Failure to Thrive Secondary to Upper Respiratory Tract Obstruction and Cor Pulmonale

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Although the syndrome of intermittent upper airway obstruction with pulmonary hypertension and cor pulmonale has been well described, recognition of the syndrome prior to the onset of heart failure remains difficult. Poor growth, which previous reports have frequently noted but have not emphasized, precedes the cardiac complications and can provide a valuable clue to earlier diagnosis and treatment.

In 1965 Menashe et al[1] and Cox et al[2] first described the childhood syndrome of intermittent upper airway obstruction with pulmonary hypertension and cor pulmonale, which has since been reported as secondary to a variety of causes including adenoidal and tonsillar hypertrophy[3] and the Pierre-Robin syndrome.[4] It is extremely important for all primary care practitioners to recognize the syndrome, because delay in diagnosis and treatment can lead to heart failure and death, while simple measures that alleviate the airway obstruction result in rapid resolution of symptoms.[5-8] The illness has not received attention in the family practice literature, and despite its description in both pediatric and otolaryngology journals, recognition of the disease prior to the onset of cardiac symptoms remains difficult. Many symptoms of intermittent obstruction (eg, mouth breathing and snoring) are commonly encountered by the family physician, who must distinguish the rare child who may have cor pulmonale from the many who do not.

If physicians are to recognize this syndrome prior to severe complications, earlier occurring symptoms need to be emphasized. Mangat et al[9] described four children with hypersomnolence, sonorous breathing, recurring upper airway infection, sleep disturbances, and enlarged tonsils and adenoids. Direct observation of apneic episodes resulted in diagnosis prior to cor pulmonale. Another clue to diagnosis can be the poor weight gain exhibited by many of these children. Although previous reports have mentioned failure to thrive, authors have not emphasized this aspect of the syndrome.[5,7,10,11]

Recently, the authors have encountered two children who presented with failure to thrive. Other signs and symptoms eventually led to the diagnosis of intermittent upper airway obstruction with cor pulmonale. Treatment resulted not only in rapid resolution of the symptoms of obstruction, but also in dramatic improvement in growth.

Case Reports

Case 1

A 14-lb, seven-month-old white boy was first seen at the University Hospital Outpatient Department with chief complaints of "funny breathing" and failure to gain weight. The product of a

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normal pregnancy, labor, and delivery, he weighed 8 lb 10 oz at birth. He was well until nine weeks of age, when he was hospitalized in a community hospital for an apparent viral pneumonia. Although the patient recovered uneventfully, he subsequently developed frequent simple upper respiratory tract infections without otitis media. His parents also began to note intercostal and substernal retractions. Respirations did not appear labored, nor did he seem to have an increased respiratory rate. Although concerned about his unusual breathing pattern, the parents did not seek additional consultation until they perceived that the child was not gaining weight properly.

On physical examination the patient appeared small for his age, breathed through his mouth, and had slight substernal and intercostal retractions. He was active, alert, and did not appear to be chronically ill. His weight was just below the 3rd percentile, while his length was at the 25th percentile. Physical examination was otherwise normal. Chest x-ray examination revealed chronic interstitial infiltration with increased right heart size. Electrocardiogram (ECG) showed right ventricular hypertrophy, as did echocardiogram. Sweat chloride was normal. Otolaryngological examination revealed adenoidal hypertrophy. Because of the child's breathing pattern, the increased heart size, and the adenoidal hypertrophy, the diagnosis of upper airway obstruction with cor pulmonale was made. After simple adenoidectomy was performed, the sternal and intercostal retractions ceased. Within two months, the infant's weight was at the 10th percentile. Four months after surgery, right heart size was normal by repeat echocardiogram. At 3 1/2 years of age, the child was at the 25th percentile for weight and was free of pulmonary and cardiac problems.

Case 2

A boy of Korean parentage was referred to the University Hospital Outpatient Department at the age of 22 months for evaluation of failure to thrive. The product of a normal pregnancy, labor, and delivery, he weighed 7 pounds at birth, was 20 inches long, and was noted to have a cleft palate.

He grew well and developed normally. At 14 months he was at the 25th percentile for length and the 15th percentile for weight. At 17 months, the child's palate was repaired, and a velopharyngeal flap was placed to facilitate development of normal speech. Examination five months after surgery revealed that the child had no linear growth and had lost weight. He was referred for evaluation.

Parental history revealed that the child was snoring, mouth breathing, and having alternate fast and slow breathing while asleep. He had dyspnea on exertion, poor appetite, nasal congestion, and recurrent colds. All these complaints were first noted after surgery. With the exceptions of the child's small size and repaired palate, physical examination was normal. Complete blood count, erythrocyte sedimentation rate (ESR), urinalysis, electrolytes, blood urea nitrogen (BUN), and glucose were also normal. Chest x-ray examination showed no evidence of cardiac enlargement. Although by history the child had indications of intermittent upper airway obstruction, a causative relationship between this and his growth failure was not appreciated. As the child appeared well and all laboratory findings were normal, the authors elected to observe him.

Over the next four months, the child's growth did not improve. Repeat chest x-ray films showed normal heart size and contour; however, ECG revealed right axis deviation and severe right ventricular hypertrophy. Blood gases were normal.

The child was admitted to the hospital. Nasal pharyngeal fluoroscopy indicated that the airway was normal on inspiration, but on expiration, the retropharyngeal soft tissues met the tongue anteriorly, producing obstruction and noisy respirations. Because of the right ventricular hypertrophy and the evidence of upper airway obstruction, the velopharyngeal flap was removed. The patient was last seen at 33 months of age, and although still below the third percentile for height and weight, his growth acceleration was remarkable (Figure 1). He had normal activity, improved appetite, and no longer snored.

Discussion

Although failure to thrive has been described in conjunction with cor pulmonale secondary to upper airway obstruction, this aspect of the syndrome has not been emphasized. Poor growth may occur in as many as 30 percent of children with significant upper airway obstruction. A literature review by Levin et al[11] indicated that 14 of 44 patients reported to have this syndrome experienced

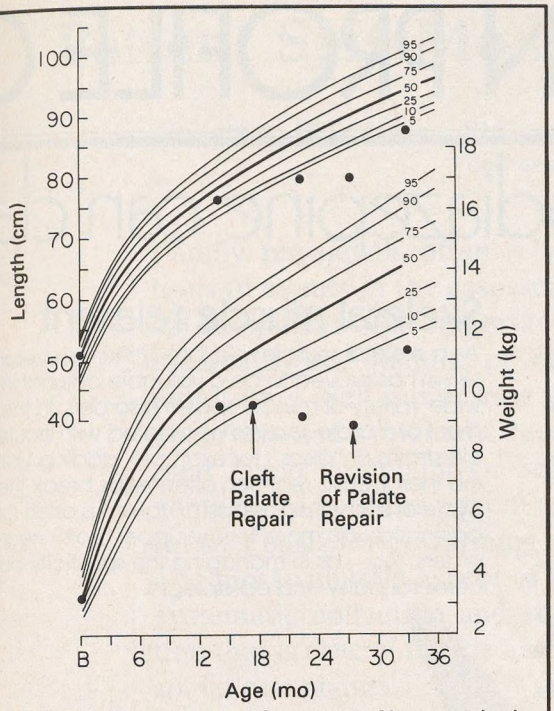

Figure 1. Growth chart for case 2. Note marked growth acceleration after relief of airway obstruction

growth failure. Three of the nine cases they presented were also failing to thrive. The only more frequently described historical or physical finding in these children was increased symptoms in the supine position.

These patients were strikingly similar to two which have been previously reported. Cox et al[2] described an infant with tracheomalacia whose growth curve was much like that seen in the first case. The child's weight, while normal at birth, fell below the third percentile and then rapidly returned to normal after the obstruction was relieved by tracheostomy. Robson et al[5] reported a four-year-old child who, like the second case, presented with failure to thrive after correction of a cleft palate. Again the patient's weight rapidly returned to normal after the airway obstruction was relieved.

Animal experiments support the correlation between intermittent respiratory tract obstruction and failure to thrive. After three weeks of alternate exposure to a mixture of hypoxic, hypercarbic air for eight hours, then to room air for 16 hours, rats had developed both pulmonary hypertension and growth failure.[12] McGrath et al[13] have shown that in rats hypoxia alone results in both decreased weight gain and right ventricular hypertrophy. Although these experiments indicate that chronic intermittent hypoxia alone could cause the growth failure seen in these children, increased caloric expenditure from the effort of breathing or decreased caloric intake may also play a role.

Certainly, recognition of the correlation between intermittent upper airway obstruction and failure to thrive would have helped the authors diagnose and treat the second case four months earlier. As these cases and those reviewed indicate, the growth failure, when it occurs, appears before the more severe complications of congestive heart failure, pulmonary edema, and death. Because the disease is easily remediated by surgery, it is the difficulty in diagnosis which leads to poor outcomes in these children. Consideration of the syndrome in children with signs of upper respiratory tract obstruction and growth impairment can lead to earlier diagnosis and treatment in as many as one third of children with this disease.

References

1. Menashe VD, Farrehi C, Miller M: Hypoventilation and cor pulmonale due to chronic upper airway obstruction. J Pediatr 67:198, 1965

2. Cox MA, Scheibler CL, Taylor WJ, et al: Reversible pulmonary hypertension in a child with respiratory obstruction and cor pulmonale. J Pediatr 67:192, 1965

3. Goodman RW, Goodman M, Goodman N, Cohen H: Cardiac and pulmonary failure secondary to adenotonsillar hypertrophy. Laryngoscope 86:1367, 1976

4. Cogswell JJ, Easton MD: Cor pulmonale in the Pierre-Robin syndrome. Arch Dis Child 49:905, 1974

5. Robson MC, Stankiewicz JA, Mendelsohn JS: Cor pulmonale secondary to cleft palate repair. Case report. Plast Reconstr Surg 59:754, 1977

6. Gvertz KK, Mattiolo L: Cardiorespiratory syndrome. Manifestations in children with chronic upper airway obstruction. J Kans Med Soc 80:126, 1979

7. Ainger LE: Large tonsils and adenoids in small children with cor pulmonale. Br Heart J 30:356, 1968

8. Kravath RE, Pollack CP, Borowiecke B, Weitzman ED: Obstructive sleep apnea and death associated with surgical correction of velopharyngeal incompetence. J Pediatr 96:645, 1980

9. Mangat D, Orr WO, Smith RO: Sleep apnea, hypersomnolence, and upper airway obstruction secondary to adenotonsillar enlargement. Arch Otolaryngol 103:383, 1977

10. Levy AM, Tabakin BS, Hanson JS, et al: Hypertrophied adenoids causing pulmonary hypertension and severe congestive heart failure. N Engl J Med 277:506, 1967

11. Levin DL, Muster AJ, Pachman LM, et al: Cor pulmonale secondary to upper airway obstruction. Chest 68: 166, 1975

12. Nattie EE, Bartlett D, Johnson K: Pulmonary hypertension and right ventricular hypertrophy caused by intermittent hypoxia and hypercapnia in the rat. Am Rev Respir Dis 118:653, 1978

13. McGrath JJ, Prochazka J, Pelough V, Ostadal B: Physiologic responses of rats to intermittent high altitude stress: Effects of age. J Appl Physiol 34:289, 1973