Persistent PAH after shunt closure

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Key issues:

1. Triple therapy
2. Implantable pump
3. Evaluation or listing for LuTx

I.	II.	III.	IV.
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... from any institution	... from any institution	... for any institution	... not related to presentation
YES ☐ / NO X	YES ☐ / NO X	YES ☐ / NO X	YES ☐ / NO X

SCORE: 01234

Case report – 16 yrs old pt

2006 - PDA closure without hemodinamic evaluation at 3 yrs of age

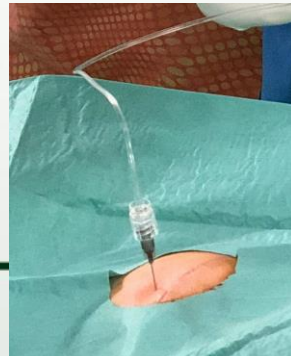
2007 - RHC 1 year later: PAP:69/25 (46) mmHg, PVRi:4.9 WU/m2
→ sildenafil

2010-2015 - slow progression in FU (↑ PAP by echo), no symptoms

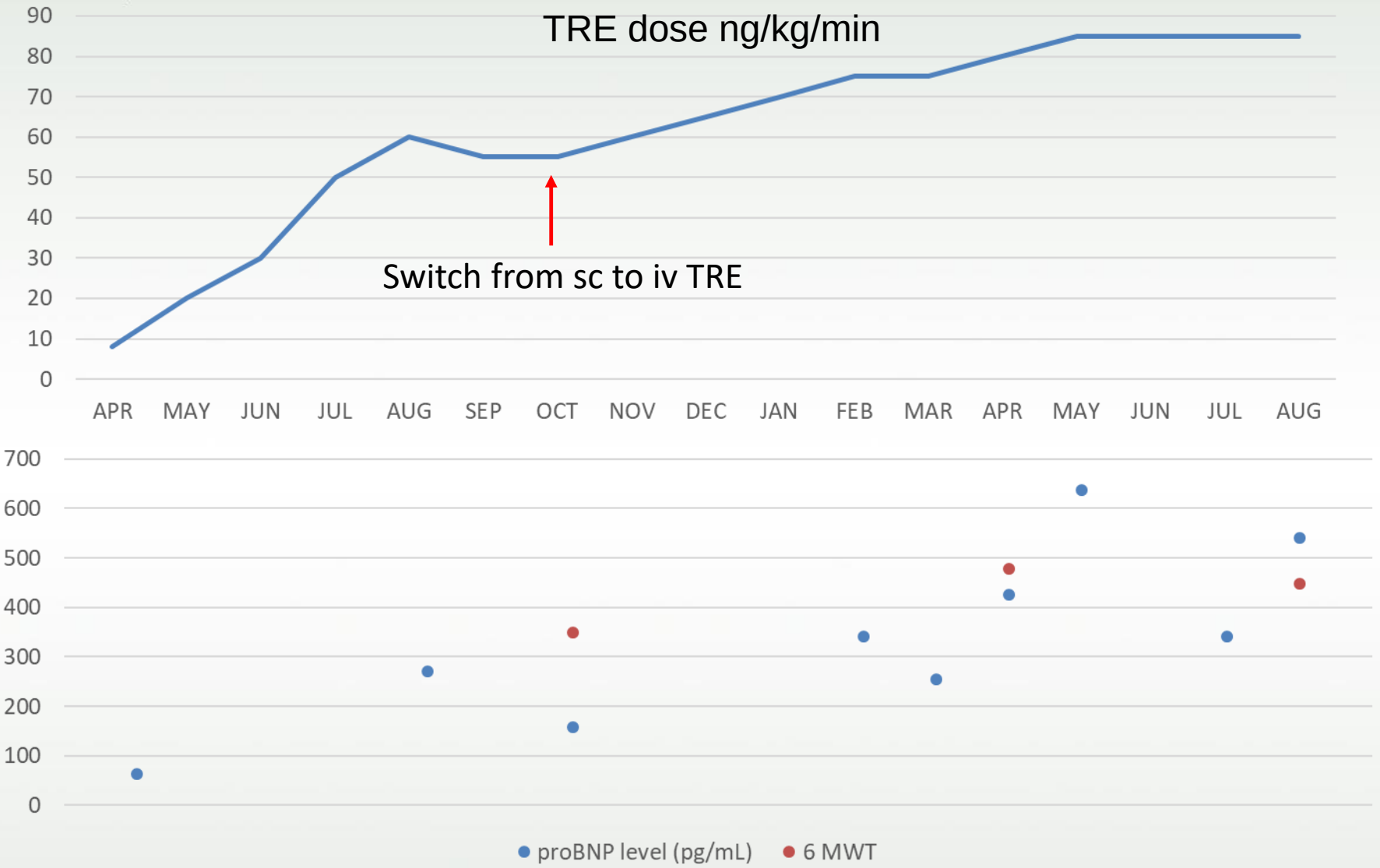
2016 - fatigue, WHO Class II, CPET - pVO2: 23 mL/kg/min at 13 yrs of age → combination with bosentan

2018 - rapid progression in the next 2 yrs (WHO Class III, syprasystemic RVP, pVO2:12.3 mL/kg/min) → *triple therapy with sc treprostinil* → ***evaluation for LuTx***

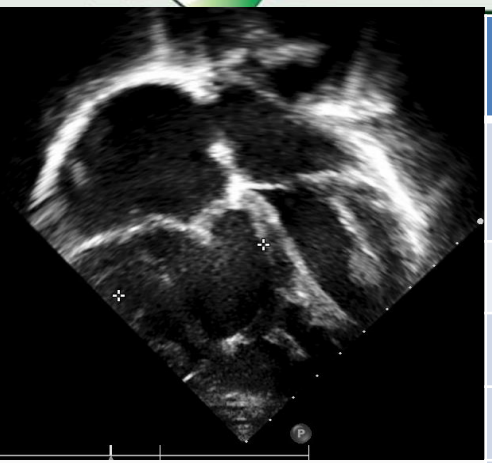
- continuous TRE dose escalation, increasing injection site pain → switch from sc to iv TRE (Lenus pump)



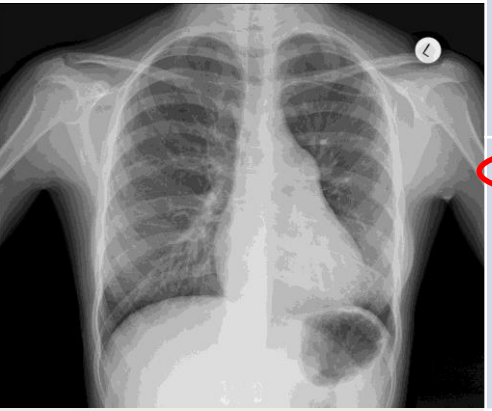
proBNP/6-MWT and TRE dose 2018-19



Lower vs higher risk



TAPSE: 19.8 mm
RVEDD: 47 mm
CI:4.0 l/min/m2



Lower risk	Determinants of risk	Higher risk
No	Clinical evidence of RV failure	Yes
No	Progression of symptoms	Yes
>350	6MWT (>6 yrs old) m	<350
Normal	Growth	Failure to thrive
I-II	WHO functional class	III-IV
Minimally elevated	SBNP/NT proBNP	Significantly elevated Rising level
	Echo	RV enlargement/dysfunction Reduced LV size, ↑ RV/LV ratio Reduced TAPSE Low RV FAC Pericardial effusion
CI>3.0 l/min/m2 SVO2 sat>65% Acute vasoreactivity	Hemodynamics	CI<2.5 l/min/m2, mRAP>10 mmHg PVRI>20 WU/m2 SVO2 sat<60% PACI<0.85 mlxmmHgxm2

Rosenzweig EB Eur Respir J. 2019 Jan;53(1):1801916

Referral and transplantation guidelines in PA

Referral guidelines

- NYHA functional class III or IV symptoms during escalating therapy
- Rapidly progressive disease (assuming weight and rehabilitation concerns not present)
- ***Use of parenteral targeted PAH therapy regardless of symptoms or NYHA functional class***
- Known or suspected pulmonary veno-occlusive disease or pulmonary capillary hemangiomatosis

Transplantation guidelines

- NYHA functional class III or IV despite a trial of at least 3 months of combination therapy including prostanoids
- Cardiac index of O_2 liters/min/m^2 (markedly reduced CI)
- Mean right atrial pressure of >15 mmHg
- 6-minute walk test of <350 m
- Development of significant hemoptysis, pericardial effusion, or signs of progressive right heart failure (renal insufficiency, increasing bilirubin, brain natriuretic peptide, or recurrent ascites).

Geert M Verleden J Thorac Dis 2017 Sep; 9(9): 3372–3384

Questions

- Maximal dose of iv treprostinil? (limitation: pump size)
- Timing of evaluation and listing for lung transplantation?
- Indicators for listing? (WHO class, BNP, CPET, 6-MWT)
- Need for FU RHC during the triple therapy?