significantly lower than that during the six days after (48.3 ± 10.03, n = 6; P < 0.001) (Figure). He did not require a higher ambient O₂ concentration and the chest x-ray was normal after theophylline was ceased. Plasma theophylline concentration on the 8th day was 17 μg/dl.

Discussion

IMV (Kirby et al., 1972)—augmentation of spontaneous ventilatory effort with a ventilator frequency sufficient to maintain normocarbia and prevention of CO₂ rebreathing by continuous gas flow—allowed recognition of dependence on low ventilator rates without which the two infants in this report developed apnoea or hypopnoea and bradycardia or hypercarbia. The minimal IMV and supplemental O₂ requirements to maintain normal arterial blood gases, and the normal or near normal chest x-rays suggested that the ventilator dependence of the infants was owing to inadequate ventilatory effort and not to pulmonary parenchymal disease. The relative ease with which they were weaned to CPAP and then extubated after treatment with theophylline supports this suggestion, since the drug is of proved value in the spontaneously breathing preterm neonate with apnoea (Shannon et al., 1975).

The mechanism whereby xanthines exert their beneficial effect in neonatal apnoea is unknown, though they have been reported not to increase alveolar ventilation, since Paco₂ was unaffected (Shannon et al., 1975; Bednarek and Roloff, 1976). However, the fact that in both infants Paco₂ was lower while they were receiving theophylline than after, and that in one infant Paco₂ was higher before theophylline was given than after, suggests an increase in alveolar ventilation during treatment with theophylline. The development of chronic pulmonary insufficiency of prematurity (Krauss et al., 1975) after theophylline was omitted in one infant also suggests that theophylline may have an effect on alveolar ventilation, since the condition is characterised by low lung volume and ventilation/perfusion unevenness (Auld, 1975).

When very low birthweight infants recovering from HMD are dependent on slow-rate IMV without which they develop apnoea or hypopnoea and/or hypercarbia, treatment with theophylline may facilitate weaning from mechanical ventilation.

Summary

Two very low birthweight infants with severe clinical hyaline membrane disease requiring mechanical ventilation were dependent on slow-rate intermittent mandatory ventilation, without which they developed apnoea or hypopnoea and hypercarbia. Their ventilator dependence was apparently owing to inadequate ventilatory effort, and treatment with oral theophylline allowed easy weaning to continuous-positive airway pressure and extubation. Paco₂ was significantly lower during theophylline treatment, suggesting that the drug may have improved alveolar ventilation.

References


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Right-sided congenital diaphragmatic herniae presenting as pleural effusions in the newborn: dangers and pitfalls

Early thoracentesis is recommended both for diagnosis and therapy in pleural effusions of the newborn. These case reports show the need for caution in placing a thoracostomy tube and describe the investigational pitfalls associated with this unusual presentation of right-sided congenital diaphragmatic hernia.
Case reports

Case 1. After a pregnancy complicated by maternal systemic lupus erythematosus, this 2778 g white baby boy was delivered at 38 weeks' gestation. Labour was spontaneous with the membranes ruptured artificially immediately before a normal vaginal delivery. Apgar score was 8 at 1 minute and 8 at 5 minutes. At age 3 hours moderate respiratory distress in 30% O₂ was noted with grunting, nasal flaring, and cyanosis. Chest x-ray was normal.

Cultures (which subsequently grew group B β-haemolytic streptococcus) were obtained and treatment with ampicillin and gentamicin was started. Hydrocortisone was also given because of maternal prednisone therapy.

When transferred to this centre at 30 hours of age, the baby was in moderate respiratory distress and the abdomen was markedly distended. A nasogastric tube was in place and he had received no feeds. A repeat chest x-ray showed a large right-sided pleural effusion with the mediastinum slightly shifted to the left and otherwise normal lung fields. The baby's condition deteriorated and a diagnostic and therapeutic thoracentesis was performed through the 4th intercostal space in the posterior axillary line. A size 12 Fr plastic (Argyle) cannula introduced with an internal trocar initially obtained 2 ml sterile sero-sanguinous fluid. The cannula was then advanced and immediately produced 35 ml grossly bloody fluid. During the next six hours 100 ml whole blood was required by transfusion to correct hypotension, the systolic BP at one time dropping from 60 to 34 mmHg. The chest tube continued to drain about 10 ml of this fluid daily. The distended abdomen was dull to percussion. A paracentesis performed at the time of the thoracentesis produced 7 ml sterile sero-sanguinous fluid.

The deterioration in his respiratory state continued and when an F₁O₂ of 0·8 was needed for satisfactory oxygenation, ventilator therapy was initiated. He subsequently developed severe disseminated intravascular coagulation, renal failure, congestive heart failure, and sclerema, all probably the result of the β-haemolytic streptococcal septicaemia, and died on the 10th day of life.

 Necropsy revealed (a) a right congenital diaphragmatic hernia of Bochdalek with the apex of the right hepatic lobe in the right hemithorax—the chest tube was inserted into the superior aspect of this lobe; (b) bilateral pulmonary hypoplasia; (c) multiple abscesses, both bacterial and mycotic; and (d) extensive hypoxic changes in the brain, kidneys, and intestine.

The next case had many similarities but, happily, not such a tragic end.

Case 2. This 2400 g white baby boy was born at 35 weeks' gestation after a normal pregnancy. He developed mild respiratory distress in the first hour of life requiring 35% O₂ to achieve satisfactory oxygenation and a chest x-ray at that time revealed mild pulmonary infiltrative changes bilaterally with normal positioning of the diaphragm. Antibiotics were started after blood cultures were obtained; these cultures later grew group B β-haemolytic streptococcus. He was transferred to the Newborn Intensive Care Unit at the University of Colorado Medical Center where his respiratory distress gradually worsened. With the F₁O₂ at 0·8 at 30 hours of age, ventilator therapy was instituted. Chest x-ray at this time showed a large right-sided pleural effusion with bilateral hilar reticular densities without mediastinal displacement. A thoracentesis was performed through the midaxillary line at the 4th intercostal space with a 23 gauge scalp vein needle. The tap produced grossly bloody fluid and a 12 Fr plastic (Argyle) cannula (without trocar) was then inserted at the same site. A small skin incision was made and a channel into the chest made by means of a curved haemostat. The chest tube, held in the clamp, was then inserted and secured. 22 ml of the same fluid was immediately drained. Despite disseminated intravascular coagulation, hyperglycaemia, hyperbilirubinaemia requiring exchange transfusion, and sclerema, the baby slowly improved after day 4. The chest tube continued to drain grossly bloody fluid until day 8 of life when clearing was noted—the subsequent fluid contained no lipid, and had a protein of 3·1 mg/100 ml (31 g/l) and the quantity increased from 10 ml to 100 ml daily. The diagnosis of congenital diaphragmatic hernia was made and a liver scan performed which revealed the right lobe of the liver in the right hemithorax.

At the age of 13 days a large right sided diaphragmatic hernia of Bochdalek was repaired—70% of the liver was found to be in the chest cavity and the liver was noted to be congested and 'weeping' from its superior surface. The lungs were normal in appearance. After surgery the baby did well and was discharged on day 22.

Discussion

The problem of right-sided congenital diaphragmatic herniae has been reviewed by Blank and Campbell (1976) but our two cases illustrate aspects of the problem which need to be emphasised. Pleural effusion is an unusual although well described
accompaniment of right-sided hernia (Canino et al., 1964; Kirchner et al., 1975; Blank and Campbell, 1976). As the liver herniates through the defect, obstruction to hepatic venous outflow may cause vascular congestion with transudative weeping of its surface. In extreme cases strangulation of the liver may occur (Ravitch, 1956).

Even with knowledge of this association, several factors may prevent correct radiological diagnosis. (a) Pulmonary inflammatory disease may be simulated by an effusion associated with concomitant atelectasis of adjacent lung tissue. Also, as herniation may be intermittent, this misdiagnosis may be perpetuated by improvement on the x-ray as the liver descends. (b) An effusion may obscure the shadow of either an abnormal diaphragm or an abnormally high liver. (c) The position of the liver is often initially normal (Canino et al., 1964) with herniation occurring gradually. (d) The liver may act as a bulwark preventing intestinal herniation, thereby producing a normal abdominal gas pattern on x-ray.

Once considered, the right-sided hernia can still be very difficult to demonstrate preoperatively. Liver scans are helpful but only in locating the liver. They do not locate the diaphragm and will not differentiate between evagination and herniation. A more definitive test is to demonstrate communication between the thoracic and abdominal cavities. This may be done by insufflating a small amount of air (or CO₂) via paracentesis and demonstrating a pneumothorax on x-ray (Baker et al., 1959; Kenny et al., 1977). Nevertheless, many cases continue to be diagnosed at operation or necropsy.

In recent reports, thoracoscopy has been advocated for the diagnosis of intrathoracic lesions. This technique requires the transthoracic insertion of a standard peritoneoscope allowing direct observation (Rodgers and Talbert, 1976). This method may prevent needless thoracotomies when such a diagnosis as congenital diaphragmatic hernia is in doubt.

Because of the difficulties in making a firm diagnosis of right-sided hernia, associated pleural effusions are likely to be investigated on the same lines as those of other aetiologies. In these effusions, thoracentesis is of paramount importance. Clearly, during this procedure, caution must be exercised to avoid laceration of the lung. Because of our experience with these cases we have changed our approach to the aspiration of such effusions, especially on the right side where transfixing the liver becomes a possibility. We now initially use the Intracath* device for diagnostic and therapeutic thoracenteses. As the needle is advanced through the intercostal space, steady pressure is applied to the catheter within the needle. When the needle enters the pleural space, there is an immediate, rapid advancement by the catheter. The blunt tipped catheter will not only displace pulmonary tissue but will also prevent laceration and puncture of any aberrant intrathoracic organs by the needle tip. The catheter has the added advantage of manoeuvrability as it can be safely moved and redirected. Furthermore, upon withdrawal of the catheter, the incidence of pneumothoraces should be nil. This technique has been used at other centres by one of the authors (J.H.T.C.) in over 75 thoracenteses and paracenteses without complication.

Also of note in these two cases is the coexistence of β-haemolytic streptococcal infection. In review of the other reported cases this organism was present in the blood culture of two other infants. In our cases and that of Kenny et al. (1977) the organism was identified as group B, in that of Kirchner et al. (1975) as 'non-group A or D'. While neonatal colonisation is common (approximately 26% of normal newborns) infection is not (approximately 1 in 100 of those colonised) (Baker and Barnett, 1973). The portal of entry is probably the respiratory tract and one can speculate that the decreased ventilation of the right side leads to an increased susceptibility to this infection. As there is no evidence to suggest genetic predisposition to right-sided CDH, the possibility of a related genetic predisposition to streptococcal infection, once respiratory colonisation has taken place, is remote.

Summary

Congenital diaphragmatic hernia presented with right-sided pleural effusion in two newborn infants.

H.W.C. was supported in part by a grant from the American Lung Association of Colorado.

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