stimulation test (Lacey and Parkin, 1974a, b). This meant that all the very short children from a population of about 8000 10-year-old children were examined. No clear case of organic growth hormone deficiency was discovered, there being good evidence of severe deprivation as the cause of the growth failure in the 3 children whose growth hormone levels were less than 10 μIU/ml. There was one patient among the children born in 1960 who possibly was an example of partial growth hormone deficiency. As her growth failure was not marked, it is unlikely that her parents will wish her to be given a trial of treatment.

Although the number of children screened in this study was not large enough to enable the incidence of growth hormone deficiency to be confirmed, it does indicate that the suggested incidence is not a gross underestimate.

Conclusion

Growth hormone deficiency is a very uncommon cause of short stature. Evidence is presented that indicates that when patients with low serum growth hormone levels associated with maternal deprivation are excluded, the approximate incidence of growth hormone deficiency is only 1 in every 30,000 births, about half of the patients having idiopathic deficiency and half having deficiency secondary to intracranial disease. If this is so only about 30 new patients who should benefit from growth hormone treatment may be expected to be diagnosed each year in England and Wales.

Summary

Evidence from the Newcastle upon Tyne region suggests that the incidence of organic growth hormone deficiency is about 1 in 30,000 births.

References


J. M. Parkin
Children's Department, Royal Victoria Infirmary, Queen Victoria Road, Newcastle upon Tyne NE1 4LP.

Cor pulmonale in the Pierre Robin syndrome

Upper airway obstruction has only recently been recognized as a cause of cor pulmonale in childhood. Tonsillar and adenoidal hypertrophy are the most common predisposing factors (Menashe, Farrehi, and Miller, 1965; Luke et al., 1966; Ainger, 1968), but laryngotraechomalacia (Cox et al., 1965) and Crouzon's disease (Don and Siggers, 1971) have also been implicated. We describe a case of the Pierre Robin syndrome with severe upper airway obstruction who developed cor pulmonale at 5 weeks of age.

Case report

A male infant was born at home weighing 2.74 kg. He was transferred to hospital at 7 days with a history of feeding difficulty, stridor, and cyanotic episodes. On examination he had marked micrognathia and a large central palatal cleft. There were no other congenital abnormalities and in particular the heart sounds were normal; there were no cardiac murmurs and chest x-ray was normal.

He was nursed prone and fed by tube. He continued to have stridor and apnoeic spells, the frequency and severity of which were influenced by his head posture. At 5 weeks of age he had a sudden rise in weight accompanied by increasing respiratory difficulty and persistent cyanosis in air. On examination he had a precordial bulge and a forceful right ventricular para-sternal thrust. The pulse rate was 180/minute. There was a third heart sound and summation gallop. A long systolic murmur was audible along the left sternal edge, coarse crepitations were heard over both lung fields, and the liver was palpable 3 cm beneath the right costal margin. Chest x-ray showed an enlarged heart, extensive soft lung shadowing consistent with pulmonary oedema, with some consolidation in the right upper lobe (Fig. 1a). ECG showed P pulmonale and changes of moderate biventricular hypertrophy. Repeated capillary blood samples taken from a warmed heel showed a persistent severe hypercapnia with Pco2 80–130 mmHg.

In an attempt to determine the posture in which respiratory obstruction was least, some studies of respiratory mechanics were carried out on the sleeping baby. Transthoracic pressure swings were recorded with an air-filled rubber balloon in the lower oesophagus; airflow and tidal volume were recorded by a pneumotachograph mounted in a face mask. Tidal volume, airflow, and oesophageal pressure swings were recorded while the baby was held in different positions. Pulmonary resistance was calculated by the pressure difference divided by the flow rate between midvolume points (Cook et al., 1957). The results obtained (Fig. 2.) showed great variability in the airway resistance with only small changes in head posture. Resistance to airflow was least in the prone position with the face straight down (total pulmonary resistance 120 cmH2O/l per sec) and rose to infinity in the supine position. Oesophageal pressure swings varied from 10 cmH2O (prone) to 40 cmH2O (supine).

The baby was nursed prone with his neck partially flexed in the position which appeared to cause least airway resistance. His head was supported in a foam mattress in which a hole had been cut for his face. The
cardiac failure was treated with digoxin, diuretics, and controlled oxygen therapy. Clinical improvement was slow and the heart failure difficult to control. By 4 months his stridor had disappeared and he was able to tolerate bottle feeding; cardiac failure had resolved, the heart sounds were again normal and the murmur was no longer audible. A repeat chest X-ray (Fig. 1b) and ECG were normal. Shortly after discharge from hospital the child died at home. Necropsy established that extensive bronchopneumonia was the cause of death. Apart from the cleft palate and micrognathia, the upper airways were normal and the heart and great vessels were entirely normal.

Discussion

Hypercapnia is very rarely a feature of primary cardiac disease and the normal cardiovascular findings at necropsy excluded a primary cardiac cause for this infant's heart failure. We believe the baby had cor pulmonale secondary to upper airway obstruction, and the systolic murmur was attributable to tricuspid regurgitation due to right ventricular dilatation.

The pathogenesis of the cardiovascular changes associated with upper airway obstruction is incompletely understood. Severe airway obstruction causes alveolar hypoventilation with resulting arterial hypoxaemia. Pulmonary vascular hyperreactivity to hypoxia does occur in about 20% of normal persons at high altitude (Grover et al., 1963), and a similar hypersensitivity may occur in some children with airway obstruction. Hypercapnia is known to potentiate the pulmonary vasoconstrictive reaction to hypoxia (Rudolph and Yuan, 1966). The increase in pulmonary vascular resistance imposes an acute high pressure load on the right ventricle, the performance of which may be further depressed by acidosis. Pulmonary oedema has previously been reported in children with upper airway obstruction and the cause is not clear. The high oesophageal pressure swings reported in this case support the hypothesis that the oedema is produced by the transcapillary gradient resulting from high swings of intrathoracic pressure.

Patients with Pierre Robin syndrome with airway obstruction usually improve spontaneously from the...
Fig. 2.—A record of lung mechanics obtained from the sleeping baby to show changes which occurred in different body positions. The top trace is the tidal volume and the middle trace a record of airflow; both these measurements were obtained by a pneumotachograph mounted in a tight-fitting face mask. The bottom trace is of oesophageal pressure swings obtained from an airfilled balloon in the lower oesophagus. The units and the scale are the same in both (a) and (b), though the paper speed was faster in (b). (a) Complete obstruction to airflow occurred when baby's posture was altered from right lateral to supine position. In the latter posture, oesophageal pressure swings of up to 40 cm H₂O were recorded, though not shown in this short trace. (b) Oesophageal pressure swings (and calculated pulmonary resistance) were minimized in the prone position with baby's face straight down. Turning the head to the right increased the airway obstruction.

Summary

An infant with the Pierre Robin syndrome is described in whom cor pulmonale developed as a result of severe upper airway obstruction.
We would like to thank Dr. D. Hull for permission to report his patient, and Dr. A. K. Mant for making available the necropsy data.

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J. J. Cogswell* and D. M. Easton
*Correspondence to Dr. J. J. Cogswell, Department of Paediatrics, Guy's Hospital, London SE1 9RT.

The Hospital for Sick Children, Great Ormond Street, London WC1N 3JH.

Serum IgG levels in feto-fetal transfusion syndrome

Comments on a discrepancy in the colour of some newborn twins have been made since Biblical times (Genesis xxv, 25), but it is only recently that the phenomenon peculiar to monozygotic twins has actually been defined and become known as the feto-fetal or twin-twin transfusion syndrome. The subject was recently reviewed by Benirschke and Kim (1973).

Details of the haematological picture found in both donor and recipient twin are well known, and Kloosterman (1963) has also discussed some of the consequences of the transfer of plasma proteins between such twins. Little reference, however, has been made to serum immunoglobulin levels, the depletion of which might well result in an imbalance in the immunological status of the two infants. A case which illustrates this problem is presented.

Case report

A 33-year-old Caucasian woman was delivered of twin male infants, by caesarean section, at 38 weeks' gestation. Apart from a transient urinary tract infection in the second trimester, this third pregnancy had been uneventful. A diagnosis of twins was confirmed radiologically in the 28th week.

At birth there was a striking difference in the appearance of the two babies. The first twin (the recipient), weighing 2.7 kg (occipitofrontal circumference 35 cm), was pale, meconium stained, and showed signs of intrauterine growth retardation.

The appearance of the monochorionic diamniotic placenta was consistent with that found in cases of feto-fetal transfusion (Strong and Corney, 1967), and on microscopical examination the villi of the recipient portion were uniformly mature and deeply congested, whereas those of the donor appeared oedematosus, with a well-preserved trophoblast layer and the vessels contained a few normoblasts and white cell precursors. The results of investigations on cord blood and blood samples taken during the first 28 weeks of life are shown in the Table and Fig.

![Fig.](image)

**Fig.**—Serum IgG levels of the two twins from birth to 28 weeks. The hatched area is the normal range after Hobbs and Davis (1967) and Hobbs (1971).

- donor twin; • recipient twin.

Total proteins were determined by the density column (Lowry and Hunter, 1945). Cellulose acetate electrophoresis was carried out, the strips scanned using a Zeiss absorbance recorder with integrating recorder, and the albumin concentration calculated using the total protein figure obtained from the density column. Transferrin, α1 antitrypsin, and immunoglobulins G, A, and M were determined by radial immunodiffusion. Commercial