Large Tonsils and Adenoids in Small Children with Cor Pulmonale

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Although small children with large inflamed nasopharyngeal lymphoid masses have been part of human ecology since the move east of Eden, the syndrome of cor pulmonale consequent to upper airway obstruction was not described until 1965 (Menashe, Farrehi, and Miller, 1965; Cox *et al.*, 1965). These and subsequent reports (Noonan, 1965; Luke *et al.*, 1966) defined the clinical syndrome of noisy, stertorous respirations, somnolence, clinical and electrocardiographic evidence of right ventricular hypertrophy, radiological findings of cardiomegaly, and occasionally pulmonary oedema and right heart failure. Like all clinical syndromes, there are numerous variations about the central unifying theme. Through recounting our experience with six of these patients, observed within a two-year period, we wish to illustrate some of these variations. Particular emphasis will be directed toward lability of the clinical, electrocardiographic, and radiological findings; and the ever present danger of death from either respiratory or cardiac arrest.

CLINICAL FEATURES

The pertinent clinical features exhibited by each of these six Negro male children are presented in Table I, and the lowest observed arterial blood oxygen saturations with the corresponding plasma carbon dioxide tension for each are presented in Table II.

The youngest patient was 17 months and the oldest 6 years of age. Signs of right heart failure (facial and peripheral oedema and abdominal protuberance) prompted hospital admission in four, and coma and seizure, respectively, in the other two. Each patient had a history of a recent antecedent respiratory infection

with onset one to three weeks before hospital admission. Each had developed noisy and laboured respiratory efforts.

All patients presented in respiratory distress with laboured and incredibly noisy respirations. Respiratory difficulty was aggravated by recumbency. In fact, three patients (Cases 2, 4, and 6) lost consciousness when recumbent. Their respirations became irregular and were interrupted by long apnoeic periods. Somnolence was a constant and striking feature. Labial and oral cyanosis was evident in some; an ashen-grey cyanosis reminiscent of the characteristic colour of infants with complete transposition of the great vessels was noted in one patient (Case 5), whose arterial oxygen saturation varied between 17 and 20 per cent. In spite of the vigour of the respiratory efforts, auscultation of the lung fields suggested poor air entry, an important diagnostic sign not emphasized in previous descriptions of this syndrome. Although each patient presented with signs of a concurrent respiratory infection (copious mucopurulent discharge, pharyngeal injection), descriptions of the oropharyngeal lymphoid ring would not suggest the degree of severity of upper airway obstruction present (Fig. 1).

Apart from 2 patients (Cases 1 and 5), the cardiovascular findings were consistently similar. Most of the patients had a bulging praecordium. All exhibited a forceful parasternal right ventricular thrust and palpable pulmonary valve closure. Several had four distinct heart sounds, often resembling a summation gallop. The second sound at the base was loud and single. Murmurs were insignificant or absent. Case 1, however, had an intermittent apical presystolic murmur and his course was so remarkable that it is described in detail below. Case 5 presented with a quadruple rhythm, a loud apical pansystolic murmur, and systolic pulsations of the liver. A diagnosis of Ebstein's malformation of the tricuspid valve was entertained until the murmur disappeared following spontaneous resolution of his respiratory infection. Four of the six patients had distended neck veins, facial oedema, hepatomegaly, and pitting oedema.

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TABLE I
CLINICAL FINDINGS

Case No.	Age (yr.)	History	Physical findings	Electrocardiology	Chest radiograph
1	2	Upper respiratory infection; noisy respirations; somnolence; facial and peripheral oedema and hepatomegaly; syncope	Noisy, laboured respiratory efforts; somnolence; cyanosis Praecordial bulge; rt. ventric. thrust; palpable pulm. closure; heart sounds; pulm. ejection murmur; intermittent apical presyst. murmur; rt. heart failure	Sinus tachycardia; rt. axis deviation, rt. atr. enlargement; rt. ventric. hypertrophy	Cardiomegaly; prominent rt. border; full pulm. art. segment; pulm. oedema, oligemia of peripheral lung fields
2	3	Upper respiratory infection; noisy respirations; somnolence; coma and irregular respirations in recumbency	Noisy, laboured respirations; poor air exchange; somnolence; coma and irregular respirations in recumbency Praecordial bulge; rt. ventric. thrust; 3 heart sounds; palpable pulm. closure; no murmurs	Sinus tachycardia; rt. axis deviation; rt. atr. enlargement; rt. ventric. hypertrophy	Cardiomegaly; prominence of rt. heart border; full pulm. art. segment; pulm. oedema; peripheral lung field oligemia
3	3	Upper respiratory infection; facial oedema and abdominal swelling; somnolence	Somnolence; difficult, laboured respiratory efforts; poor air exchange Praecordial bulge; rt. ventric. thrust; 4 heart sounds; palpable pulm. closure; no murmurs; facial and peripheral oedema; hepatomegaly	Sinus tachycardia; rt. axis deviation; rt. atr. enlargement; rt. ventric. hypertrophy	Cardiomegaly; prominence of rt. heart border; full pulm. art. segment; pulm. oedema; peripheral lung field oligemia
4	22 mth.	Upper respiratory infection; noisy difficult breathing; convulsive seizure	Somnolence; laboured, noisy respiratory efforts; apnoea in recumbency; poor air exchange Praecordial bulge; rt. ventric. thrust; palpable pulm. closure; 4 heart sounds; no murmurs	Rt. axis deviation; rt. atr. enlargement; rt. ventric. hypertrophy	Cardiomegaly; prominence of rt. border; full pulm. art. segment; pulm. oedema; peripheral oligemia
5	18 mth.	Iron-deficiency anaemia; upper respiratory infection	Laboured respiratory efforts with poor air exchange; somnolence; ashen-grey cyanosis Praecordial bulge; rt. ventric. heave; quadruple rhythm; pansyst. murmur of tricuspid insufficiency; syst. pulsation of liver edge; facial and peripheral oedema; hepatomegaly	Rt. axis deviation; combined atr. and ventric. enlargement	Cardiomegaly; prominence of rt. border; full pulm. art. segment
6	6	Mouth-breather for 2 yr.; upper respiratory infection; laboured, noisy respiration; generalized oedema	Laboured, noisy respirations; apnoea in recumbency; somnolence Forceful rt. ventric. lift; 3 heart sounds; palpable pulm. closure; no murmurs; hepatomegaly; peripheral oedema	Rt. axis deviation; rt. atr. enlargement; rt. ventric. hypertrophy	Cardiomegaly; prominence of rt. border; full pulm. art. segment; peripheral oligemia

Electrocardiograms demonstrated right axis deviation, right atrial enlargement, and right ventricular hypertrophy (Fig. 2). The electrocardiogram of Case 5 had additional electrocardiographic evidence suggesting left atrial and left ventricular overload, presumably the changes induced by a recent severe iron-deficiency anaemia.

Chest radiographs demonstrated cardiomegaly (Fig. 3), with prominence of the right heart border and a full pulmonary artery segment. Some patients had perihilar congestive changes compatible with the presence of pulmonary oedema. In general, the periphery of the lung fields was oligemic.

TABLE II
LOWEST OBSERVED PERIPHERAL ARTERIAL OXYGEN SATURATION AND HIGHEST OBSERVED CARBON DIOXIDE TENSION IN 6 CHILDREN WITH COR PULMONALE SECONDARY TO UPPER AIRWAY OBSTRUCTION

Case No.	Oxygen saturation (%)	Carbon dioxide tension (mm. Hg)
1	42.0	Not obtained
2	31.0	68
3	68.3	55
4	59.2	54
5	17.0	68
6	87.0	52

FIG. 1.—Nasopharyngogram demonstrating the severity of upper airway obstruction of Case 5.

Blood gas analyses revealed arterial oxygen unsaturation, often severe (Table II), and raised carbon dioxide tensions. Two patients (Cases 1 and 2) were studied by cardiac catheterization, which revealed arterial unsaturation and wide phasic swings in the intravascular

FIG. 3.—Initial chest x-ray film of Case 2 with moderate cardiomegaly, prominent pulmonary artery segment, and perihilar congestion (pulmonary oedema).

FIG. 2.—Initial electrocardiogram of Case 2, showing right axis deviation. P wave peaking and R/S ratio in V5 and V6 < 1.0.

and intracardiac pressure pulses that were synchronous with the laboured respiratory efforts. Mean pulmonary arterial pressure was raised.

Relief of obstruction by tonsillectomy and adenoidectomy was followed by a prompt return of arterial blood gases to normal; heart size often decreasing rapidly as is shown in Fig. 4 and 5. Although the technique is not comparable in the two films (the initial was an antero-posterior portable radiograph and the latter a standard 6-foot postero-anterior film), the change in heart size 18 hours after insertion of a nasotracheal tube is dramatic. Apart from P wave amplitude, which rapidly returned to normal, the other electrocardiographic abnormalities resolved more slowly; all returned to normal within six months.

CASE REPORT

Case 1, our introduction to the chronic upper airway obstruction syndrome, presented as a perplexing and dramatic diagnostic problem.

At the age of 2 years he was admitted to hospital with noisy breathing, facial and peripheral oedema, abdominal protuberance, and signs of a mild febrile upper respiratory infection. He had a praecordial bulge, a forceful right ventricular lift, a triple rhythm, a loud pulmonary closure sound, and an insignificant short pulmonary

FIG. 4.—Chest x-ray film of Case 6 before nasotracheal intubation (portable antero-posterior film).

FIG. 5.—Chest x-ray film of Case 6, 18 hours after nasotracheal intubation (standard 6-foot postero-anterior film).

ejection murmur. The electrocardiogram showed right atrial enlargement and right ventricular hypertrophy, and the chest radiograph demonstrated cardiomegaly with hilar congestion. Response to conventional anti-congestive therapy seemed slow. During a syncopal episode characterized by profuse sweating, very laboured respiratory efforts, and intense cyanosis, a presystolic apical rumble was heard and was recorded during subsequent episodes (Fig. 6). In the intervals between episodes of syncope, four heart sounds only were present

(Fig. 7). Cardiac catheterization revealed arterial unsaturation, wide phasic swings in the vascular and intracardiac pressure pulses, and a mean pulmonary arterial pressure of 53 mm. Hg. Cine-angiocardiology revealed intermittent filling of the left ventricle though no tumour mass was observed. Because the clinical picture followed the classical textbook description for left atrial myxoma, exploration on cardiopulmonary bypass was carried out and revealed, to the bewilderment of all, only an inversion of the atrial appendage

FIG. 6.—Phonocardiogram taken during presence of murmur in Case 1. PS, presystolic murmur; OS, opening snap; SM, systolic murmur; DM, diastolic murmur.

FIG. 7.—Case 1. Phonocardiogram showing the 4 distinct heart sounds.

into the left atrial cavity. Gentle traction applied to the tip of the appendage caused it to protrude through the mitral valve. The appendage was amputated and, judging by the patient's initial post-operative course, this seemed a curative procedure. During the second post-operative week he had a mild upper respiratory infection and recurrence of signs of right heart failure, which proved intractable to medical management; his subsequent course was one of inexorable progression to fatal termination. Permission for necropsy was not obtained. Shortly after the patient's death we became aware of the syndrome of cor pulmonale secondary to chronic upper airway obstruction, and the diagnosis was

made retrospectively. No rational explanation has been found to account for the inversion of the left atrial appendage into the left atrial cavity.

DISCUSSION

As a rule the clinical features of the chronic upper airway obstruction syndrome are sufficiently characteristic to lead to early bedside recognition and definitive and curative treatment. Certain exceptions, however, do occur. The unusual course of Case 1 suggested the diagnosis of left atrial myxoma, and the presenting cardiovascular manifestations in Case 5 suggested the diagnosis of Ebstein's malformation of the tricuspid valve. The correct diagnosis of this syndrome, therefore, may be obscured by a physical finding associated with congenital or acquired cardiac lesions. In such cases radiological examination of the nasopharynx following the nasal instillation of "lipiodol" proves a simple, quick, and readily available method for demonstration of the nasopharyngeal obstruction.

Previous reports of this syndrome have not emphasized the labile and changing physical findings and electrocardiographic and radiological manifestations. None has emphasized its potential lethal outcome. Signs of right heart failure usually appeared in close temporal association with an antecedent respiratory infection. Heart failure did not respond to conventional anticongestive medical measures but seemed to improve spontaneously as the severity of nasopharyngeal obstruction was relieved or after the establishment of an adequate airway, either through nasotracheal intubation or through surgical removal of the obstructing lymphoid tissue. Indeed, the frequent ventricular premature contractions observed in Case 5 and the bradycardia observed in Case 1 following incomplete digitalization suggested that these patients might be unduly sensitive to the toxic effects of digitalis glycosides. Because relief of the upper airway obstruction is mandatory and the signs of cardiac decompensation rapidly abate following establishment of an adequate airway, there is little need to employ such measures in the management of these patients.

When obstruction was severe, four heart sounds were frequently audible. With establishment of an adequate airway the fourth sound always disappeared, the third sound often became inaudible, the intensity of the pulmonary component of the second sound diminished, and the normal respiratory split of the latter sound became discernible.

Electrocardiographic P wave peaking, interpreted perhaps erroneously as right atrial enlargement, disappeared rapidly with relief of airway obstruction. This P wave lability was mentioned by Luke *et al.*

(1966), and P wave peaking without right atrial hypertrophy also occurs in cyanotic congenital heart disease (Sodi-Pallares *et al.*, 1958). In this syndrome it probably reflects hypoxia. The variation in P wave morphology can be correlated with variations in the level of arterial blood oxygen saturation. The other electrocardiographic abnormalities, right axis deviation and right ventricular hypertrophy, resolved over a period of several weeks following relief of nasopharyngeal obstruction.

The degree of cardiomegaly revealed by chest x-rays varied from time to time in the same patient. This variation in heart size also correlated with the severity of nasopharyngeal obstruction. When the obstruction was most severe, radiological evidence of pulmonary oedema was manifest. Heart size decreased rapidly following relief of obstruction, often within a few hours to a few days.

Blood gas analyses before relief of airway obstruction always revealed some degree of arterial oxygen unsaturation, sometimes of an alarming degree (Table II). On the other hand, hypercarbia appeared only after severe obstruction had been present for some time, a function of the greater diffusibility of carbon dioxide. Somnolence, a clinical manifestation of hypercarbia rather than of anoxia, suggests that the obstruction is very severe.

The two fatalities that occurred in this group of six patients emphasize the serious consequences of this syndrome. In addition to the fatalities, two patients experienced apnoeic episodes when recumbent. Therefore, there should be no delay in relieving the obstruction either surgically or through nasotracheal intubation if immediate surgical removal of the obstructing tissue is not feasible. Therapy with atmospheres of high oxygen content is not adequate because one of the deaths occurred while the patient was in such an atmosphere. There is ample evidence that in patients suffering from severe hypoxaemia the respiratory centre becomes insensitive to carbon dioxide and hypoxia becomes the driving stimulus. Relief of hypoxia without relief of the hypercapnoea abolishes all central stimulation with consequent respiratory arrest.

Some details concerning the pathogenesis of the cardiovascular manifestations of the chronic upper airway obstruction syndrome have not been completely elucidated. It is clear that alveolar hypoventilation with attendant hypoxaemia in certain susceptible subjects leads to the development of pulmonary arterial hypertension. Whether the increase in pulmonary vascular resistance results from precapillary arteriolar constriction or postcapillary venular constriction or both has not been resolved (Houston, 1960; Hultgren *et al.*, 1961; Fred *et al.*, 1962; Singh *et al.*, 1965). The occur-

rence of pulmonary oedema in this and related syndromes (high altitude sickness, the Pickwickian syndrome) favours a venular constrictive response. However, it has been postulated that if the vasoconstrictive response is confined to the precapillary arterioles, pulmonary oedema may be due to a transcapillary gradient, created by the wide phasic swings of intrapleural pressure coupled with alternating Valsalva and Muller manoeuvres.

The increase in total pulmonary vascular resistance imposes an acute high pressure load on the capacitance-adapted right ventricular chamber. If the load is imposed acutely, it is undoubtedly sufficient to cause dilatation and right heart failure. Most authors, however, when discussing this syndrome, have ignored the possible deleterious effects of the abnormal acid-base balance and hypoxia on ventricular performance in the pathogenesis of this form of right heart failure. Acidosis, *per se*, does not influence ventricular performance (Talner, Garner, and Downing, 1966), but the degree of depression of ventricular performance imposed by a given level of hypoxia is potentiated by acidosis (Downing, Talner, and Gardner, 1965). An increase in the left ventricular end-diastolic pressure has been described in some forms of hypoxic heart disease (Kuida *et al.*, 1963; Hecht, Kuida, and Tsagaris, 1962). Although the clinical picture in these children was dominated by signs of right heart failure, cine-angiographic studies in some suggested that there might also have been an element of left ventricular dysfunction. Not only does hypoxaemia depress myocardial function, but the hypercarbia also potentiates the pulmonary vasoconstrictive reaction to hypoxic stimulation (Rudolph and Yuan, 1966).

Although cor pulmonale secondary to chronic upper airway obstruction constitutes a syndrome only recognized within recent years, it is somewhat disconcerting to realize that it cannot be a "new disease". Recognition of a syndrome undoubtedly increases its apparent frequency but other factors seem to be involved in the apparent frequency with which this syndrome is encountered. One of these may be a more conservative attitude leading to the preservation of nasopharyngeal lymphoid tissue and replacing the former custom of its mass removal in childhood. All our patients were Negroes from a low income group occupying the core region of a large metropolitan centre and not having ready access to medical care. That this sociological factor is of importance is suggested by the symptoms that led to their admission to hospital—coma in one, convulsive seizure in one, and right heart failure in the remainder. Children from the more affluent section of our society would have had earlier medical attention.

SUMMARY

Cor pulmonale as a consequence of chronic upper airway obstruction by hypertrophied tonsils and adenoids seems relatively common among young children in the less affluent section of our society.

The syndrome is sufficiently characteristic to permit rapid clinical diagnosis. Its features are: (1) noisy, laboured respiratory efforts accompanied by poor pulmonary air exchange; (2) somnolence, particularly marked in recumbency; (3) cardiomegaly; (4) right ventricular hypertrophy; and (5) signs of right heart failure. Although characteristic, the severity of these signs varies directly with the degree of obstruction, at least in the early stages.

Prompt relief of obstruction, either by the establishment of an adequate airway or surgical removal of the tonsils and adenoids, is indicated to avoid potentially lethal consequences.

The cardiovascular findings appear to be completely reversible.

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