Reversible pulmonary hypertension post adenotonsillectomy

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ABSTRACT

An 18-month-old boy presented with signs and symptoms of obstructive sleep apnea and pulmonary hypertension of 12 months duration. Confirmatory laboratory studies, in the form of echocardiography and overnight oximetry, were carried out, which showed hypoxemia and severe pulmonary hypertension. He had adenotonsillectomy, which resulted in complete resolution of signs and symptoms of pulmonary hypertension. Chronic upper airway obstruction should not be overlooked as it is a reversible cause of pulmonary hypertension.

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The association between upper airway obstruction and reversible lung changes is well known and previously reported in several cases.[1,2] This results in changes in the early morning blood gases in the form of hypoxia and hypercapnia. Pulmonary heart disease occurring in children with chronic upper airway obstruction, resulting from adenotonsillar hypertrophy was first reported in 1965.[1] Since then, cor pulmonale, secondary to upper airway obstruction by adenotonsillar hypertrophy has been well documented in children. This is thought to occur as a result hypoxic pulmonary vasoconstriction during periods of apnea, which may occur many times at night.[3] However, some cases may escape recognition, even though they have severe pulmonary hypertension when daytime arterial blood gases are within normal range. Here, we report such a case to alert clinicians to this possibility, and to investigate this further by tests, such as overnight oximetry, if there is any suspicion.

Case Report. An 18-month-old boy seen at the Outpatient Clinic of King Khalid University Hospital (KKUH), with the complaints of poor sucking, irritability, mouth breathing, night sweating, night snoring, and poor weight gain of 8 months duration. He had multiple admissions in different local hospitals with the same complaint, treated as upper respiratory tract infection, and received multiple courses of different antibiotics. However, he did not show any clinical improvement. He was a product of an uneventful pregnancy, and full term normal spontaneous vaginal delivery with no immediate natal or postnatal problems. He was vaccinated up-to-date with no side effects. He is the sixth child in his family with 4 normal sisters and one brother of non-consanguineous parents. On examination, he looked well and active, but in mild respiratory distress. He was pale, with no dysmorphic features. His head and neck were supple with no lymphadenopathy. He did not have clubbing. His vital signs included heart rate 118 beat/min, respiratory rate 40 breath/min, axillary temperature 36.4^{O}C, blood pressure 102/45 mm Hg, and pulse oximetry in room air approximately 95%. His growth parameters were: weight 7.2 kg, height

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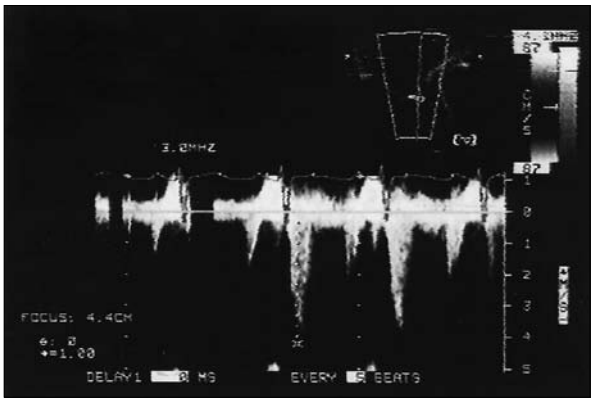

Figure 1 - Pre-operative 4 chamber view, using Doppler waves at tricuspid valve regurgitation jet, showing flow of 79 mm Hg.

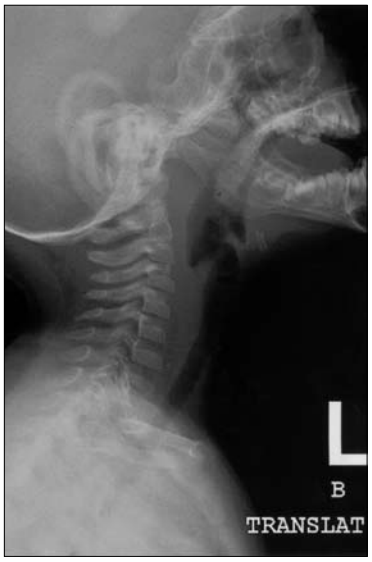

Figure 2 - Left post nasal space view demonstrating enlarged soft tissue of the adenoid.

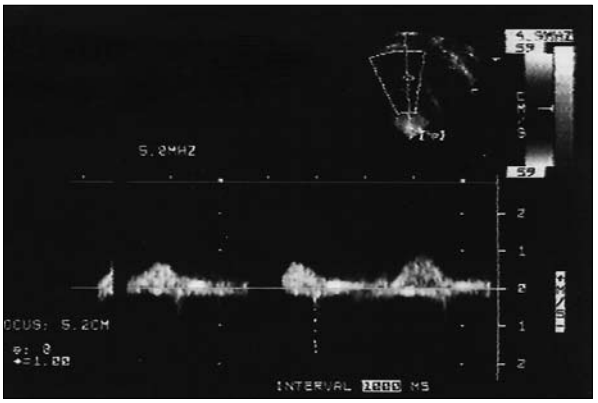

Figure 3 - Postoperative, 4 chamber view, using Doppler waves at tricuspid valve regurgitation jet, showing a peak systolic flow of 10 mm Hg

71 cm, head circumference 46.5 cm, all below the 5th percentile for his age. The ear, nose, and throat examinations revealed large adenoids. Chest examination revealed no chest deformity, and his breath sounds were equally heard. Cardiovascular examination showed hyperactive pericardium with normal first heart sound, accentuated second heart sound with no murmurs. The abdomen was soft and lax, liver was 4 cm below the right costal margin, and the liver span was 8 cm. Spleen was just palpable. Central nervous and locomotor systems were grossly intact. Investigation revealed hemoglobin of 93 g/L with a microcytic hypochromic picture. His arterial blood gas was within normal range. His chest radiograph revealed moderate cardiomegaly. Electrocardiography showed right atrial and ventricular hypertrophy. Echocardiography revealed normal heart anatomy, right ventricular hypertrophy and dilatation, mild tricuspid valve regurgitation with an estimated right ventricular systolic pressure of 70 mm Hg. A small patent ductus arteriosus with minimal left to right shunting (**Figure 1**) was noted. Overnight oximetry revealed desaturation down to 70%, polysomnography was attempted but he was not cooperative. The clinical findings were highly suggestive of obstructive sleep apnea (OSA) complicated by pulmonary hypertension. Lateral neck radiograph revealed complete obliteration of his nasopharynx with adenoidal tissue (**Figure 2**) and fibre optic nasopharyngoscopy revealed large adenoidal tissue obstructing the posterior choana. Therefore, adenotonsillectomy was recommended. Three months later after adenotonsillectomy, repeated echocardiography showed trivial tricuspid regurgitation, which showed a gradient of only 10 mm Hg, namely, decreased right ventricular systolic pressure (**Figure 3**).

Discussion. Obstruction at any site along the upper airway, may cause pulmonary hemodynamic disturbances. This is seen in craniofacial abnormalities, micrognathia, cleft palate, glottic web, vocal cord paralysis, glossoptosis, nasal septal deviation, and the Pierre-Robin syndrome.[4] However, the most common cause of chronic airway obstruction and sleep related obstructive phenomena in children, is hypertrophy of the nasopharyngeal lymphoid tissue, specifically the adenoids and tonsils. As the case in our patient, repeated episodes of obstruction resulted in increased pulmonary vascular resistance that becomes later persistently elevated, with resultant dilatation and hypertrophy of the right side of the heart, and the eventual development of congestive heart failure.[5] Hypoxemic pulmonary hypertension, cor pulmonale,

pulmonary edema and failure to thrive, as seen in our patient, are well documented sequela of chronic upper airway obstruction, requiring multiple admissions, antibiotic therapy, intubation, and assisted ventilation in the intensive care unit.[2] Maurizi et al[6] reported that 65.7% of clinically normal children with adenoid hypertrophy showed pulmonary function abnormalities. There was significant reduction in mid-inspiratory flow of (forced inspiratory flow 50%) and oxygen saturation, and these improved after adenotonsillectomy.[7] The association between OSA, and cardiovascular disease was first raised by observational studies linking snoring, a surrogate for OSA, with increased cardiac events. A recent sleep clinic study, reported a linear relationship between hypertension and severity of OSA with each extra apneic episode per hour increasing the odds of hypertension by 1%.[8] Our patient did not show any hypercapnia in early morning blood gas, but did show desaturation down to 70% in the overnight oximetry. Brouillette et al,[9] found that a positive pulse oximetry result increased the probability of a patient having OSA from 60-97%. Tricuspid valve regurgitation can be calculated easily using Doppler echocardiography; its mean pressure reflects the mean pulmonary artery pressure. It was easily measured in our case before, and 3 months post adenotonsillectomy. This noninvasive cardiac evaluation is an alternative to cardiac catheterization, offers a reliable method for routine evaluation and can be used to assess pulmonary hypertension in any suspected case.[10] Resolution of pulmonary arterial hypertension as documented by Doppler echocardiography, confirmed the response to adenotonsillectomy. Our case also demonstrates the usefulness of combined overnight oximetry and Doppler echocardiography, in evaluation of suspected OSA when polysomnography is difficult to perform. We therefore recommend the use of these 2 simple and reproducible tests in children with a suggestive history of OSA, secondary to tonsillar or adenoidal tissue hypertrophy, or both, prior to its surgical removal.

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