

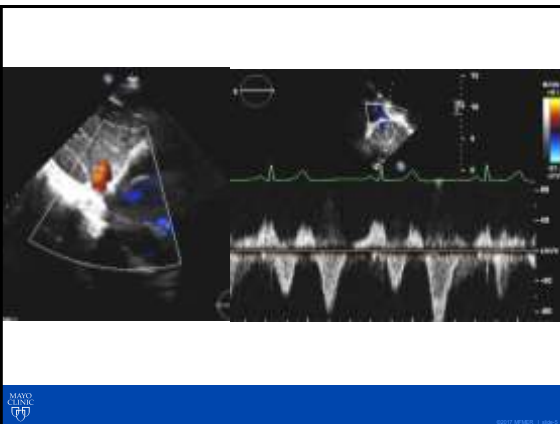
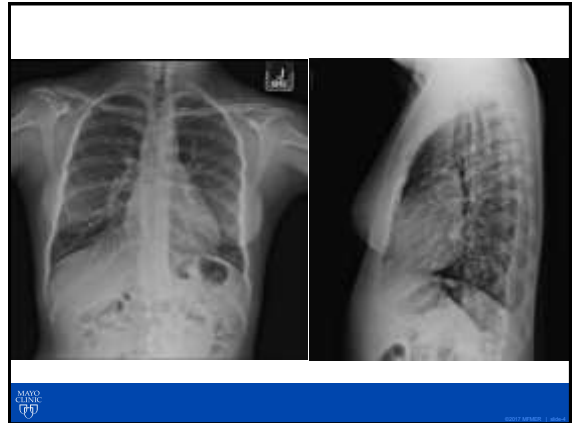
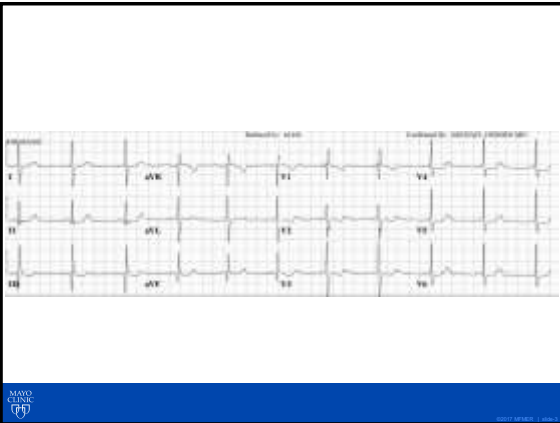


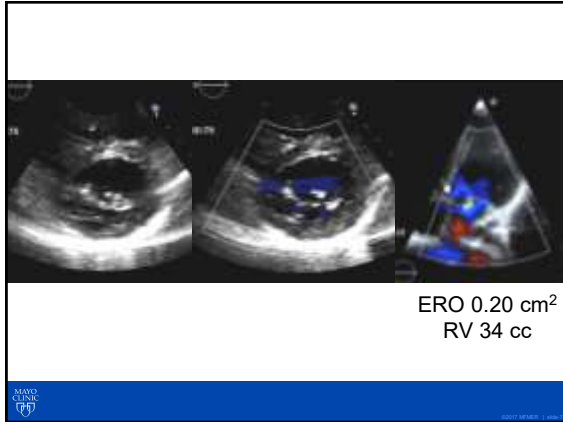
Heart Failure Case - Reversible

Heidi M. Connolly

27-Year-Old Female with Down Syndrome

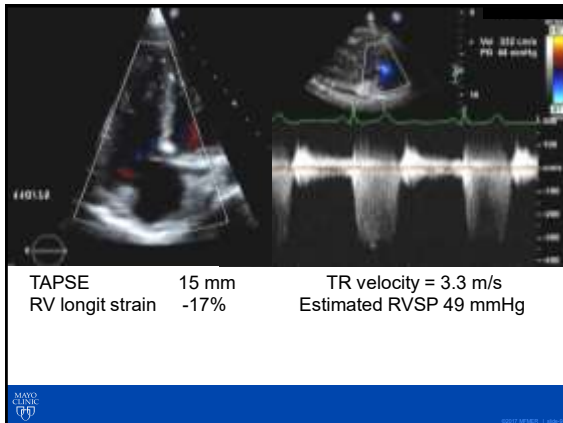
- Transitional AV canal defect
- 1990 – age 2 years
 - Right heart cath complicated by entrapment of catheter in MV chordae requiring urgent surgical removal, ASD and VSD also closed
 - Anterior MV cleft not addressed
- Regular FU – returns May 2015
 - No complaints
 - Aspirin and synthroid
- Group home, works 7 hours per day, HS graduate





Diastology

- E velocity 0.9 m/s
- A velocity 0.5 m/s
- E-A ratio 1.8
- Medial e' 0.12 m/s
- Lateral e' 0.22 m/s
- Medial E/e' 7.5
- Lateral E/e' 4.1



Serial Echocardiograms

	05/18/15	05/02/17	05/15/11	04/25/09
Pulm. HTN				
TR Velocity (m/sec)	3.90	3.60	3.80	3.70
RA Pressure (mmHg)	5	7	10	10
RVSP (mmHg)	49	57	68	65
RV E/A Ratio	27	26	26	26
RV E/e' (Medial)	8.25	8.33	8.14	8.27
RV E/e' (Lateral)	4.37	3.62	3.52	2.99
TV Coaptation (mmHg)			1.03	
TV Leaflet Vel (m/sec)	0.89		0.13	0.13
TAPSE (mm)	21	18	14	17
RV Area Peak Systolic Strain (%)				

PH clinic evaluation
No symptoms
No PH therapy now

What would you recommend?

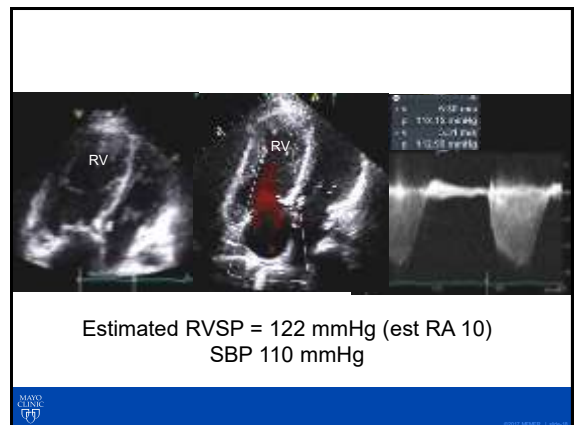
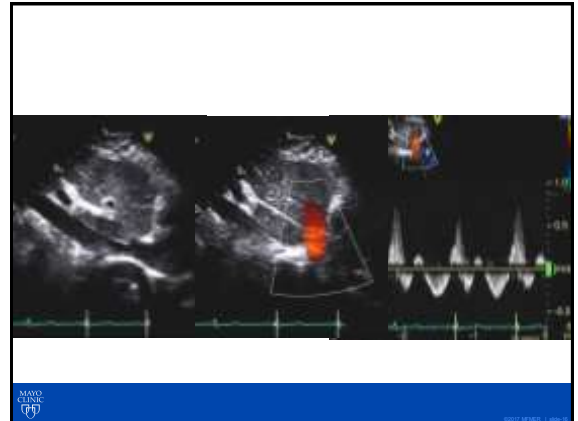
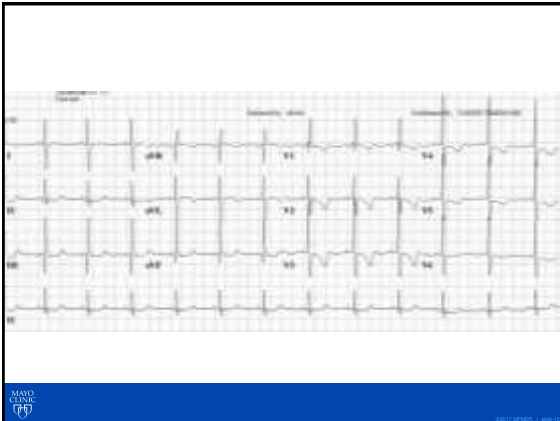
1. Observation, follow-up in one year
2. MV repair or replacement
3. PH consultation for advanced therapy
4. Other

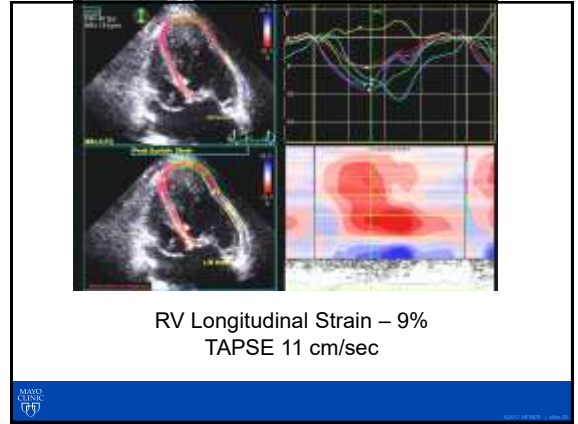
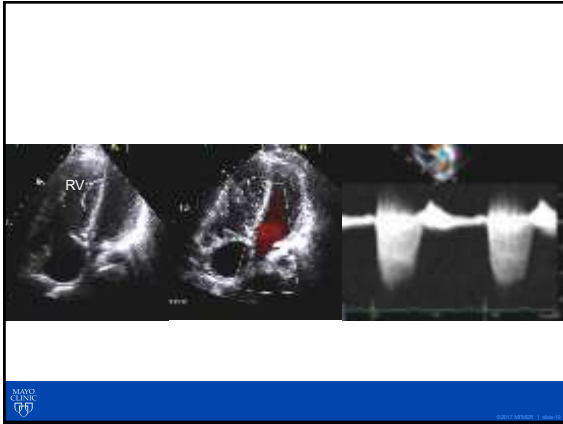
27-Year-Old Female with Down Syndrome

- 2/16 – syncope related to foot pain
 - Subsequent surgery – accessory navicular removed
- Progressive dyspnea, now with 1 flight
 - 4/16 CT PE – negative
 - Started on diuretics – no help
- Returns early – dyspnea, edema

Examination

- JVP 15 cm, A and V visible
- Parasternal impulse, normal S_1 , increased P_2
- Grade 2 HSM LSB, increases with inspiration
Grade 3 HSM at apex, increases with expiration, no DM
- NT-proBNP 6505 pg/mL (normal < 124)





Param. HTN	06/21/16	09/18/15	05/02/13	03/15/11
RA Pressure (cmH ₂ O)	18	9	9	10
RVSP (mmHg)	122	99	77	68
RVAP (mmHg)	34			20
Mid-EVID (d) 4-C (mm)	40	27		
RA volume index (MOD) (ml/m ²)	25			
RMP	0.19	0.28	0.33	0.24
LV Cardiac Index (L/min/m ²)	1.81	3.57	3.82	3.12
RA Capacitance (ml/mmHg)	0.67			0.83
TV Lat Area Vol (L)(ml/m ²)	0.11	0.09		0.10
TAPSE (cm)	15	21	18	14
RV Avg Peak Systolic Strain (%)	-8			-17

Note: Only the last 4 studies are shown.

- ### Cardiac Catheterization
- Severe PH
 - mPA = 91 mmHg (FA 118/63)
 - PVR = 30 WU
 - RA = 15 mmHg
 - 100% inhaled oxygen
 - mPA = 88 mmHg
 - PVR = 25 WU
 - 60 ppm inhaled NO
 - mPA = 89 mmHg
 - PVR = 28 WU
 - CI = 1.7 L/min/m²
 - PCWP = 14 mmHg

- ### What would you recommend?
- Diuretics and reassess
 - MV repair or replacement
 - PH consultation, advanced therapy
 - Refer for transplant
 - Other

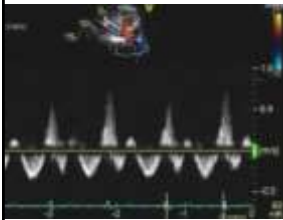
- ### PH Consultation
- Right heart failure
 - Medical therapy
 - Furosemide 20 mg daily
 - Sildenafil (Revatio) 20 mg TID
 - PDE5 inhibitor
 - Macitentan (Opsumit) 10 mg QD
 - Endothelin receptor antagonist (ERA)
 - Treat sleep apnea

Where did I go wrong?

Four Months Later – Reevaluation

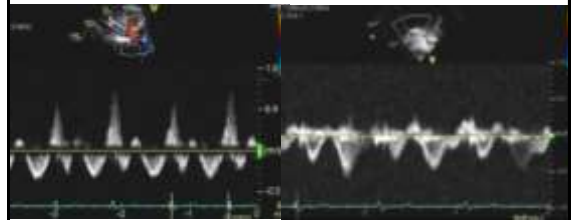
- Improved exercise capacity – no edema
- NT-proBNP 168 pg/mL (4 months prior 6505)

April 2016



April 2016

August 2016

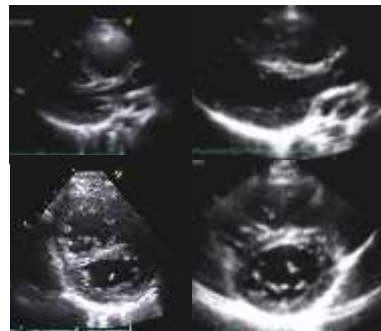


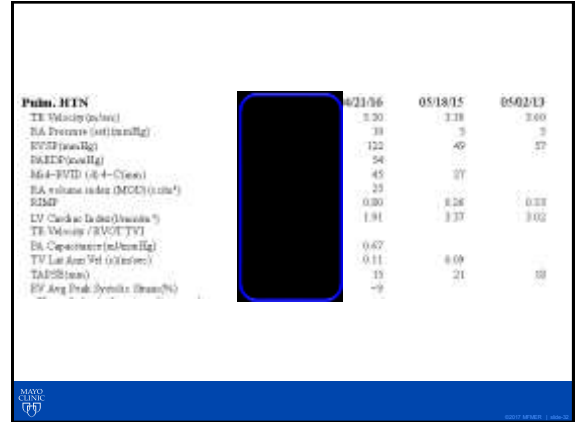
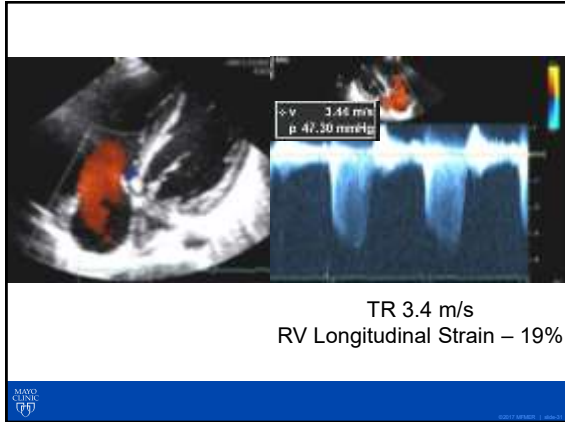
August 2016



April 2016

August 2016





Follow-up

- Return visit April 2017
 - RH size, function and pressures stable
 - Continue dual PH therapy, CPAP
 - Decrease furosemide
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Lessons Learned

1. Hepatic vein PW Doppler helps assess RA hemodynamics, analogous to RAP
 2. PAH can develop rapidly
 3. PAH can be effectively treated by modern medical therapy
 4. Medicine is fascinating – and humbling
 5. Never stop learning!
- MAYO CLINIC



Questions & Discussion
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