Pulmonary hypoplasia presenting as persistent tachypnoea in the first few months of life

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See editorial by Le Souef

Consider pulmonary hypoplasia in infants with unexplained persistent tachypnoea

Over the past two years four infants with persistent tachypnoea have been referred to this hospital. All were subsequently found to have pulmonary hypoplasia. Two (cases 1 and 2) were self referred from the local population. The third had been admitted under the care of a general paediatrician in this hospital (CMMS), and the fourth case was discovered by chance. All the infants had been born in this hospital, for which the delivery rate is about 3500 a year.

Case reports

CASE 1

This boy was born by normal delivery at 39 weeks' gestation after an uneventful pregnancy; he weighed 2960 g. He was seen on the postnatal ward at 12 hours of age for grunting and poor feeding. A chest x ray taken at the time was consistent with transient tachypnoea of the newborn. He was discharged at 5 days of age but remained tachypnoeic. At 3 weeks of age he was admitted with fever and coryzal symptoms, pronounced tachypnoea, and poor feeding. Examination was unremarkable apart from a respiratory rate of 70/minute with mild intercostal recession.

The coryzal illness resolved but he continued to be tachypnoeic. An electrocardiogram and an echocardiogram did not disclose any abnormalities. He was referred for lung function testing at the age of 4 months because of parental anxiety. His functional residual capacity measured by whole body plethysmography was 16.9 ml/kg indicating pulmonary hypoplasia. In our laboratory the normal mean is 29.8 ml/kg (range 17.6-42 ml/kg). Over the first year his respiratory rate gradually settled to 30-35/minute at rest. A repeat measurement at 12 months of age was 20 ml/kg, showing "catch up" lung growth.

CASE 2

Born at term with a birthweight of 3370 g after an uneventful pregnancy, this baby remained in hospital for seven days because of initial poor feeding. He subsequently fed well and gained weight satisfactorily. Despite repeated reassurance from health professionals his parents remained concerned because of persistent tachypnoea. At 4 months of age examination was normal apart from a respiratory rate of 60-80/minute and mild chest wall recession. Chest x ray picture (reported as normal) showed small lung fields. Functional residual capacity measured with the inert gas dilution method using a rebreathing bag was 13.1 ml/g, indicating pulmonary hypoplasia. Normal mean in our laboratory is 24.7 ml/kg (range 14.9-34.5 ml/kg). Follow up at 14 months of age showed that his respiratory rate was still 66/minute but the functional residual capacity had risen to 17.8 ml/kg.

CASE 3

This child was born at 35 weeks' gestation by caesarean section for premature rupture of the membranes and fetal tachycardia. He weighed 2570 g at birth. His mother had generalised tonic-clonic seizures, which were successfully controlled with

carbamazepine. He started grunting soon after birth and needed a maximum of 35-40% inspired oxygen administered through a head box. The chest x ray picture was compatible with mild surfactant deficiency.

His respiratory illness resolved and he was discharged at 6 days of age. He was readmitted at 9 days with persistent tachypnoea and recession. An infection screen was negative. Chest x ray picture, an electrocardiogram, and echocardiography were all normal. His respiratory rate did not settle, and lung size measured using the inert gas dilution method at 3 months of age indicated a reduced functional residual capacity of 14.4 ml/kg, consistent with pulmonary hypoplasia.

CASE 4

This girl was born at 38 weeks by planned caesarean section for breech presentation; she weighed 4200 g at birth. The pregnancy had been complicated by gestational diabetes and polyhydramnios. The infant required intravenous dextrose for hypoglycaemia and was moderately tachypnoeic, with a respiratory rate of 50/minute. By chance she was recruited to a research study involving the measurement of functional residual capacity by inert gas dilution, which was found to be 11 ml/kg, indicating pulmonary hypoplasia. She remained asymptomatic apart from moderate tachypnoea and was discharged home at 5 days of age. Outpatient follow up showed that her weight had remained relatively static for the first three months of life despite appropriate feeding. Repeat testing at 8 months showed an increase in the functional residual capacity to 19 ml/kg.

Discussion

All of these infants presented with persistent tachypnoea which lasted several months and they all had clinically important pulmonary hypoplasia on lung function testing. The normal ranges given are for our own laboratory and agree closely with those of other published normal ranges.¹ The association between pulmonary hypoplasia and oligohydramnios is well known. The mother in case 4 had polyhydramnios. As far as we are aware, this has only been reported once before as an associated finding² except in conjunction with Pena-Shokeir syndrome³ (where fetal swallowing is impaired) and pleural effusions.⁴

Pulmonary hypoplasia is normally divided into two main types—primary and secondary—and is traditionally associated with severe lung disease at birth and a high mortality. Secondary pulmonary hypoplasia is the more common form and is usually associated with causes of oligohydramnios or congenital malformations such as diaphragmatic hernia.⁵ Primary pulmonary hypoplasia, however, is thought to be very rare and published reports are few.^{6,7} There is a large discrepancy, however, with the incidence reported in one post mortem series: pulmonary hypoplasia was the commonest single abnormality present in 15-20% of early neonatal deaths.⁽⁸⁾ The true incidence of milder degrees of pulmonary hypoplasia is probably obscured

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by the prevalence of respiratory disease at birth from other causes, particularly in those born prematurely.

The diagnosis of pulmonary hypoplasia should be based on the measurement of lung volume and the exclusion of other pathology which can cause tachypnoea, such as congenital heart disease, obstructive lung disease, interstitial lung disease, and infection. Several different techniques are available for the measurement of lung volumes in infants⁹ and measurement should ideally be undertaken in laboratories used to performing these tests. In case 1 functional residual capacity was assessed by whole body plethysmography. This measures all the air present in the chest including that contained in non-ventilated alveoli (and also a small amount of intra-abdominal air). Values are therefore higher than those obtained from measurement of functional residual capacity by inert gas dilution, where only the volume of the lung taking part in gas exchange is measured. Measurement of functional residual capacity by gas dilution can therefore underestimate lung volume in the presence of airways obstruction. None of the babies described here had wheezing on auscultation.

All of the children had chest x ray pictures taken which were reported by radiologists as normal, except the first x ray in case 1, described as being consistent with transient tachypnoea of the newborn. It is important to remember, however, that the possibility of pulmonary hypoplasia had not been raised by the clinicians when these examinations were performed and that chest radiography is a relatively insensitive method of diagnosing pulmonary hypoplasia.

Of interest is that two of these infants (cases 1 and 2) were the children of doctors. Perhaps parental concerns about persistent tachypnoea were taken more seriously because of this?

Little is known about the prognosis of babies with pulmonary hypoplasia: long term follow up studies have been performed on only a few babies with diaphragmatic hernia, in whom important lung

function abnormalities are still evident many years later.^{10,11}

Persistent respiratory morbidity has also been described in infants born after prolonged premature rupture of the membranes during pregnancy.¹² The infants reported here were not severely ill in the neonatal period and showed good "catch up" lung growth in relation to body weight during the first year of life to around the lower end of the normal range of values. It may be important to identify children with this condition, which might be more common than previously thought, as it is not yet known whether they are at increased risk of respiratory problems later in life.

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Management of HIV infected health care workers: lessons from three cases

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Three cases in which doctors in Glasgow were diagnosed as having HIV infection were all handled differently in relation to telling patients and the media. In the first patients were not told because the doctor had been doing administrative work and there was thought to be no risk to patients; although the media did report the case, it accepted the assurances given. In the second case, where a doctor had done many jobs in different specialities and places, the media identified the doctor before most patients had been informed: most calls to the helpline subsequently set up by the health authority were from patients who had not been treated by this doctor. This episode, however, allowed the incident team to be prepared for the next case, enabling the helpline to be established swiftly. In this case the doctor voluntarily identified himself, and this served to allay public fears and reduce the number of inappropriate calls to the helpline.

The risk of transmission of HIV from an infected health care worker to a patient lies between 1 in 4000 and 1 in 40 000.¹ HIV antibody tests have been

performed on more than 22 000 patients treated by infected health care workers.^{2,3} Although more than 100 patients tested positive, in only one incident was the health care worker, a dentist, implicated as the possible source of infection.²³ The United Kingdom guidelines on the management of HIV infected health care workers encompass three main principles: a duty to protect patients, a duty of confidentiality towards infected health care workers, and the concept that the risk of HIV transmission is restricted to certain "exposure prone" procedures from which infected staff should refrain (see box).⁴⁵ There have been three recent incidents in Scotland involving HIV infected doctors, all in the Glasgow area. The nature and management of these varied. This paper describes our experience and the lessons learnt.

Case 1

In January 1990 a consultant in accident and emergency medicine was admitted with *Pneumocystis carinii* pneumonia. HIV was diagnosed, and he died two weeks later. He had had hepatitis B one year

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