A therapeutic strategy for ASD patients with pulmonary hypertension or masked restriction

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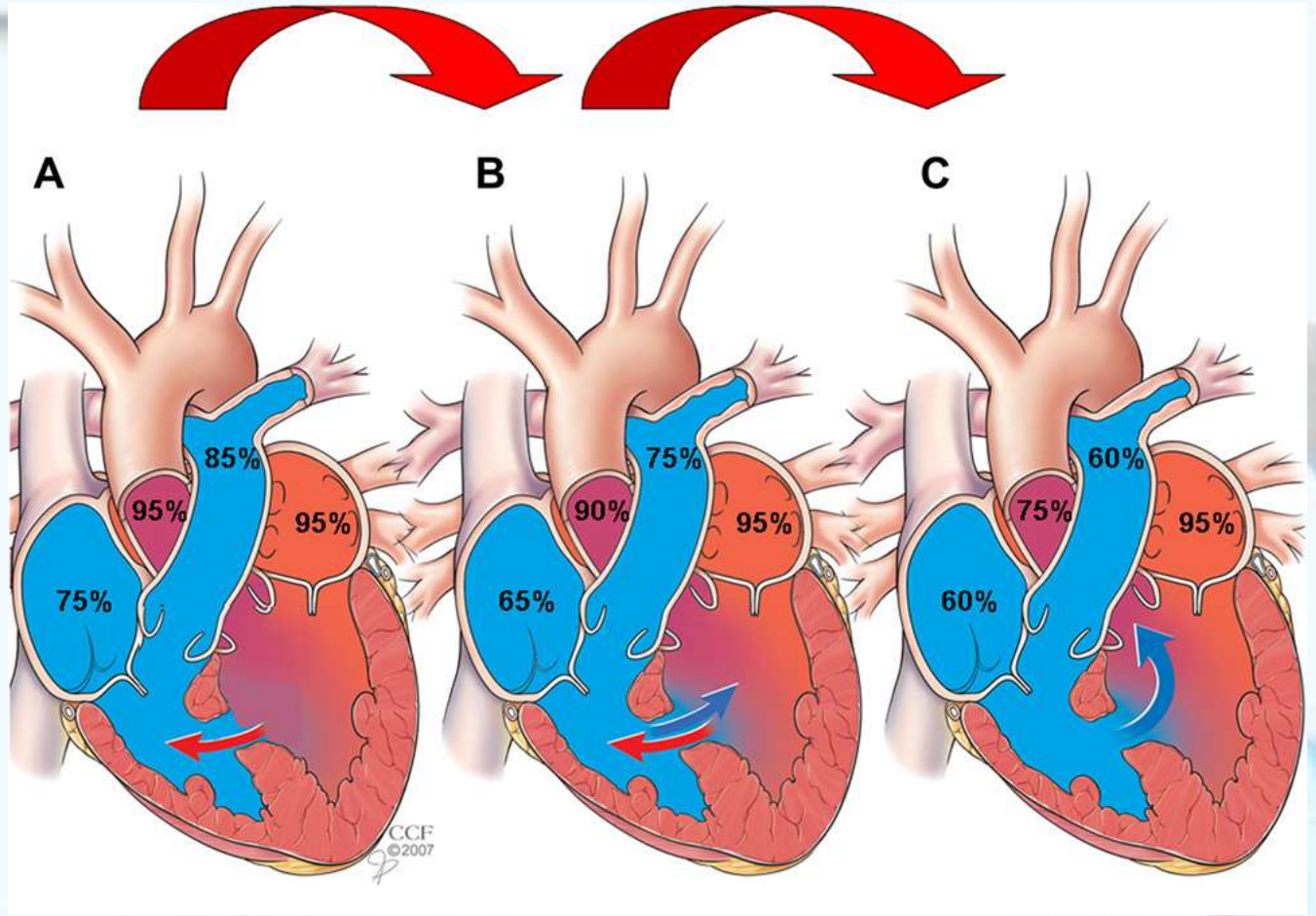
National Taiwan University Hospital
TCTAP 2015 April 30 7:20PM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©2007

Masked LV restriction in ASD patients

- Pulmonary edema secondary to LV dysfunction & increase in LA pressure after closure above 25 mmHg
- Elderly patients: 2-3.6%
- Detection: balloon test occlusion>> check PCWP & Doppler echocardiography

Swan L, Int J Cradiol 2006;107:207, Schubert S, CCI 2005;64:333,
Elshershari H J invasive Cardiol 2008;20:173
Ewert P, CCI 2001;52:177, Choi JY KCJ;2013;

ASD with LV failure

LV failure in ASD >> diastolic dysfunction most common,

- age, hypertension, LV hypertrophy, CAD, arrhythmias

ASD closure in patients with LV failure

- increase in LV filling, LV dimension, after ASD closure >> heart failure, pulmonary edema

Strategy of ASD closure in the presence of LV failure

- Periprocedural anticongestive medication; dopamine, diuretics, milrinone >>preconditioning
- Balloon test occlusion, check PCWP or LVEDP,LAP > 25 mmHg needing preconditioning
- Fenestrated device?

Severe PAH vs. Eisenmenger

- Safe upper limit of closure?

  PVR < 5 wood units >> safe

  *PVR <6 wood units,

  PAP/Ao < 2/3, PVR/SVR < 2/3, after vasodilator, net Qp/Qs > 1.5

  ESC guidelines Eur Heart J 2010 ; 31:2915

- Grey zone ?

Indications	Class[a]	Level[b]
Patients with significant shunt (signs of RV volume overload) and PVR <5 WU should undergo ASD closure regardless of symptoms	I	B[26]
Device closure is the method of choice for secundum ASD closure when applicable	I	C
All ASDs regardless of size in patients with suspicion of paradoxical embolism (exclusion of other causes) should be considered for intervention	IIa	C
Patients with PVR ≥5 WU but <2/3 SVR or PAP <2/3 systemic pressure (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy) and evidence of net L–R shunt (Qp:Qs >1.5) may be considered for intervention	IIb	C
ASD closure must be avoided in patients with Eisenmenger physiology	III	C

Evaluation of operability in shunt lesions with severe PAH

- Echocardiography
- PVR
- Test occlusion
- Vasodilators to test vasoreactivity
- Pulmonary wedge angiogram
- Biopsy?
- Exercise test
- Pretreatment with targeted therapies

ELSEVIER

International Journal of Cardiology 129 (2008) 163–171

International Journal of Cardiology

www.elsevier.com/locate/ijcard

Review

Evaluating operability in adults with congenital heart disease and the role of pretreatment with targeted pulmonary arterial hypertension therapy

Konstantinos Dimopoulos *, Ana Peset, Michael A. Gatzoulis

Adult Congenital Heart Centre and Centre for Pulmonary Hypertension, Royal Brompton Hospital and National Heart & Lung Institute, Imperial College School of Medicine, London, United Kingdom

Received 19 February 2008; accepted 22 February 2008
Available online 26 March 2008

Table 1

Evaluating operability of cardiac defects in adults with CHD and PAH

For	Against
– Abort right-to-left shunting	– Potential conversion of Eisenmenger physiology to iPAH physiology (and thus worse long-term outcome)
– ↓ Cerebrovascular events (stroke /abscess)	
– Prevent cyanosis	
↑ Exercise capacity	– High perioperative risk
↓ Erythrocytosis	
↓ Hemostatic problems	– Very limited experience and no long-term data available
↓ Systemic organ failure	
– Protect pulmonary circulation	

CHD = Congenital heart disease, PAH = pulmonary arterial hypertension, iPAH = idiopathic pulmonary arterial hypertension.

Optimal cut-off value for determining operability

- Rp/ Rs < 0.42 with O2 alone
- Rp/ Rs < 0.27 with O2 + NO

Dimopoulous et al. Int J Cardiol 2008;129:163

Acute vasodilator testing

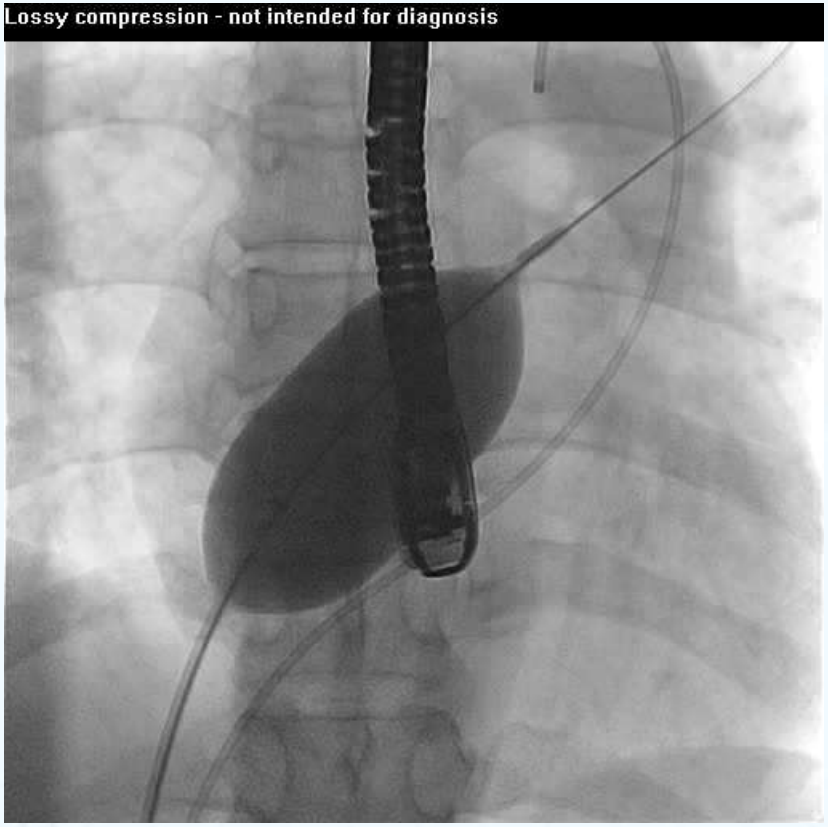
- O_2 100 % , NO, iloprost
- reduction of mean PAP by at least 10 mmHg to a value < 40 mmHg with an increase or unchanged cardiac index
- iloprost: reduction PVR > 20 % & PVR index < 11 w.u. m^2

Douwes JM, EHJ 2011;32:3137

Hill KD, CCI 2010;76:865

Gorenflo Cardiology 2010;116:10

Balloon test occlusion



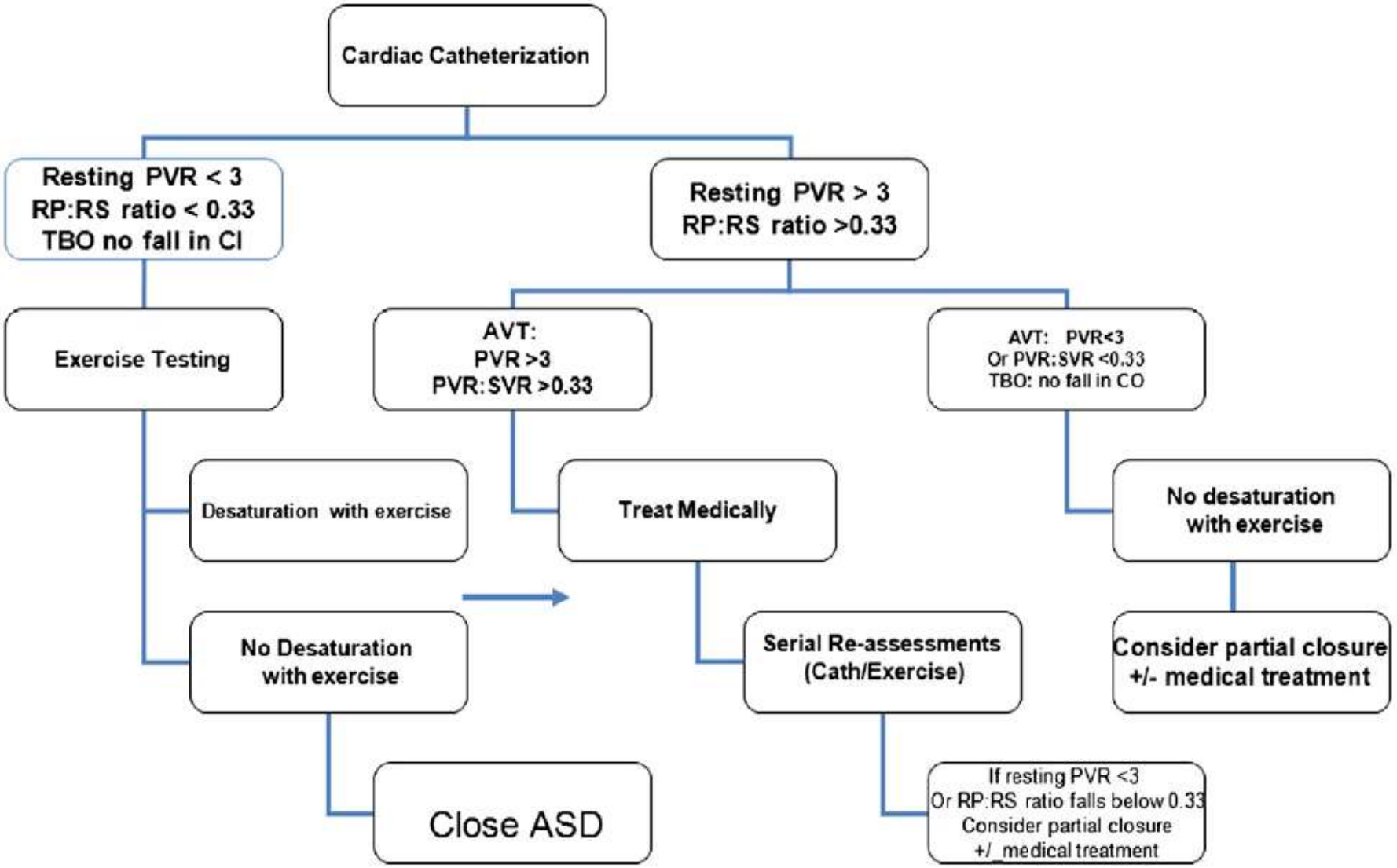


Fig. 1. CHD/PH (ASD) Clinical management algorithm: Individualized case approach.

Can "Inoperable" Congenital Heart Defects Become Operable in Patients with Pulmonary Arterial Hypertension? Dream or Reality?

Maurice Beghetti, MD,* Nazzareno Galiè, MD,† and Damien Bonnet, MD‡

*Pediatric Cardiology Unit, Department of the Child and Adolescent, Children's University Hospital, Geneva, Switzerland; †Institute of Cardiology, University of Bologna, Bologna, Italy; ‡French Reference Center for Complex Congenital Heart Diseases, Necker Hospital, University Paris Descartes, Paris, France

Strategy for closing ASDs with PAH

- Treat-and-repair strategy
- partial repair using fenestrated device followed by targeted therapy

PA wedge angiogram

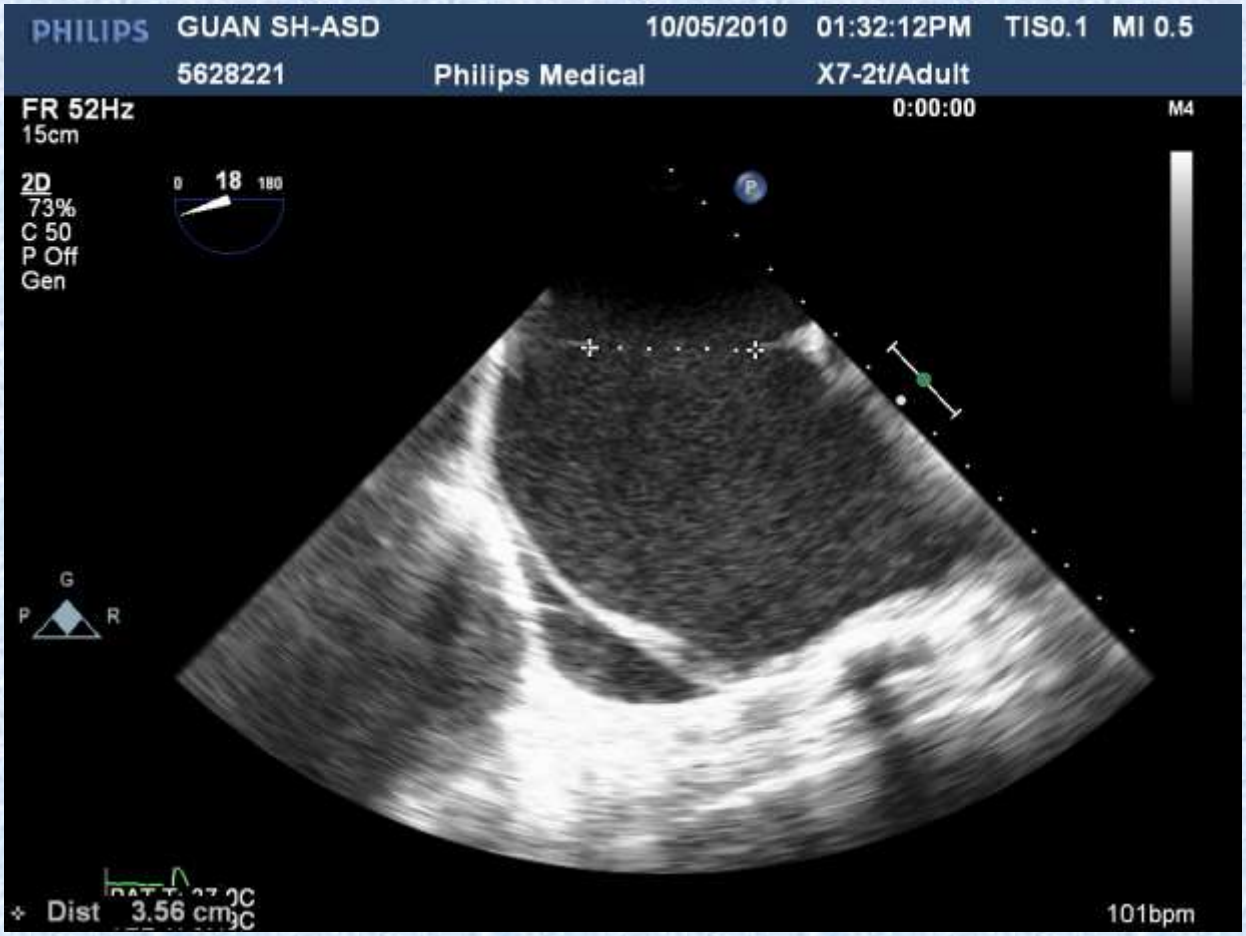
Case 2 Hemodynamics

	O_2 saturation(%)	pressure (mmHg)
SVC	56	
RA	70	mean 11
IVC	66	
RV	65	104/12
MPA	65	105/31 mean 61
LA	83	mean 12
RPV	92	

Qp/Qs = 1.28 PVR 10.8 WOOD UNIT

67 y/o F, NYHA III-IV

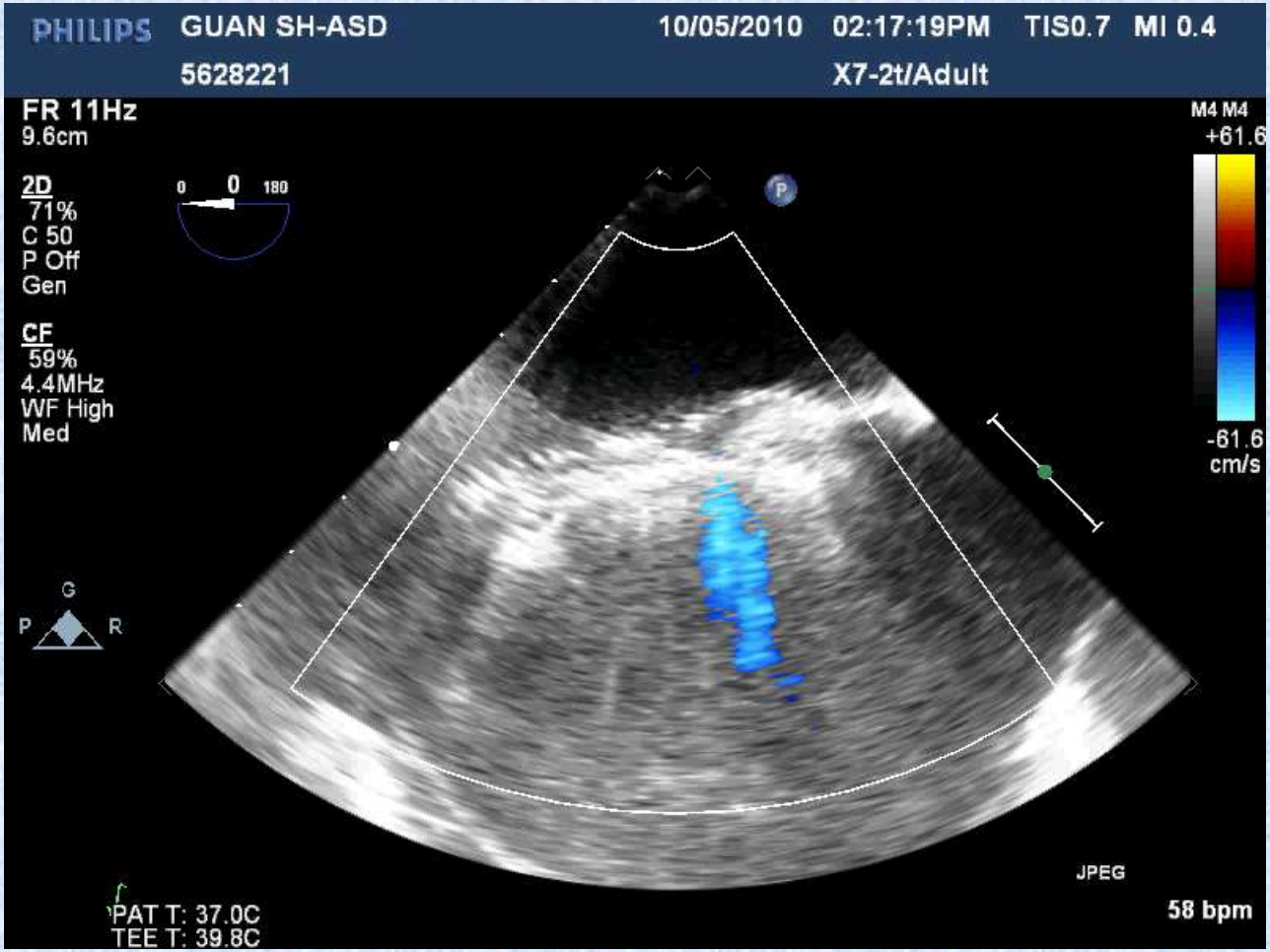
a large defect measured 35.6mm



Large ASD PAH

PA angio

Immediately after closure



Fenestrated device indications

- presence of severe pulmonary hypertension (mean PAP > 35 mmHg or PA systolic pressure > 60 mmHg)
- presence of heart failure (right or left)
- Elderly (> 80 years) or those with unpredictable risks for catheter closure

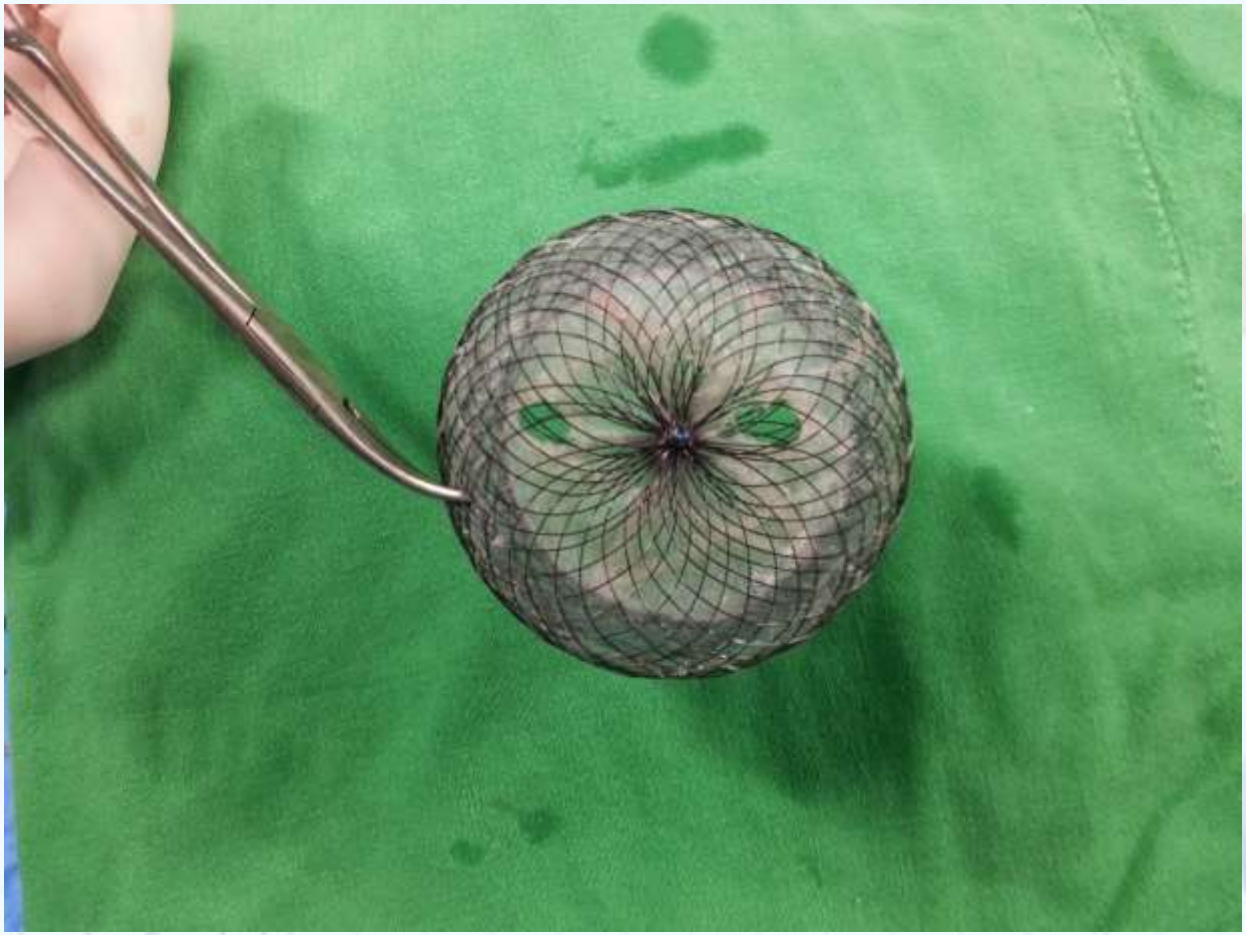
Exclusion criteria

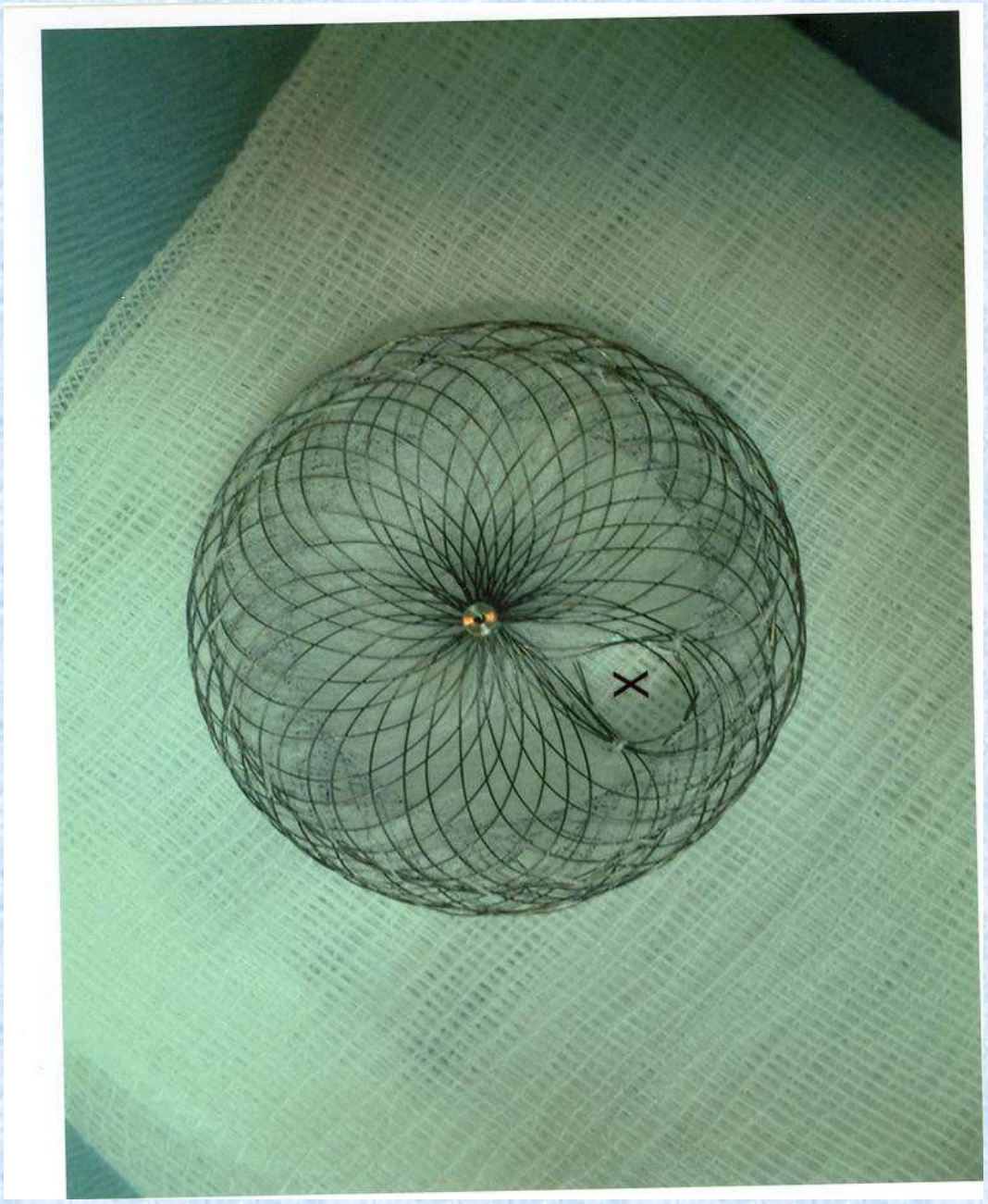
- O_2 saturation < 90%
- Pulmonary artery wedge angiogram: severe & diffuse obliteration of pulmonary arterioles (subjective)
- Multi-fenestrated defects
- Severe COPD
- PA resistance >12 Wood units

Methods

- Test balloon occlusion was performed after hemodynamic study.
- A self-fabricated fenestration 1/3- 1/4 of the diameter of Amplatzer septal occluder was created.
- In some occasions, two fenestrations were created.
- In 3 patients, a customized device from AGA was used.

Two fenestrations

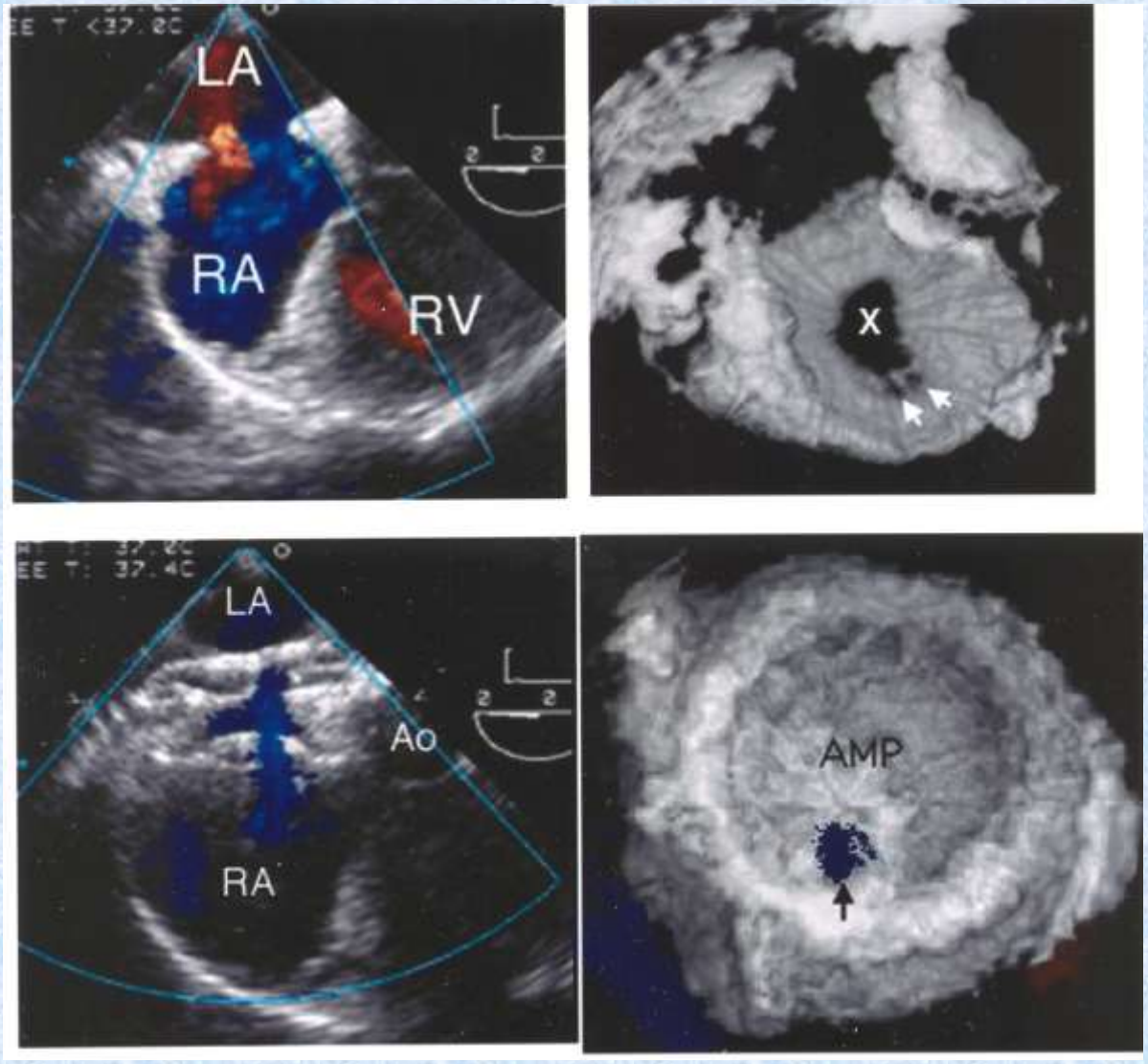




Patients

- 11.5-year period, 52 PAH patients (15 males & 37 females) with ages 7 - 81 years (mean 45 ± 20 years) underwent transcatheter closure of ASD with a fenestrated device.

- The indications of using a fenestrated device were moderate-to-severe pulmonary hypertension, (mean PAP> 35mmHg, PASP > 60 mmHg) n=48, with associated heart failure n=4,


LA
RA
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LA
Ao
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AMP

Results

- Hemodynamic data: mean Qp/Qs ratio was 2.7±1.4 & pulmonary artery systolic pressure was 66 ± 18 mmHg, median 65 mmHg, mean PAP 42 ± 15 mmHg .

- The mean device diameter used was 30 ±6 mm. (median 30 mm)

Results II

- Implantation was initially successful in all 52 patients.
- One patient who had CHF & AF developed significant increase in central venous pressure (13 → 24 mmHg) after device implantation & distal embolization of the device occurred several hours later.

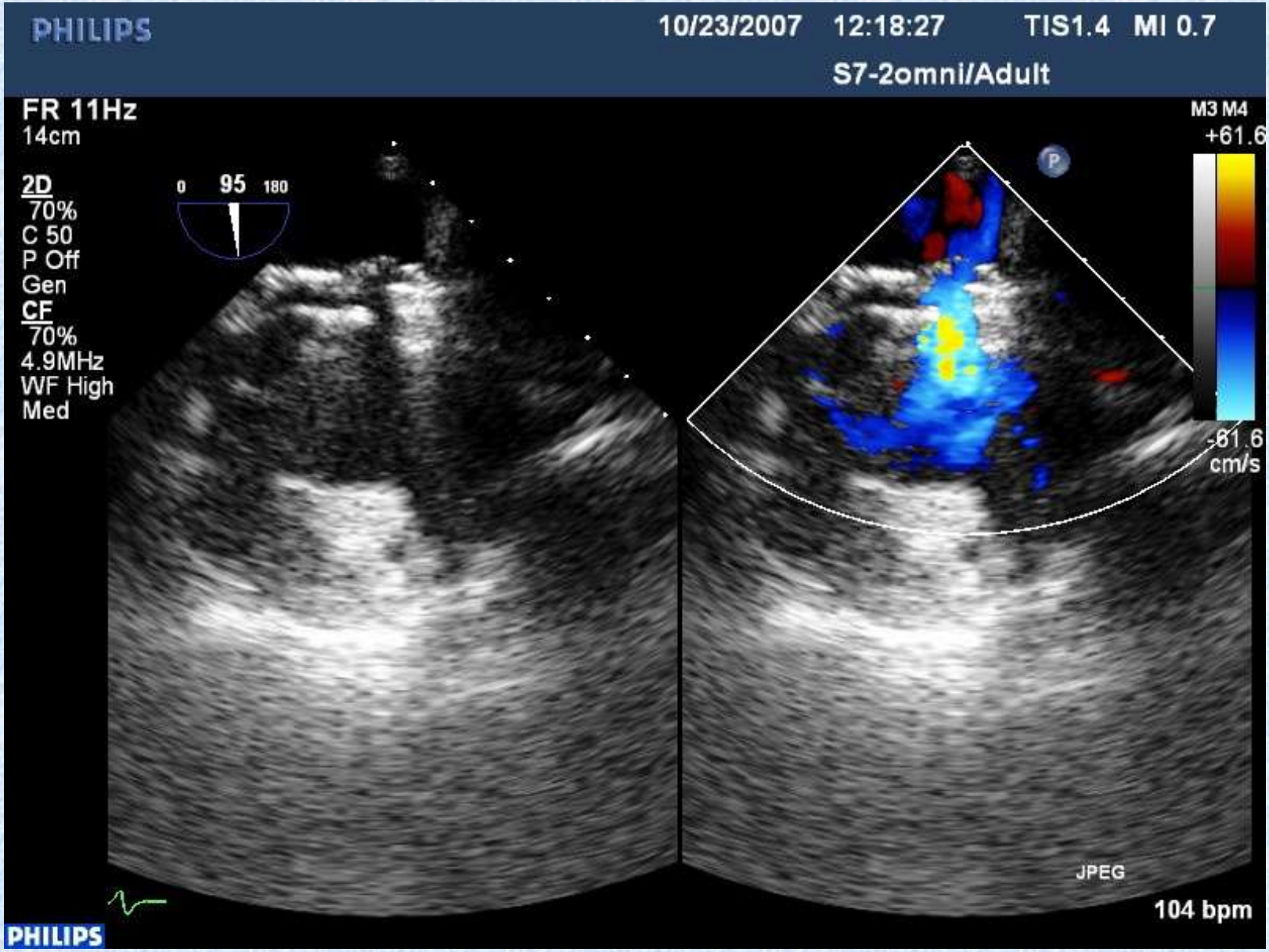
Follow up

- All except 1received at least 6 months follow-up, mean f/u period 46 ± 22 months.
- 31 took Sildenafil 20-25 mg bid or tid, 2 received Bosentan.
- 3 underwent a second procedure to close the fenestration because presence of significant shunt (Qp/ Qs > 1.5) & cardiomegaly
- The mean PASP estimated by echo was 45 ± 19 mmHg at the latest F/U vs. preclosure measured by cath 66 ± 18 mmHg (p< 0.01)

.

Fate of the fenestration

- Spontaneous closure in majority of cases
- A second device was deployed in 3 cases because of Qp/Qs ratio > 1.5. All 3 patients received implantation with a custom-made fenestrated device (7 mm fenestration).
- persisted small shunt in 7 patients.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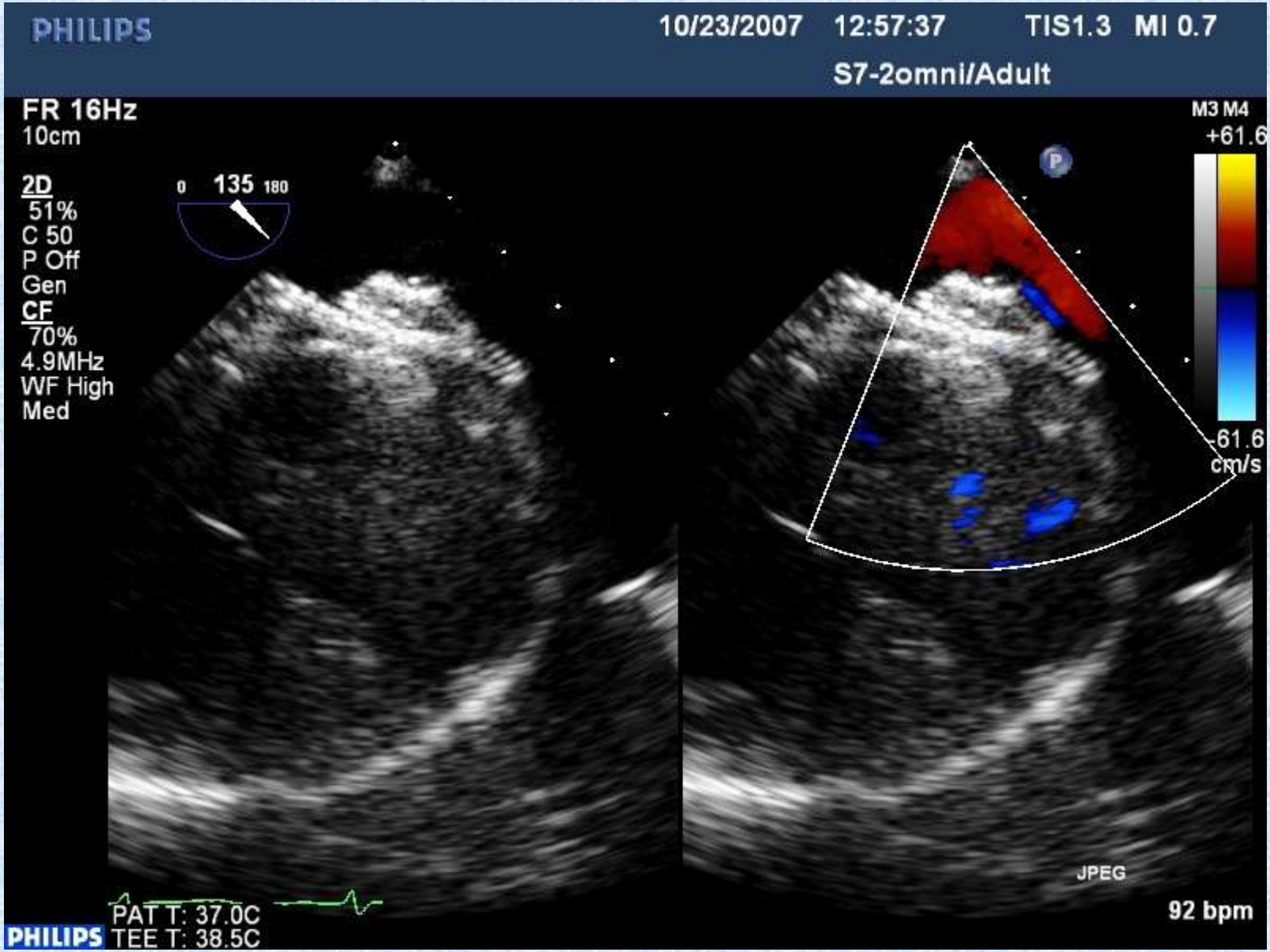
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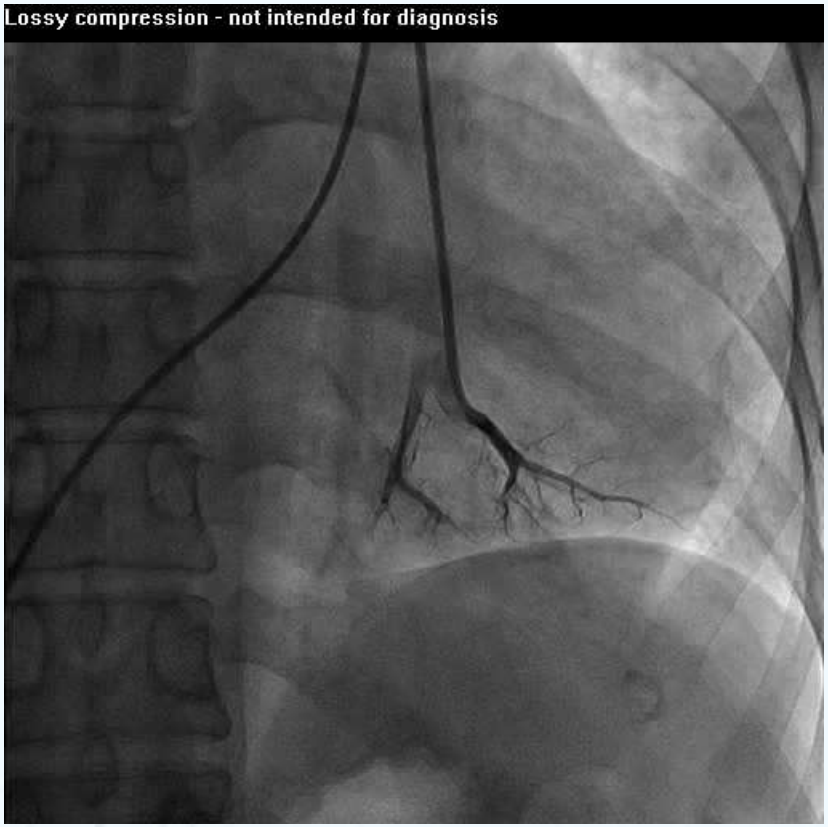
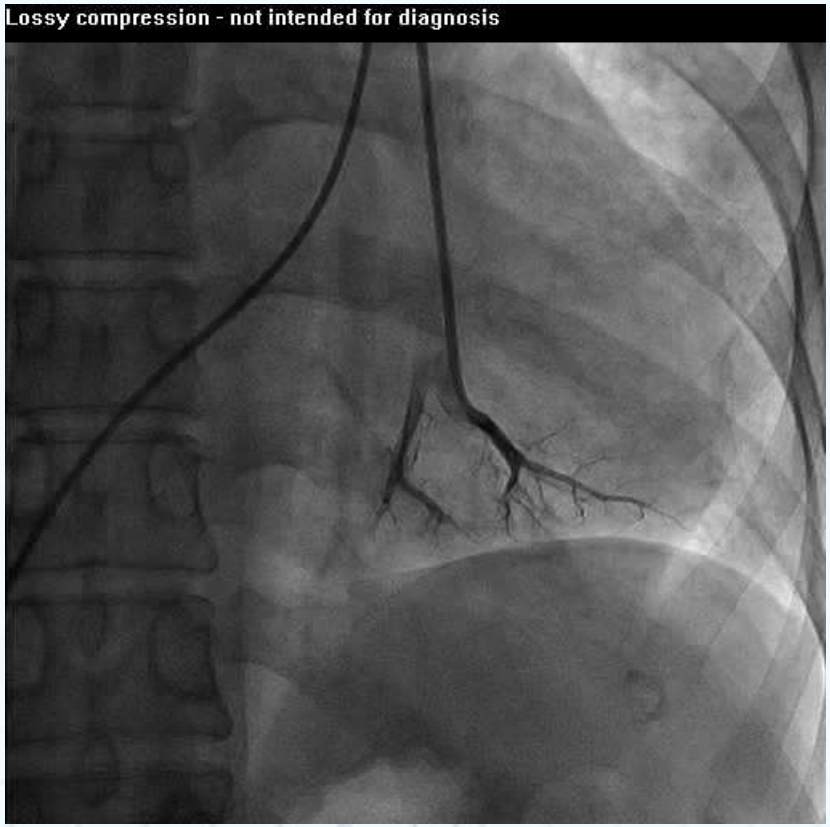
Is PA wedge angiogram useful in predicting operability?



Wedge angio useful?

60/39 (39) PVR 3.79 WU



97/33 (55) PVR 9.81 WU

Advantages of creating fenestrations

- allows for gradual reduction in left-to-right shunt
- avoids sudden changes in hemodynamics in compromised patients

Kretschmar et. Al. CCI 2010;76:564-71
Schneider et.at. J Interv Cardiol 2011;24:485-90

Disadvantages of fenestration

- residual shunt, small enough?

- need for closure if significant shunt

  Qp/Qs ≧ 1.5 is present without severe pulmonary hypertension

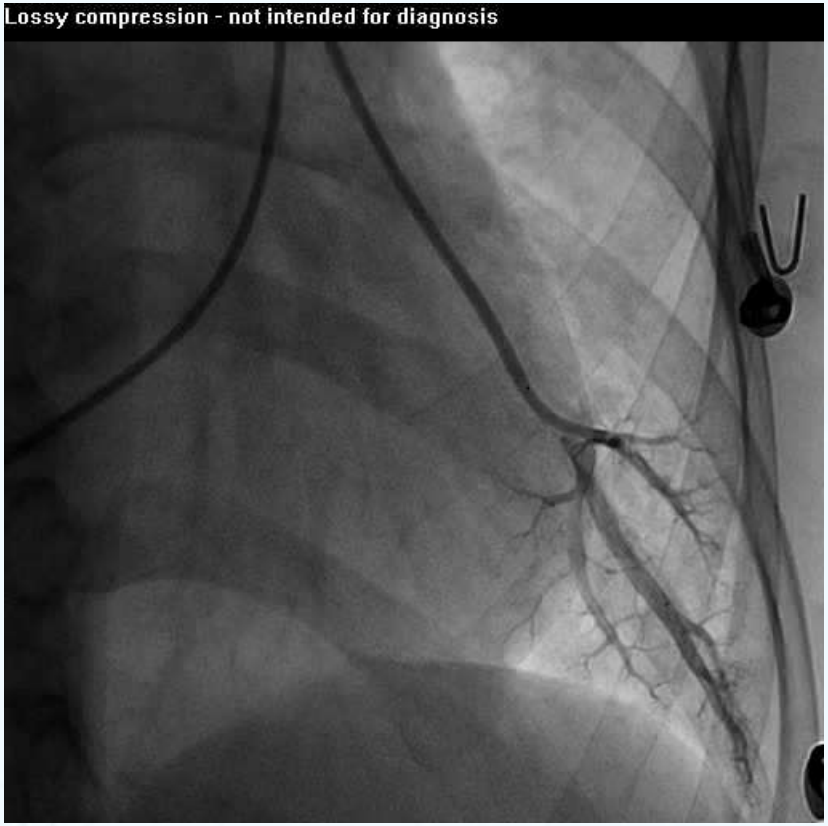
Discussions

- In patients with ASD with PAH, closure can be performed by using a fenestrated device after evaluations.
- Spontaneous closure of fenestrations occurred in majorities of patients at follow up.
- 3 patients underwent a second intervention to close the fenestration because persistence of significant shunt, too large fenestration (7 mm)?

Conclusions

- Fenestrated device is an ideal device to use in ASD patients with severe pulmonary hypertension or heart failure. It is safe & beneficial in most cases, if the pulmonary vascular resistance is not too high. Balloon occlusion is helpful in unmasking restricted LV function.
- Targeted therapy is needed for those with persisted pulmonary hypertension.
- Long-term follow-up is mandatory.

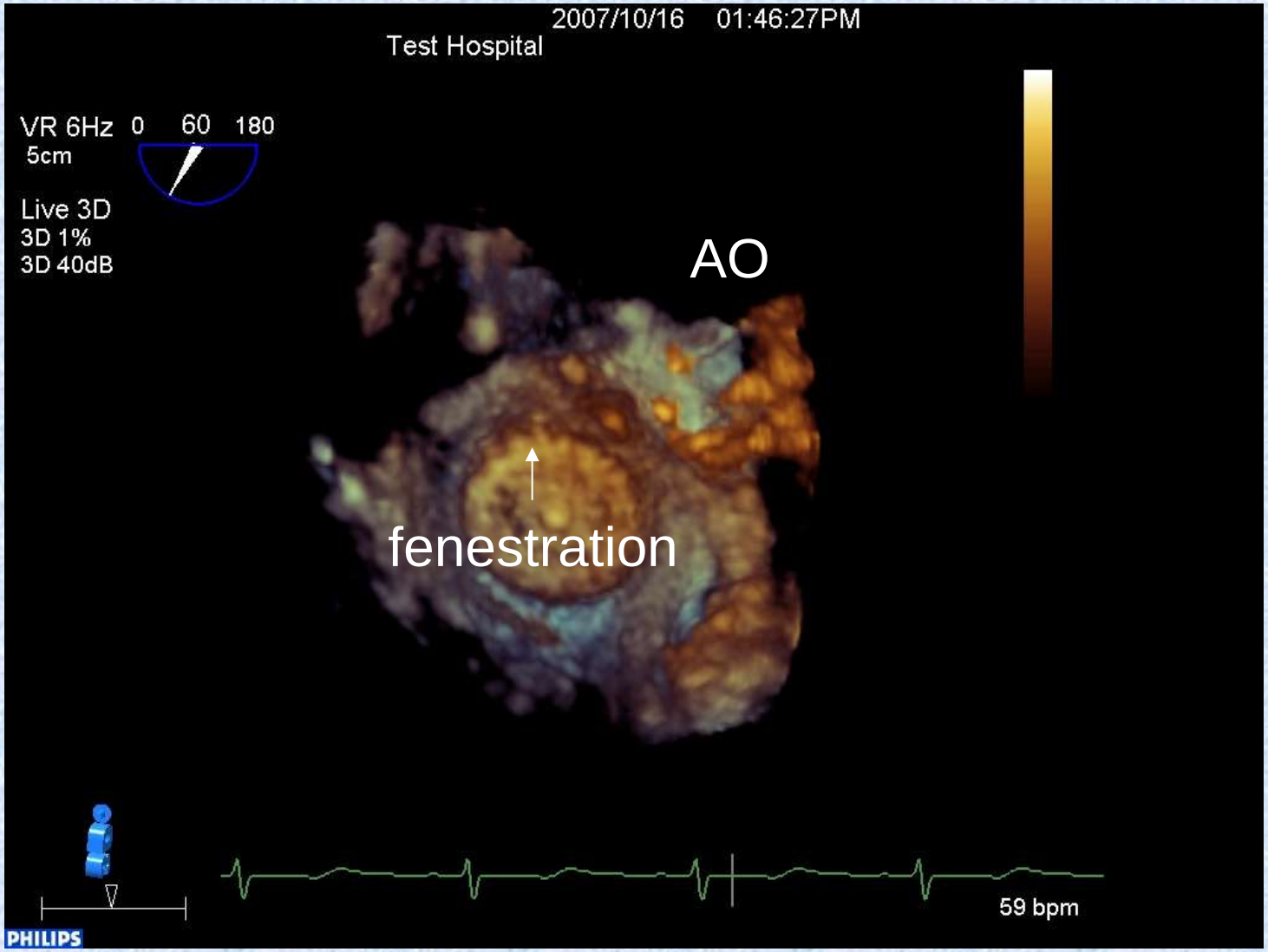
Wedge angio 7.13WU, 37 y, F



Home-made fenestration

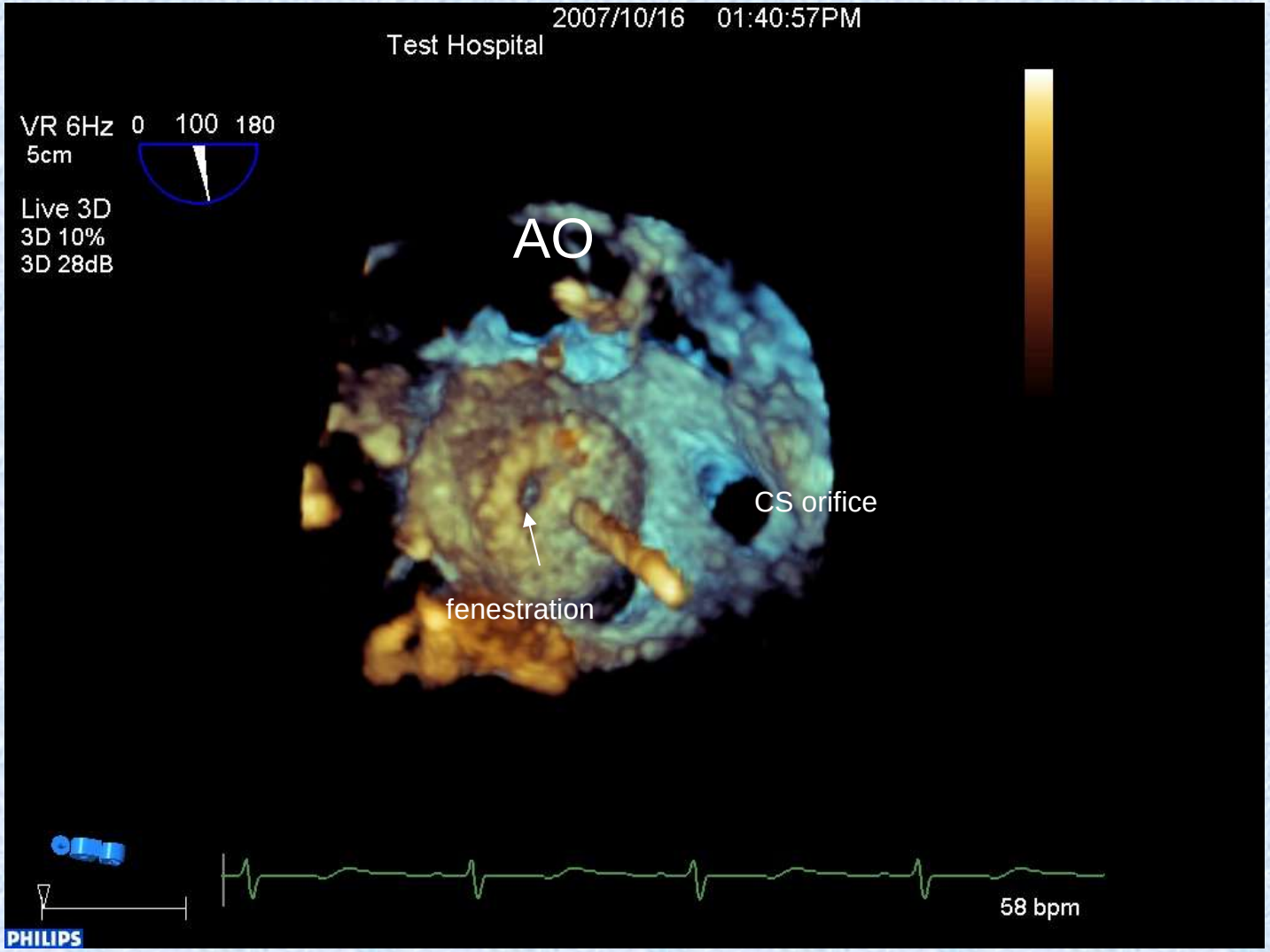
- create a defect around 1/3 -1/4 of the diameter of device eg 7 mm fenestration in a 28 mm Amplatzer septal occluder

- create 2 smaller defects

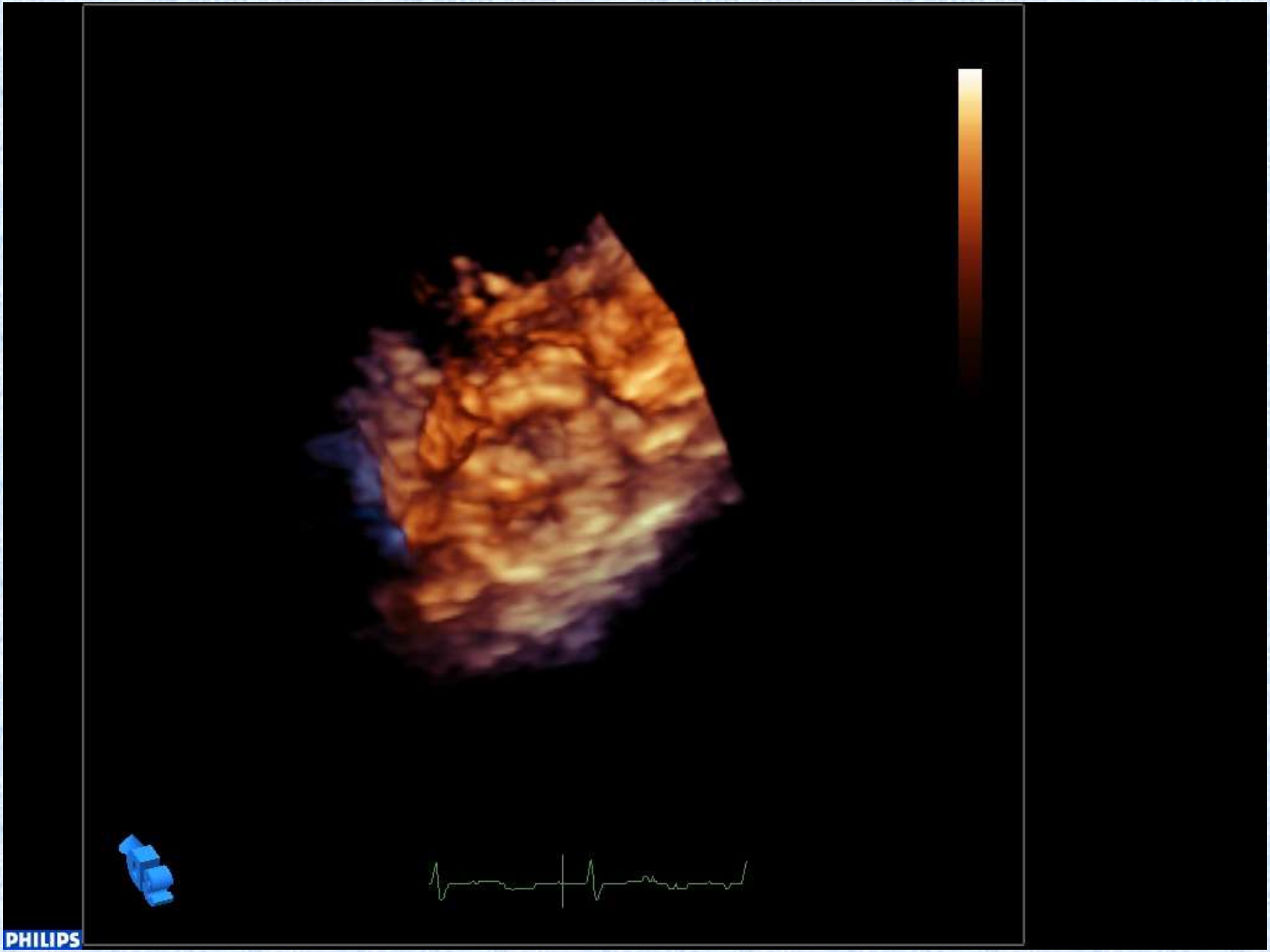


3D image post detachment of occluder, RA side

28# Fenetrated Amplatzer



Before detachment of 2nd occluder, view from RA side


PHILIPS

Outcomes of closure in ASD patients with PAH

(ECHO)	PASP (mmHg)	Reduction ≧ 5 mmHg	Normalization of PAP
No PAH	＜ 40	33.7 %	90.2 %
Mild	40-49	73.9 %	71.7 %
Moderate	50-59	79.2%	66.7 %
Severe	≧60	100 %	23.5 %

Yong et al.
Cir Cardiovascular Intervent 2009;2:455

PAH with systemic-to-pulmonary shunts: close or not to close

Kaplan-Meier survival estimates a 20 years

ES	87% (77-93%)	n = 90	ASD 11 %
PAH-shunt	86% (60-96 %)	n = 48	ASD 46 %
PAH after defect correction	36 % (12-72 %)	n = 44	ASD 27 %

Manes et al. Eur Heart J 2014;35:716-24

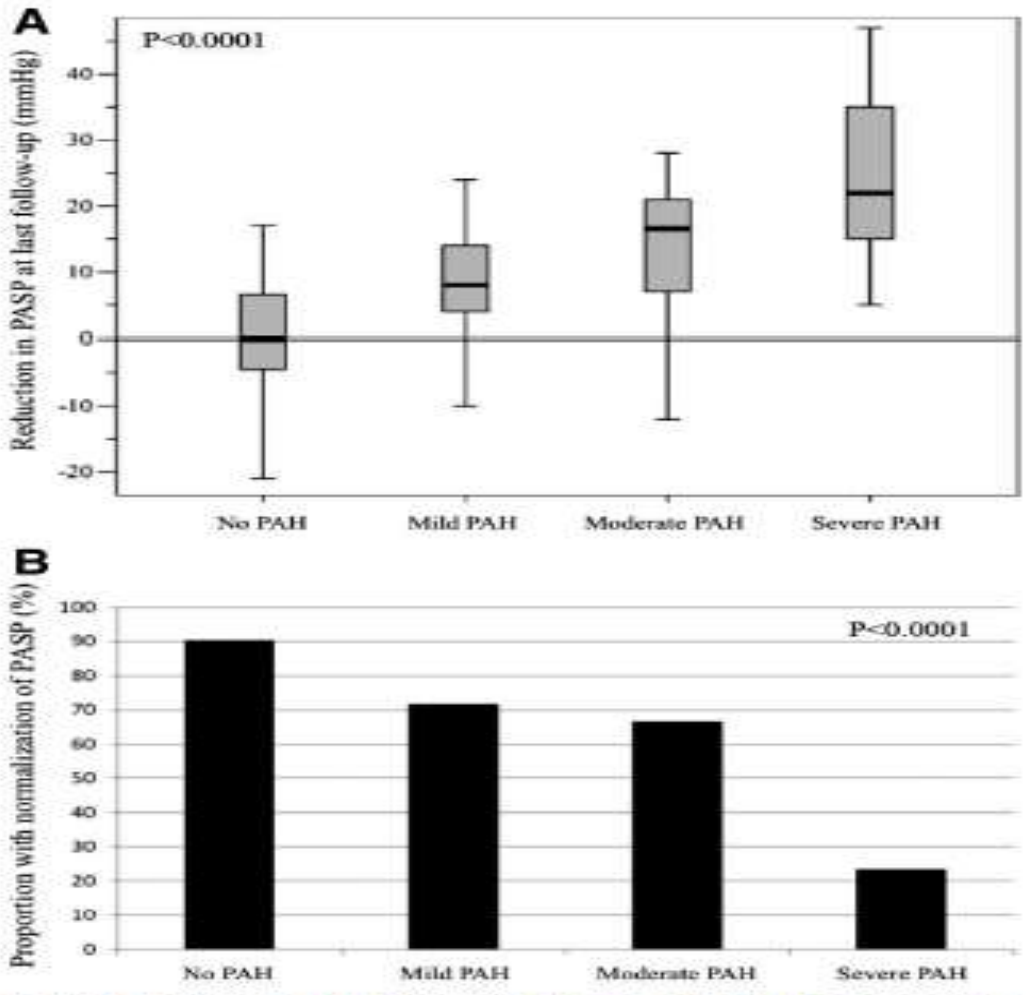
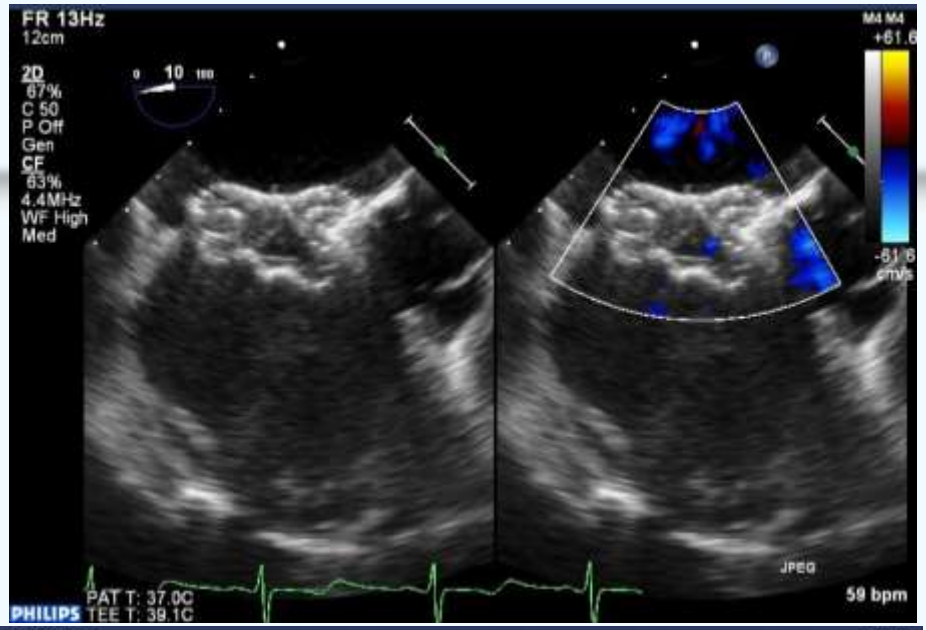
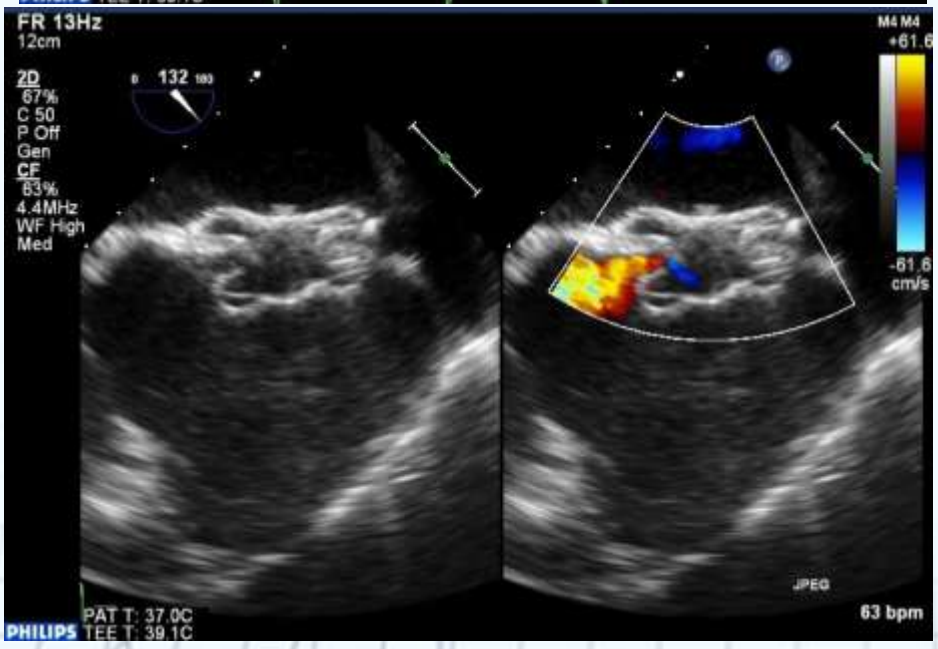


Figure 2. Reduction in PASP (A) and normalization of pressures (B) after transcatheter closure of ASD. A, Degree of reduction in PASP after transcatheter ASD closure according to whether patients had no, mild, moderate, or severe PAH at baseline. B, Proportion of patients with normalization of PASP (<40 mm Hg) after ASD closure, as stratified by the baseline degree of PAH.

Cir Cardiovascular Intervent 2009;2:455 Yong et al





ASD post occlusion

complications

- Embolization in 1, cause, malfunction of self fabricated device? elevation in CVP to 24 mmHg. Emergent operation including a Maze procedure was performed.

- Tachyarrhythmia n=4, cardioversion n=2

- No other serious complications

28# Fenetrated Amplatzer

Treatment for L-R shunt lesions with severe PAH

- Corrective interventions in patients with L-R shunt & severe PAH are generally contraindicated.
- Borderline cases: pulmonary vasoactivity test
- Partial repair & targeted therapy

Aims

- To evaluate the safety & efficacy of using the strategy of "partial repair-and-treat" in patients with ASD & severe PAH

background

1. Adult patients with unclosed ASD may develop various degree of pulmonary vascular obstructive disease with or without heart failure.

2. Surgical closure in those patients with severe PAH carries risks of pulmonary hypertension crisis or low cardiac output.

3. Presence of heart failure, arrhythmias or advanced ages are frequently associated.

Background

- In patients with old ages, heart failure or pulmonary hypertension, partial closure by surgery with a fenestrated patch can be safely performed with good mid-term results.

  Heart surg Forum 2012;15:111

- Transcatheter closure of ASD with a fenestrated device may be a superior alternative to surgery in those patients with a high risk.

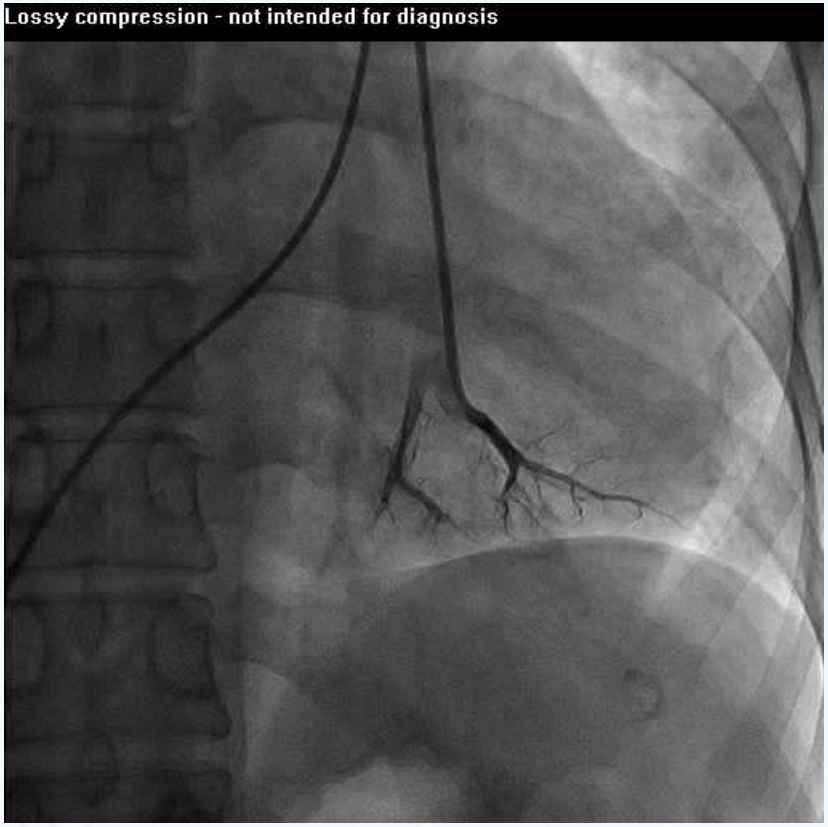
  CCI 2005;64:513, J Interv Cardiol 2008;21:44

Objectives

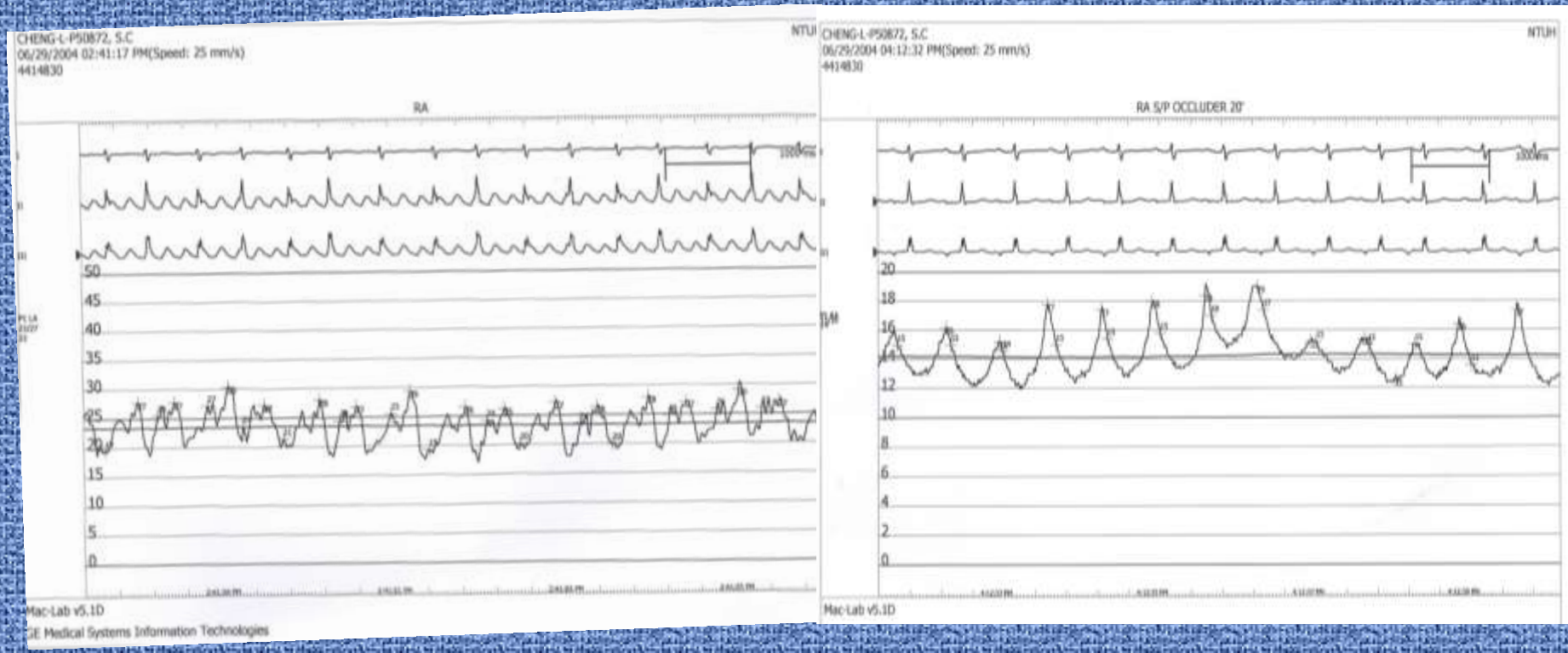
1. Can we stop the progression of pulmonary vascular obstructive disease & restore cardiac function by using a fenestrated device & followed by advanced treatment in patients with moderate to severe pulmonary hypertension or heart failure?

2. Will those patients be benefited from the strategy of treatment?

Wedge angio PAP 66/23 PVR3.8WU



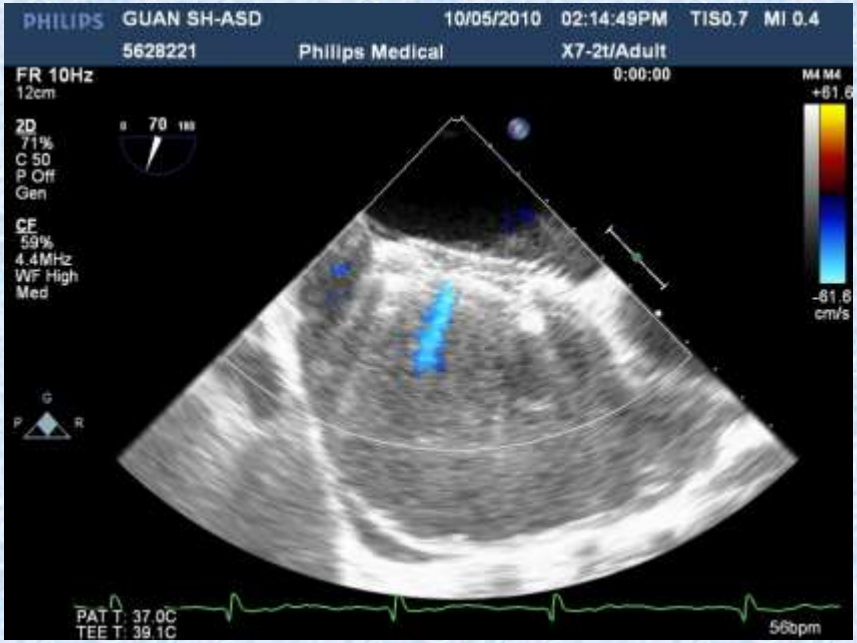
Atrial flutter before closure



Large ASD & severe PAH

pre

post

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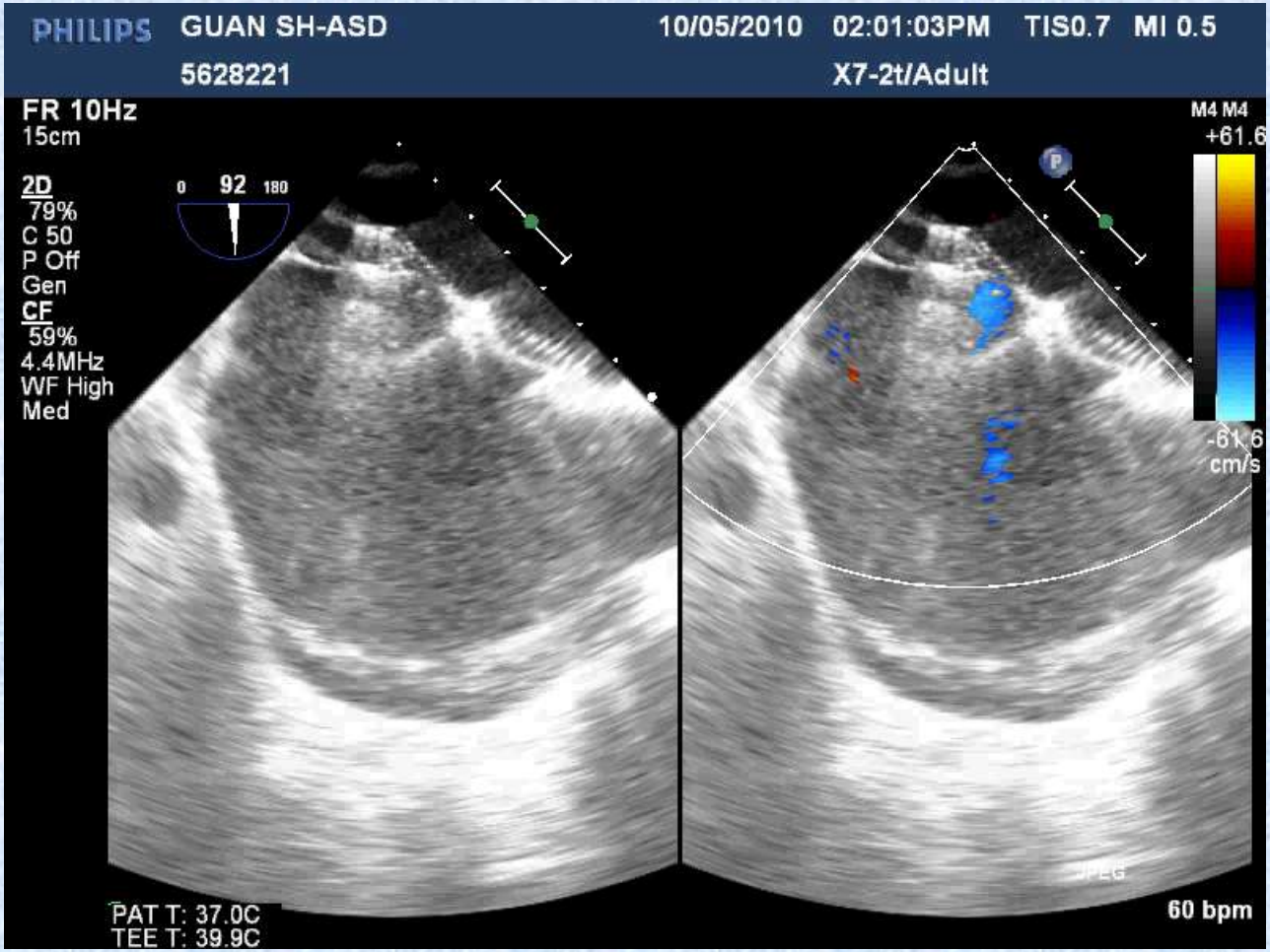
Pediatric and Adult Interventional Cardiac Symposium, Asia-Pacific
TAIPEI
April 1-4, 2015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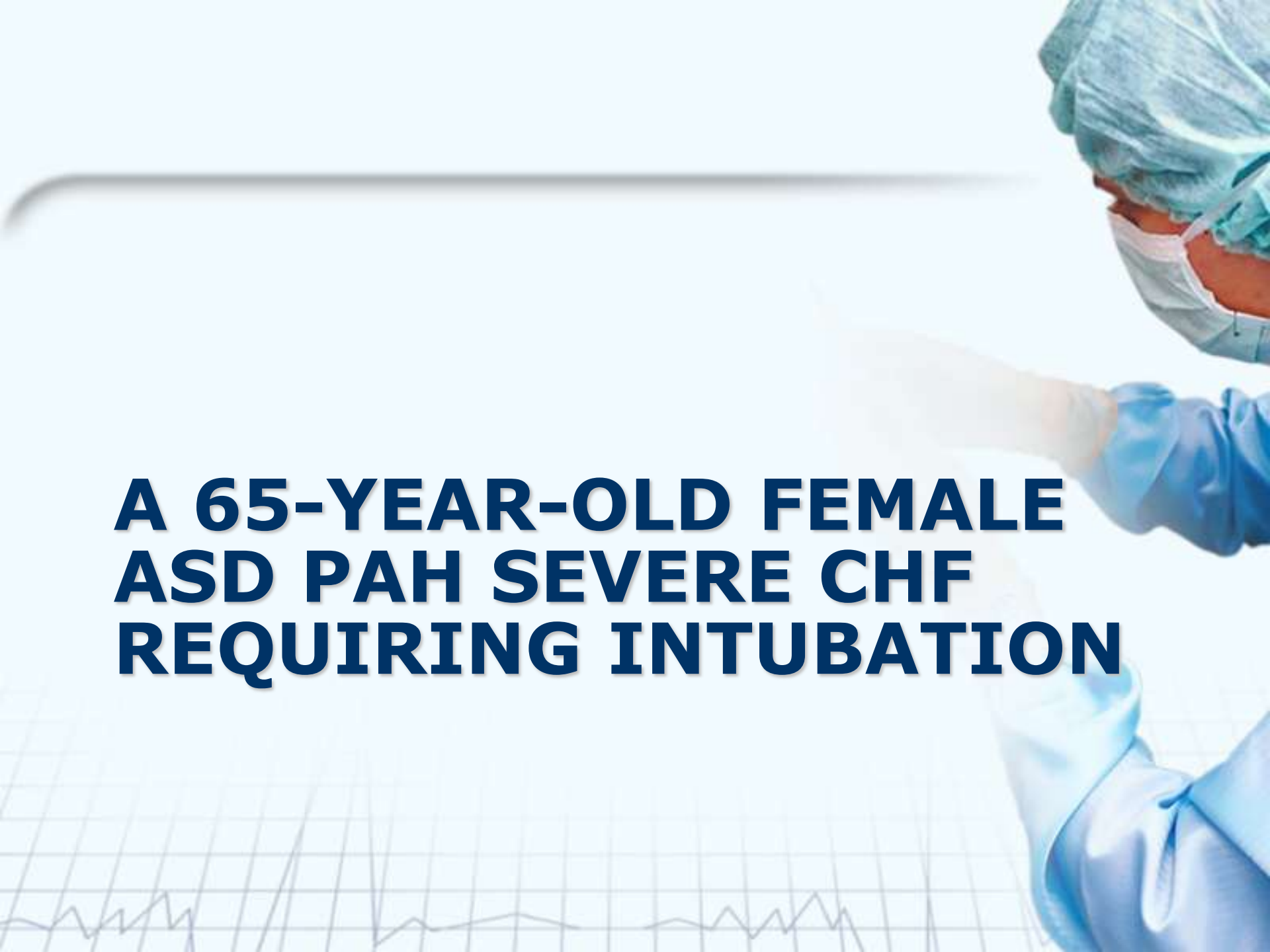

A 65-YEAR-OLD FEMALE ASD PAH SEVERE CHF REQUIRING INTUBATION


CHEST AP


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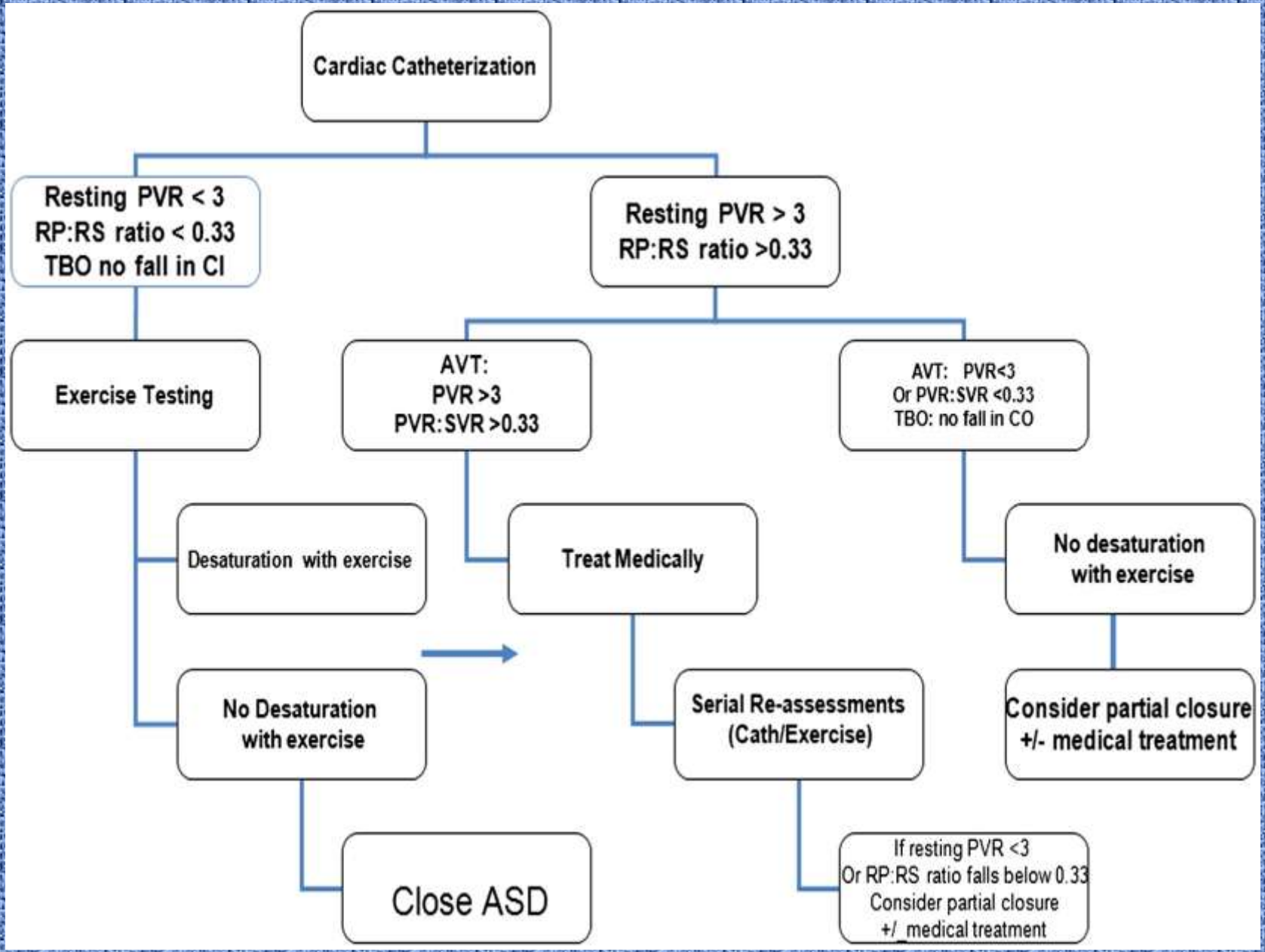
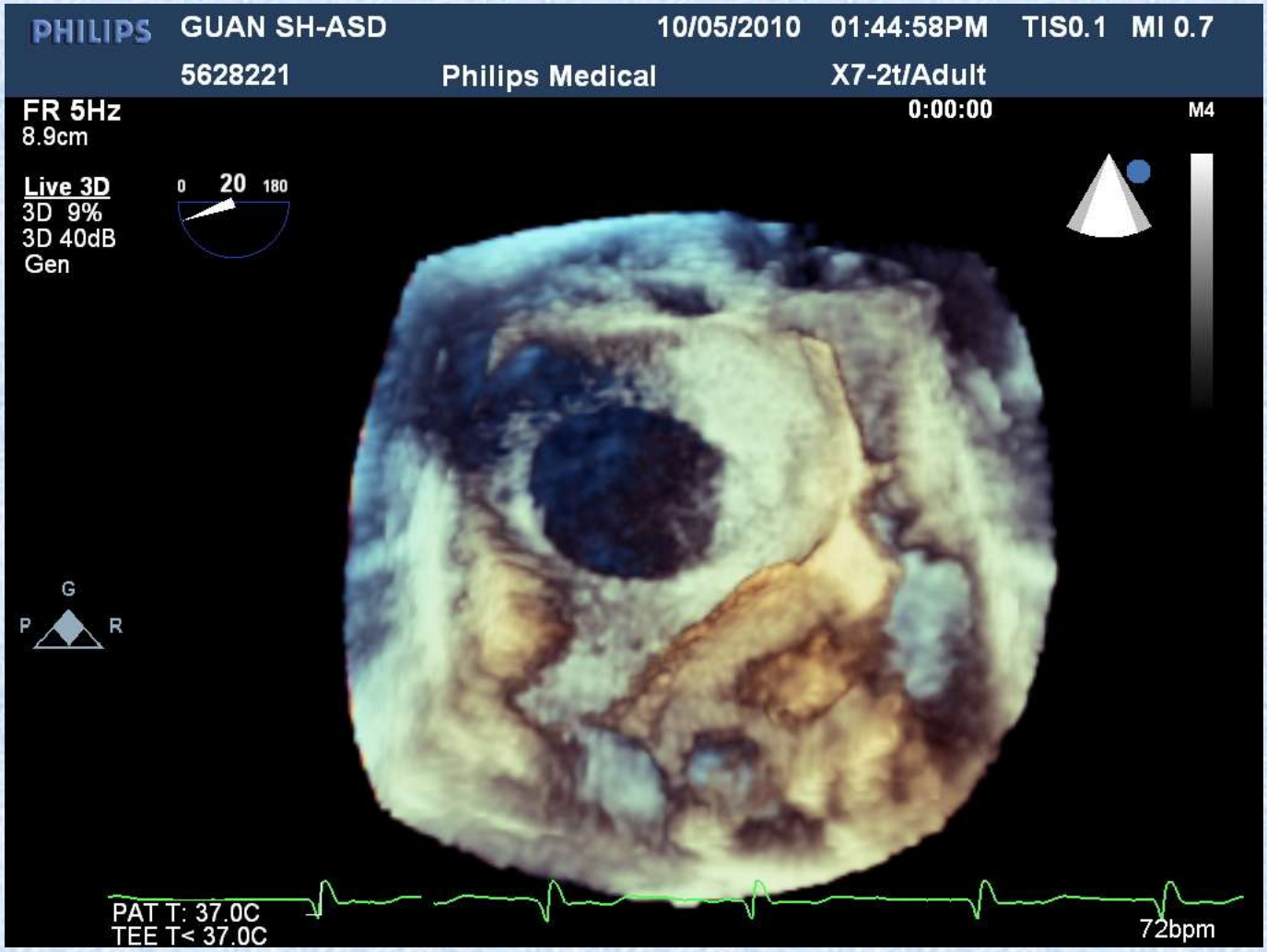
3D 50dB
HGen
M2
JPEG
100 bpm
PHILIPS


Cardiac Catheterization
Resting PVR < 3
RP:RS ratio < 0.33
TBO no fall in CI
Resting PVR > 3
RP:RS ratio >0.33
Exercise Testing
AVT:
PVR >3
PVR:SVR >0.33
AVT: PVR<3
Or PVR:SVR <0.33
TBO: no fall in CO
Desaturation with exercise
Treat Medically
No desaturation
with exercise
No Desaturation
with exercise
Serial Re-assessments
(Cath/Exercise)
Consider partial closure
+/- medical treatment
Close ASD
If resting PVR <3
Or RP:RS ratio falls below 0.33
Consider partial closure
+/ medical treatment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