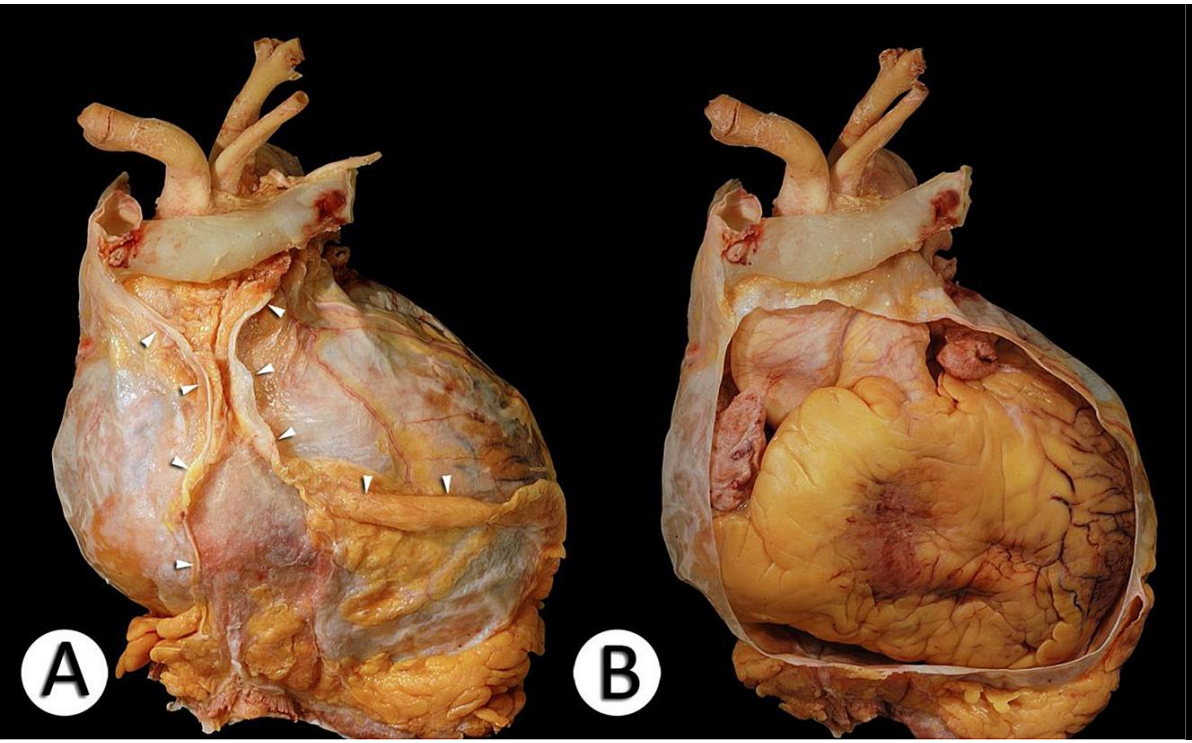
How to evaluate pericardial disease in congenital heart disease

부천세종병원 소아청소년과
최은영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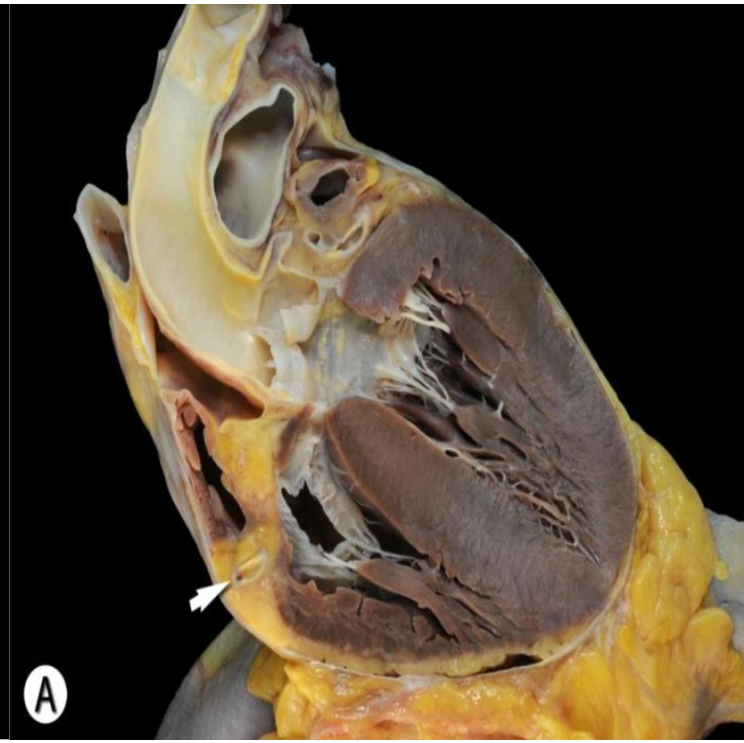
B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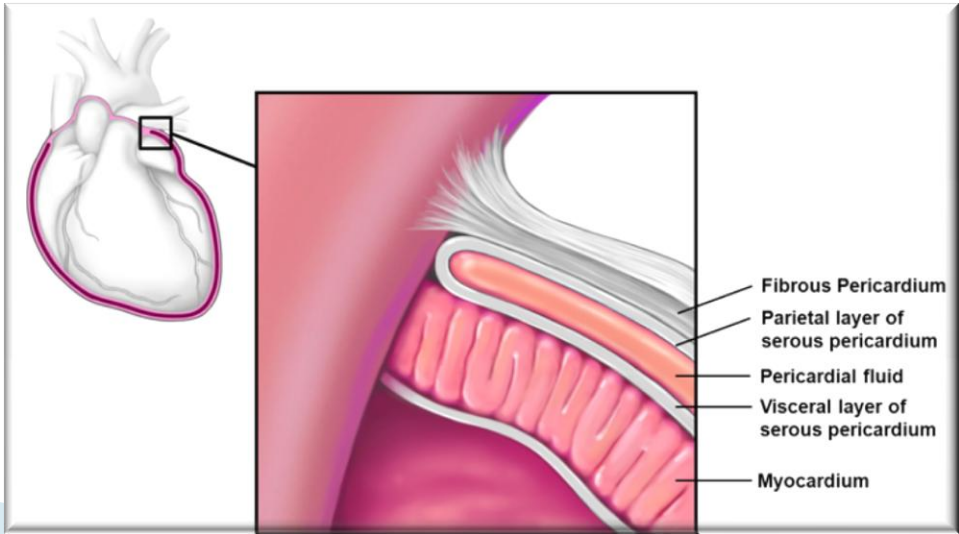
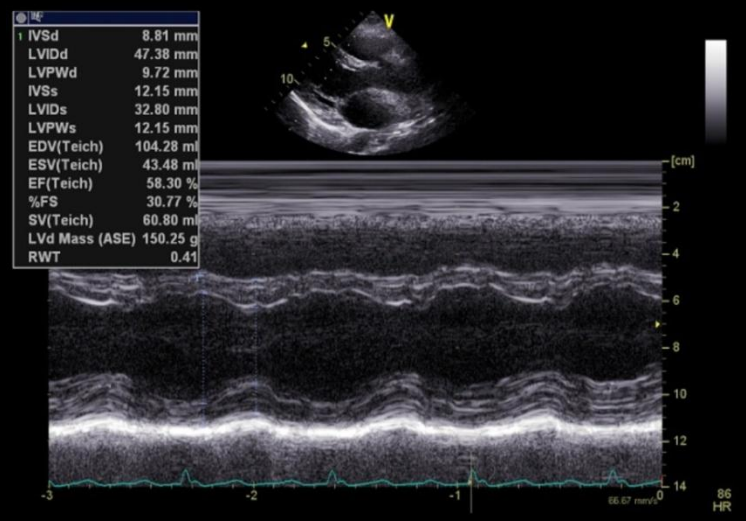
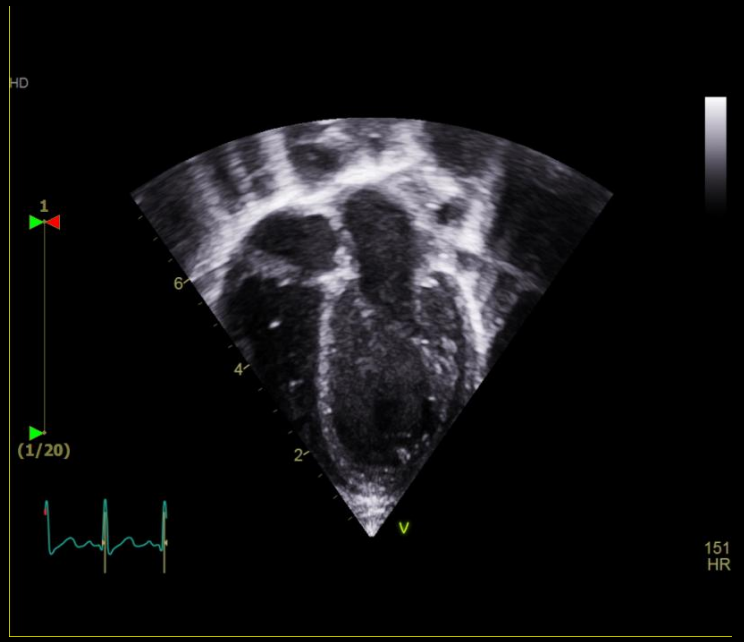
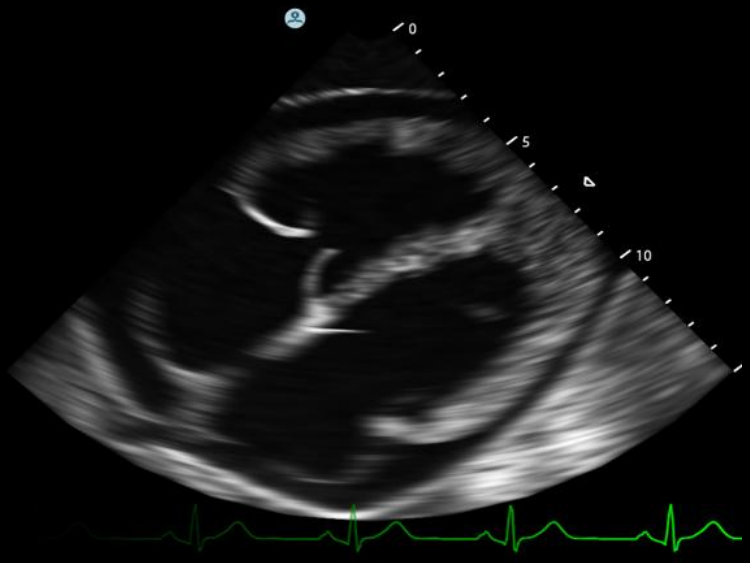
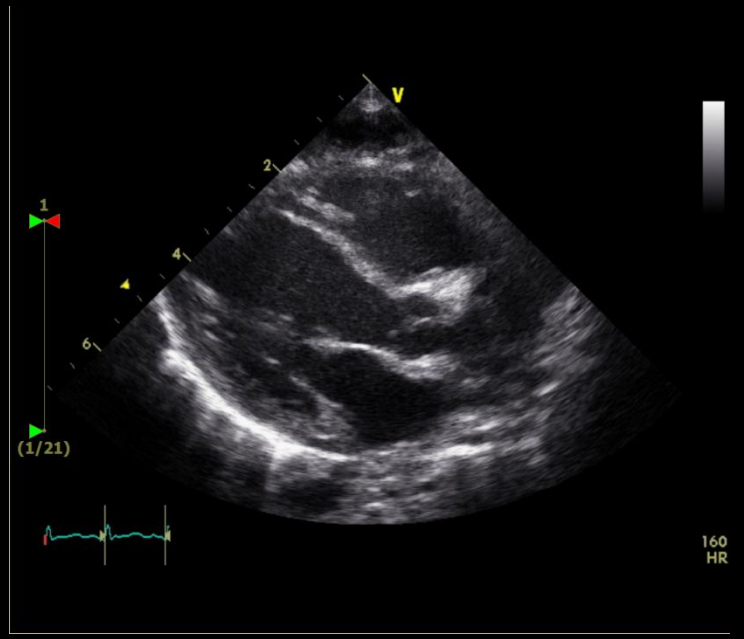
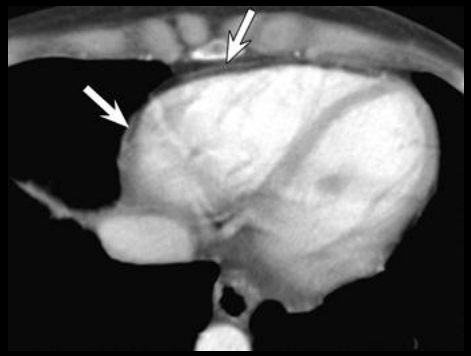

Fibrous Pericardium
Parietal layer of serous pericardium
Pericardial fluid
Visceral layer of serous pericardium
Myocardium

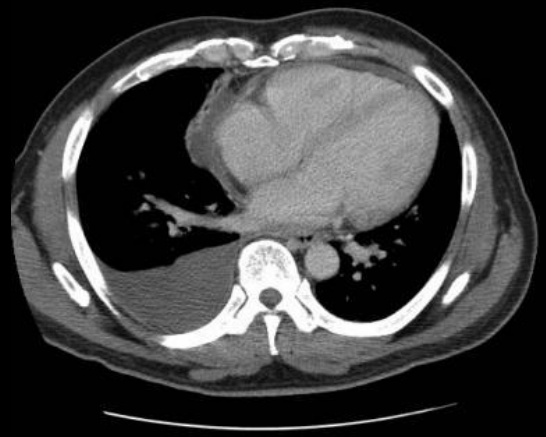

IVSd 8.81 mm
LVIDd 47.38 mm
LVPWd 9.72 mm
IVSs 12.15 mm
LVIDs 32.80 mm
LVPWs 12.15 mm
EDV(Teich) 104.28 ml
ESV(Teich) 43.48 ml
EF(Teich) 58.30 %
%FS 30.77 %
SV(Teich) 60.80 ml
LVd Mass (ASE) 150.25 g
RWT 0.41
[cm]
86
HR

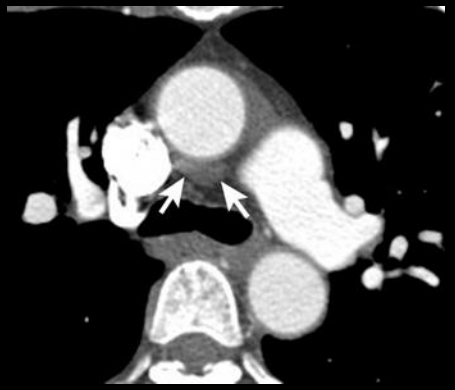

HD
(1/20)
151
HR

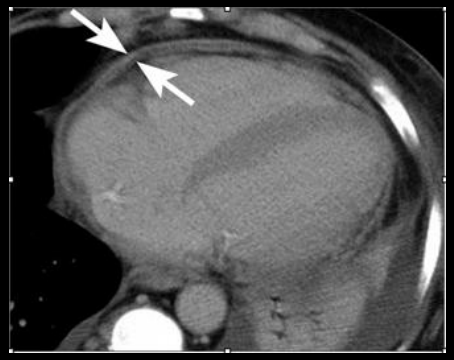


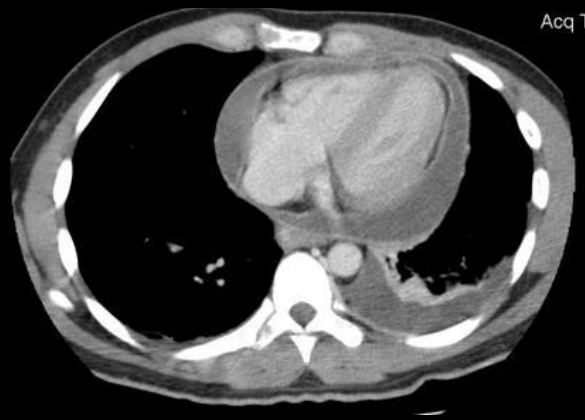

(1/21)
160
HR

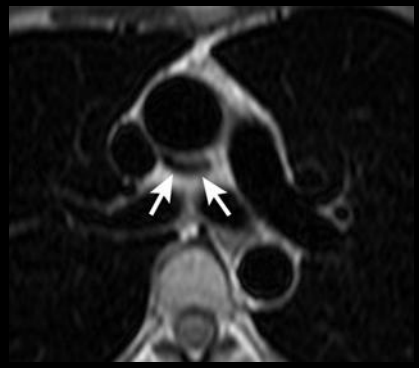


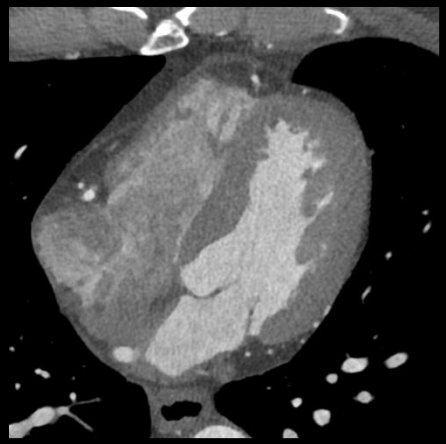


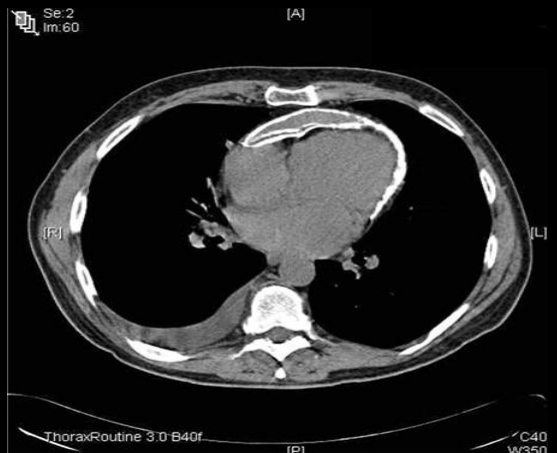


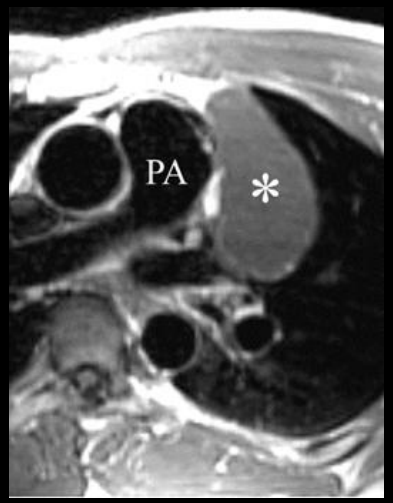


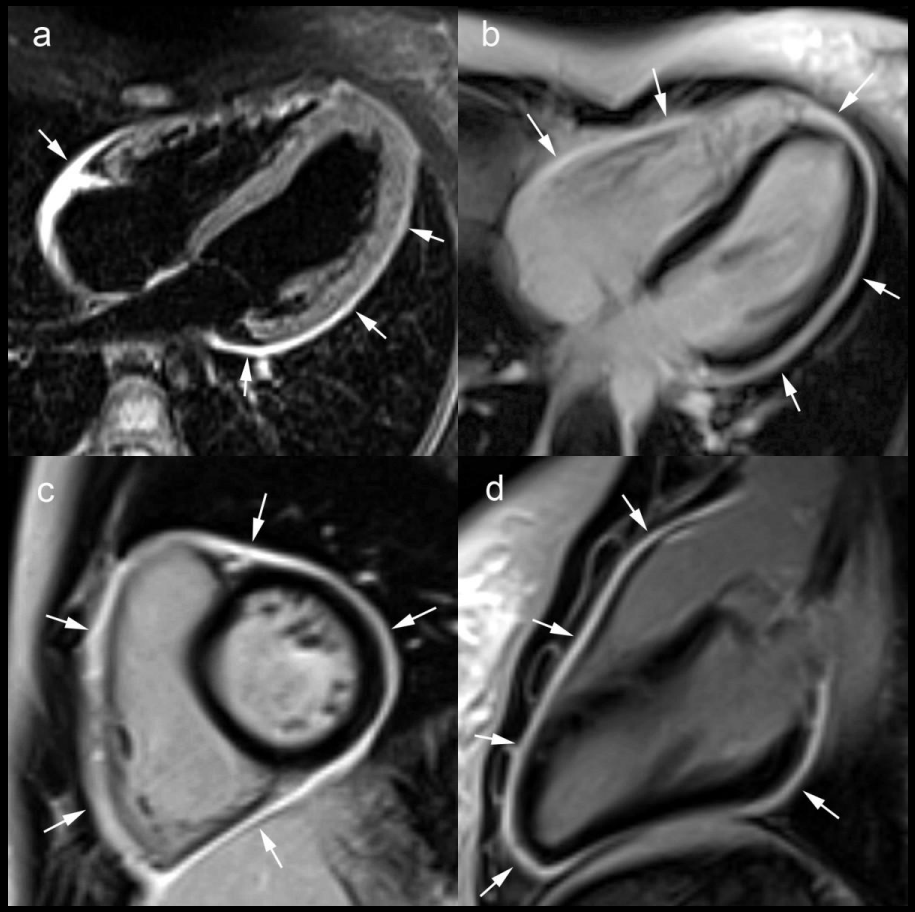

Acq Tm






Se:2
Im:60
[A]
[R]
[L]
ThoraxRoutine 3.0 B40f
[P]
C40
W350


PA
*


a
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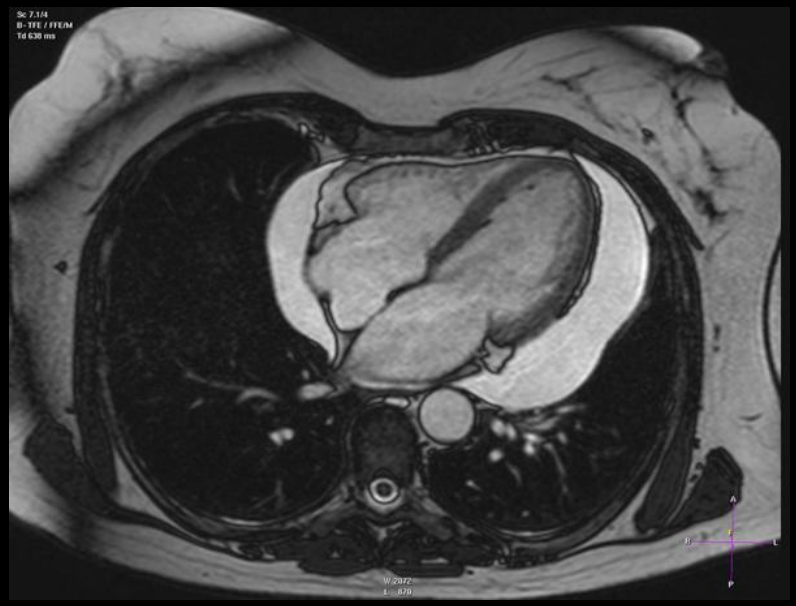


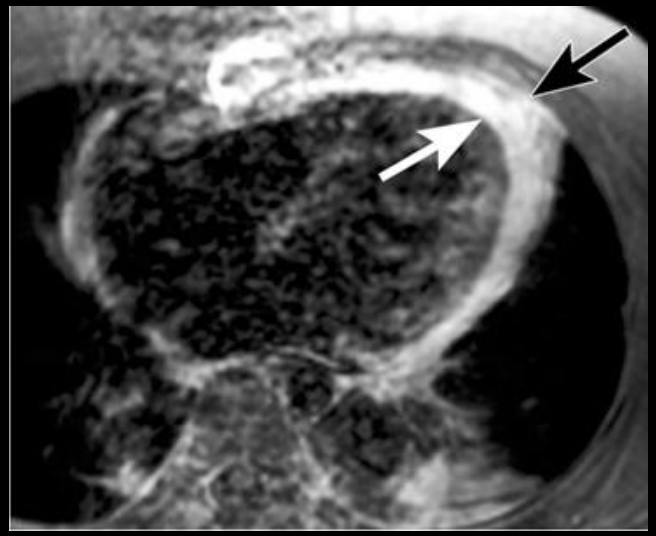
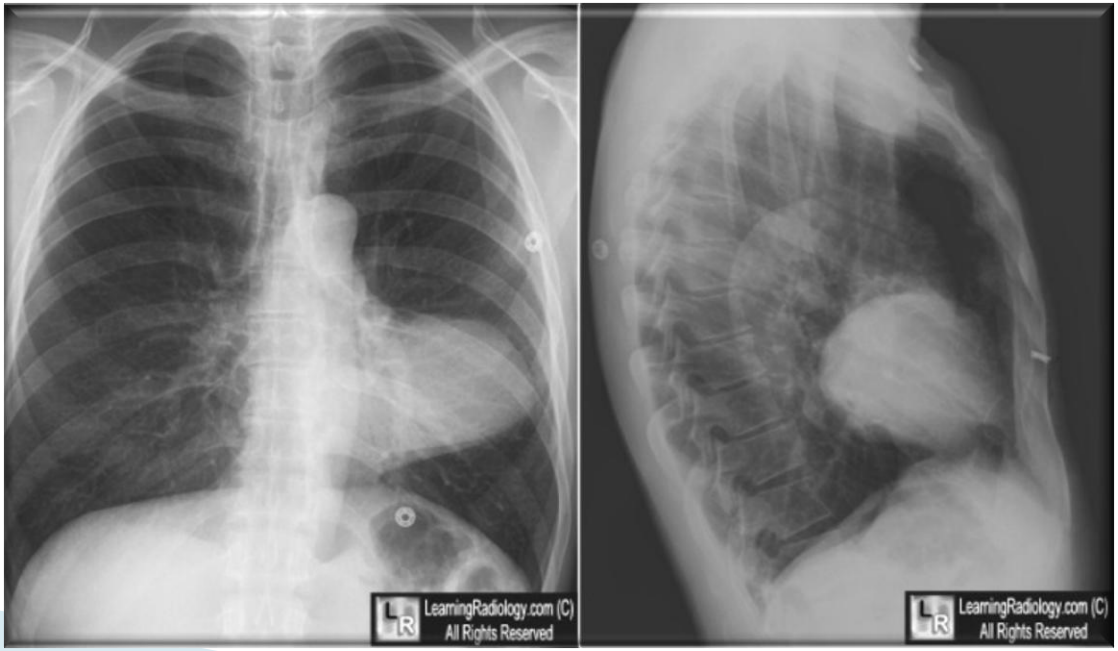


Table 1 Comparison of multimodality imaging modalities in the evaluation of pericardial diseases

	Echocardiography	CT	CMR
Main strengths	• First-line imaging test in the diagnostic evaluation of pericardial disease • Readily available • Low cost • Safe • Can be performed at bedside or urgent situations • Portable • TEE available for better evaluation • High frame rate • Can be performed with respirometer	• Second-line for better anatomic delineation • Evaluation of associated/ extracardiac disease • Preoperative planning • Evaluation of pericardial calcification	• Second-line for better anatomic delineation • Superior tissue characterization • Evaluation of inflammation
Main weaknesses	• Limited windows, narrow field of view • Technically limited with obesity, COPD, or postoperative setting • Relatively operator dependent • Low signal-to-noise ratio of the pericardium • Limited tissue characterization	• Use of ionizing radiation • Use of iodinated contrast • Functional evaluation only possible with retrospective gated studies (higher radiation dose, suboptimal temporal resolution) • Difficulties in case of tachycardia or unstable heart rhythm (particularly for prospective gated studies) • Need for breath-hold • Hemodynamically stable patients only	• Time-consuming, high cost • Preferably stable heart rhythms • Relatively contraindicated in case of pacemaker or ICD • Lung tissue less well visualized • Calcifications not well seen • Use of gadolinium contrast contraindicated in case of advanced renal dysfunction (glomerular filtration rate <30 mL/min) • Use of some breath-hold sequences • Hemodynamically stable patients only

1. Congenital complete or partial absence of the pericardium

- Common cardinal veins supply blood to the pleuropericardial membrane during embryonic development. The right common cardinal vein persists as the superior vena cava which results in closure of the right pleuropericardial membrane. If the left common cardinal vein atrophies early, this could result in defects in left sided pericardium.

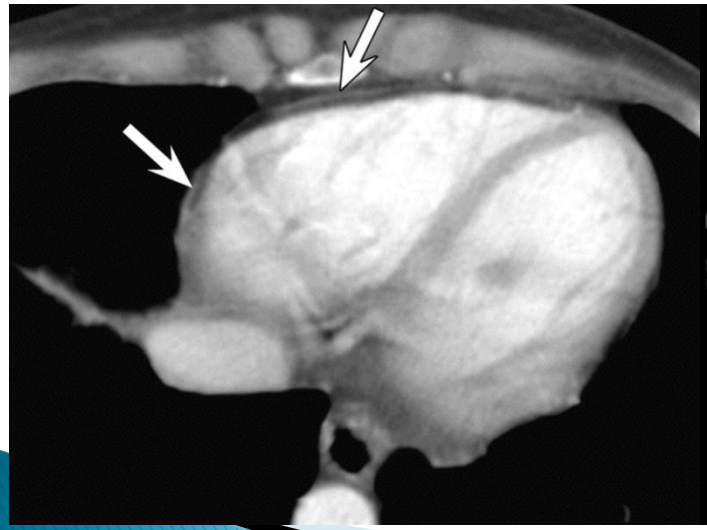


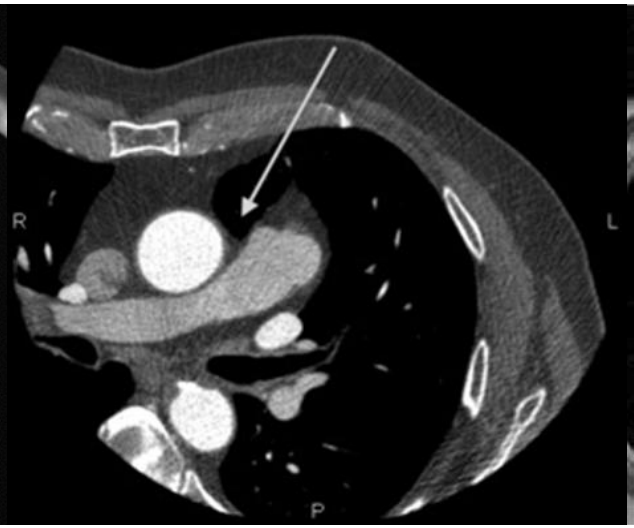
1. Congenital complete or partial absence of the pericardium

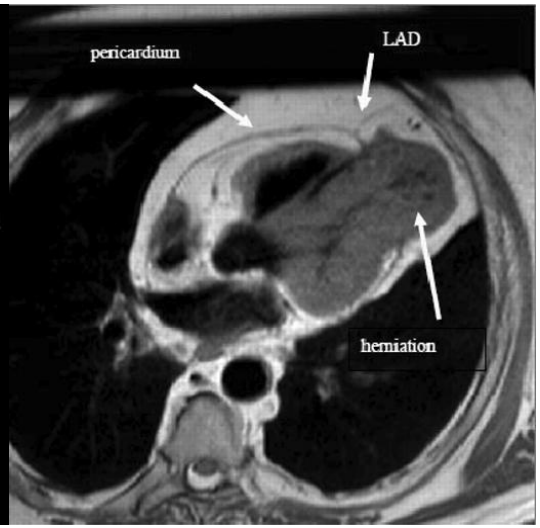
- Right ventricle is visualised more in left parasternal views
- Enlargement of the right ventricle
- Paradoxical septal motion
- Abnormal swinging motion of the heart in the absence of pericardial effusion.
- Can mimic the appearance of right ventricular fluid overload in conditions like atrial septal defect.

1. Congenital complete or partial absence of the pericardium

- CT and MR imaging provide excellent visualization of the pericardium in most patients. The thickness of the normal pericardium, measured on CT scans and on MR images, is less than 2 mm.







2. Pericardial Effusion in pulmonary arterial hypertension

- PAH is a serious condition that can lead to right heart failure and death.
- PAH is mean PA pressure ≥25mmHg at rest, PA occlusion pressure ≤15mmHg, PVR ≥3Wood units.
- Long-term PAH disease management showed that pericardial effusion worsens the prognosis in patients with PAH.

Table 1. Studies describing the prevalence of pericardial effusion in pulmonary arterial hypertension (PAH)

Study	Prevalence of pericardial effusion, proportion (%) of cases	Type of PH, proportion (%) of cases	PH-specific treatment, %
Benza et al.[3]	532/2,105 (25)	IPAH: 1,262/2,716 (47); CTD PAH: 648/2,716 (24); other: 806/2,716 (30)	PA: 42; ERA: 47; PDE-5 I: 50
Hinderliter et al.[6]	43/79 (54)	IPAH: (100)	PA alone: 50; PA plus conventional therapy: 50
Batal et al.[8]	11/72 (15)	IPAH: 4/58 (8); CTD PAH: 7/14 (54)	PA: 20 (8 of 47 cases)
Zhang et al.[11]	45/276 (16)	IPAH: 23/173 (13); CTD PAH: 22/103 (21)	PDE-5 I (IPAH): 79; PA (IPAH): 2; PDE-5 I (CTD PAH): 74; PA (CTD PAH): 1
Eysmann et al.[9]	17/26 (65)	IPAH: (100)	Survivors: CCB in 100; nonsurvivors: CCB in 42
Raymond et al.[10]	42/79 (53)	IPAH: (100)	PA alone: 50; PA plus conventional therapy: 50
Hemnes et al.[45]	6/6 (100)	IPAH: 3/6 (50); CTD PAH: 1/6 (17); other: 2/6 (33)	PA: 67 (4 of 6 cases)
Shimony et al.[12]	81/154 (53)	IPAH: 44/154 (29); CTD PAH: 29/154 (19); other: 8/154 (5)	PA: 42; ERA: 61; PDE-5 I: 28
Park et al.[37]	27/41 (28)	CTEPH: 25/41 (61); PPH: 2/41 (5)	NA
Bossone et al.[38]	8/51 (15.7)	PPH	NA

Pulm Circ. 2013 Sep; 3(3): 467–477.

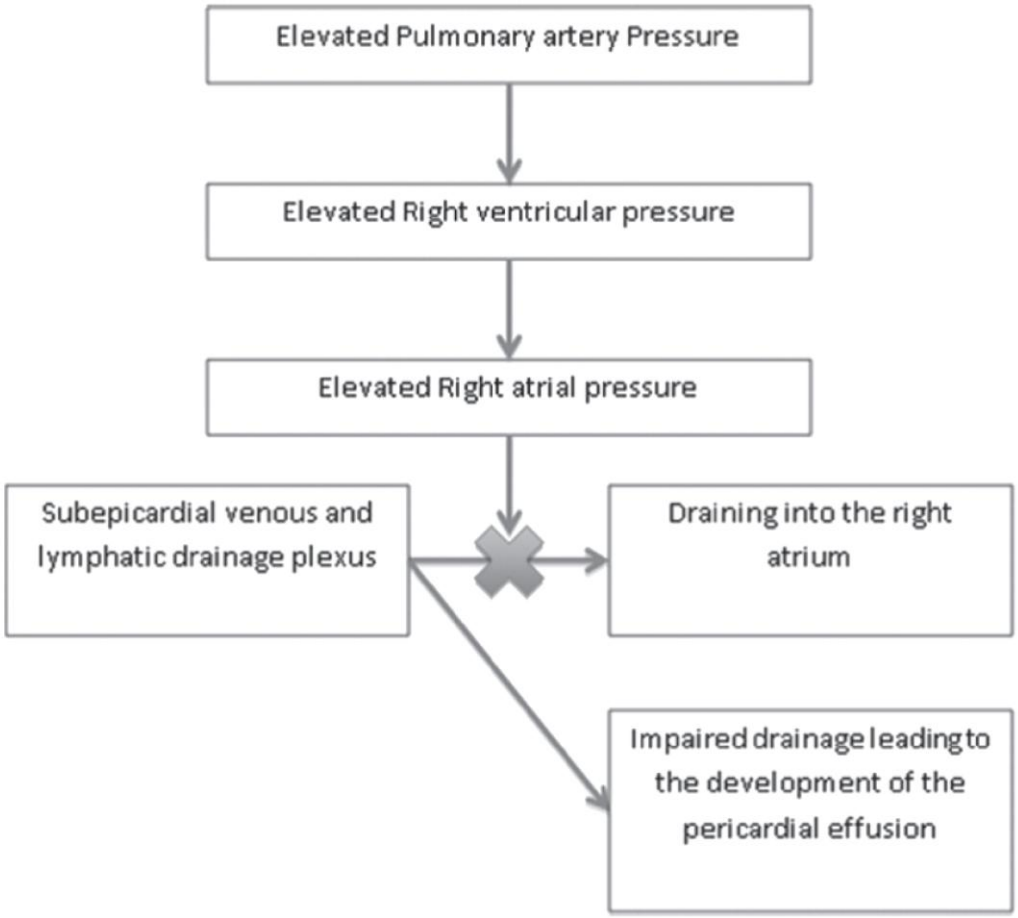
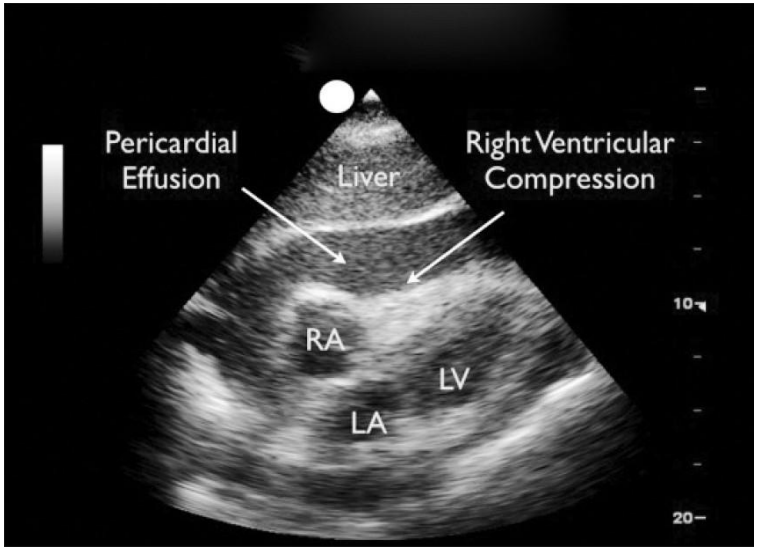
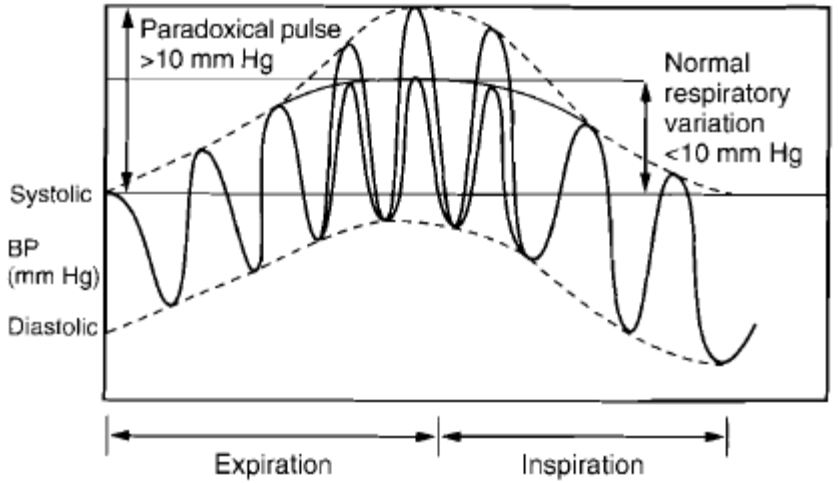
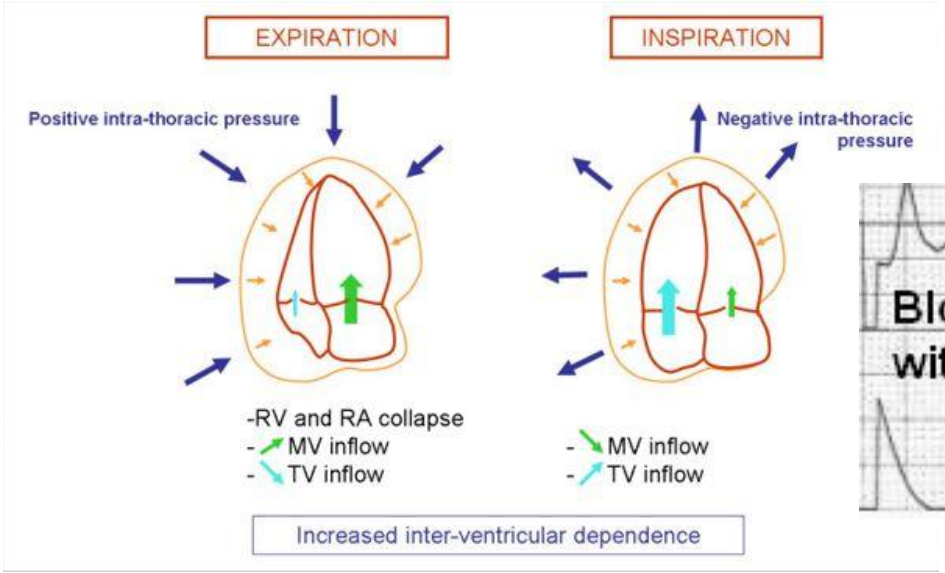


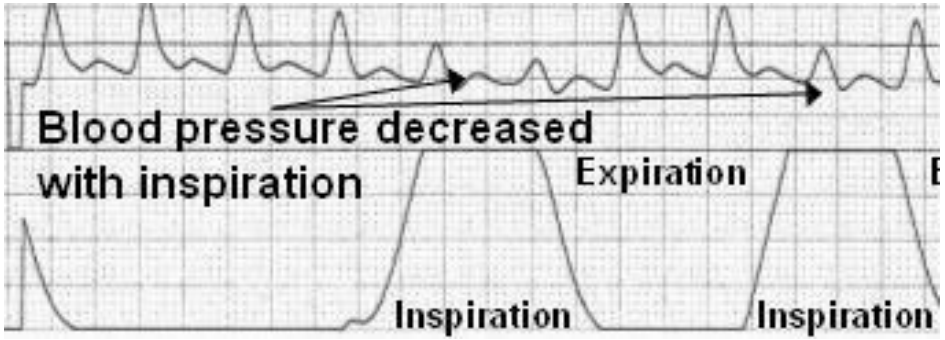
Figure 1. Mechanisms for the development of the pericardial effusion in patients with pulmonary arterial hypertension. A color version of this figure is available in the online edition.

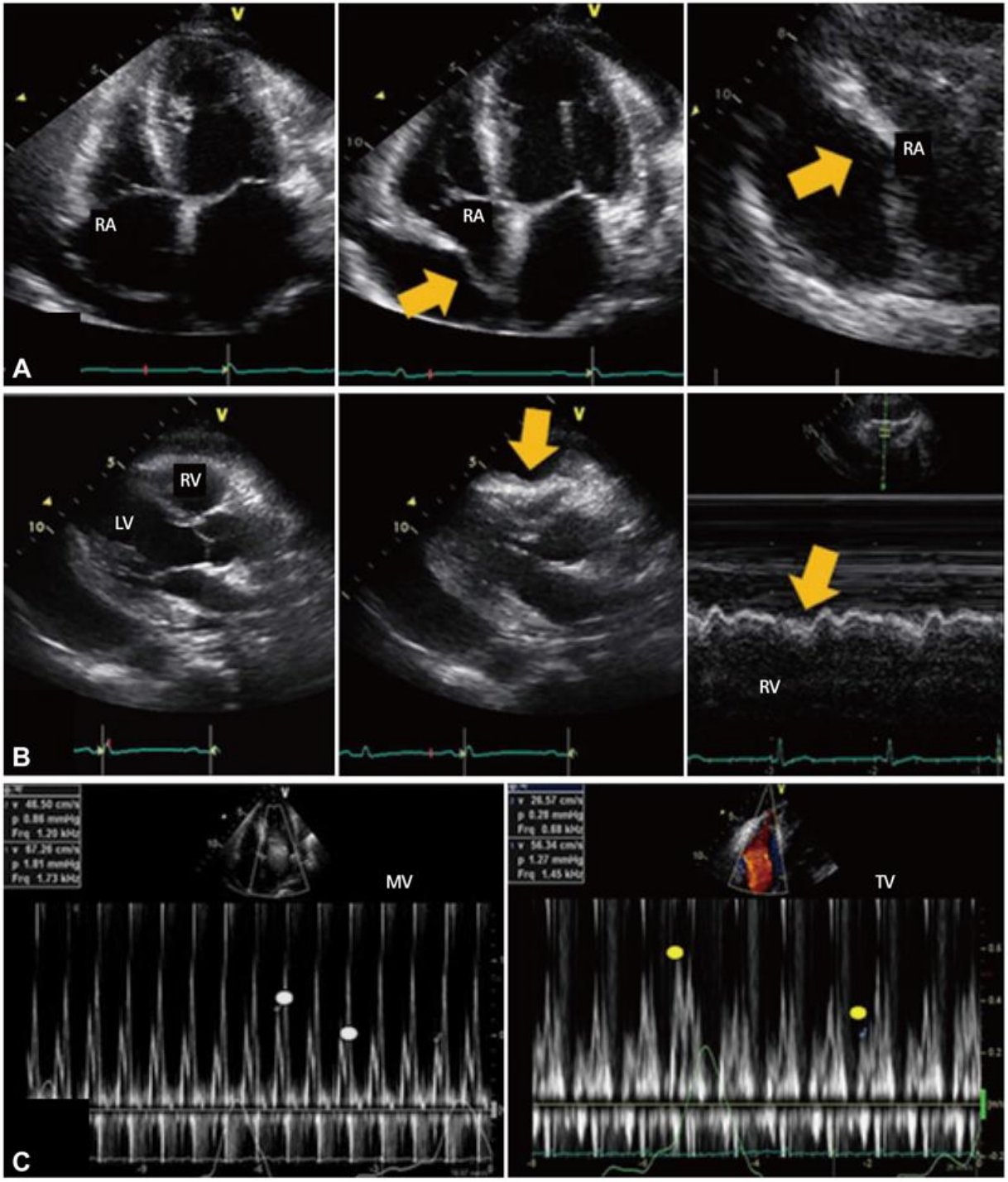
Pulm Circ. 2013 Sep; 3(3): 467–477.

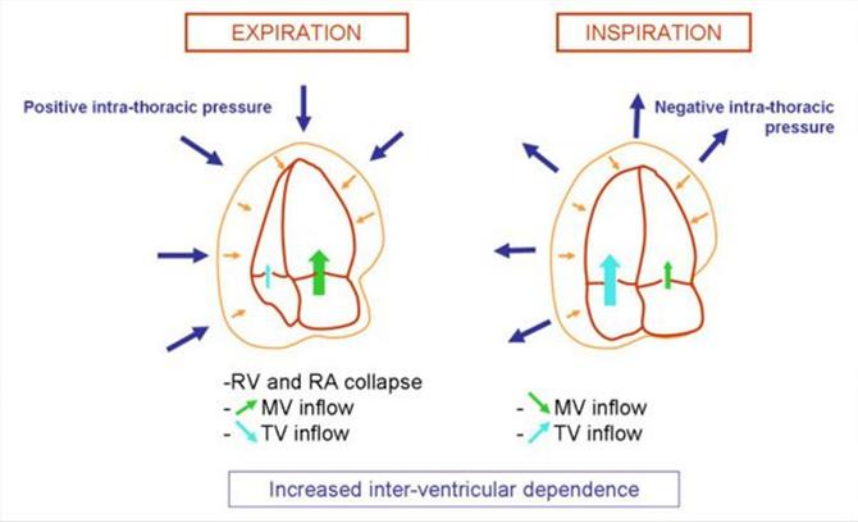


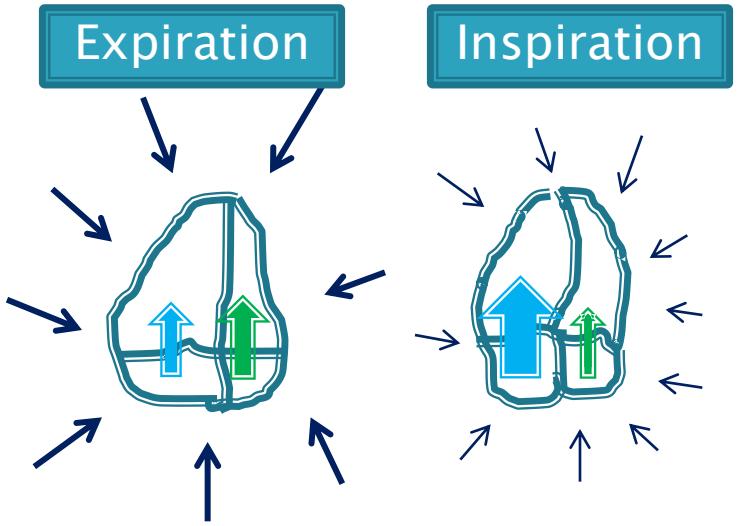


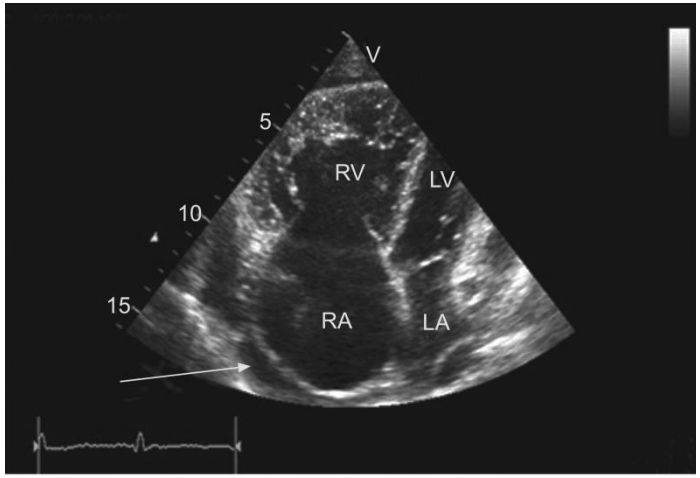


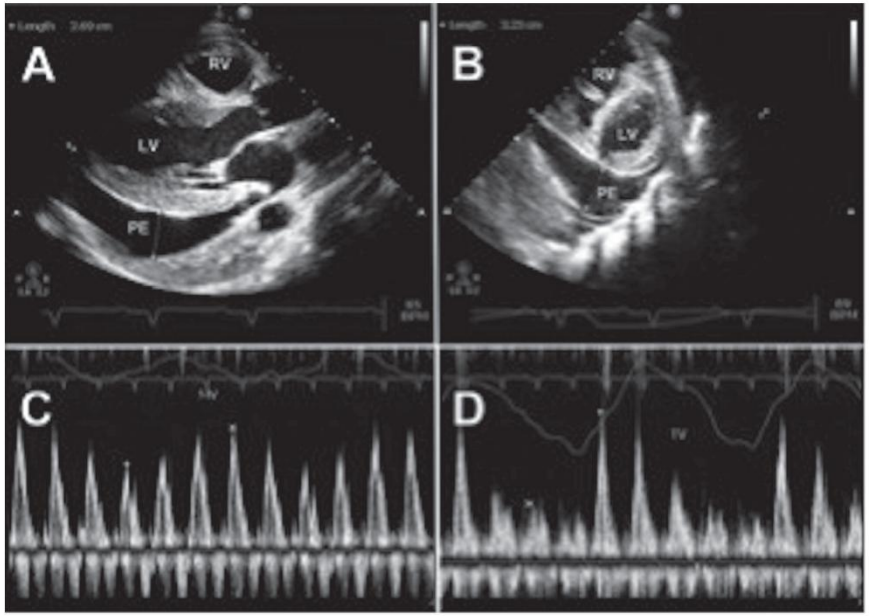



RA
RA
RA
A
RV
LV
RV
B
MV
TV
C

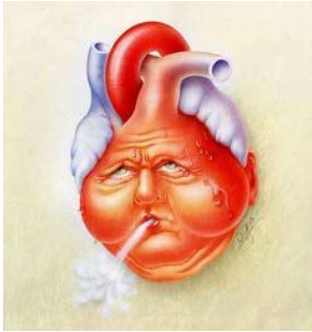

EXPIRATION
INSPIRATION
Positive intra-thoracic pressure
Negative intra-thoracic pressure
-RV and RA collapse
- MV inflow
- TV inflow
- MV inflow
- TV inflow
Increased inter-ventricular dependence

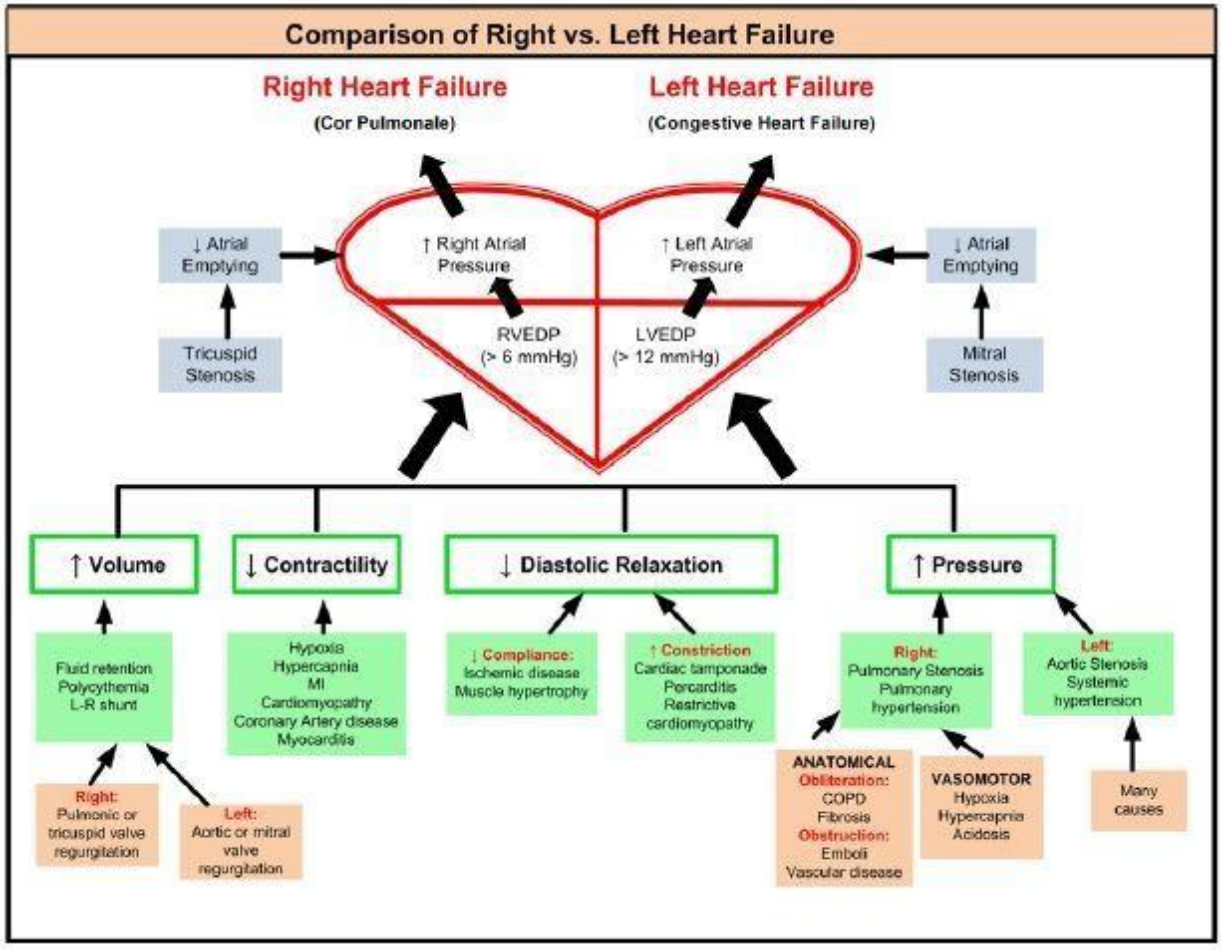

Expiration
Inspiration

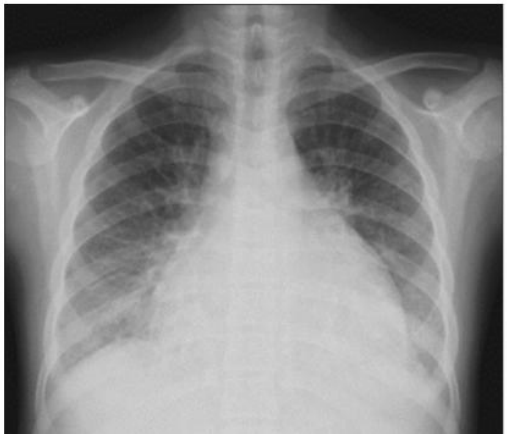

V
5
10
15
RV
LV
RA
LA

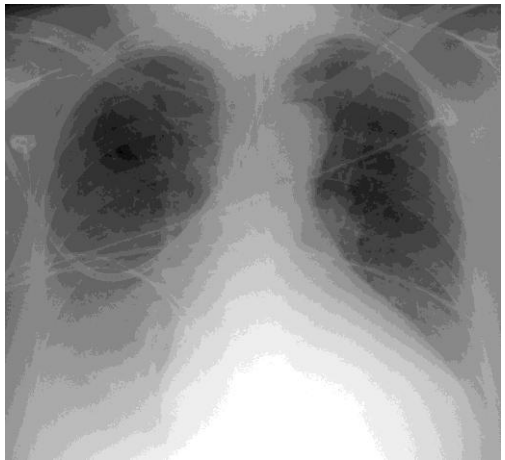

A
RV
LV
PE
B
RV
LV
PE
C
D

3. Pericardial and pleural effusion in congestive heart failure









4. Postpericardiotomy synsrome

- Postpericardiotomy syndrome (PPS) is a febrile illness secondary to an inflammatory reaction involving the pleura and pericardium.
- It is more common in patients who have undergone surgery that involves opening the pericardium.
- Its frequency varies from 2% to 30%.
- The syndrome is also characterized by pericardial or pleuritic pain, friction rubs, pleural effusions, pneumonitis, and abnormal ECG and radiography findings.
- It is believed that PPS results from a heightened immune response to injury following cardiothoracic surgery .

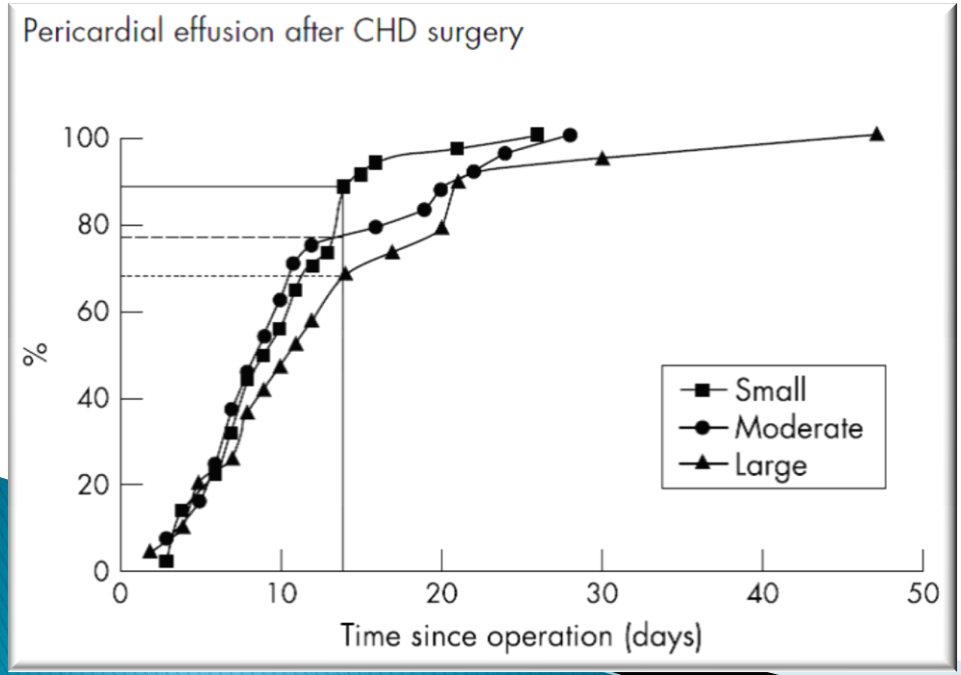
4. Postpericardiotomy synsrome

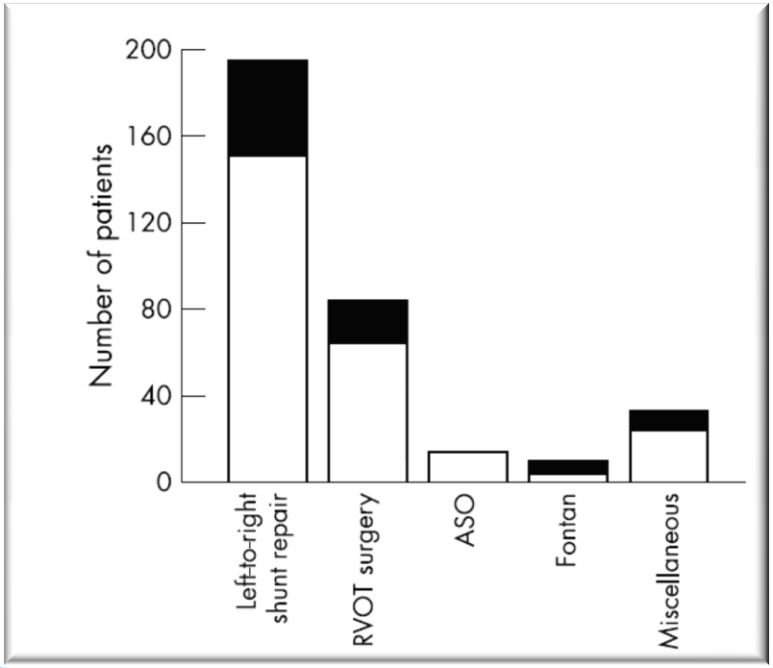
CONGENITAL HEART DISEASE

Pericardial effusion after open heart surgery for congenital heart disease

E W Y Cheung, S A Ho, K K Y Tang, A K T Chau, C S W Chiu, Y F Cheung

Heart 2003;**89**:780–783





European Heart Journal (2010) **31**, 2749–2754
doi:10.1093/eurheartj/ehq319

FASTTRACK
ESC CLINICAL TRIAL UPDATE

COlchicine for the Prevention of the Post-pericardiotomy Syndrome (COPPS): a multicentre, randomized, double-blind, placebo-controlled trial

Massimo Imazio[1]*, Rita Trinchero[1], Antonio Brucato[2], Maria Elena Rovere[3], Anna Gandino[4], Roberto Cemin[5], Stefania Ferrua[6], Silvia Maestroni[2], Edoardo Zingarelli[3], Alberto Barosi[4], Caterina Simon[2], Fabrizio Sansone[3], Davide Patrini[4†], Ettore Vitali[4†], Paolo Ferrazzi[2], David H. Spodick[7], and Yehuda Adler[8], on behalf of the COPPS Investigators

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Received 19 July 2010; revised 8 August 2010; accepted 10 August 2010; online publish-ahead-of-print 30 August 2010

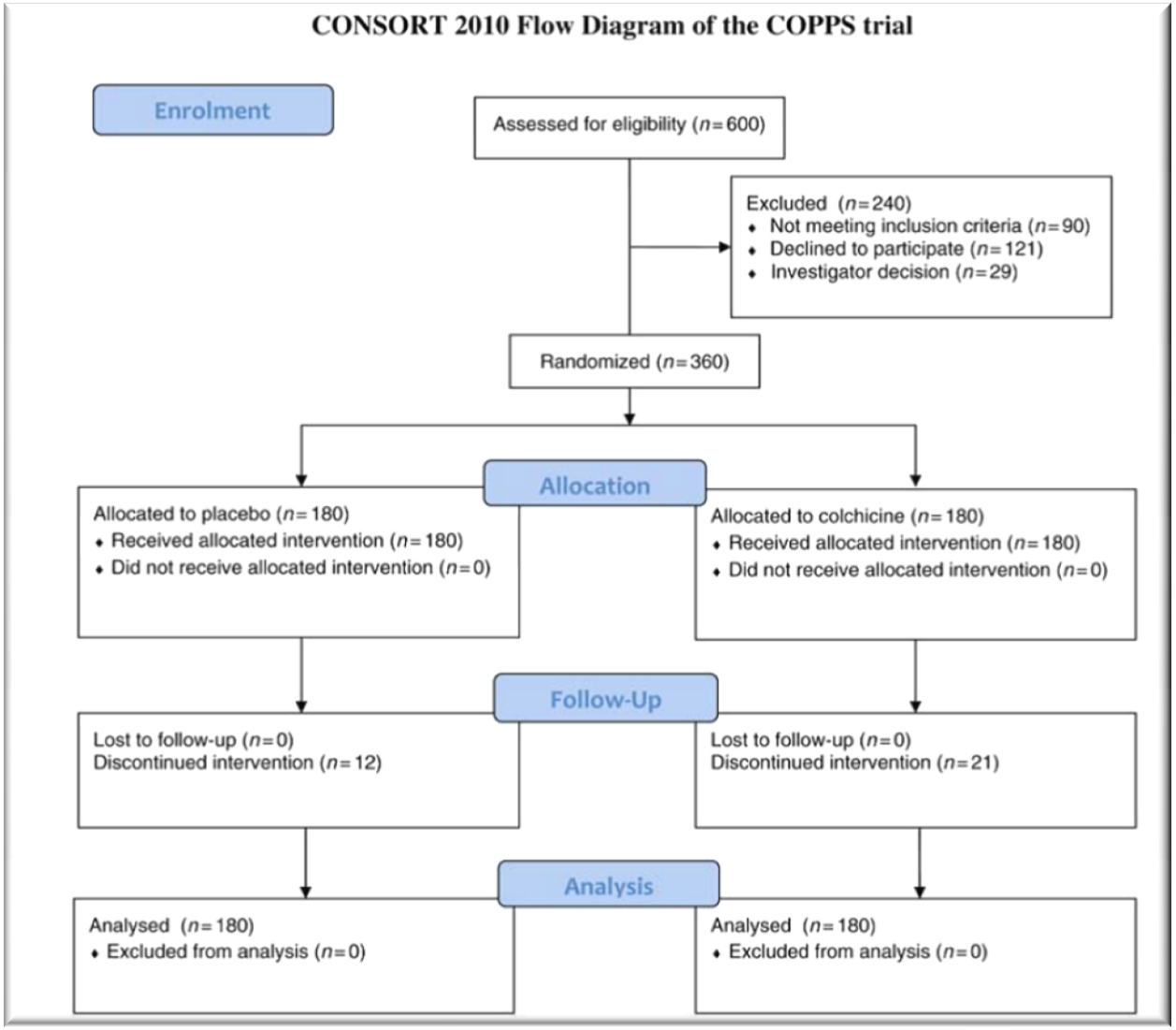
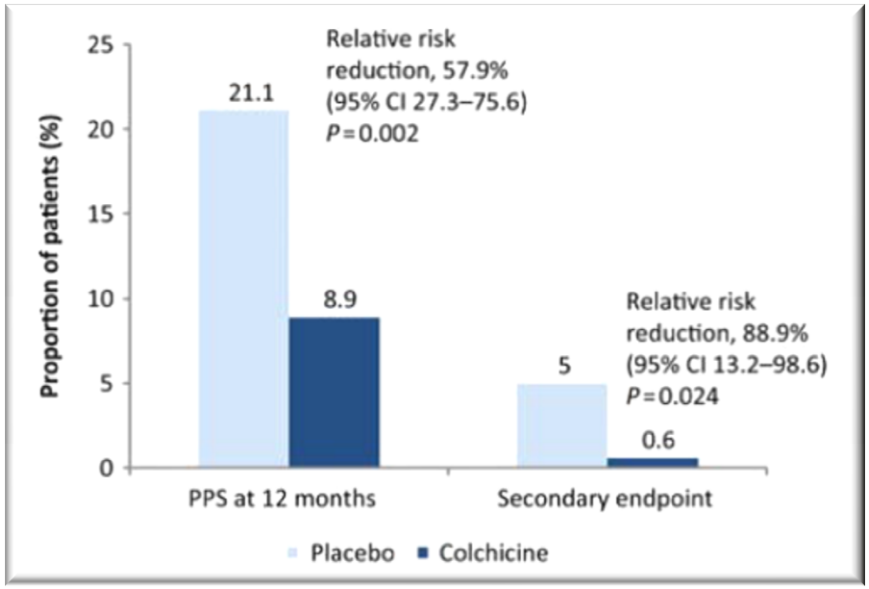

CONSORT 2010 Flow Diagram of the COPPS trial
Enrolment
Assessed for eligibility (n=600)
Excluded (n=240)
Not meeting inclusion criteria (n=90)
Declined to participate (n=121)
Investigator decision (n=29)
Randomized (n=360)
Allocation
Allocated to placebo (n=180)
Received allocated intervention (n=180)
Did not receive allocated intervention (n=0)
Allocated to colchicine (n=180)
Received allocated intervention (n=180)
Did not receive allocated intervention (n=0)
Follow-Up
Lost to follow-up (n=0)
Discontinued intervention (n=12)
Lost to follow-up (n=0)
Discontinued intervention (n=21)
Analysis
Analysed (n=180)
Excluded from analysis (n=0)
Analysed (n=180)
Excluded from analysis (n=0)

Table 3 Study endpoints and diagnostic criteria for the post-pericardiotomy syndrome

Event	Placebo (n = 180)	Colchicine (n = 180)	P-value
Primary endpoint (%)			
PPS at 12 months	38 (21.1)	16 (8.9)	0.002
Fever beyond first post-operative week[a]	7 (3.9)	6 (3.3)	0.982
Pleuritic chest pain	23 (12.8)	7 (3.9)	0.004
Friction rub	15 (8.3)	5 (2.7)	0.036
Pleural effusion	46 (25.6)	22 (12.2)	0.002
New or worsening pericardial effusion[b]	41 (22.8)	23 (12.8)	0.019
Secondary endpoint[c] (%)	9 (5.0)	1 (0.6)	0.024
Recurrence	2 (1.1)	0 (0.0)	0.485
Cardiac tamponade	1 (0.6)	0 (0.0)	0.992
Constrictive pericarditis	0 (0.0)	0 (0.0)	0.982
PPS-related hospitalization	6 (3.3)	1 (0.6)	0.130
Mean follow-up (months)	18.5	20.2	0.252



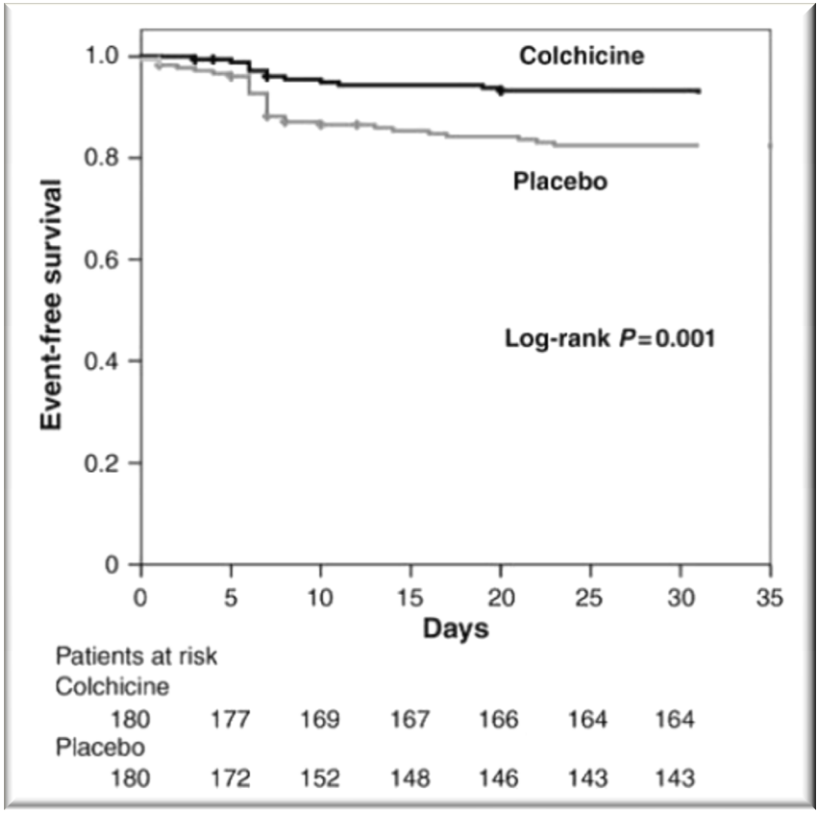


Table 4 Side effects and drug withdrawal

Event	Placebo (n = 180)	Colchicine (n = 180)	P-value
Side effects (%)	9 (5.0)	16 (8.9)	0.212
Severe side effects	0 (0.0)	0 (0.0)	0.212
Other side effects	9 (5.0)	16 (8.9)	0.133
Gastrointestinal	8 (4.4)	16 (8.9)	0.939
Alopecia	0 (0.0)	0 (0.0)	
Anorexia	0 (0.0)	0 (0.0)	
Hepatoxicity	0 (0.0)	0 (0.0)	
Myotoxicity	1 (0.6)	0 (0.0)	
Bone marrow toxicity	0 (0.0)	0 (0.0)	
Other	0 (0.0)	0 (0.0)	
Drug withdrawal (%)			
Overall	12 (6.7)	21 (11.7)	0.145
Related to side effects	9 (5.0)	16 (8.9)	0.212
Patient or medical decision	3 (1.7)	5 (2.8)	0.728

Research

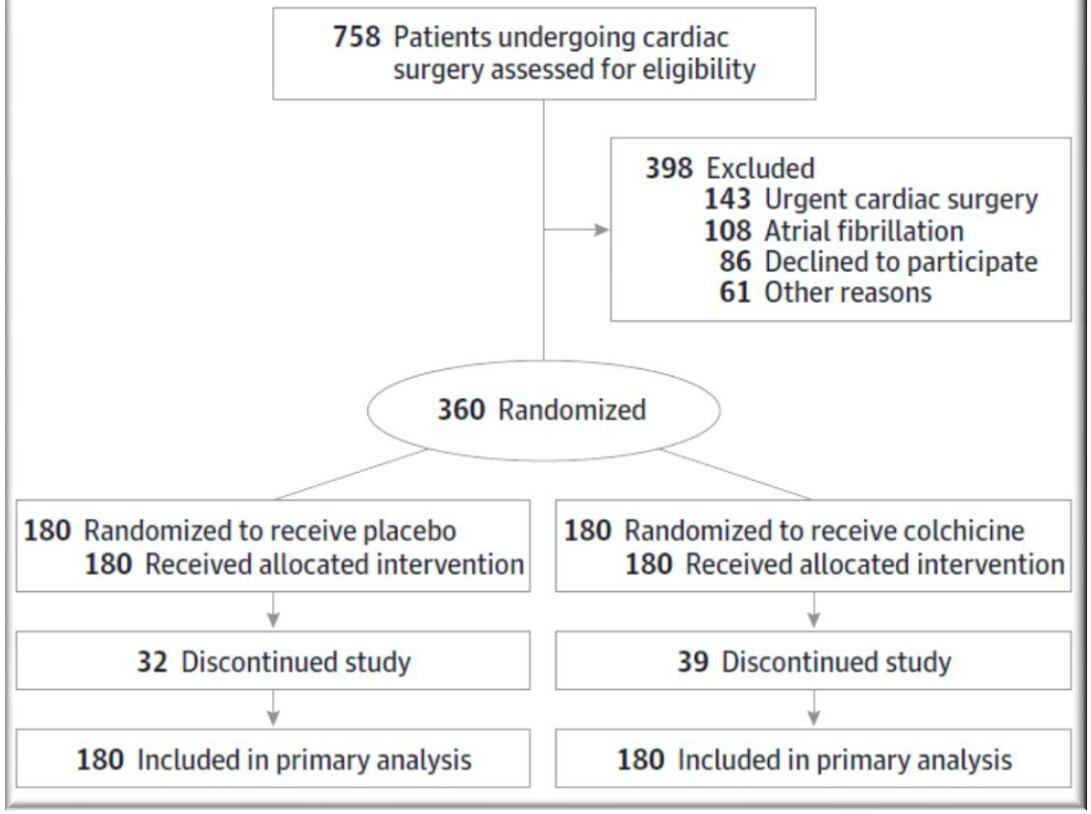
Original Investigation

Colchicine for Prevention of Postpericardiotomy Syndrome and Postoperative Atrial Fibrillation
The COPPS-2 Randomized Clinical Trial

Massimo Imazio, MD; Antonio Brucato, MD; Paolo Ferrazzi, MD; Alberto Pullara, MD,; Yehuda Adler, MD; Alberto Barosi, MD; Alida L. Caforio, MD; Roberto Cemin, MD; Fabio Chirillo, MD; Chiara Comoglio, MD; Diego Cugola, MD; Davide Cumetti, MD; Oleksandr Dyrda, MD; Stefania Ferrua, MD; Yaron Finkelstein, MD; Roberto Flocco, MD; Anna Gandino, MD; Brian Hoit, MD; Francesco Innocente, MD; Silvia Maestroni, MD; Francesco Musumeci, MD; Jae Oh, MD; Amedeo Pergolini, MD; Vincenzo Polizzi, MD; Arsen Ristić, MD; Caterina Simon, MD; David H Spodick, MD; Vincenzo Tarzia, MD; Stefania Trimboli, MD; Anna Valenti, MD; Riccardo Belli, MD; Fiorenzo Gaita, MD; for the COPPS-2 Investigators

JAMA. 2014;312(10):1016-1023. doi:10.1001/jama.2014.11026

Figure 1. Screening, Enrollment, Randomization, and Follow-up of Study Participants in the COPPS-2 Trial

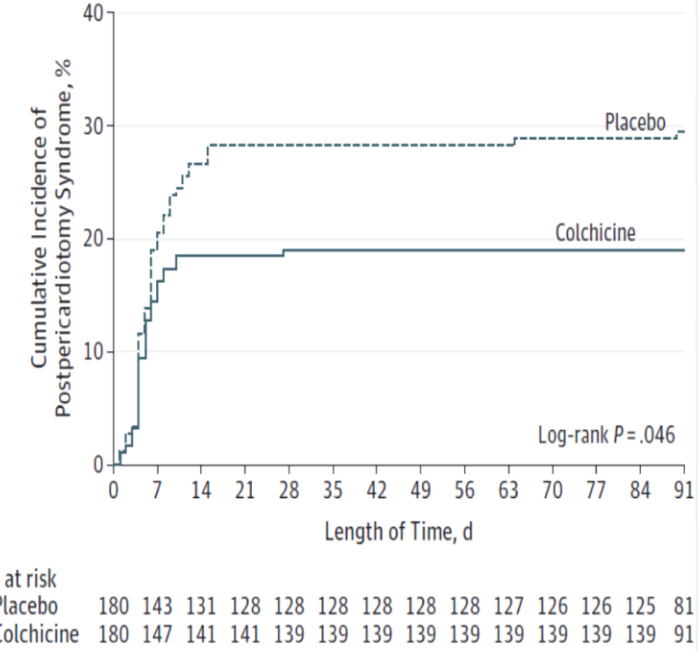


JAMA. 2014;312(10):1016-1023. doi:10.1001/jama.2014.11026

Table 2. Primary and Secondary Study Outcomes at 3-Month Follow-up According to Treatment Assignment

Outcomes	No. (%) of Participants		Absolute Difference (95% CI), %
	Placebo (n = 180)	Colchicine (n = 180)	
Primary end point (postpericardiotomy syndrome) within 3 mo	53 (29.4)	35 (19.4)	10.0 (1.1 to 18.7)
Main secondary end points			
Postoperative atrial fibrillation[a]	75 (41.7)	61 (33.9)	7.8 (−2.2 to 17.6)
Postoperative pericardial/pleural effusions	106 (58.9)	103 (57.2)	1.7 (−8.5 to 11.7)
Cardiac tamponade	3 (1.7)	1 (0.6)	1.1 (−1.6 to 4.3)
Pericardiocentesis or thoracentesis	13 (7.2)	13 (7.2)	0.0 (−5.6 to 5.6)
Postpericardiotomy syndrome recurrence	3 (1.7)	3 (1.7)	0.0 (−3.3 to 3.3)
Disease-related readmissions[b]	2 (1.1)	2 (1.1)	0.0 (−2.7 to 2.7)
Overall mortality[c]	2 (1.1)	6 (3.3)	2.2 (−1.6 to 6.1)
Stroke	1 (0.6)	2 (1.1)	0.50 (−2.1 to 3.4)

Figure 2. Results of Kaplan-Meier Analysis of the Primary Outcome (Incidence of Postpericardiotomy Syndrome)



JAMA. 2014;312(10):1016-1023. doi:10.1001/jama.2014.11026

Table 3. Adverse Events in COPPS-2 According to Treatment Assignment at 3-Month Follow-up According to Treatment Assignment[a]

Adverse Events	No. (%) of Participants[b]		Absolute Difference (95% CI), %
	Placebo (n = 180)	Colchicine (n = 180)	
Any adverse events	21 (11.7)	36 (20.0)	8.3 (0.76 to 15.9)
Gastrointestinal intolerance[c]	12 (6.7)	26 (14.4)	7.7 (1.4 to 14.3)
Hepatotoxicity[d]	2 (1.1)	1 (0.6)	0.50 (−2.1 to 3.4)
Drug discontinuation	32 (17.8)	39 (21.7)	3.9 (−4.4 to 12.5)

JAMA. 2014;312(10):1016-1023. doi:10.1001/jama.2014.11026

5. Cardiac calcification in CHD

Cardiac Calcifications in Adults with Congenital Heart Defects

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ABSTRACT

Objective. We investigated the type and extent of calcification in a series of heart specimens from adult congenital heart disease patients because recent autopsy observations suggested a high prevalence of calcification.

Design. We used computed tomography to examine seven heart specimens from adults (>18 years old) with a congenital heart defect collected with permission from the family during a recent 3-year period. Clinical data regarding diagnosis, history, and imaging studies were recorded. The 3D data sets were reviewed after reformatting as maximum intensity projection and volumetric renderings to determine the pattern and extent of calcium deposition.

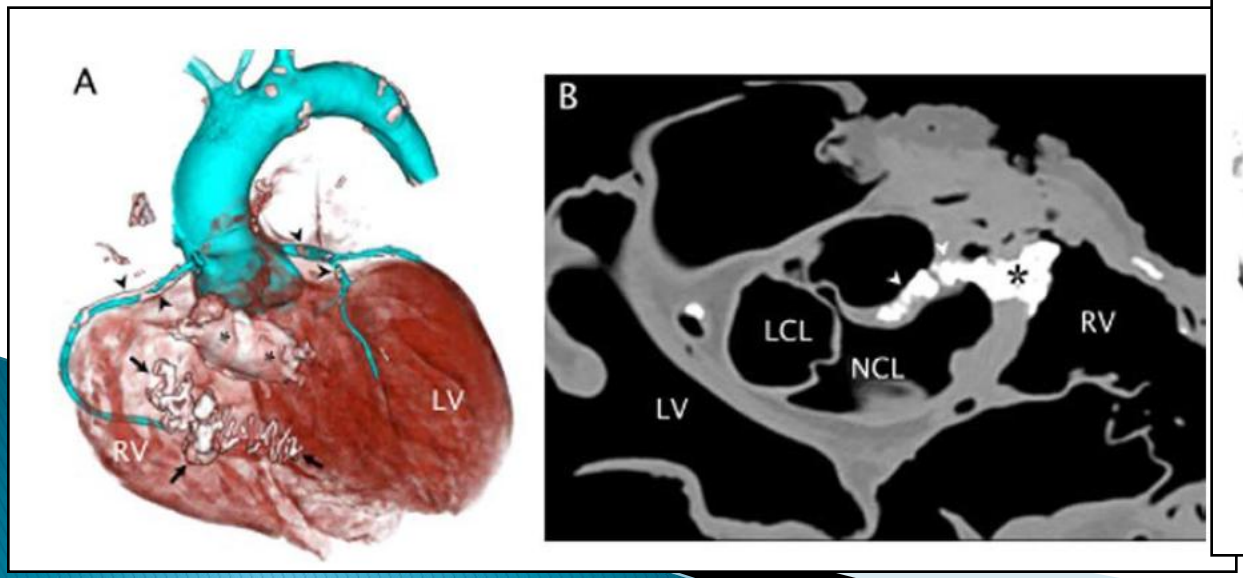
Results. Five of the seven hearts had extensive calcifications in one or more of three patterns: atherosclerosis associated in the three oldest cases; surgery associated in four of five hearts that had undergone heart surgery; and myocardial calcification remote from surgical sites in two cases. Myocardial calcification was associated with regional dysfunction and was present in the three patients that died suddenly and unexpectedly.

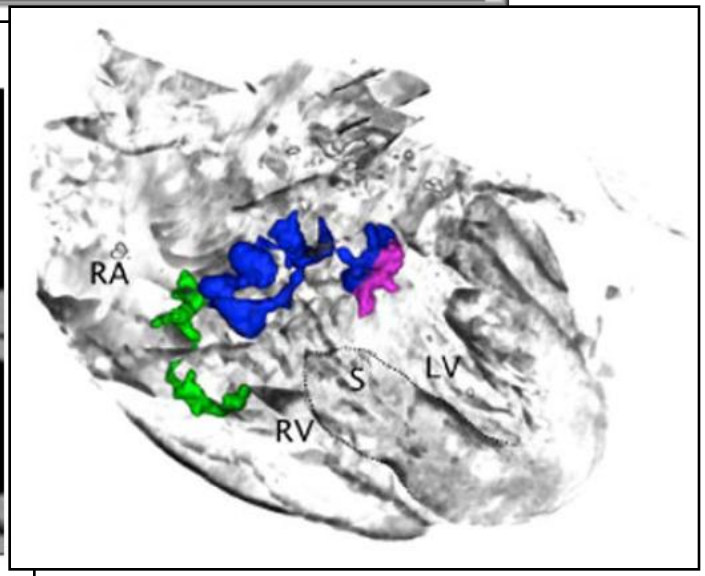
Conclusion. Cardiac calcification was frequent in our series of heart specimens from adults with congenital heart defects, was often but not uniformly associated with prior surgery, and, in our small series, was associated with regional dysfunction and sudden death.

Congenit Heart Dis. 2015;10:396–402

Table 1. Summary of Cases

Case	Age (years)	Sex	Diagnosis	Prior cardiac surgery (age in years)	Calcification sites	Cause of death
1	66	M	TOF	Potts shunt (8) Complete repair (17)	Ventriculotomy site VSD patch, aortic valve, coronary arteries, aorta	Sudden, unexpected Presumed arrhythmia
2	51	F	TOF and PA, multiple AP collaterals	None	Aortic valve, mitral valve, tricuspid valve, myocardial extension	Heart failure
3	46	F	TOF and PA, multiple muscular VSDs, LAD from RCA	Waterston shunt (2) Complete repair with RV–PA conduit (9) Conduit revision (12, 23) Balloon dilatation (43) Melody valve (45)	RV–PA conduit, VSD patch, pericardium, myocardium, lungs	Sudden, unexpected Presumed arrhythmia Recent decline with signs of restrictive pattern of heart failure
4	23	F	CAVC, subaortic stenosis	CAVC repair (<1) Mitral valvuloplasty (1)	Pledgets in: anterior mitral leaflet, tricuspid valve, ASD patch	Acute lymphoblastic leukemia
5	52	M	Heterotaxy, DORV, hypoplastic LV, CAVC, PS	Blalock–Taussig shunt (1) Central shunt (19) Bidirectional Glenn (32) Extracardiac Fontan (40)	Papillary muscle, left AV valve, right aortic leaflet, RCA, aorta, Fontan conduit	Sudden, unexpected Presumed arrhythmia Recent leg and viral infection
6	38	F	VSD, cleft MV, PDA	None	None	Eisenmenger syndrome
7	20	F	Scimitar syndrome, right lung hypoplasia, L-SVC to CS	RPV to left atrium (12) Stent of RPV (15)	None	Eisenmenger syndrome

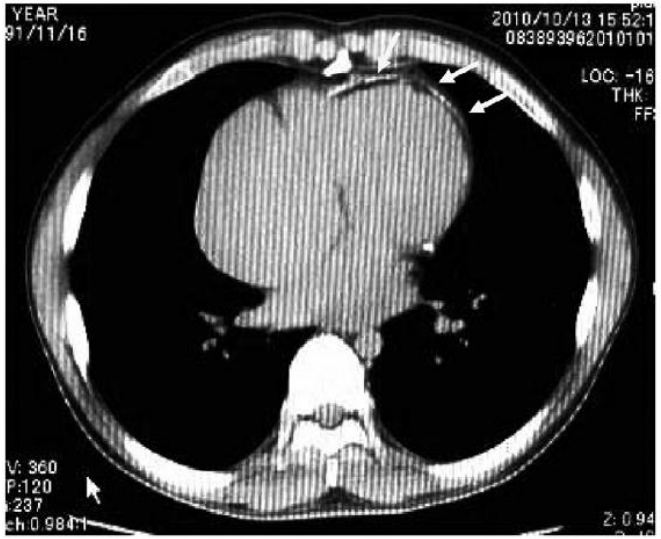




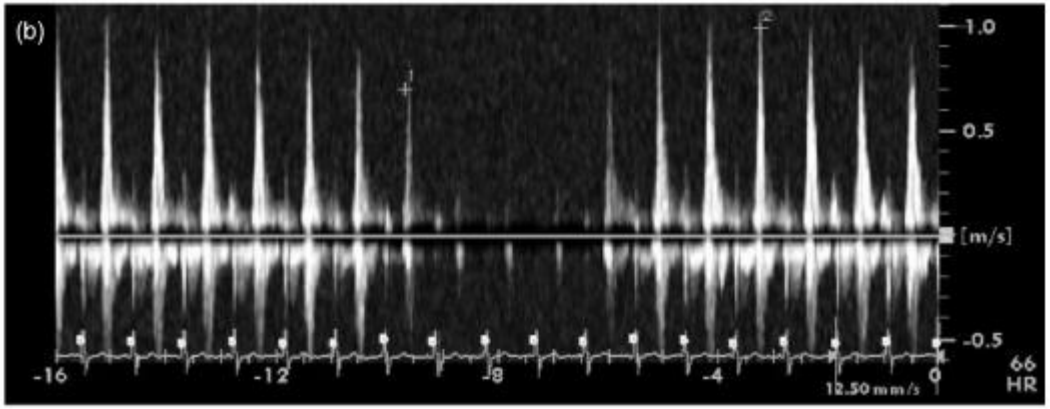
PEDIATRICS INTERNATIONAL Official Journal of the Japan Pediatric Society

Constrictive pericarditis developed after childhood repair of ventricular septal defect

Hirofumi Saiki, Satoshi Masutani, Masanori Tamura and Hideaki Senzaki
Department of Pediatrics and Pediatric Cardiology, Saitama Medical University, Saitama, Japan



- 19 years old male
- Heart failure and atrial flutter induced by constrictive pericarditis 15 years after VSD closure



Inspiration Expiration

Take Home massage

- First line imaging modality is echocardiography.
- CT is good for evaluation of cardiac calcification and extracardiac disease.
- MRI is good for evaluation of inflammation.

- Patients with pulmonary arterial hypertension and circumferential pericardial effusion can develop atypical cardiac tamponade.
- Cardiac calcification with or without heart surgery should be followed.