Dry lung syndrome after oligohydramnios

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SUMMARY We studied four infants whose mothers had had leaks of amniotic fluid for varying periods before birth. Three were associated with compression deformities. Oxygenation was achieved only by overriding the pressure limit safety devices on the resuscitation apparatus and ventilating after birth with high inflation pressures. In all cases the ventilation pressures had to be further increased for 12 hours but rapid improvement followed on the second day so that all four were spontaneously breathing air after a few days. It is postulated that collapse of the airways resulted from oligohydramnios, and that the resulting 'dry lungs' would only respond to high inflation pressures.

It seems that adequate quantities of amniotic fluid are required for the satisfactory development of the fetal lung.^{1 2} Oligohydramnios associated with renal agenesis or gross dysplasia is well recognised as a cause of pulmonary hypoplasia that is so severe that the infants die very shortly after birth.^{3 4} More recently the association between pulmonary hypoplasia and prolonged leakage of liquor following both spontaneous membrane rupture and amniocentesis has been described, again usually with fatal outcome.^{5 6}

We report the association between severe early respiratory distress in four babies after prolonged leak of amniotic fluid in whom the respiratory distress improved remarkably on the second or third postnatal day. We postulate that this was due to secondary reduction in the lung fluid and effective lung volume (dry lungs) due to the external compression and the leak of fluid. Physical distension of the lung by higher pressures than normal may allow survival.

Patients

Of 115 premature babies of less than 37 weeks' gestation with premature and prolonged rupture of membranes (longer than 99 hours) who were managed in the neonatal unit at this hospital between January 1980 and January 1987, 11 died shortly after birth from respiratory insufficiency associated with hypoplastic lungs, six confirmed at necropsy. In many cases extremely high inflation pressures had been required to achieve oxygenation at the resuscitation, and further increases in ventilator pressures had been necessary during life before the almost

invariable development of air leaks. In many cases hypoxia occurred gradually when it was impossible to provide higher airway pressures on the ventilators (Bournes BP200 or Sechrist). All the babies who died had skeletal deformities consistent with oligohydramnios tetrad.²

In four infants a similar early course was seen, but on the second or third days of life, after we had used extremely high peak inflation pressures, a sudden and dramatic improvement occurred and the infants rapidly ceased to require ventilatory assistance or supplementary oxygen. These four cases are described below. During the period of this study no infants were seen with a similar clinical story in whom the membranes had been ruptured for less than 99 hours.

CASE 1

Mrs D had premature rupture of fetal membranes at 31 weeks' gestation. She was treated with ritodrine, betamethasone, and amoxycillin and continued to leak large volumes of liquor until delivery eight days later. The baby's heart rate at delivery was 50, but despite intubation and intermittent positive pressure ventilation that led to a prompt increase in the heart rate, the infant remained blue. There was poor air entry (worse on the left) and although the infant began spontaneous respiration he remained blue. There was a fixed flexion deformity at the left elbow and deformed feet. In view of the persistent cyanosis and the poor chest movements a diagnosis of diaphragmatic hernia was considered, but the chest radiograph showed only airless lungs compatible with congenital pneumonia. After two hours in 100% oxygen the baby became pink but carbon

dioxide retention continued for 12 hours. He was then extubated, and did not require supplementary oxygen after 21 hours. No pathogens were grown from the sputum and he made an uncomplicated recovery.

CASE 2

Mrs K had an amniocentesis at 16 weeks because a previous infant had had a neural tube disorder and she had a high plasma concentration of α fetoprotein. She had a continuous leak of amniotic fluid following this procedure, and oligohydramnios was diagnosed on ultrasonography. A caesarian section was performed electively at 33 weeks' gestation. At birth the baby's heart rate was unrecordable and there was no respiratory effort. The heart rate improved after intubation and intermittent positive pressure ventilation, but the baby remained blue with poor air entry. The baby was reintubated three times because of apnoea and bradycardia, and only became pink when the pressure release button on the resuscitation apparatus was overridden. Continuous inflation with a bag compressed by hand at high pressure finally achieved oxygenation. On transfer to the special care baby unit the chest radiograph showed small, airless, 'hypoplastic lungs'. There were Potter's facies and many limb deformities including bilateral genu recurvatum. The baby was stabilised on extremely high mean airway pressure of 22 cm H₂O and 100% oxygen but over the next 24 hours it was possible to reduce these to 12 cm H₂O and 30% oxygen. At 48 hours the baby was transferred to continuous positive airway pressure and at 60 hours she was extubated and breathing air. No pathogens were grown from the sputum, the respiratory rate was normal by 10 days of age, and she made an uncomplicated recovery.

CASE 3

A baby girl was born at 27 weeks' gestation by caesarean section for breech preterm labour 23 days after the spontaneous rupture of fetal membranes. Oligohydramnios was confirmed by ultrasonography. There was no spontaneous respiratory effort at birth and the Apgar score was 3 at one minute, so she was intubated and given intermittent positive pressure ventilation. Because of bradycardia, cyanosis, and poor chest movements with the pressures normally used for resuscitation, the pressure release valve on the resuscitation bag was overridden and the infant was transferred to the neonatal unit where high inflation pressures were used. There were pronounced contraction deformities of all four limbs. During the next five hours

because of persistent cyanosis despite being given 80–100% oxygen, the peak airway pressures were gradually increased to a maximum of 32 with an end pressure of 6 cm H₂O (a mean airway pressure of 19). The chest radiograph showed airless lungs compatible with surfactant deficiency or severe bilateral congenital pneumonia. Using a variety of conventional ventilator rates (20 to 60) she had persistent dusky spells on 100% oxygen and inflation with a bag compressed by hand was required. Precarious oxygenation was eventually achieved using a peak airway pressure of 32, an end pressure of 4, a 1:1 inspiratory ratio, and a ventilator rate of 140/minute. The mean airway pressure was 18 or more because of probable inadvertent end pressure at this ventilator frequency. Tolazoline was given without notable improvement in oxygenation. Frequent inflation with a bag compressed by hand at high pressure was required during the first 12 hours. After 16 hours the infant was stabilised on a peak airway pressure of 26, end pressure of 7, ratio of 1:1, a ventilator rate of 35/minute, and a mean airway pressure of 16.5. By 24 hours of age the mean airway pressure had been reduced to 10 and the oxygen to 75%. There was then steady and rapid improvement until she was extubated on the fourth day of life, and she was spontaneously breathing air on the fifth day. No pathogens were cultured from the secretions at any stage in the illness. Over the five weeks until discharge from hospital she had a respiratory rate which was on occasions over 60/minute at rest, but at follow up at 4 months old there were no chest problems or signs.

CASE 4

Mrs L had several small antepartum haemorrhages after 21 weeks' gestation. At 27 weeks a further haemorrhage was followed by a pink watery loss. Ultrasound examination after this showed 'no liquor' and examination using a speculum confirmed the loss of blood stained amniotic fluid. She continued to leak fluid freely over the next 10 days when a further ultrasound examination showed a liquor column of only 1 cm. Amniocentesis showed a lecithin:sphingomyelin ratio of 2:2 in the liquor, so at 28 weeks and six days a caesarean section was performed. Intermittent positive pressure ventilation and 100% oxygen were required to resuscitate the baby girl. No contractual deformities were apparent. By 8 hours of age a mean airway pressure of 14 cm H₂O and 90% oxygen were required to achieve oxygenation. The chest radiograph at six hours showed a 'white out' compatible with severe surfactant deficiency or bilateral congenital pneumonia. Over the next six hours, however, it was possible to reduce the oxygen to 35%. A repeat

chest radiograph taken at 18 hours of age showed complete aeration and no obvious disease. Forty eight hours later ventilatory support was withdrawn and on the fifth day she was breathing spontaneously on 25% oxygen. She had a pronounced tachypnoea and needed 25% to 35% oxygen in the incubator for the two months, repeated chest radiographs showing slight haziness bilaterally but no definite evidence of consolidation or infection. Since discharge she has been admitted several times with wheeziness and even when her chest sounds clear there is some recession.

Discussion

The diagnosis of anatomical pulmonary hypoplasia during life depends on the clinical finding of oligohydramnios, a low Apgar score, the presence of deformities, and an unusual postnatal pulmonary course.⁶ This resembles severe respiratory distress syndrome with persistent pulmonary hypertension, but differs in that there is immediate onset of severe pulmonary insufficiency requiring high ventilator pressures.⁶ Chest radiographs are variable, in some infants showing airless lungs compatible with the respiratory distress syndrome, in some small clear lung fields and in some compression deformity of the thorax so that it is bell shaped.⁷

In three of the infants reported here the association between positional deformities of the limbs and prolonged leakage of liquor amnii made it reasonable to assume that their respiratory problem was pulmonary hypoplasia. Despite the absence of postural deformity, the fourth patient had severe oligohydramnios shown on ultrasonography during the 13 days before delivery. Hypoplasia of the lungs seemed to be confirmed in the delivery room in each case by the need to override the pressure control on the resuscitation apparatus to achieve oxygenation. Once the infants had reached the neonatal unit and the compression deformities and Potter's facies were evident we decided to use high mean airway pressures to ventilate them. In all cases antibiotics were given in case they had congenital pneumonia, but no bacteriological confirmation was ever obtained. The fourth case, despite the absence of positional deformities, seems to have had pulmonary hypoplasia similar to that described by Perlman *et al.*⁸

In all cases the ventilation pressures had to be increased further in the first 12 hours of life to achieve adequate oxygenation. It was difficult then to know whether additional surfactant deficiency was compounding the problems, particularly as the chest radiographs suggested little aeration. The high lecithin:sphingomyelin ratio in the fourth

infant, and the improvement in all the infants on the second day at a time when surfactant deficiency would be worsening, made this diagnosis unlikely. All the infants had low Apgar scores but we believe that this was largely because of our inability to expand the lungs at the resuscitation rather than because of pre-existing asphyxia.

It seems possible that a temporary 'functional hypoplasia' was the cause of this clinical condition where the lungs had leaked dry of the normal volumes of lung fluid and collapsed, necessitating high pressures to reopen them. This partial pulmonary hypoplasia has been alluded to before. Perlman *et al* described a case in which an infant had facial and limb deformities after a leak of liquor for 16 days together with a pulmonary condition which, though it did not require ventilation, was felt to be consistent with anatomically and functionally small lungs at birth with 'compensatory hyperplasia later'.⁸ In a group of 21 infants reported by Thibeault *et al* who required ventilation and who had positional deformities following prolonged leakage of liquor three survived; these may also have been instances of 'dry lung syndrome' but the details of the care and course are not given.⁶

Alcorn *et al*⁹ showed that although chronic drainage of liquor for three to four weeks in fetal sheep reduced the amount of lung tissue and its anatomical development, the biochemical maturation was enhanced leading to increased numbers of type 2 cells with lamellar material present both within the cells and within the alveolar lumen. Richardson *et al* showed that in the human fetus prolonged rupture of the fetal membranes led to acceleration of maturation of the fetal lung over a few days.¹⁰ Wigglesworth and Desai showed that the hypoplastic lungs associated with oligohydramnios had narrow airways and there was a failure of elastic tissue development round the airways and terminal sacs.¹¹ It is conceivable therefore that physical distention of the airways with high pressures over the first days of life might encourage comparatively mature biochemical mechanisms to promote satisfactory oxygenation. This seems a more reasonable explanation of the time course than the rapid hyperplasia of the lungs that was postulated by Perlman.⁸

Recently Blott *et al* described how pregnancies with oligohydramnios associated with rupture of the membranes during the second trimester have a satisfactory outcome if fetal breathing movements continue.¹² The presence or absence of fetal breathing movements is not recorded in our cases, but the duration of rupture was shorter, being 8,13[?+42], and 23 days in the first three cases though not in the fourth (119 days). It seems

unlikely that the moment that fetal breathing stops the infant develops anatomical hypoplasia, but it might be that with the cessation of breathing movements the lungs collapse leading to temporary dry lungs due to unrestrained compression.

With modern obstetric policies more mothers with early premature rupture of the membranes are being managed conservatively. It would seem appropriate not only to monitor the volume of liquor and the strength of fetal breathing movements obstetrically, but to deliver these infants electively with paediatricians in attendance who are of sufficient seniority to assess immediately whether the normally accepted inflation pressures may need to be overridden.

I thank junior staff able to intelligently and appropriately overrule policy. These infants would not have survived without them.

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Received 1 September 1987