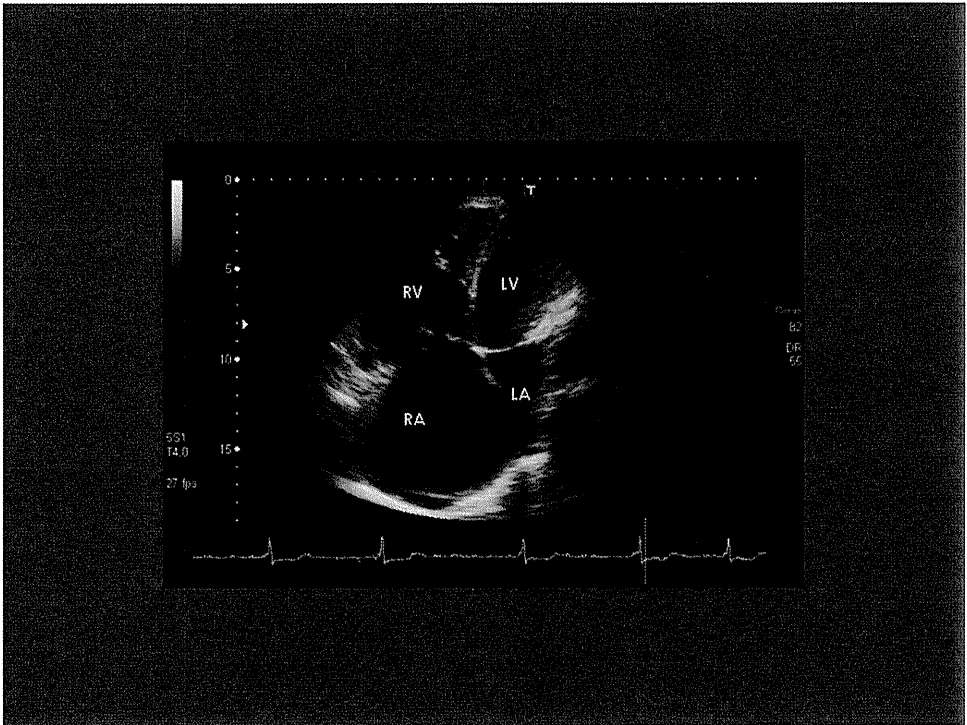
Hirabayashi A, Akagi T, Catheter Cardiovasc Interv 2009

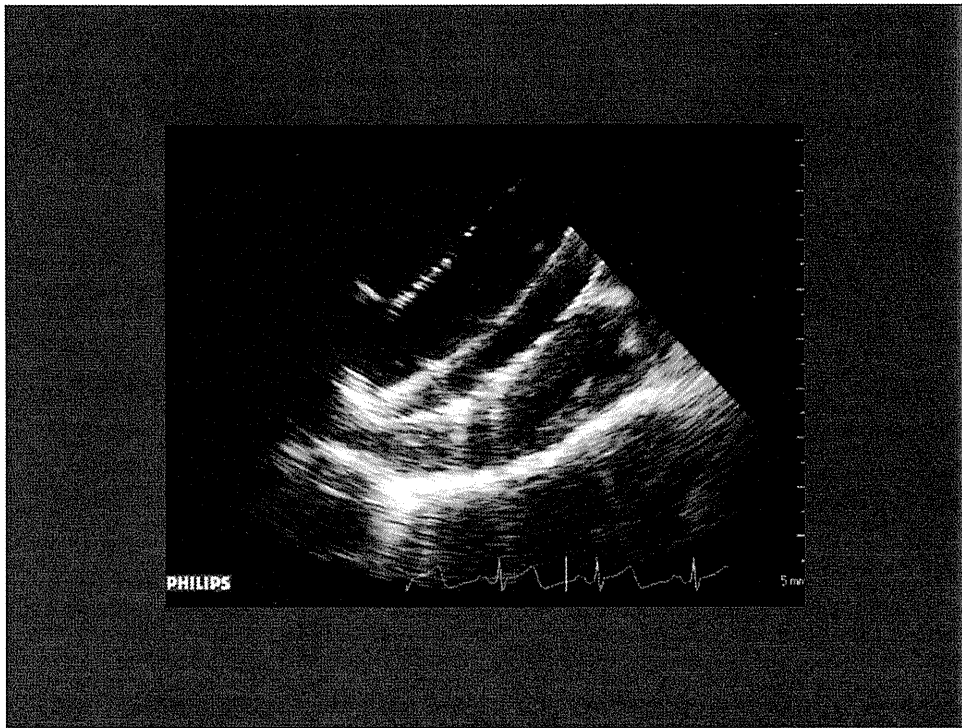
Case# . 82 years old, female.

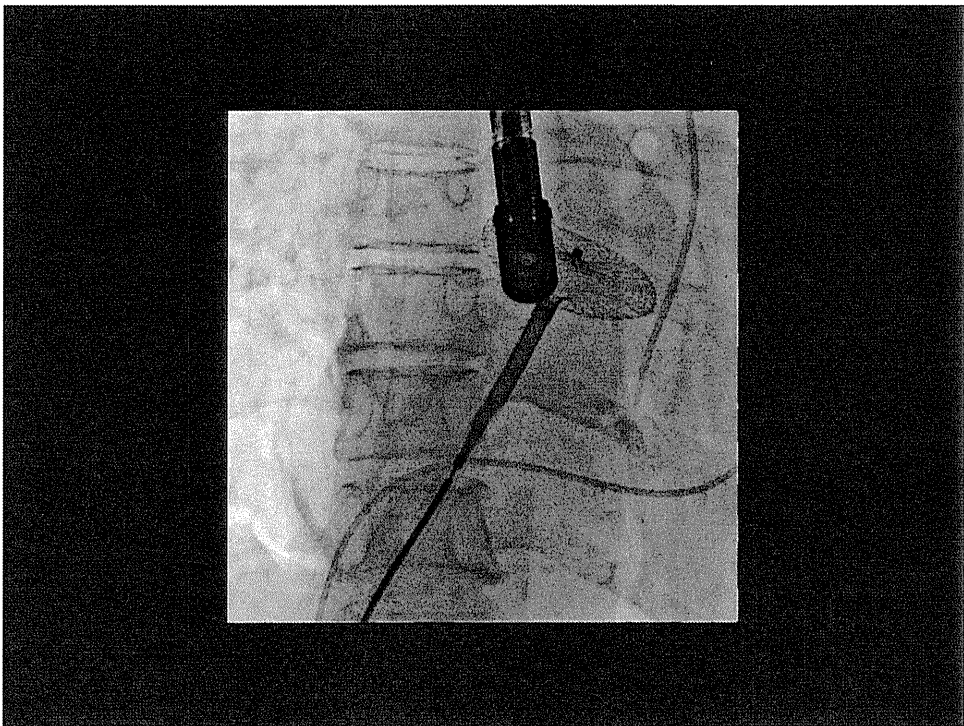
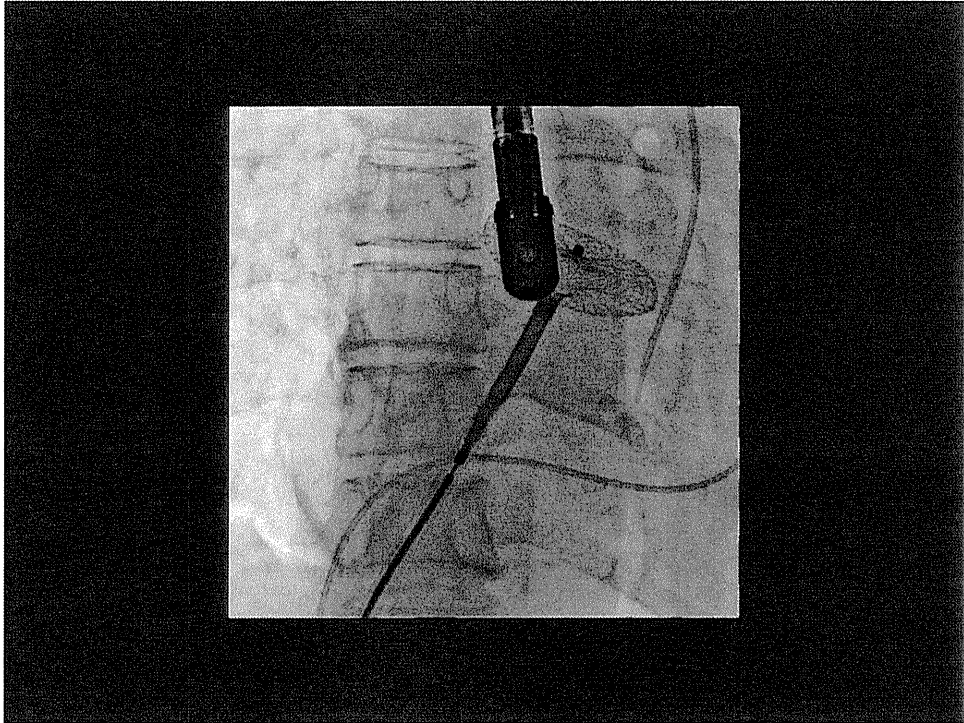
History:

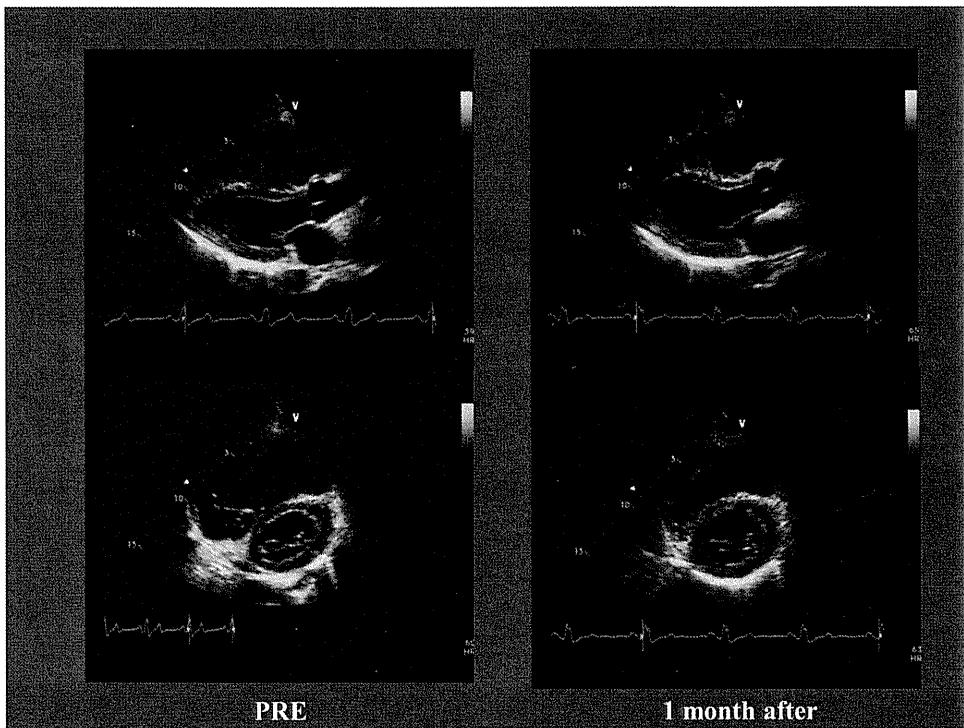
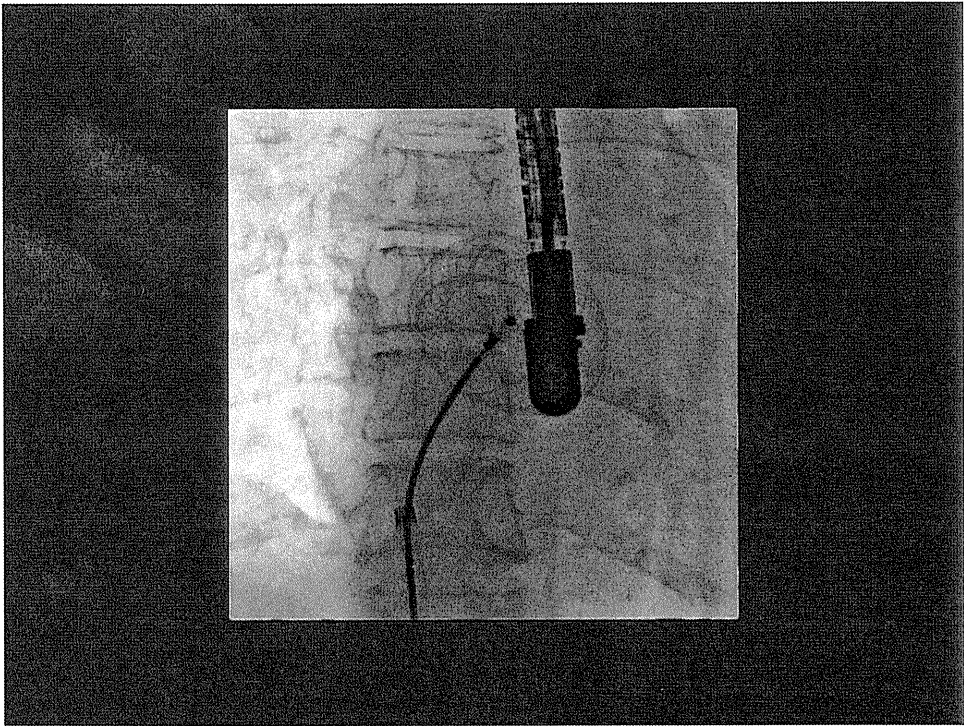
ASD was diagnosed at the time of admission of recurrent congestive heart failure and PH.

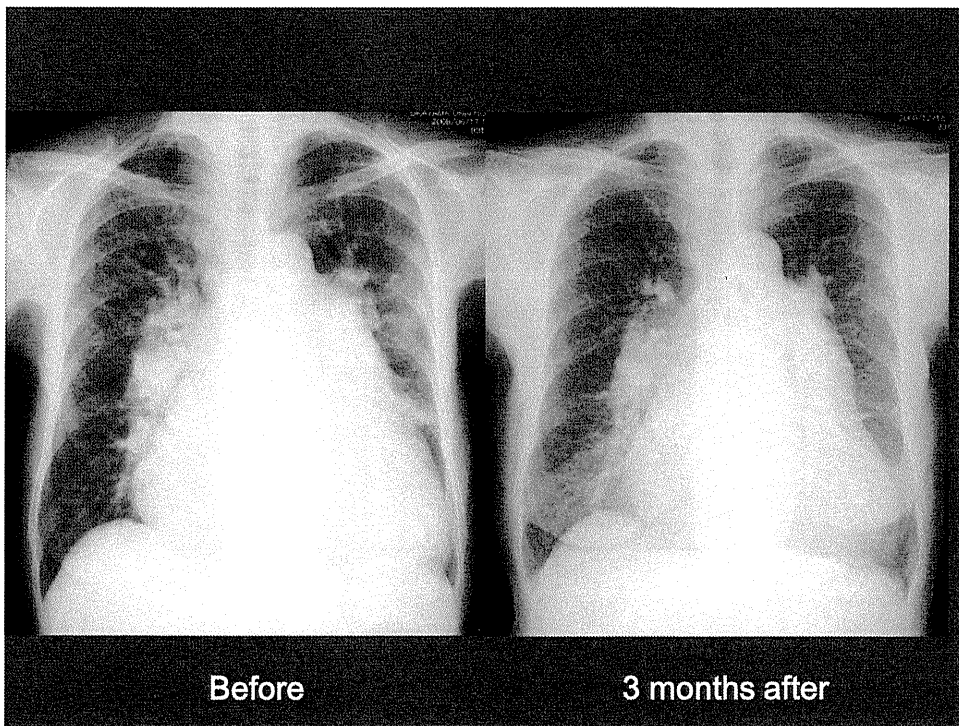
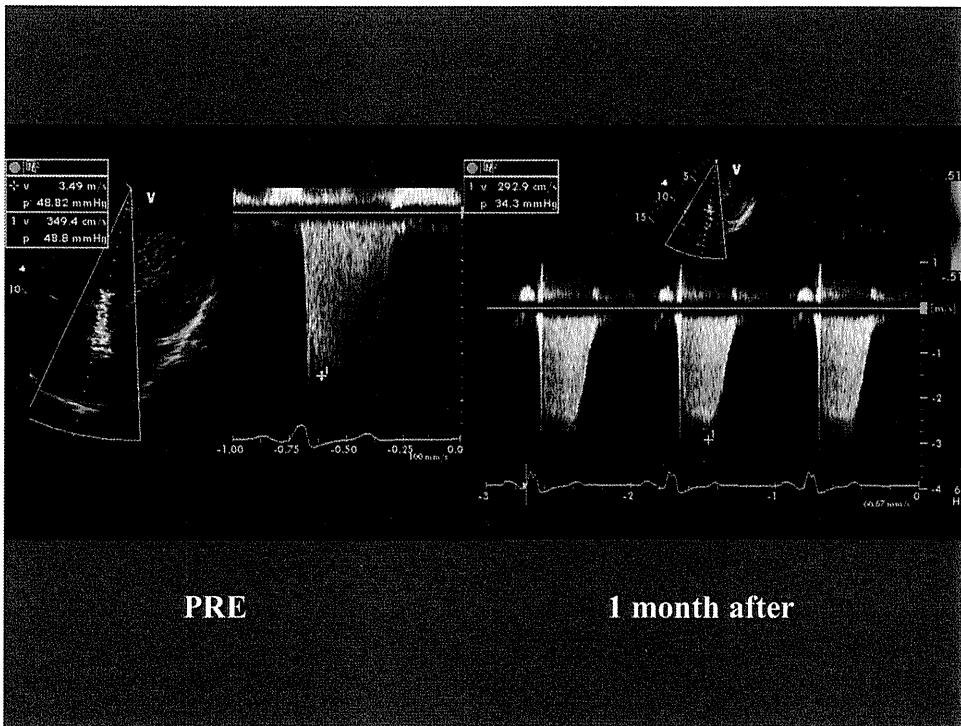










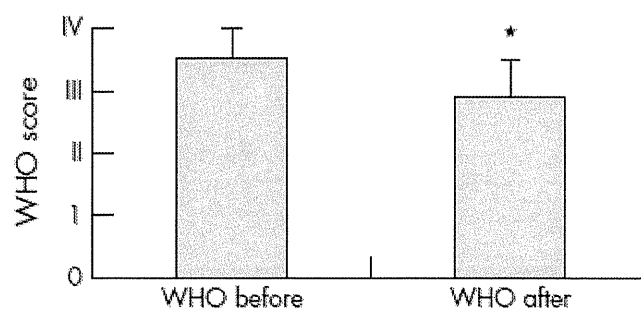


Various Conditions of ACHD with PH

- Eisenmenger Syndrome
- Left to Right Shunt disease with PH
- Post operative PH (without shunt)

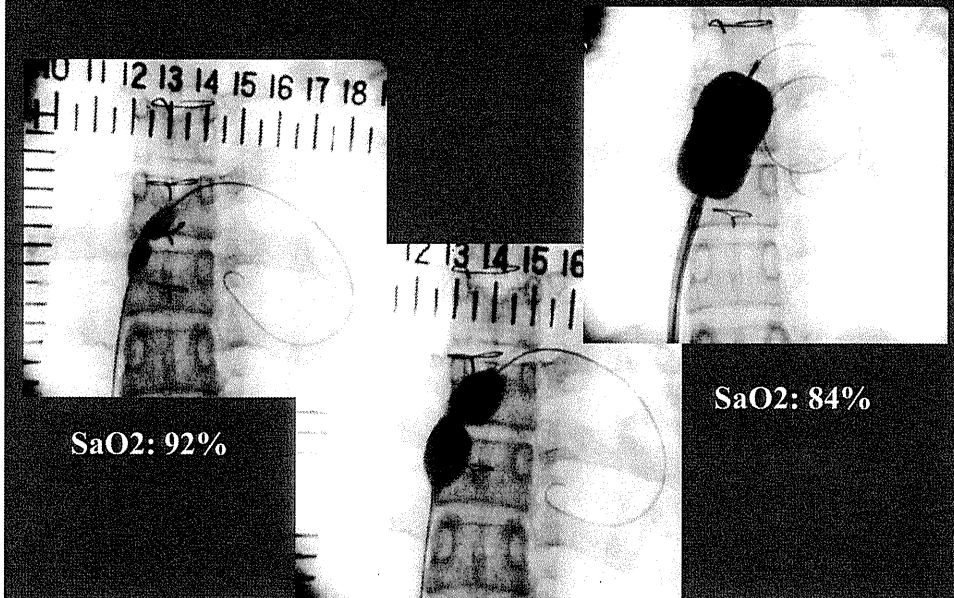
Role of atrial septostomy in the treatment of children with pulmonary arterial hypertension

A Micheletti, A A Hislop, A Lammers, P Bonhoeffer, G Derrick, P Rees, S G Haworth



Heart 2006;92:969-972. doi: 10.1136

ASD creation

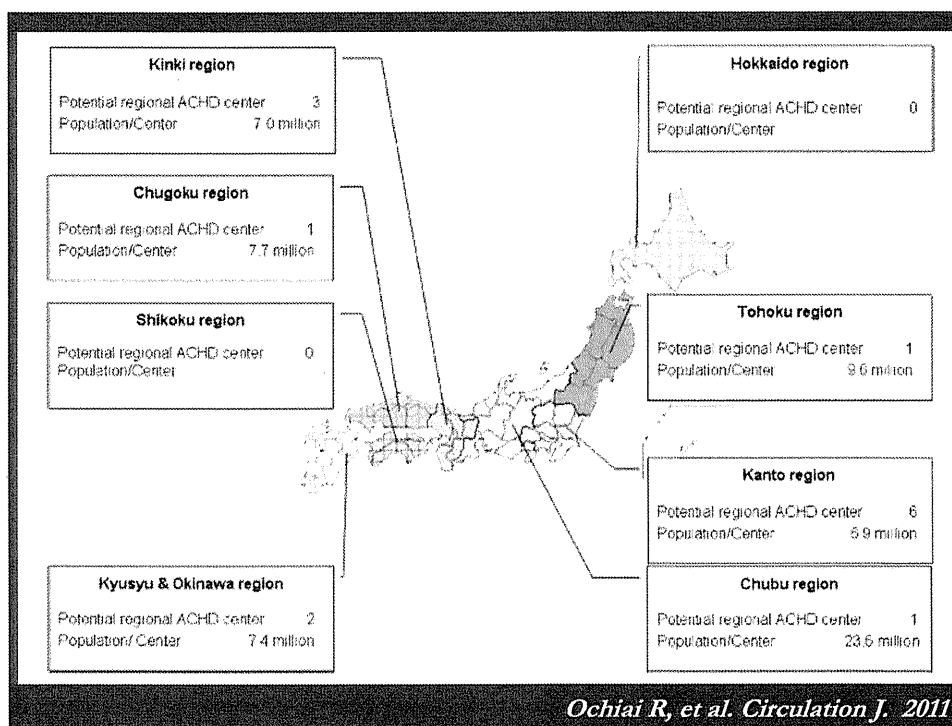


What should we inform the prognosis of ACHD with severe PH

- Effectiveness of advanced therapy
- Possibility of complete repair
- Morbidity and Mortality
- Natural course
- Quality of life
- When?, Whom? How?

Optimal ACDH Care Center

- Staff ACHD specialists at least 1, preferably 2.
- Connection with pediatric cardiology and cardiac surgery.
- Sufficient numbers of patients and perform a sufficient number of procedures to maintain high levels of performance.
- Established referral relationship with a specialist center.
- A minimum of 2 cardiac surgeons practicing adult and pediatric cardiac surgery
- >125 operations/year for CHD. >50 per year for ACHD
- Fully equipped electrophysiology laboratory staffed experience in pacemaker, ablation, and defibrillator.
- At least 1 nurse specialist for care of ACHD patients.



Clinical Issues of Adult CHD

- Establishment of medical staffs
- Establishment of facilities
- Establishment of management guideline
- Establishment of medical group (society)
- Recognition by society

症例1

27歳, 女性

VSD, PDA根治術後, Eisenmenger症候群

在胎40週, 3150gにて出生.

4生日にVSDの診断.

4歳時に根治術.

肺高血圧が残存 (PAP:103/61(76)mmHg, PVRI: 22)

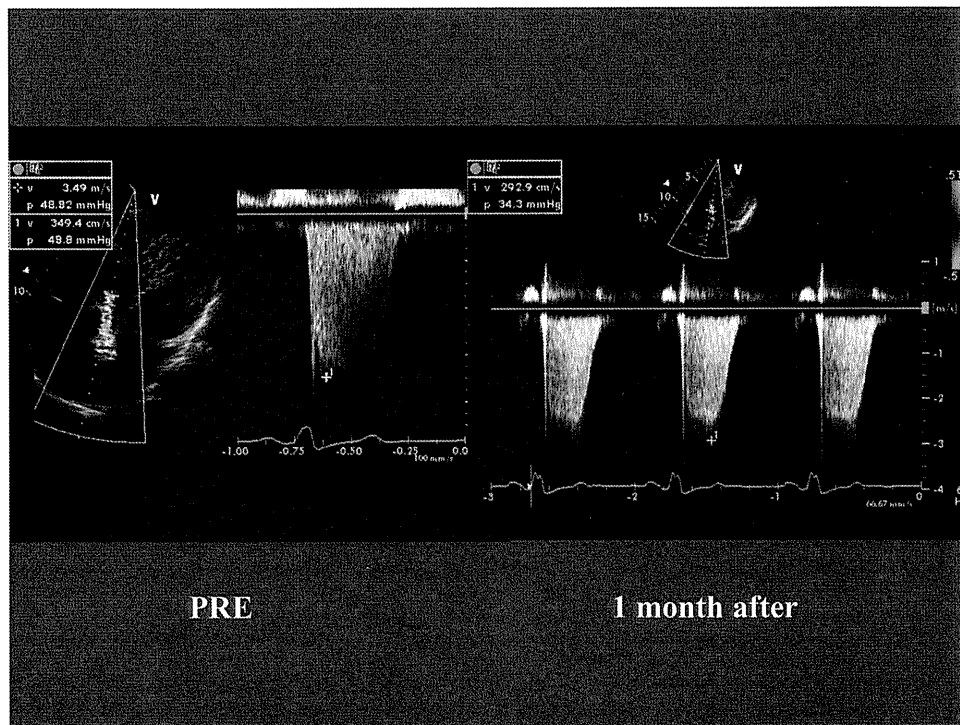
10歳時に運動中に失神.

18歳時に全身倦怠感が増強し歩行時にも失神.

Beraprost経口投与するも症状改善せず.

21歳時に, 経カテーテル的ASD作成術.

23歳時に, 2回目の経カテーテル的ASD作成術.



症例2

45歳, 女性.

PA with VSD, APCA

生後6ヶ月時に上記診断.

22歳時に上行大動脈右肺動脈短絡術.

42歳時より喀血を認めるようになる.

44歳時にAPCAに対する

経カテーテル的コイル閉鎖術.

Coil occlusion to aortopulmonary collateral arteries

