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PULMONARY HYPERTENSION DUE TO CHRONIC UPPER AIRWAY OBSTRUCTION: A CLINICAL REVIEW AND REPORT OF FOUR CASES*

*Dolli Yafet Aji MD***, Ayşe Sarıoğlu MD***, Lale Sever MD****
*Nil Arısoy MD*****

SUMMARY: Aji DY, Sarıoğlu A, Sever L, Arısoy N. (Department of Pediatrics, Cerrahpaşa Faculty of Medicine, and Institute of Cardiology, İstanbul University, İstanbul, Turkey). Pulmonary hypertension due to chronic upper airway obstruction: clinical review and report of four cases. Turk J Pediatr 33: 35-41, 1991.

Partial airway obstruction due to the enlargement of the tonsils and adenoids is a well recognized clinical entity, but cardiorespiratory changes due to chronic obstruction have infrequently been reported. Four children with severe nasopharyngeal obstruction due to tonsil and adenoid hypertrophy, who developed pulmonary hypertension and cardiac failure, were studied. Relief of upper airway obstruction by adenotonsillectomy resulted in a regression of the presenting signs and symptoms.
Key words: adenoid hypertrophy, tonsil hypertrophy, pulmonary hypertension, adenotonsillectomy.

Hypoventilation is listed as one of the causes of pulmonary hypertension. Obstructive respiratory tract disease with hypoxia, hypercapnia and acidosis is extremely rare in childhood, except in patients with chronic asthma and cystic fibrosis[1]. Pulmonary hypertension resulting from upper airway obstruction has recently been reported in children[2-7]. The present report describes the clinical course of four children with tonsil and adenoid hypertrophy.

Case Reports

Case 1

A 3-year-old boy was admitted to our hospital in January 1987, with complaints of frequent upper respiratory tract infection, sleep associated stertorous breathing and shortness of breath since infancy.

On examination, while awake, the child appeared dull. The mouth was held open and there was an audible snoring sound with respiration. Adenoid facies, perioral cyanosis and pretibial edema were present. He was 89 cm in height and weighed 14.3 kg. The heart rate was 140/min, and respiration rate, 38/min. The blood

* From the Department of Pediatrics, Cerrahpaşa Faculty of Medicine, and Institute of Cardiology, İstanbul University, İstanbul.

** Chief Resident in Pediatrics, Cerrahpaşa Faculty of Medicine, İstanbul University.

*** Chief Resident in Pediatric Cardiology, Institute of Cardiology, İstanbul University.

**** Professor of Pediatrics, Cerrahpaşa Faculty of Medicine, İstanbul University.

pressure was 110/60 mmHg, and axillary temperature, 37°C. During sleep the child was cyanotic and diaphoretic, and exhibited supraclavicular and intercostal retractions which disappeared while awake. The tonsils and adenoids were almost occluding the oropharynx.

On auscultation, the pulmonary valve closure sound was moderately accentuated, and a short grade II systolic murmur was heard over the pulmonary area. Crepitations were present on both lung fields. The firm liver edge was palpable four cm below the right costal margin.

Laboratory data revealed a hemoglobin level of 8 g/dl, hematocrit 42% and white blood cell count 11,200/mm^3. The urine analysis was normal.

The chest X-ray showed a cardiothoracic ratio of 0.66, with pulmonary congestion. The electrocardiogram (ECG) revealed right axis deviation, peaked p waves of right atrial hypertrophy and right ventricular hypertrophy (Fig. 1). Two-dimensional echocardiography revealed no intracardiac pathology, except for right atrial and ventricular enlargement. M-mode tracings showed horizontal positioning of the pulmonary valve and disappearance of the "a" wave, leading to a diagnosis of pulmonary hypertension.

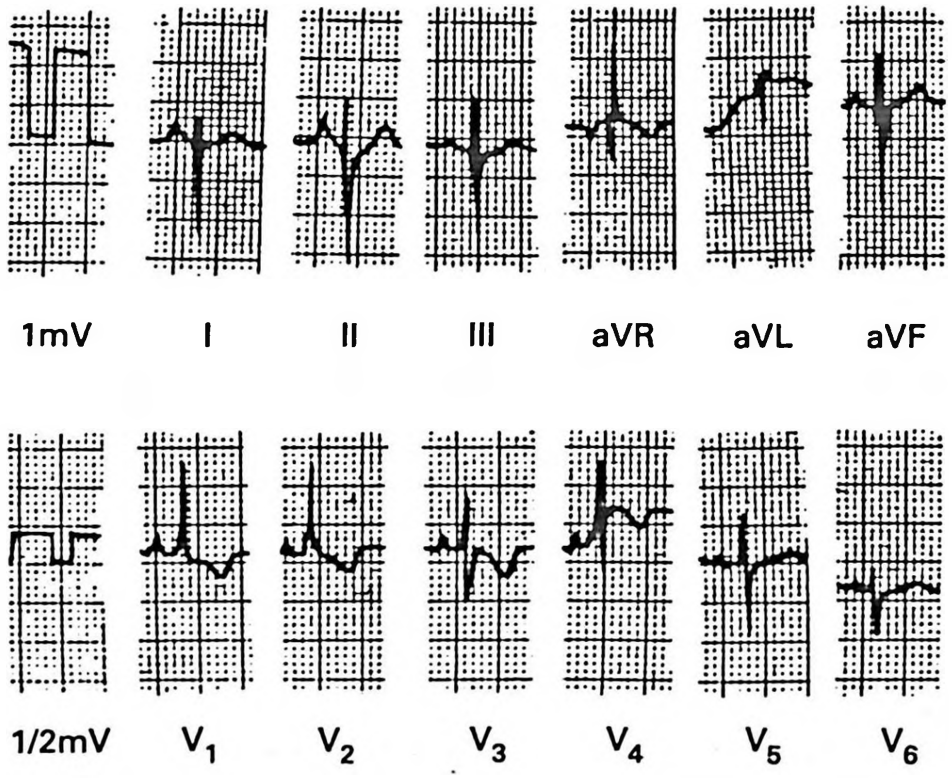

Fig. 1: Electrocardiogram of Case 1.

A diagnosis of pulmonary hypertension and congestive heart failure secondary to upper airway obstruction was made, and the patient was treated with digoxin and diuretics. On the seventh day of admission, adenotonsillectomy was performed which was followed by rapid improvement. The medical treatment was discontinued. One year later, the clinical and laboratory findings were completely normal (Fig. 2).

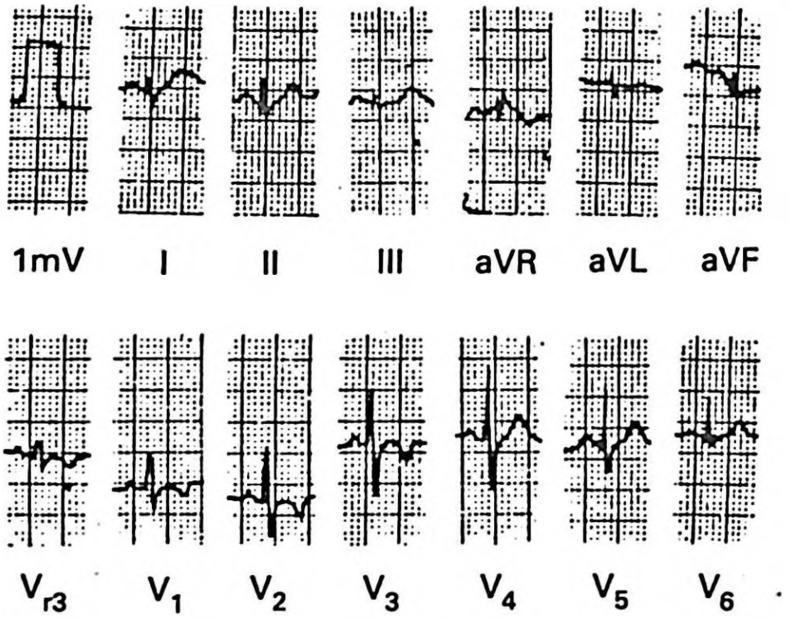

Fig. 2: Electrocardiogram of Case 1 following medical treatment.

Case 2

A 2 1/2-year-old boy was admitted to our hospital in March 1988, with complaints of shortness of breath and difficulty in breathing, especially at night, of two months' duration. He had been referred from Trabzon Community Hospital (a mountainous city on Turkey's Black Sea Coast) for cardiologic evaluation.

Physical examination revealed a boy who was 86 cm in height and weighed 14 kg. He had adenoid facies and a plethoric appearance. During sleep he snored, and apnea and cyanosis developed which awakened the child.

The heart rate was 120/min, and respiration rate 42/min. The blood pressure was 110/80 mmHg, and axillary temperature 38°C. The adenoids and tonsils were extremely enlarged, and acute tonsillitis was present. The pulmonary valve closure sound was accentuated but there were no heart murmurs. Coarse rales were detected over the lungs, especially at the bases. The liver was palpable two cm below the right costal margin.

The hemoglobin was 14 g/dl, hematocrit 55% and white blood cell count 7000/mm^3. The chest X-ray revealed increased interstitial markings and a cardiothoracic ratio of 0.61. ECG showed right axis deviation and peaked p waves.

The two-dimensional echocardiogram revealed no intracardiac pathology other than right ventricular enlargement. M-mode tracings showed pulmonary hypertension with horizontal positioning of the pulmonary valve in diastole and disappearance of the "a" wave.

The patient was treated with antibiotics for the tonsillitis. On the sixth day of admission, adenotonsillectomy was performed, followed by dramatic clinical improvement, a decrease in heart size on X-ray, and a regression of electrocardiographic findings.

Case 3

A 22-month-old boy was admitted to our hospital in November 1988, with complaints of shortness of breath, orthopnea and stertorous sleep of over one year's duration. Swelling of the face, neck and legs had developed one week before his admission to a local hospital. A diagnosis of congestive heart failure was made, and he was treated with digoxin, prednisolone and antibiotics, with no clinical improvement noted. He was then referred to our hospital for cardiologic evaluation.

Physical examination revealed an underdeveloped boy who was 74 cm in height and 8 kg in weight. He was lethargic, and cyanosis and edema on the face and legs were present. The respiration rate was 40/min and irregular. The pulse rate was 100/min, and a grade II systolic murmur was heard over the pulmonary area. The pulmonary valve closure sound was accentuated. The liver was palpable six cm below the right costal margin. The tonsils and adenoids were enormous, and postnasal purulent dripping was present.

Laboratory data showed a hemoglobin of 10 g/dl, hematocrit 38%, and white blood cells 9400/mm^3.

Arterialized blood showed a normal pH, PCO_2 of 85.7 mmHg, PO_2 61.4 mmHg and HCO_3, 58 mmol/1.

ECG showed right axis deviation, p pulmonale, right ventricular hypertrophy and negative T waves in V_5 and V_6. The chest X-ray showed an enlarged heart with a cardiothoracic ratio of 0.59. Pulmonary congestion was present.

Right ventricular and right atrial enlargement were the only intracardiac pathological features which appeared in two-dimensional echocardiography. M-mode tracings of the pulmonary valve illustrated a horizontal "e-f" slope in diastole and absence of the "a" wave, evidence of pulmonary hypertension.

A diagnosis of pulmonary hypertension and congestive hert failure secondary to upper airway obstruction was made. The patient was treated with antibotics, digoxin and diuretics, and was maintained in an upright position. On the ninth day of admission, he underwent adenotonsillectomy, followed by improvement in his clinical status, chest X-ray and electrocardiogram.

Case 4

A 6-year-old girl was admitted to our hospital in May 1989, with complaints of shortness of breath, chest pain and swelling of the face and feet. Frequent upper respiratory infections, stertorous breathing and a disturbed sleep pattern were described.

Physical examination revealed a girl who was 100 cm in height and 22 kg in weight. Dyspnea, perioral cyanosis, increasing during sleep, edema on the face and legs and ascites were present. The tonsils and adenoids were extremely enlarged. The respiration rate was 52/min and the heart rate, 124/min. The pulmonary valve closure sound was accentuated but there were no heart murmurs. The blood pressure was 110/70 mmHg. Crepitations were present on both lung fields. The liver was enlarged five cm below the right costal margin.

Laboratory data showed a hemoglobin level of 11.6 g/dl, hematocrit 37% and white blood cell count 7800/mm^3. Urine analysis was within normal limits.

The chest X-ray showed a cardiothoracic ratio of 0.62, with pulmonary congestion. ECG revealed right axis deviation, peaked p waves and right ventricular hypertrophy.

The cross-sectional echocardiogram showed right atrial and ventricular enlargement and leftward bulging of the atrial septum. In M-mode echocardiograms of the pulmonary valve, the "a" wave was missing, the "b-c" slope appeared more vertical and the "e-f" slope more horizontal than normal which suggest a diagnosis of pulmonary hypertension.

The child was treated with digoxin and diuretics for heart failure. The dyspnea and edema disappeared on the fourth day of treatment, and adenotonsillectomy was performed on the tenth day of admission, followed by rapid improvement. The patient is now leading a normal life, and does not use any medication.

Discussion

Partial upper airway obstruction due to tonsil and adenoid hypertrophy is frequently encountered in children between two and five years of age[6]. Infection and allergy effect its severity.

Cor pulmonale due to chronic upper airway obstruction in children was first reported by Menashe et al[7] in 1965. However, children with enlarged tonsils and adenoids do not all develop pulmonary hypertension. Talsat et al[8] reported only two cases of cor pulmonale among thirty such cases.

Wilkinson et al[9] have examined 92 patients with tonsil and adenoid hypertrophy and diagnosed pulmonary hypertension in only 3.3 percent of them. Macartney et al[10] have suggested that pulmonary vasculature is hyperreactive towards hypoxemia in some children, as seen in 20% of people who live at high altitudes.

Other factors, such as race and central nervous system pathology leading to abnormal respiration control have also been suggested[3].

Figure 3 shows the mechanism involved in upper airway obstruction which leads to pulmonary hypertension and heart failure.

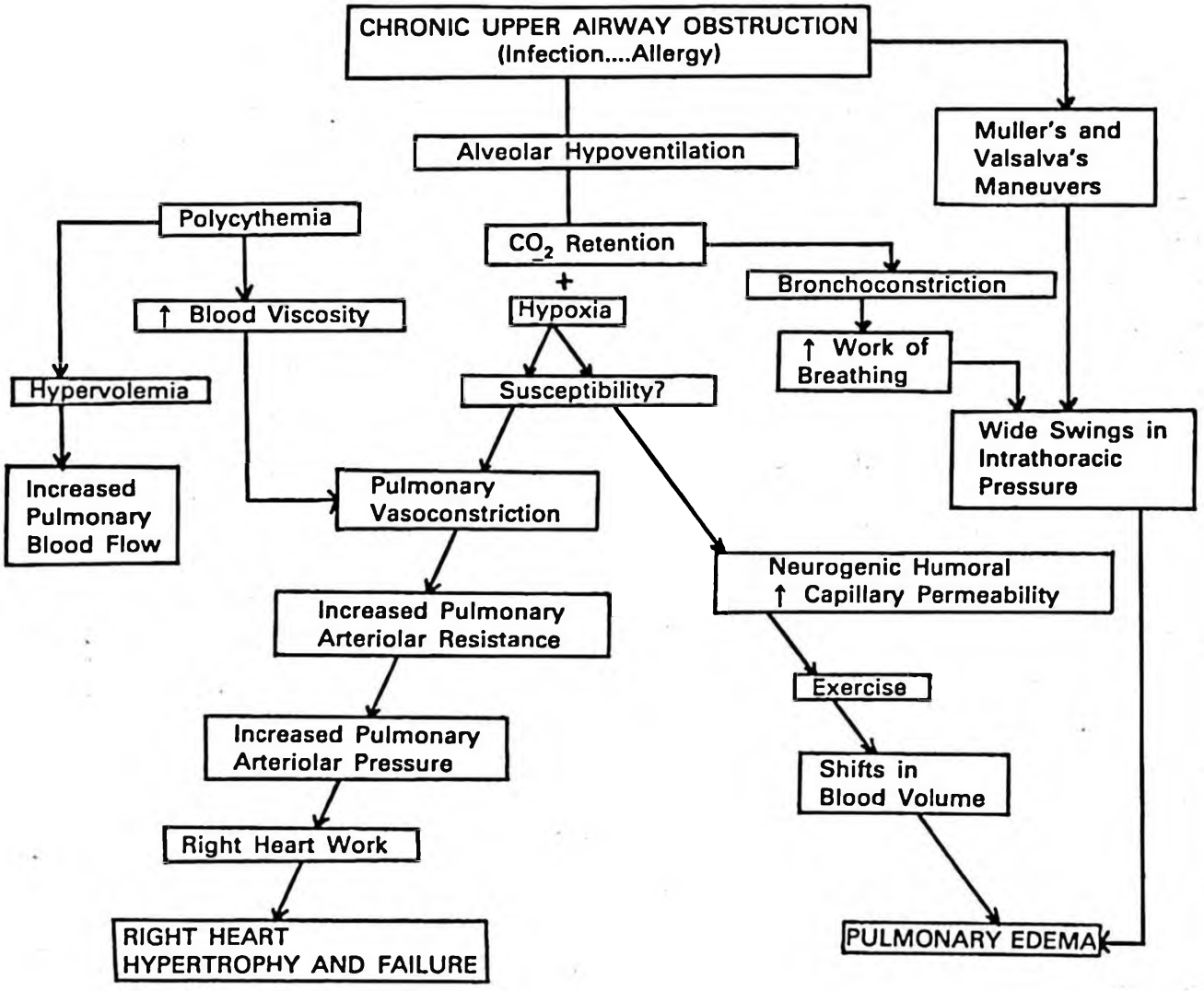

Fig. 3: Flow chart showing the mechanism of pulmonary hypertension and heart failure in upper airway obstruction.

Patients with tonsil and adenoid hypertrophy who develop pulmonary hypertension present with noisy mouth breathing, dangerous nocturnal disturbance of sleep leading sometimes to sleep-associated apnea, lethargy, rhinitis, cyanosis, clubbing, respiratory distress, cardiomegaly, right axis deviation, congestive heart failure, pulmonary accentuated valve closure, hypoxemia and hypercapnia, as demonstrated in our four cases.

It is well known that polycythemia which results from residing at high altitude causes pulmonary hypertension[11,12]. Therefore, polycythemia may well have played a role in the development of pulmonary hypertension in our second case,

who lived in the mountainous region of the Black Sea Coast, in addition to hypoxemia, due to chronic upper airway obstruction.

Cor pulmonale due to tonsil and adenoid hypertrophy is reversible. After adenotonsillectomy, clinical and laboratory findings rapidly improve, as was observed in our four cases. The only risk factor reported during the operation is sudden death caused by the anesthesia[9].

On clinical grounds, the relation between tonsil and adenoid hypertrophy and cardiorespiratory pathology can easily be overlooked. Assessing these children as primary heart patients and postponing adenotonsillectomy for thorough cardiologic evaluation could lead to loss of time, and is important in prognosis.

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