Respiratory function after repair of congenital diaphragmatic hernia

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SUMMARY Respiratory function studies were carried out in 22 infants who had successful repair of diaphragmatic herniae of the Bochdalek type. Thoracic gas volume was initially reduced in only 3 of these, but subsequent studies showed that improvement occurred. There were no consistent abnormalities in either dynamic compliance or mean pulmonary conductance. This is evidence that there is rapid adaptation which compensates for any alteration in the parenchymatous tissue in the lungs or abnormalities in the bronchial tree in infants soon after the repair of congenital diaphragmatic herniae. Further studies are necessary to determine the changes in these lungs with growth.

There is considerable pathological evidence that significant hypoplasia of both lungs is present in infants who die soon after birth with congenital diaphragmatic herniae of the Bochdalek type. However, there have been conflicting reports concerning the type of hypoplasia present. One suggestion has been that the hypoplasia is predominantly due to a reduction in the number of bronchial generations (Areechon and Reid, 1963), the other that it results from a failure of development of the alveolar region of the lung (de Lorimier, 1970).

Areechon and Reid studied 2 newborn infants with diaphragmatic herniae who died soon after birth, and found a reduction in the number of bronchial generations in both the ipsilateral and contralateral lung, the reduction being greater in the ipsilateral. Bronchial and bronchiolar regions were both affected, with the alveoli relatively less so. The alveoli were probably present in normal numbers for the number of airways but reduced in size.

de Lorimier studied 2 lungs from babies dying with congenital diaphragmatic herniae. He was unable to confirm the findings of Areechon and Reid. He counted the number of bronchial generations in one ipsilateral lung and found this to be within normal limits. The bronchi in all the lungs were quite distorted and he considered that this may have been a factor in the poor expansion of the lungs. In both ipsilateral and contralateral lungs there was an increase in the proportion of parenchymatous tissue. There was a marked reduction in alveolar surface area, the reduction being greater in the ipsilateral lung, and in addition its alveolar ducts were short and poorly formed. On the basis of these findings he postulated that the major abnormality was a failure of alveolar sac and alveolar duct development rather than a reduction in the total number of bronchial generations.

These pathological studies were made in infants who died soon after birth, some still quite premature. The initial clinical problems presented by surviving infants support the pathological evidence that there is significant pulmonary hypoplasia. However, gross hypoplasia may not always persist in patients with diaphragmatic herniae because within one week of operation the ipsilateral lung radiologically may appear to fill the whole hemithorax (Moore et al., 1957).

Apart from the occasional report that the chest x-ray can appear normal within a few months (Young, 1969), little is known of the behaviour of these lungs in infancy, and there have been no physiological studies to define the function of the lungs in the babies who survive. The present study was undertaken to determine whether there was physiological evidence of pulmonary hypoplasia in infants surviving successful repair of a diaphragmatic hernia. Further, it was hoped to document lung growth in those infants with demonstrable reduction in lung volume soon after the repair of the hernia. The only other reports of physiological studies of this type are of measurements of lung volume in older children, 6 to 19 years after operation (Chatrath et al., 1971; Wohl et al., 1973).
Patients

In the 6-year period, 1968 to 1973, 64 infants with congenital posterolateral diaphragmatic defects presented to the Royal Children's Hospital, Melbourne, and were treated surgically. 48 presented and were operated on in the first day of life. 31 (64·6%) survived. 16 presented after 24 hours, and 14 (87·5%) of these survived. The combined mortality was 30%. Gross (1964) reported an overall mortality of 32% since 1950. Young (1969) reported a mortality of 35% at The Hospital for Sick Children, London, with a mortality of over 50% in those who presented in the first 24 hours.

Of the 19 deaths at the Royal Children's Hospital, 16 occurred in the first 3 days of life. All had significant hypoplasia of the ipsilateral lung and in most cases the contralateral lung was also smaller than normal for the infant's weight. 3 died later—1 with a cardiac anomaly and 2 from sepsis. 2 of these 3 patients also had definitive necropsy evidence of pulmonary hypoplasia with lungs one-half to one-third of normal weight.

From a survey of all perinatal deaths in Victoria during this same 6-year period it was found that approximately 50 babies died with a congenital diaphragmatic hernia and these would include many with other abnormalities. The surviving children represent approximately 50% of all babies born with a congenital diaphragmatic hernia and a representative cross-section of these were studied. They certainly included many with massive diaphragmatic herniae in whom there were large amounts of abdominal viscera in the thorax (Fig. 1).

Twenty-two infants with repaired diaphragmatic herniae of the Bochdalek type were studied. Of this group, 18 were from the group of 31 who presented before 24 hours and survived. 9 of these presented within the first 6 hours of birth. The other 4 presented after 24 hours. 19 were tested within 4 weeks of birth, and 3 were first tested after 4 weeks. Repeat studies were done where possible at approximately 4-monthly intervals during the first year of life. Many had a normal study by 6 months of age and were not restudied, and some children could not return for further tests, so that 15 were restudied between 1 and 6 months and 8 between 6 and 12 months. All children were reasonably well when studied, so that those who had a stormy postoperative course were not tested until they were feeding well and breathing satisfactorily in room air. Children with other abnormalities were not included.

Methods

A 49-litre whole body plethysmograph was used to measure tidal volume and thoracic gas volume (TGV) which are the only measurements of lung volume possible in a subject unable to co-operate with the investigator. TGV is the volume of air in the lungs at the end of a normal expiration and is equivalent to functional residual capacity. It has been shown that TGV can be related to body weight in healthy infants and a range of normal values has been published (Phelan and Williams, 1969).

By recording simultaneously, during spontaneous respiration, tidal volume flow at the mouth and intrapleural pressure by means of an intraoesophageal balloon, values for dynamic compliance and pulmonary conductance (reciprocal of resistance) were derived. Full details of the techniques are given elsewhere (Phelan et al., 1969). Dynamic compliance is a measure of the elastic properties of the lung, though it can be reduced with bronchiolar disease, and pulmonary conductance measures predominantly (inversely) resistance to flow in the larger airways.

Normal values and 95% confidence limits for the parameters measured are (1) TGV/kg 30·0±6·6 ml/kg; (2) specific compliance 0·056±0·02 l/
cmH₂O per l (TGV); (3) conductance/length = 0·22 l ± 0·132 l/s per cmH₂O per cm².

Results

Fig. 2 shows the normal range and the percentage of children either within or outside this range for each test in 3 different age groups of less than 4 weeks, 4 to 25 weeks, greater than 25 weeks. Of the 19 patients studied within the first month, 13 had normal measurements of TGV. 4 were in fact normal when studied in the first week of life. Of the 6 with abnormal TGV, 3 had a marked reduction and 3 an increase. By 6 months of age no patients had reduced TGV. 3 still had evidence of hyperinflation, which was not evident when studied after 6 months. One child studied after 25 weeks had a TGV which was then less than predicted. This patient could not be restudied as he was too big for the plethysmograph by this stage, but all other parameters measured were normal.

Thirteen of the babies under 4 weeks of age had normal dynamic compliance while 6 were reduced at the initial study, but only 3 were reduced at 4 to 25 weeks, and all were within normal limits after 25 weeks. Of those with reduced dynamic compliance, two-thirds had associated overinflation. No abnormality in the measurement of pulmonary conductance was found at the initial study. Only 1 patient had a low conductance on subsequent examination.

Discussion

This preliminary study of respiratory function in infants after successful surgical repair of a diaphragmatic hernia indicates that the majority of babies achieve normal lung volumes in the first weeks of life. Some have reduced lung volumes which tend to become normal later in the first year of life and a small number have hyperinflation, probably due to gas trapping. These physiological observations support the radiological findings. In most children chest x-rays appear normal, though in some there may be a raised diaphragm while others appear emphysematous (Chatrath et al., 1971).

TGV is an indication of the aerated lung volume but a measurement of total lung capacity (TLC) would be a more definitive index of lung hypoplasia if present. Unfortunately, it is not possible to measure TLC in infants because of the need for active patient co-operation. However, normal TGV suggests that compensation has occurred and gross hypoplasia is no longer functionally apparent.

Oxygen therapy may have contributed to the abnormal lung volumes in a few of the infants as some were initially in considerable respiratory distress and were nursed in high oxygen concentrations. This can cause alteration in surfactant production leading to atelectasis and airway disease with gas trapping (Northway et al., 1967). However, not all patients with an abnormal TGV had high oxygen concentrations for a significant period.

The initially low dynamic compliance in 6 babies may have been due to one of two factors. Increased lung stiffness can result in a low compliance; however, dynamic compliance can also be reduced due to maldistribution of ventilation if an airway abnormality is present (Woolcock et al., 1969). The low compliance in some of these babies could be evidence of an initial increase in the proportion of parenchymatous tissue as suggested by de Lorimier (1970). However, 4 out of 6 babies with low dynamic compliance had hyperinflation as well, suggesting an abnormality in the small peripheral airway due to either the disease or to therapy. It appears that with alveolar development during the first year of life, both lung volumes and lung compliance became normal, often very soon after birth and certainly by 6 months.

The measurement of conductance generally reflects the calibre of major airways and the normal pulmonary conductance in these studies suggests that there was no reduction in the total cross-sectional
area of larger airways. However, Hogg et al. (1970)
have shown that peripheral airways contribute
up to 50% to this measurement in infants. Con-
sequently, it is highly unlikely that there was
significant airway abnormality in most of these
children. In addition, the total cross-sectional area
of peripheral airways is probably not markedly
reduced in these babies as this should have led to
some decrease in conductance. Areechon and Reid
(1963) found a decrease in the number of bronchial
branches in the lungs of 2 babies who died with
diaphragmatic herniae. If such a reduction is present
the size of the individual bronchi and bronchioli
may be larger and lead to relatively normal numbers
of alveolar ducts during the first year of life which
could produce normal conductance. Marked hyper-
inflation of reduced numbers of alveoli is unlikely
in these children as this should lead to more marked
abnormalities in dynamic compliance.

It is not possible from these studies to differentiate
between the differing conclusions of de Lorimier
and Areechon and Reid. From their studies and our
data it appears that a spectrum of severity of pul-
mmonary hypoplasia occurs. Any reduction in
bronchial branching in surviving babies is not
likely to be gross. The lungs studied by the patho-
logists were those of babies who died within a few
hours of birth or those associated with experimentally
induced herniae in lambs (Starrett and de Lorimier,
1935) and most of the 19 babies who presented
to the Royal Children's Hospital and died probably
fall into this group with gross hypoplasia. However,
70% survived and the babies studied appear to
represent a wide spectrum of those who presented,
and the physiological measurements in these babies
suggest that rapid development of the alveolar duct
and alveolar region occurs during the early weeks
of life and results in normal tests of pulmonary
function, even when marked hypoplasia was
initially present.

Further detailed studies in children over 8 years of
age after alveolar multiplication has stopped may be
necessary to determine whether structural
abnormalities in the airways or parenchyma exist in
these patients. Chatrath et al. (1971) assessed ventila-
tory function in children between 6 and 12 years of
age after repair of a diaphragmatic hernia. Lung
volumes were normal. Forced vital capacity (FVC)
and forced expiratory volume in 1 second (FEV₁)
were reduced. These results were interpreted as
evidence of abnormal lungs probably due to emphy-
sema. However, the slow vital capacities were nor-
mal and FEV₁/FVC ratios had a mean of 91%,
suggesting that the reduction in forced manoeuvres
were more likely to be due to a diaphragmatic
of abnormality or to a technical error. Most
the children they studied had normal chest x-rays.

Wohl et al. (1973) found that patients between
6 and 19 years of life had normal lung volumes and
forced expiratory manoeuvres including maximum
expiratory flow volume curves. However, perfusion
to the side of the hernia was decreased when assessed
by scanning after administration of radioactive
xenon. The vascular abnormality suggests that some
change in lung structure persists into late childhood.
Abnormalities of perfusion to the side of the hernia,
both local and generalized, were also reported by
Reid (1975).

The results of this study suggest that while the
lungs of infants surviving the repair of a congenital
diaphragmatic hernia may be stiff soon after birth,
they have the potential to develop apparently normal
pulmonary function within the early weeks of life.
Structural abnormalities may persist in these lungs,
but they are unlikely to be gross and do not affect
lung function. Further studies are necessary to
determine whether this status is maintained into adult
life.

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