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Cor Pulmonale in Crouzon's Disease

Cor pulmonale resulting from upper airway obstruction secondary to hypertrophy of the tonsils and adenoids has now been well documented (Macartney, Panday, and Scott, 1969; Menashe, Farrehi, and Miller, 1965; Ainger, 1968; Levy *et al.*, 1967; Cox *et al.*, 1965). The features of this syndrome are (i) noisy stertorous respiration and stridor in the supine position, (ii) somnolence, (iii) pulmonary hypertension, (iv) right heart failure, (v) electrocardiographic changes of right atrial and right ventricular hypertrophy and strain, (vi) arterial hypoxia and hypercapnia, and (vii) radiographic appearances of cardiomegaly, dilatation of the pulmonary artery, and often pulmonary oedema.

Cox *et al.* (1965) also described an infant with laryngotracheomalacia who went on to develop cor pulmonale. We describe a further case of cor pulmonale secondary to chronic nasopharyngeal obstruction, this time in a patient with Crouzon's

disease. It is felt that the abnormal facial bony development in Crouzon's disease, with consequent narrow nasopharyngeal and oropharyngeal airway, has contributed to the nasopharyngeal obstruction in this patient.

Case Report

This 3-year-old child presented with a 3-day history of ankle oedema. She had been diagnosed as having Crouzon's disease at the age of 8 weeks when she was referred for paediatric opinion, because of unusual facies.

Bilateral craniectomy was performed at the age of 7 months for craniostenosis. At the age of 2 years she was referred to an ear, nose, and throat surgeon because of noisy breathing. The parents were informed that the child had narrow air passages and that no further treatment was possible.

Neither of the parents was clinically affected by Crouzon's disease, but a paternal uncle and the paternal grandmother were both affected.

On examination on admission she was in right heart failure with central cyanosis, peripheral pitting oedema, tachycardia, tachypnoea, hepatomegaly, and noisy respiration. Chest x-ray showed gross cardiomegaly with early pulmonary oedema. The electrocardiogram (Fig. 1) showed an axis of +150 with evidence of right atrial and right ventricular hypertrophy. Blood gases breathing oxygen, soon after admission, showed P_{aO_2} 70 mm Hg, P_{aCO_2} 98 mm Hg, and HCO_3 of 36.5 mEq/l. She was treated with digoxin, frusemide, and oxygen with good improvement. An oral airway was needed to maintain a good colour. Cardiac catheter studies were performed to exclude a cardiomyopathy or intracardiac shunt. These investigations, which unfortunately were limited because of a cardiac arrest, showed pulmonary hypertension with a pulmonary artery pressure of 75/50 mm Hg, but with considerable decrease in pressure on inspiration (Fig. 2). Arterial gases at rest, breathing room air, showed hypoxia and hypercapnia with P_{aO_2} 31 mm Hg and P_{aCO_2} of 66 mm Hg. After intubation and inflation with 100% oxygen, after cardiac arrest, the P_{aO_2} improved to 425 mm Hg and the P_{aCO_2} came down to 45 mm Hg. These findings supported the diagnosis of cor pulmonale.

The patient improved considerably on medical treatment, though she still had noisy respiration and respiratory obstruction when sleeping.

She had several episodes of upper respiratory tract infection, associated with croup between discharge from hospital in June and readmission in September for adenotonsillectomy. At operation she had moderately enlarged tonsils and small adenoids. The ear, nose, and throat surgeon and anaesthetist commented on the very small postnasal space.

Postoperatively she no longer had noisy respiration, and a femoral vein puncture a month later showed a normal PCO_2 .

Comment

Crouzon in 1912 was the first to describe the features of the syndrome that now bears his name

FIG. 1.—The electrocardiogram taken on admission in congestive heart failure showing right atrial and right ventricular hypertrophy.

FIG. 2.—The pulmonary artery pressure recording showing raised pulmonary artery pressure and marked inspiratory swing.

(Crouzon, 1912). These features were (i) scaphocephalic and trigonocephalic deformities of calvarium, (ii) facial dysostosis with hooked parrot nose, small upper maxilla, and prognathism, (iii) bilateral exophthalmos, and (iv) genetic transmission and familial incidence.

Since the original description, other features and complications have been added (Flippen, 1950; Lemarié, Paquelin, and Thomas 1962); these include progressive exophthalmos and loss of vision, and increasing intracranial pressure giving rise to mental retardation.

Narrowing of the upper airways in Crouzon's disease has been reported previously (Lemarié *et al.*, 1962). The case we describe has all the features of the syndrome of cor pulmonale secondary to chronic upper airway obstruction, and we feel that the bony abnormality of Crouzon's disease has played a major part in the development of this obstruction.

Summary

A case of Crouzon's disease is reported in which the congenital narrowing of the oral and nasal airways gave rise to carbon dioxide retention and hypoxia which resulted in the development of

pulmonary hypertension and severe cor pulmonale. Adenotonsillectomy resulted in improvement.

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