noninvasiveness, its low false-positive rate, its robustness, and simplicity it should be seriously considered as a possible method of screening for cystic fibrosis. It appears to be the best method currently available.

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Upper airways obstruction

Presentation with systemic hypertension

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SUMMARY Of 14 patients whose final diagnosis was upper airways obstruction associated with heart failure, 3 presented with systemic hypertension (up to 200/100 mmHg). In 2 the hypertension was so severe that at first it had to be considered as a possible cause of the presenting symptoms. The subsequent history indicated that it was an effect of the upper airways obstruction with heart failure.

Upper airways obstruction can lead to pulmonary arteriolar vasoconstriction, pulmonary arterial hypertension, cor pulmonale, and right heart failure.¹⁻⁴ We describe 3 children who were part of a group of 14 children who presented consecutively with upper airways obstruction associated with heart failure.

Case report

Case 1. A 2-year-old boy was admitted to hospital, semi-comatose, with a 3-day history of upper respiratory infection. Blood pressure (BP) was 160/120 mmHg* and there were signs of severe congestive heart failure. Chest x-ray film showed

gross cardiomegaly and signs of pulmonary oedema. Electrocardiogram (ECG) showed a normal QRS axis of +70°, right atrial P-waves, and left ventricular hypertrophy pattern. He was treated with intravenous diuretics and digitalis, and was given artificial ventilation using a respirator. Before artificial ventilation arterial Pco₂ was 72.5 mmHg (9.6 kPa), Po₂ 63.6 mmHg (8.5 kPa), and pH 7.23. After 3 hours of artificial ventilation, Pco₂ was 46 mmHg (6.1 kPa), Po₂ 149.5 mmHg (19.9 kPa), and pH 7.45. BP remained high (150/120 mmHg) and he remained semi-comatose. His fundi were normal but a lumbar puncture showed an opening pressure of 400 mm cerebrospinal fluid. BP fell after antihypertensive medication. By day 3 he was awake with no respiratory distress and his BP had stabilised (about 110/60 mmHg) without treatment. Renal arteriogram and vanillyl-mandelic acid (VMA) levels (0.7 mg/24 h) were normal. Because of large tonsils and adenoids it was felt that upper airways obstruction might be responsible for the symptoms, and removal of tonsils and adenoids was planned but the appointment was not kept.

The boy did not return until age 3½ when he was again admitted with a history of upper respiratory infection, in severe respiratory distress, semi-comatose, and in congestive heart failure. Again he recovered, removal of tonsils and adenoids was again planned, but again the appointment was not kept. He

*The equipment used in this and in the subsequent cases was a standard mercury sphygmomanometer and a cuff that covered at least two-thirds of the upper arm. Systolic and diastolic pressures were recorded at the appearance and disappearance (first and fifth phases) of Korotkoff's sounds.

was brought in with a similar history at age 5 and developed cardiorespiratory arrest, needing resuscitation and 5 days of artificial ventilation. After weaning from the respirator he was intolerant of extubation and a tracheostomy was performed. BP on this occasion fluctuated between 130/90 and 100/60 mmHg. His ECG showed right axis deviation of $+130^\circ$, right atrial P-waves, and combined ventricular hypertrophy pattern. He was still hypertensive and hypercarbic at cardiac catheterisation which was done 3 weeks after admission (Table). He underwent removal of tonsils and adenoids but his condition did not improve. Laryngoscopy showed subglottic stenosis, confirmed by contrast x-rays, and was treated by progressive dilatation of the trachea. His BP is now about 90/60 mmHg and he remains asymptomatic.

Case 2. This obese 3-year-old boy was brought to hospital because of dizziness, vomiting, diarrhoea, and gradually increasing respiratory distress over 24 hours. He had been falling asleep very easily and preferred to sleep with 2 or 3 pillows; he occasionally snored. On admission he was asleep but could be roused; he was in moderate respiratory distress, and had signs of congestive heart failure. BP was 200/100 mmHg. There was a grade 2/6 systolic murmur at the left sternal border to apex. The tonsils were not enlarged. The ear drums were inflamed. Chest x-ray film showed cardiomegaly and signs of pulmonary oedema. ECG showed right axis deviation of $+150^\circ$, right atrial enlargement, and right ventricular hypertrophy pattern.

Arterial Po_2 was 47 mmHg (6.2 kPa), Pco_2 44 mmHg (5.8 kPa), and pH 7.48. Electrolytes, Addis count, 17-hydroxycorticosteroids, and 17-ketosteroids were normal. Cortisol was 38.8 $\mu\text{g}/100$ ml (1070 nmol/l) in the evening, and 46.7 $\mu\text{g}/100$ ml (1314 nmol/l) in the morning. VMA was 1.6 mg/24 h; metanephrine and normetanephrine 0.09 mg/24 h. Renin from right renal vein was 3.2 ng/ml per hour

and from left renal vein 4 ng/ml per hour, these high renin levels being consistent with 8 days of diuretic therapy. Timed intravenous pyelogram was normal. Tomograms of pharynx showed adenoidal hypertrophy and enlarged posterior pharyngeal soft tissues. He was treated with digitalis and diuretics, and placed on a salt-restricted diet, after which BP decreased to about 120/70 mmHg. The congestive heart failure slowly responded to treatment. At cardiac catheterisation (Table) he was normotensive but hypoxic and hypercarbic in room air. Angiography showed a morphologically normal heart, and a normally contracting left ventricular myocardium—mean rate of circumferential fibre shortening 1.31 circumference/second;⁵ however the child was receiving digitalis at the time of the study. After review of upper airway tomograms it was agreed that adenoidal hypertrophy and enlarged posterior pharyngeal tissues were the cause of upper airways obstruction. Removal of tonsils and adenoids was done and the condition of the patient was improving at the time of discharge; his heart was still enlarged on x-ray film but his exercise tolerance had greatly increased. At this time ECG showed right axis deviation of $+120^\circ$ and combined ventricular hypertrophy pattern. One month after discharge BP was 100/60 mmHg.

Case 3. This 4½-year-old boy was admitted to the paediatric cardiology department after elective surgery to remove his tonsils and adenoids had been postponed because he had been found to be in congestive heart failure. During transfer he had an apnoeic spell and on arrival was sweating and stuporose; he was in severe congestive heart failure with moderate respiratory distress. BP was 150/100 mmHg. He had a grade 2/6 systolic ejection murmur at the apex. Chest x-ray film showed cardiomegaly and signs of pulmonary oedema. ECG showed right axis deviation of $+160^\circ$, right atrial P-waves, and right ventricular hypertrophy pattern. Arterial Po_2

Table Condition of patients at cardiac catheterisation

Case	Age (years)		Pressures (mmHg)		Arterial sat %	Arterial blood gases (mmHg)		
			Pulmonary artery	Aorta		pH	Po_2	Pco_2
1	5	Intubation, room air	33/9 (19)	116/87 (90)	88–94	7.36	91	57
2	3	Room air	43/22 (15)	103/73 (78)	94	7.38	56	46
		Crying	—	—	—	7.49	82	38
		Added O_2	—	—	—	7.36	238	45
3	5	Awake	—	—	—	7.35	62	57
		Asleep	—	—	—	7.15	53	85
3*		Room air	25/7 (13)	92/56 (75)	98	—	—	—
		Room air	—	—	—	7.37	93	40

*5 months after operation. Mean pressure is shown in brackets.

was 62 mmHg (8.2 kPa), P_{CO_2} 57 mmHg (7.6 kPa), and pH 7.35. The boy's condition improved after treatment with digitalis and diuretics. There was a weight loss of 2.5 kg but BP remained raised, at least up to the day of surgery, in the region of 150/115 mmHg. The tonsils and adenoids were removed on his 3rd day in hospital with marked symptomatic relief. Five months later he underwent cardiac catheterisation (Table). BP was normal, 92/56 mmHg.

Discussion

The initial presence of systemic hypertension in these patients complicated the diagnosis. Was the hypertension a cause or an effect? The subsequent history suggested that it was an effect.

In a study of patients with hypoxaemia related to sleep apnoea, an increase in arterial pressure was noted with each apnoeic episode.⁶ Possible mechanisms were increased sympathetic tone caused by the hypoxaemia, or by respiratory acidosis or hypercapnia, or both, the last causing increased intracranial pressure leading in turn to the hypertension. Heart failure itself, not caused by systemic hypertension, is well known to induce peripheral vasoconstriction by increased sympathetic tone,⁷ and by increased renin production.⁸ With all these mechanisms of peripheral vasoconstriction in operation it is not surprising that some patients manifest systemic hypertension.

Whether or not peripheral vasoconstriction leads to systemic hypertension would depend on the corresponding flow; only if it were high would vasoconstriction raise the pressure substantially. Whether or not peripheral vasoconstriction causes actual hypertension, it would still promote left heart failure and in turn pulmonary oedema. In this way a self-perpetuating cycle could be established which is broken only when the original cause, hypoxia, and perhaps hypercapnia and respiratory acidosis, is removed.

With the complex clinical findings presented by

these three patients, which included systemic hypertension and left ventricular hypertrophy pattern on the ECG, it would be easy to miss the basic problem—upper airways obstruction. Therefore, upper airways obstruction should always be considered in the differential diagnosis of children presenting with heart failure, respiratory distress, and systemic arterial hypertension.

Case 2 was under the care of Dr A R Hastreiter.

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