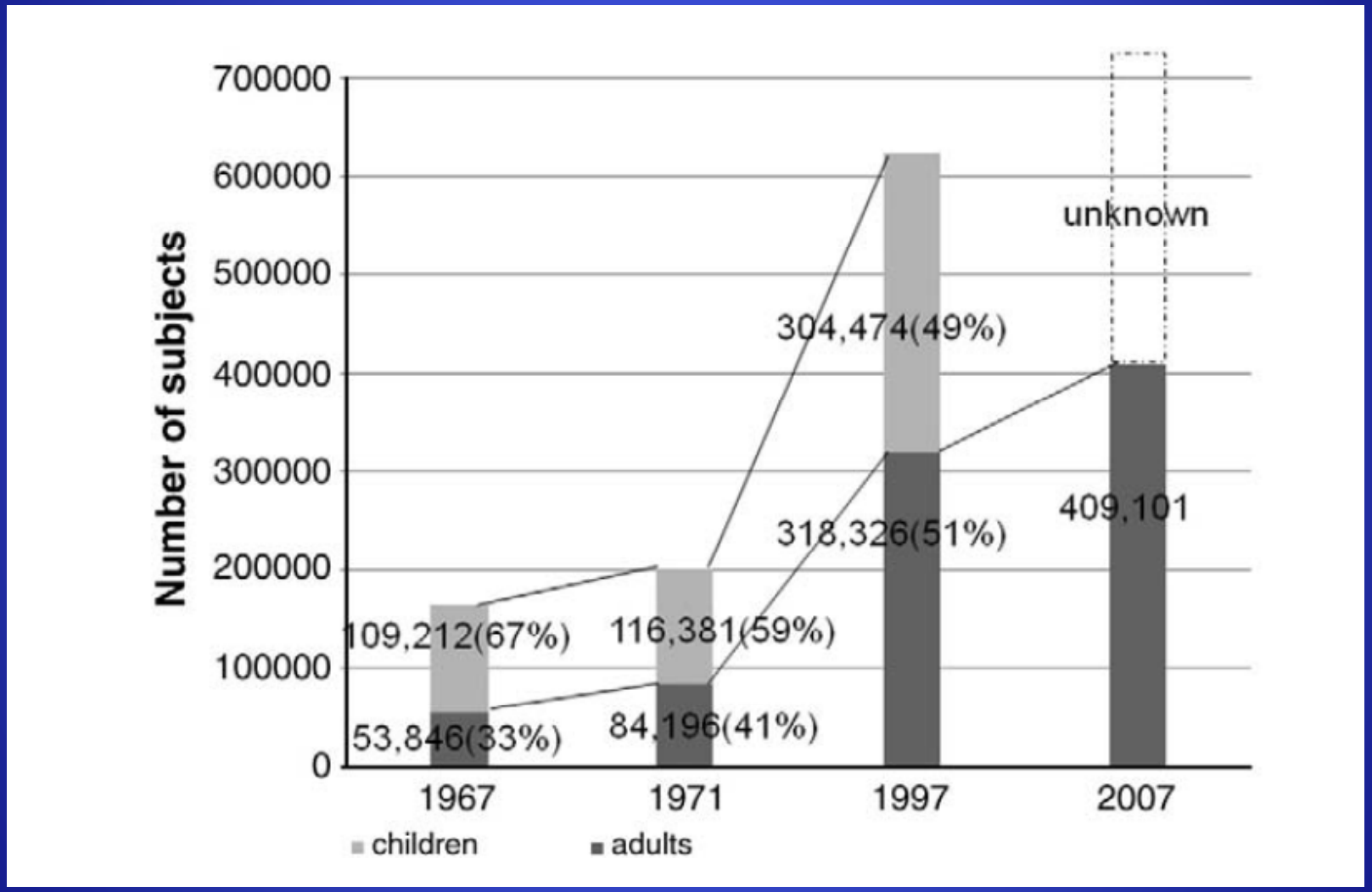
How to Collaborate with Pediatric and Adult Cardiologists for Management of Adult Congenital Heart Disease

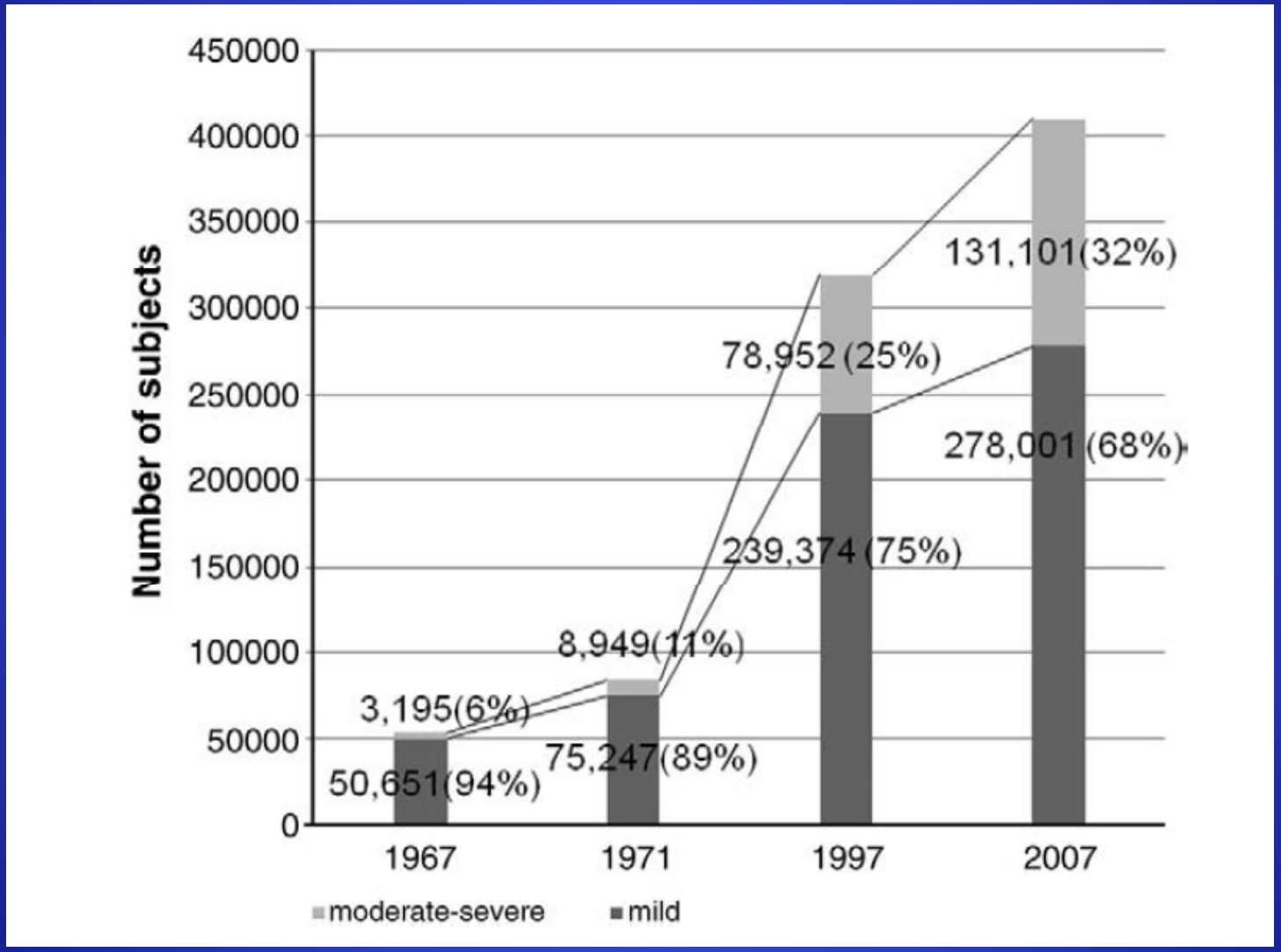
Teiji Akagi, MD, PhD, FACC FSCAI.
Okayama University Hospital,
Okayama, Japan

Prevalence of adult patients with CHD in Japan



Shiina Y, et al. Inter J Cardiol 2009

Prevalence of adult patients with CHD in Japan



Shiina Y, et al. Inter J Cardiol 2009

Who care the adult patients with CHD

- Pediatric cardiologists
- Adult cardiologists

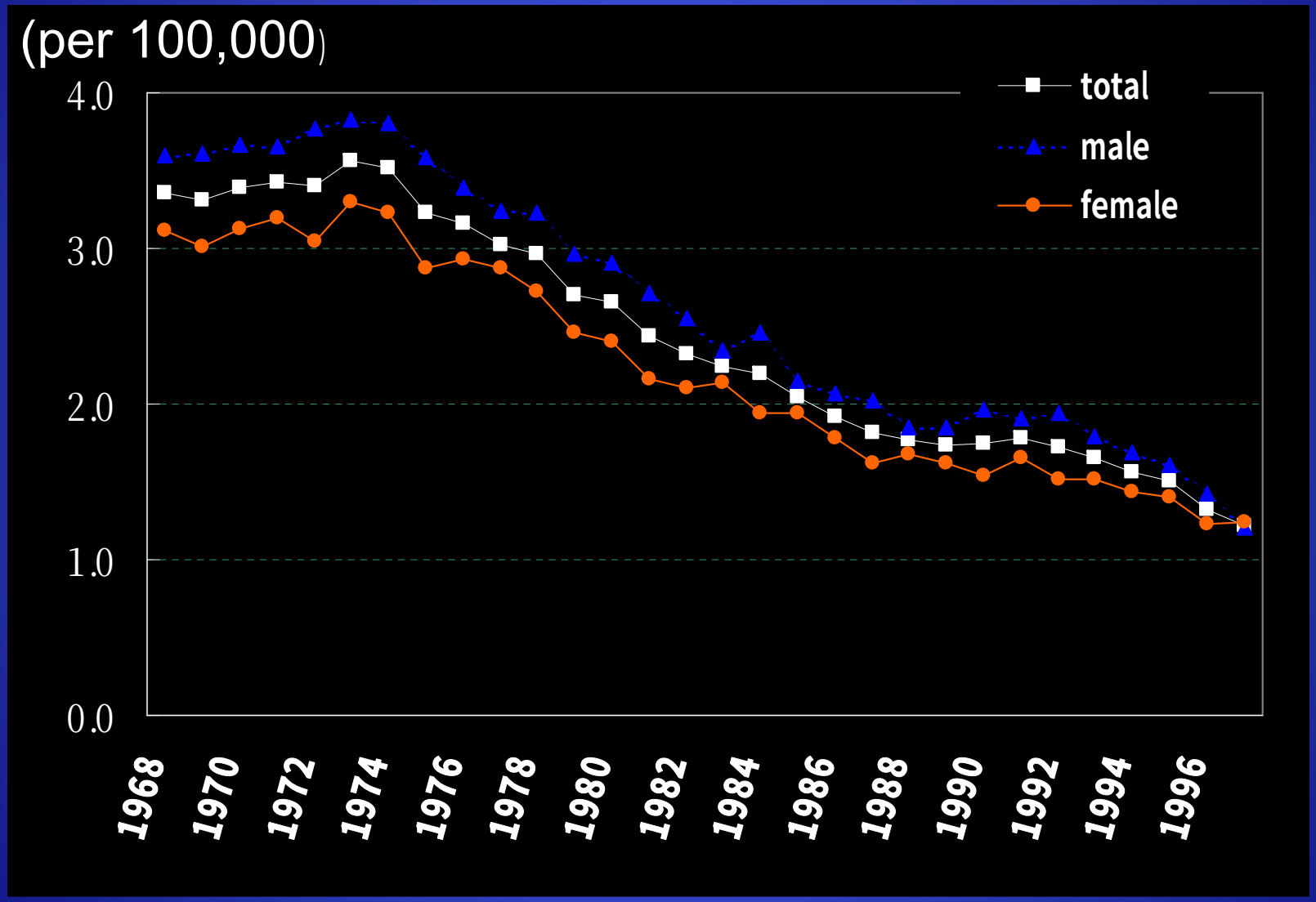
  many subspecialties
- Cardiac surgeons

  many subspecialties

How about the situation of adult cardiology

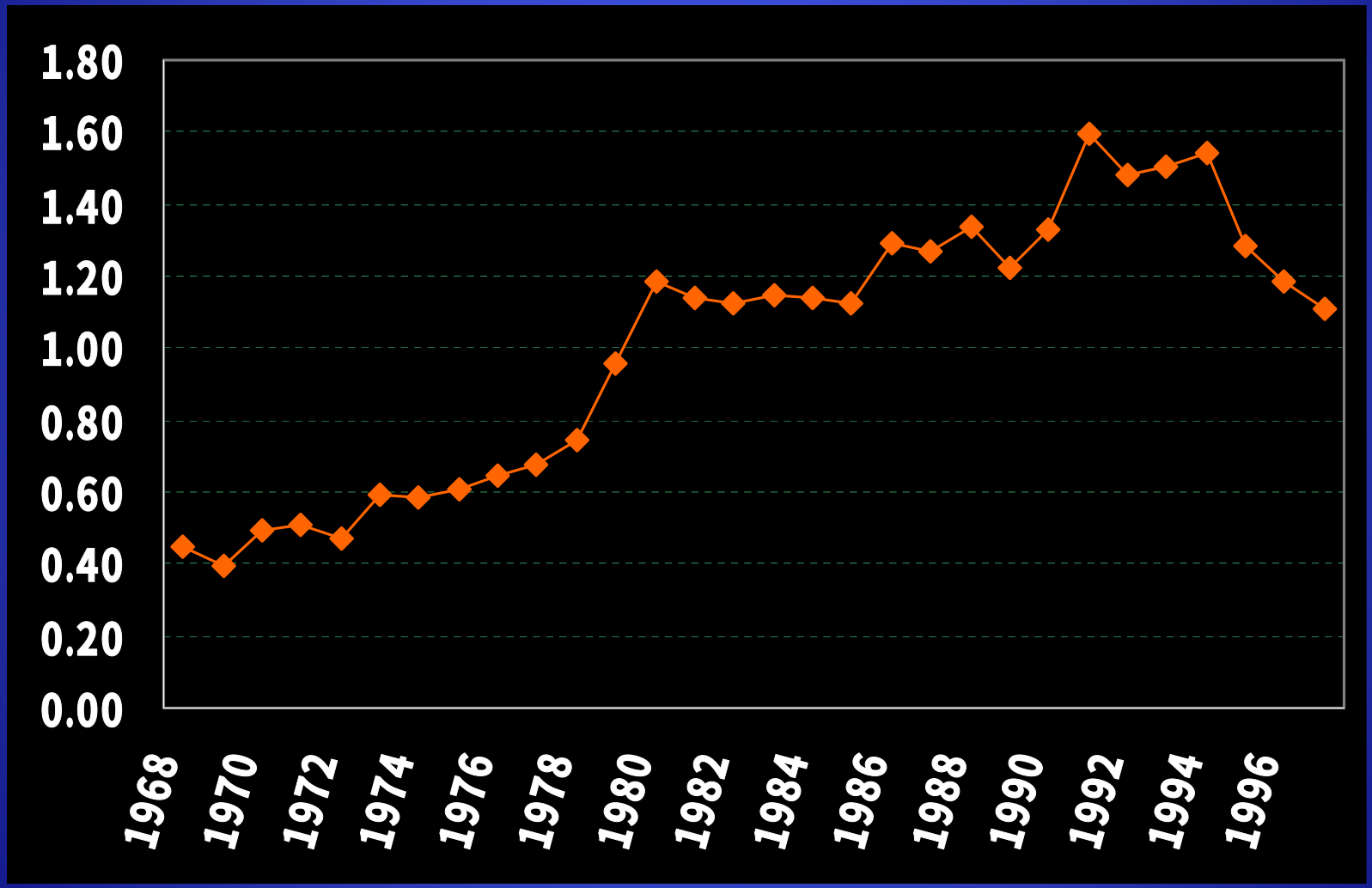
- already have the patients with non repaired CHD.
- already encountered the adult CHD patients.
- Most management done by individual base, number of ACDH specialists is very limited.

Mortality of Congenital Heart Disease in Japan

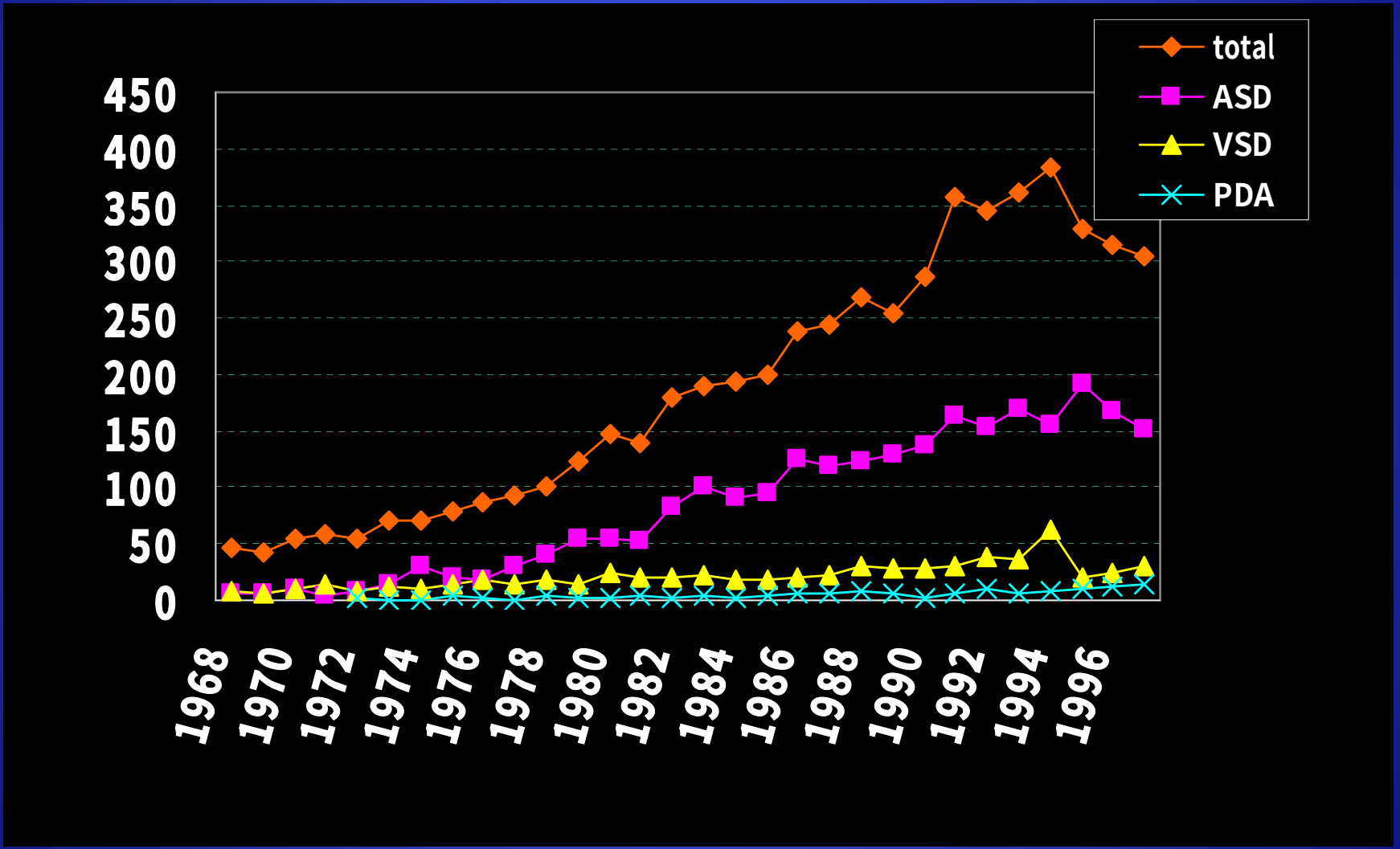


Terai M, Nakazawa M. Circulation J 2001

Mortality of Congenital Heart Disease aged > 60 years

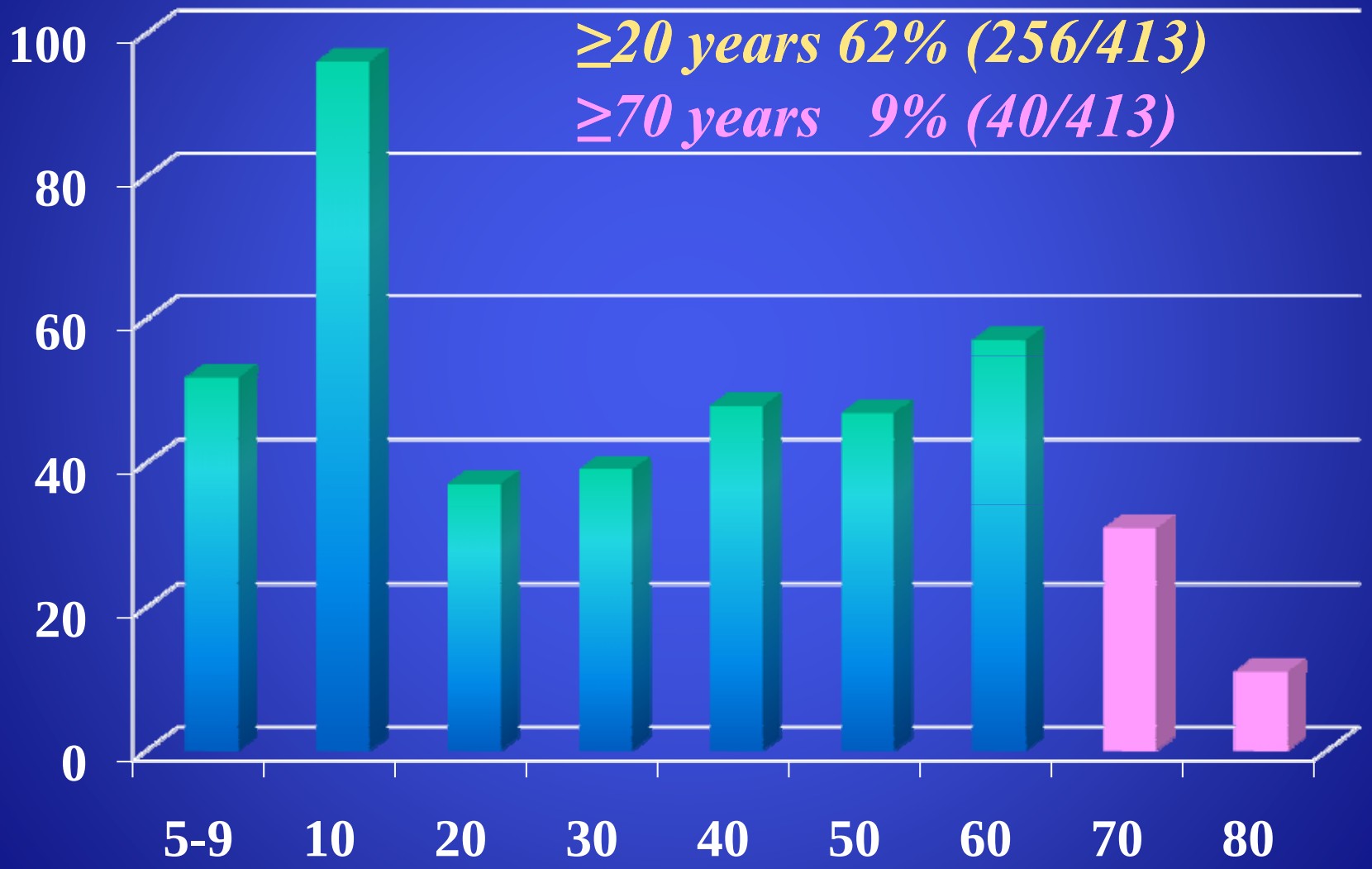


Mortality of Congenital Heart Disease aged > 60 years



Terai M, Nakazawa M. Circulation J 2001

Age distribution (n=413)



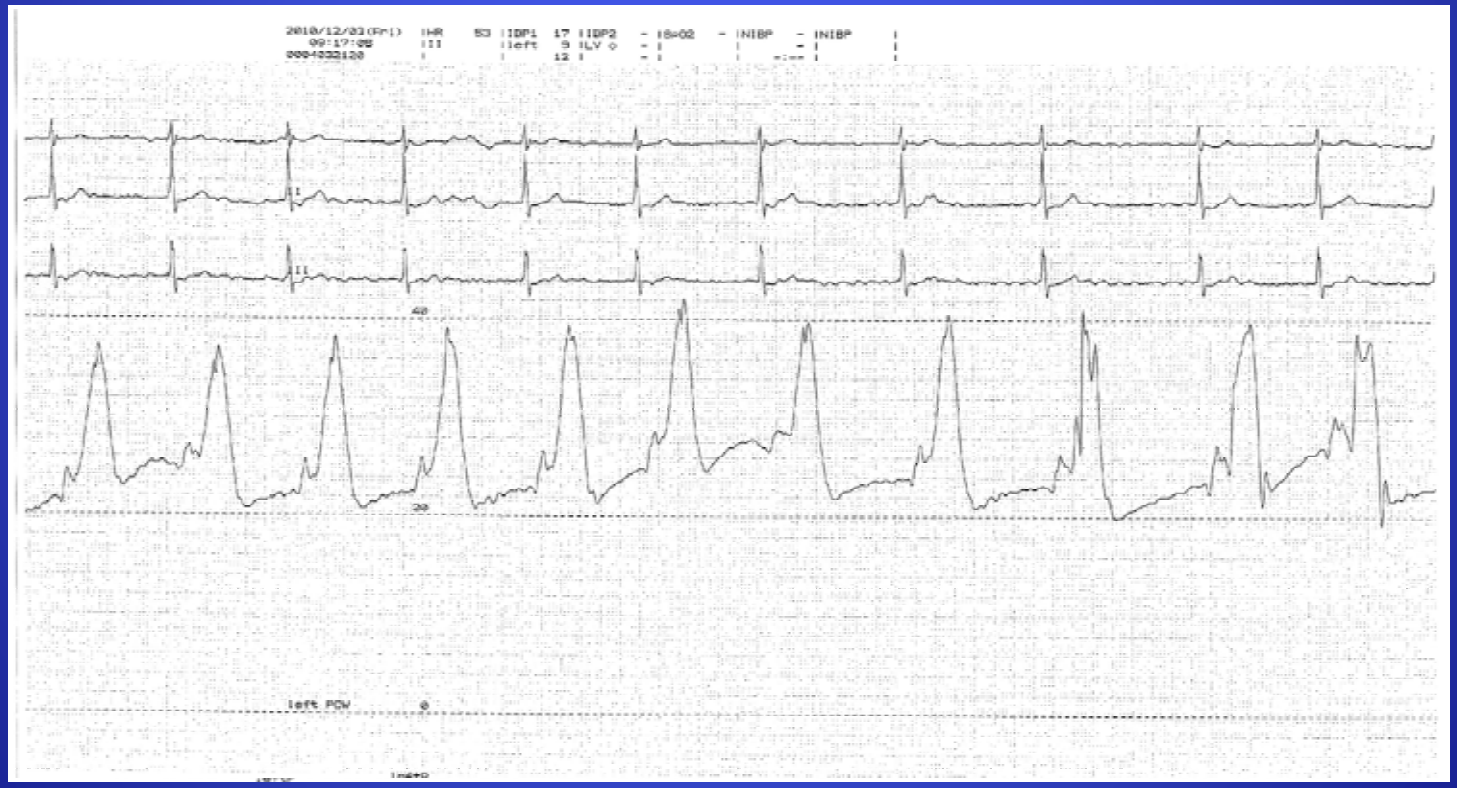
Case: 75 M

- ✓ *ASD diameter: 24mm*
- ✓ *Mitral regurgitation (mild)*
- ✓ *Qp/Qs: 3.18*
- ✓ *Hypertension, permanent Afib, CKD*
- ✓ *NYHA class III HF despite medication*
- ✓ *PA pressure: 57/19/32 mmHg*
- ✓ *BNP 351 pg/ml*

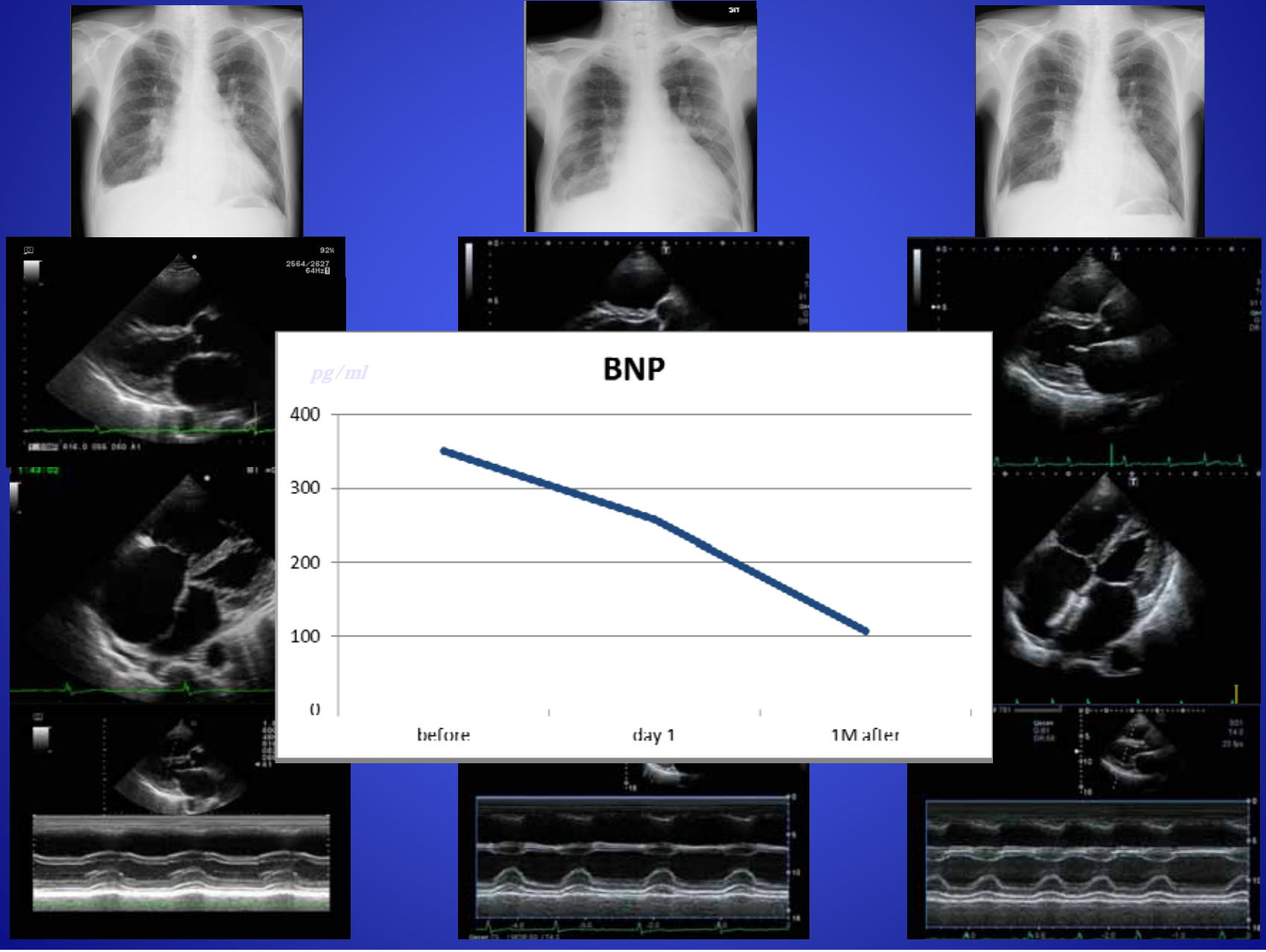
Chest X-ray

Change in PCWP

by test balloon occlusion

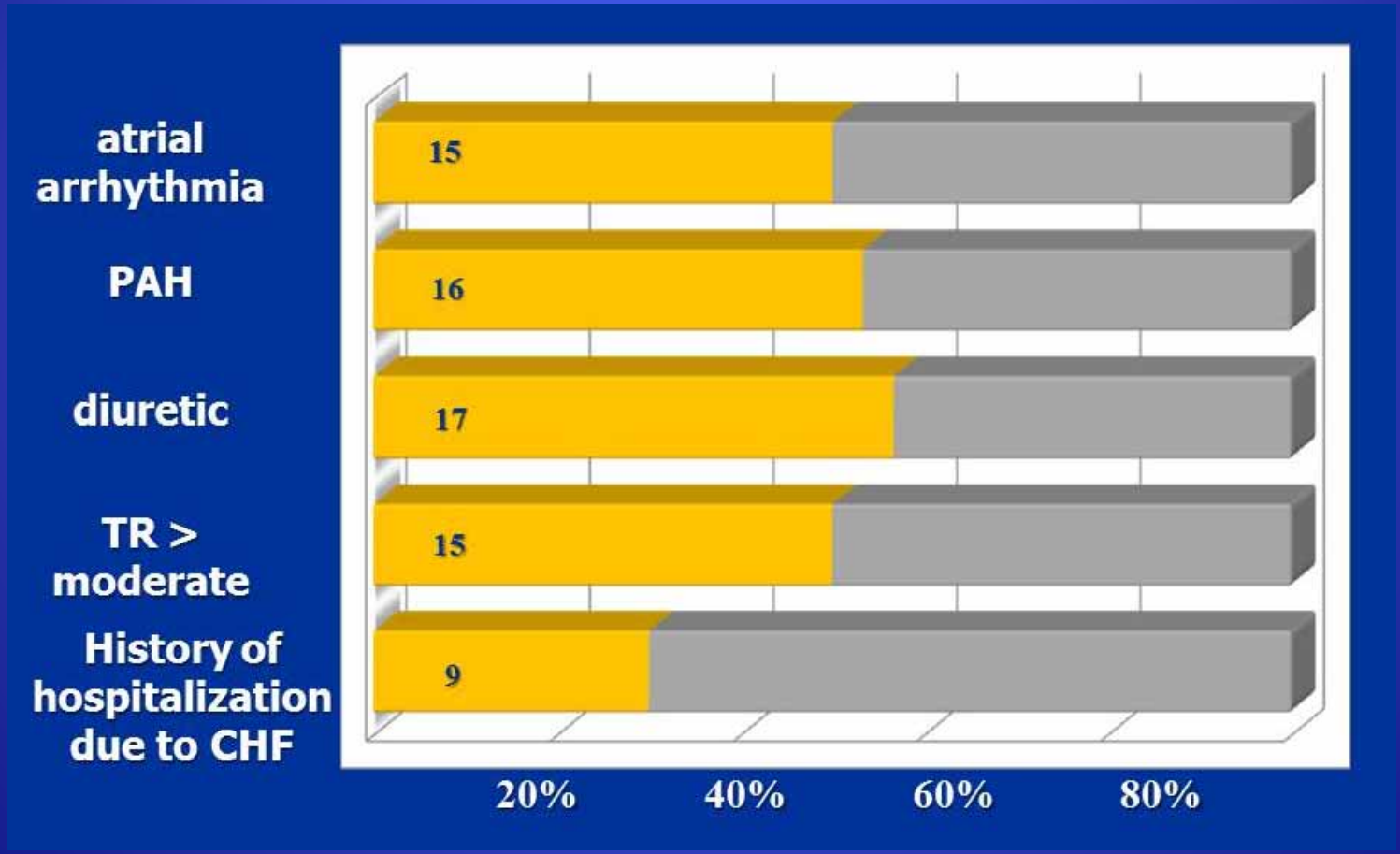


before *day 1* *1M after*



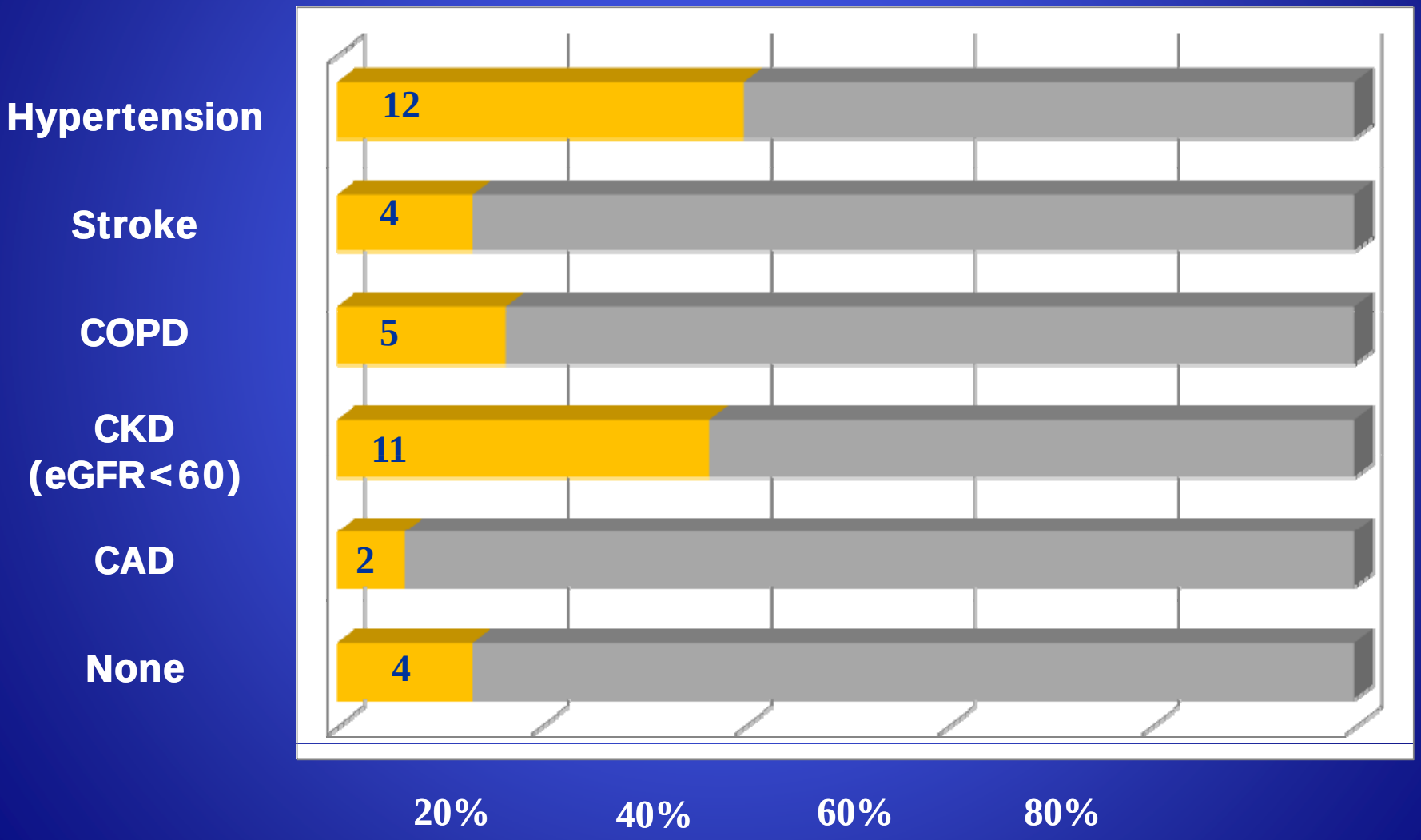
Transcatheter Closure of Atrial Septal Defect in a Geriatric Population

Koji Nakagawa,[1] MD, Teiji Akagi,[2*] MD, PhD, FSCAI, Manabu Taniguchi,[2] MD, PhD, Yasufumi Kijima,[1] MD, Keiji Goto,[3] MD, PhD, Kengo F. Kusano,[1] MD, PhD, Hiroshi Itoh,[1] MD, PhD, and Shunji Sano,[4] MD, PhD



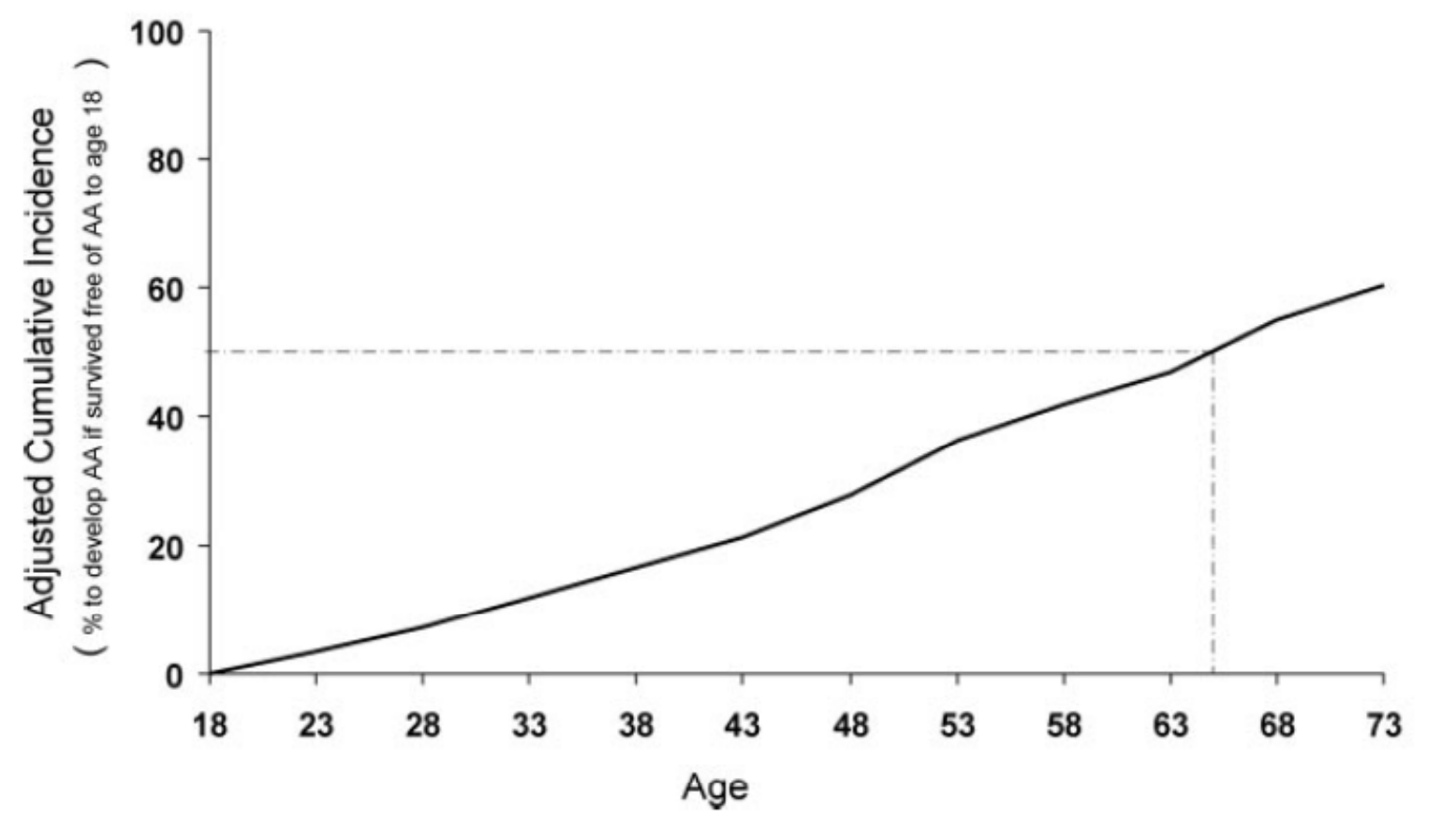
Catheter Cardiovasc Interv 2012 (in press)

Comorbidities



Atrial Arrhythmias in Adults With Congenital Heart Disease

Judith Bouchardy, MD; Judith Therrien, MD; Louise Pilote, MD, MPH, PhD;
Raluca Ionescu-Ittu, MSc; Giuseppe Martucci, MD; Natalie Bottega, MD; Ariane J. Marelli, MD

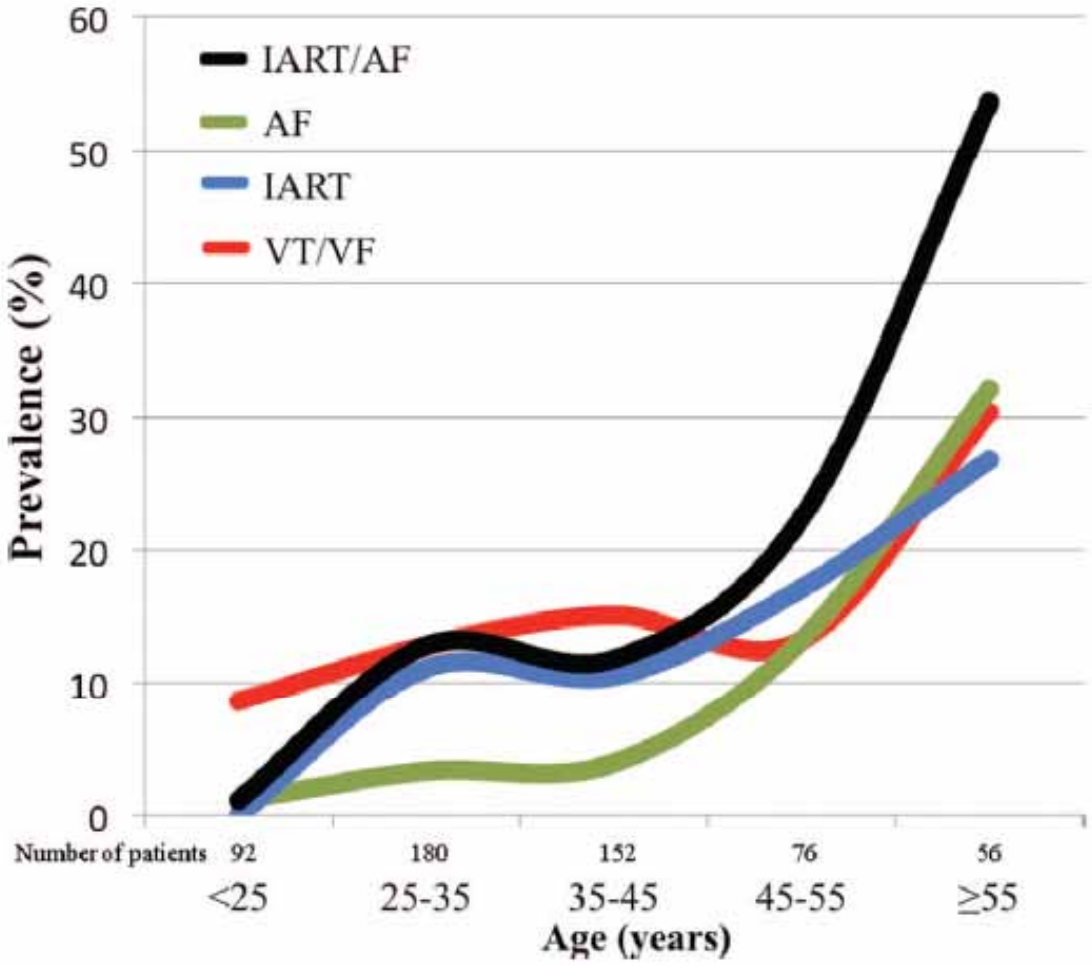


(*Circulation*. 2009;120:1679-1686.)

Arrhythmia Burden in Adults With Surgically Repaired Tetralogy of Fallot

A Multi-Institutional Study

Pa
Alexa
Anne Marie V
Stephen Cook
Michael J.
D;
, MD;
Gersony, MD;
y Webb, MD;
Research in



(*Circulation*. 2010;122:868-875.)

Team approach for ACHD patients

- Role of Pediatric cardiologists

  morphological diagnosis, surgical information hemodynamic features,

- Role of Adult cardiologists

  diagnosis and management of co-morbidities, management of in-patient's care

- Role of ACDH specialists

  overall management and follow-up

Circulation Journal
Official Journal of the Japanese Circulation Society
http://www.j-circ.or.jp

Status and Future Needs of Regional Adult Congenital Heart Disease Centers in Japan

– A Nationwide Survey –

Ryota Ochiai, PhD; Atsushi Yao, MD, PhD; Koichiro Kinugawa, MD, PhD;

Table 1. Recommendations for Optimal ACHD Care[18]*

1. An ACHD referral center must employ at least 1, preferably 2, cardiologist(s) specifically trained and educated in the care of adults with CHD.
2. Specialized ACHD centers should provide care in connection with pediatric cardiology and/or congenital cardiac surgery.
3. Specialist centers† must treat sufficient numbers of patients and perform a sufficient number of procedures to be effective as well as develop and maintain high levels of performance.
4. General adult cardiac facilities and non-specialist centers should have an established referral relationship with a specialist center.
5. A minimum of 2 cardiac surgeons trained in and practicing adult and pediatric cardiac surgery are required.
6. The optimal activity for a pediatric and congenital heart surgeon is 125 operations per year. Specifically, for ACHD, a minimum of 50 operations per year is recommended.
7. A fully equipped electrophysiology laboratory staffed by properly trained electrophysiologists with experience in detecting arrhythmias inherent to CHD and with experience in pacemaker technology, ablation technology, and defibrillator implantation must available.
8. An ACHD referral center must employ at least 1 nurse specialist that is trained and educated in the care of ACHD patients.

Why pediatric cardiologists keep to care adult CHD patients ?

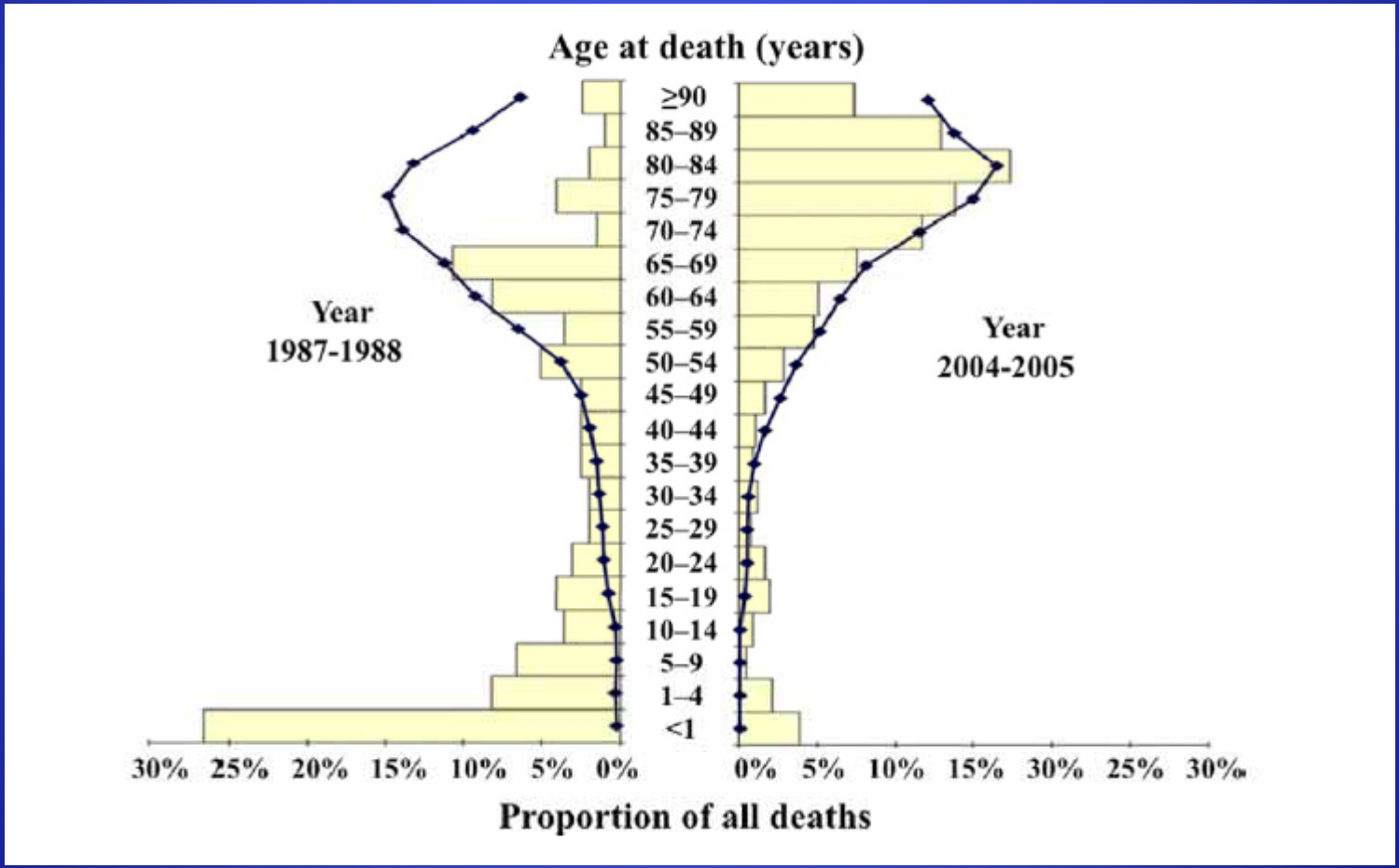
- They know long-history, condition, events….
- Patients prefer to stay same doctor (clinic).
- There is no adult cardiologists to transfer.
- Adult cardiologists have no interests.
- Patients are following at children’s hospital.

How long adult CHD patients are followed by pediatric cardiologists?

No one knows.

Changing Mortality in Congenital Heart Disease

Paul Khairy, MD, PhD,* Raluca Ionescu-Ittu, MSc,†§ Andrew S. Mackie, MD, SM,†
Michal Abrahamowicz, PhD,§ Louise Pilote, MD, MPH, PhD,‡§ Ariane J. Marelli, MD†



(J Am Coll Cardiol 2010;56:1149–57)

Background

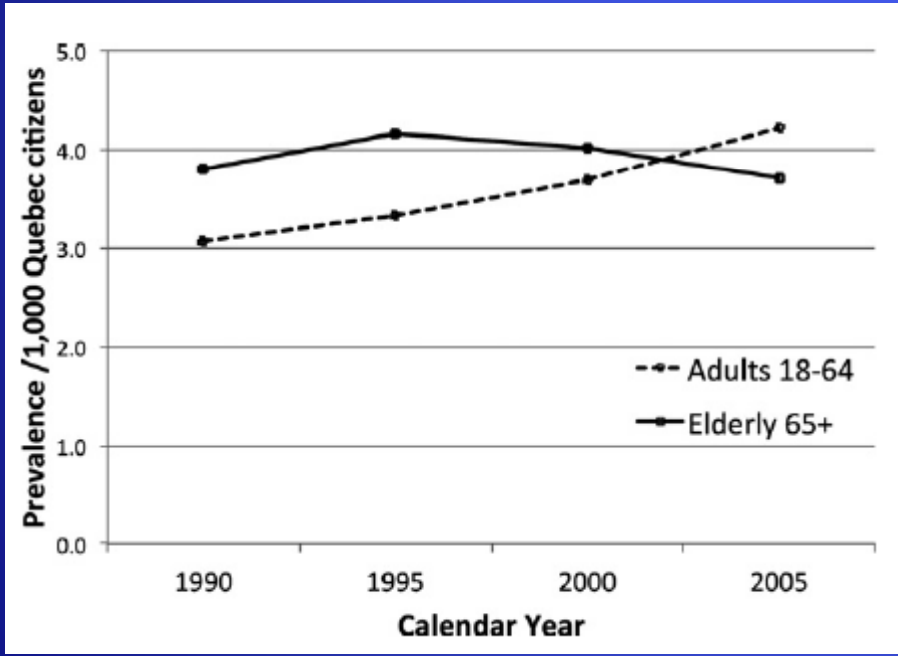
- Population of adult congenital heart disease patients are increasing 9000/years in Japan.
- Population of pediatric cardiologists has not increased yet, rather the majority of hospitals are struggling to keep the specialists.

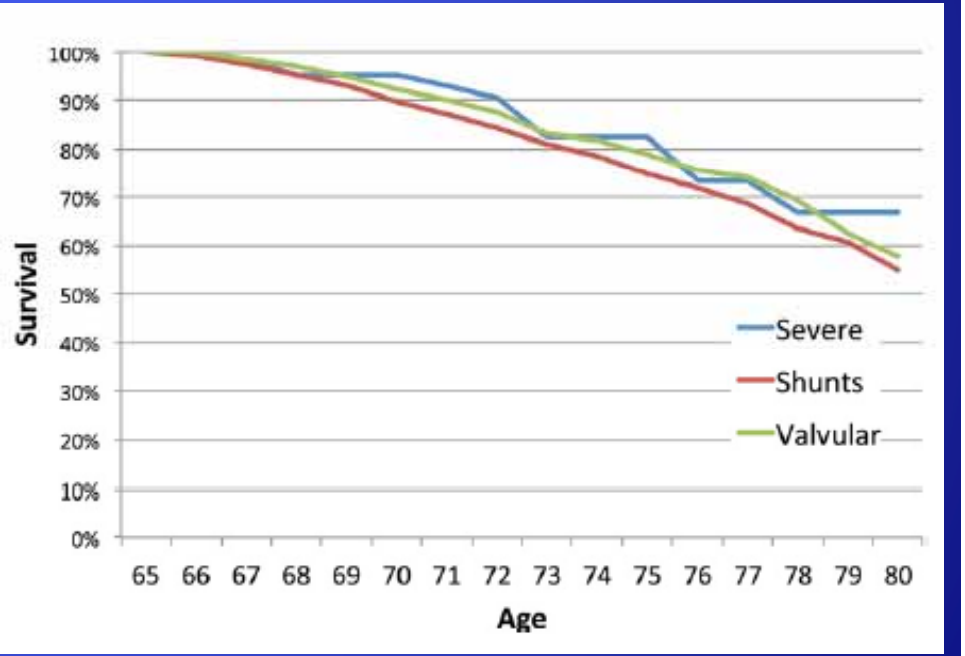
Geriatric Congenital Heart Disease

Burden of Disease and Predictors of Mortality

Jonathan Afilalo, MD, MSc,* Judith Therrien, MD,*‡ Louise Pilote, MD, MPH, PhD,†
Raluca Ionescu-Ittu, MSc,‡ Giuseppe Martucci, MD,‡ Ariane J. Marelli, MD, MPH‡

Montreal, Quebec, Canada





J. Am. Coll. Cardiol. 2011;58;1509-1515

循環器病の診断と治療に関するガイドライン（2004－2005年度合同研究班報告）

成人先天性心疾患診療ガイドライン(2006年改訂版)

Guidelines for Management of Congenital Heart Diseases in Adults (JCS 2006)

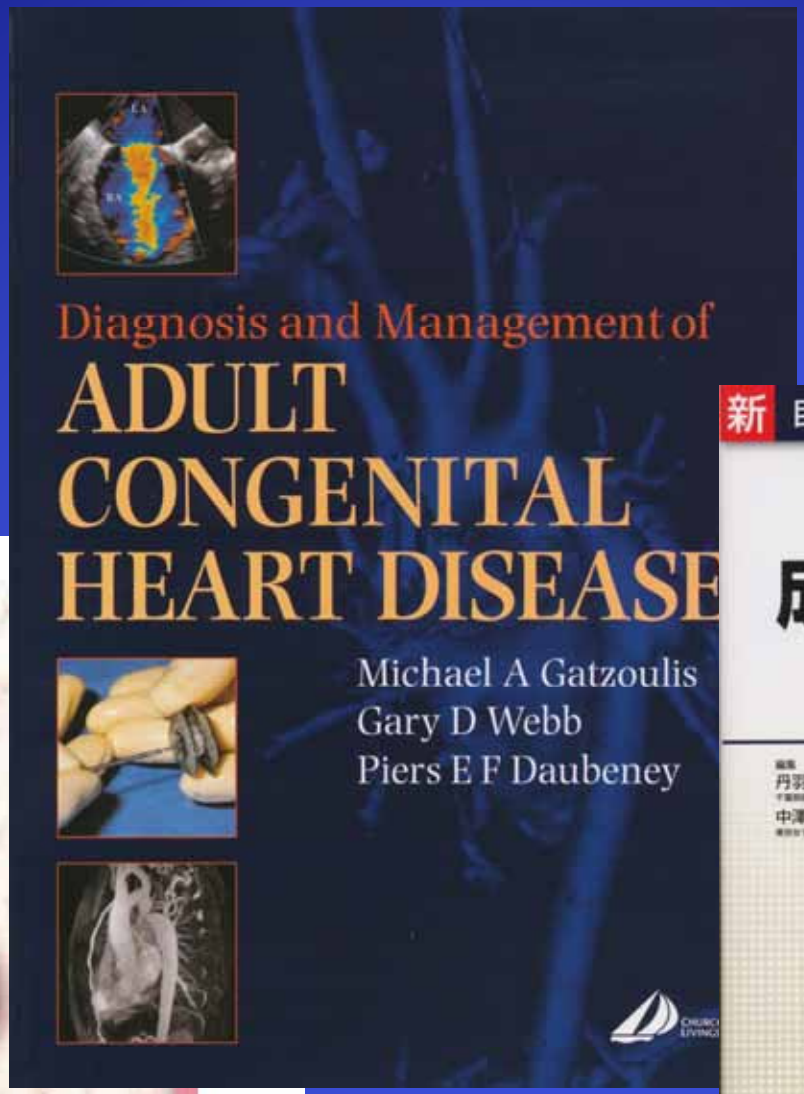
合同研究班参加学会：日本循環器学会，日本胸部外科学会，日本産科婦人科学会，日本小児循環器学会，
日本心臓病学会

班　長　黒　澤　博　身　東京女子医科大学心臓血管外科

班　員　赤　木　禎　治　岡山大学循環器疾患治療部
石　澤　　瞭　国立成育医療センター循環器科
市　田　蕗　子　富山大学小児科
越　後　茂　之　国立循環器病センター小児科
大　嶋　義　博　兵庫県立こども病院心臓血管外科
角　　秀　秋　福岡市立こども病院心臓血管外科

協力員　市　川　　肇　大阪大学臓器制御外科
牛ノ濱　大　也　福岡市立こども病院循環器科
川　俣　和　弥　国立循環器病センター周産期
小　垣　滋　豊　大阪大学小児科
高　橋　一　浩　東京女子医科大学循環器小児科
立　野　　滋　千葉県循環器病センター小児科


先天性心疾患の方のための
妊娠・出産ガイドブック
編著
丹羽公一郎
中外医学社


Diagnosis and Management of
ADULT CONGENITAL HEART DISEASE
Michael A Gatzoulis
Gary D Webb
Piers E F Daubeney


新 目でみる循環器病シリーズ
14
成人先天性心疾患
編集
丹羽 公一郎
中澤 誠
MEDICAL VIEW

Japanese Society for ACHD

JSACHD

日本成人先天性心疾患学会

Japanese Society for Adult Congenial Heart Disease

- HOME
- 学会について
  - 学会のご案内
  - 成人先天性心疾患とは
  - 沿革
  - 役員
  - 学会会則
  - 会則施行細則
- 入退会
  - 入会について
  - 退会について
- 総会・学術集会、セミナー
  - 総会・学術集会
  - セミナー
  - 過去に開催された学術集会（プログラム）
  - 過去に開催された学術集会（プログラム・抄録　会員専用）

HOME

会員継続・入会申込はこちらからお手続き下さい

新着情報

第14回 日本成人先天性心疾患学会終了の御礼　2012年01月25日

第14回 日本成人先天性心疾患学会終了のお礼とご報告を掲載しました

第14回日本成人先天性心疾患学会　総会・学術集会のご案内　2011年12月20日

日時　2012年1月14日（土）・15日（日）

場所　聖路加看護大学 Alice G St John's Memorial Hall　東京

Educational Seminar

- Annually (usually June and October)
- Target: young doctor, nurse, co-medical staff
- 1 to 1.5 days seminar
- Lecture, Case discussion, Free discussion
- Meeting venue: Tokyo and Osaka
- Attendees: 150 (2009), 200 (2010), 300 (2011)

JSACHD

International Society

- Grown-Up Congenital Heart disease

Grown Up Congenital Heart Patients Association
Supporting Young People and Adults with Congenital Heart Disease

- International Society for Adult Congenital Heart Disease

International Society for
Adult Congenital Heart Disease

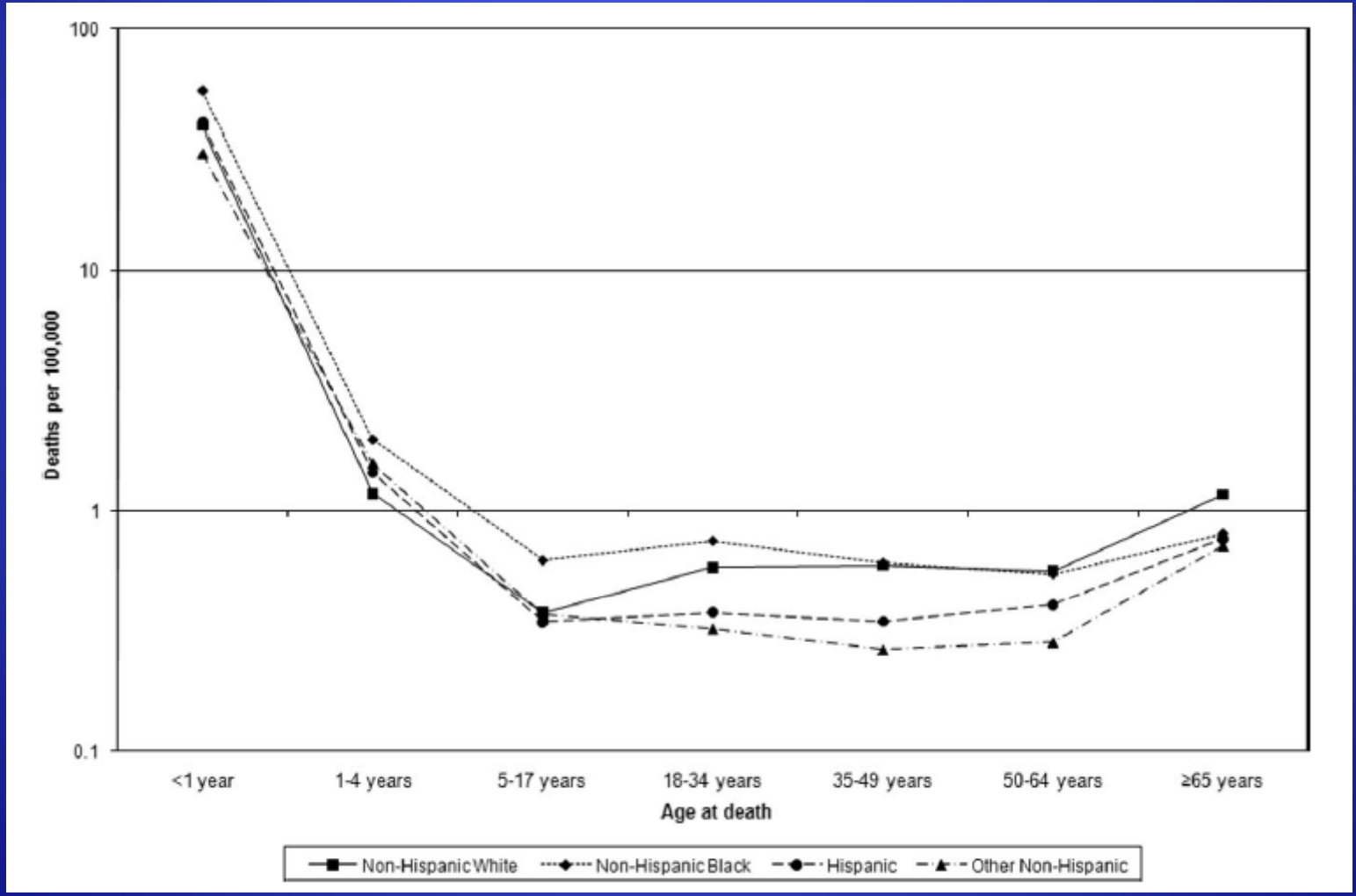
- Asia-Pacific Society for Adult Congenital Heart Disease

Asia Pacific Society for ACHD

- Established in 2008
- 1st meeting was held in Jeju, Korea as a conjunction meeting with 2nd APPCCS.
- 3rd meeting will be held in Taipei, 2012.

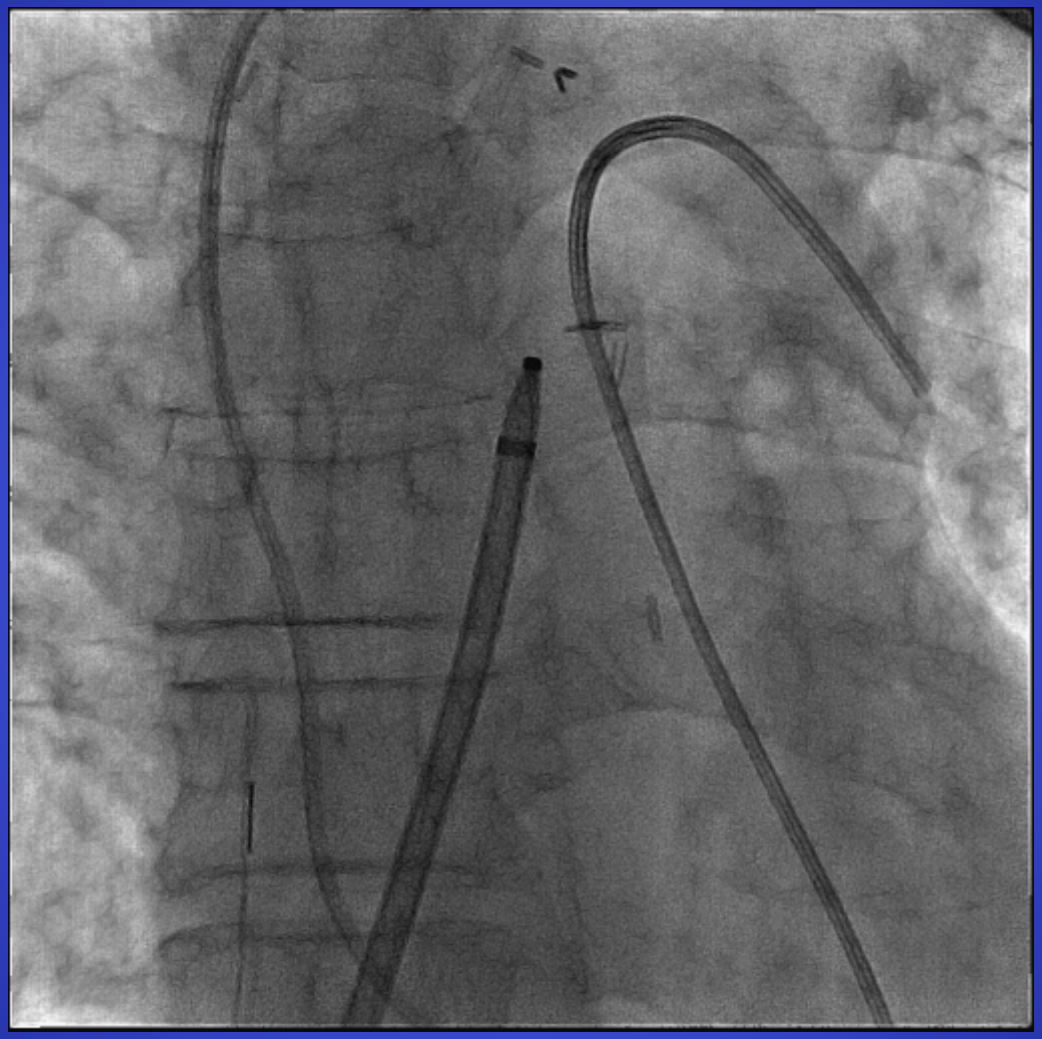
Mortality Resulting From Congenital Heart Disease Among Children and Adults in the United States, 1999 to 2006

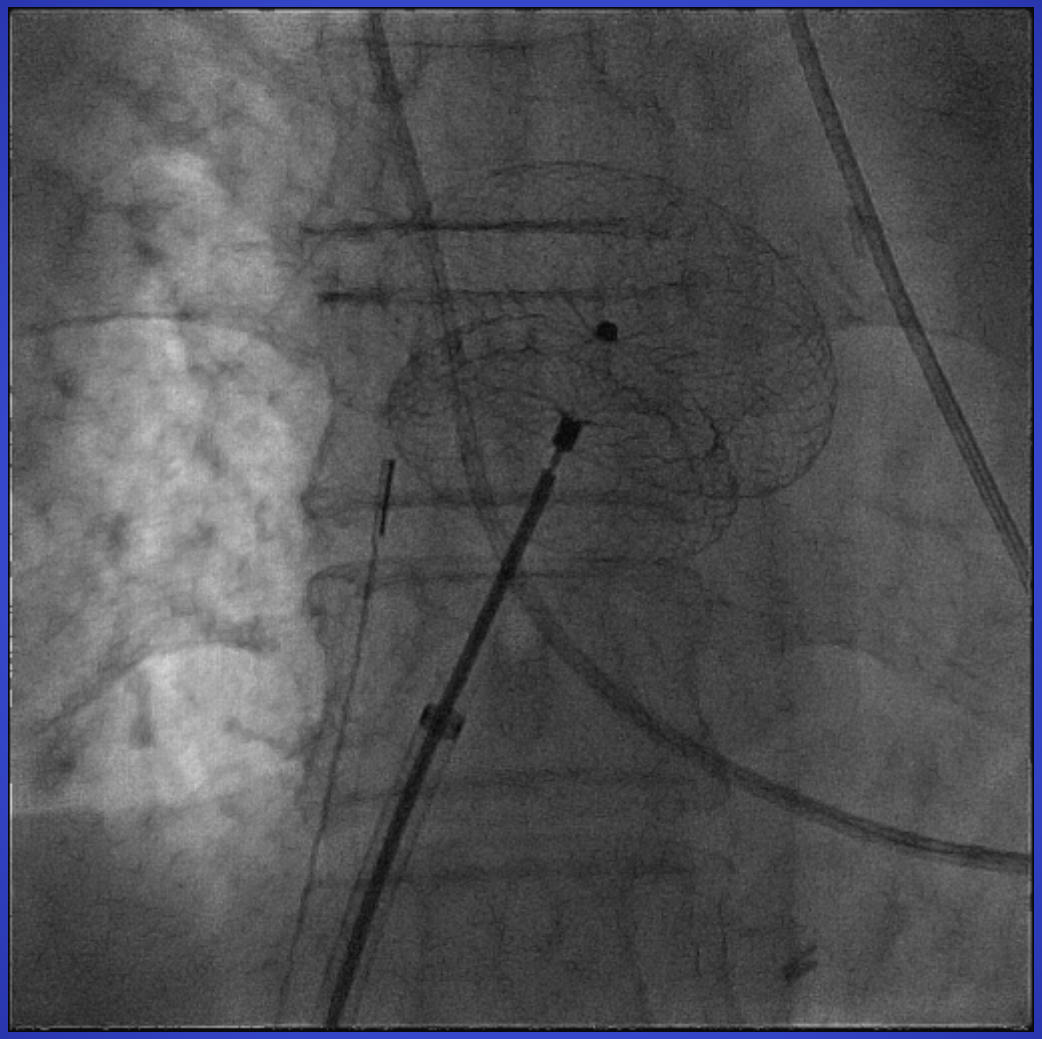
Suzanne M. Gilboa, PhD; Jason L. Salemi, MPH; Wendy N. Nembhard, PhD;
David E. Fixler, MD; Adolfo Correa, MD, PhD



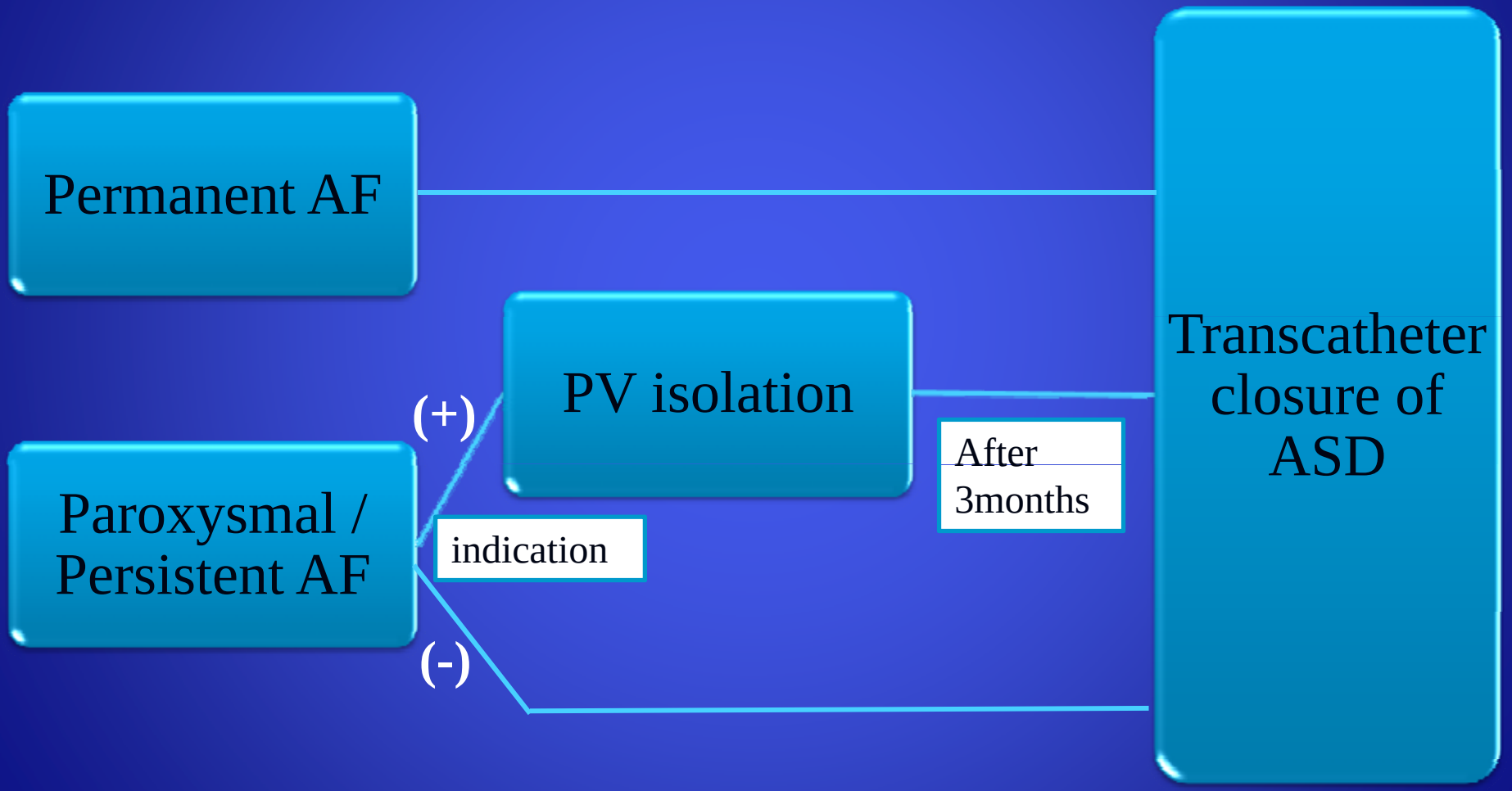
60 years male

- ASD was pointed out 10 years ago, however he refused to surgery. He developed congestive heart failure with pulmonary hypertension.
- At the time of hemodynamic evaluation, esophageal cancer was detected.
- GI team requested ASD closure before the surgical esophageal resection.
- However, he developed hemorrhagic shock due to massive GI bleeding at the day of catheter closure.





Therapeutic Strategies for Patients with ASD and Atrial Arrhythmia



Cardiovascular function and aging

- Pomp function
- AV valve regurgitation
- Atherosclerosis
- Co-morbidities
  - Coronary stenosis
  - Respiratory disease
  - Kidney disease
  - Cancer, etc

岡山大学病院における
成人ASDカテーテル閉鎖術までの流れ

他院、他科からの紹介、検診後精査依頼
自身での来院　(internetなどの情報)

外来

循環器内科外来　または　心臓血管外科外来

経胸壁心エコー、胸部Xp、ECG、採血、CPXなど

入院

クリニカルパス

3泊4日

4泊5日(土日挟む)

経食道心エコー　(全例)

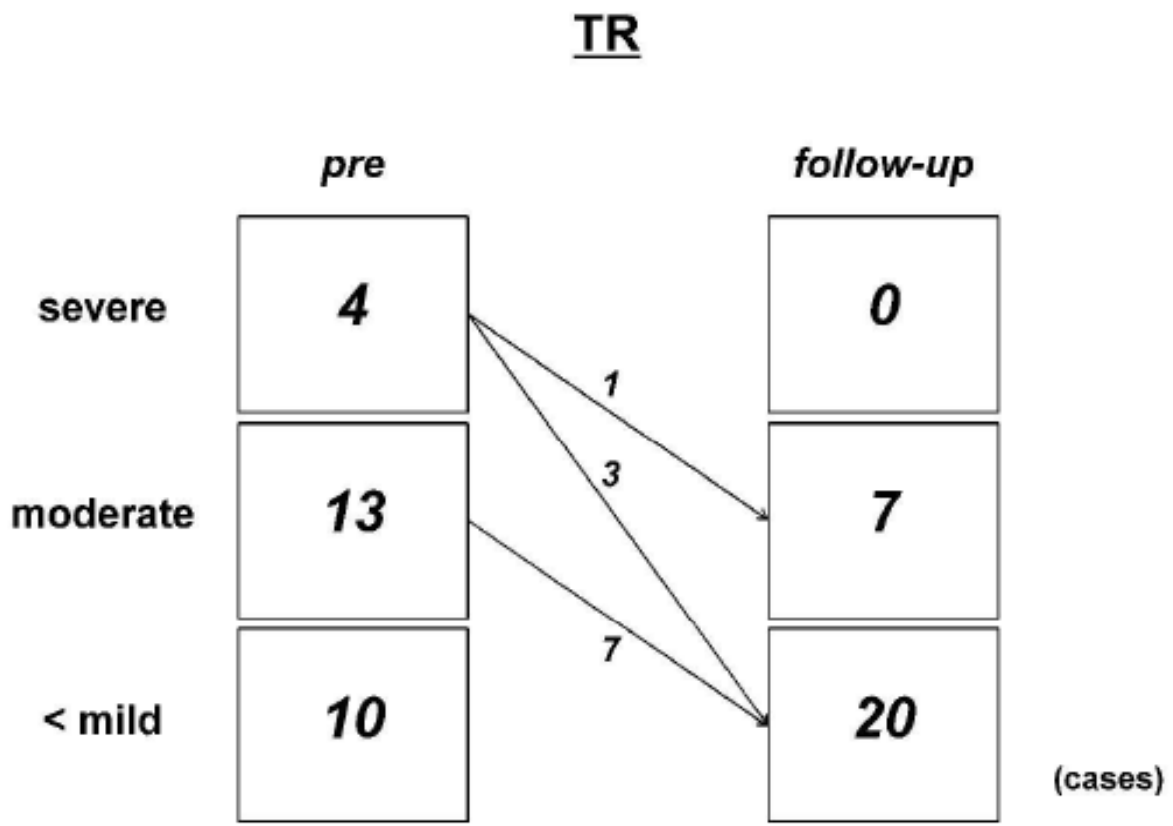
心臓カテーテル検査
(40歳以上で心血管リスクのある患者は冠動脈造影)

経皮的ASD閉鎖術

心臓血管外科医
循環器内科医
循環器小児科医
心エコー専門医
心エコー専門技師

Adult Congenital Heart Disease

Same name, but different disease


TR
pre
follow-up
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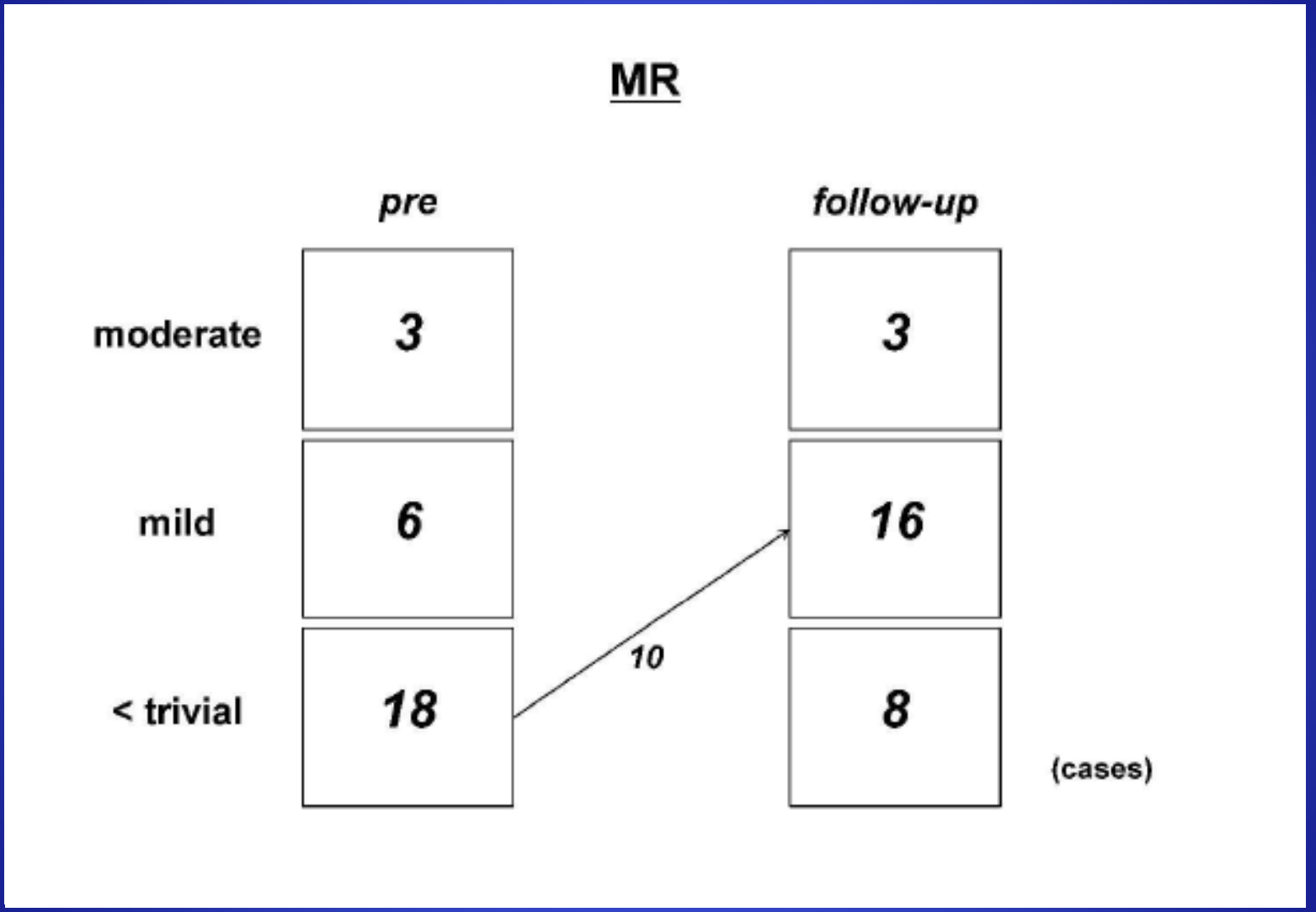
moderate
< mild
4
13
10
0
7
20
1
3
7
(cases)


MR
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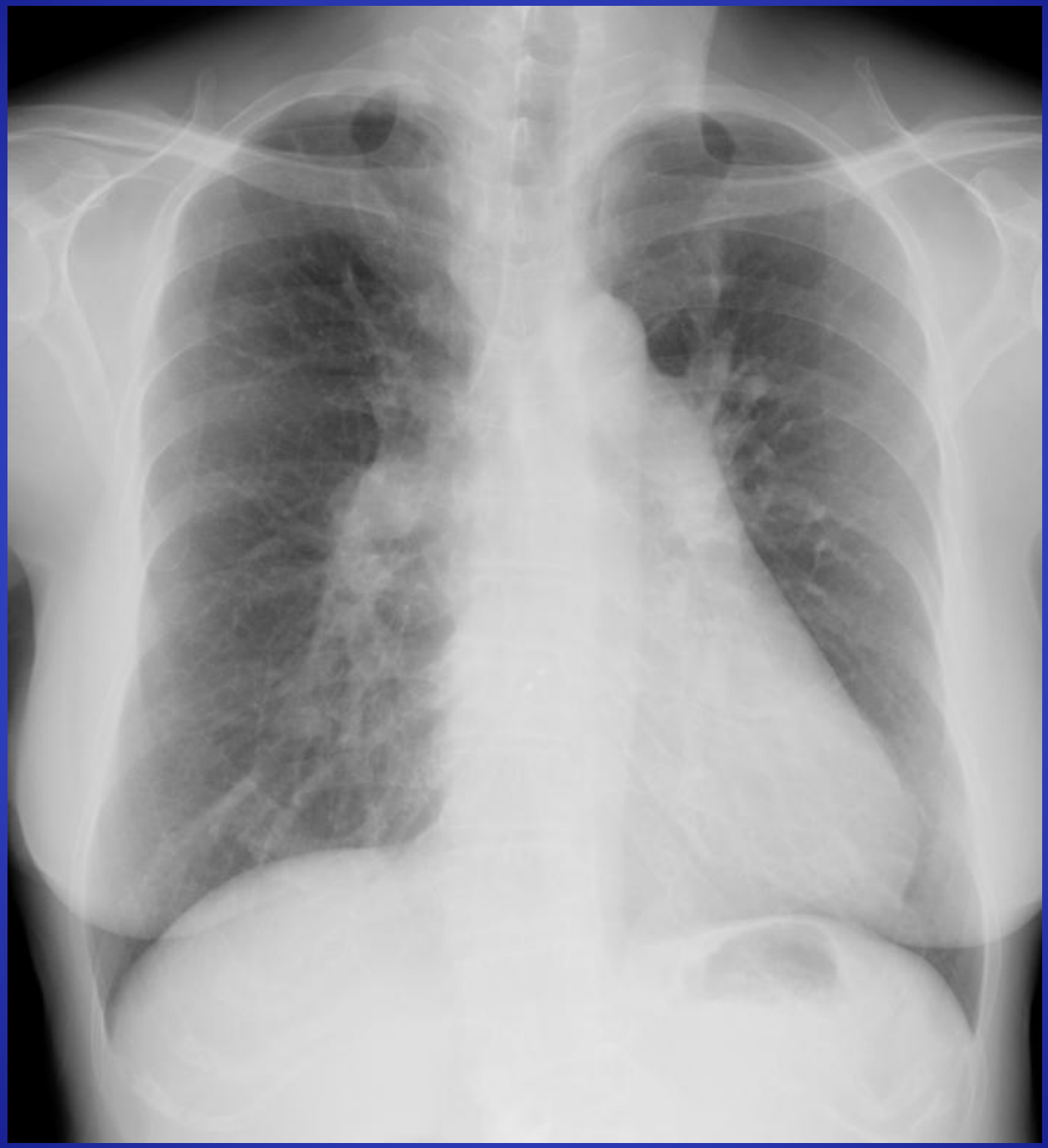

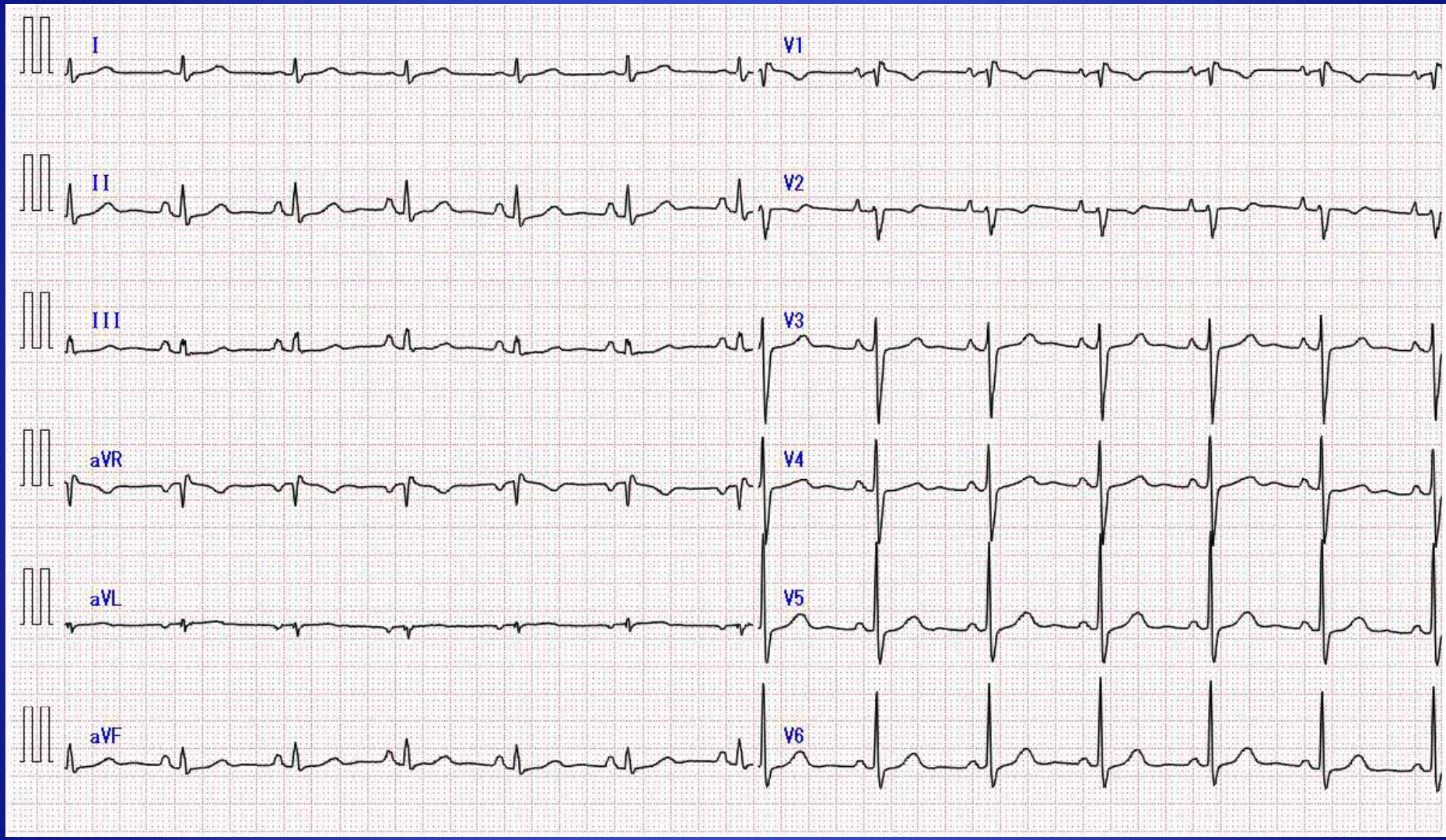
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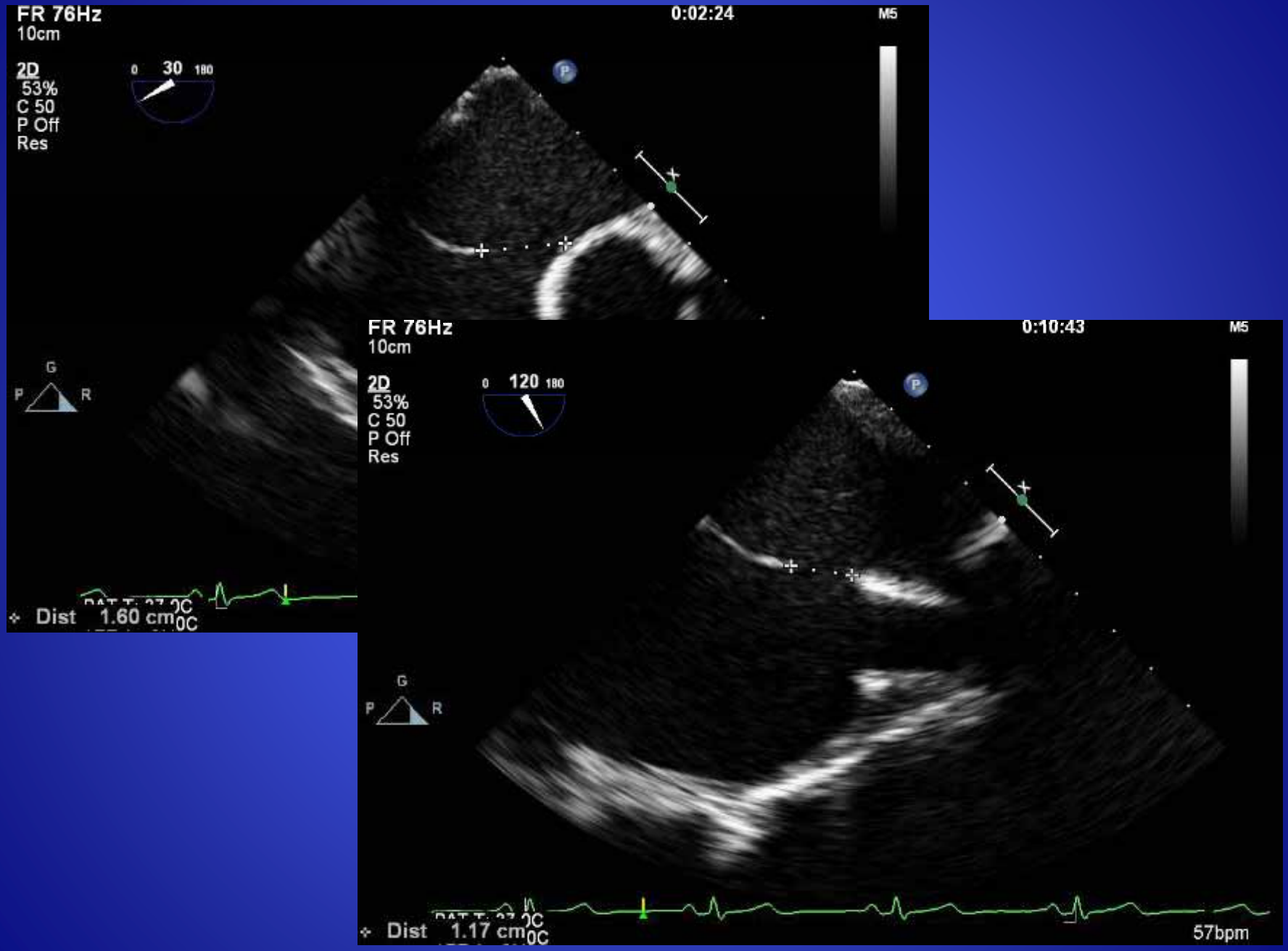
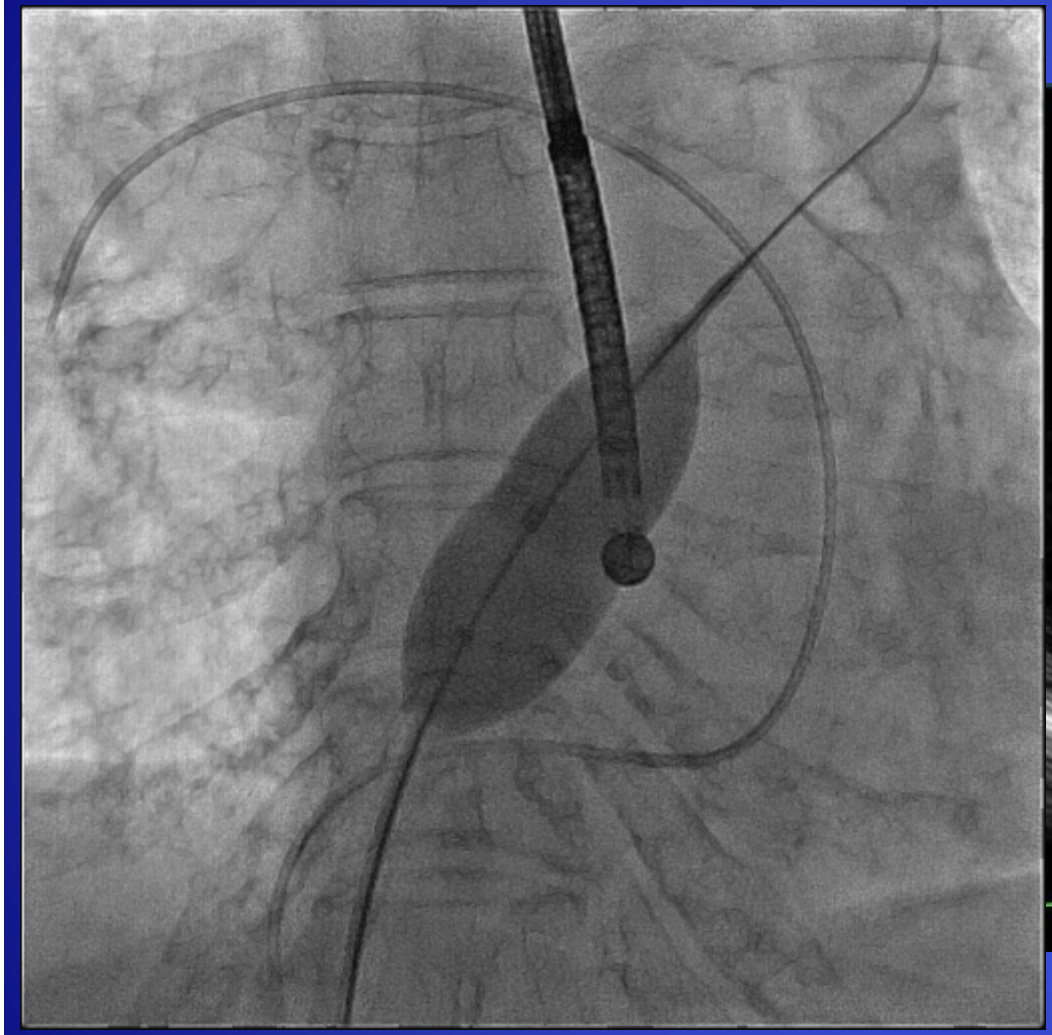
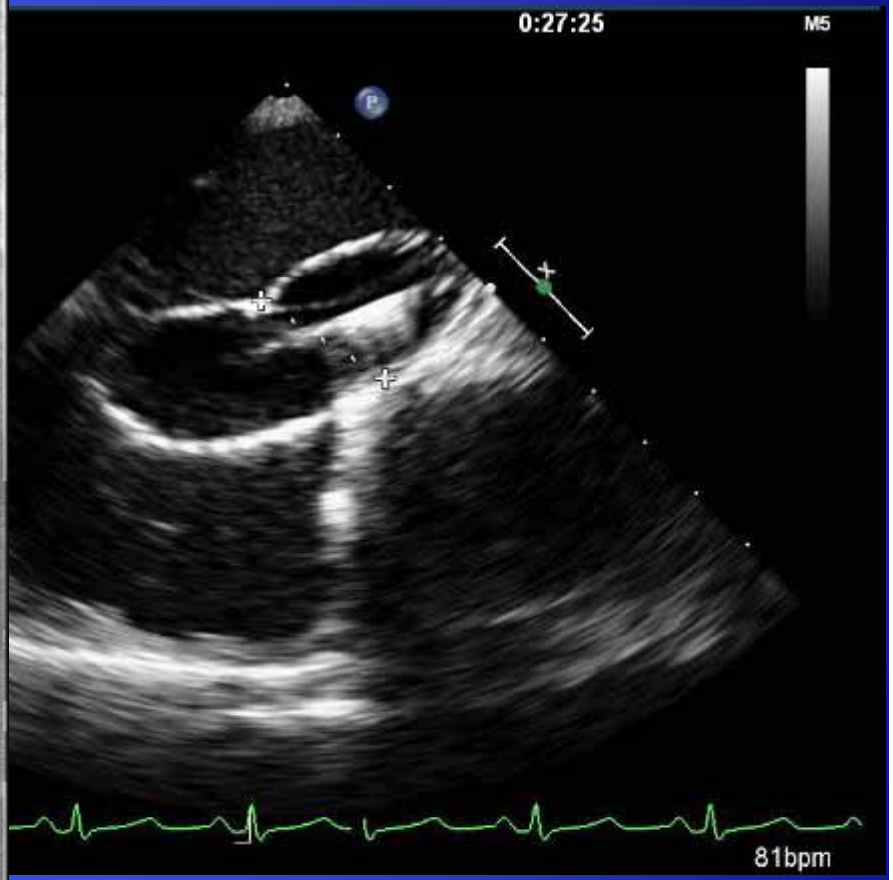
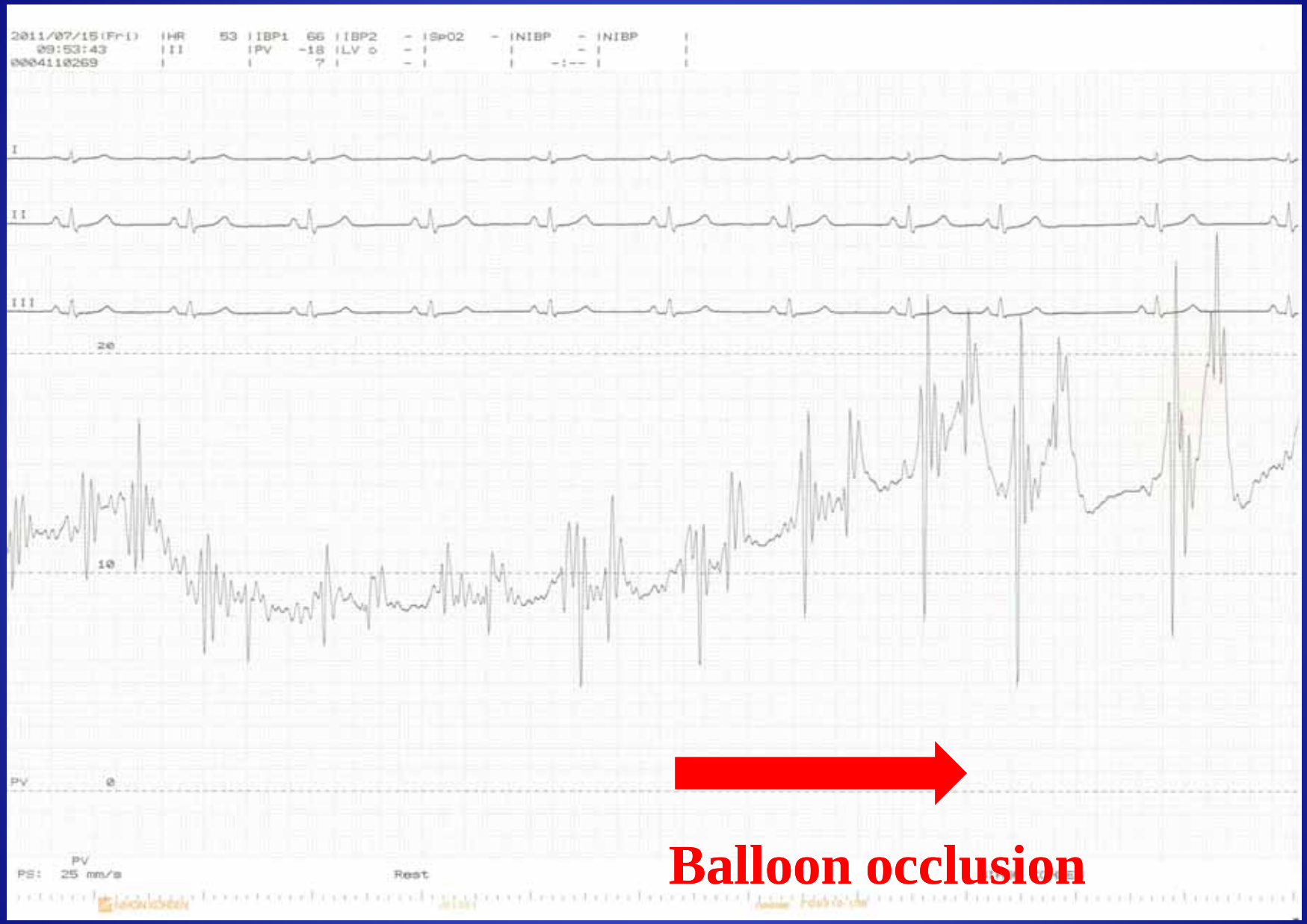
V6

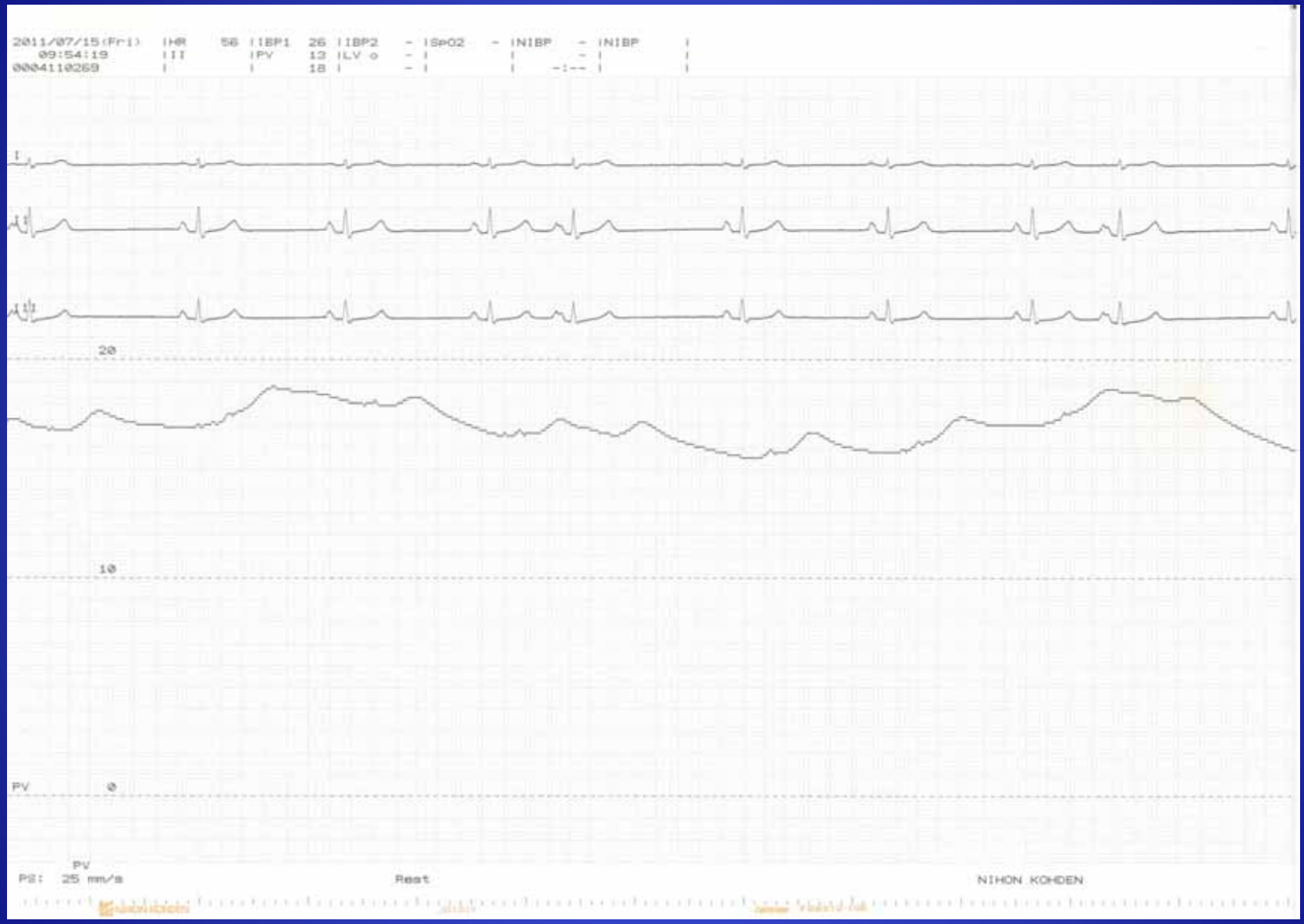
項目名	結果	コメント	項目名	結果	コメント	項目名	結果	コメント	項目名	結果	コメント
血液学的検査			APTT	31.4		Mg	2.0		ﾕｹﾂﾏISAB	1000.0 +	↑
WBC	4.07		Fibg	354		UN	21.0		ﾕｹﾂﾏICAB	82.4 +	
RBC	3.93		PT(%)	92		CRTN	0.57		ﾕｹﾂﾏIHCV	0.1 -	未満
Hb	12.3		PT-INR	1.05		UA	4.5		ﾕｹﾂﾏICｺｱ	49.9	以下
Ht	36.4		生化学的検査			T.CHO	201		ﾕｹﾂﾏIHIV	0.1 -	未満
PLT	203		TP	7.6		UN/CRTN	36.8		分野なし		
MCV	92.6		Alb	4.5		CK	150		TIME-14		
MCH	31.3		TTT	3.0		eGFR>=18	78.3				
MCHC	33.7		ZTT	16.8		H(ﾖｳｹﾂ)	1				
RDW	12.8		T.Bil	0.96		L(ｺﾝﾀﾞｸ)	0				
Pct	0.160		D.Bil	0.16		I(ｵｳﾀﾞﾝ)	1				
MPV	8.1		AST	24		FBS	85				
PDW	41.8		ALT	19		A1Cｲﾗｲ					
HDW	2.53		ALP	284		A1c(JDS)	4.9				
WBCｿﾞｳ			LAP	51		A1c(NGSP	5.3				
Ly	29.6		G-GT	27		内分泌的検査					
Mon	5.8		CHE	295		BNP	127.4				
Eos	0.9		LD	222		感染症血清					
Bas	0.6		A/G	1.45		STS(RPR)	0.3	－			
NE	63.2		Na	141		TP(TPLA)	<5.0	－			
NE#A	2.6		K	4.3		血漿蛋白					
LY#	1.2		Cl	109		CRP	0.05				
出血・凝固			Ca	8.8		輸血感染症					
PT	12.2		IP	3.6		ﾕｹﾂﾏISAG	0.1 -	未満			


FR 76Hz
10cm
2D
53%
C 50
P Off
Res
0 30 180
0:02:24
M5
G
P
R
Dist 1.60 cm
FR 76Hz
10cm
2D
53%
C 50
P Off
Res
0 120 180
0:10:43
M5
G
P
R
Dist 1.17 cm
57bpm

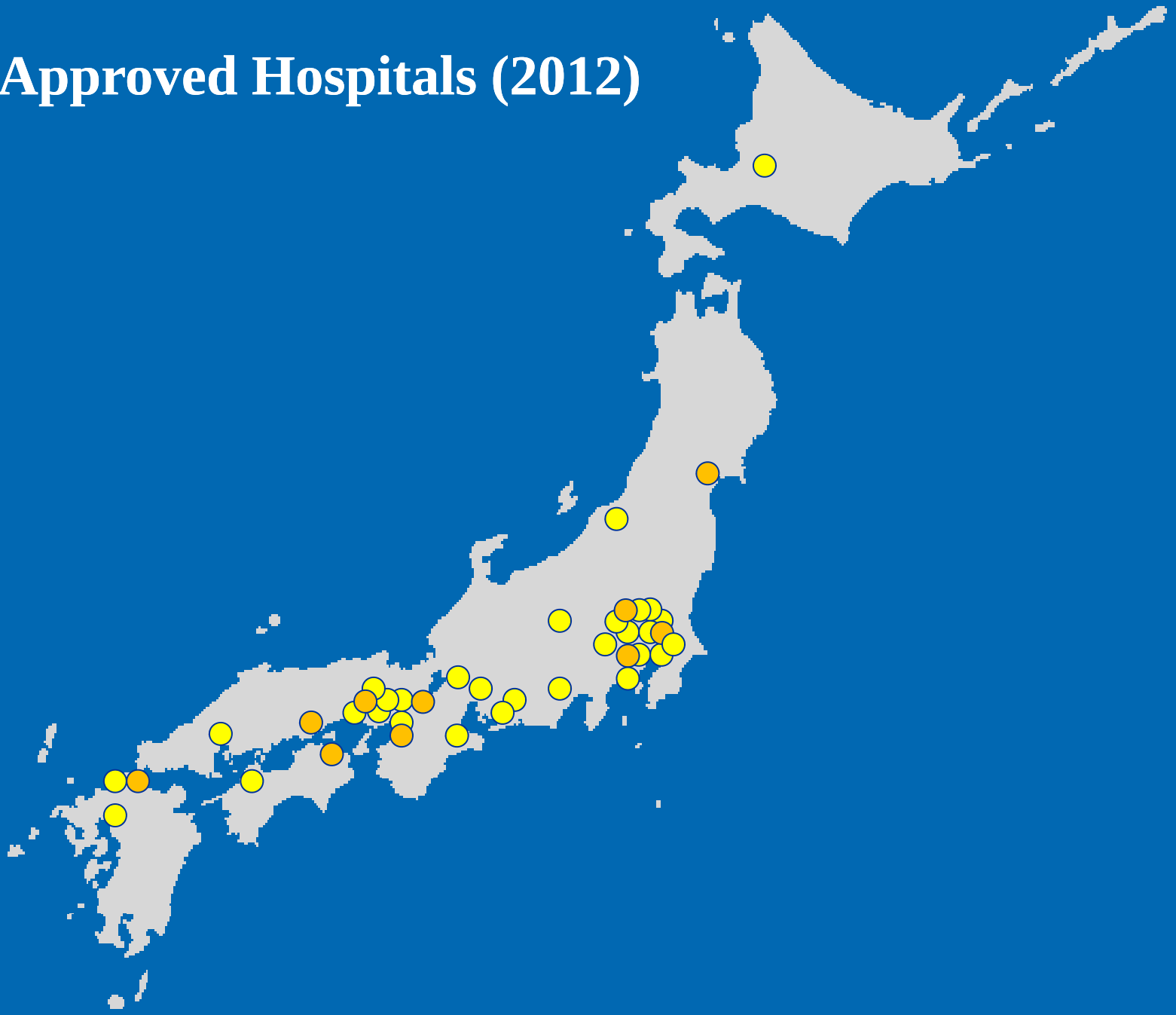



0:27:25
M5
81bpm


2011/07/15(Fri) HR 53 IBP1 66 IBP2 - SpO2 - NIBP - NIBP
09:53:43 II PV -18 LV o - - -:--
0004110269 7
I
II
III
20
10
PV
0
PV
PS: 25 mm/s
Rest
Balloon occlusion


2011/07/15(Fri) HR 56 IBP1 26 IBP2 - SpO2 - NIBP - NIBP
09:54:19 II PV 13 LV o - - -:-- -
0004110269 18 -
I
II
III
20
10
PV
0
PV
PS: 25 mm/s
Rest
NIHON KOHDEN

Approved Hospitals (2012)



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

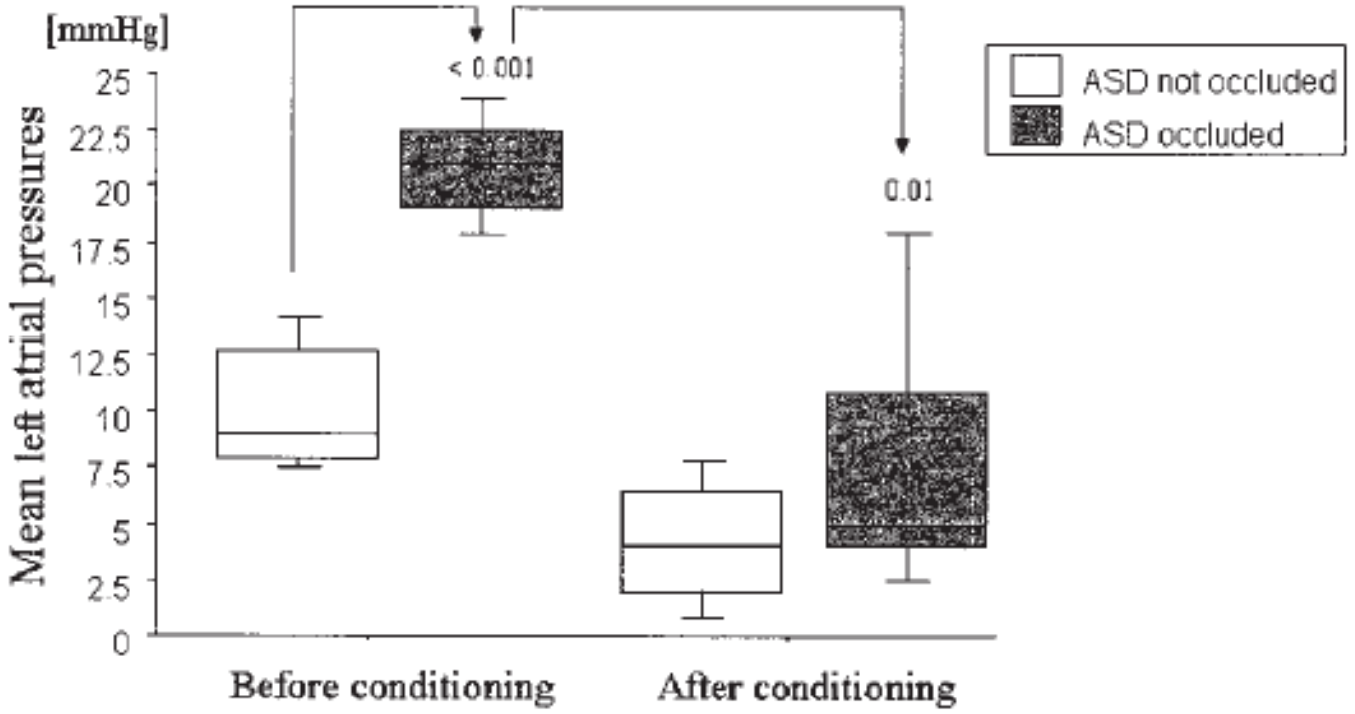
	Nonresponders (n = 11)	Responders (n = 7)	P
Age (median, years)	70	70	NS
Gender (f:m)	6:1	9:2	NS
Atrial flutter/fibrillation	4	4	NS
Systemic hypertension	5	3	NS
Coronary heart disease	0	1	NS
Defect diameter (mm)	24	25	NS
Shunt (Qp/Qs)	1.6	1.8	NS
Mean arterial pressure (mm Hg) before/during occlusion	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

Catheterization and Cardiovascular Interventions 52:177–180 (2001)

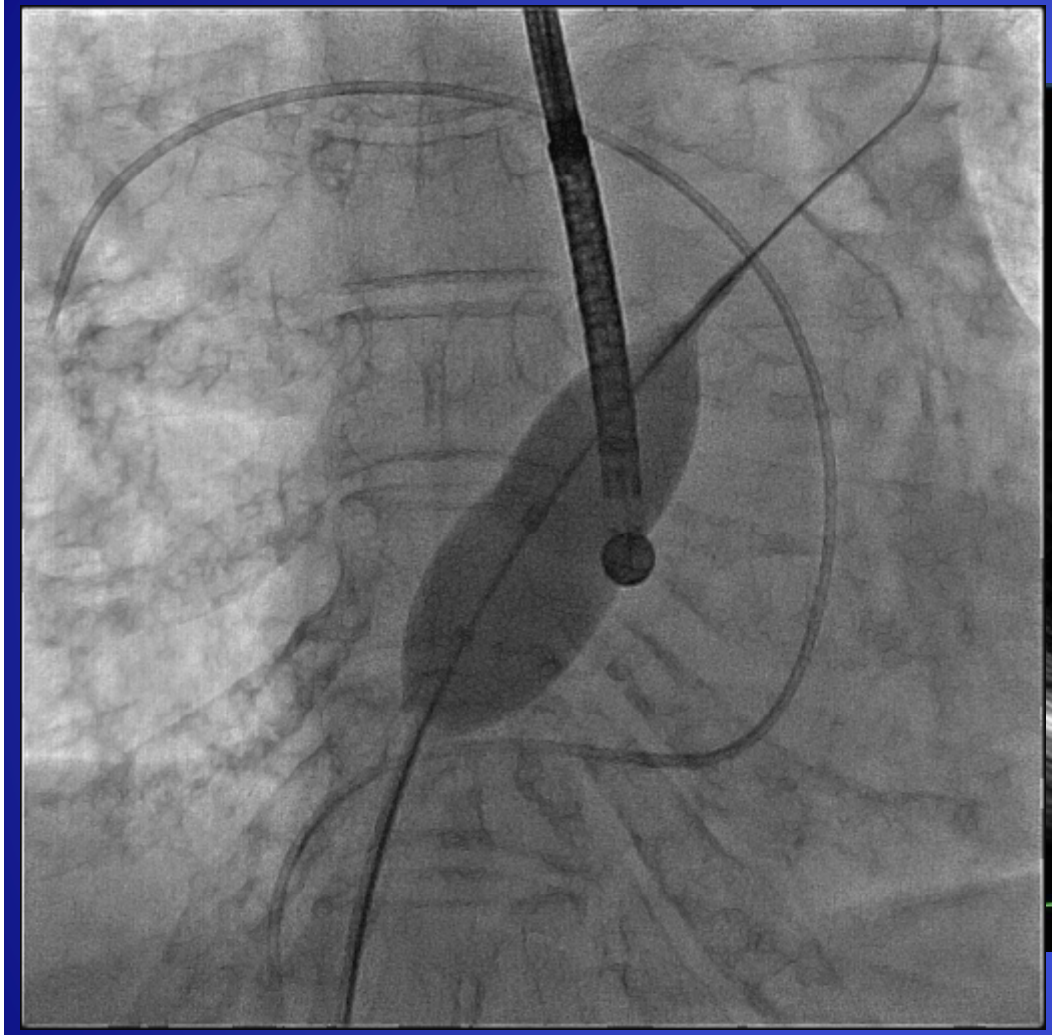
Left Ventricular Conditioning in the Elderly Patient to Prevent Congestive Heart Failure After Transcatheter Closure of Atrial Septal Defect

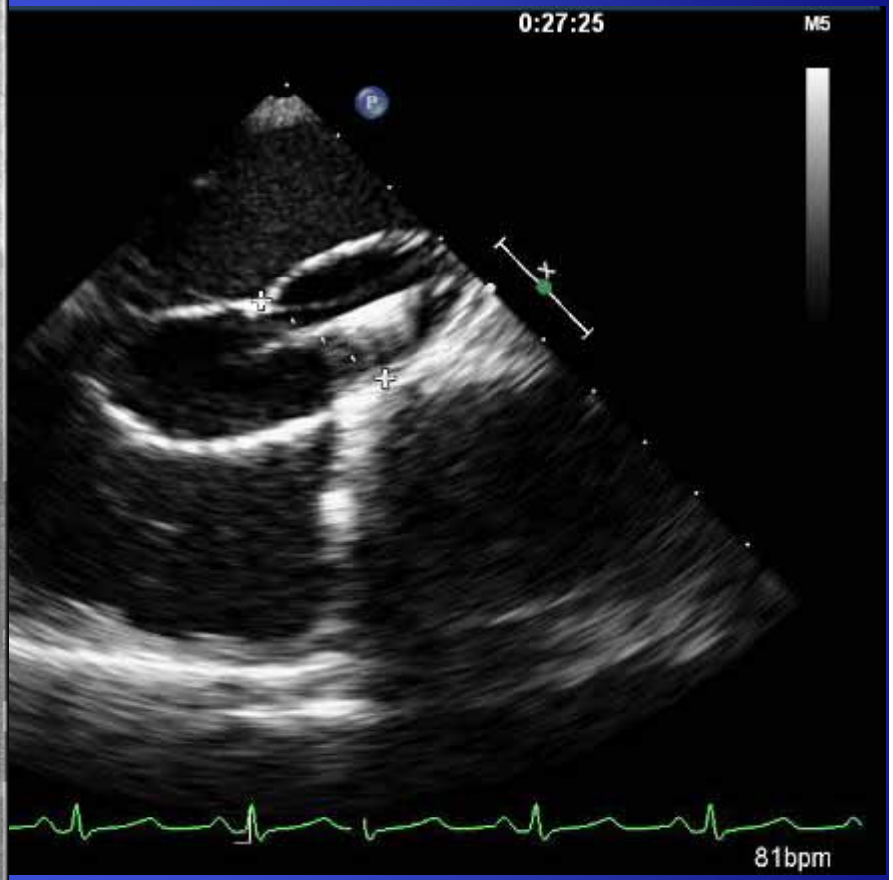
S. Schubert, MD, B. Peters, MD, H. Abdul-Khaliq, MD, PhD, N. Nagdyman, MD, P.E. Lange, MD, PhD, and P. Ewert,* MD, PhD

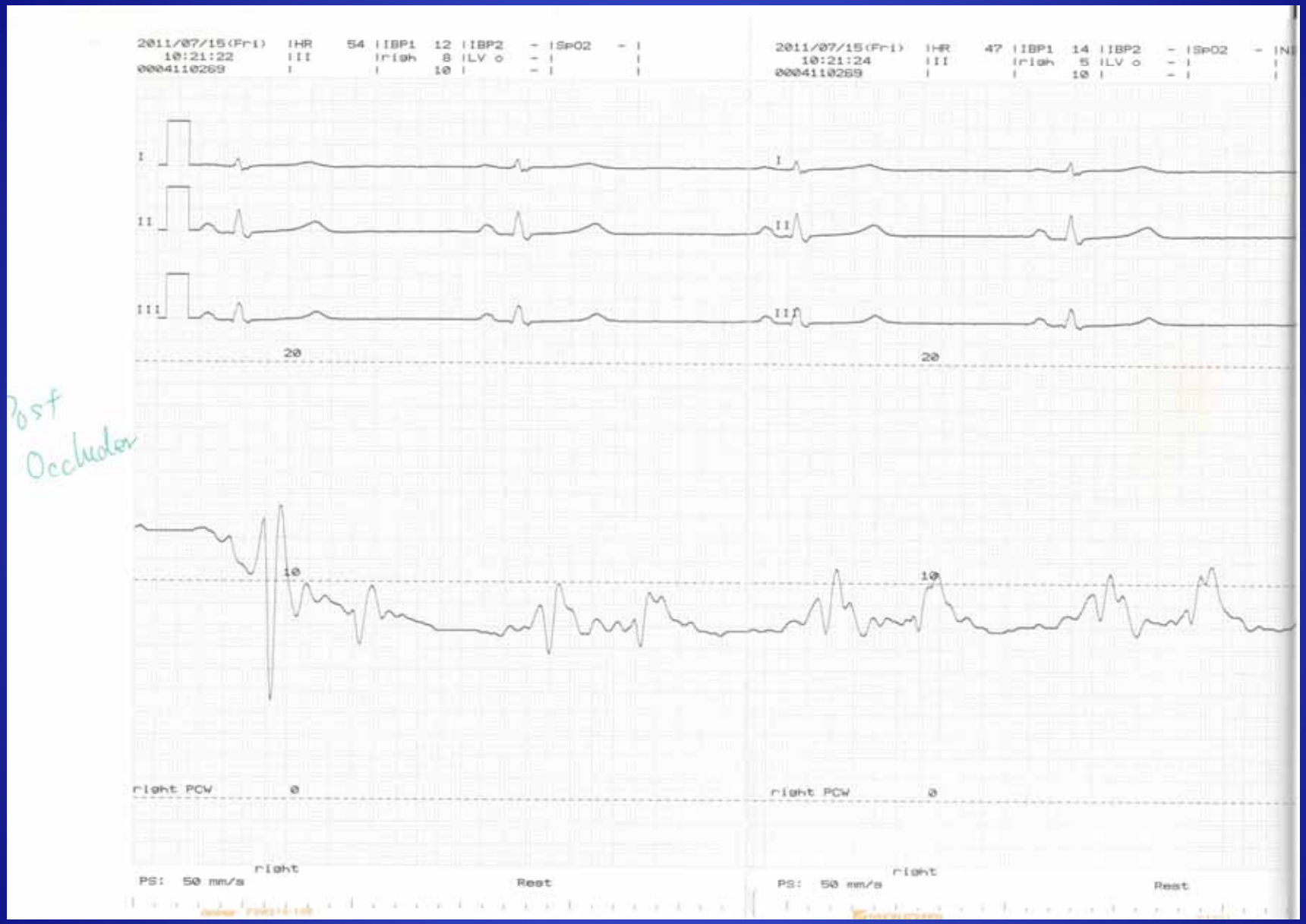
Pressure site	*P*
Left atrial pressur	<0.001[a]
Left atrial pressur	<0.001[a]
Right atrium (mea	0.05
Pulmonary artery	0.02[b]



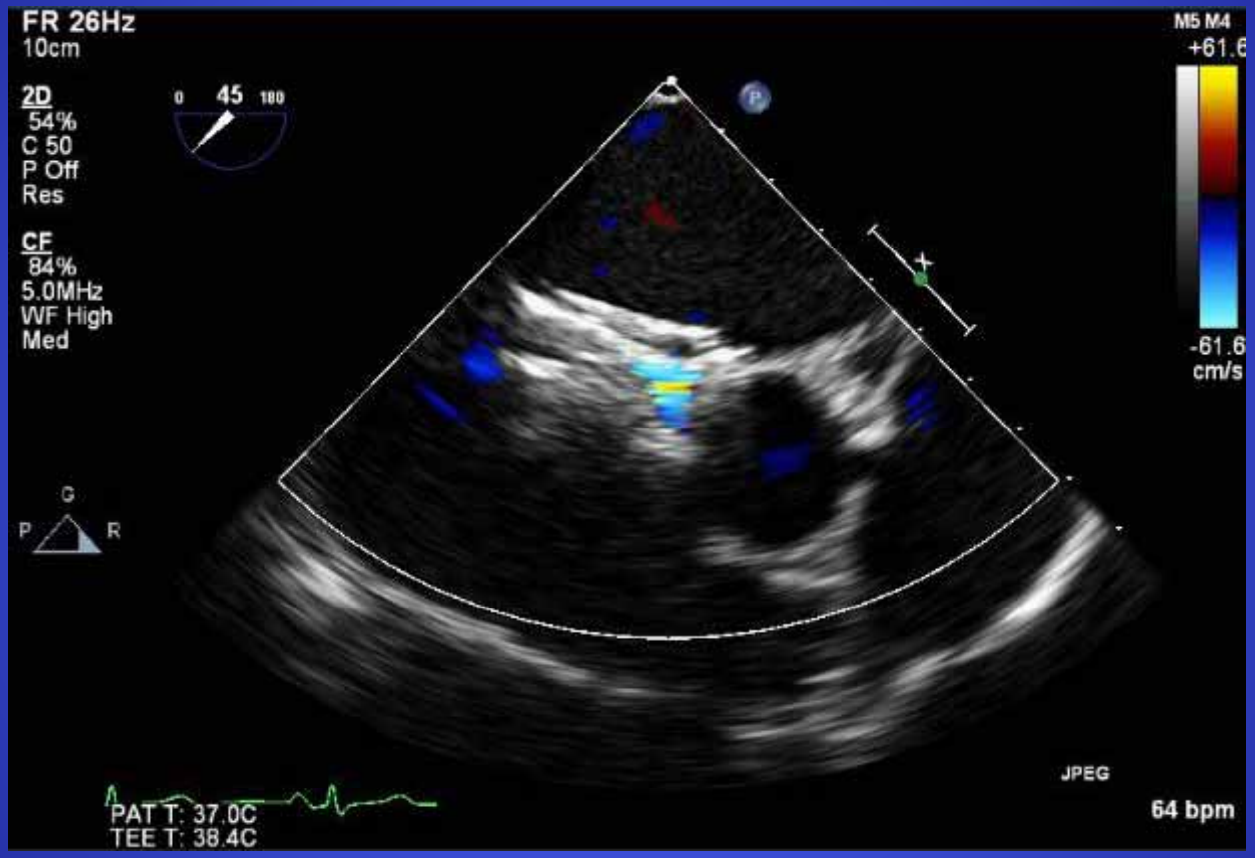
Catheterization and Cardiovascular Interventions 64:333–337 (2005)

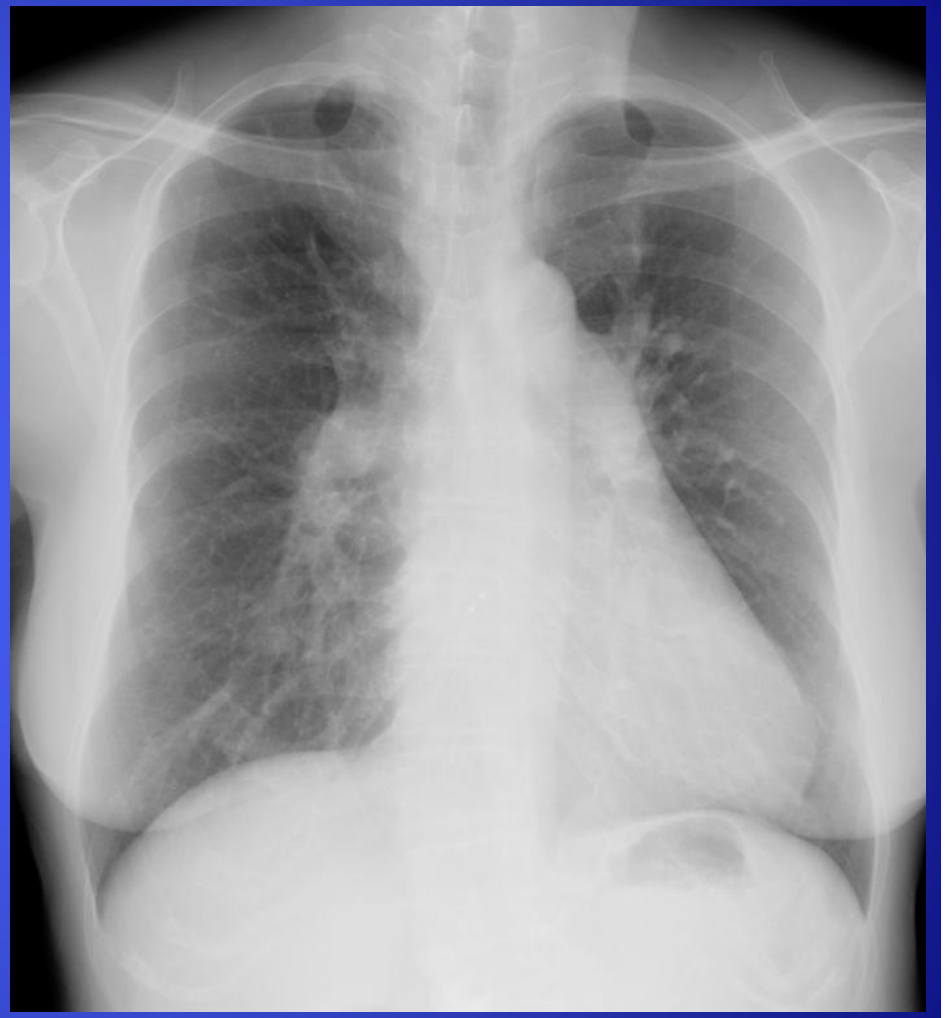



0:27:25
M5
81bpm



Post Occlusion


FR 26Hz
10cm
2D
54%
C 50
P Off
Res
CF
84%
5.0MHz
WF High
Med
0 45 180
M5 M4
+61.6
-61.6
cm/s
G
P
R
JPEG
64 bpm
PAT T: 37.0C
TEE T: 38.4C

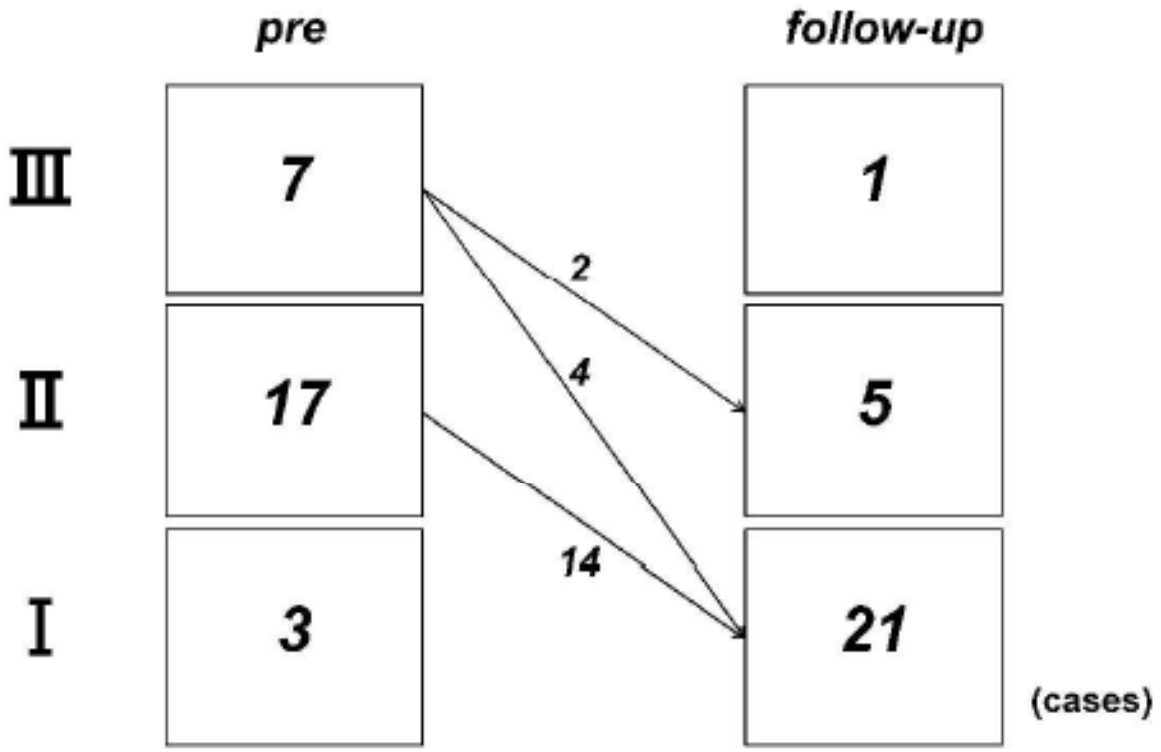


Before

24 hours after

項目名	結果	コメント	項目名	結果	コメント	項目名	結果	コメント
血液学的検査			APTT	30.0		H(ﾖｳｹﾂ)	0	
WBC	4.50		PT(%)	101		L(ｺﾝﾀﾞｸ)	0	
RBC	4.01		PT-INR	0.99		I(ｵｳﾀﾞﾝ)	1	
Hb	12.5		生化学的検査			内分泌的検査		
Ht	36.8		TP	7.8		BNP	106.2	
PLT	146		Alb	4.4		血漿蛋白		
MCV	91.7		T.Bil	1.09		CRP	0.13	
MCH	31.2		D.Bil	0.16				
MCHC	34.0		AST	30				
RDW	13.1		ALT	24				
Pct	0.120		ALP	259				
MPV	8.1		G-GT	23				
PDW	56.0		CHE	298				
HDW	2.62		LD	218				
WBCｿﾞｳ			A/G	1.29				
Ly	29.9		Na	140				
Mon	5.2		K	3.8				
Eos	2.4		Cl	107				
Bas	0.5		Mg	1.9				
NE	62.1		UN	14.4				
NE#A	2.8		CRTN	0.54				
LY#	1.3		UN/CRTN	26.7				
出血・凝固			CK	56				
PT	11.6		eGFR>=18	83.1				

NYHA functional class

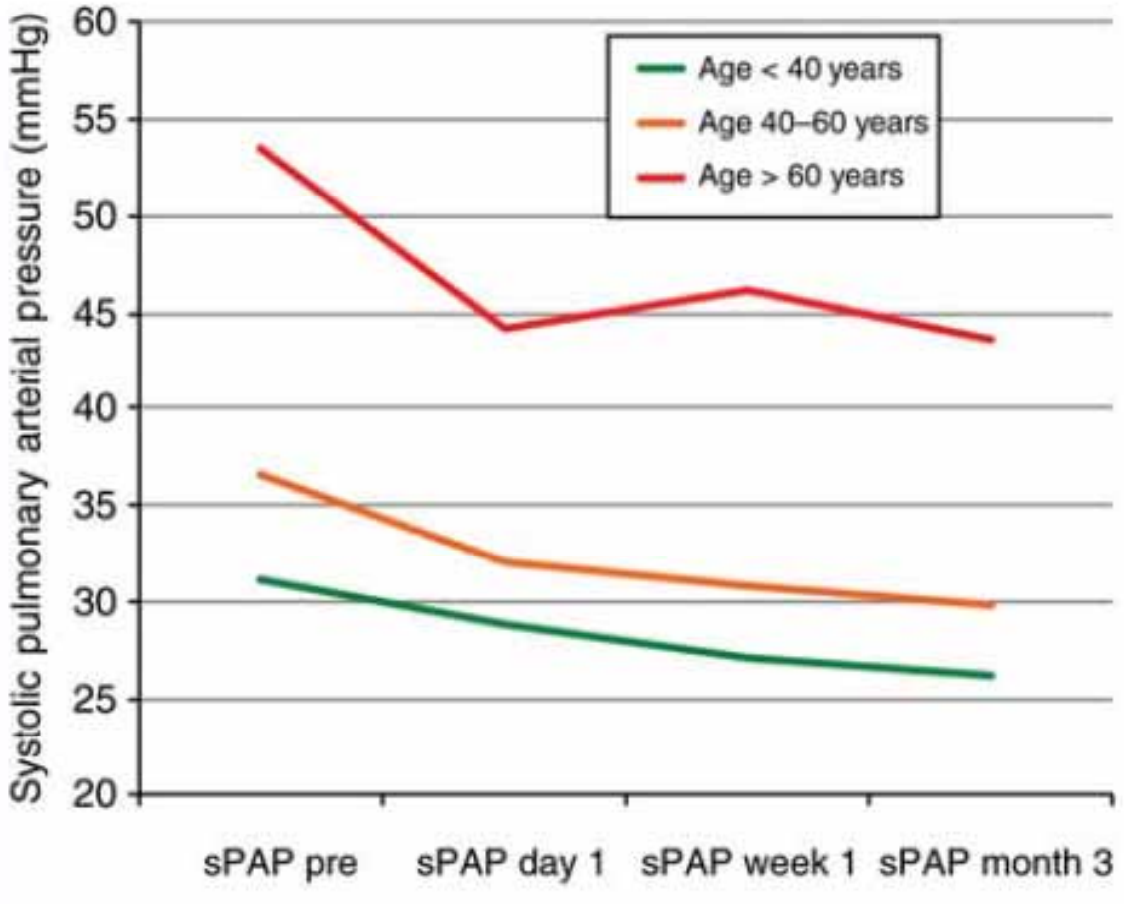


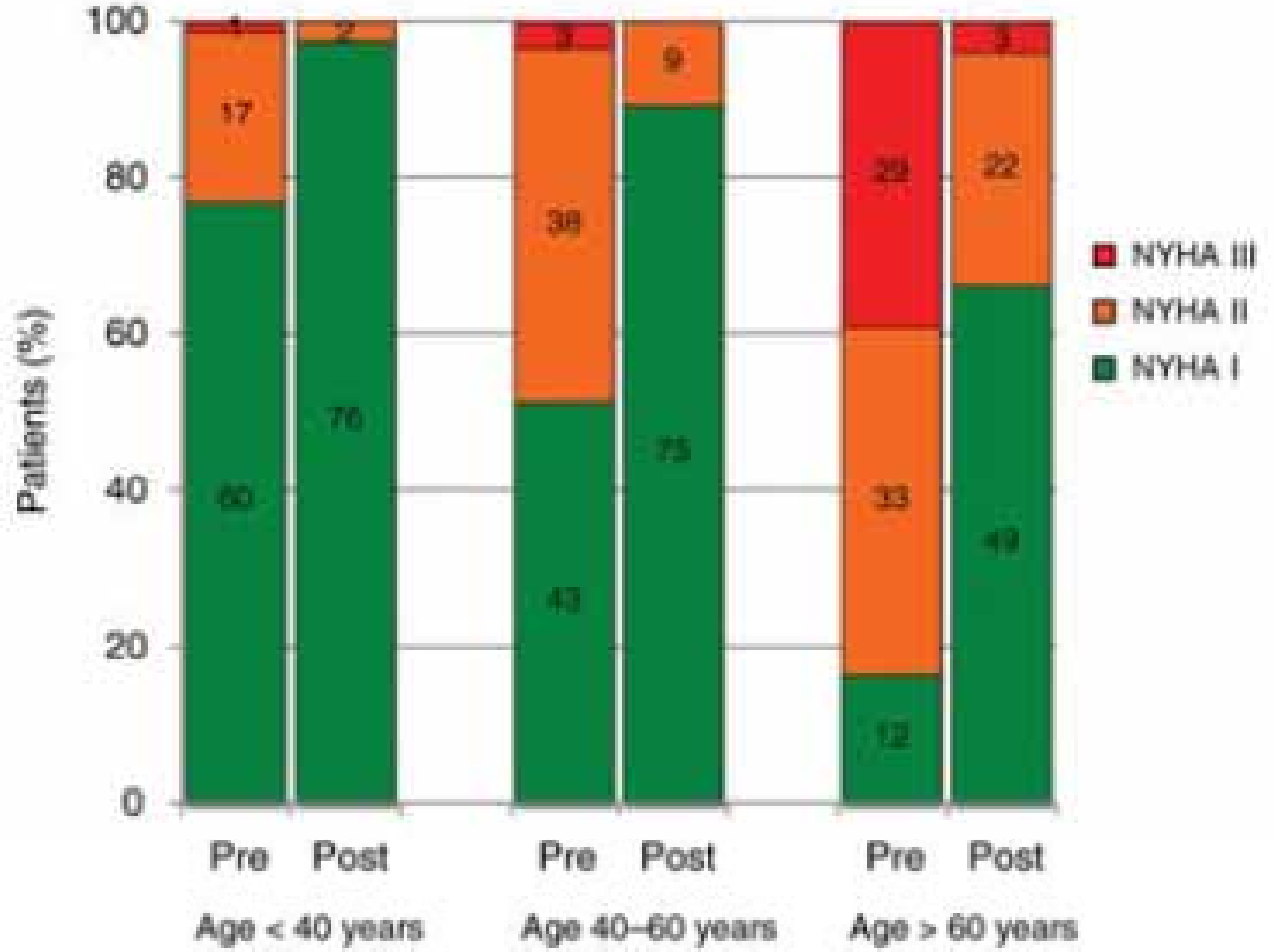
European Heart Journal (2011) **32**, 553–560
doi:10.1093/eurheartj/ehq352

CLINICAL RESEARCH
Congenital heart disease

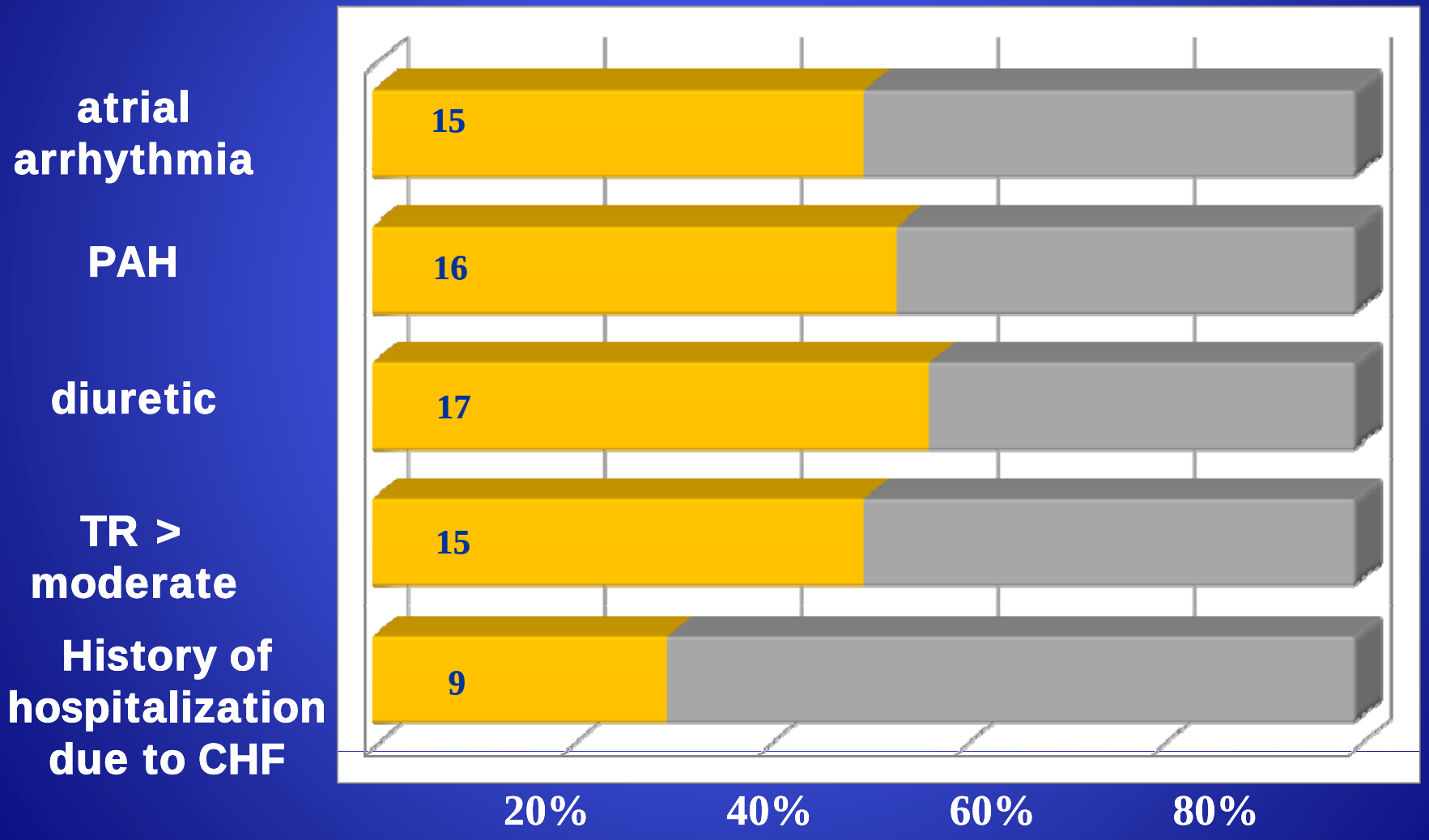
Benefit of atrial septal defect closure in adults: impact of age

Michael Hun **Rader**[1],
Maria Heger **leinze**[2],
Gerald Maur

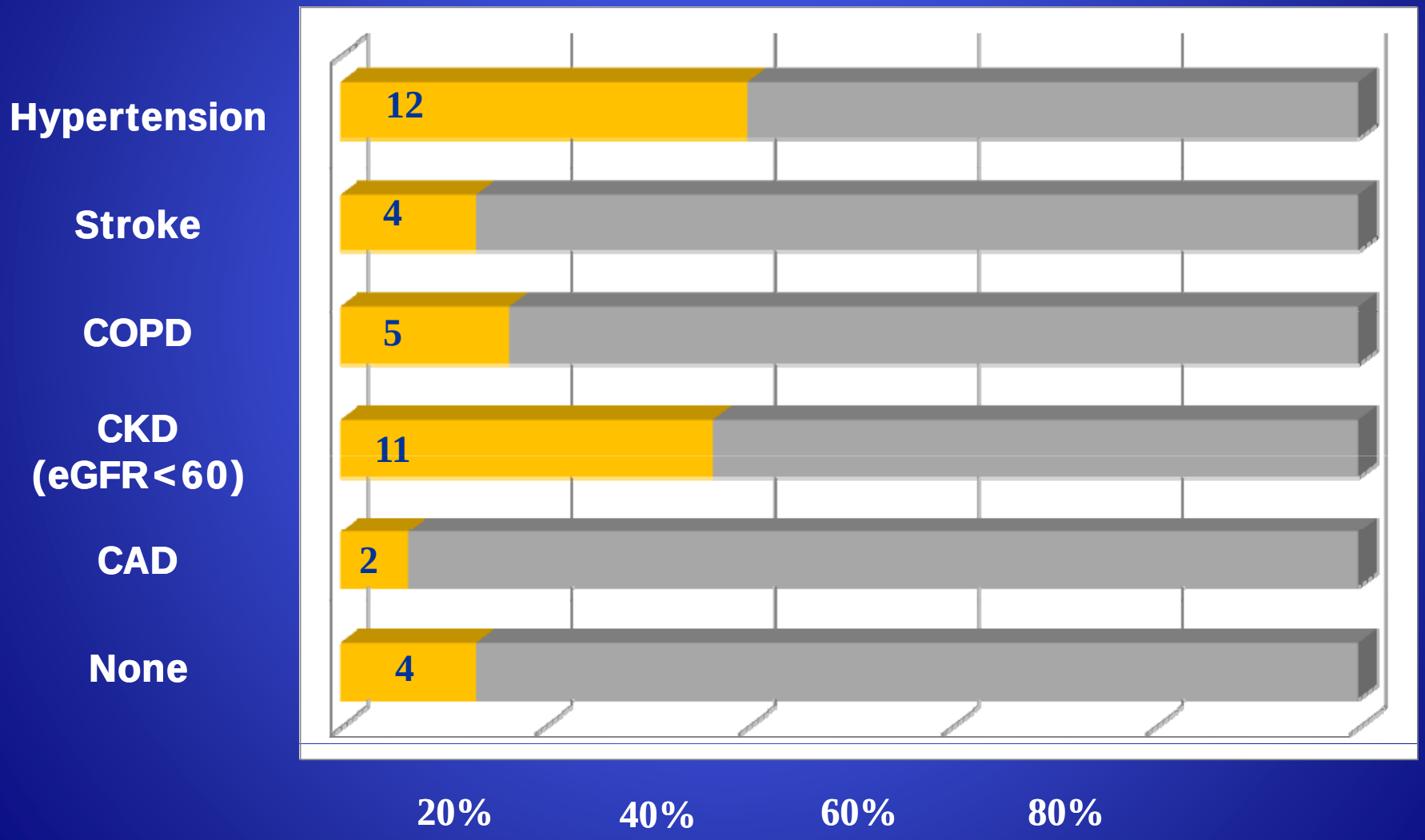



100
80
60
40
20
0
Patients (%)
17
60
2
76
38
43
9
75
29
33
12
22
49
3
NYHA III
NYHA II
NYHA I
Pre
Post
Pre
Post
Pre
Post
Age < 40 years
Age 40–60 years
Age > 60 years

高齢者ASDの合併症


atrial arrhythmia
15
PAH
16
diuretic
17
TR > moderate
15
History of hospitalization due to CHF
9
20%
40%
60%
80%

高齢者ASDの合併症


Hypertension
Stroke
COPD
CKD
(eGFR<60)
CAD
None
12
4
5
11
2
4
20%
40%
60%
80%