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Fate of Hypoplastic Lungs After Repair of Congenital Diaphragmatic Hernia

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Chatrath, R. R., El Shafie, M., and Jones, R. S. (1971). *Archives of Disease in Childhood*, **46**, 633. **Fate of hypoplastic lungs after repair of congenital diaphragmatic hernia.** Ventilatory function was assessed in 14 children between 6 and 12 years of age who had been operated on in infancy for correction of a congenital diaphragmatic hernia. The FEV_1 and FVC were significantly below normal ($P < 0.001$), but the values for lung volume compartments were within normal limits. Low figures occurred particularly in those in whom the lungs had been noted at operation to be markedly hypoplastic. There is, therefore, a residual defect in ventilatory function in these patients despite a satisfactory repair of the hernia and apparently normal *x*-ray appearances of the chest in most instances.

Hypoplasia of the lungs is thought to be one of the main causes of death in newborn infants with congenital diaphragmatic hernia (Snyder and Greaney, 1965; Allen and Thompson, 1966; Campanale and Rowland, 1955; Johnson, Deaner, and Koop, 1967). Some degree of hypoplasia on the ipsilateral side is certainly present in varying degree in some of the infants who survive with or without surgical correction. No published information is available, however, about the ultimate function of the lungs in later childhood. We report here pulmonary function studies in a group of these children followed since infancy.

Material and Methods

The ages of the 14 children studied ranged from 6 to 12½ years (Table I); 12 had a left-sided hernia through the foramen of Bochdaleck and one had a right-sided hernia all of which were operated on in infancy. The 14th child had an eventration of the diaphragm and was not operated on.

The age at operation varied from a few hours to 6 weeks, and in all but the right-sided hernia the abdominal approach was used.

In 5 there was evidence at operation that at least one lung was hypoplastic, in 4 the evidence was doubtful, and in the remaining 5 the lungs appeared normal.

The FEV_1 was measured on a Gaensler type spirometer* which was calibrated daily (McKerrow, McDermott, and Gilson, 1960). Lung volumes were measured using a closed circuit helium dilution apparatus.† All volume measurements were corrected to BTPS.

TABLE I
Clinical Details of 14 Cases of Congenital Diaphragmatic Hernia

	Age (yr)	Height (cm)	Diagnosis	Lung Appearances at Operation	*X*-ray of Affected Lung on Follow-up
1	7·0	114	LDH	Hypoplastic	Normal
2	10·0	132	LDH	Hypoplastic	Emphysema
3	12·6	157	LDH	Hypoplastic	Normal
4	10·0	134	LDH	Hypoplastic	Normal
5	11·0	140	LDH	Hypoplastic	Emphysema
6	8·6	122	LDH	?Normal	L-diaphragm elevated
7	7·6	126	LDH	?Normal	Normal
8	12·0	140	LDH	?Normal	Normal
9	6·0	110	LDH	?Normal	Emphysema
10	11·6	134	LDH	Normal	Normal
11	8·6	127	EVD	—	—
12	7·0	130	LDH	Normal	Normal
13	9·6	130	RDH	Normal	Normal
14	9·0	125	LDH	Normal	Normal

LDH = Left diaphragmatic hernia; RDH = Right diaphragmatic hernia; EVD = Eventration of diaphragm.

Predicted normal values for FEV_1 and FVC were those of Strang (1959), and for lung volumes Blackhall (1969). A full clinical examination and *x*-ray of the chest were carried out before pulmonary function tests.

Results

In 8 cases the *x*-rays of the chest appeared normal, in 1 the *x*-ray was not available. In 3 the left lung field appeared slightly emphysematous by comparison with the right (Cases 2, 5, and 9). There was some elevation of the diaphragm in the remaining 2 (Cases 6 and 8).

The difference between the mean observed FEV_1 (1·39 l.) and the predicted (1·85 l.) is highly

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significant ($P < 0.001$) (Table II). The difference between the mean observed FVC and the predicted is also highly significant ($P < 0.001$).

The values for the observed and predicted functional residual capacity (FRC), residual volume (RV), and total lung capacity (TLC) were not significantly different.

In Cases 1 to 5 the left lung was noted at operation to be very hypoplastic. Though the numbers are too small for statistical analysis it is noteworthy that in all instances the figures for FEV_1 and FVC are appreciably below predicted levels in these patients, but no such difference is evident in lung volume measurements.

Discussion

The most striking finding in this study is the reduction of ventilatory function as measured in terms of FEV_1 and FVC. This contrasts with the normal values for the lung volume compartments (FRC, RV, and TLC). The evidence of hypoplasia in the neonatal period, the normal lung volume measurements at the time of re-examination, and the evidence of emphysema on the affected side on *x*-ray in some cases, suggests that the hypoplastic lung has not grown normally, despite having expanded to fill the pleural cavity. It is presumably overdistended or emphysematous and this would account for the impaired values for FEV_1 and FVC.

This evidence of a persistent abnormality is not surprising in view of the histological studies which have been carried out on these lungs at necropsy in infancy. Butler and Claireaux (1962) reported an excessive number of bronchial structures, while the alveoli and alveolar ducts were reduced in number. Areechon and Reid (1963) found, however, that the number of bronchial branchings was greatly reduced and that the alveoli were relatively less affected. Though the histological evidence is not consistent, clearly there are abnormalities present at the time of birth which might be expected to persist.

Many studies of growth in the postnatal period have been made. Short (1950) thinks that growth takes place by an increase in the linear dimensions of existing lung units, whereas Emery and Mithal (1960) state that there is an increase in the number of units. Dunnill (1962) showed that the number of alveoli increases over tenfold, mainly during the first 8 years of life, and that after this age growth takes place by an increase in the linear dimensions of existing alveoli.

Hypoplasia of the lung has been produced experimentally in fetal lambs following the creation of a diaphragmatic hernia *in utero* (De Lorimier, Tierney, and Parker, 1967). Snyder and Greaney (1965) reported a series of neonates with congenital diaphragmatic hernia in whom at necropsy the ipsilateral lung weighed on average 15 g and the contralateral lung 19·5 g. The combined weight of the two was 30% below normal. Allen and Thomson (1966) reported similar findings.

All the evidence points, therefore, to a defect in growth of lung structure which may have been

TABLE II

Respiratory Function on Follow-up

Case No.	FEV_1		FVC		FRC		RV		TLC	
	Obs.	Pred.	Obs.	Pred.	Obs.	Pred.	Obs.	Pred.	Obs.	Pred.
1	0·88	1·35	1·18	1·55	1·15	0·70	0·35	0·35	2·11	1·65
2	1·42	1·90	1·83	2·13	1·29	1·05	0·71	0·53	2·73	2·45
3	1·64	3·03	1·90	3·54	1·49	1·75	0·69	0·85	3·40	3·90
4	1·71	1·93	1·82	2·10	1·09	1·15	0·51	0·55	2·42	2·50
5	1·16	2·13	1·45	2·37	1·35	1·25	0·82	0·63	2·95	2·80
6	1·16	1·54	1·20	1·78	0·62	0·85	0·33	0·43	1·69	1·95
7	1·24	1·67	1·56	1·90	0·94	0·92	0·51	0·47	2·24	2·20
8	2·12	2·17	2·32	2·47	2·10	1·20	1·20	0·63	3·50	2·80
9	1·00	1·28	1·20	1·47	0·73	0·60	0·39	0·37	2·64	1·30
10	1·60	2·00	1·82	2·25	1·52	1·15	1·09	0·55	3·34	2·50
11	1·40	1·70	1·71	1·90	0·87	0·95	0·126	0·48	2·20	2·20
12	1·50	1·81	1·95	2·04	1·07	1·00	0·59	0·51	2·67	2·35
13	1·34	1·73	1·48	1·91	1·12	0·98	0·59	0·50	2·40	2·30
14	1·22	1·11	1·41	1·81	0·67	0·92	0·15	0·47	1·65	2·20
Mean	1·39	1·85	1·63	2·09	1·14	1·03	0·58	0·52	2·57	2·39
SD	0·308	0·414	0·322	0·485	0·381	0·265	0·303	0·120	0·565	0·580
Diff. of mean	−0·461		−0·454		+0·110		+0·053		+0·189	
t	−5·165		−4·265		1·31		0·73		1·37	
P	0·001		0·001		0·20		0·20		0·20	

induced in fetal life by the presence of the hernia. Postnatally, in view of the evidence presented here, the growth process presumably never catches up despite repair of the hernia, so that a residual defect of ventilatory function persists. Further follow-up studies will be important in order to ascertain whether the affected lung will remain in a stable condition or whether further deterioration will occur with age.

The nature of the ventilatory defect will require elucidation. There could be a reduction in the number of elastic fibres associated with over-distension of the lung and 'trapping' of gas during the expiratory phase of respiration. The possibility of progressive loss of elastic fibre exists. On the other hand, impairment may be due primarily to the abnormally small number of functioning lung units.

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