

Cyanosis in a patient with corrected pulmonary atresia due to pulmonary arterial hypertension and nocturnal apnea effectively treated by non-invasive CPAP

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Objectives: Case report of a 41 year old male, with pulmonary atresia and ventricular septal defect, presenting with chronic cyanosis after pulmonary valve replacement and closure of the VSD at the age of 23 years and later interventional pulmonary valve replacement. Methods: His physical capacity is declining due to dyspnea, equivalent to NYHA Class III. Resting SpO₂ is 85 %. He is overweight (BMI 28.5). Echocardiography shows mildly decreased biventricular systolic function and right ventricular systolic pressure of 70-80 mmHg and paradox ventricular septal movement. Mean pulmonary valve gradient is 23 mmHg. No right to left shunt. Bodyplethysmography shows low vital capacity (24.2% of normal) and low total lung capacity (21.4% of normal). CPET shows a peak oxygen consumption of 8.6 ml/min/kg (25% of normal), low breathing reserve of 10% and a drop in SpO₂ to 74%. Computertomography and MRI showed no pulmonary thrombosis, but RV hypertrophy, dilated proximal but rarefied distal pulmonary arteries and right sided diaphragmatic paresis. Polysomnography revealed episodes of hypopnea 10.6/h and obstructive apnea 0.6/h, snoring 101 min (24.2%). SpO₂ during sleep averaged 81% (minimum 68%). Resting CO₂ was 49 mmHg; with increase at sleep to average 62 mmHg (maximum 73 mmHg). A polysomnography using a non-invasive CPAP mask showed significant reduction of obstructive apnea to 0.1/h, CO₂ was much less with a mean of 56 mmHg (maximum 60 mmHg) and SpO₂ level was higher (mean 85.9%). Results: A cyanotic patient with no remaining right to left shunt and secondary pulmonary arterial hypertension and relevant restrictive lung disease due to diaphragmatic paresis and multiple thoracotomies shows moderate to severe respiratory and mild circulatory restriction, a drop of SpO₂ at exercise. Hypopnea and obstructive apnea during sleep cause relevant worsening of CO₂ and SpO₂. All of these symptoms are improved by use of a CPAP-mask. Conclusion: Sleep-apnea may aggravate the symptoms of chronic lung disease and pulmonary arterial hypertension, this should be recognized by polysomnography. Symptoms and long term prognosis may effectively be treated using non-invasive CPAP.