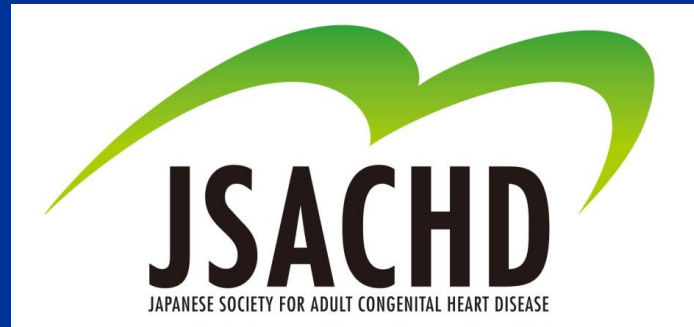
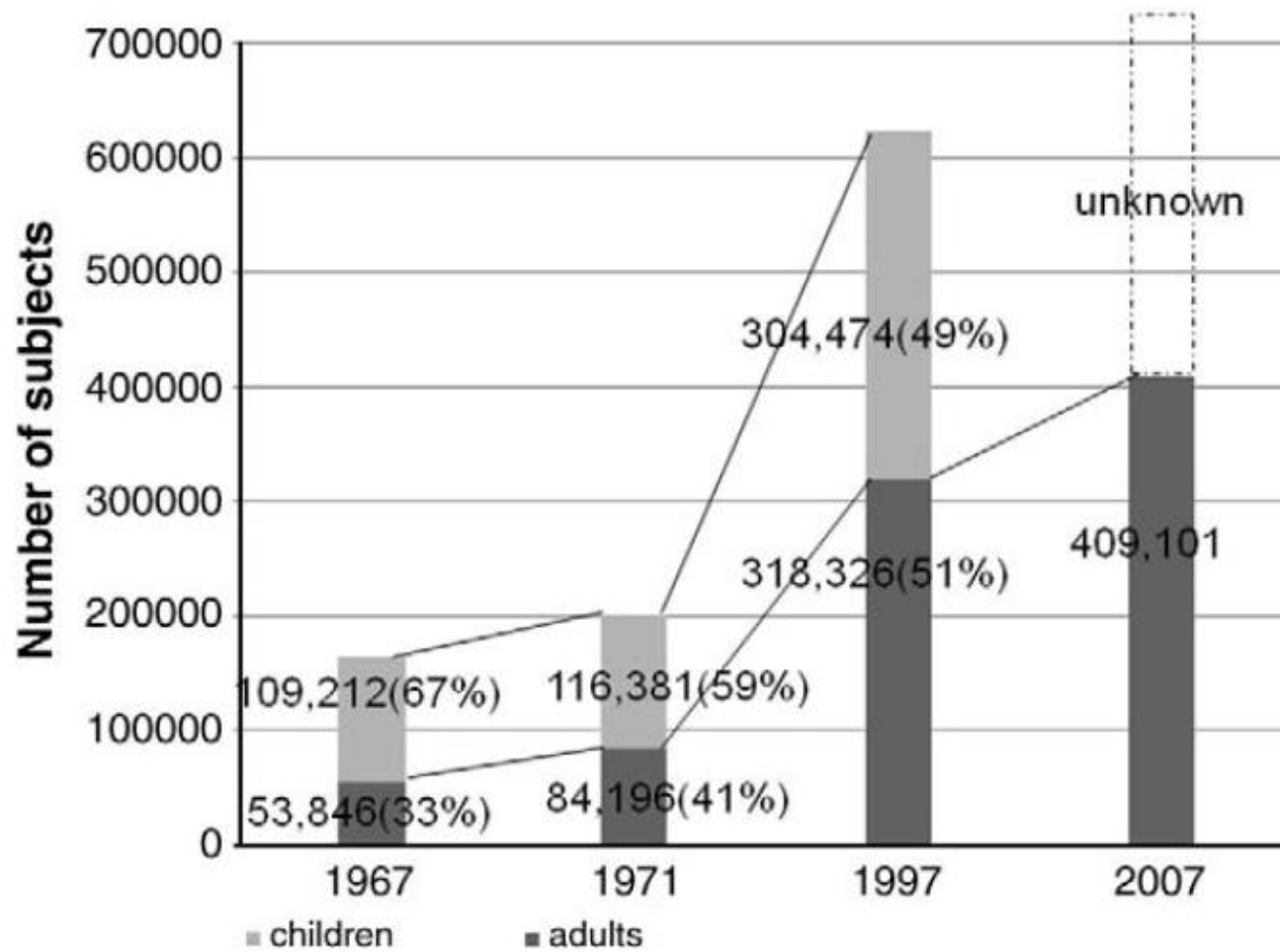


Management of Adult Patients with Congenital Heart Disease and Severe Pulmonary Hypertension

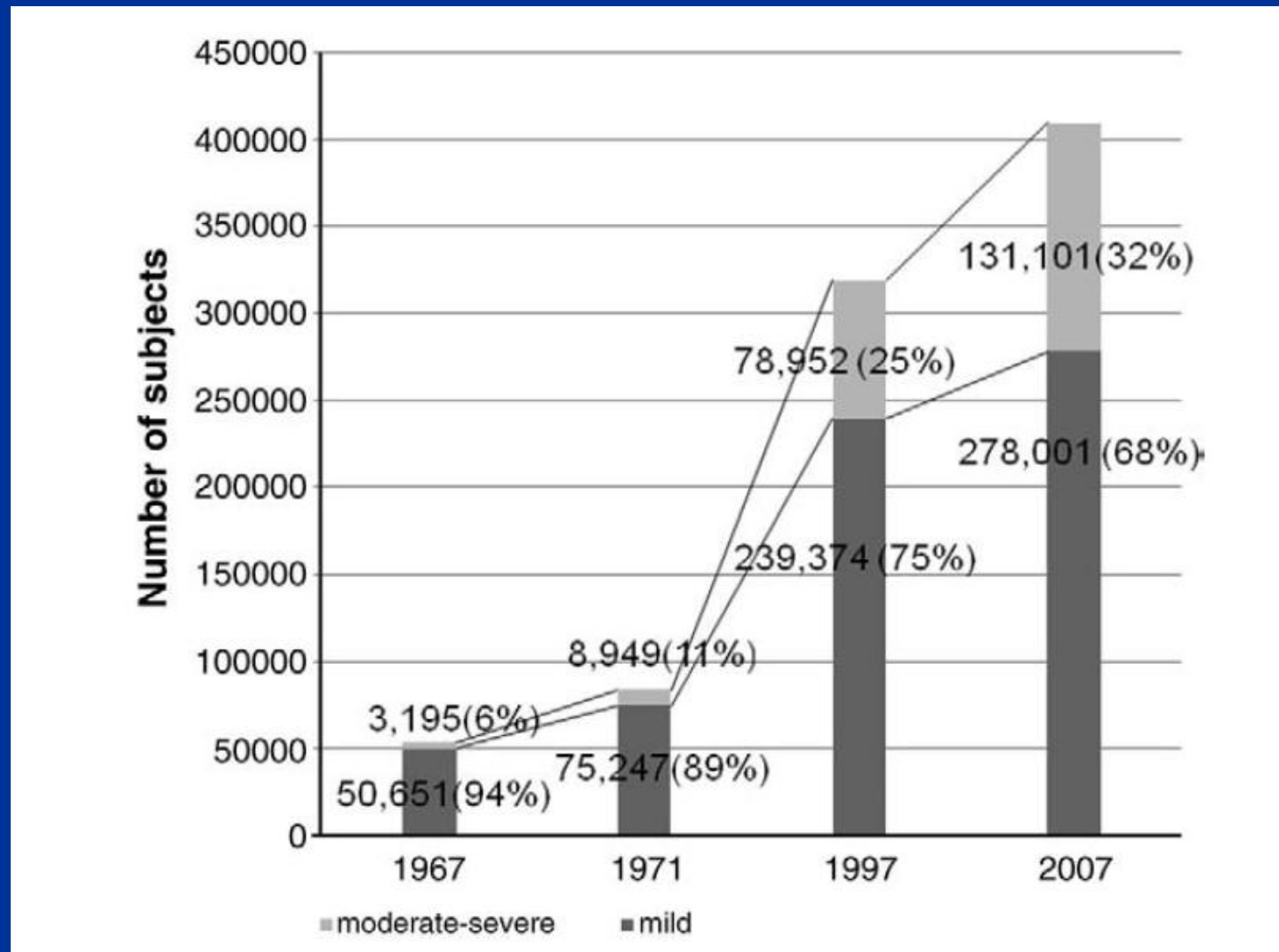


Teiji Akagi, MD, PhD
Cardiac Intensive Care Unit,
Okayama University, Okayama, Japan

Prevalence of adult patients with CHD in Japan



Prevalence of adult patients with CHD in Japan

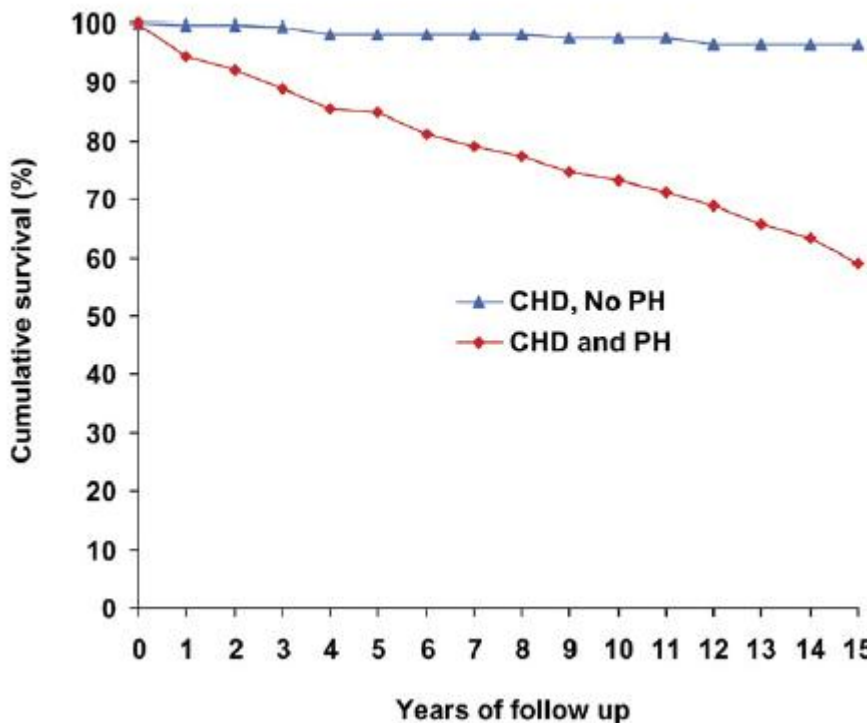


Diagnosis of Pulmonary Hypertension in the Congenital Heart Disease Adult Population

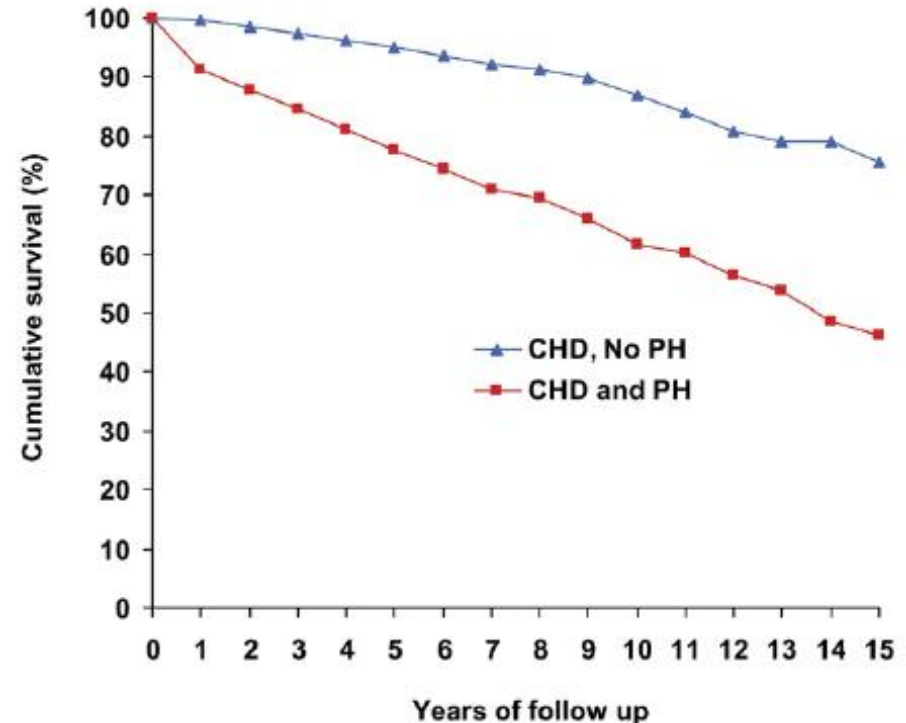
Impact on Outcomes

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*

Age 18 – 39 years



Age 40 – 64 years



Various Conditions of ACHD with PH

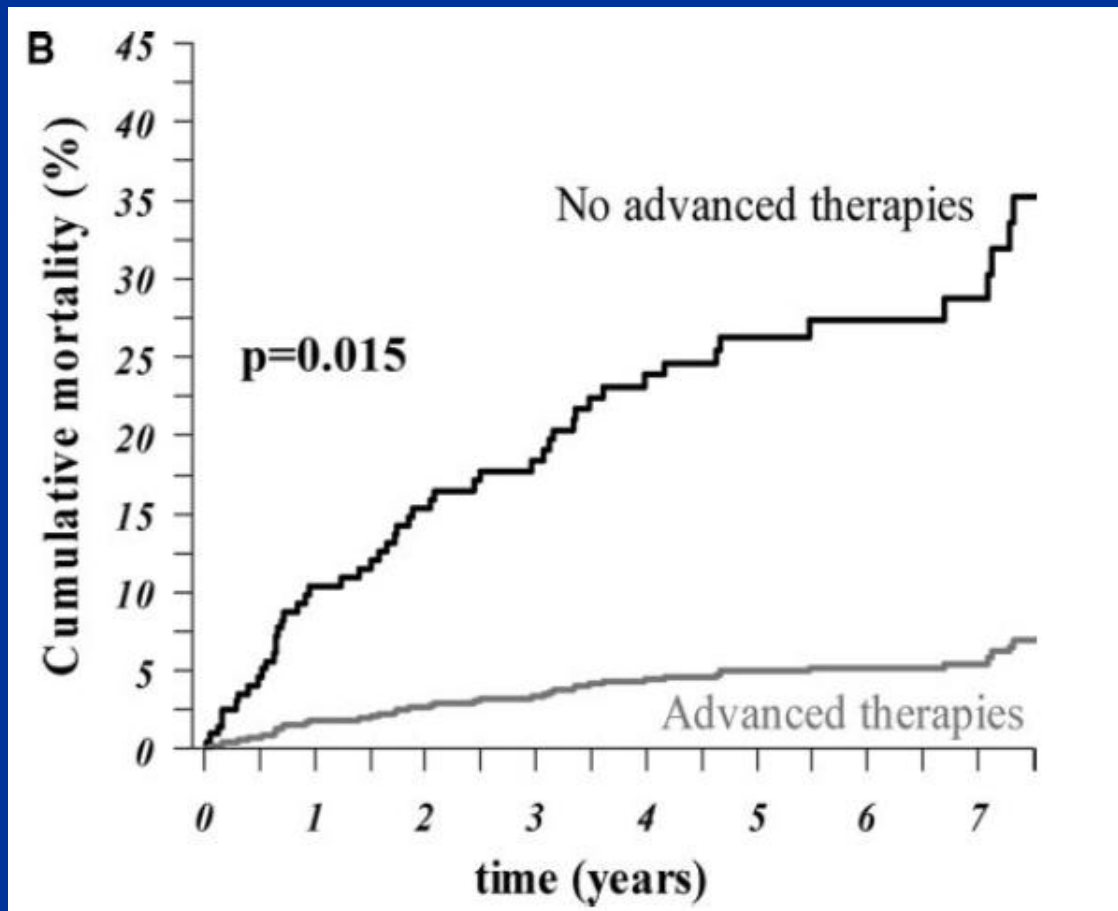
- Eisenmenger Syndrome
- Left to Right Shunt disease with PH
- Post operative PH (without shunt)

Various Conditions of ACHD with PH

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

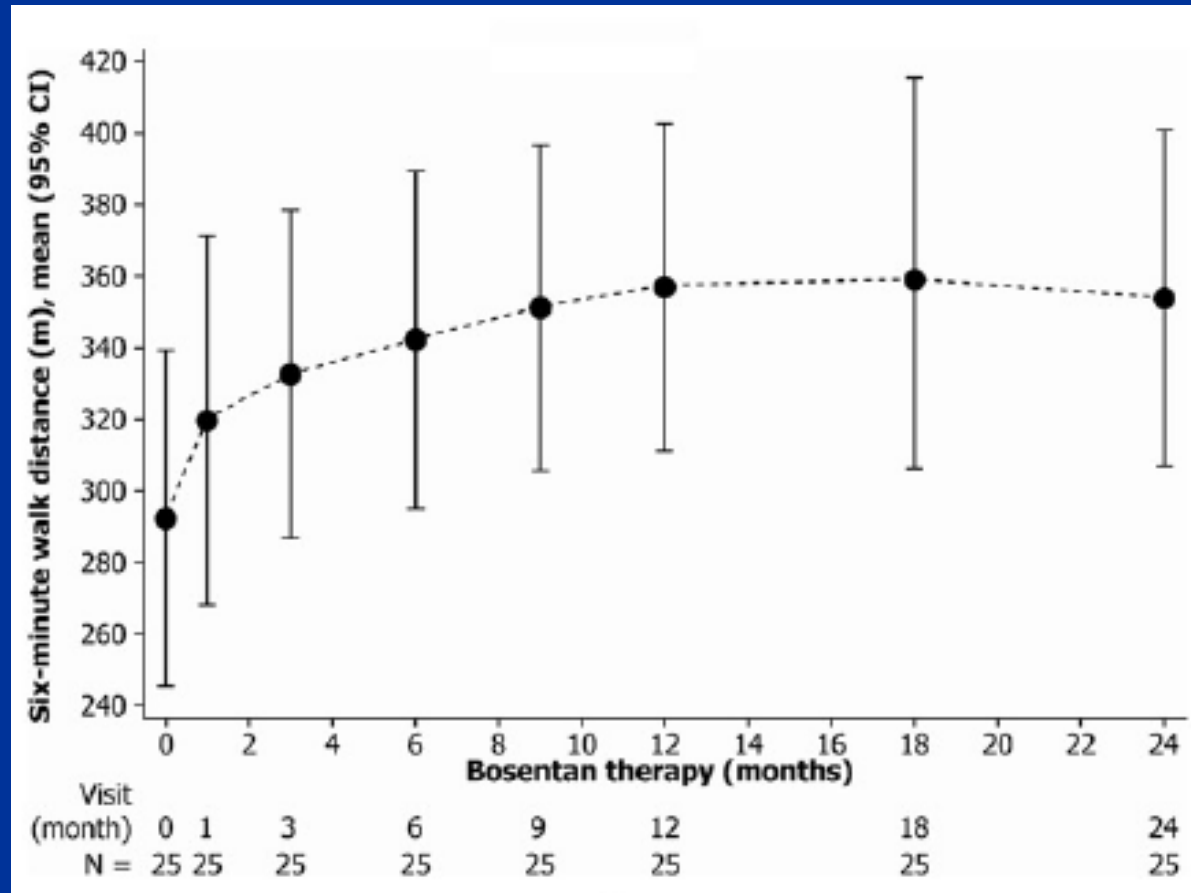
Improved Survival Among Patients With Eisenmenger Syndrome Receiving Advanced Therapy for Pulmonary Arterial Hypertension

Konstantinos Dimopoulos, MD, MSc, PhD, FESC*; Ryo Inuzuka, MD*; Sara Goletto, MD; Georgios Giannakoulas, MD, PhD, FESC; Lorna Swan, MD, MRCP; Stephen J. Wort, BA, MBBS, MRCP, PhD; Michael A. Gatzoulis, MD, PhD, FESC



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,*}



What is the goal of management in ACHD patients with PH

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

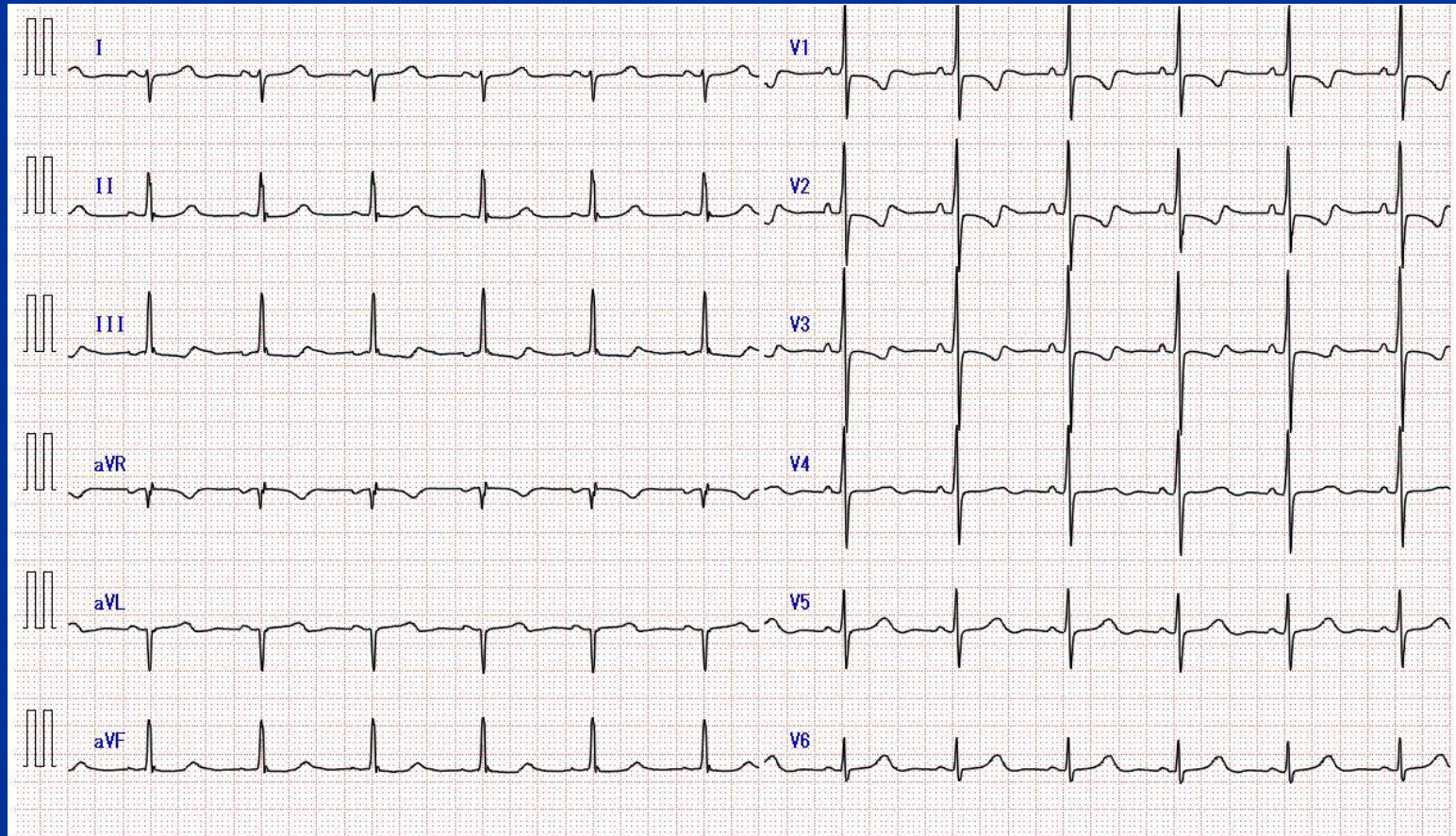
Various Conditions of ACHD with PH

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

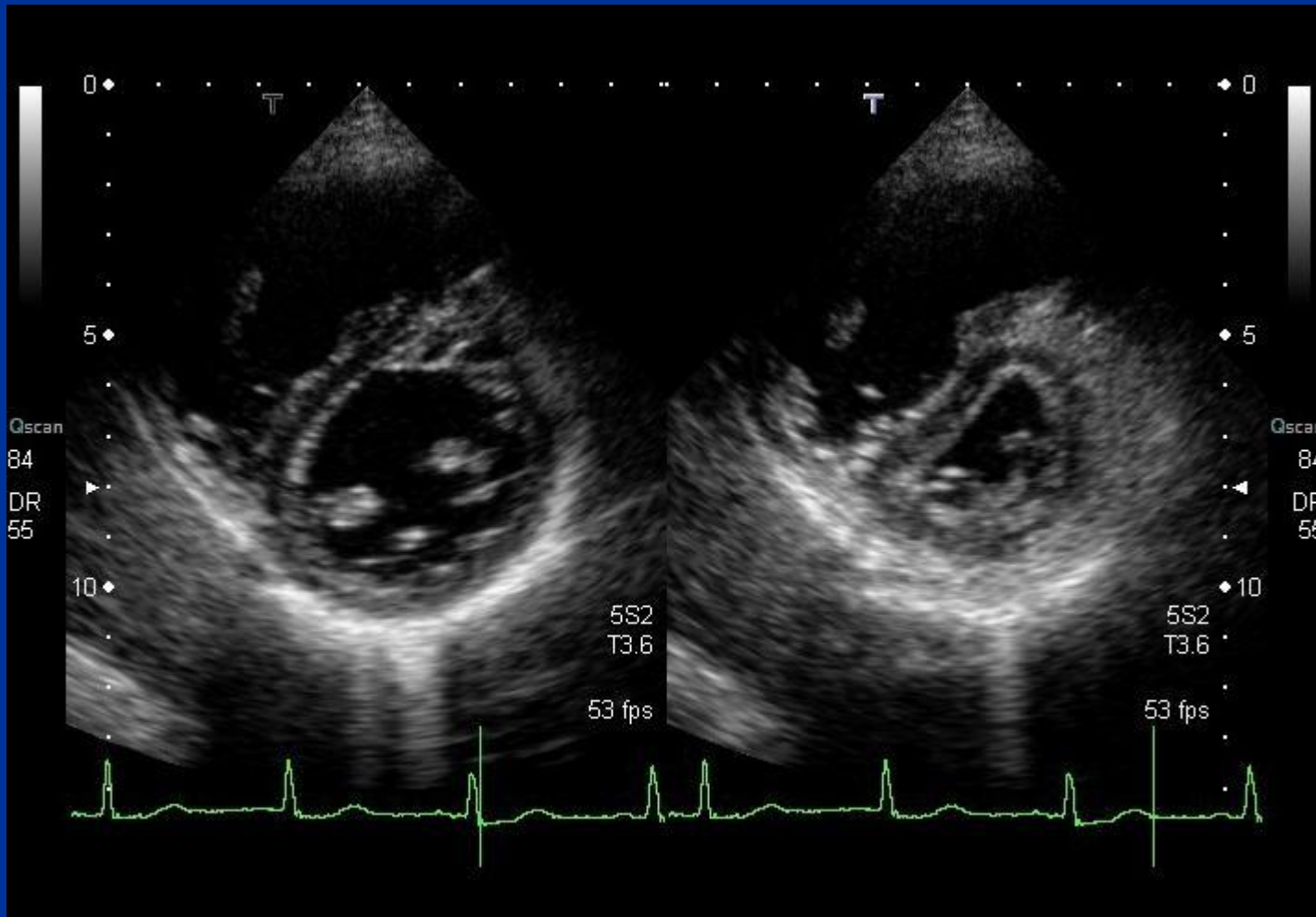
What is the goal of management in ACHD patients with PH

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

32 years female







PA: 82/31 (m=51) Ao: 96/44 (m=58)

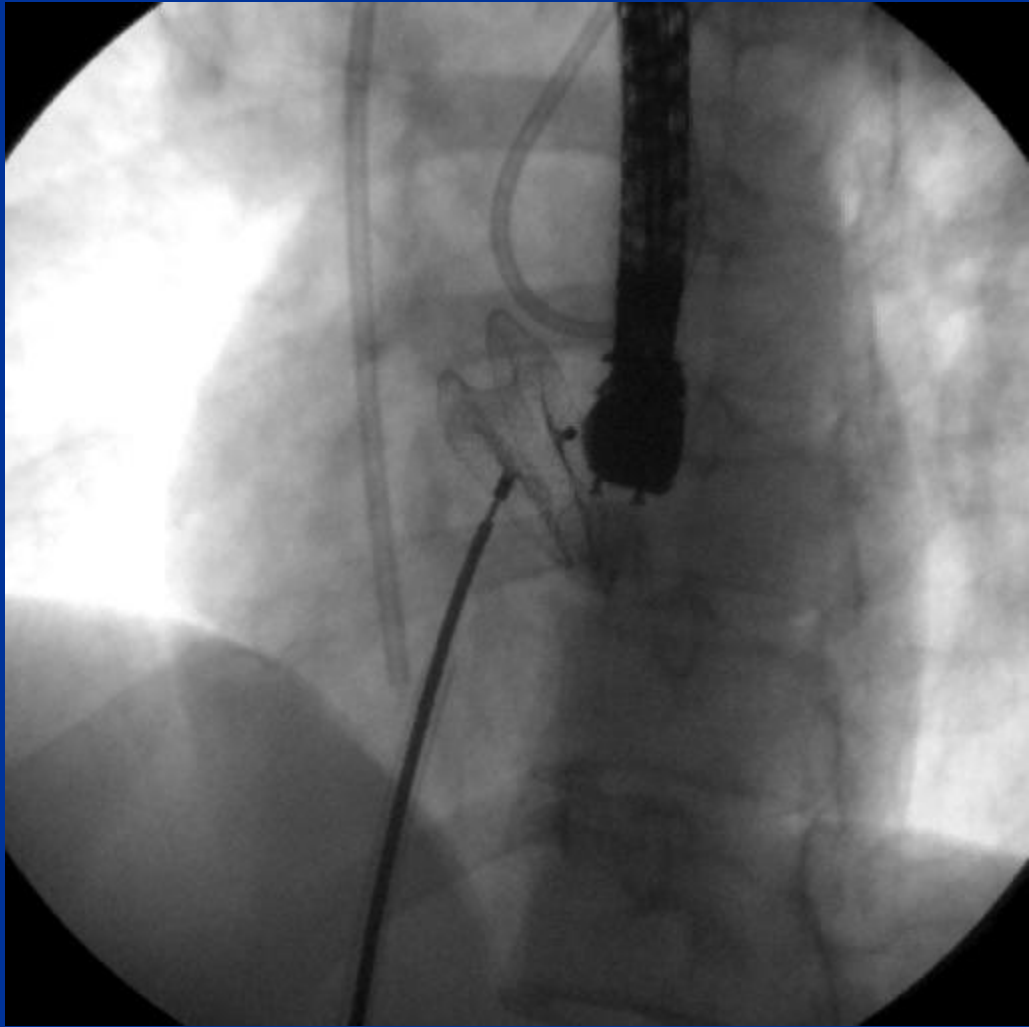
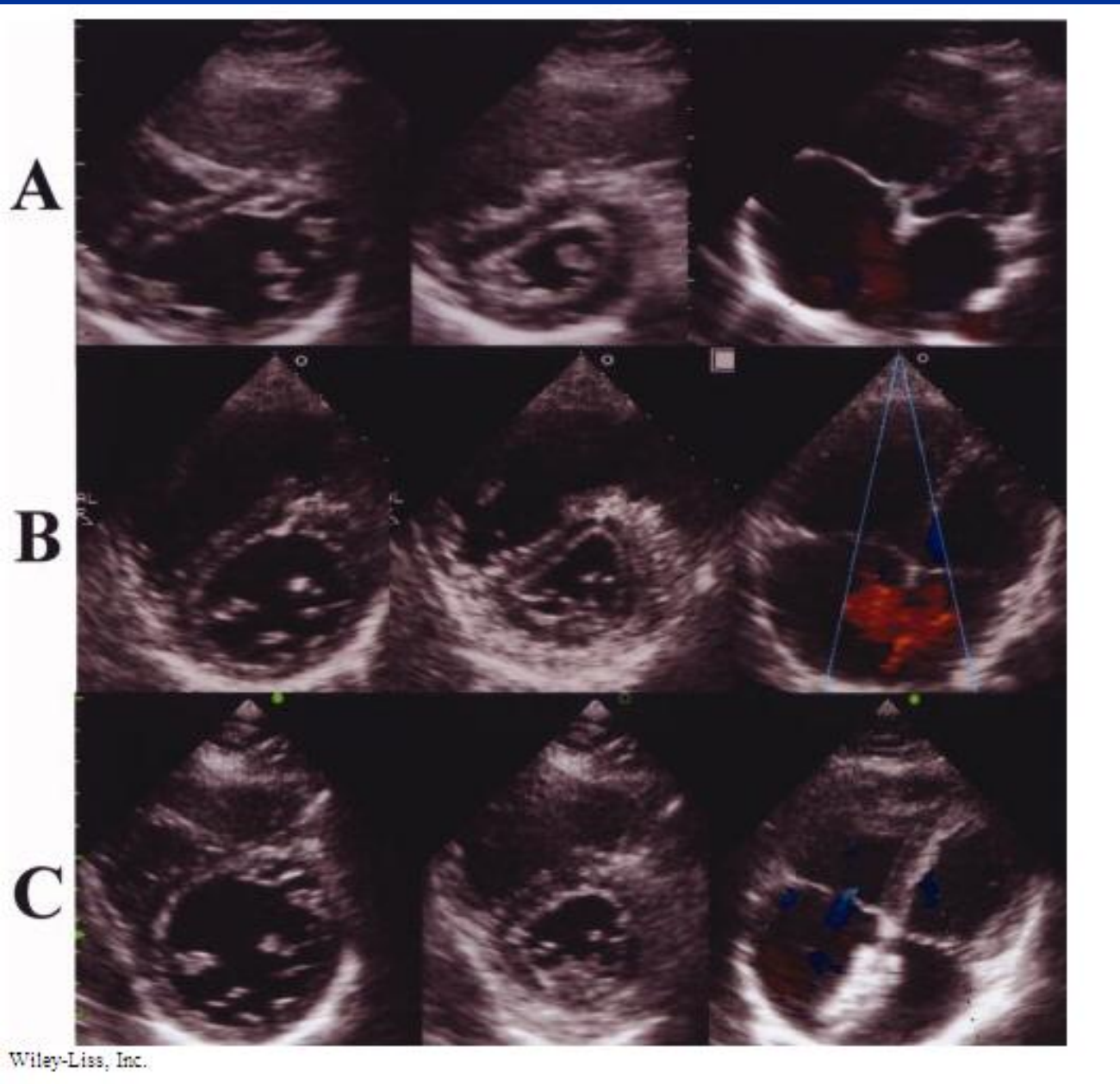
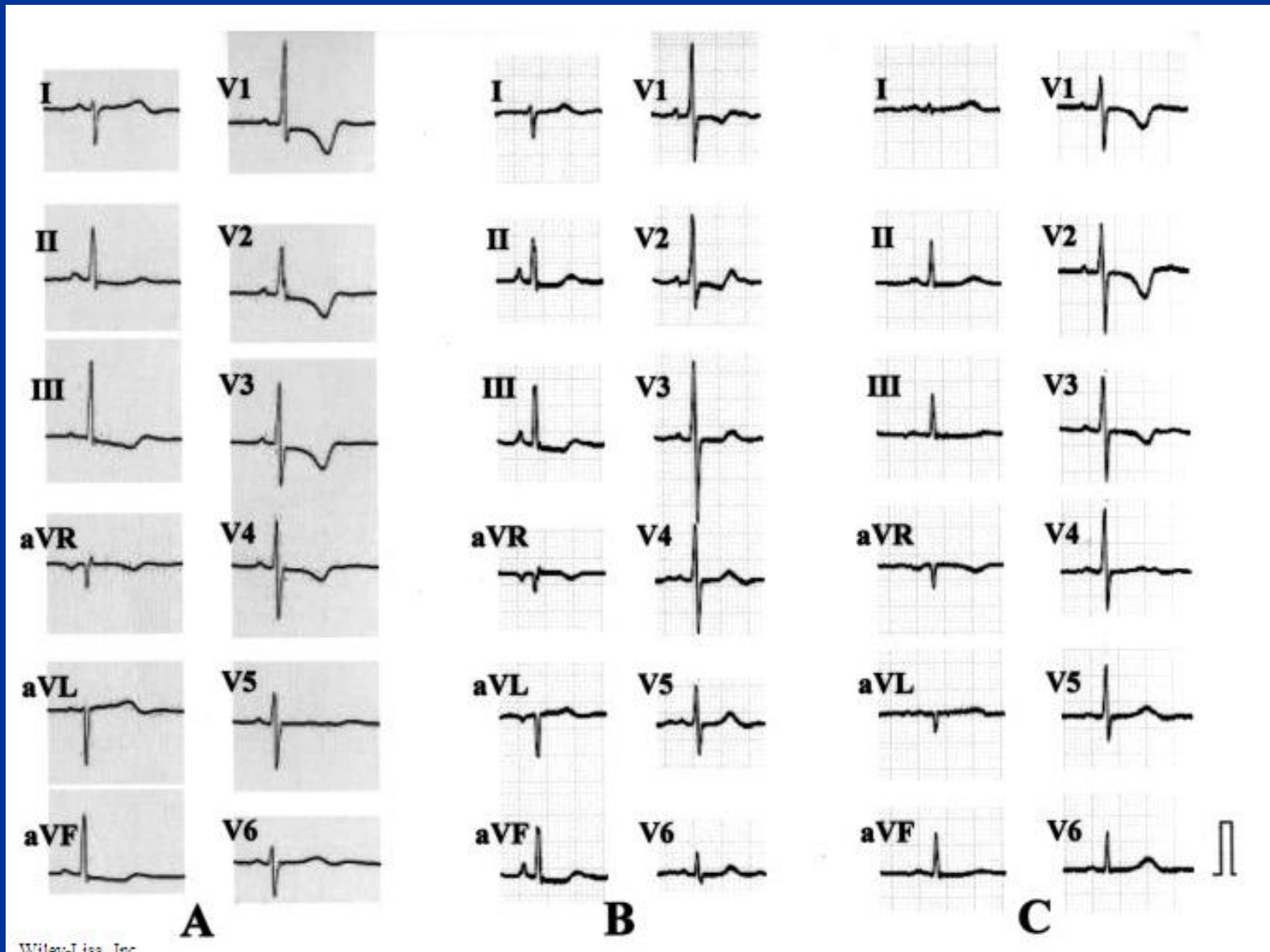


Table I. Sequential Hemodynamic Data by Cardiac Catheterization

	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



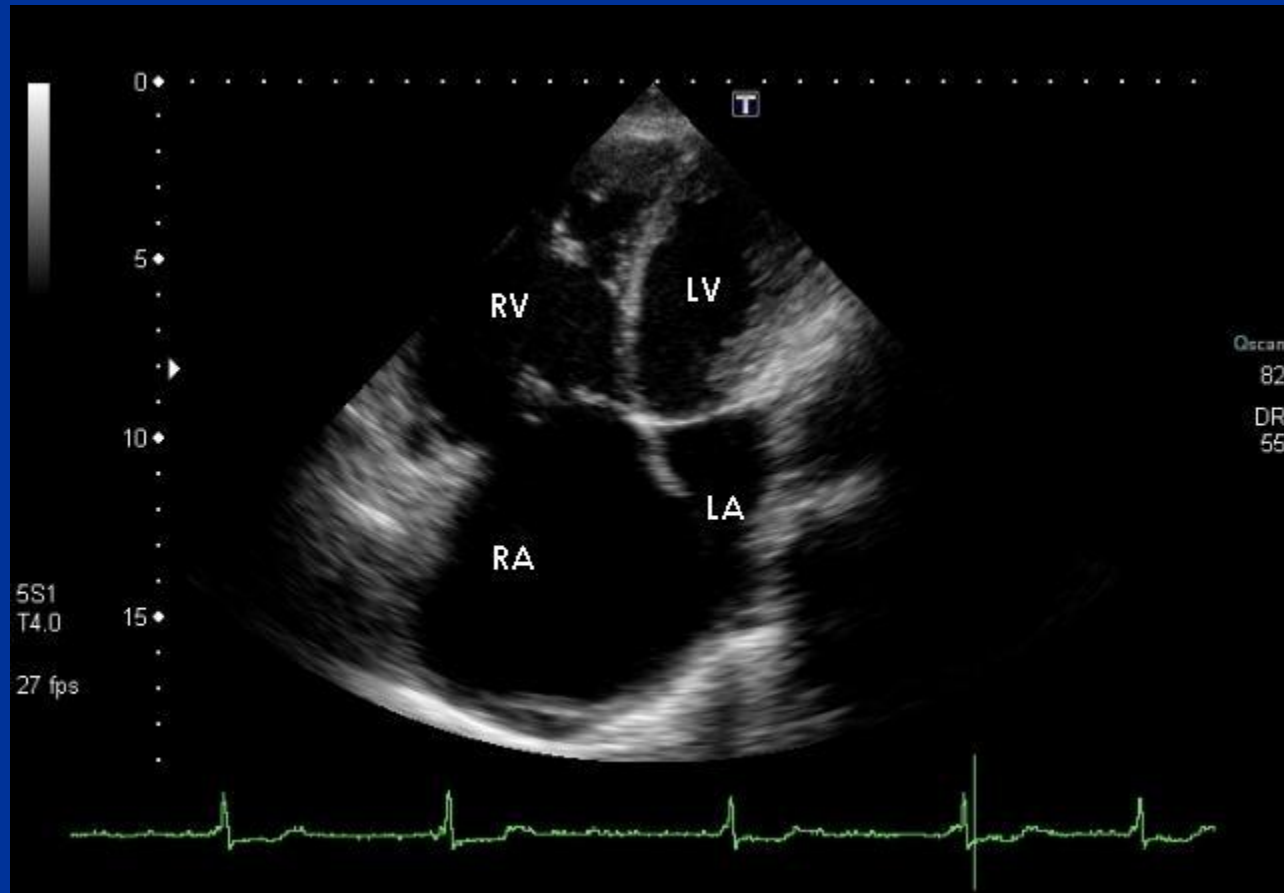
Wiley-Liss, Inc.

Case# . 82 years old, female.

History:

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





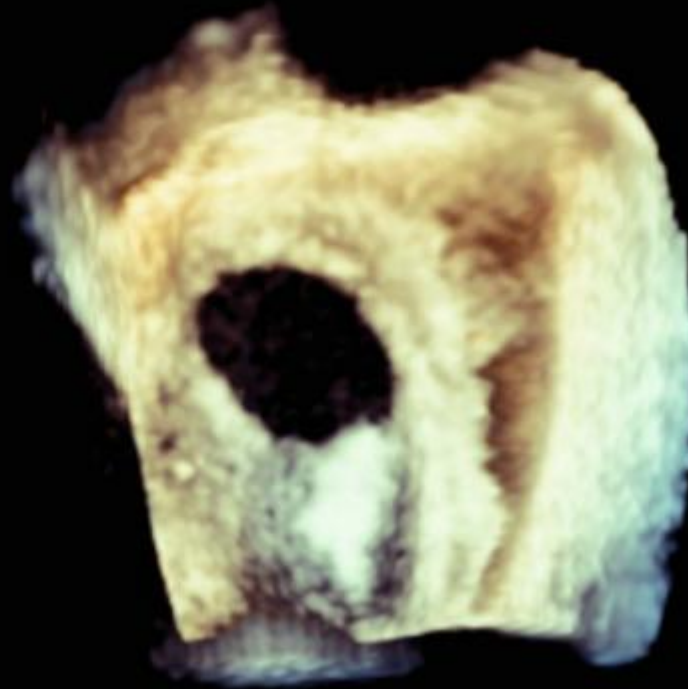
0005660901

X7-2t/3DTEE

FR 9Hz
4.5cm

M4

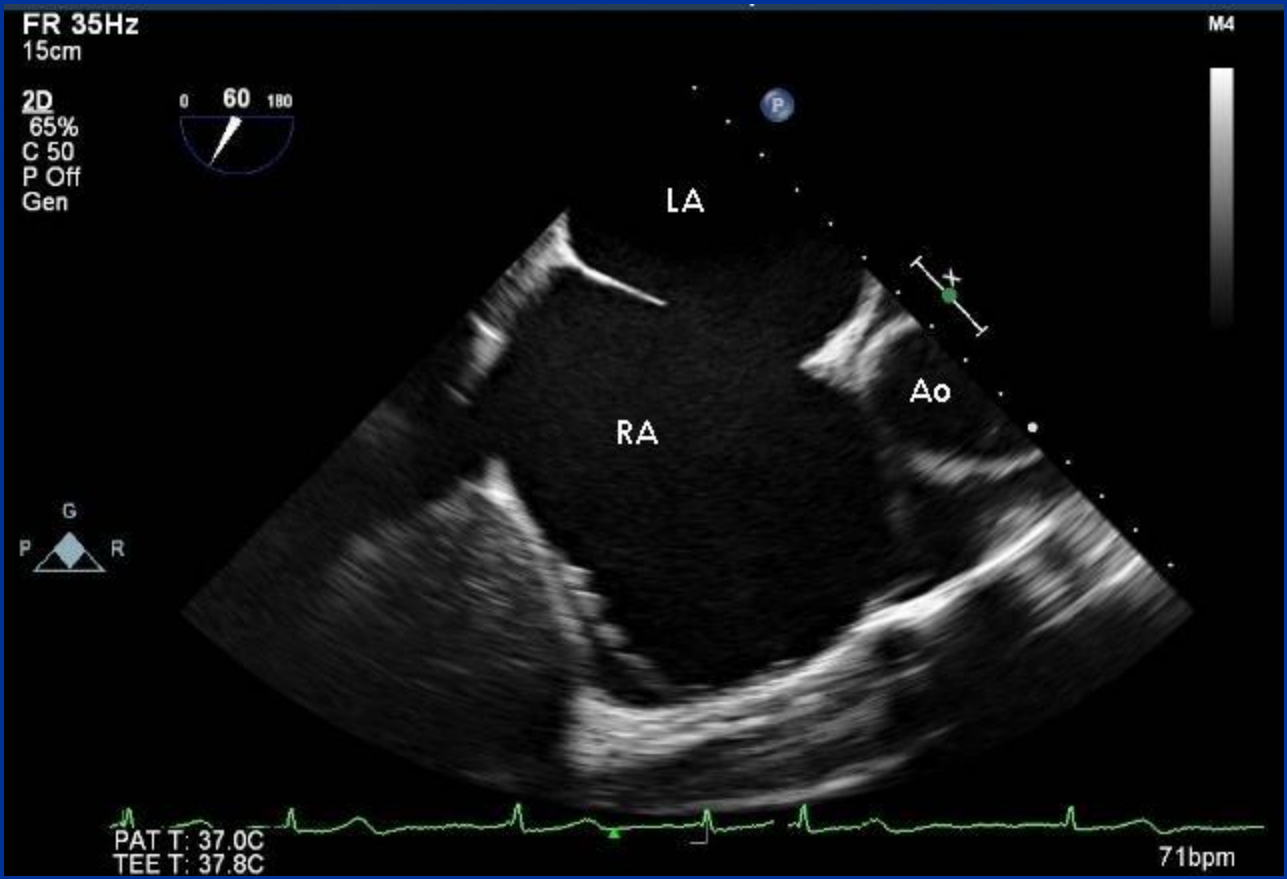
Live 3D
3D 47%
3D 40dB
Gen

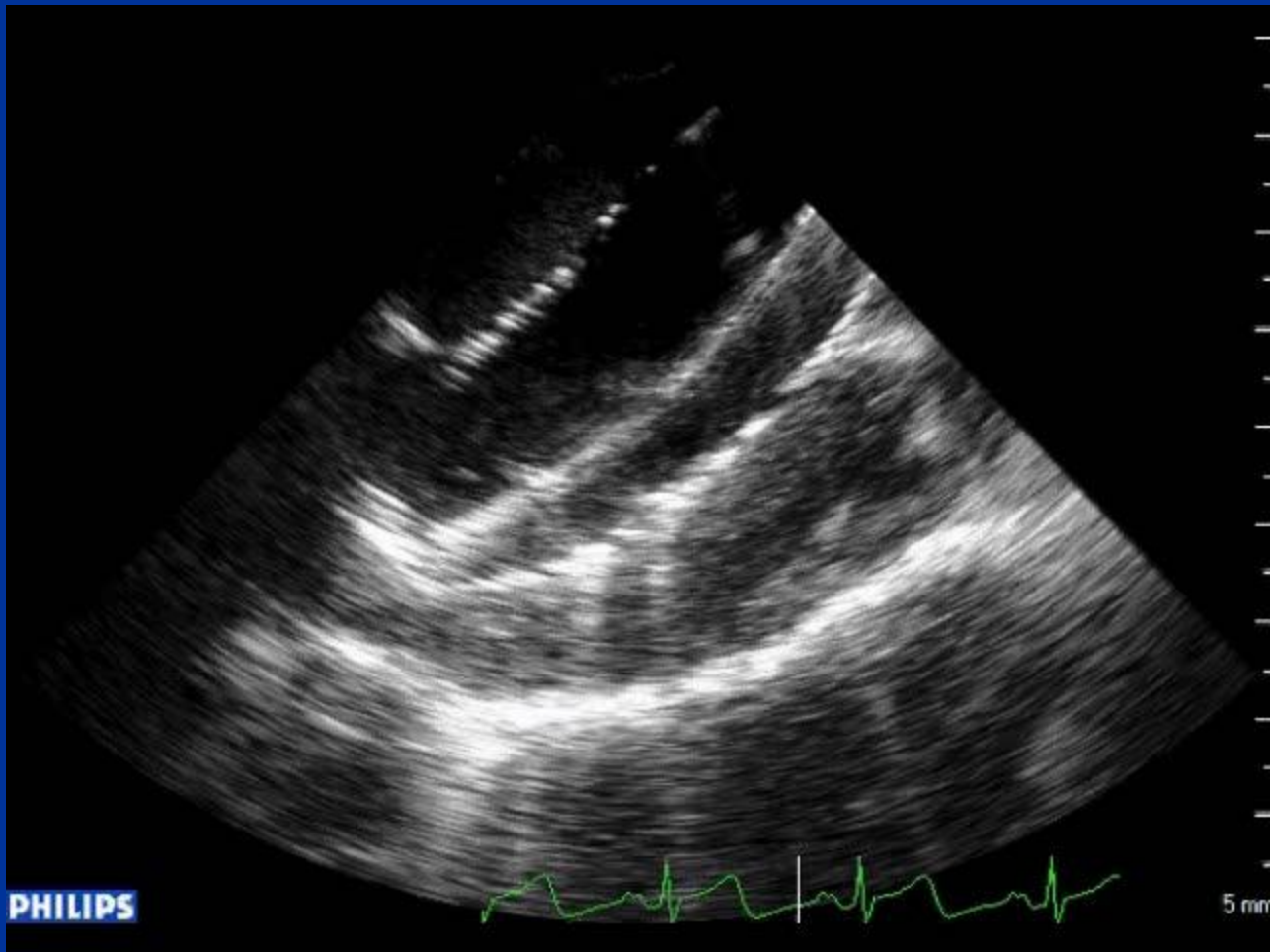


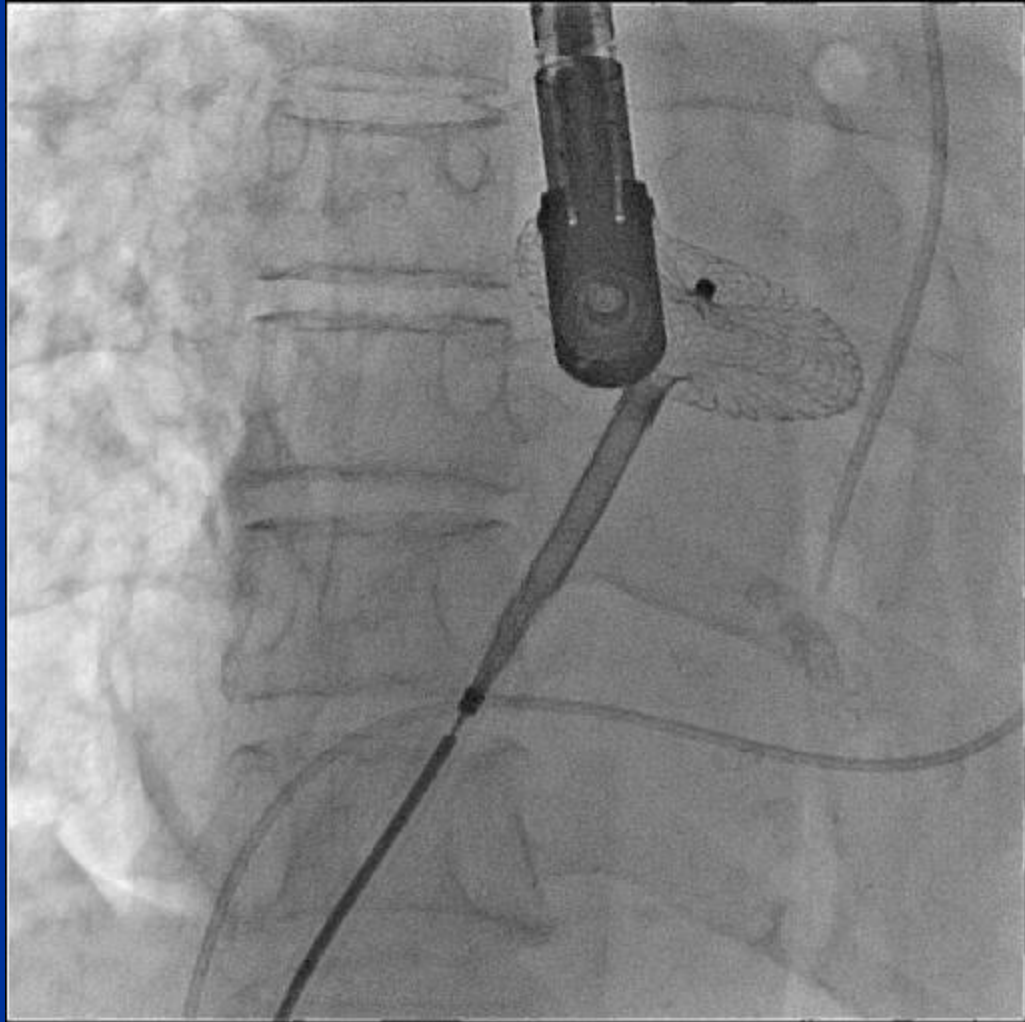
JPEG

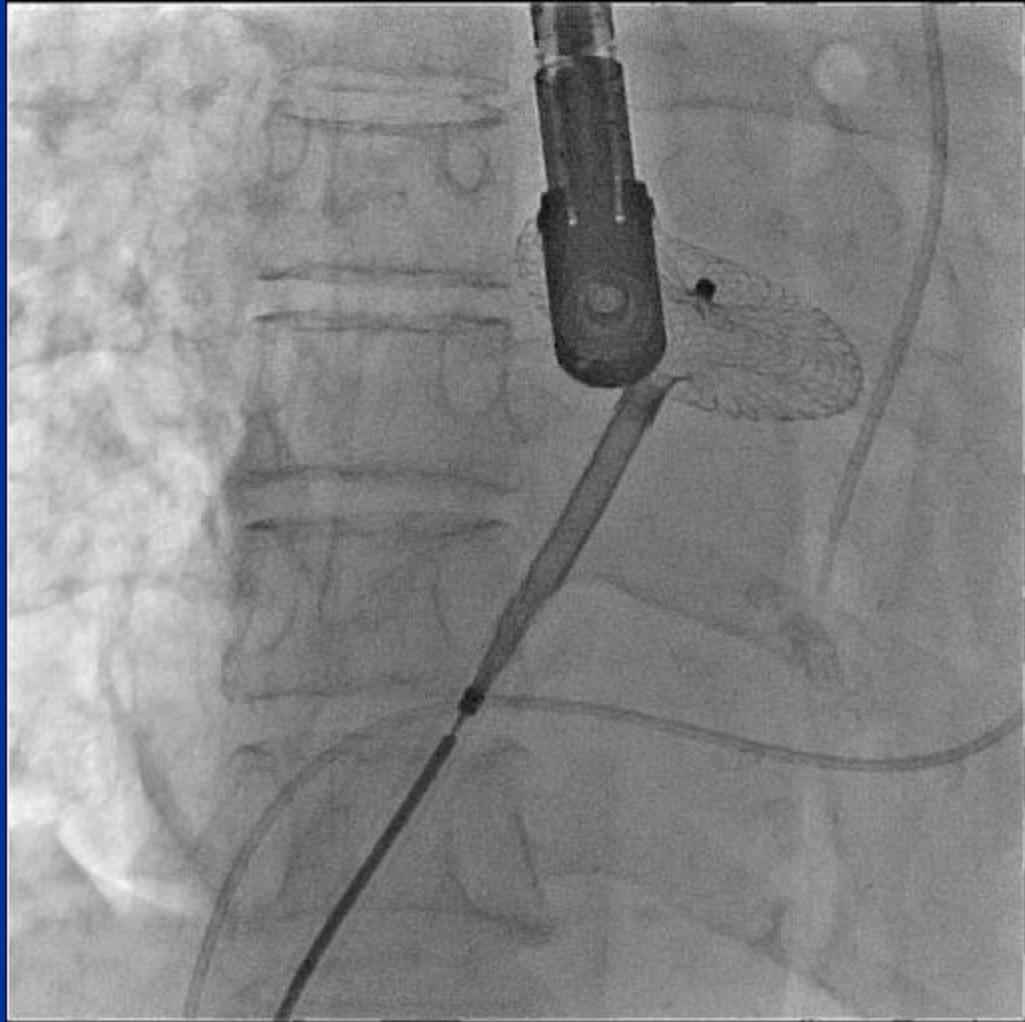
PAT T: 37.0C
TEE T: 38.9C

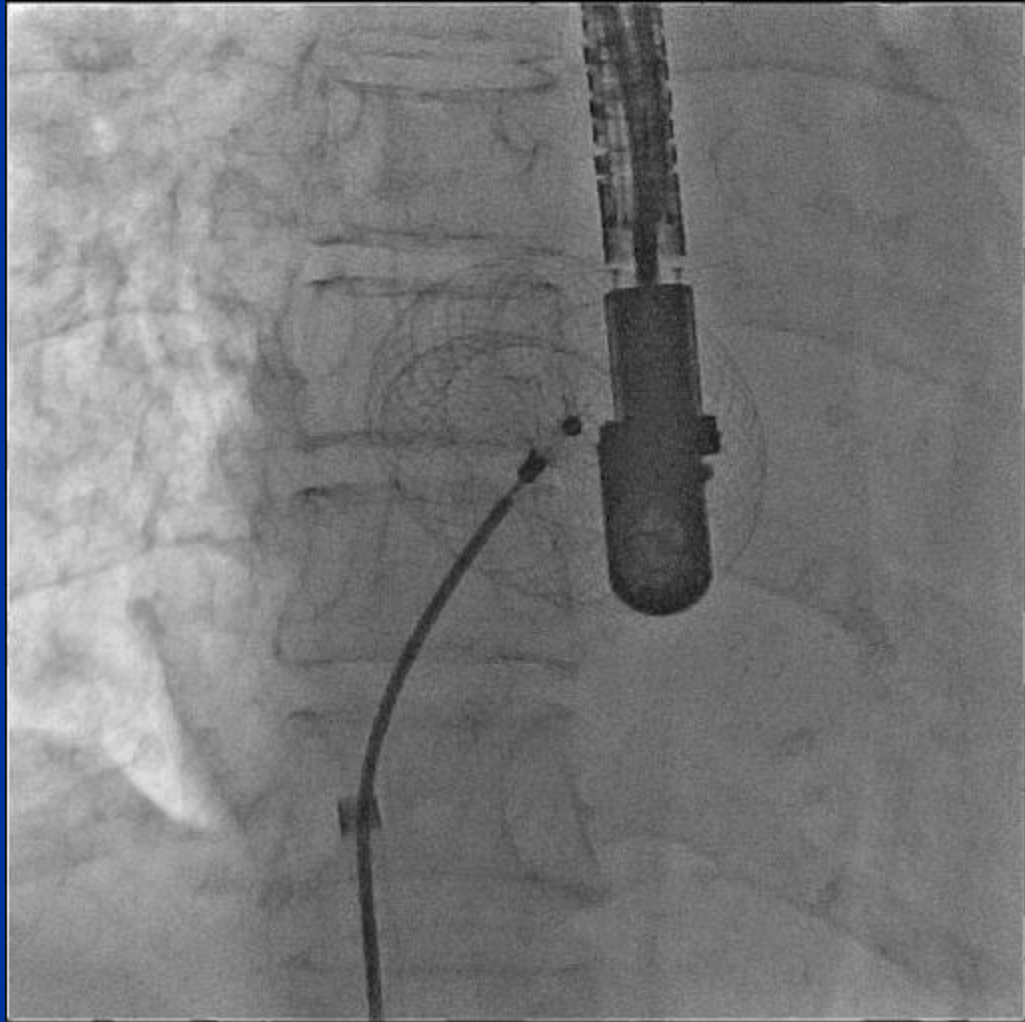
48 bpm

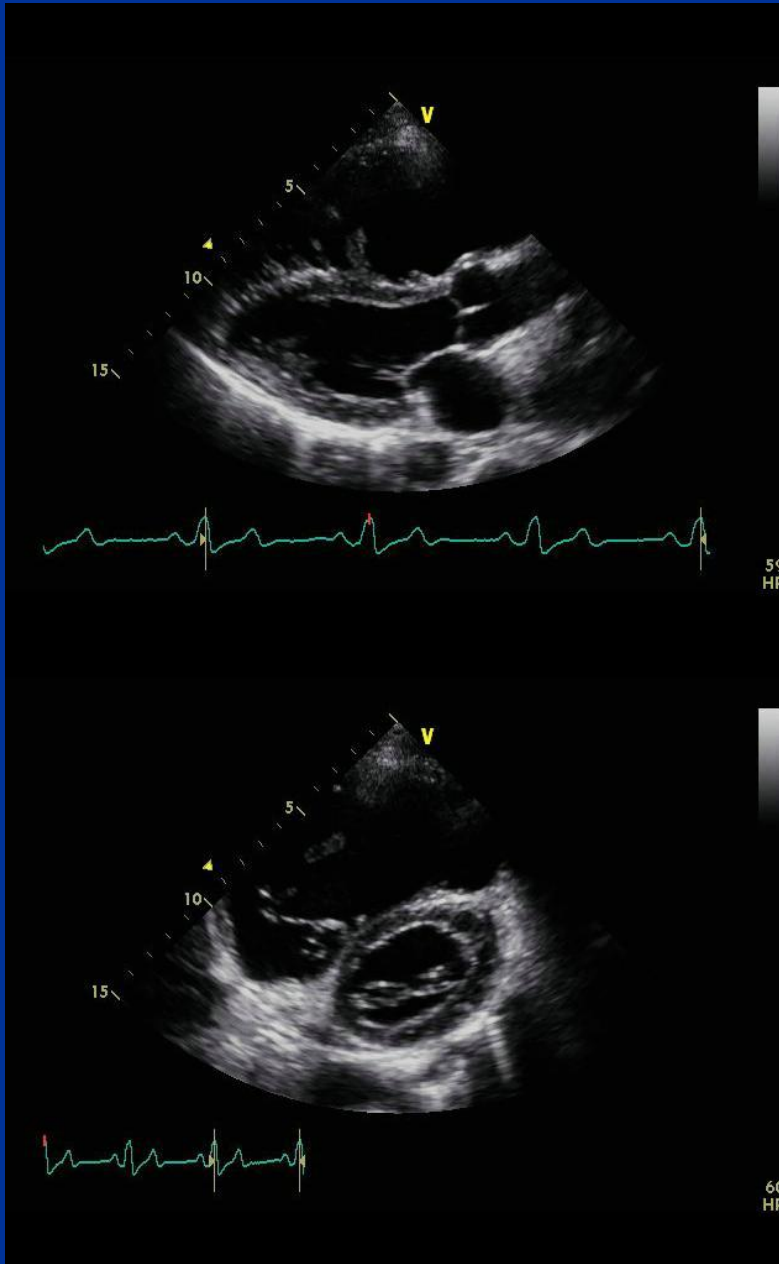




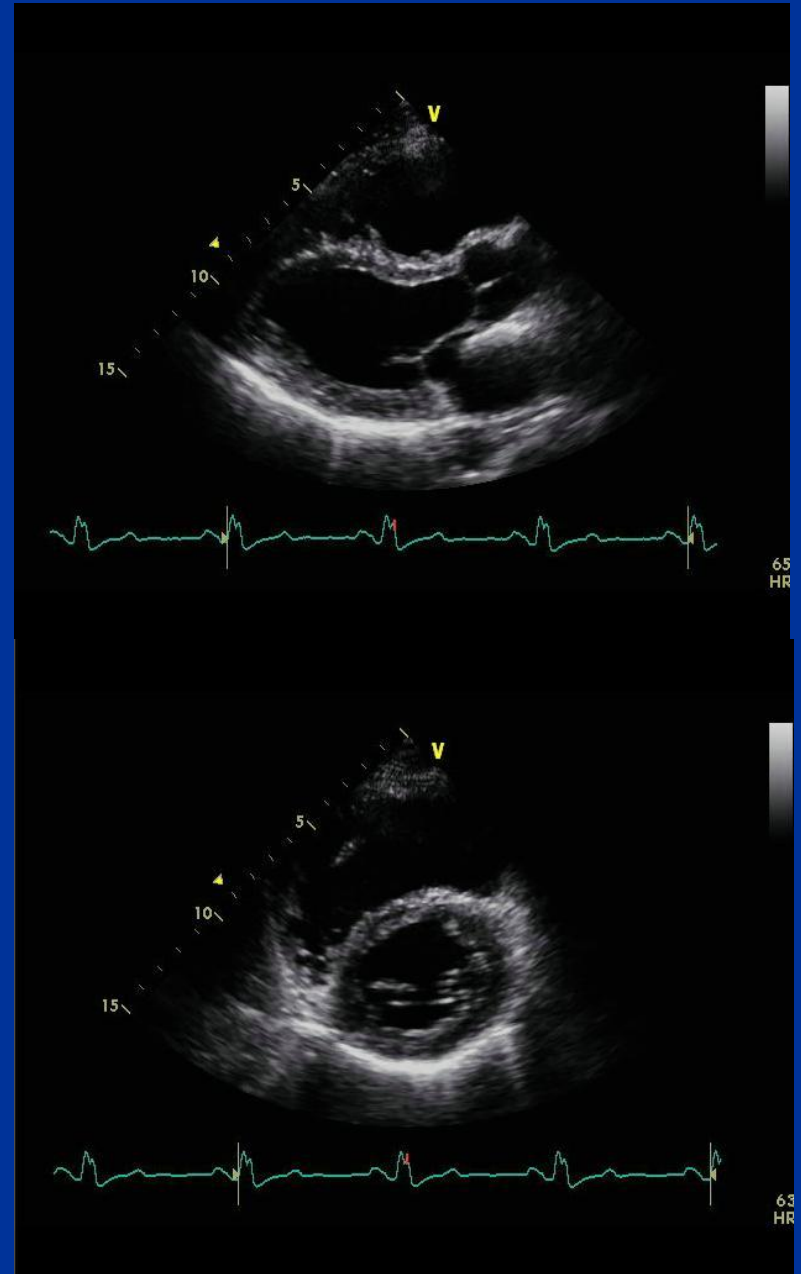




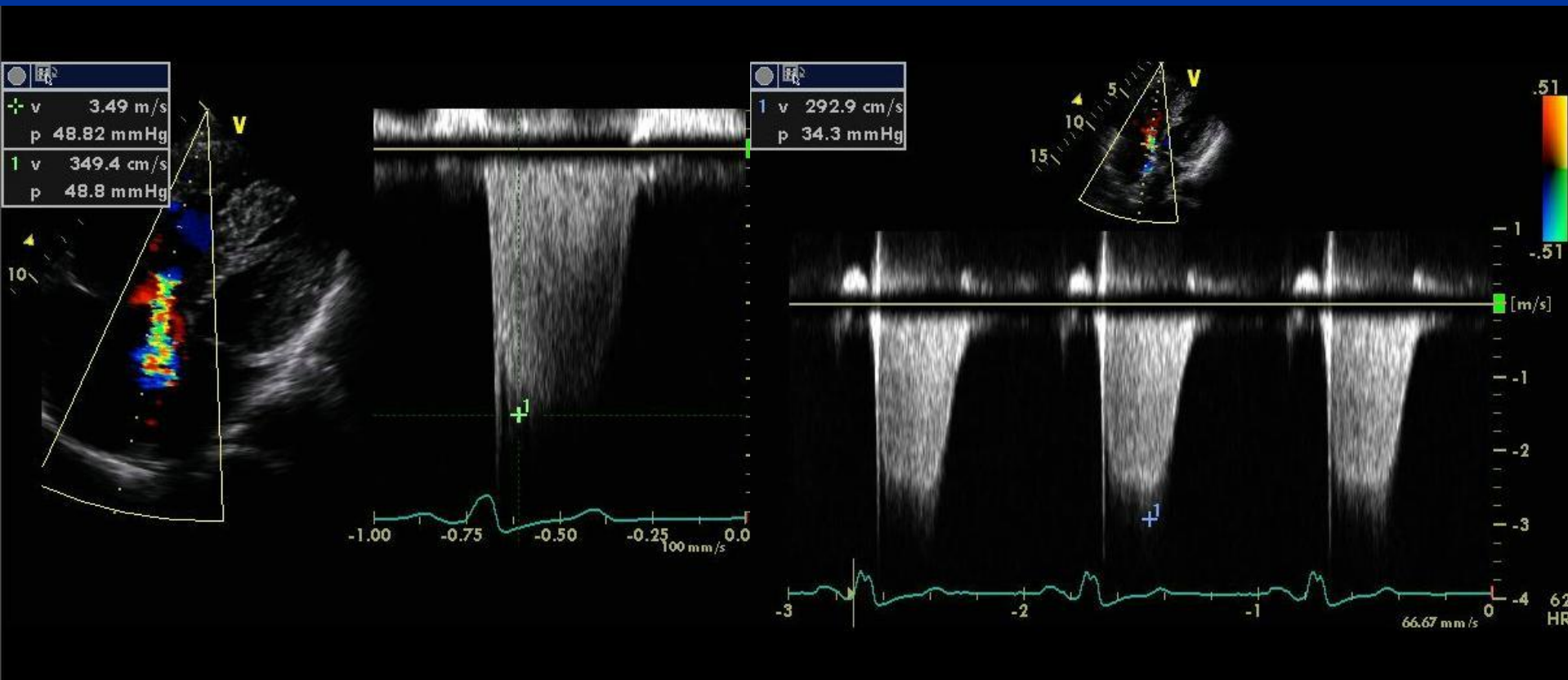




PRE



1 month after



PRE

1 month after



Before

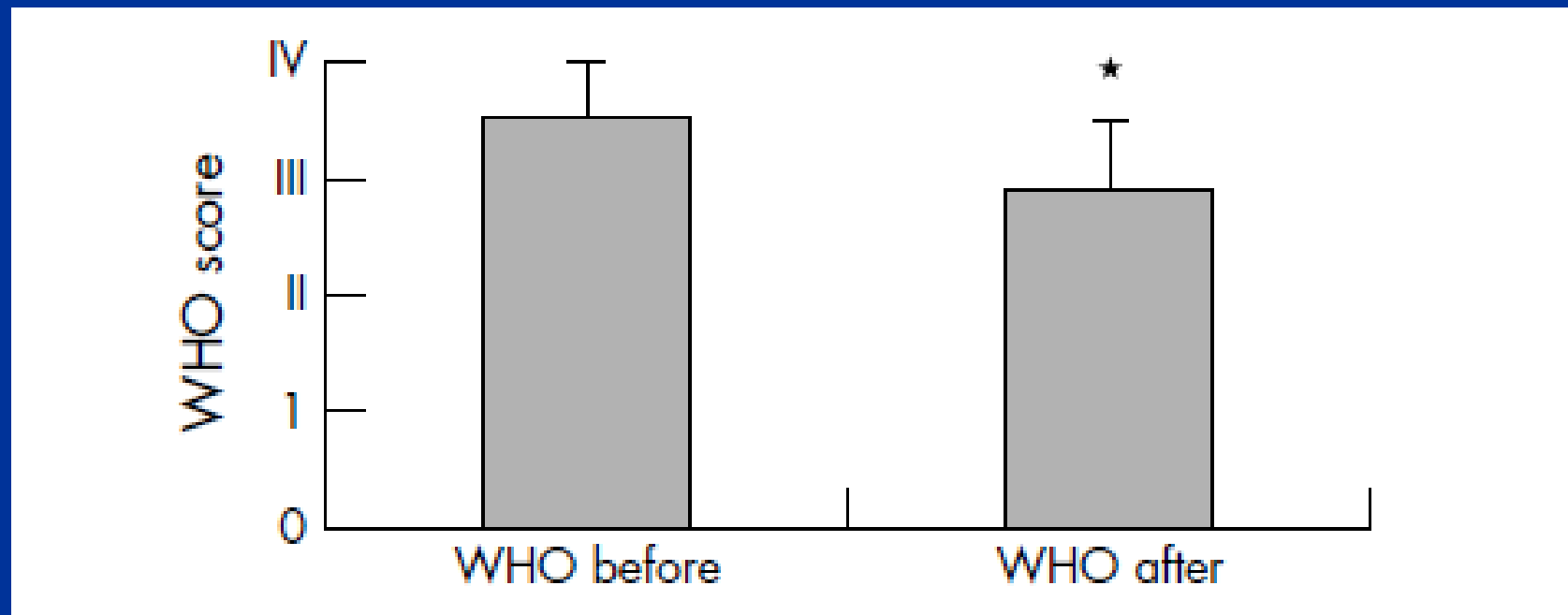
3 months after

Various Conditions of ACHD with PH

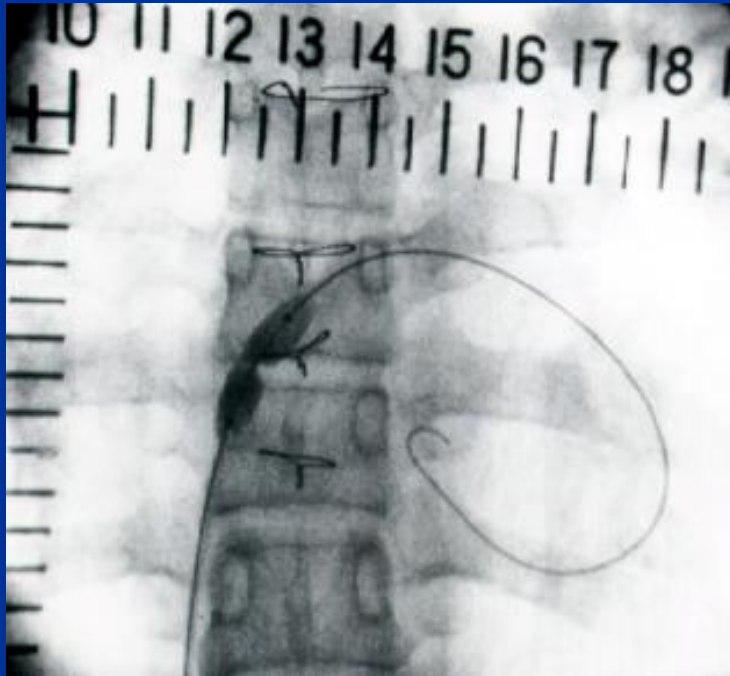
- Eisenmenger Syndrome
- Left to Right Shunt disease with PH
- Post operative PH (without shunt)

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

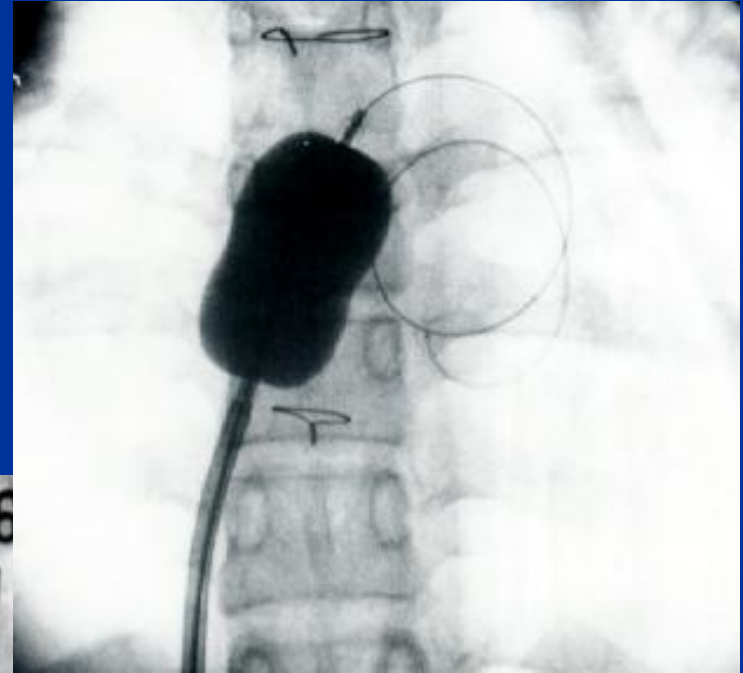
A Micheletti, A A Hislop, A Lammers, P Bonhoeffer, G Derrick, P Rees, S G Haworth



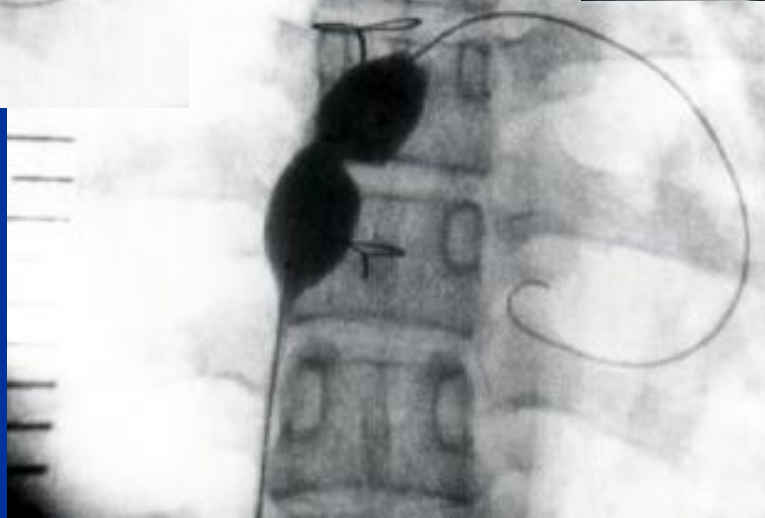
ASD creation



SaO₂: 92%



SaO₂: 84%

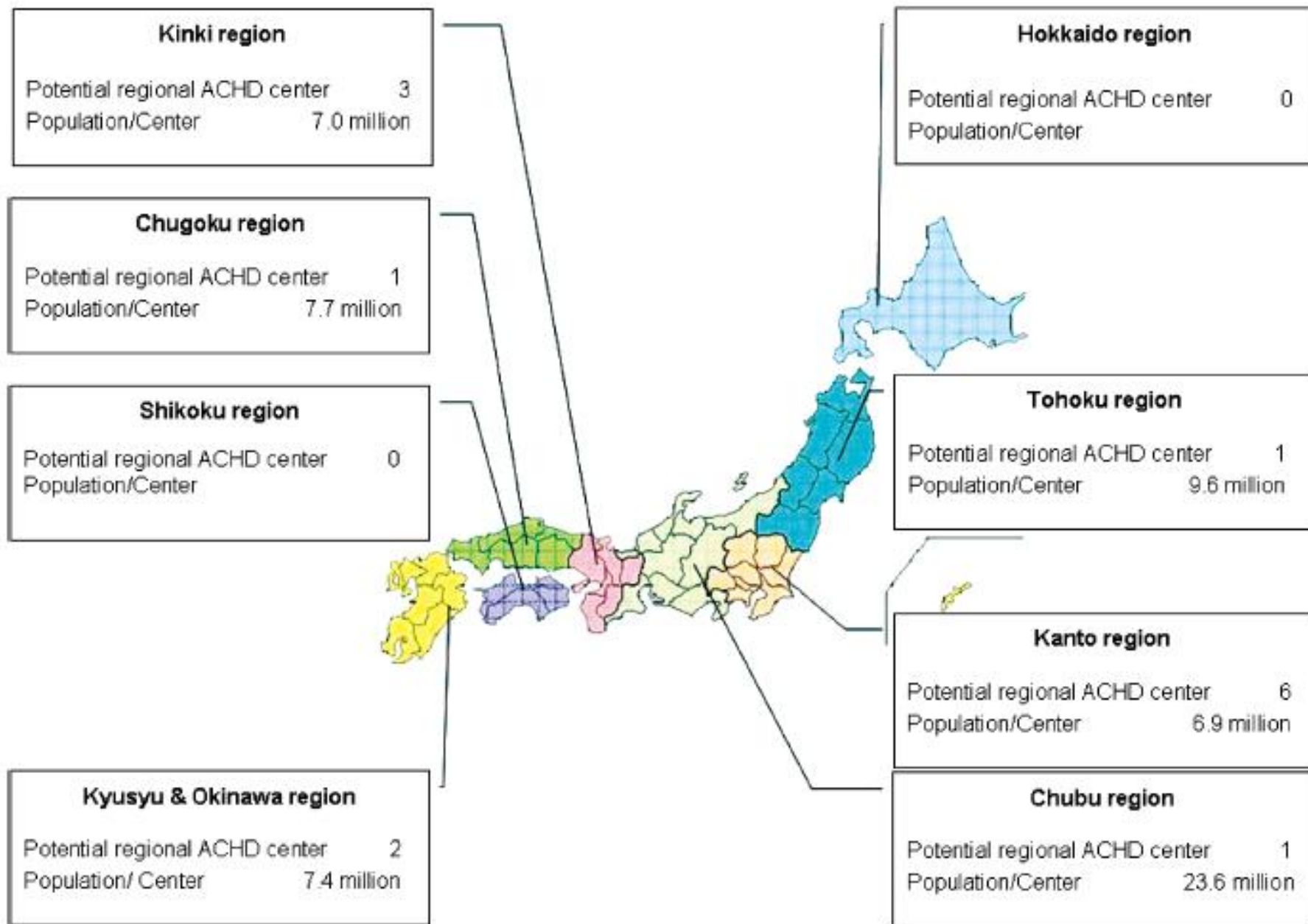


What should we inform the prognosis of ACHD with severe PH

- 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.



To establishment for optimal management system for ACHD patients

「成人先天性心疾患診療体制の構築に向けて」

招待講演

Prof. Philip J. Steer
(Imperial College London)

Prof. Joseph K. Perloff
(UCLA)

Prof. Ad Bogers
(Rotterdam University)

January 9-10, 2012
Tokyo





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)

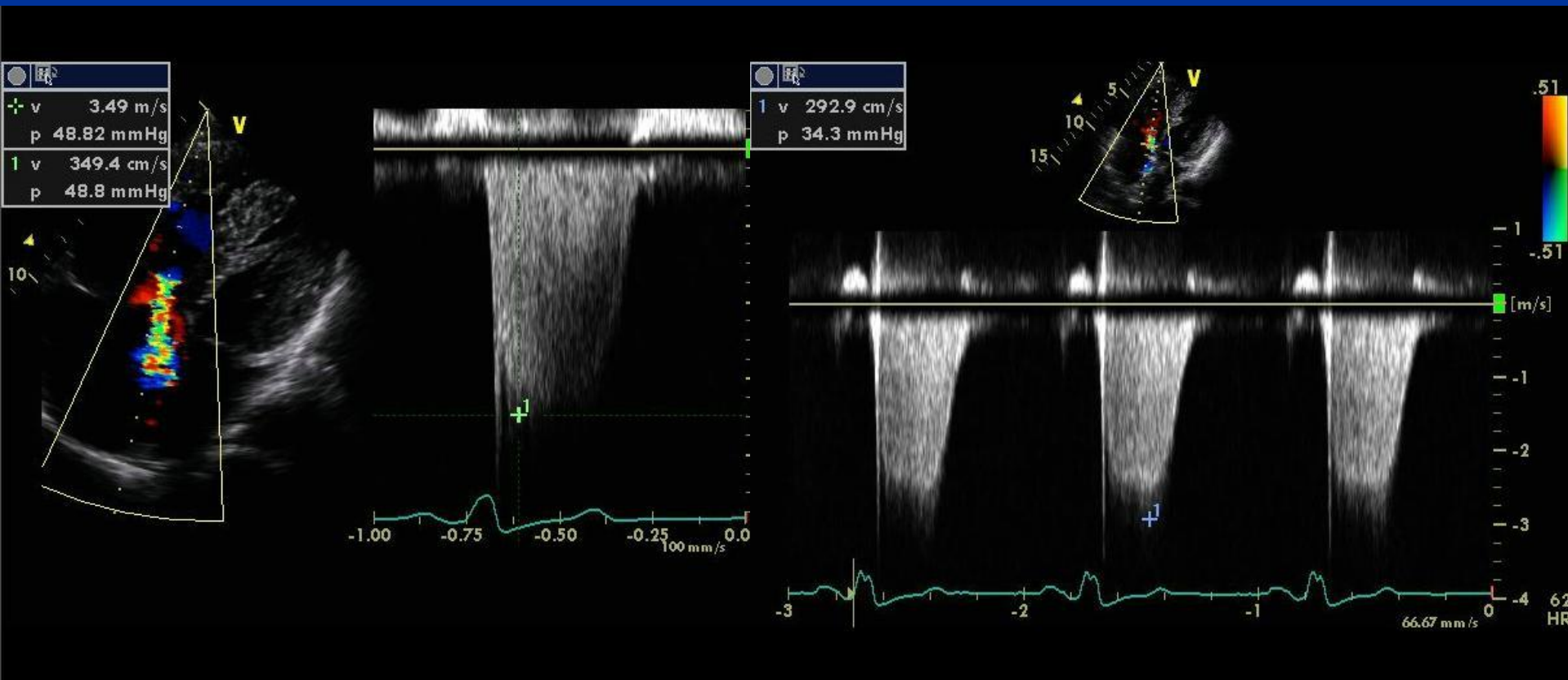
10歳時に運動中に失神.

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.



PRE

1 month after

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．

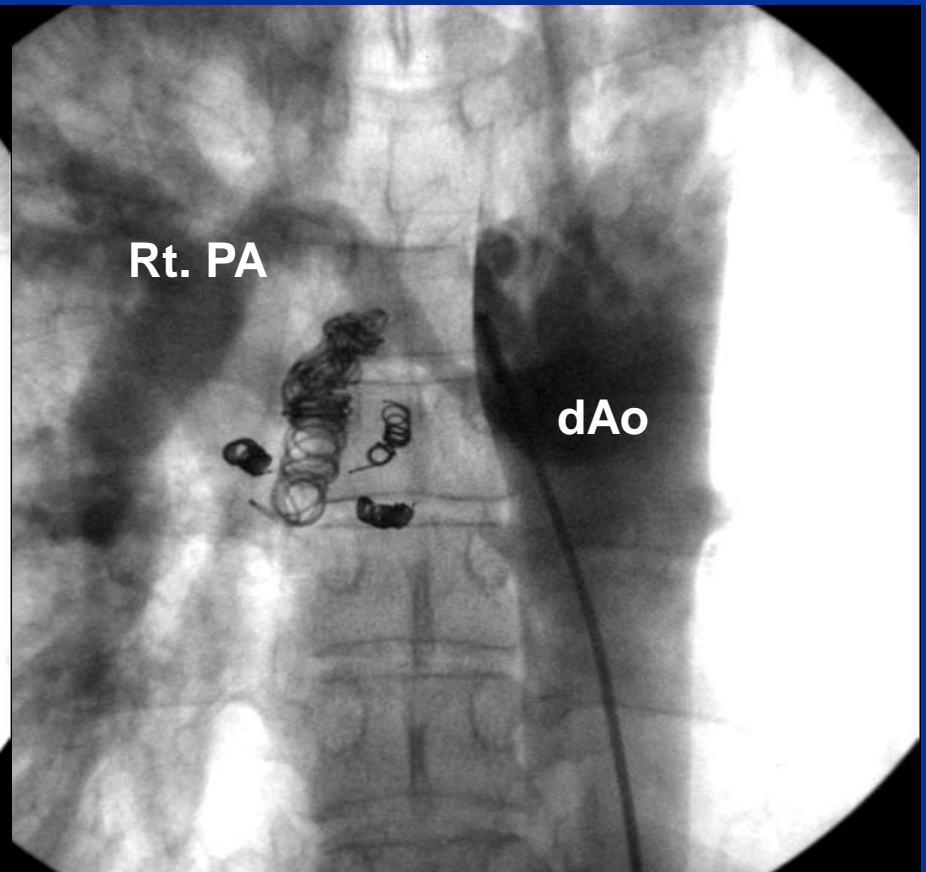
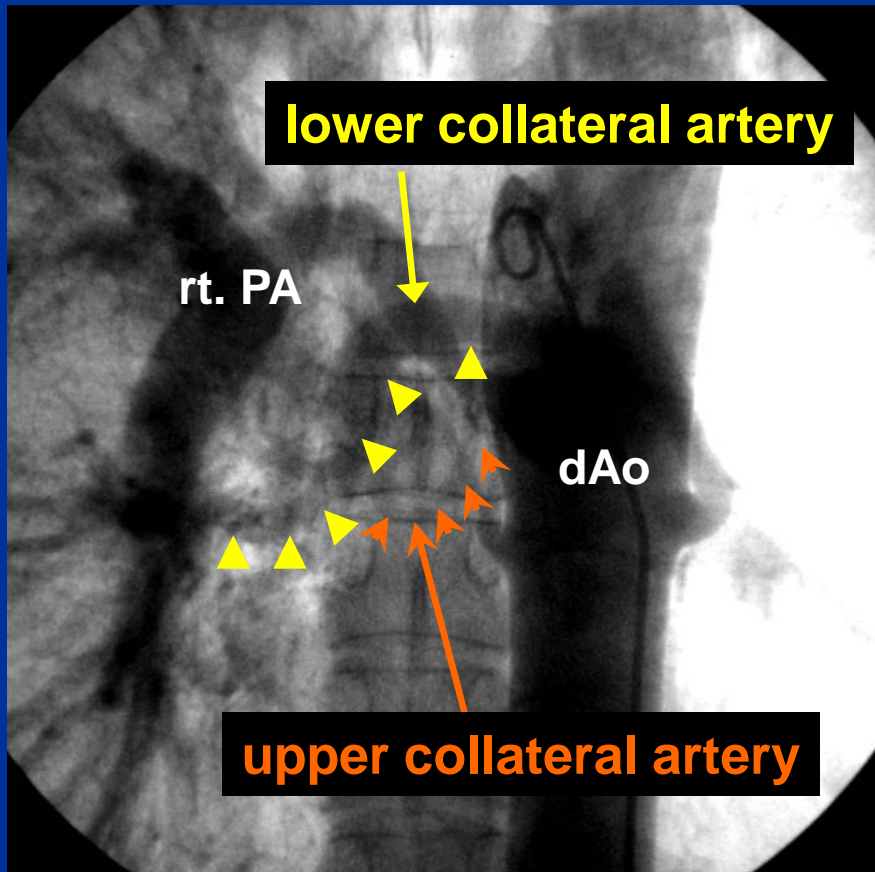
44歳時にAPCAに対する

経カテーテル的コイル閉鎖術．

Coil occlusion to aortopulmonary collateral arteries

before

after



before

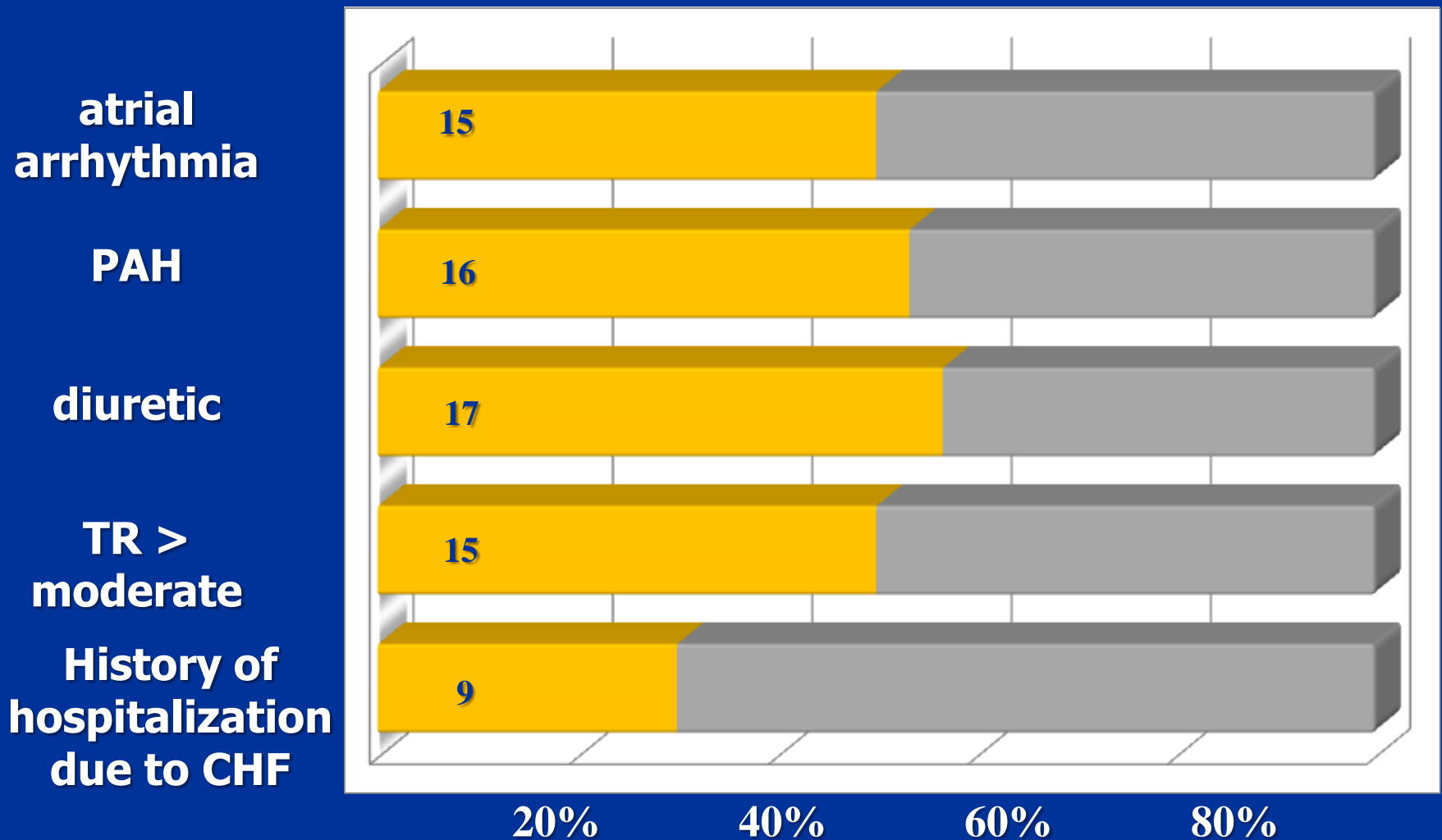
after



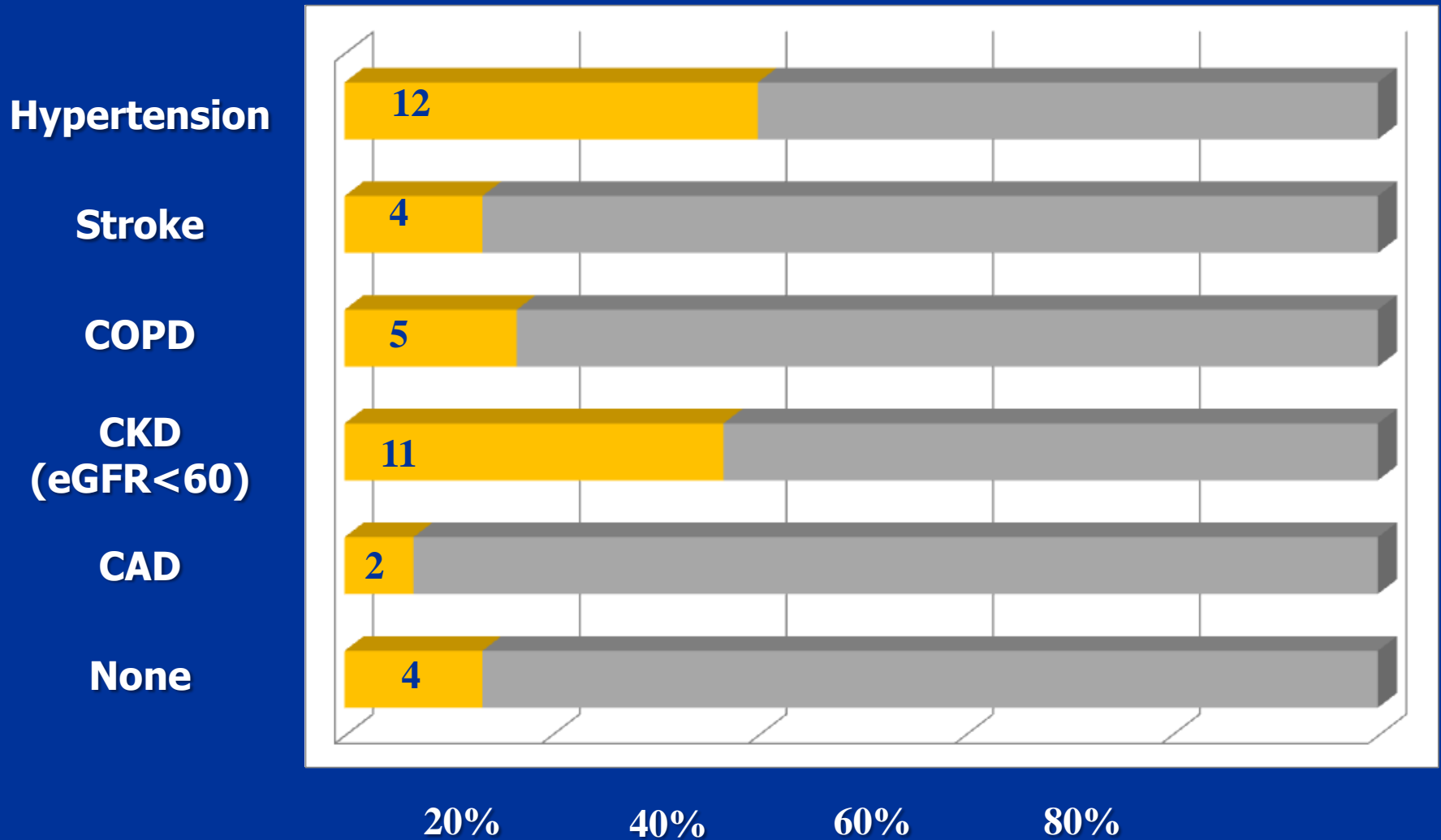
Japanese Society for Adult CHD

- Established in 1999
- Subgroup of Japanese Society for Pediatric Cardiology and Cardiac Surgery
- Active members: 200
 - 80% Pediatric Cardiologist
 - 10% Cardiac Surgeon
 - 5% Adult Cardiologist
 - 5% Obstetrician and Gynecologist

Comorbidities & Medication



Comorbidities



Catheter intervention

■ 症例1

VSD, PDA, Eisenmenger症候群



transcatheter ASD creation

■ 症例2

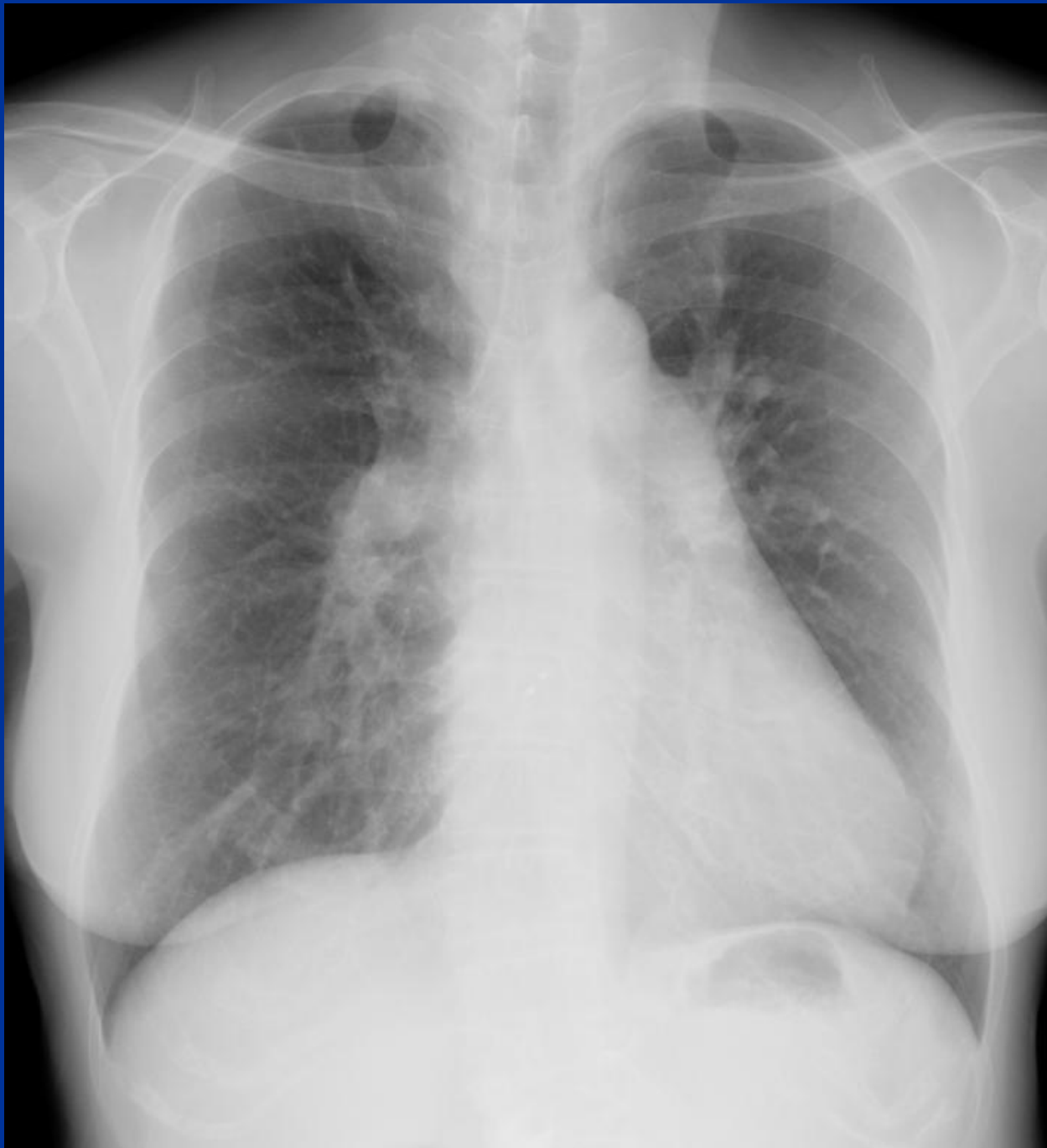
PA with VSD, APCA

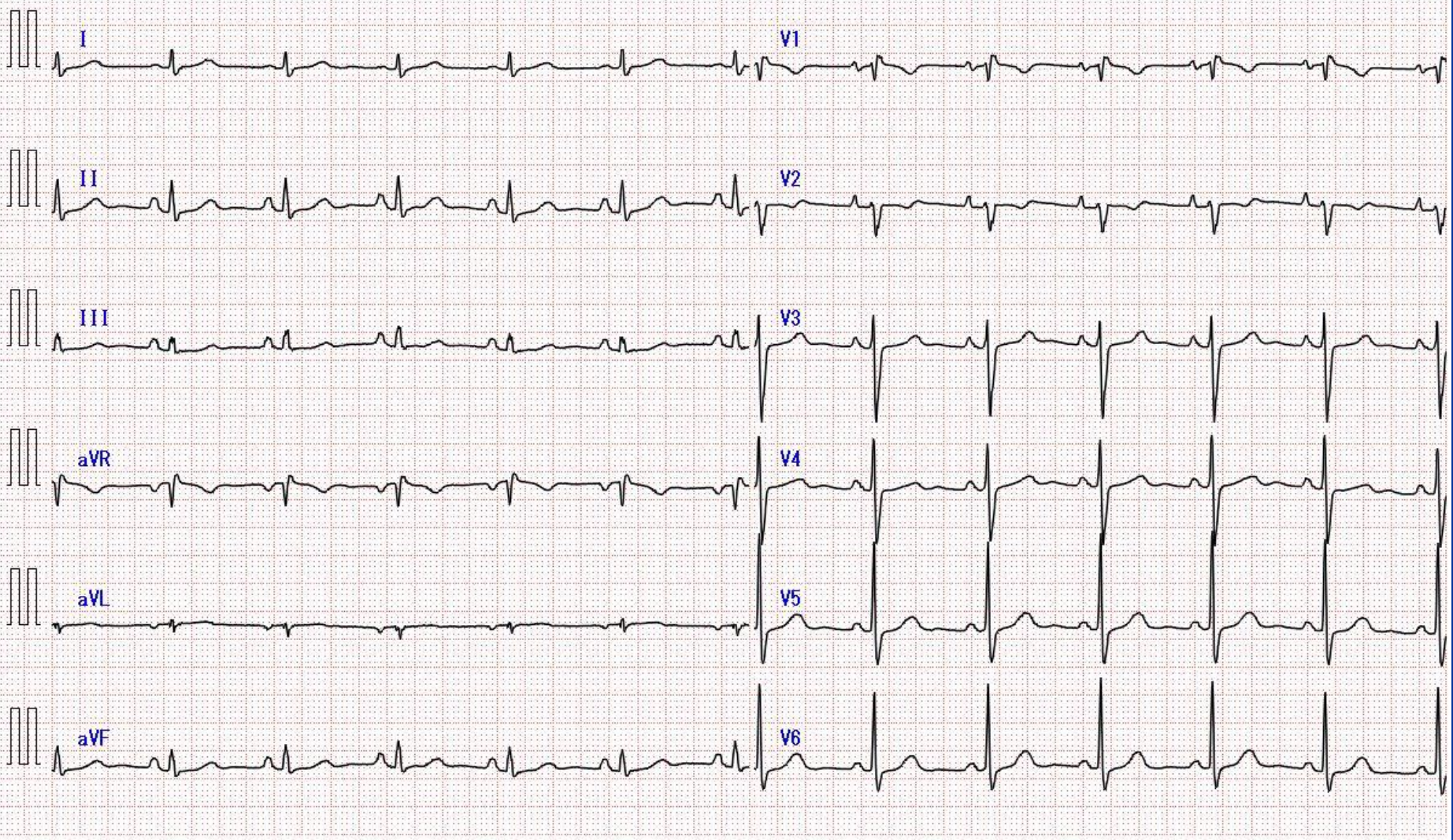


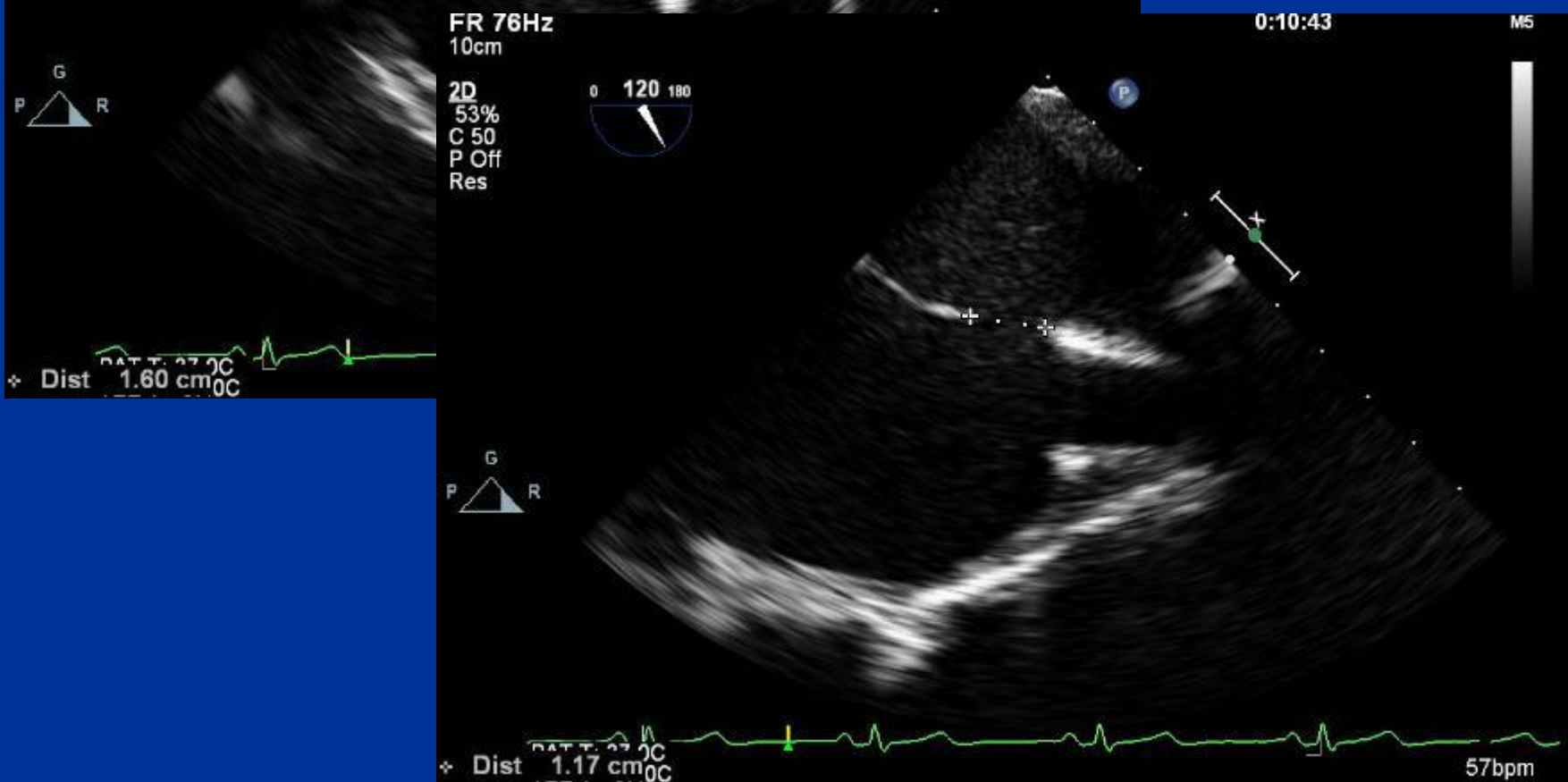
transcatheter APCA occlusion

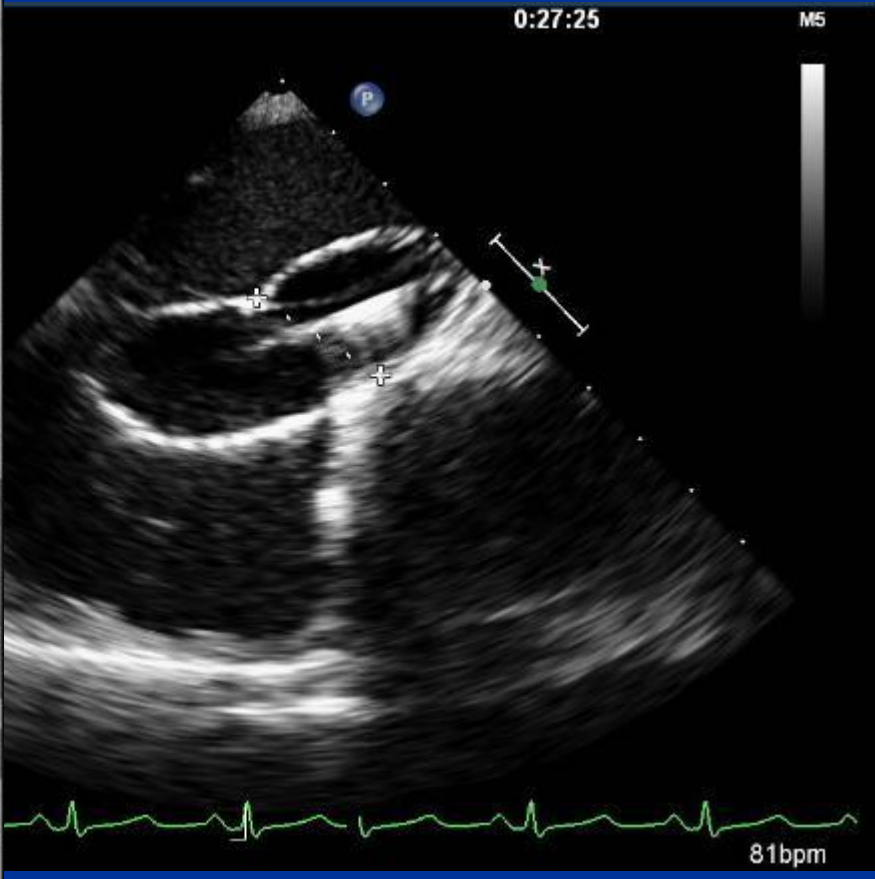
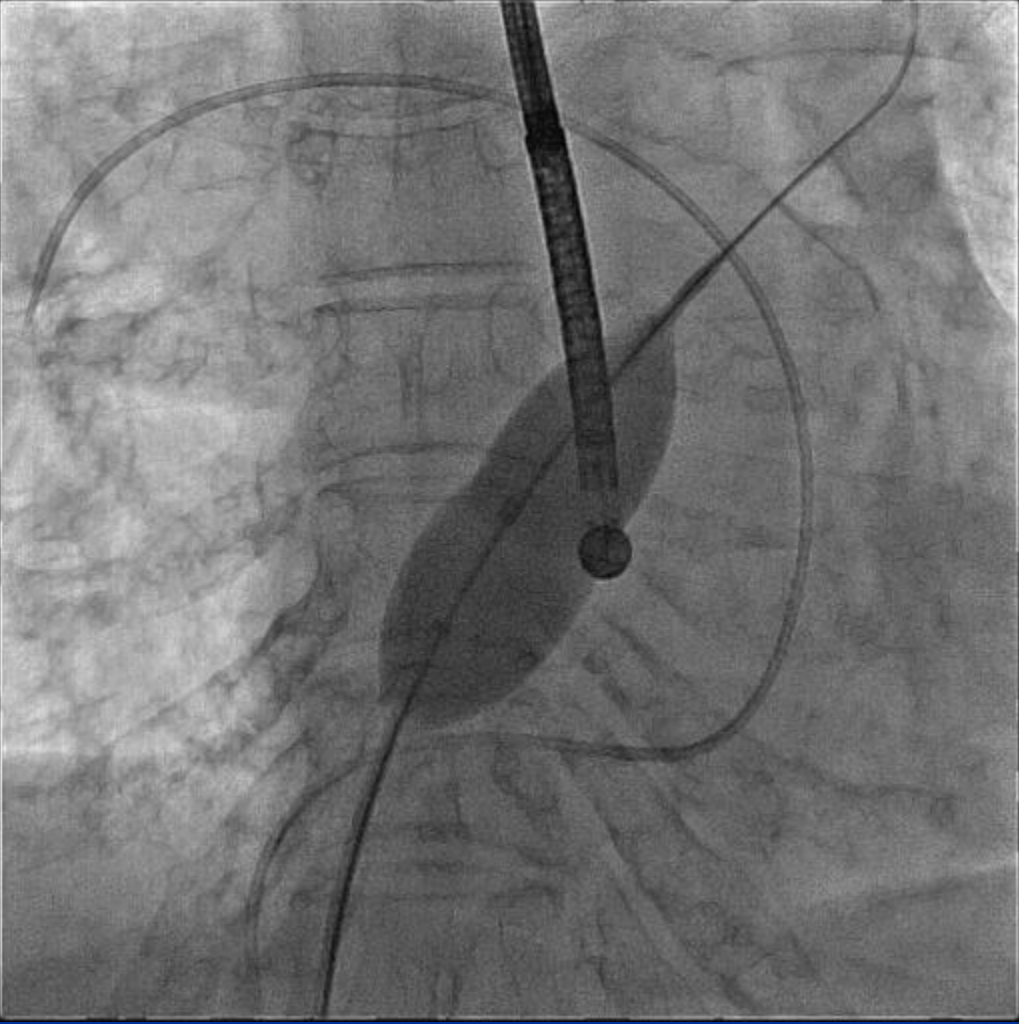
70 years female

- 10年ほど前の健康診断で胸部レントゲン異常を指摘され、心房中隔欠損症と診断された。手術を勧められたが、躊躇したまま経過をみられていた。
- 現在の処方
 - amlodipine 5mg/day
 - valsartan 80mg/day









2011/07/15 (Fri) IHR 53 IIBP1 66 IIBP2 - ISpO2 - INIBP - INIBP |
09:53:43 III IPV -18 ILV 0 - | | | | | | | | | |
0004110269 | | | ? | - | | -:- | | |



Balloon occlusion

PV
PS: 25 mm/s

Rest

2011/07/15 (Fri) IHR 56 IIBP1 26 IIBP2 - ISpO2 - INIBP - INIBP |
09:54:19 III IPV 13 ILV o - - - - - |
0004110269 | | 18 | - | | -:- | |



20



10

PV 0

PV
PS: 25 mm/s

Rest

NIHON KOHDEN

NIHON KOHDEN

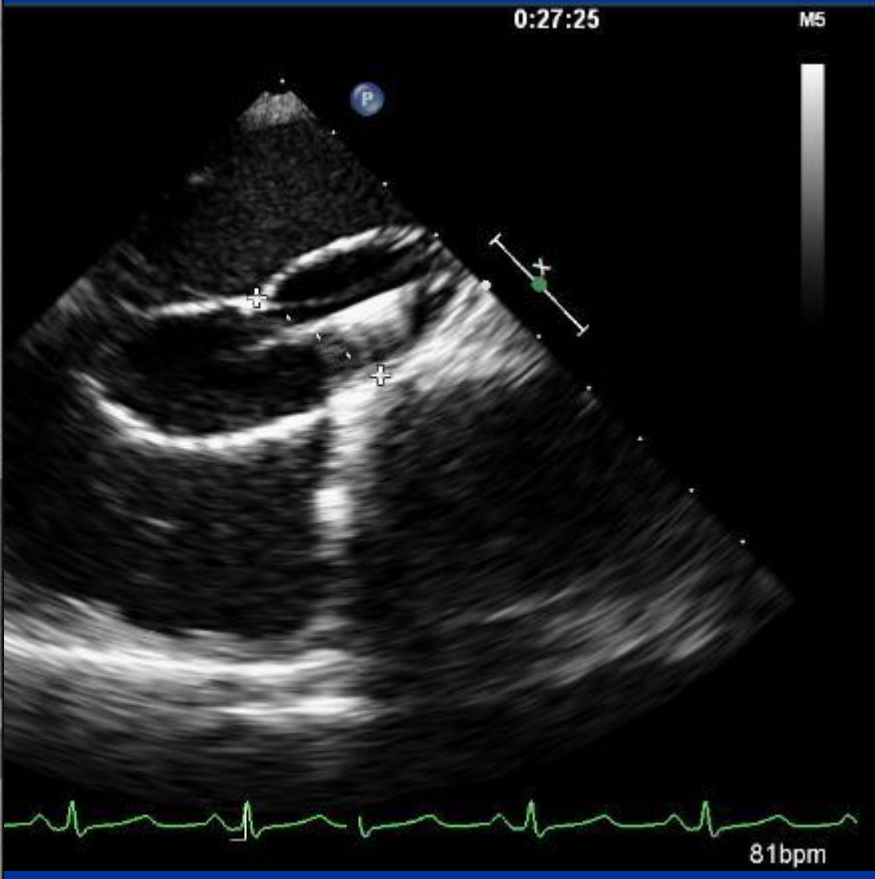
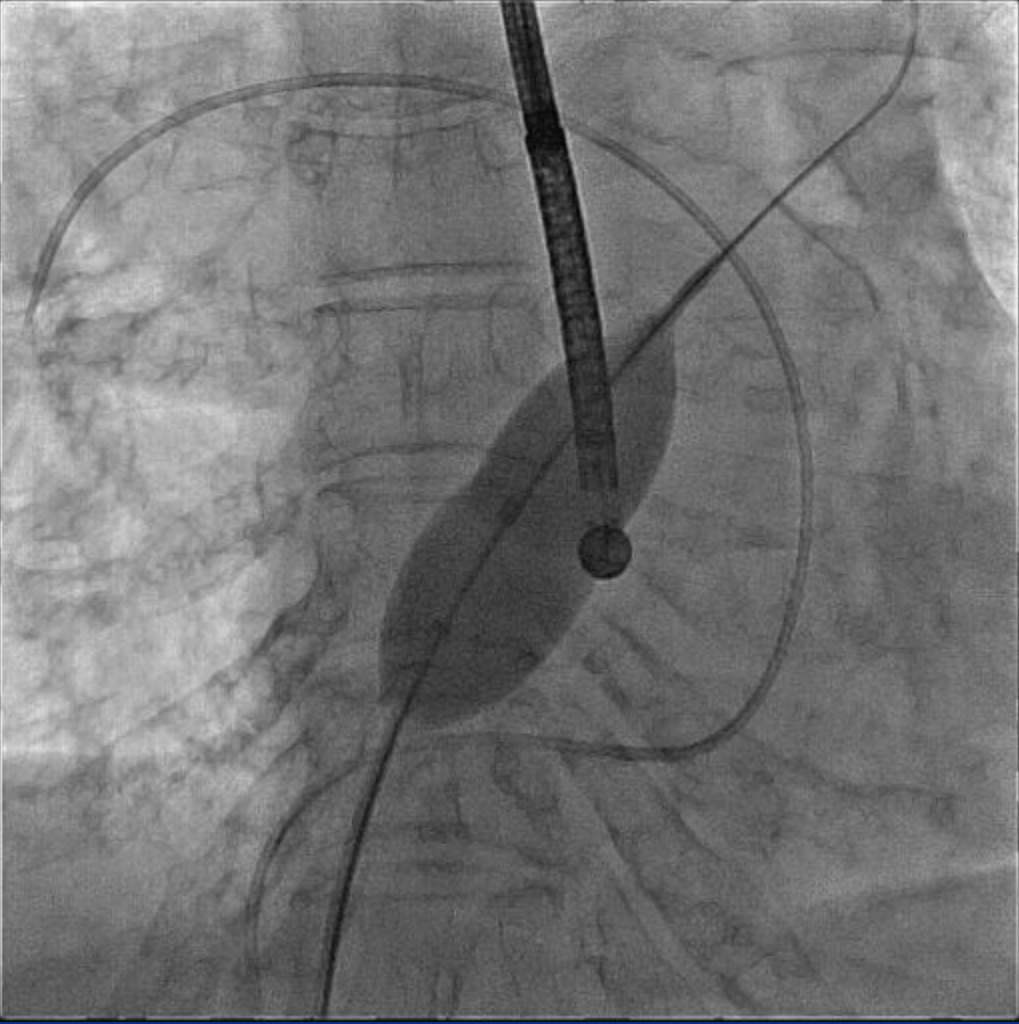
J01301

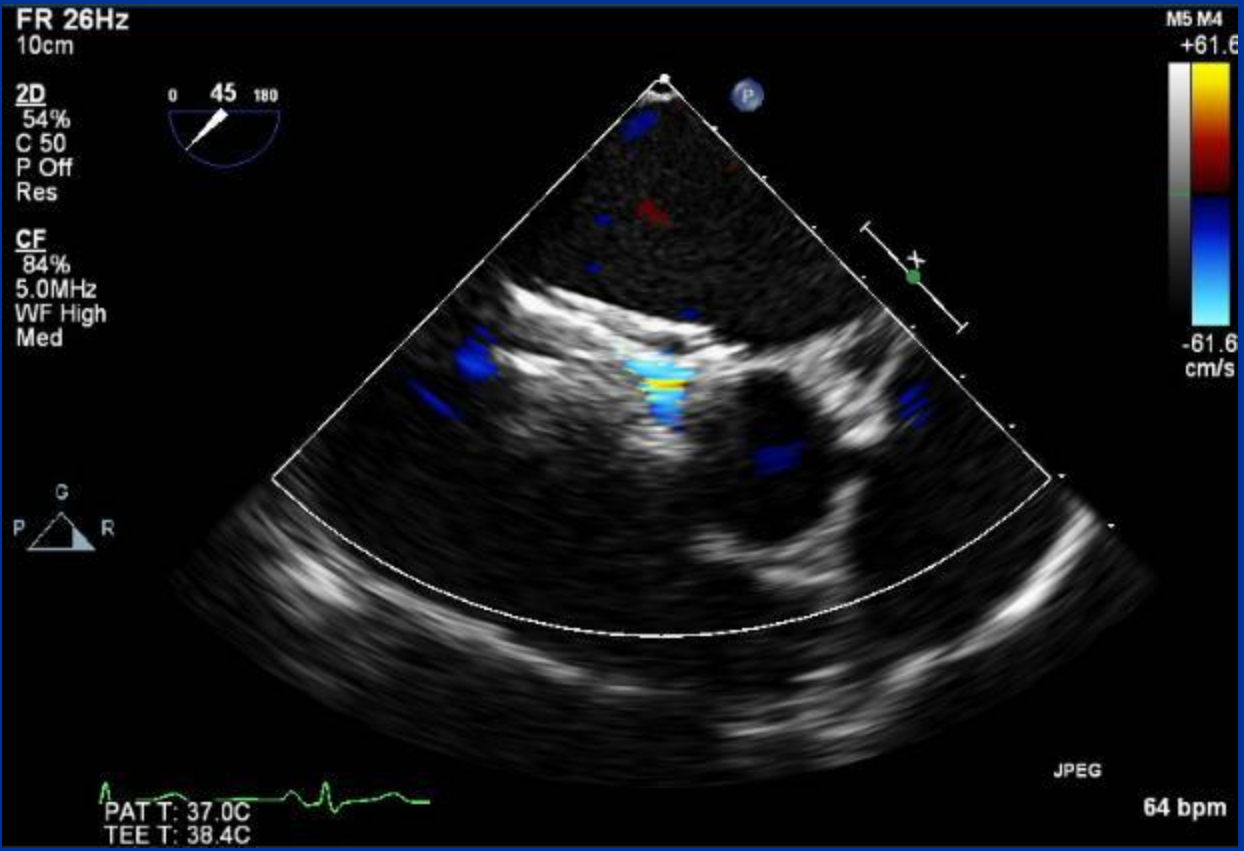
F6W210-108

Masked Left Ventricular Restriction in Elderly Patients With Atrial Septal Defects: A Contraindication for Closure?

Peter Ewert,* MD, Felix Berger, MD, Nicole Nagdyman, MD, Oliver Kretschmar, MD, Sven Dittrich, MD, Hashim Abdul-Khaliq, MD, and Peter E. Lange, PhD

	Nonresponders (n = 11)	Responders (n = 7)	P
Age (median, years)	70	70	NS
Gender (f:m)	6:1	9:2	NS
Atrial flutter/fibrillation	4	4	NS
Systemic hypertension	5	3	NS
Coronary heart disease	0	1	NS
Defect diameter (mm)	24	25	NS
Shunt (Qp/Qs)	1.6	1.8	NS
Mean arterial pressure (mm Hg) before/during occlusion	94/95	94/93	NS
LA pressure (mean, mm Hg) before/during occlusion ^a			
a-wave	7/7	18/26	0.02
v-wave	6/7	24/41	< 0.001
Mean	3/4	14/23	< 0.001





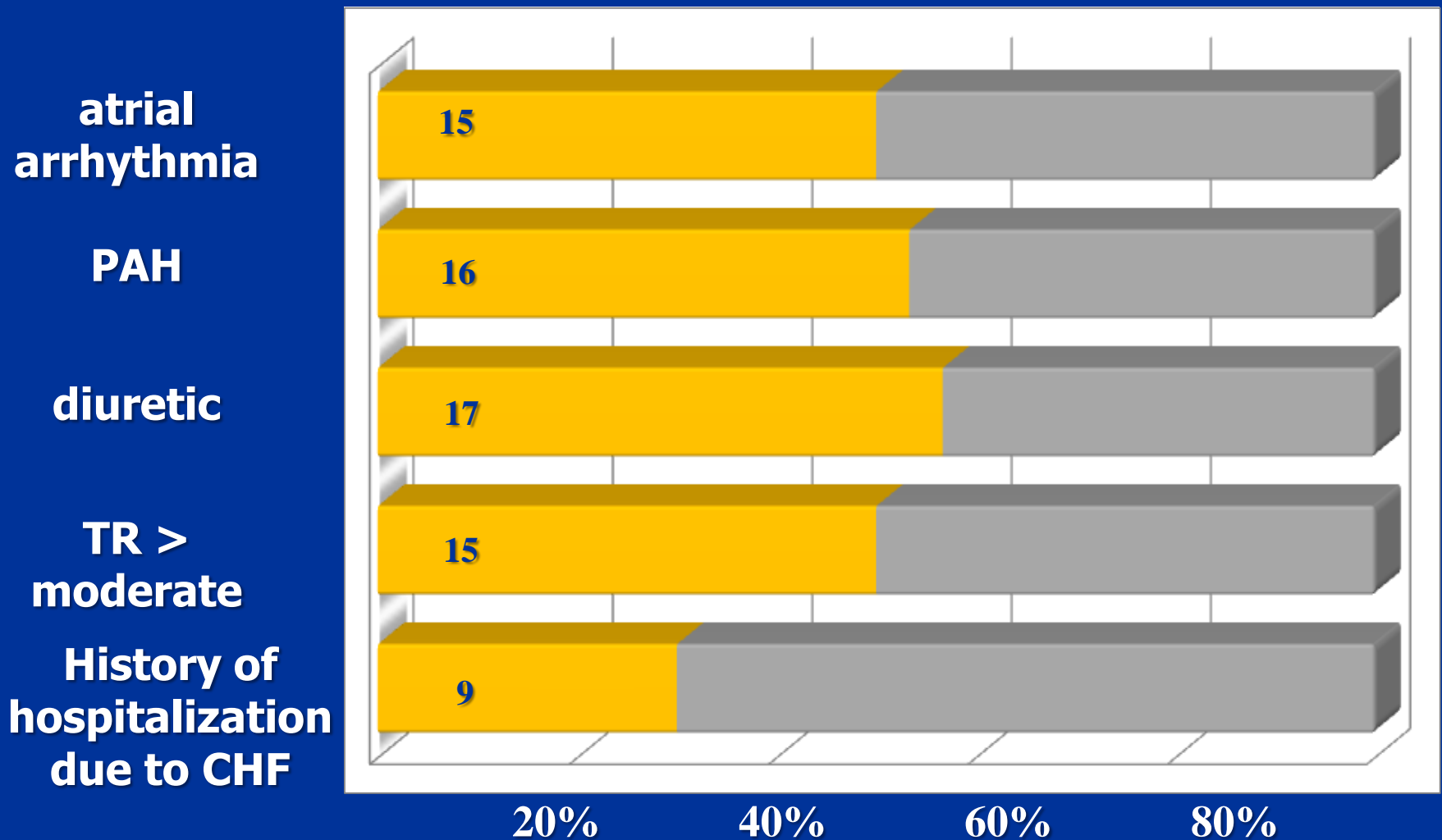


Before

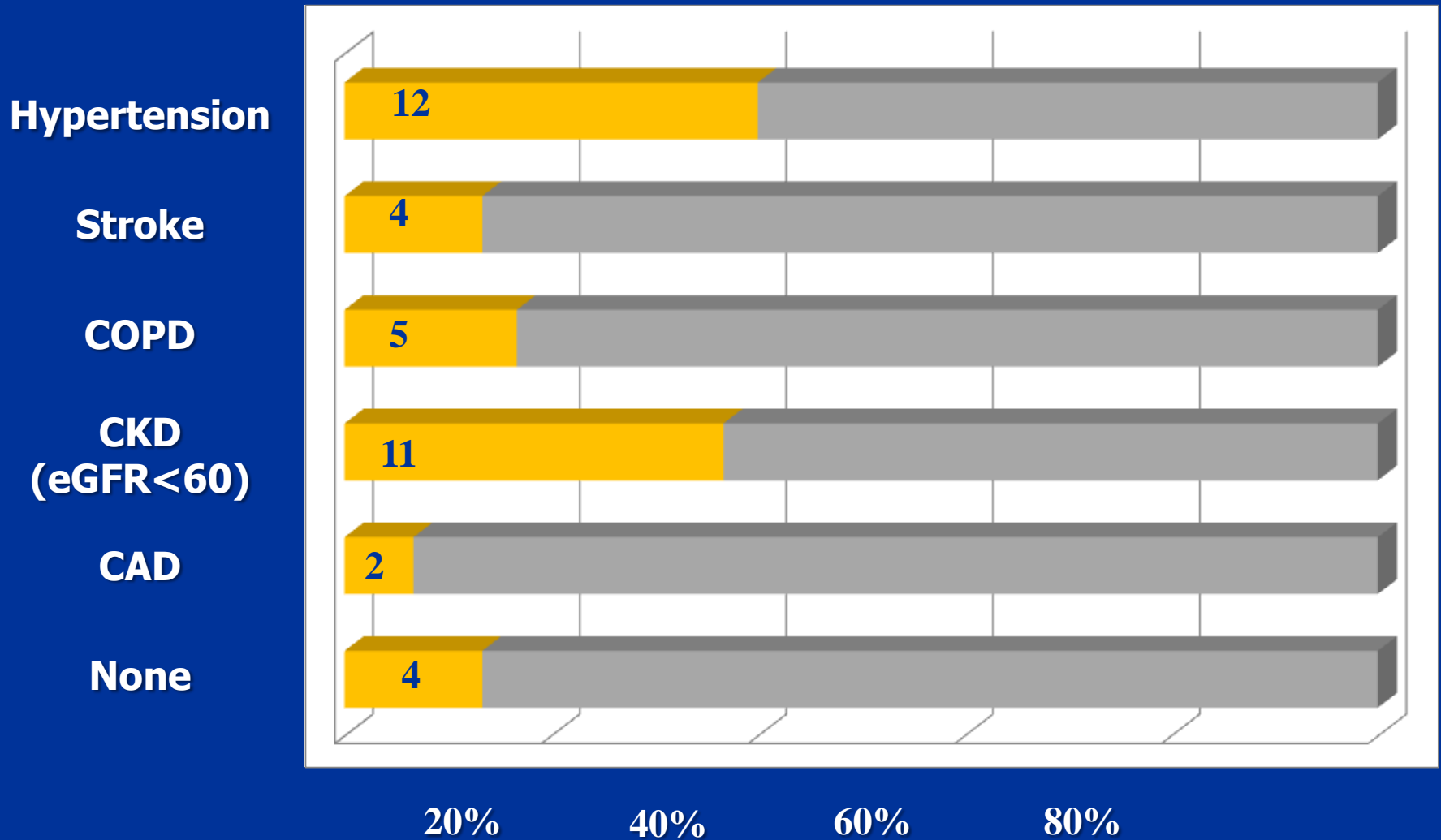


24 hours after

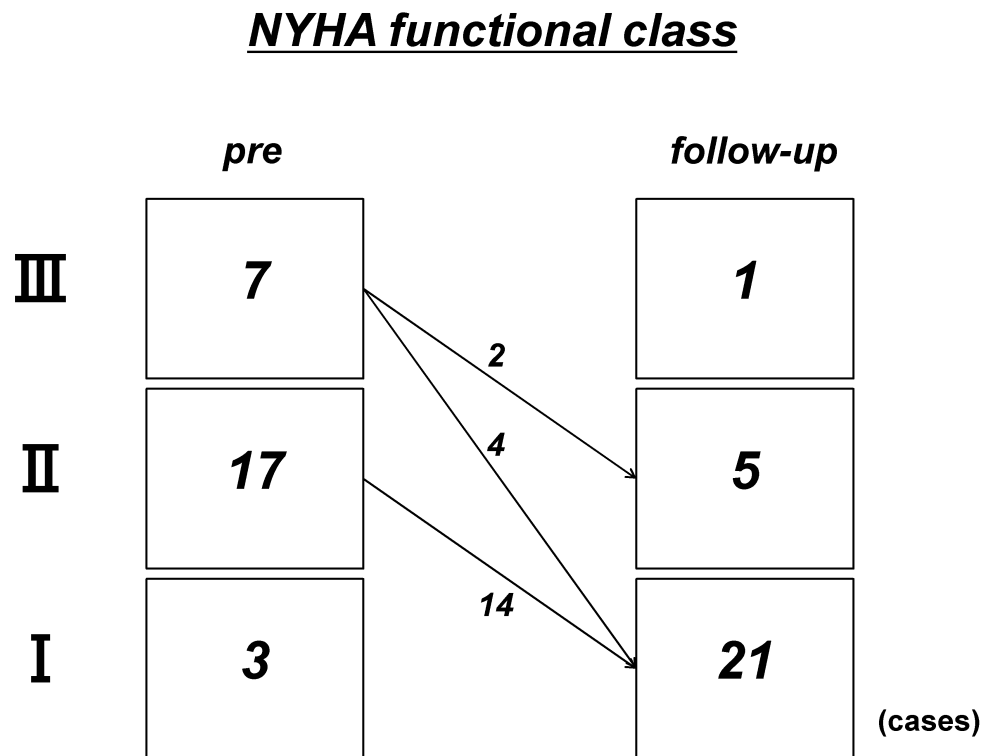
Comorbidities & Medication



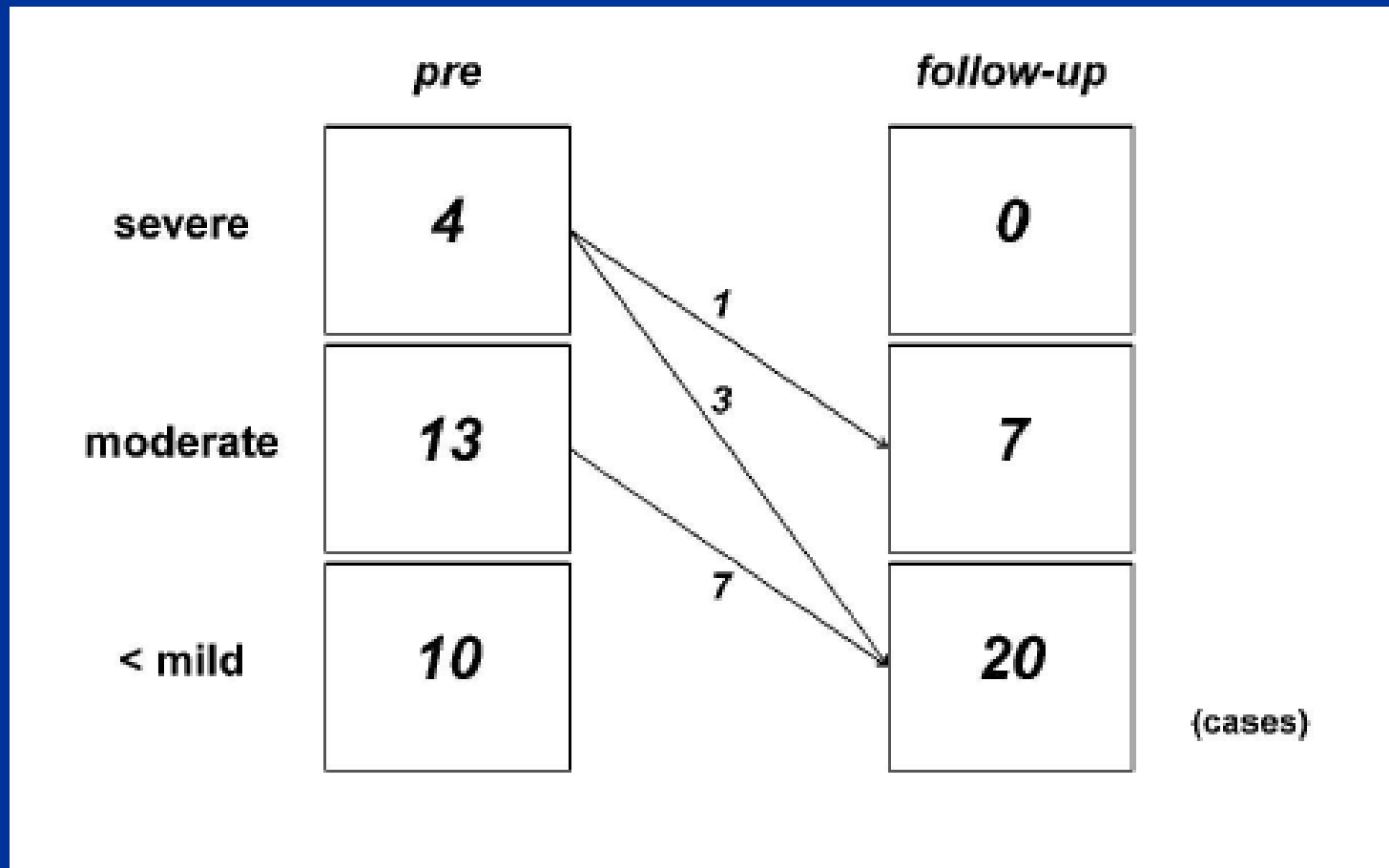
Comorbidities



Improvement of NYHA Class in Patients >70 years



Tricuspid Regurgitation after ASD closure >70 years old



Mitral Regurgitation after ASD closure >70 years old

