

# 2 year old boy with pre and post-capillary pulmonary hypertension

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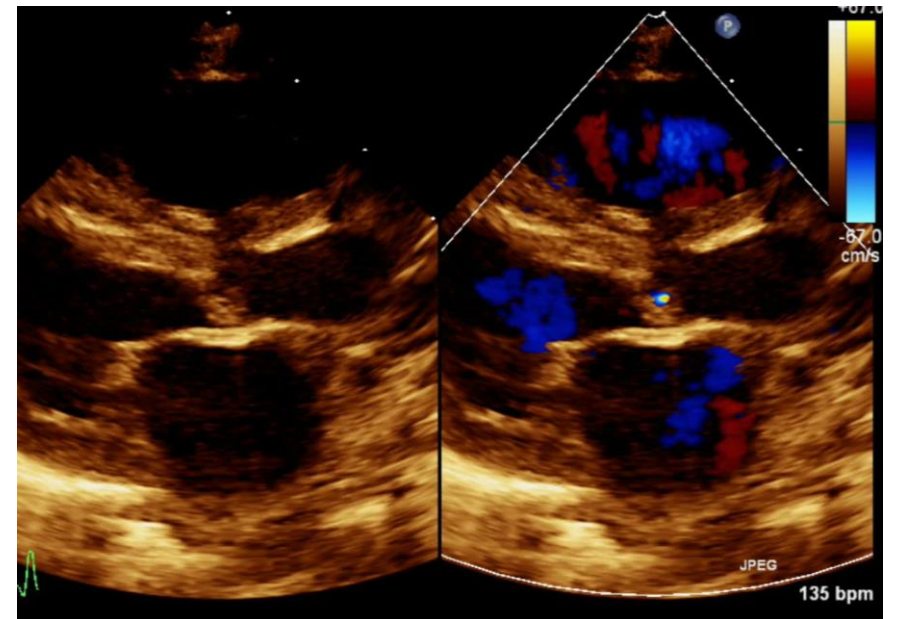
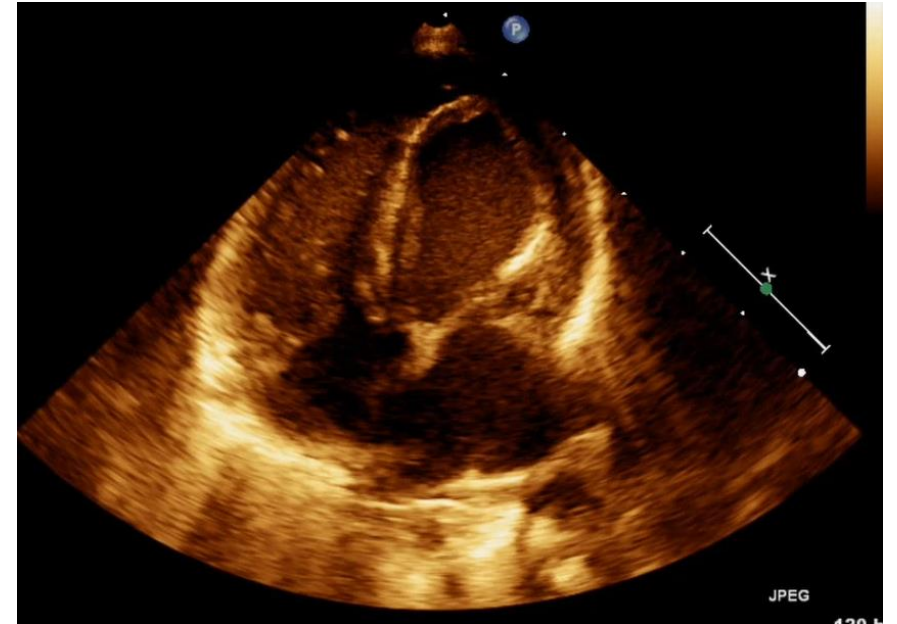
2 year old child:

- prenatal diagnosis: aortic stenosis with HF
- premature closure of foramen ovale from 26th week of pregnancy
- successful fetal balloon aortic valvuloplasty

ICU, respiratory failure

Echo:

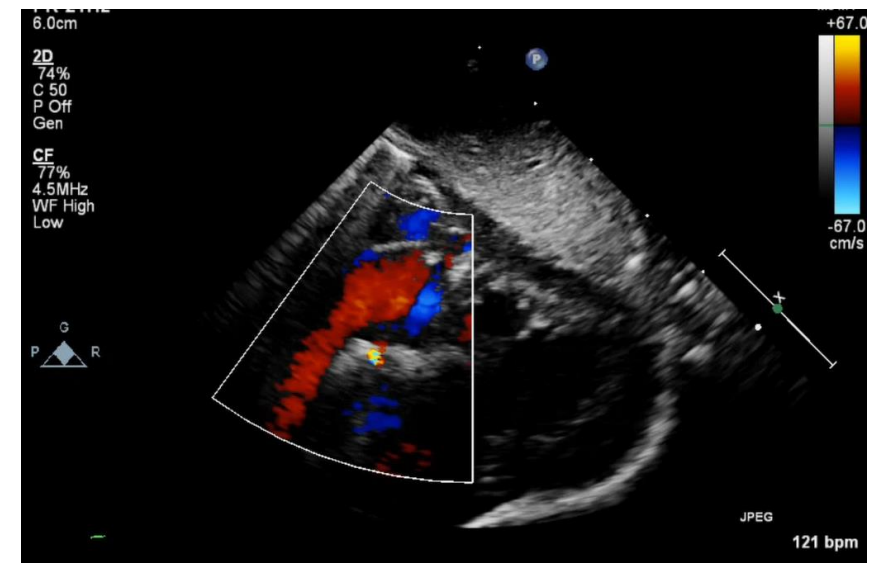
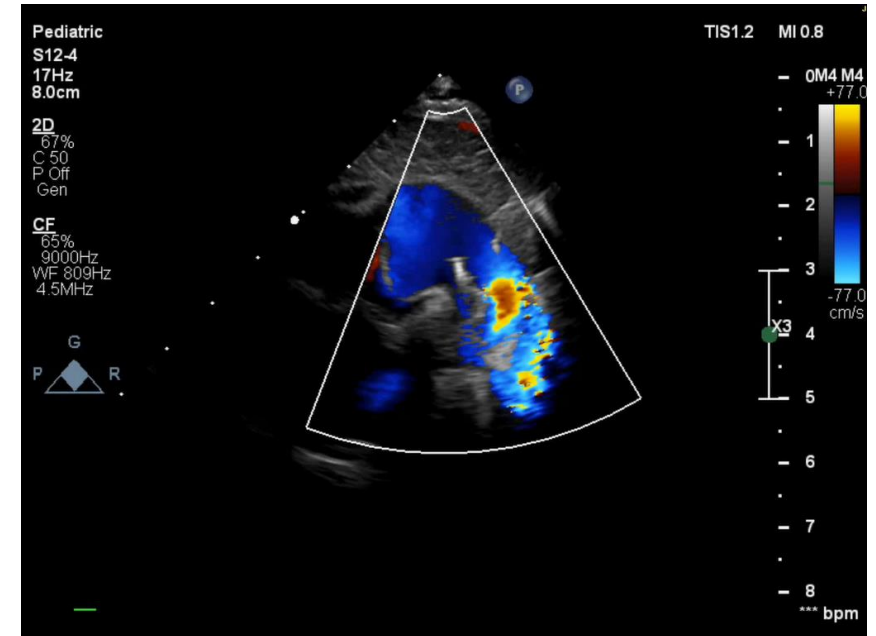
- mild SA (LV - Ao=28mmHg)
- mitral valve with thickened leaflets, E/A 2.2 IM II
- thickened left ventricular muscle
- mild endocardial fibroelastosis
- DA – right to left flow-PPHN?



PH crisis after DA closure → NO,  
epoprostenol, prostin  
->percutaneous transseptal puncture and  
balloon atrioseptostomy  
->stent to DA implanted

PA 55/22/39, Ao 42/23/32, DPG 6, TPG 23  
LVEDP 16mmHg, PCWP 16mmHg

sildenafil and then bosentan added as a  
second treatment



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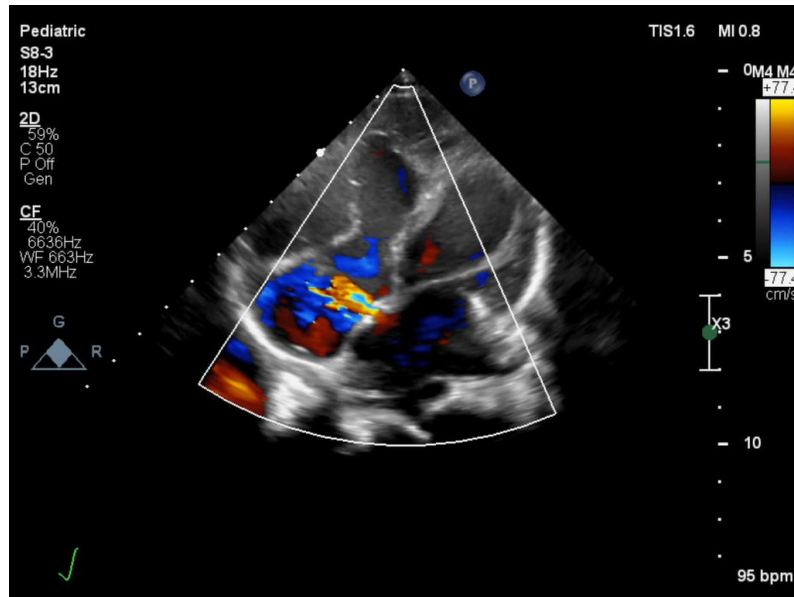
During 2 years:

- two balloon re-atrioseptostomy due to restrictive flow through ASD
- didn't require aortic valve intervention

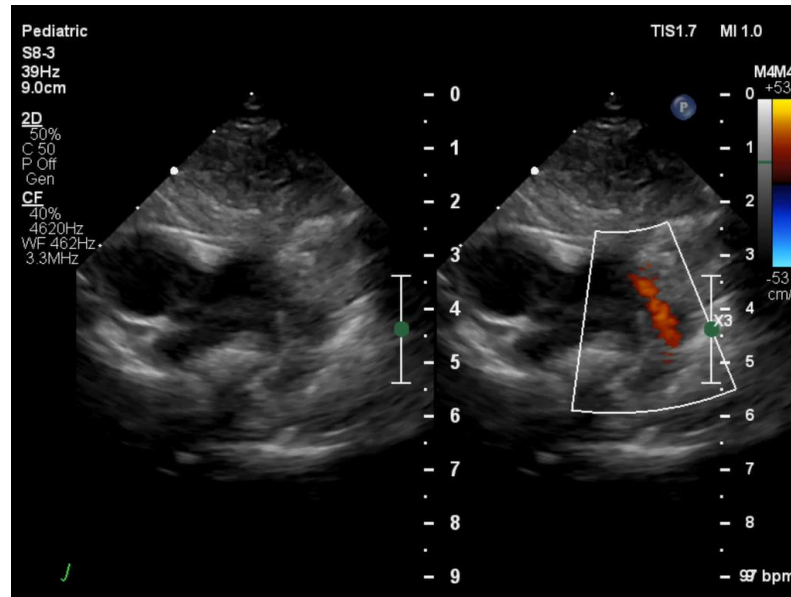
# WHO FC I / II

Sat lower limb 85-90%, upper limb 100%

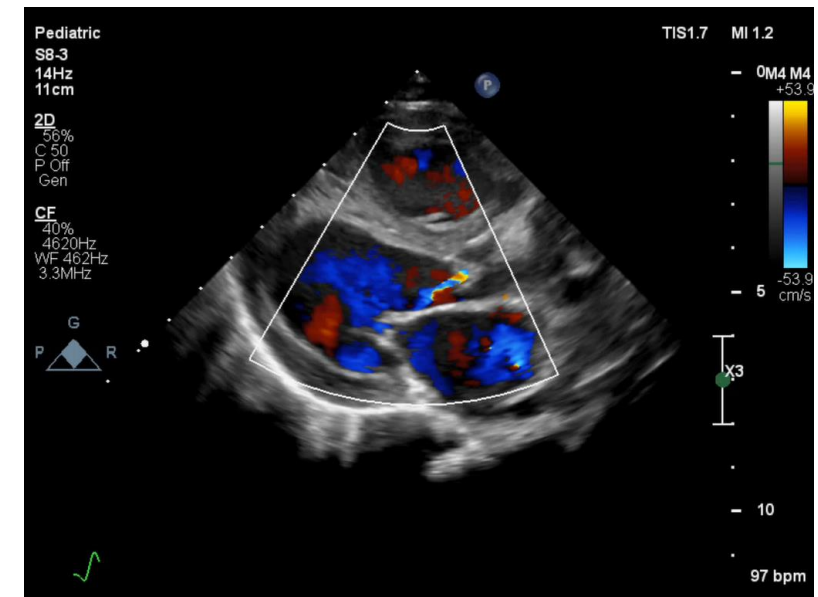
NTproBNP 800pg/ml



LV mild fibroelastosis, ↑LA  
L-R ASD 2, 1 m/s.



Bidirectional DA flow



thickened leaflets  
LV-Ao 7mmHg  
vMV 2l, opening 11mm

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PA 53/28/40,

Ao 54/27/40,

PVR 11 WU ( 5,6WUxm<sup>2</sup>)

PCWP mean 23 mmHG,

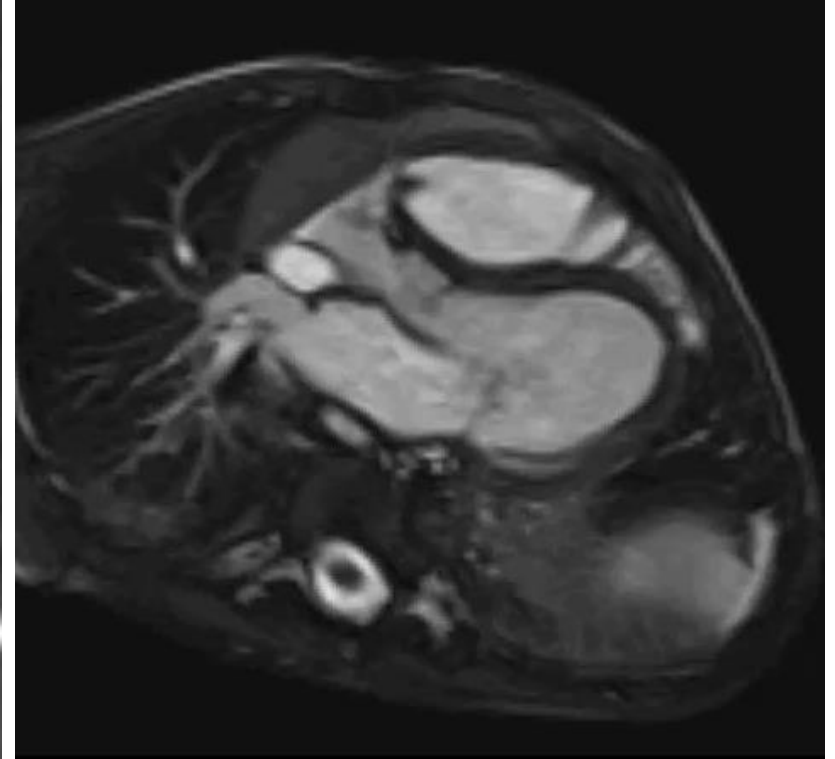
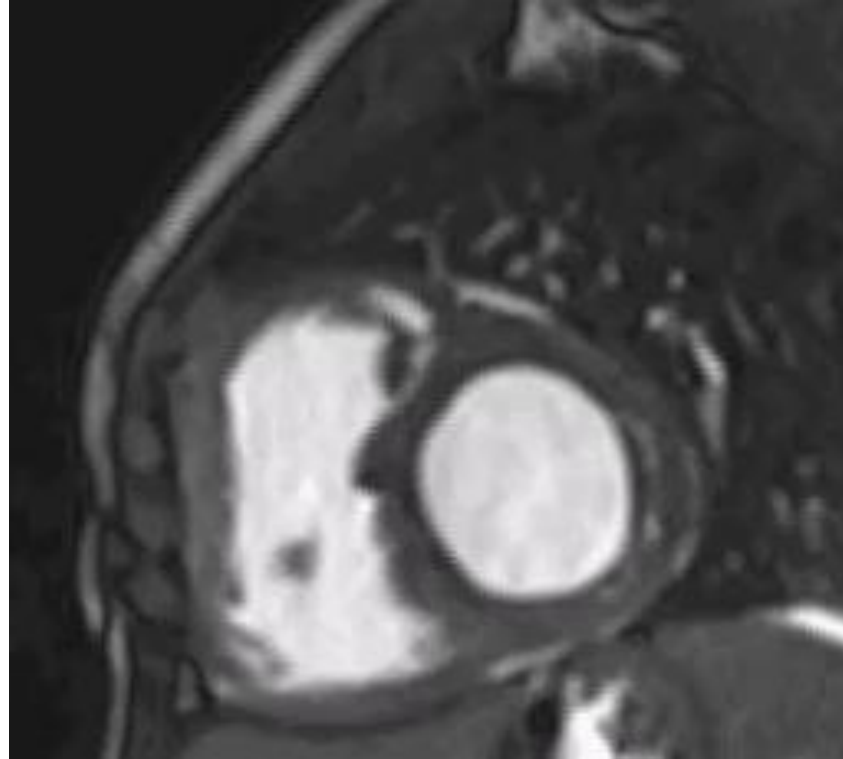
LVEDP 25 mm Hg,

ASD restriction: LA 22mmHg, RA 4 mmHg

Bidirectional DA flow

TPG 17mmHg

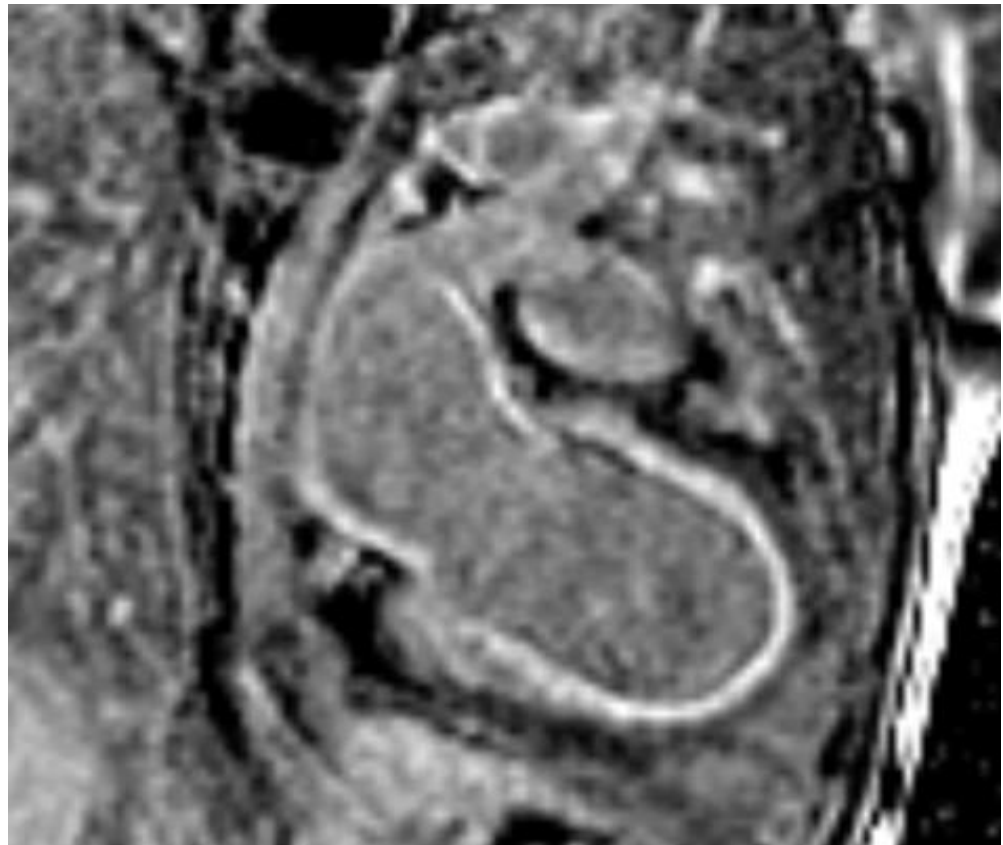
DPG 5mmHg



EF LV 60%, RV 58%  
↑LA, RA

LV ESVi 30ml/m<sup>2</sup>, EDVi 71ml/m<sup>2</sup>  
RV ESVi 48ml/m<sup>2</sup>, EDVi 127ml/m<sup>2</sup>





LGE ++ LV, LA, RA

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What next? Should we:

- continue to treat him with PAH specific therapy despite LV high end diastolic pressure?
- do re-atrioseptostomy or may implant stent/ASD fenestrated device due to recurrent ASD restriction (AFR device)?
- perform any surgical intervention?
- consult him for transplantation in future?