Management of a Child with Severe Pulmonary Hypertension Presenting with Severe Systemic Hypertension: A Case Study

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Purpose: We describe the presentation and management of a 12-year-old female with severe pulmonary hypertension (PH) who initially presented with severe systemic hypertension.

Background: We describe the presentation and management of a 12-year-old female with severe pulmonary hypertension (PH) who initially presented with severe systemic hypertension. While at times critically ill, successful therapy included traditional pulmonary vasodilators and an atrial septostomy, while balancing adequate maintenance of her systemic vascular resistance (SVR) to maintain cardiac output.

Case Report: Her neonatal history was significant for meconium aspiration syndrome and multiple small VSDs, resolved by 3 months. There was no family history of PH. The first significant event was at age 6 with a syncopal episode, ascribed to dehydration. Subsequently, she had a gradual decline in exercise tolerance, and for 3 years prior to presentation, had decreased appetite with frequent emesis. During a routine pediatric visit on the day of admission, her upper extremity BP measured 200/135. An echo performed shortly thereafter demonstrated normal LV function (but with reversed E/a ratio), an hypertrophied RV with severe dysfunction, and significant bowing of the ventricular septum into the LV during systole. The TR Doppler velocity predicted an RV pressure of 140mmHg (5.9m/s) + CVP, and pulmonary regurgitant end-diastolic gradient of 47mmHg (3.4m/s) + CVP. The initial ECG was notable for sinus rhythm, right atrial enlargement, RVH and T- wave inversion in inferior leads. Cardiac catheterization under GA the following day revealed RV and LV systolic pressures of 146 and 165mmHg respectively, thermodilution CI of 3 l/min/m², mean PA pressure of 116mmHg, and PVR and SVR of 33 and 51Wu x m² respectively. Her mean RA pressure was 3mmHg, and mean wedge pressures normal at 10mmHg.

Results: IV epoprostenol was commenced during the catheterization, and titrated up to a dose of 23ng/kg/min over the first 48 hours, then converted to treprostinil (with continued up-titration). There was no evident impact on her systemic blood pressure (systolic greater than 210mmHg). Oral enalapril was then commenced at incremental doses of 2.5mg every other dose (q12h). After 4 days, at a dose of 15mg, her systolic pressure had decreased to the mid-180s, but her LV was hyperdynamic, with a new left ventricular intra-cavitary gradient on echo interrogation (65mmHg). Two days later, approximately 1 hour after receiving an enalapril dose of 25mg, she developed acute hypotension (BP 60/35) and low cardiac output, severe ECG evidence of ischemia, and began to vomit. IV vasoconstrictor therapy was immediately instituted (vasopressin, nor-epinephrine and phenylephrine), with a return to baseline blood pressure and resolution of ischemic changes after 4 hours of accelerating therapy. Over 24 hours, most medications were weaned, but she remained on phenylephrine. Cardiac catheterization the following day demonstrated some drop in her resistances compared to her initial assessment, but PA systolic pressures were greater than systemic. An atrial septostomy was also performed. Thereafter, therapy concentrated on escalating her prostanooid dose, with no attempt made to directly treat her systemic hypertension. For the ensuing three weeks, she required low dose phenylephrine, with any attempt to wean manifested by the recurrence of the intra-cavitary LV obstruction and ischemic ECG changes. Her systemic BP did decline slowly with escalating prostanooid dosage, and ultimately, on a dose of 100ng/kg/min of treprostinil, the phenylephrine was weaned. She was discharged home with a systemic BP of 155/90, and echo estimated RV pressure of 118mmHg, with improved RV function and no intra-cavitary obstruction. During the course of her admission, diagnostic tests to exclude secondary causes of systemic hypertension were all negative, including evaluation of her renal arteries and exclusion of neuro-endocrine tumors.

Conclusion: This patient represents an extreme situation where intrinsic mechanisms to maintain systemic pressure in the face of compromising PH overcompensated and resulted in a massive elevation of SVR. Systemic vasodilators to relieve the SVR were not tolerated as intra-cavitary LV obstruction developed. Appropriate therapy in this case centered on effective pulmonary vasodilation and SVR support, with the hemodynamic milieu adjusting appropriately over time. Rapid changes in SVR under any circumstances, especially those described in this report, should be avoided if at all possible in patients with severe PH.