CONGENITAL (BOCHDALEK) DIAPHRAGMATIC HERNIA: PROBLEMS IN DIAGNOSIS AND MANAGEMENT*

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SUMMARY

A review of 24 children with posterolateral (Bochdalek) diaphragmatic hernia over a five-year period was carried out to highlight the problems of diagnosis and management. Nine children were delivered in the Maternity Hospital Kuala Lumpur, giving an incidence of 1 : 10,000 live births which is half the expected incidence. Difficulty in diagnosis is apparent from the large number of initially misdiagnosed cases (29%) and those not detected soon after birth (71% diagnosed after 24 hours). Less than half the babies had associated anomalies, commonest being malrotation and ipsilateral lung hypoplasia.

Mortality (20.8%) appears to be related to the degree of lung hypoplasia and shunting, and the birthweight of the babies. Current evidence indicates that pulmonary hypertension is the main factor in the chain of events beginning with lung hypoplasia, which ultimately leads to their demise. Various methods to overcome this complication have been evolved which appear to give some hope for these high-risk infants.

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INTRODUCTION

Vincent Bochdalek described the posterolateral hernia in 1848,[1] but it was only a hundred years later that the first case was reported in this region by Shanmugaratnam and Haridas.[2] Damodaran *et al.*,[3] reported the first successfully treated case in Malaysia in 1973 and Somasundaram[4] gave a brief synopsis of 20 cases in his inaugural lecture in 1979. Hitherto, no detailed study of this condition has been made in this country.

The purpose of this paper is to highlight the problems of diagnosis and treatment and to review recent developments in the management of Bochdalek diaphragmatic hernias in childhood.

MATERIALS AND METHOD

Case notes of children diagnosed as having congenital diaphragmatic hernia admitted to the General Hospital Kuala Lumpur, Universiti

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Kebangsaan Malaysia (UKM) Unit over the five-year period from January 1978 to December 1982 were reviewed. 24 children had a foramen of Bochdalek hernia. Nine had been delivered in the Maternity Hospital Kuala Lumpur, which had 92,842 live births over that period (an incidence of 1: 10,427 LB). Nine were referred from other hospitals around Kuala Lumpur and six came from outside the city.

There were 13 boys and 11 girls: 12 Malays, 4 Chinese and 8 Indians. Their ages ranged between six hours to one-and-a-half years. 21 had left-sided hernia whilst three had right-sided hernia.

RESULTS

19 infants (79%) presented with respiratory distress and 14 (58%) with cyanotic attacks (Fig. 1). Apparent 'dextrocardia' was observed in 14 patients (58%). Chest x-rays were done on all the babies, confirming the diagnosis in 17 cases (Fig. 2a, b).

Of the 20 neonates, seven were initially diagnosed as having pneumonia and treated as such. Four infants presented at the ages of three, six, ten and 18 months; two with recurrent chest infections, one each with bronchopneumonia and failure to thrive.

Barium meal was required for diagnosis in seven patients. Three of these had late onset of symptoms (Fig. 3), two were previously misdiagnosed cases and

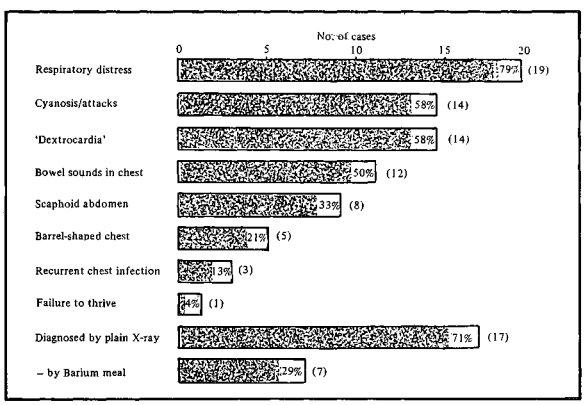

Fig. 1 Features noted in 24 cases of Bochdalek Hernias.

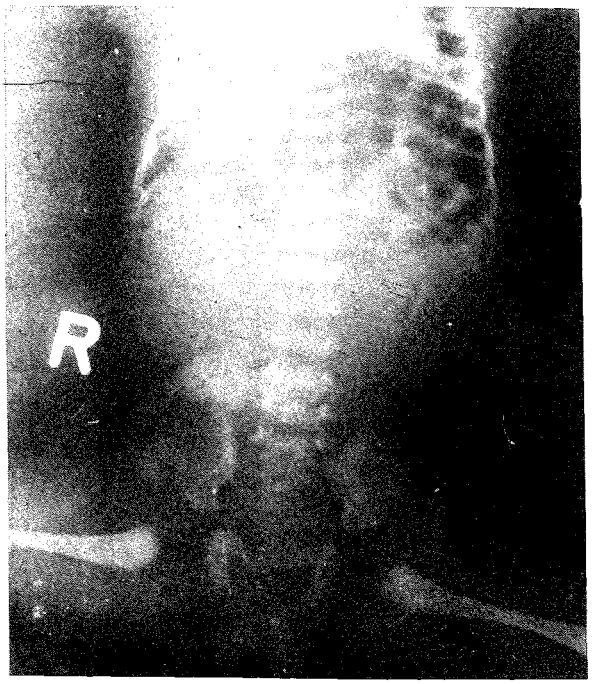

Fig. 2(a) Left-sided posterolateral diaphragmatic hernia. Note a paucity of gas shadows in abdomen and shift of mediastinum.

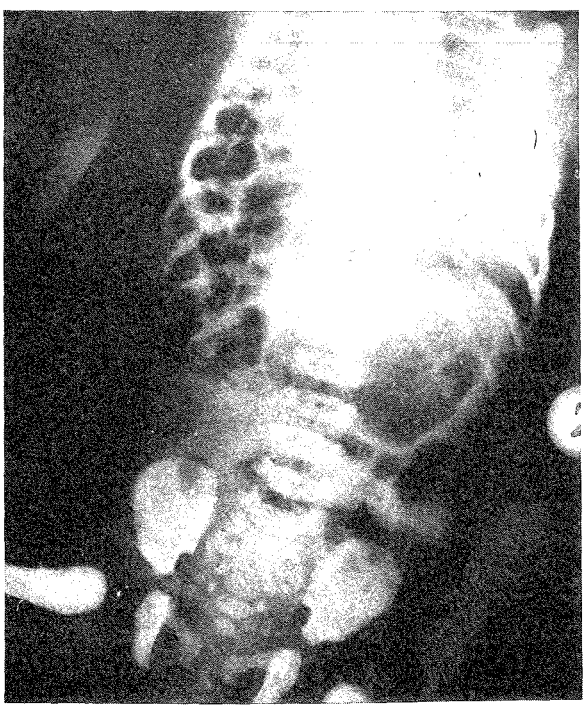

Fig. 2(b) Right-sided posterolateral diaphragmatic hernia.

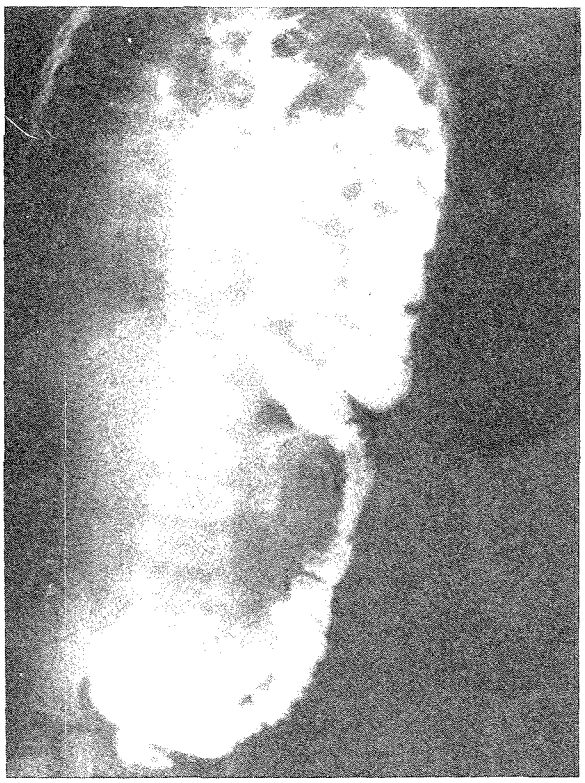

Fig. 3 **Barium meal in a one-and-a-half-year-old boy with left diaphragmatic hernia.**

one had a right-sided hernia. One left-sided hernia had the barium study done elsewhere.

Five of the neonates (25%) had birth weight less than 2.5 kg. There were associated anomalies in 11 babies. Malrotation, the commonest anomaly, was found in seven patients (Table I). One baby had a combination of exomphalos, cleft-lip and palate, imperforate anus and malrotation.

All operations were performed *via* a transverse upper abdominal incision except for one right-sided hernia which required a thoracoabdominal approach because of pneumohepatic adhesion. After the contents were reduced into the peritoneal cavity, malrotation, where present, was corrected. Hernial sacs, found in only 12 patients (50%), were excised. A chest drain was routinely inserted on the ipsilateral side and removed on the third to fifth postoperative day. The diaphragmatic defects were closed with interrupted 3–0 silk. In one patient, a patch of lyophilised human dura was used to bridge the gap. Broad spectrum antibiotics were given routinely. Mean operating time was 93.2 minutes.

TABLE I
ASSOCIATED ANOMALIES IN BOCHDALEK HERNIA IN 11 PATIENTS

Malrotation	7
Congenital heart disease	3
Polydactyly/Syndactyly	2
Exomphalos	1
Cleft lip and palate	1
Imperforate anus	1
Others	2

All infants were put on IPPV postoperatively except for two who were extubated within two hours of operation. Inflation pressure of 30 cm of water or less was used, and FIO_2 ranging from 30% to 100% was given. Once stable, as determined clinically by blood gases and x-rays, they were weaned to IMV and then to 2–3 cm water of CPAP before extubation. The mean period of endotracheal intubation was 3.3 days for survivors. Non-survivors were left intubated up to the time of death, the longest being 28 days.

Five infants died postoperatively (20.8%). All but one death occurred in babies who were symptomatic shortly after birth and were operated on within 24 hours of birth (Table II). One preterm baby with exomphalos major and other major anomalies was operated on at 42 hours and died. Average time of death for non-survivors was 10.6 days postoperatively with a range of two to 28 days.

TABLE II
COMPLICATIONS OF BOCHDALEK HERNIA REPAIR IN 11 PATIENTS

Pneumonia	7
Difficult to wean off ventilation	6
Wound infection	4
Contralateral pneumothorax	2
Cardiac failure	3
Septicaemia	2
Others	3

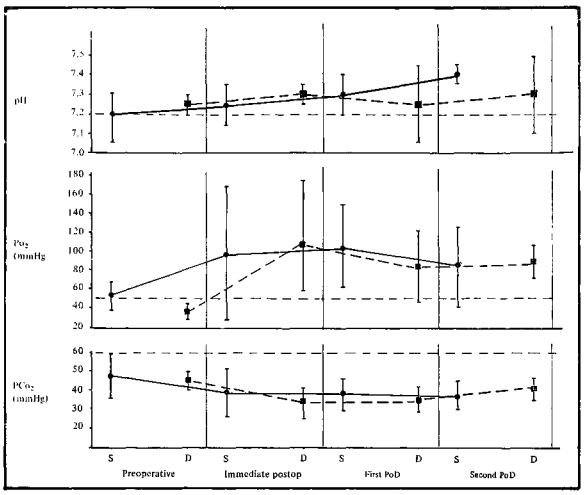

Fig. 4 **Blood gases compared between survivors and non-survivors: the first few days.**
S = Survivor (●); D = Death (■); Mean ± ISD.

Blood gases were determined preoperatively in some of the more severely ill newborns (12 survivors and two non-survivors). The mean PO_2 level for non-survivors was 36 mm Hg whilst survivors had a mean of 52.5 mm Hg (Fig. 4). No applicable difference was found in the pH, PO_2 or PCO_2 in the first two post-operative days in survivors and non-survivors.

Virtual shunt was determined by using Benatar's isoshunt curves.[5] Significant adverse factors in prognosis were a left-to-right shunt of over 20%, severe ipsilateral lung hypoplasia, and birth weight under 2.5 kg (all p values < 0.05 using Fisher's exact test) while defect size and the presence of sac were not.

Eleven patients (45.8%) developed 27 complications postoperatively (Table III). Of these patients, eight had more than one complication all of which were cardiopulmonary in origin. Of six patients who had endotracheal tubes for more than five days, four died. All five patients who succumbed could not be weaned off their ventilators. Despite using low inflation pressures, two patients acquired pneumothorax of the contralateral side but both survived. One patient developed a recurrence after two days while another had adhesion obstruction ten months later.

TABLE III
AGE GROUP AND MORTALITY OF DIAPHRAGMATIC HERNIA

Age Group	Number	Deaths
<24 hr	7	4
$24 \leqslant 48$ hr	5	1
48 hr $\leqslant 1$ month	8	0
1 month – 1 yr	3	0
>1 yr	1	0
Total	24	5

DISCUSSION

Congenital diaphragmatic hernia (CDH) continues to be amongst the most poorly recognised congenital conditions in this country. In most advanced countries, the incidence is 1 : 5,000 live births[6,7] and 1 : 2,000 total births.[8] In a recent survey in West Malaysia, the rate was estimated at only 1 : 18,000 live births (unpublished data). This may partly be due to the fact that more than half of deliveries are carried out at home and some 50% of these babies have not been referred to the hospital.[7] The lack of x-ray facilities in the district hospitals may further prevent detection. However, the situation is only slightly better in our large and relatively well-equipped Maternity Hospital where the incidence is 1 : 10,000 livebirths. In other words, at best we may be identifying only 50% of the cases.

There is an obvious lack of a high index of suspicion for the condition: as many as one-third of the neonates were initially misdiagnosed and only 29% of babies were diagnosed within 24 hours of birth (Table II).

Since about 90% of CDH occur on the left side,[8] a child with respiratory distress and apparent 'dextrocardia' should always be suspect (Fig. 1). A plain chest film is the single most useful diagnostic tool (Fig. 2) and barium meal reserved for the unusual cases only.

The four patients presenting after the neonatal period form an interesting group. Though only two

of them had symptoms from birth to the extent that one had failed to thrive, the other two may have had mild dyspnoea which was not recognised until they had the episode of respiratory infection. On the other hand, several authors have reported diaphragmatic hernia appearing in children who previously had normal x-rays.[9,10]

Associated anomalies in live-born CDH babies, apart from the PDA, pulmonary hypoplasia and malrotation are relatively rare.[8,11] The intestines returning from the physiological umbilical hernia are unable to undergo the normal rotation and fixation processes, hence the high incidence of gut malrotation.[12,13] Concomitant exomphalos with CDH has only been reported twice previously.[8,14] The coexistence of these two conditions implies that CDH may be the result of delayed closure of the diaphragm and not the premature return of the gut from the umbilicus.

Immediate management of the baby with CDH entails urgent admission to the ICU for the purpose of stabilisation prior to surgery.[15] Since these babies are poor travellers,[7,11] the fastest and shortest route should be taken. A nasogastric tube is used to decompress the herniated hollow viscera, the head is elevated and, if indicated, an endotracheal tube is inserted prior to transportation to assist ventilation. Since frequent blood gas estimations are usually required, either an arterial line or a continuous percutaneous oxygen monitor is connected to the baby while in ICU. Operation is usually undertaken only after acidosis, hypoxia and hypothermia are at least partially corrected, which usually takes between one to three hours.

The defect in the diaphragm can often be closed primarily but in a few instances, this is not possible in which case either a muscle flap[16] or a prosthetic material is used to close gap.[12,17,18] We have used lyophilised human dura in such situations and have found it to be completely satisfactory. The creation of a ventral hernia and the use of gastrostomy for abdominal decompression as suggested by some authors[15] have not been found to be necessary in our cases.

Postoperative ventilation is normally required but the inflation pressure should not exceed 25–30 cm of water.[19] This is to minimise the risk of causing pneumothorax in the opposite chest since the normal lung tend to be more compliant.

It is well known that those babies who become symptomatic soon after birth and required urgent surgery are associated with a high postoperative mortality.[4,7,12,15,17,18,20-23] In fact the overall mortality in any institution is largely dependent upon the proportion of these high risk cases (Table IV). The low mortality (20.8%) in this report is also probably attributed to this fact.

A number of prognostic indicators have been suggested to identify patients at particularly high risk.[18,20,21] Among them has been the finding of a lowered PO_2 preoperatively,[20] which was also found in our patients. Whilst low birth weight was not considered as an adverse factor by Boles *et al.*[15] We found a significantly higher mortality in this group perhaps reflecting the inadequacy of our

TABLE IV
SELECTED STUDIES SHOWING RELATIONSHIP BETWEEN PERCENTAGE OF HIGH RISK BABIES AND OVERALL MAJORITY

Studies	No. of patients	Percentage of high risk cases <24 hrs old	Overall mortality (%)
Raphaely and Downes (1973)[19]	58	85	41
Ehrlich and Salzberg (1978)[18]	12	92	50
Ruff *et al.*, (1980)[17]	36	72	44
Rose-Spencer *et al.*, (9181)[16]	43	58	50
Marshall and Sumner (1982)[12]	62	74	29
Present Study (1984)	24	29	21

perinatal support. Grotte *et al.*,[13] in a large series of 125 cases had a mortality of 72% among LBW babies compared to 38% for normal birth-weight babies.

Hypoplasia particularly of the ipsilateral lung is almost always present to a certain degree.[8,12,21-22] However, the fact that majority of the non-survivors could attain normal blood gas values immediately postoperatively (Fig. 4) and that death does not occur till after several days of repair seems to suggest that these lungs despite being hypoplastic are capable of maintaining adequate ventilation.[20-23] Recent evidence shows the presence of thick-walled arterioles which remain in a state of tonic spasm hence raising the pulmonary arterial pressure.[21-23] Once this pressure supercedes systemic pressure, right to left shunting can take place at the ductus as well as at the foramen ovale, giving rise to a state of persistent foetal circulation. As the venous admixture deteriorates, a vicious cycle is set up which aggravates the pulmonary hypertension to the point of myocardial failure.

This hypothesis explains the brief 'honeymoon' period wherein the baby seems well and then declines inexorably and dies usually within the first two weeks. Since the recognition of this syndrome there has been an upsurge of interest to try to reverse this process particularly with the advent of vasodilator drugs e.g. tolazoline[12,17,18] and dopamine.[24] Ligation of the PDA has met with limited success[23] because it removes only one shunt and does not alter the pulmonary hypertension.

More recently Hardesty *et al.*,[25] have used extracorporeal membrane oxygenators to briefly tide these babies over, and results have so far been encouraging. Experimental *in utero* hernia repair has been successfully carried out in animals[26] but its clinical application remains uncertain. However, if this becomes possible it would then provide the best way of ensuring subsequent normal lung development.

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