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Complex Case:

PH Associated with an ASD

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I.	II.	III.	IV.
A	A	A	A
YES: ☐	YES: ☐	YES: ☐	YES: ☐
NO: X	NO: X	NO: X	NO: X
B	B	B	B
YES: ☐	YES: X	YES: ☐	YES: ☐
NO: X	NO: ☐	NO: X	NO: X

SCORE: 01234

January 2010

6 yo girl referred to The Hospital for Sick Children (SickKids) when she was 4 yo, from a peripheral Children's Hospital

- born term
- diagnosed with secundum ASD at 9 months of age
- followed by cardiologist at peripheral Children's Hospital
- symptoms:
    - feeling tired at school and not keeping up with her peers
    - at times requiring being carried afterschool to the car due to fatigue, quit cheerleading
    - episodes of self-resolving chest pains "heart hurts
    - no history of cyanosis or syncope
- echo showed mPAP

Cardiac cath at The Hospital for Sick Children:

baseline:	mPAP 43	PVRi 7.1	wedge 6	Qp:Qs 1.79:1
100% O2:	mPAP 46	PVRi 5.5	wedge 8	Qp:Qs 2.2:1
100% O2 + iNO:	mPAP 41	PVRi 4.7	wedge 8	
iNO:	mPAP 45	PVRi 6.6	wedge 7	Qp:Qs 1.97:1

Subsequently started on oral sildenafil and repeat cath in

1 year later – **January 2011**

baseline:	mPAP 52	PVRi 8.8	wedge 9	Qp:Qs 1.5:1
100% O2:	mPAP 46	PVRi 4.29	wedge 6	Qp:Qs 2.4:1
100% O2 + iNO:	mPAP 44	PVRi 3.8	wedge 7	Qp:Qs 2.05:1
iNO:	mPAP 47	PVRi 5.7	wedge 7	Qp:Qs 1.74:1

presented for possible ASD closure with reliable fenestration

7 months later – **August 2011**

- ASD closure with fenestration in cath lab
- Exhibited signs of stroke in recovery
- MRI - left MCA stroke; resulting in right hemiparesis
- Immediately brought to IGT for thrombectomy, decompressive craniotomy - bone flap removed

4 months later – **December 1, 2011**

- pulmonary hypertension crisis during anesthesia induction for elective autologous cranioplasty replacing bone flap.
- Procedure cancelled
- Echo - suprasystemic , with estimated mPAP of 87 mmHg and RVSP of 109 mmHg + RA pressure of 13 mmHg (by CVP tracing). Severely dilated RV with moderate to severe systolic dysfunction and IVS bowing into the LV. Underfilled LV with qualitatively good systolic function. Well seated ASD device with a small device fenestration shunting LA to RA

5 days later – **December 6, 2011**

Chest CT with contrast:

- Severe pulmonary artery hypertension with markedly dilated central pulmonary arteries, right ventricular hypertrophy and flattening of the interventricular septum.
- Pulmonary veno-occlusive disease, unlikely but cannot be completely excluded.
- Ground-glass appearance of peripheral parts of both lungs, nonspecific findings of any airspace/interstitial pathology including infection and hemorrhage.

3 days later – **December 9, 2011**

- Cannulated on ECMO

3 days later – **December 12, 2011**

- Decannulated from ECMO
- Echo - severely dilated right ventricle with severely reduced systolic function, similar to previous study. IVS is flat in systole. Estimated RVSp by incomplete TR jet is at least 80mmHg + RA pressure (cuff BP 103/67). Estimated mean PA pressure by PI is at least 37mmHg +Rap. Good LV systolic function. Mild MR.

Other issues:

- Epistaxis - since extubation from left nare. Seen by ENT -silver nitrate no effect, therefore, inserted absorbable pack with good effect

- Anticoagulation therapy

- FTT - receiving TPN via CVL, remains on ranitidine, unable to insert N/G due to nasal packing

- Left vocal cord paresis due to intubation

2 days post off ECMO – **December 14, 2011**

- Started on bosentan and epoprostenol IV via CVL
- Echo - severely dilated RV with severely reduced systolic function. Septum flat in systole. Incomplete TR jet, unable to estimate RVsp. Estimated mean PA pressure by PI is at least 53mmHg + RAp. Good LV systolic function. Mild MR.

3 days later – **December 17, 2011**

- Weaned off iNO and milrinone.
- Echo - Severely dilated RV with moderately reduced systolic function, similar to previous study. Septal flattening in systole and diastole. Incomplete TR jet to estimate quantitatively the RVsP. Estimated mean PA pressure is at least 62mmHg + RAp. Good LV systolic function. Device fenestration shunting L-R

****Listed for double lung transplant****

10 days later – **December 27, 2011**

- transferred to ward from CCCU (resp NP = Janette Reyes)
- switched from IV epoprostenol to SC treprostinil
- continued on sildenafil and bosentan
- Physical assessment: Neuro – alert, whining, does not want to be touched, unable to sit up to void. Resp - good air entry bilat with no adventitious sounds. Cardiac - periph warm with brisk cap ref <3 sec, radial pulses palp at 2+, left fem pulse palp at 2+, right pedal pulse weak and present by doppler (dressing to right groin), normal S1, loud and single S2, no murmur, RV heave, palpable P2. Abd - soft, no HSM. NYHA Class III-IV

2 days later – **December 29, 2011**

- NT-proBNP = 500 pmol/L
- echo - severely dilated RV with mod reduced systolic function. Mild TR, estimated RVSP is at least 102 mmHg + Rap. Mild PI, estimated mPAP of 79 mmHg +RAp. IVS bowing towards LV in systole. Good LV systolic function. ASD device fenestration shunting mainly L-R. RV z-score +7.2
- Good spirits compared to yesterday, playful, more alert, increased energy, and communicative.
- Physical exam: sitting up in bed, alert, laughing, hoarse voice, playful with NP, good a/e bilat. No cyanosis, normal S1, single loud S2, hyperactive precordium (unchanged from previous), periph warm with brisk cap refil<3 sec, brach bilat and left pedal pulse palp at 2+, right pedal pulse palp at 1+, abd soft, no HSM
- treprostinil dose at 22.2 ng/kg/min, sildenafil at 7.65 mg po Q6H, bosentan at 31.25 mg po bid

increased energy

Listed for double lung transplant

treprostinil

mom unsure about transplant

2 days later – **December 31, 2011**

- double lung transplant

2 days later – **January 2, 2012**

- Echo post transplant - Mildly reduced RV systolic function with moderate to severe RVH, similar c/w previous. Good LV function. Trivial TR unable to estimate RVSp. Normal septal curvature.

3 weeks after transplant – **January 23, 2012**

- Feeling well and able to walk around the unit with family. Tried ice cream (unthickened) with OT - tolerated well
- Saturating well on room air.
- Tolerating 10 hour continuous feeds on NJ tube.