Prostacyclin Withdrawal in a Pediatric Idiopathic Pulmonary Hypertension Patient: A Case Report

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**Purpose:** To describe one unique pediatric patient scenario where hemodynamics demonstrated the ability to wean parenteral prostacyclin therapy.

**Background:** Intravenous prostacyclin is believed to require therapy of indefinite duration. To our knowledge, weaning off parenteral prostacyclin therapy in pediatric patients has not been well characterized. We describe the presentation and management of a 14-year-old female with idiopathic pulmonary hypertension (PH) and complex social situation, who initially presented profoundly symptomatic, yet without significant hemodynamic changes. Despite severe presentation, this patient was highly responsive to traditional pulmonary vasodilators. Due to remarkable tolerance of a prolonged abrupt discontinuation of treprostinil as well as clinical symptom resolution, the patient was able to be successfully weaned off parenteral prostacyclin therapy.

Patient was previously healthy until symptoms onset at age 13, including worsening activity intolerance, shortness of breath, and syncope. No family history of PH was reported. An echocardiogram performed during a second-opinion cardiology visit on the day of admission demonstrated severe right atrial and right ventricular dilation, severely depressed right ventricular function, septal flattening. The initial EKG was notable for right atrial enlargement, right axis deviation and right ventricular hypertrophy. Cardiac catheterization revealed hemodynamics consistent with diagnostic criteria for PH. An atrial septostomy was also performed. IV prostacyclin, bosentan and sildenafil therapies were initiated within four days of admission. IV epoprostenol was commenced during the catheterization, and titrated up to a dose of 13 ng/kg/min, then converted to IV treprostinil (with continued up-titration to 57 ng/kg/min). Clinical data showed rapid improvement to medication therapies.

**Methods:** Four months after prostacyclin therapy initiation, patient was admitted due to an episode of abrupt discontinuation of treprostinil lasting 24 hours. Patient remained remarkably asymptomatic throughout this episode. Prostacyclin infusion was reinitiated and titrated up to 25 ng/kg/min, 50% of previous treprostinil maintenance dose. Three months later central line infection was identified and decision was made to discontinue prostacyclin therapy due to improvement of PH on cardiac catheterization. The total duration of prostacyclin therapy prior to weaning was seven months.

IV Treprostinil wean was performed in the cardiac intensive care unit. The dose was decreased by increments of 5 ng/kg/min (20% of baseline dose) every 8 hours as clinically tolerated.

**Results:** Overall, patient tolerated rapid wean well over 30 hours. Hemodynamics at the time of original diagnosis of PH, immediately prior to wean, and one month post-discontinuation of prostacyclin: PVR (5; 3.3; 3.63), mPAP (27; 25; 27), mPAWP (10; 10; 8) and CI (3.6; 4.56; 5.23), respectively. Regarding exercise intolerance, she was unable to perform six minute walk distance (6MWD) at time of diagnosis, 6MWD was 440 meters at one month post pulmonary vasodilator initiation and exercise tolerance continued to improve with time. One month post-prostacyclin discontinuation, graded exercise reported patient achieved 89% of max predicted heart rate.

**Conclusion:** This is a complex case in which a pediatric patient with PH experienced dramatic symptom resolution. This unique pediatric patient scenario illustrates tolerance of a controlled rapid parenteral prostacyclin therapy withdrawal.