



# **Pulmonary Arterial Hypertension and Congenital Heart Disease: Role of Interventional Cardiology**

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**Table 1 Updated Classification of Pulmonary Hypertension\***

**1. Pulmonary arterial hypertension**

- 1.1 Idiopathic PAH
- 1.2 Heritable PAH
  - 1.2.1 BMPR2
  - 1.2.2 ALK-1, ENG, SMAD9, CAV1, KCNK3
  - 1.2.3 Unknown
- 1.3 Drug and toxin induced
- 1.4 Associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 HIV infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart diseases
  - 1.4.5 Schistosomiasis

1' Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomas

1'' **Persistent pulmonary hypertension of the newborn (PPHN)**

**2. Pulmonary hypertension due to left heart disease**

- 2.1 Left ventricular systolic dysfunction
- 2.2 Left ventricular diastolic dysfunction
- 2.3 Valvular disease
- 2.4 **Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies**

**3. Pulmonary hypertension due to lung diseases and/or hypoxia**

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental lung diseases

**4. Chronic thromboembolic pulmonary hypertension (CTEPH)**

**5. Pulmonary hypertension with unclear multifactorial mechanisms**

- 5.1 Hematologic disorders: **chronic hemolytic anemia**, myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure, **segmental PH**

\*Modified as compared with the Dana Point classification. Reprinted with permission from Simonneau G, Gatzoulis MA, Adatia I. Updated clinical classification of pulmonary hypertension. *J Am Coll Cardiol* 2013;62:D34-41.

BMPR2 = bone morphogenetic protein receptor type II; CAV1 = caveolin 1; ENG = endoglin; KCNK3 = potassium channel K3; PAH = pulmonary arterial hypertension; PH = pulmonary hypertension; PPHN = persistent pulmonary hypertension of the newborn.

**Table 3**

**Clinical Classification of Congenital Heart Disease Associated With Pulmonary Arterial Hypertension**

**1. Eisenmenger Syndrome**

**2. Left to right shunts**

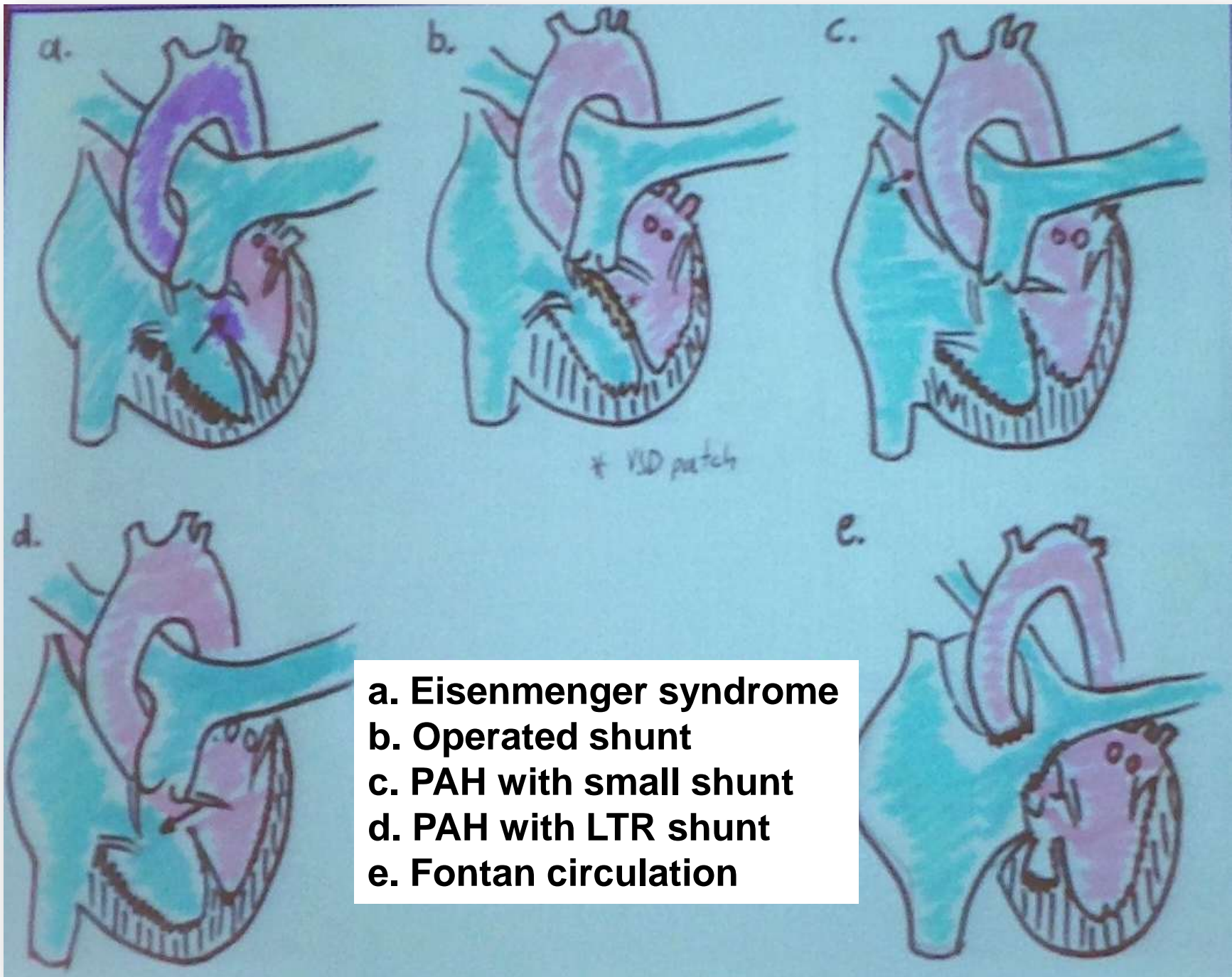
*Operable*

*Inoperable*

**3. PAH with co-incidental CHD**

**4. Post-operative PAH**

Definition of PAH based on mean PAP  $>25$  mm Hg and PVR  $>3$  Wood units  $\times$  m<sup>2</sup>.



**a. Eisenmenger syndrome**  
**b. Operated shunt**  
**c. PAH with small shunt**  
**d. PAH with LTR shunt**  
**e. Fontan circulation**

# Pediatric Pulmonary Hypertension

## Biventricular circulation:

mean PAP  $\geq$  25 mmHg  
and

PVRI  $>$  3 WUm<sup>2</sup>

Nice 2013  
Panama 2011\*

PCWP  $\leq$  12 mmHg

Nice 2013

## Univentricular circulation:

PVRI  $>$ 3 WUm<sup>2</sup>

or

Trans-pulmonary  
gradient  $>$ 6 mmHg

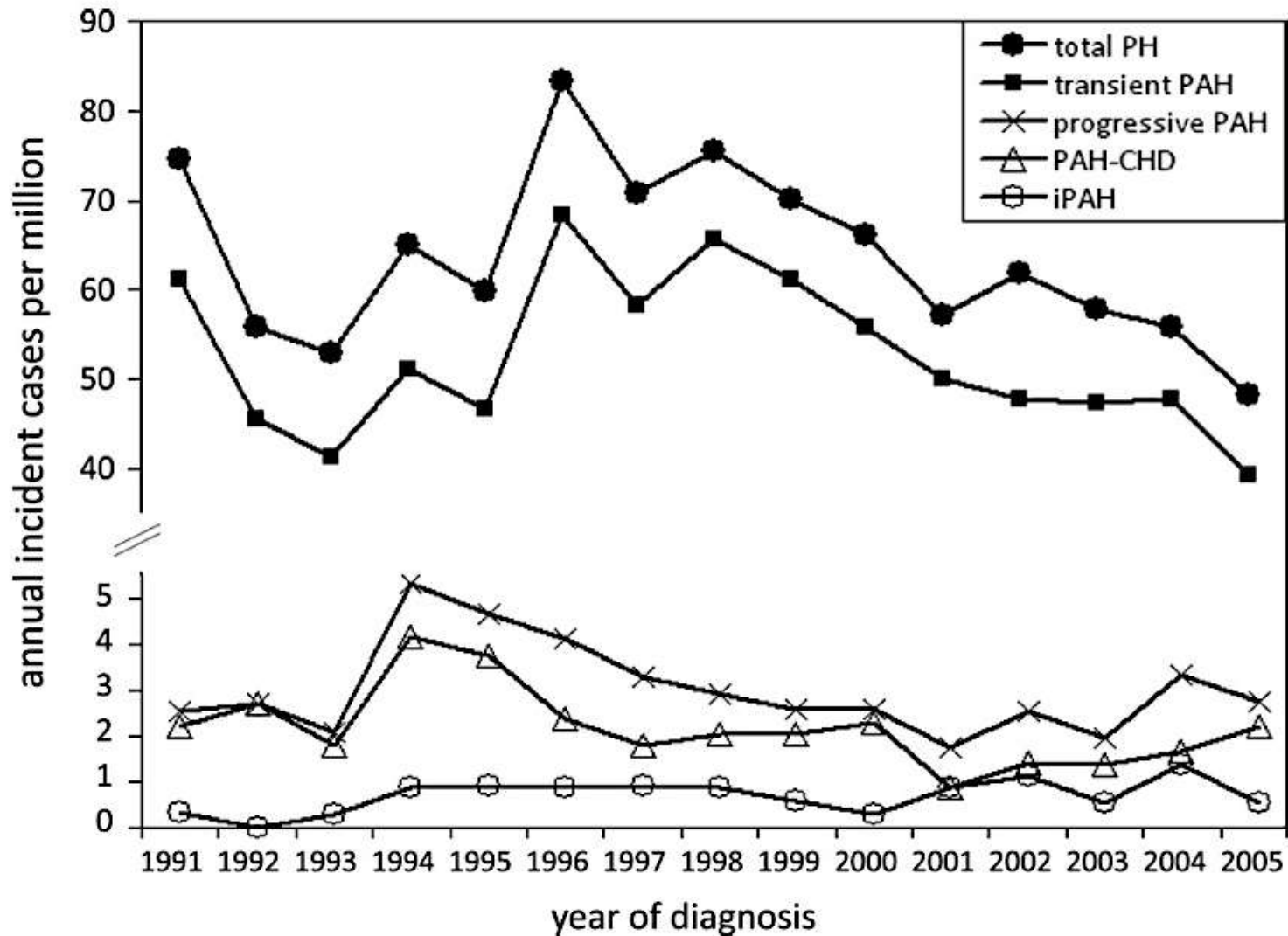
(mPAP – mLAP)

(even if mPAP  $<$ 25 mmHg)

Panama 2011\*

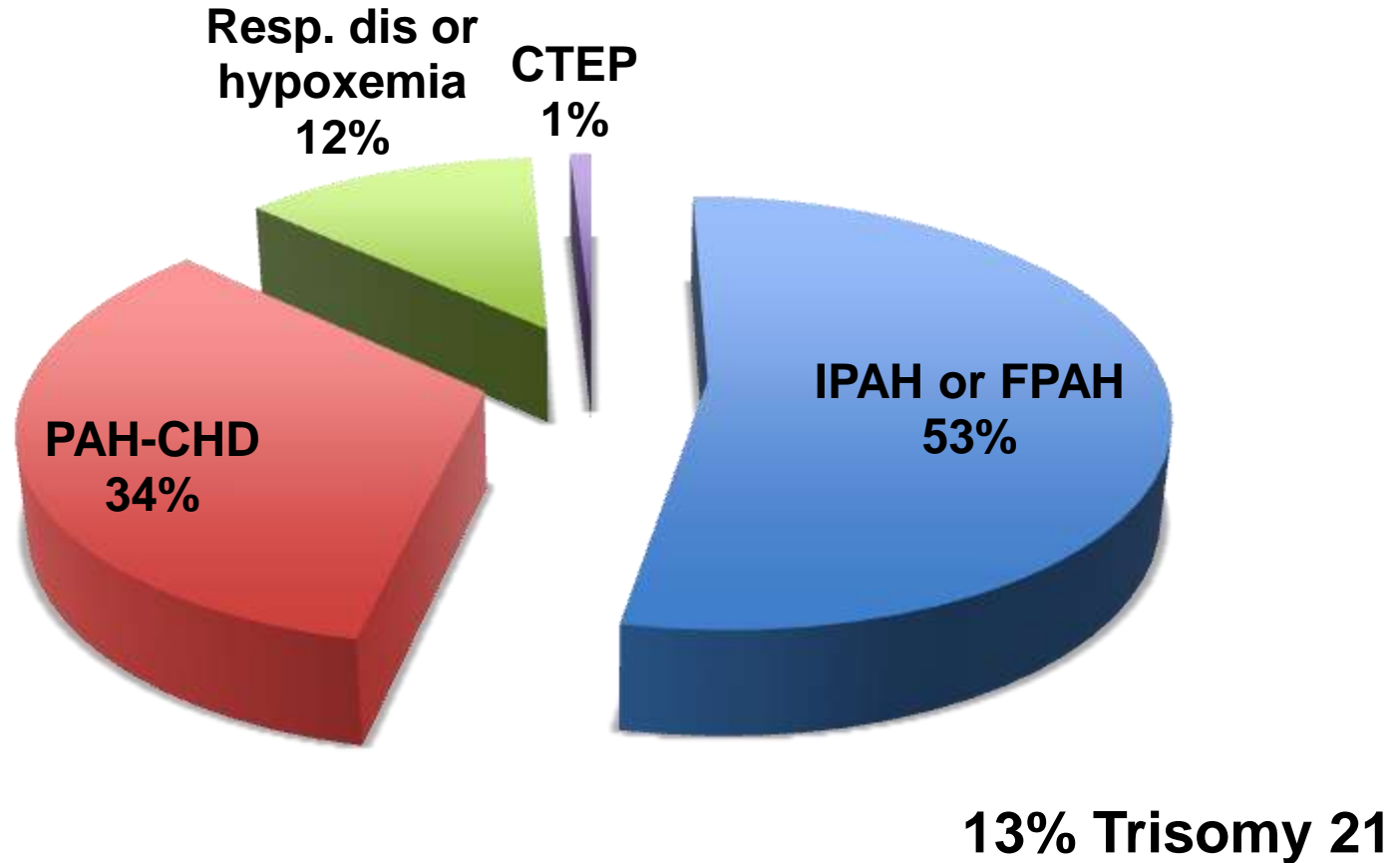
\*Cerro MJ, et al. Pulm Circ 2011;1:286–98.

# Annual incidence rates for pediatric pulmonary hypertension: Netherland Registry



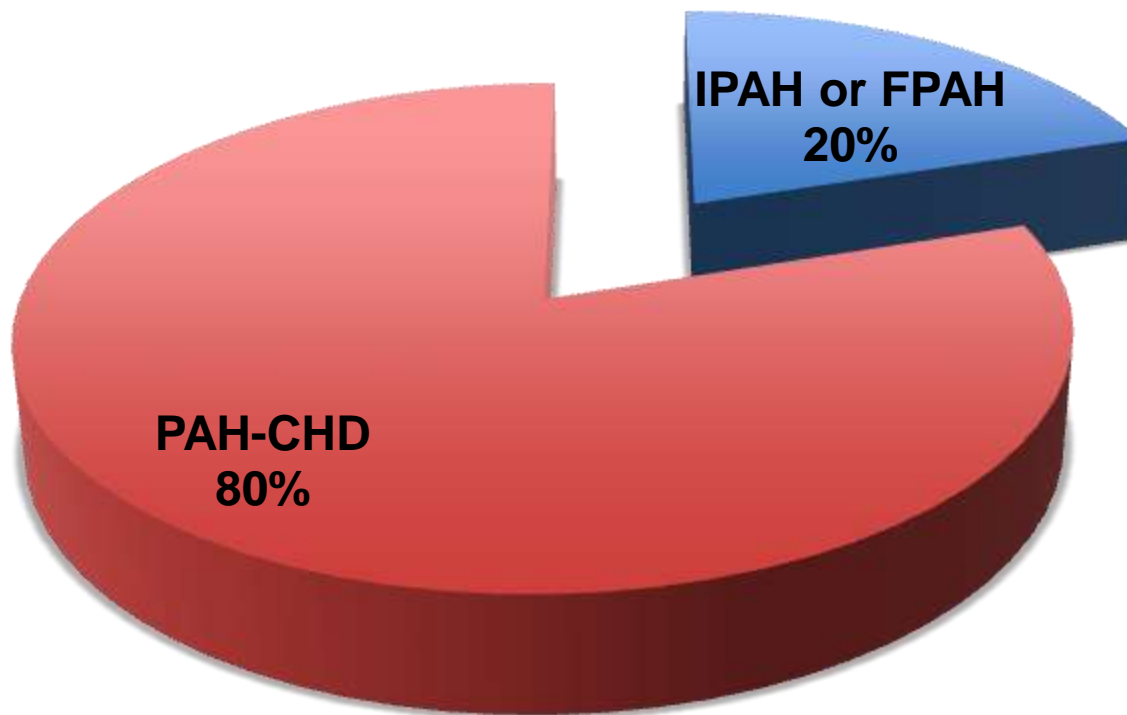
# TOPP Registry

(19 countries, 362 patients, age <17 yrs)





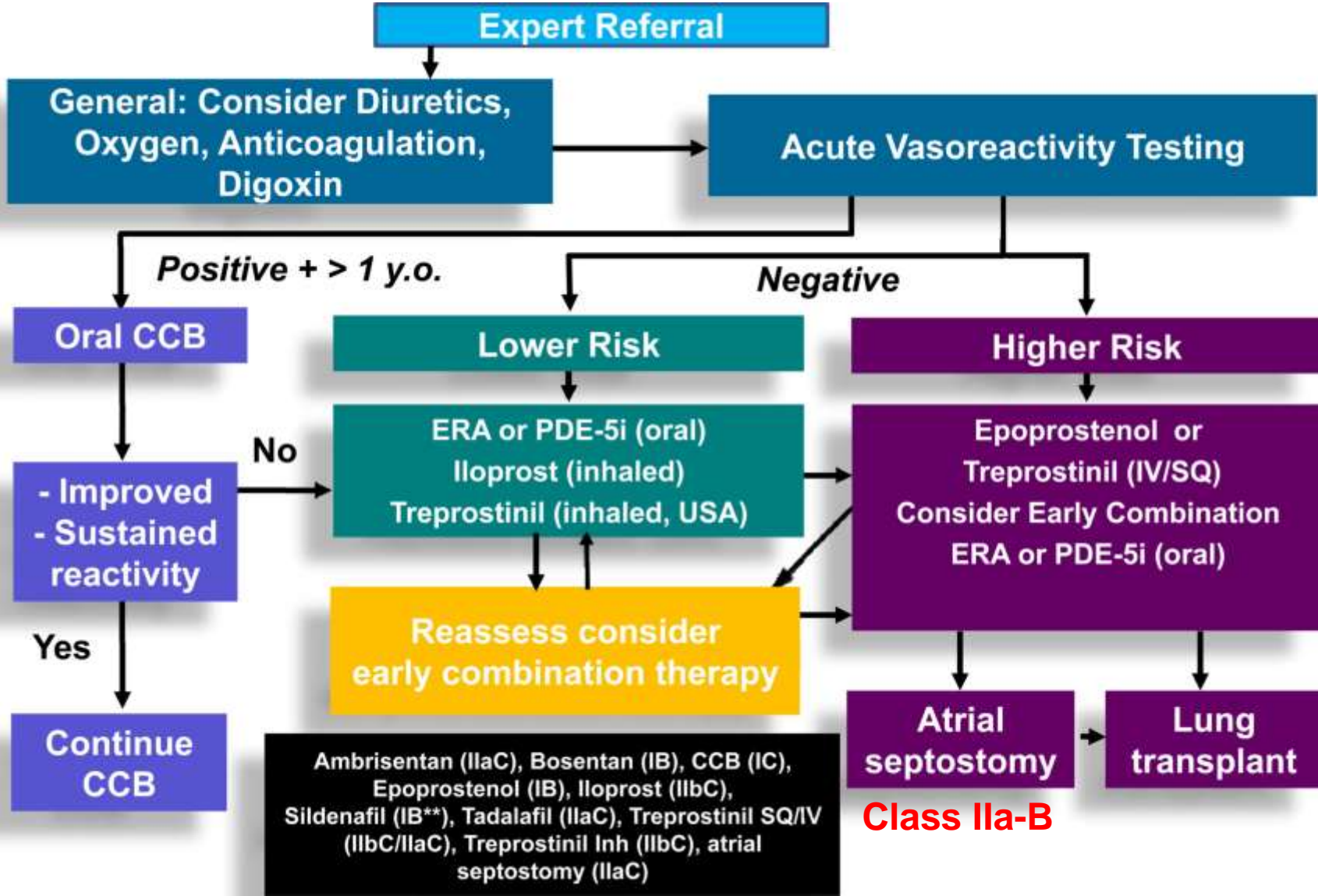
# Thai PHA Registry



**N = 41**



# World Symposium on Pulmonary Hypertension 2013 Consensus Pediatric IPAH/FPAH Treatment Algorithm\*



# **Role of Interventional Cardiology**

# Eisenmenger Syndrome

## **Indication:**

Deterioration of SpO<sub>2</sub> or RV performance.

## **Aim:**

To decompress RV and provide adequate systemic blood flow.

## **Methods:**

1. Creation of atrial communication.
2. Creation of PA-Ao connection.

with right-to-left shunting and systemic desaturation

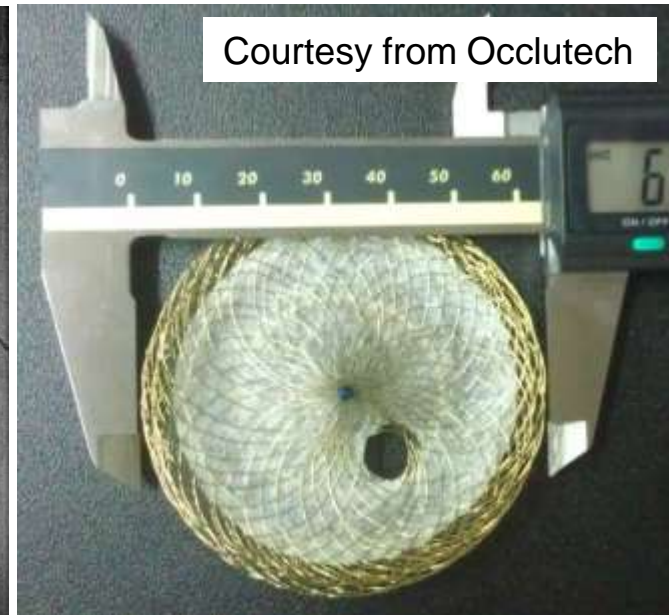
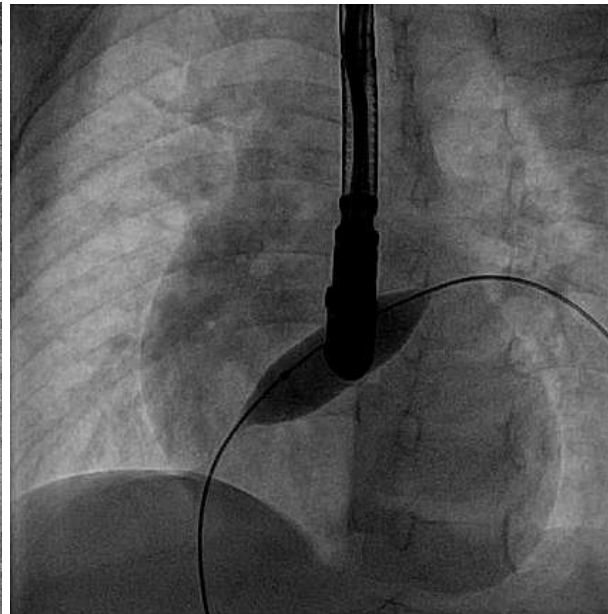
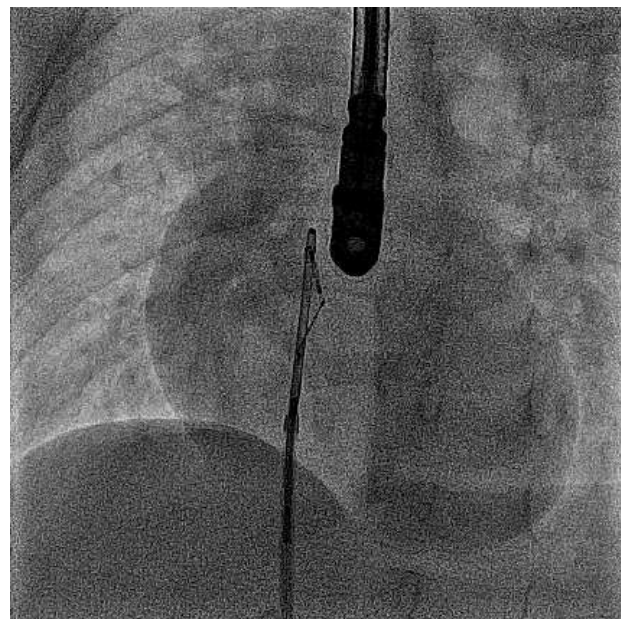
# Creation of atrial communication

- Blade atrial septostomy
- Graded balloon dilation
- Fenestrated Device

RAP  $\geq$  20 mm Hg  
LVEDP  $>$  18 mm Hg  
PVRI  $>$  55 U/m<sup>2</sup>  
Baseline SpO<sub>2</sub>  $<$  90% in room air

as these factors have been shown to be predictive of a 25% procedural mortality rate.

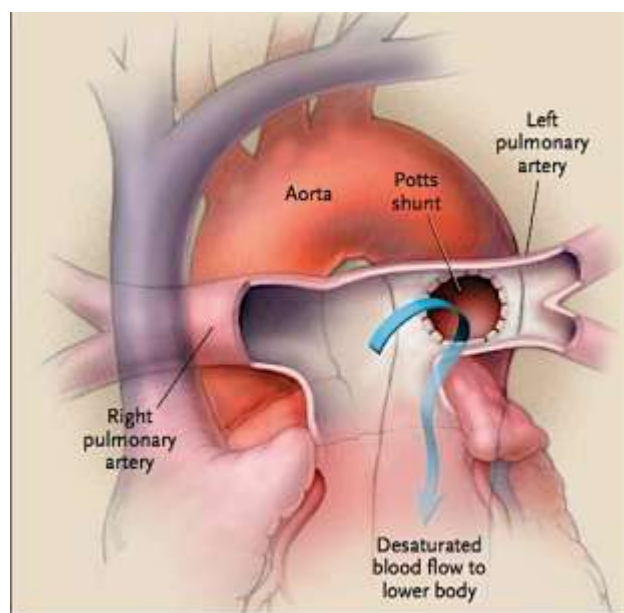
Keogh AM, et al. J Am Coll Cardiol 2009;54 Suppl 1:S67-77.



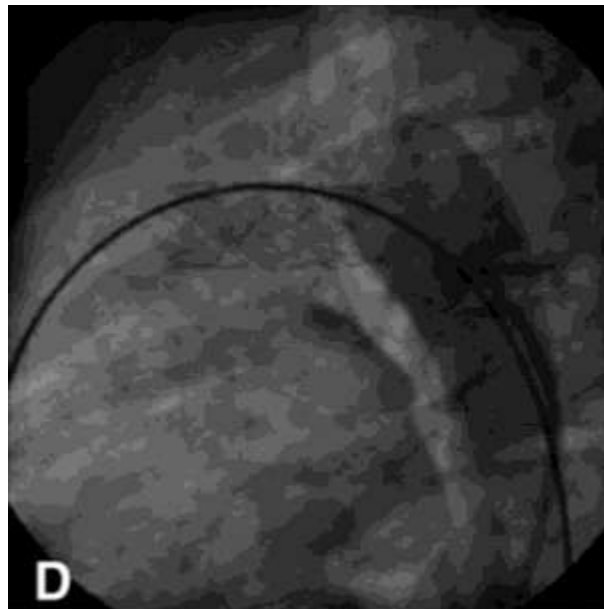
# Creation of PA-Ao Connection

- Pott's Shunt
- PDA Stenting

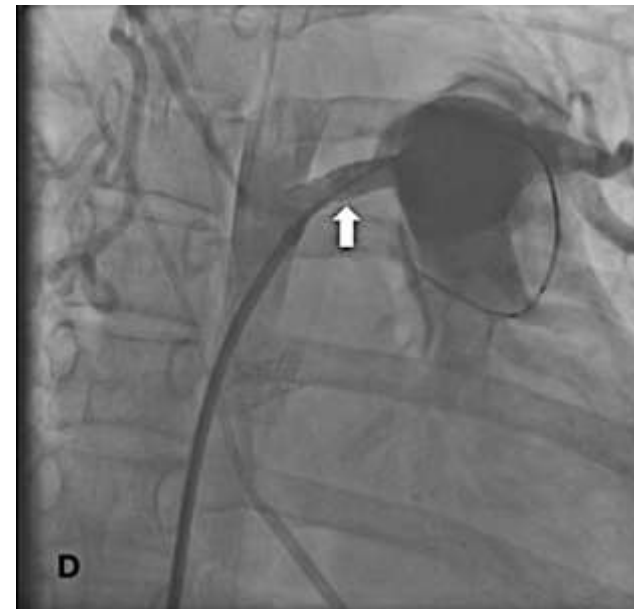
Supra-systemic PA pressure  
Severe RV dysfunction



Bonnet D. NEJM 2004 350;6:623.



Boudjemline Y et al. Circ Cardiovasc Interv. 2013;6:e18-e20



Esch JJ, et al. JHLT 2013; 32: 381-7.

**Patient 2:** 3.3 yrs old male, 14 kg. Trisomy 21 with PAVC diagnosed @ 32 mo.

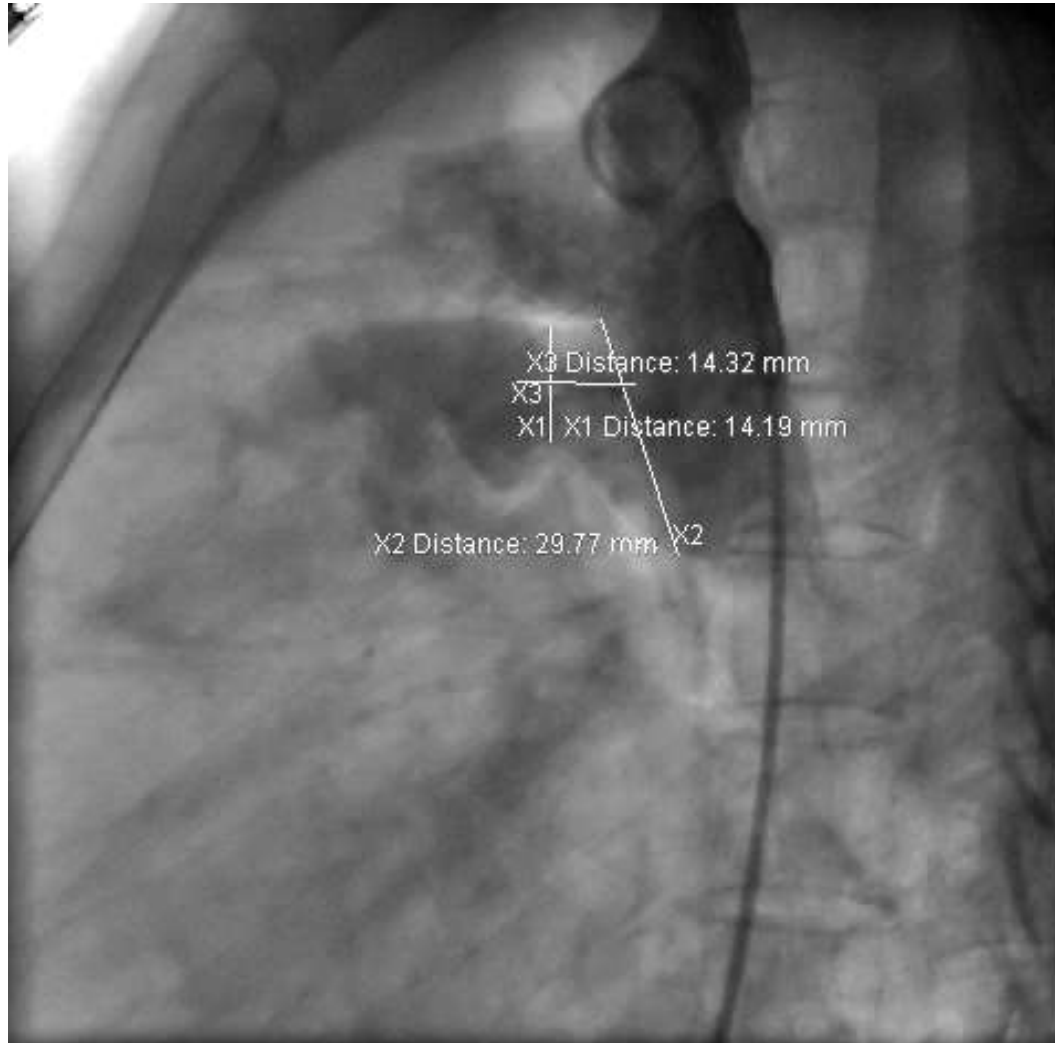
	Pre	Post
Meds	Sildenafil	none
Symptoms	Fatigue	none
SpO <sub>2</sub> (arm/leg)	91/86	89/84
BNP, ng/mL	203	<5
Hb, g/dL	12.3	18.5
PDA shunting	Rt. to Lt. small	Large
Septal curvature	Inverse	Flat
TAPSE, mm	18	21
Pulmonary VTI, cm	14.5	12
F/U PDA gradient	NA	1.5 m/s
PDA size, mm	1.4	
sPA/sAoP, mmHg	131/98	
Type of stent	Valéo 7x26	
Follow-up, mo/outcome	10/ alive	

# Left to Right Shunts

- **Single lesion:** ASD, VSD, PDA
- **Combine/Complex lesion**

The shunts may be either operable or inoperable but are characterized by increased PVR.

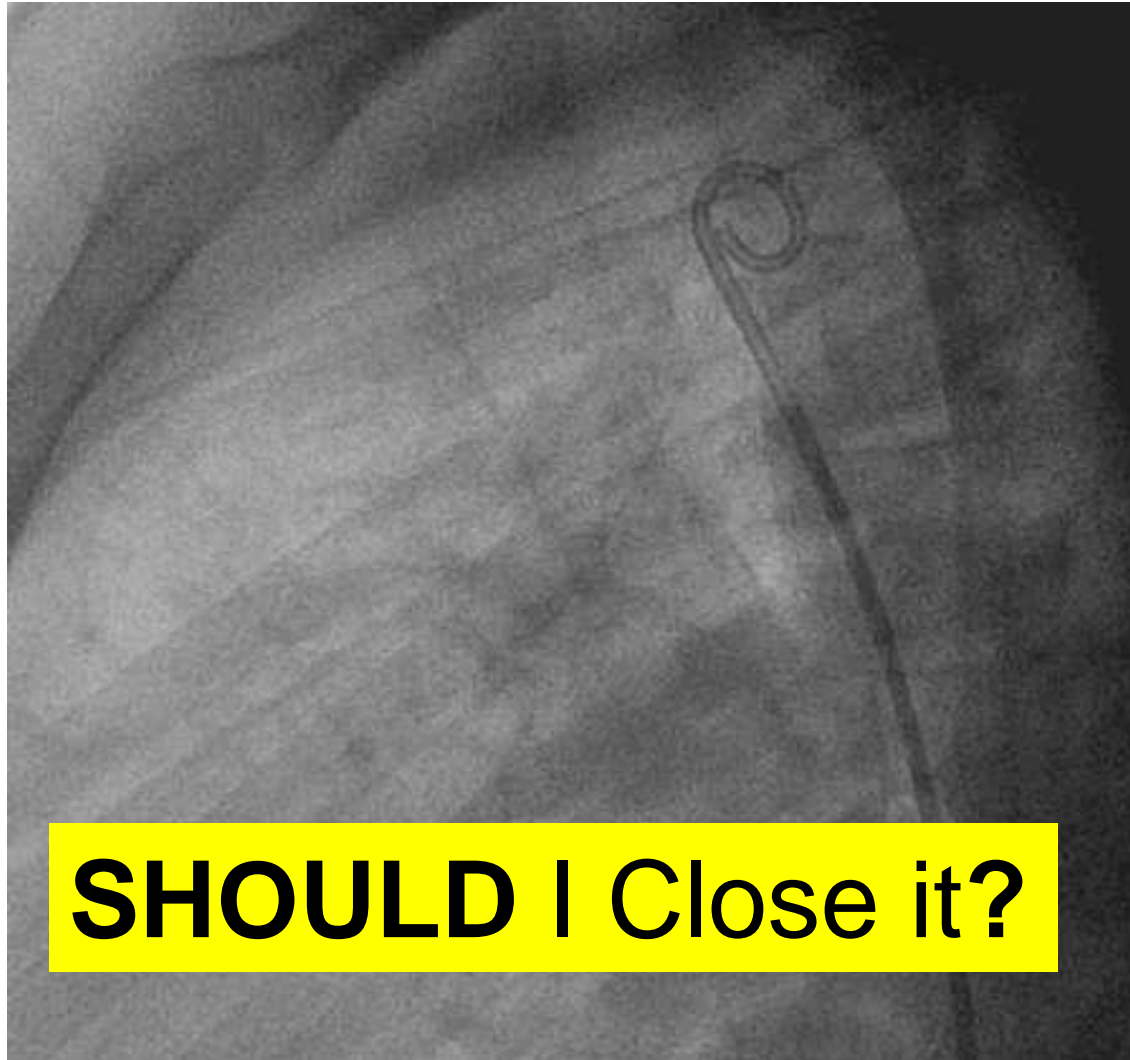
# Left to Right Shunt: Large PDA



30 years old with PDA. Systemic PA pressure.



# Left to Right Shunt: Large PDA



19 years old, Down syndrome, large PDA. Systemic PA pressure.

# Problems and Risks

## Problems

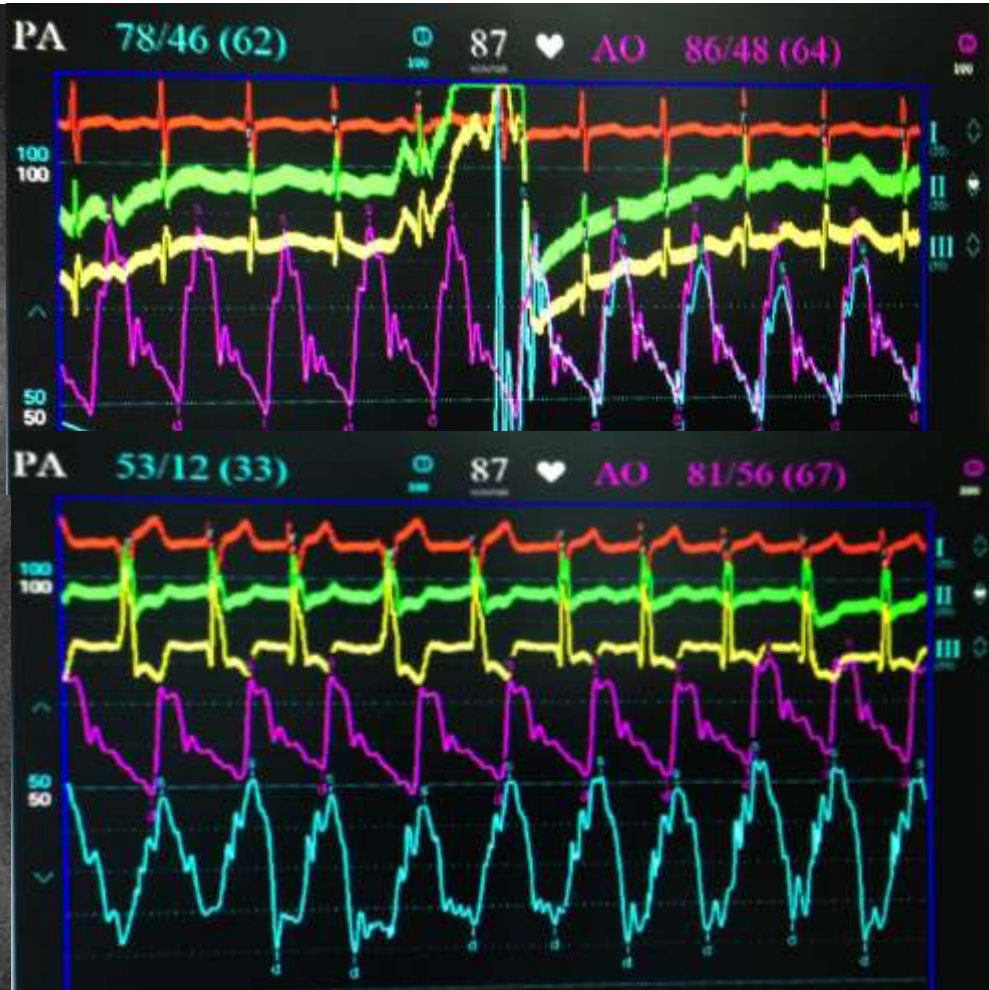
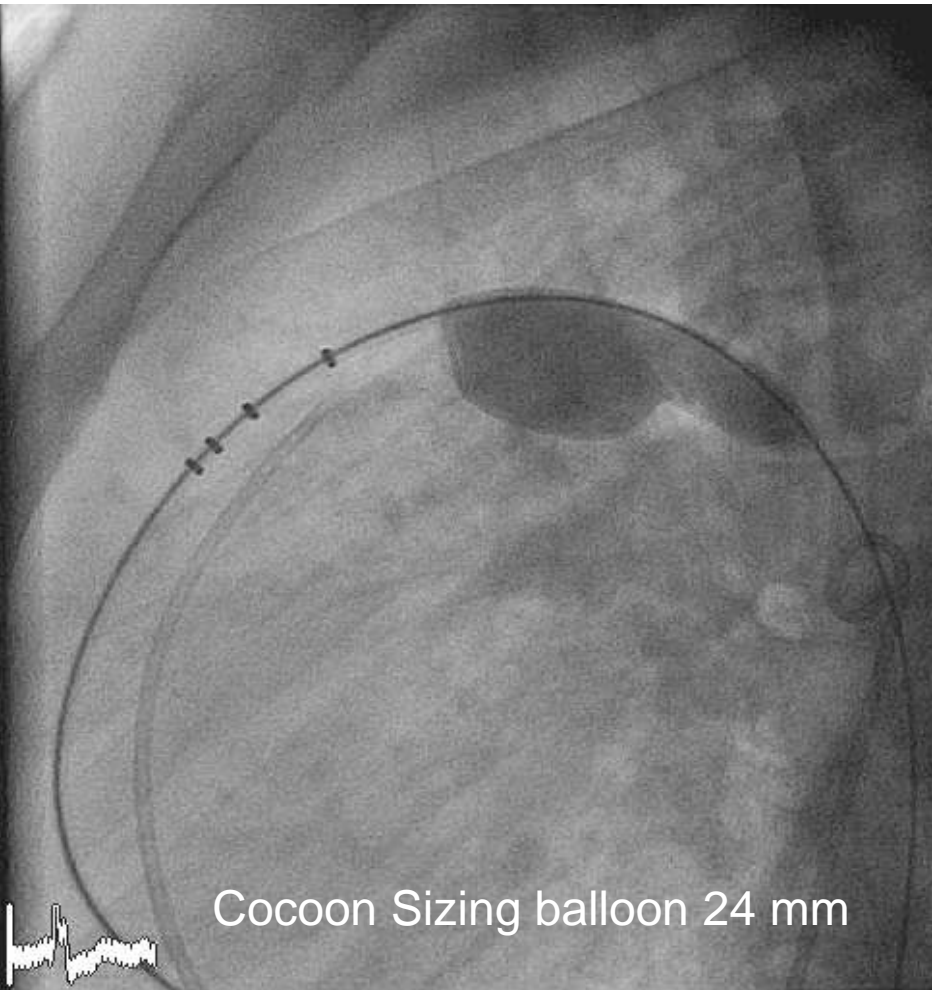
- High PA pressure
- High PVR
- Large/huge defect

## Risks

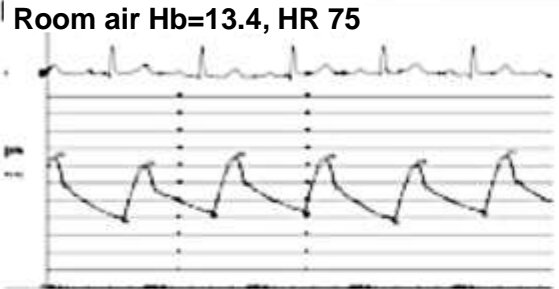
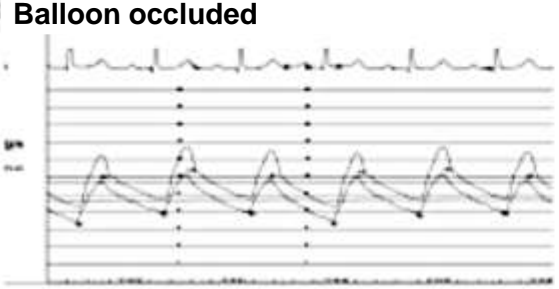
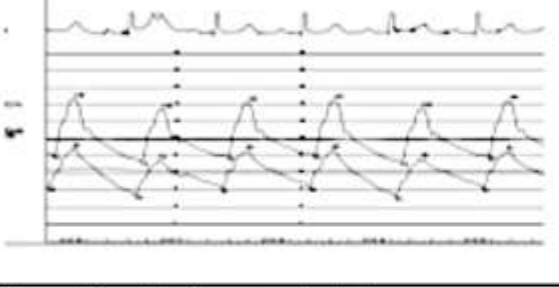
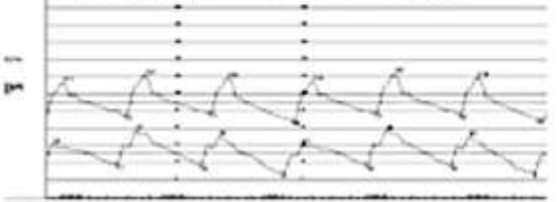
Increase risk of complications:

- Procedure related:  
vascular injury
- Disease related:  
Pulmonary hypertensive  
crisis
- Device related:
  - Residual shunt
  - Device embolization
  - Device migration
  - Adjacent organs  
obstruction

# Test Occlusion



19 years old, Down syndrome, large PDA. Systemic PA pressure.

Hemodynamic wave form	Pressure data	Oxygen saturation	Hemodynamic profile
<p>Room air Hb=13.4, HR 75</p> 	<p>LV 132/11 Ao 132/69/88 DAO 131/67/90</p> <p>RA 9/7/9 RV 117/7 PA 131/64/88</p>	<p>LV 87 Ao 87 DAO 88 SVC 51 RA 47 RV 58 PA 58</p>	<p><math>Qp/Qs\ 36/29 = 1.24</math></p> <p><math>Rp/Rs\ (88/89)/1.24 = 0.79</math></p> <p><math>Qp = 150/1.34 \times 10 \times 13.4 \times (0.87 - 0.58) = 2.88</math></p> <p><math>PVR = (88 - 11)/2.88 = 26.73</math></p>
<p>Balloon occluded</p> 	<p>AO 140/77/100</p> <p>PA 82/39/60</p>		
<p>100% Oxygen and balloon occluded</p> 	<p>AO 140/77/100</p> <p>RA 11/9/7</p> <p>RV 90/8</p> <p>PA 82/44/62</p>	<p>Ao 95 SVC 54 RA 50 IVC 53 RV 67 PA 81</p>	<p><math>Qp/Qs\ 41/14 = 2.92</math></p> <p><math>Rp/Rs\ (62/100)/2.92 = 0.21</math></p> <p><math>Qp = 150/1.34 \times 10 \times 13.4 \times (0.95 - 0.81) = 6.00</math></p> <p><math>PVR = (62 - 11)/6.00 = 8.5</math></p>
<p>Iloprost and balloon occluded</p> 	<p>AO 118/73/92</p> <p>PA 49/13/32</p>	<p>Ao 92 SVC 46 PA 80</p>	<p><math>Qp/Qs\ 46/12 = 3.83</math></p> <p><math>Rp/Rs\ (32/92)/3.83 = 0.08</math></p> <p><math>Qp = 150/1.34 \times 10 \times 13.4 \times (0.92 - 0.80) = 7.00</math></p> <p><math>PVR = (32 - 11)/7.00 = 3.0</math></p>

**HOW** to close it?

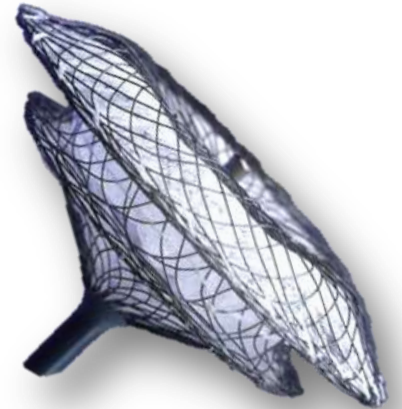
# Off-Label Use Devices for PDA

For

1. Large, hypertensive or tubular duct

Usually 4-6 mm. oversize

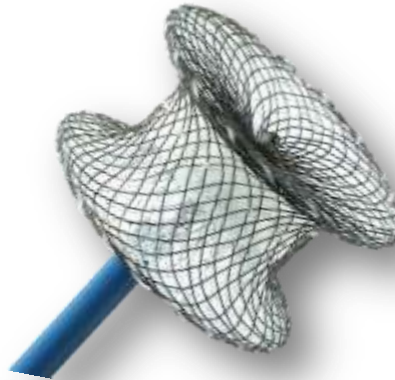
2. Unusual appearance



ASO



Amplatzer™ mVSD (SJM)

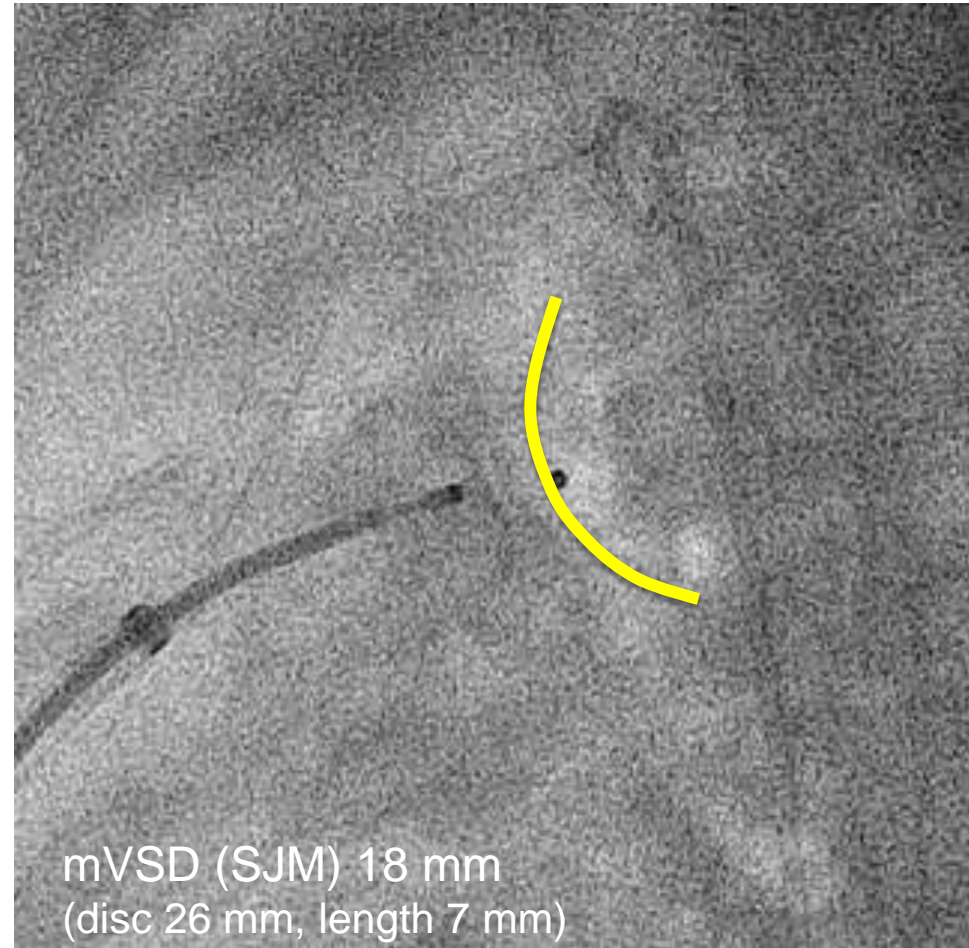
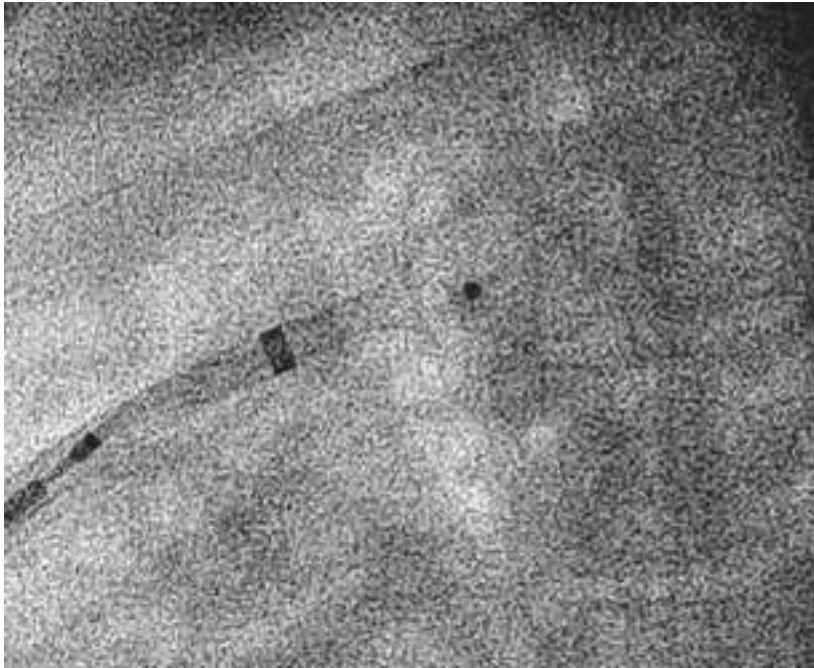


Amplatzer™ PI mVSD (SJM)



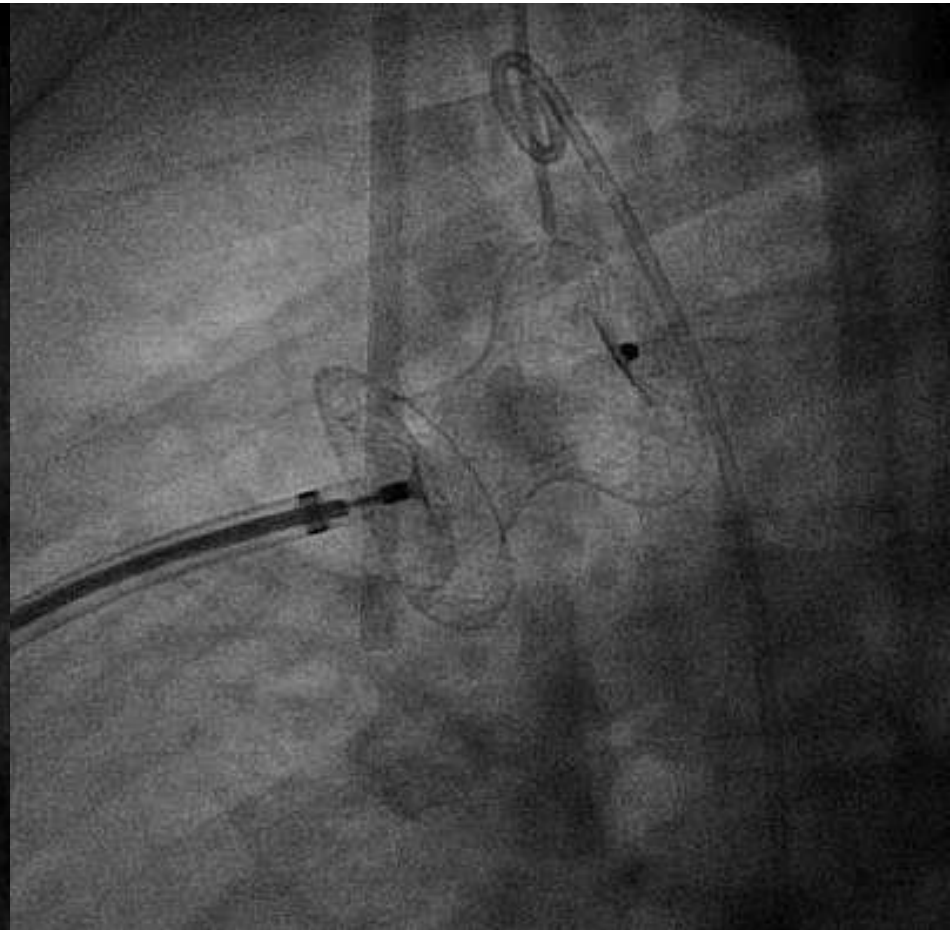
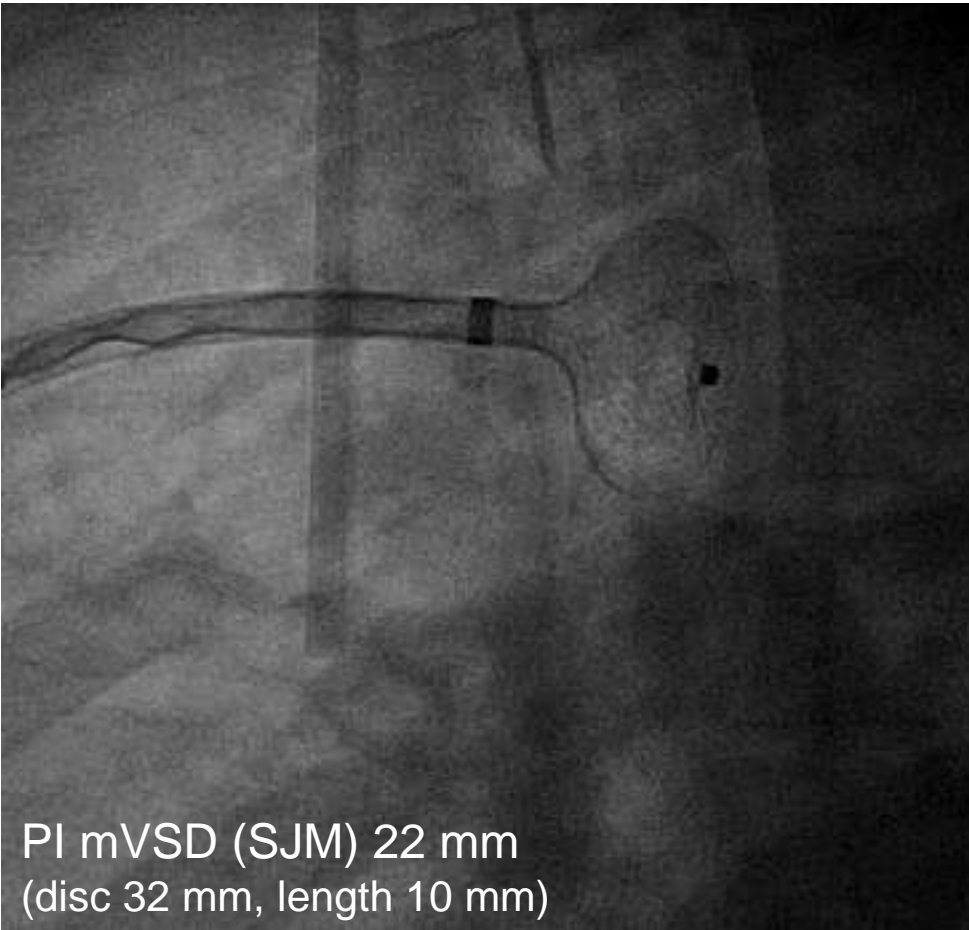
AVP II™ (SJM)

# mVSD and PI mVSD Devices



**Down syndrome 19 yrs with large PDA  
Sildenafil 2 mg/kg/day given 6 mo.**

# mVSD and PI mVSD Devices

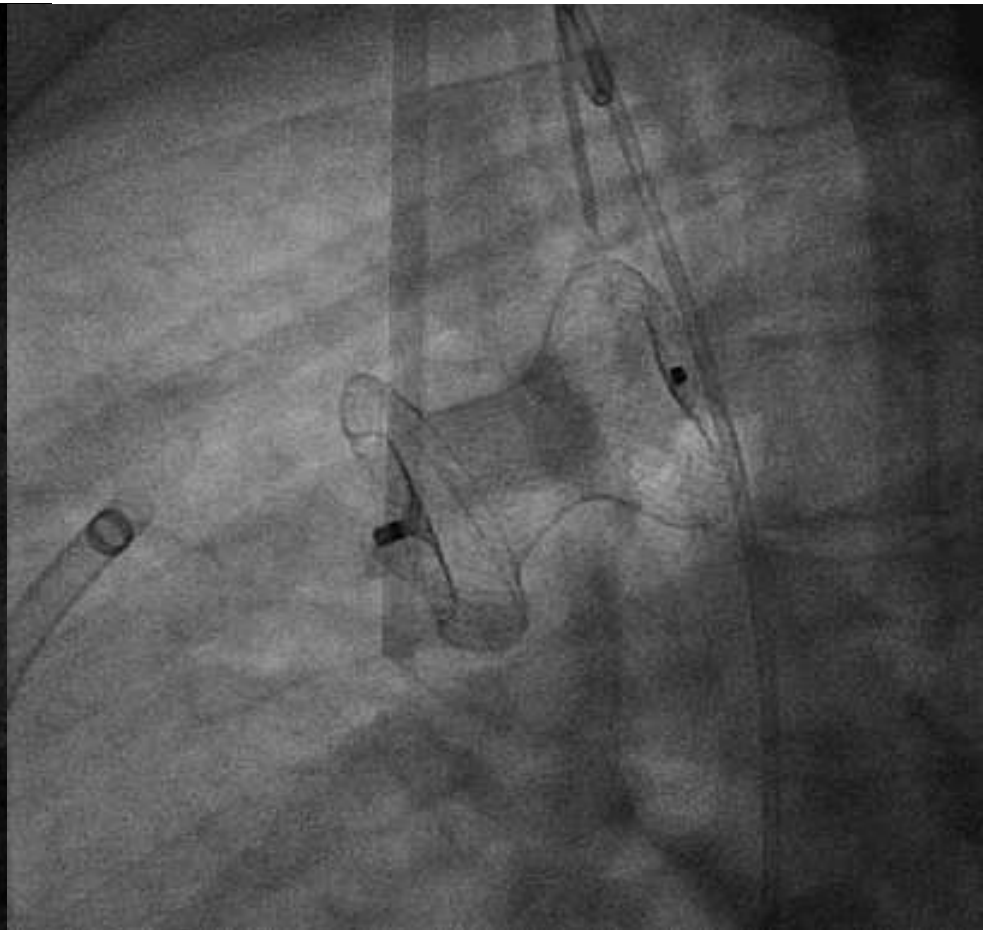
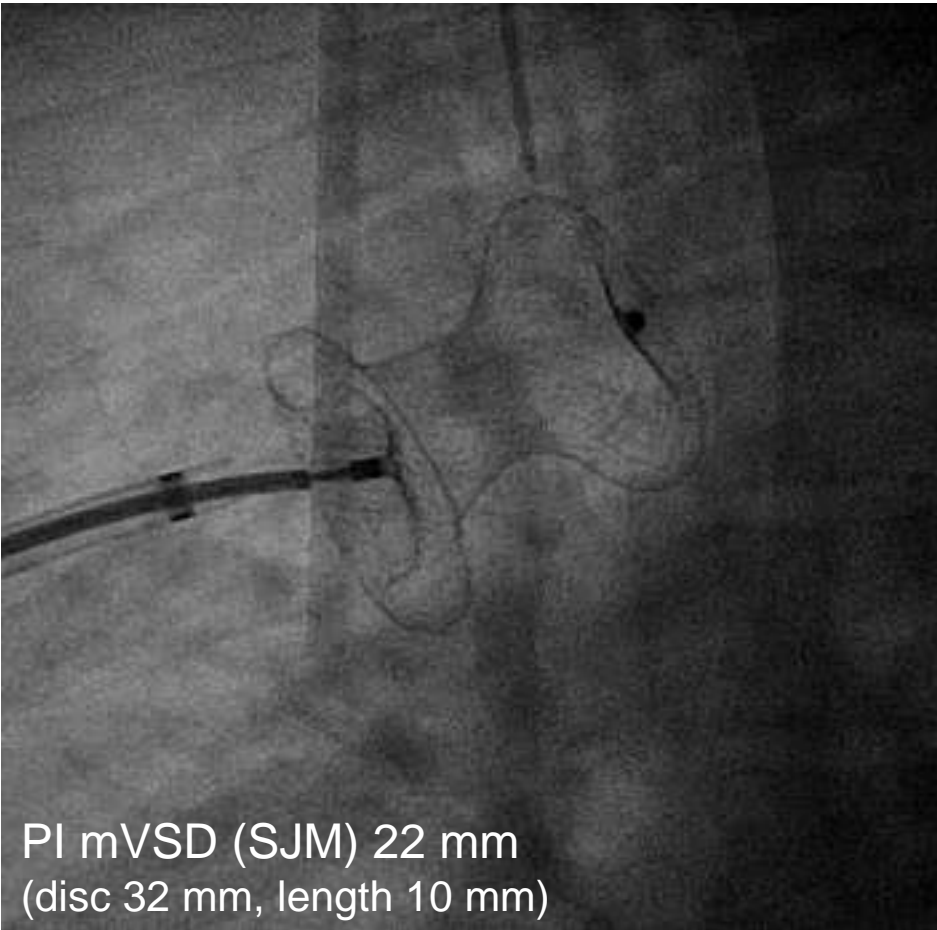


PI mVSD (SJM) 22 mm  
(disc 32 mm, length 10 mm)

**Down syndrome 19 yrs with large PDA  
Sildenafil 2 mg/kg/day given 6 mo.**



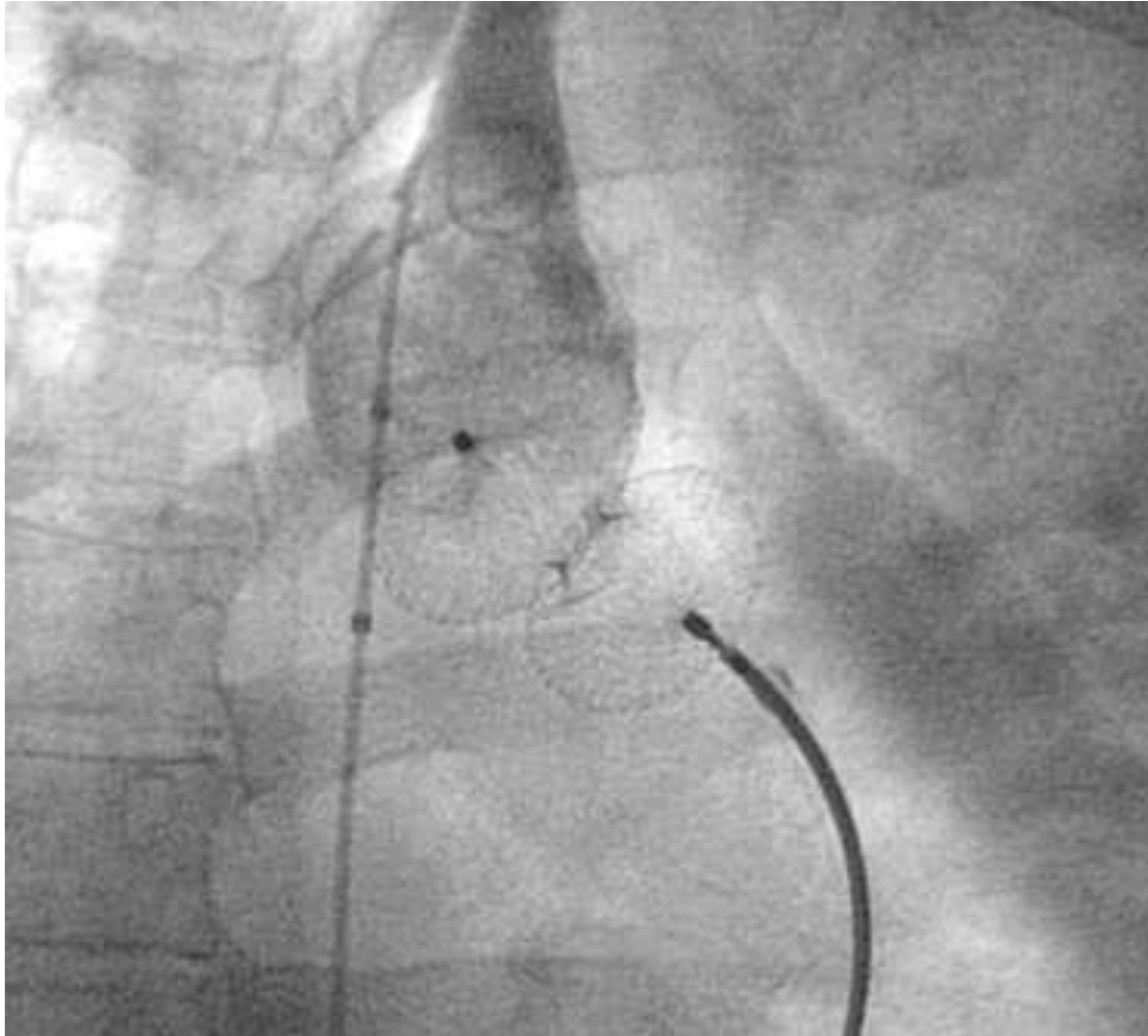
# mVSD and PI mVSD Devices



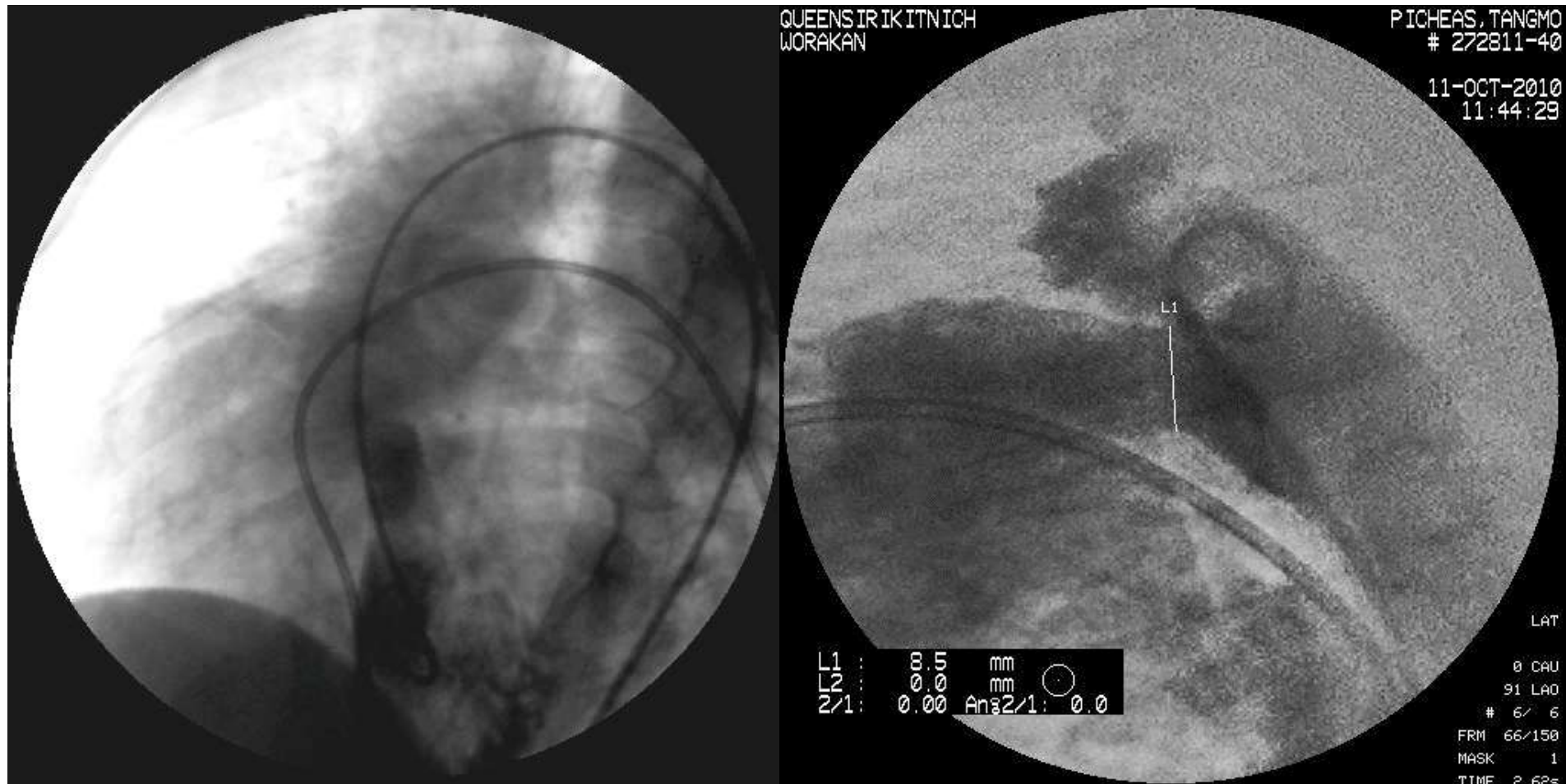
PI mVSD (SJM) 22 mm  
(disc 32 mm, length 10 mm)

**Down syndrome 19 yrs with large PDA  
Sildenafil 2 mg/kg/day given 6 mo.**

# Amplatzer™ Septal Occluder



# Left to Right Shunt: Combine lesions



14 years old boy with large PM, PDA. Systemic PA pressure.

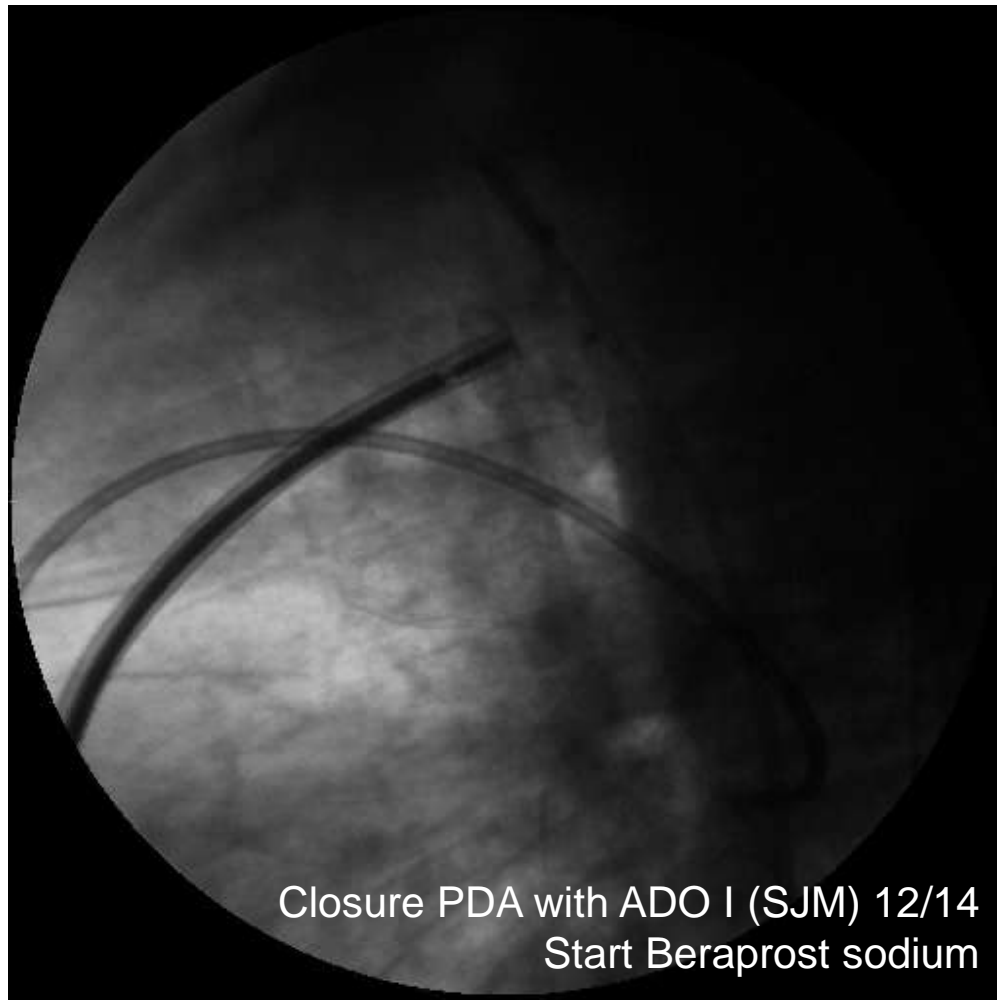
# Eisenmenger?

## What should I do?

	Room air		Oxygen 100%		Iloprost		Test PDA Occlusion	
	Pressure (mmHg)	O <sub>2</sub> sat (%)	Pressure (mmHg)	O <sub>2</sub> sat (%)	Pressure (mmHg)	O <sub>2</sub> sat (%)	Pressure (mmHg)	O <sub>2</sub> sat (%)
RSVC	Qp/Qs = 1.86 PVR=11.55 Rp/Rs = 0.51		Qp/Qs= 1.83 PVR=10.283 Rp/Rs = 0.49		Qp/Qs = 1.5 PVR = 10.24 Rp/Rs = 0.58			
LSVC								
RA								
IVC		68						
RV	119/16	79.3						
LPA	<b>119/75</b> <b>[96]</b>	<b>82.6</b>	<b>114/62</b> <b>[89]</b>	<b>96.5</b>	<b>112/58</b> <b>[88]</b>	<b>87.6</b>	<b>110/46</b> <b>[79]</b>	<b>83.7</b>
LV	130/16	94						
Ao	<b>124/62</b> <b>[96]</b>	<b>91.1</b>	<b>122/68</b> <b>[93]</b>	<b>100</b>	<b>120/67</b> <b>[94]</b>	<b>91.7</b>	<b>126/88</b> <b>[104]</b>	<b>92.8</b>

14 years old boy with large PM, PDA. Systemic PA pressure.

# Left to Right Shunt: Combine lesion



**Clinically improved**  
SpO2 94-96 %.

**Sent for closure VSD**  
@ 15 year 2 month

(1 yr after PDA closure)

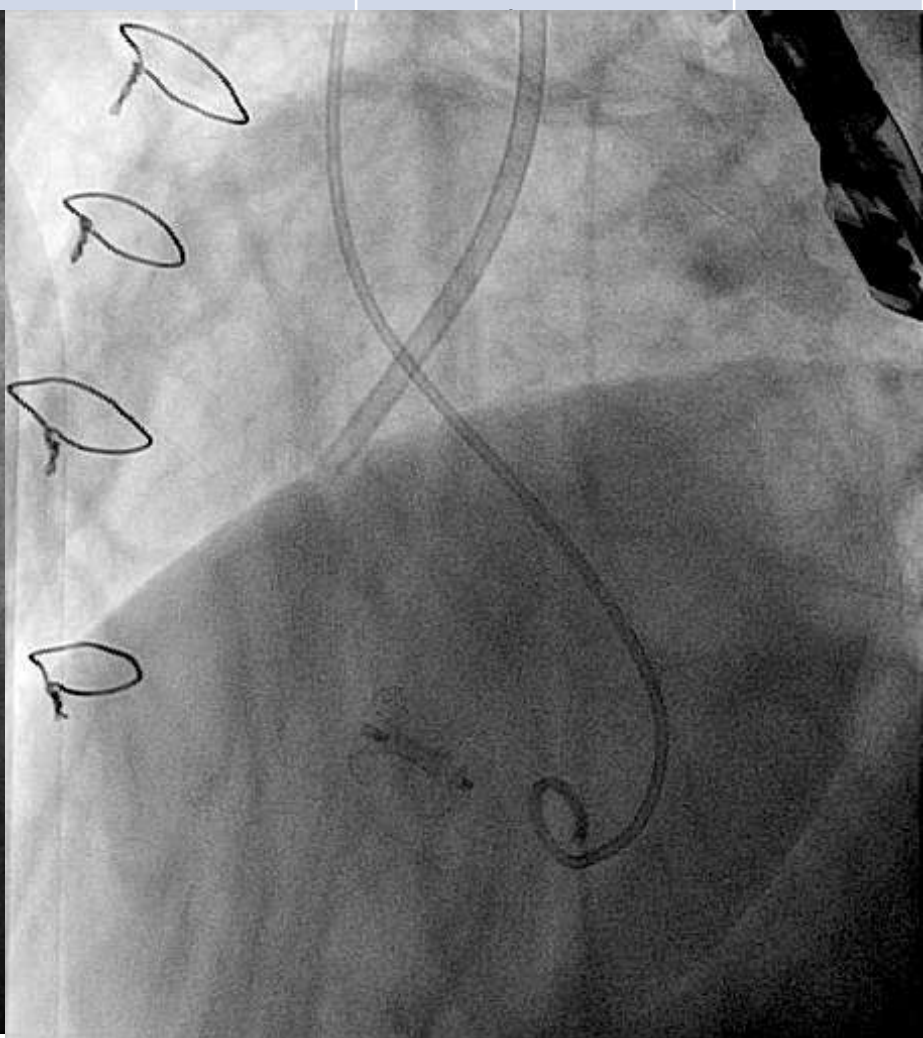
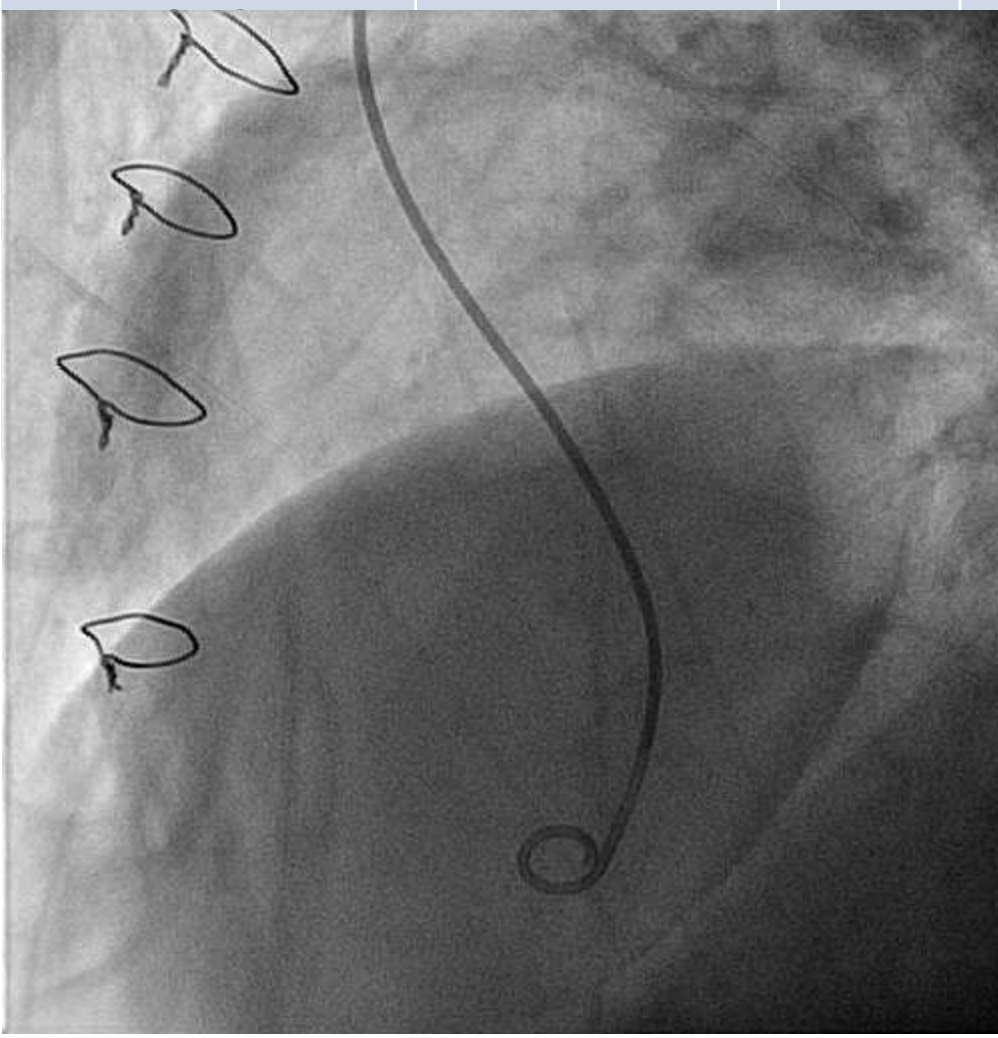
1. mVSD left open
2. Sildenafil 3 mg/kg/day +  
Beraprost 3.6 mg/kg/day

6 MWD increased from 240  
to 360 m

14 years old boy with large PM, PDA. Systemic PA pressure.

# Catheterization 1 year after surgery

	Pressure (mmHg)	O <sub>2</sub> sat (%)		Pressure (mmHg)	O <sub>2</sub> sat (%)
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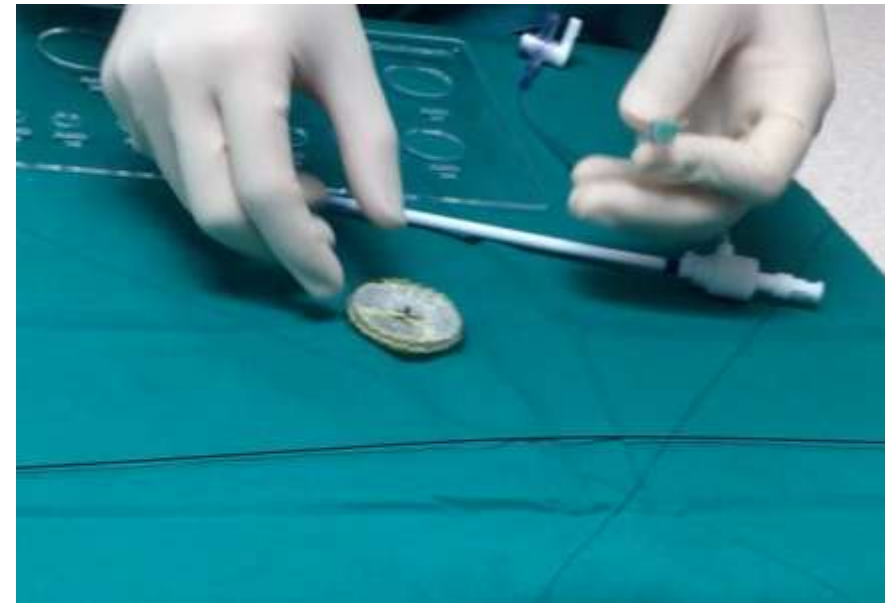
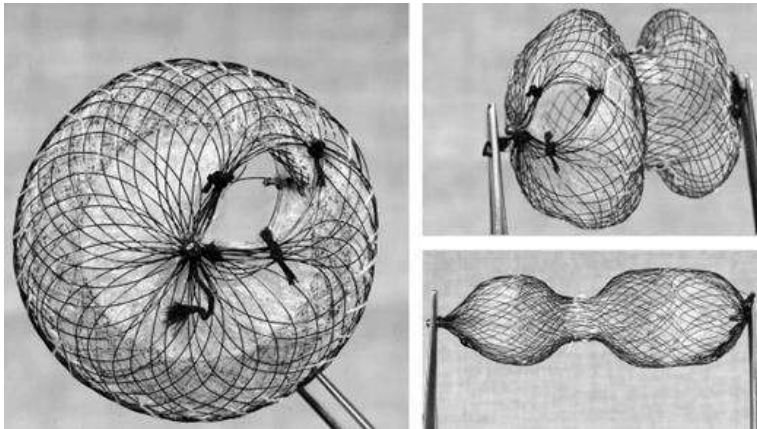
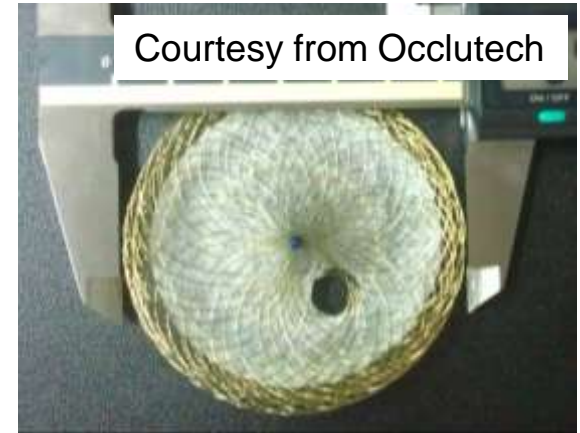


14 years old boy with large PM, PDA. Systemic PA pressure.

# Fenestrated ASD device for PAH-CHD

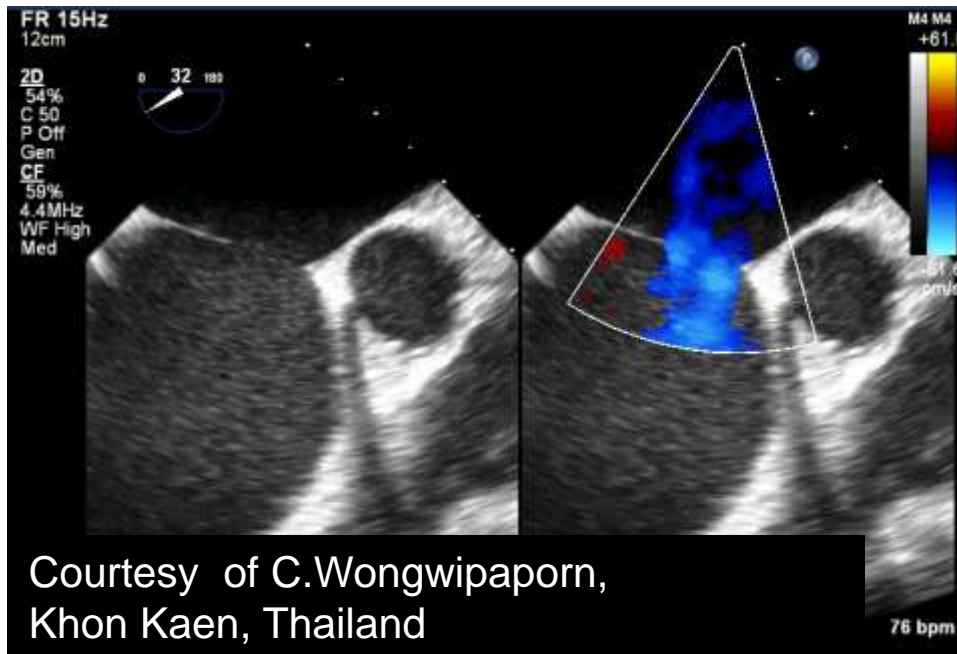
## Indications:

- As for step down approach in ASD with sig. PAH.
- RV decompression in suprasystemic PAH

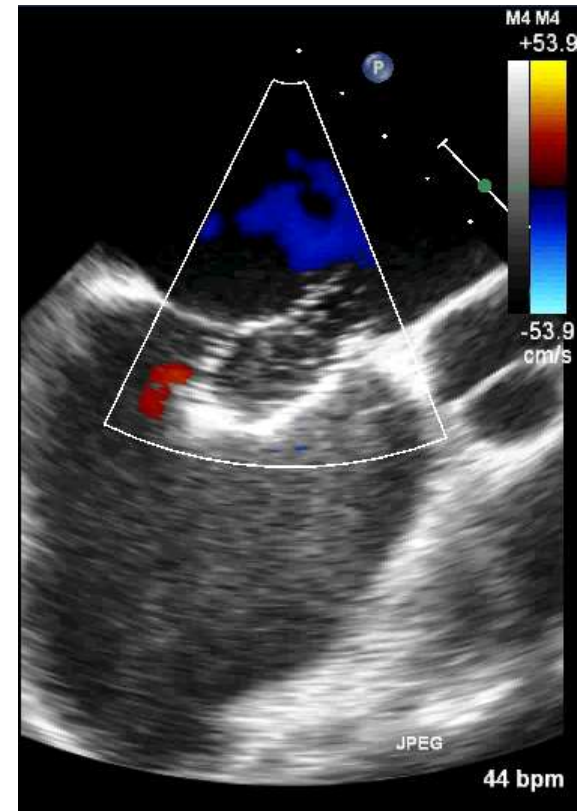


# PAH with coincidental CHD

Test occlusion is the key to elucidate this type of disease



Courtesy of C.Wongwipaporn,  
Khon Kaen, Thailand



small atrial or ventricular septal defects that do not cause severe PAH and follow a course similar to IPAH.



# Post-operative PAH

This includes patients with single ventricle physiology who have undergone bidirectional Glenn or Fontan-type procedures\* .

Cerro MJ, et al. Pulm Circ 2011;1:286–98.

with repaired CHD of any type who develop PHVD.

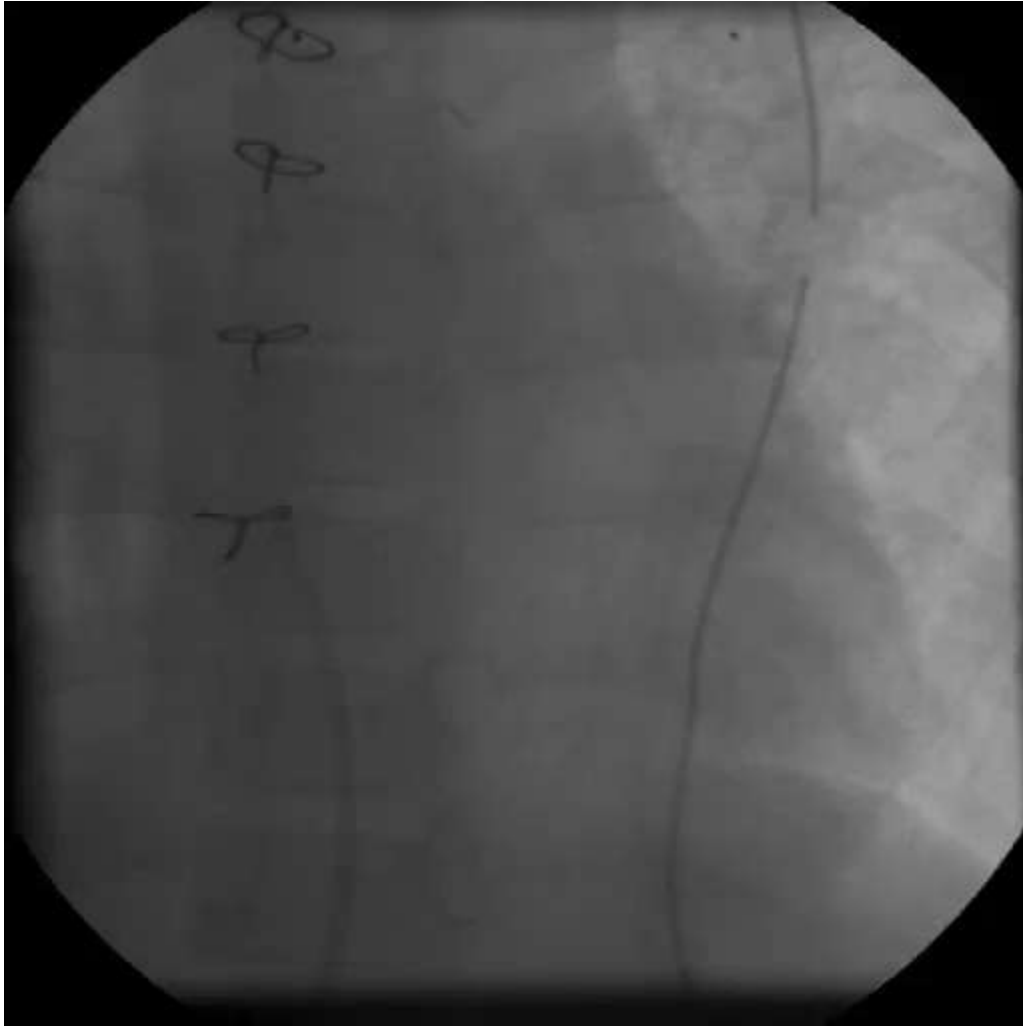


7 yo. S/P Fenestrated  
Fontan with  
intracardiac conduit  
SpO<sub>2</sub> 95%  
Massive pleural  
effusion 1,000 mL/day

CXR: mild pulmonary  
congestion

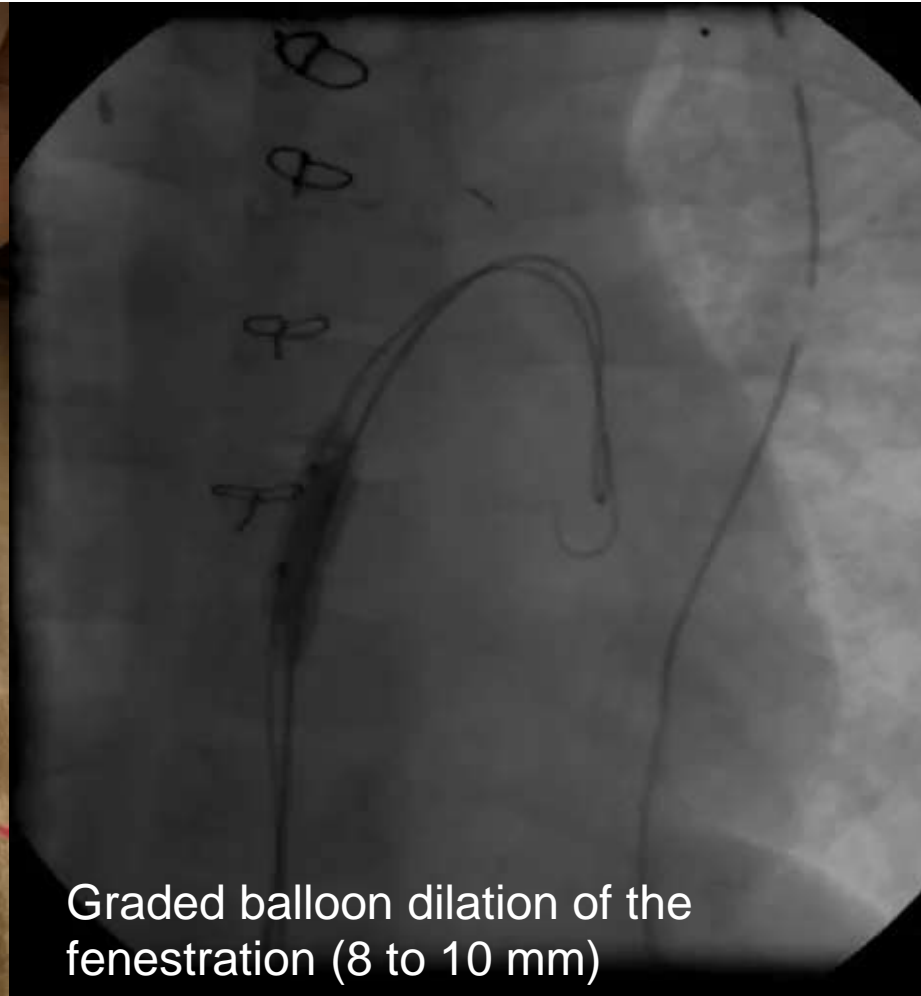
Echo: **Moderate**  
**AVVR**, good  
ventricular systolic  
function, no pathway  
obstruction, **small**  
**fenestration**, mPG 8  
mmHg.

# High Pressure in Fontan Circuit



No pathway obstruction  
mPAP 18 mmHg,  
Atrial pressure 12 mmHg  
Fenestration 4 mm  
Saturation 97% (with oxygen)

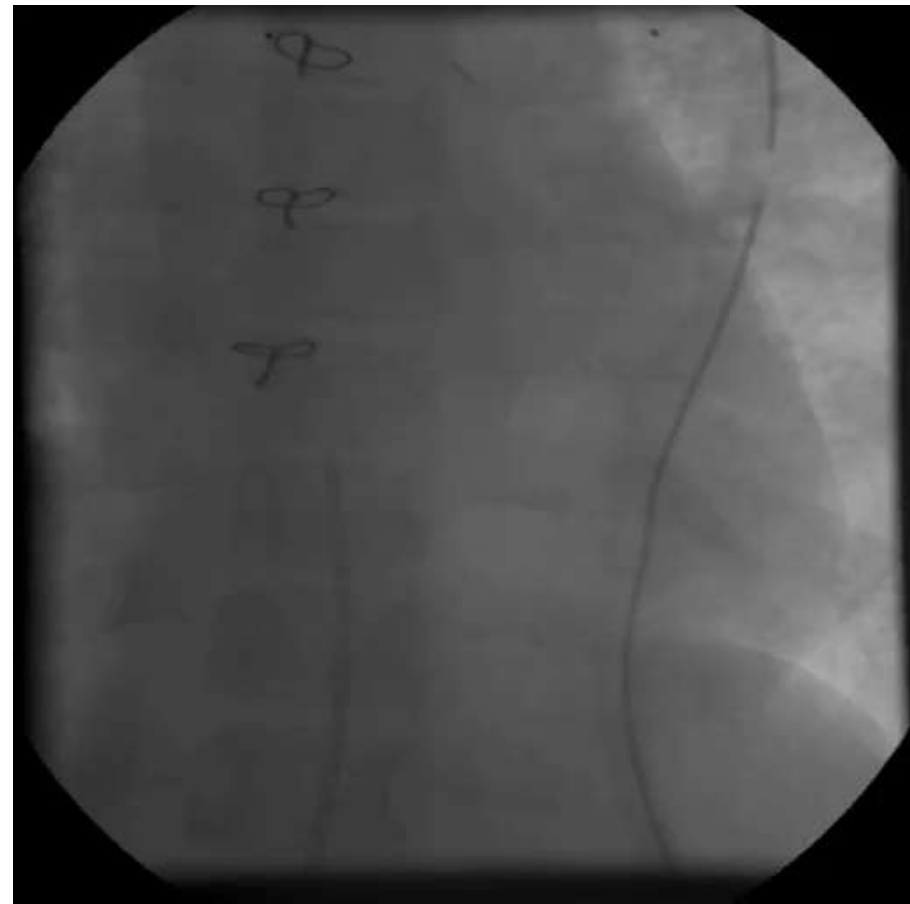
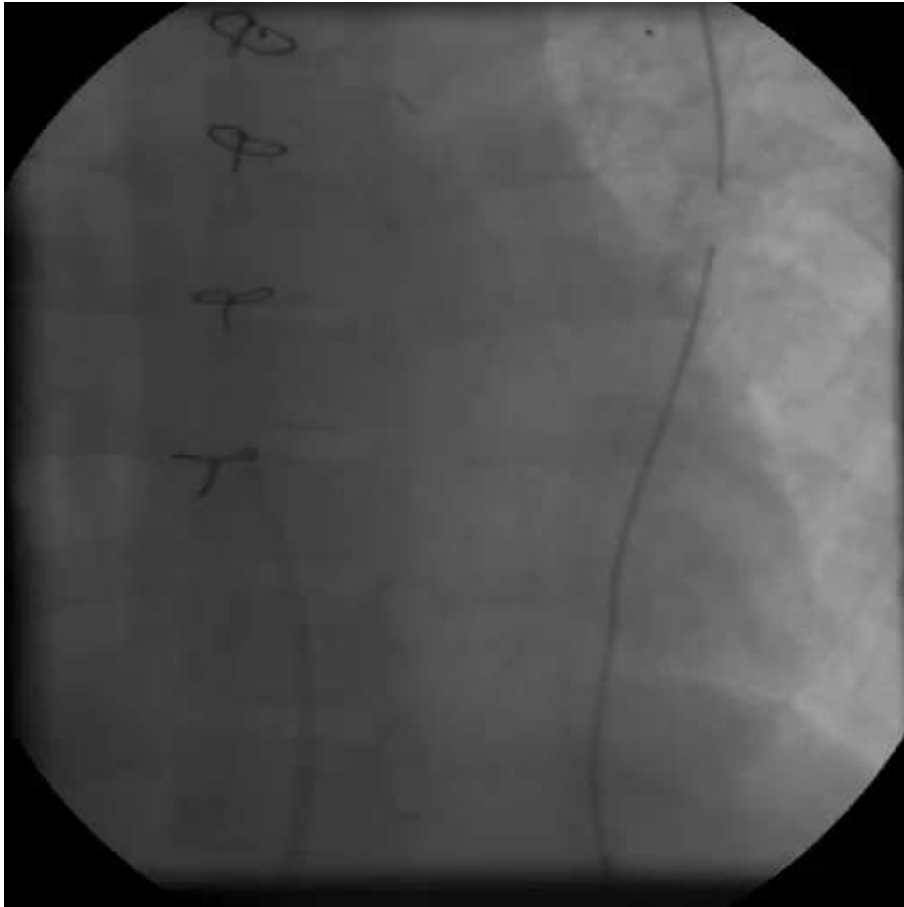
7 yo. S/P Fenestrated Fontan with intracardiac conduit  
Massive pleural effusion



Graded balloon dilation of the fenestration (8 to 10 mm)

7 yo. S/P Fenestrated Fontan with intracardiac conduit  
Massive pleural effusion

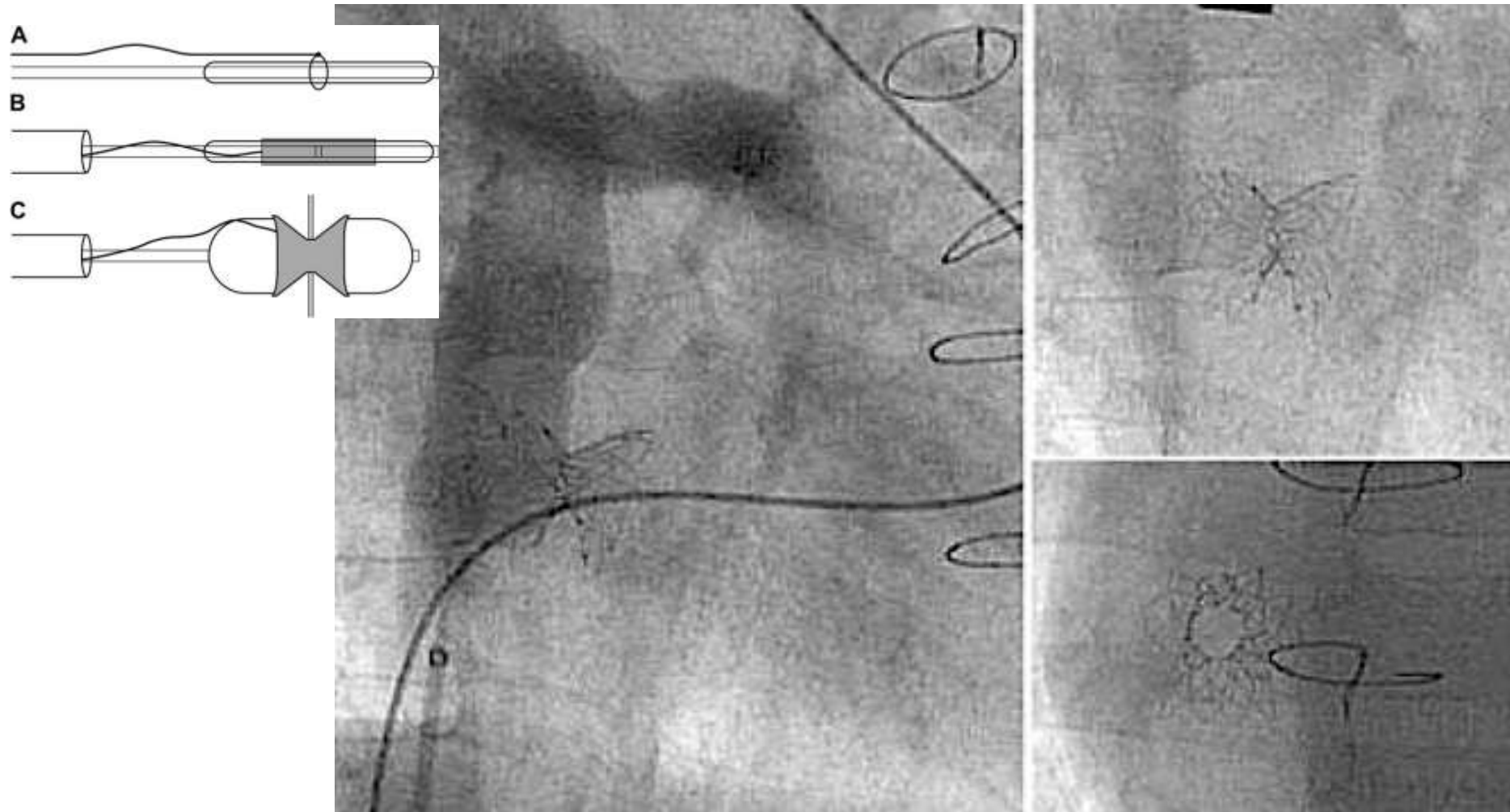
# High Pressure in Fontan Circuit



SpO<sub>2</sub> 89% (room air), mPAP 16 mmHg

7 yo. S/P Fenestrated Fontan with intracardiac conduit  
Massive pleural effusion

# High Pressure in Fontan Circuit



# **Role of Interventional Cardiology**

- Need a comprehensive assessment for decision making.**
- Can be used for palliation or stepwise therapy in complicated cases.**
- It's a high risk procedure.**
- Definitely.... It's a team work approach!**

감사합니다

