Congenital diaphragmatic hernia

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ABSTRACT

Objective: To study the pattern of congenital diaphragmatic hernia in the Assir Province of Saudi Arabia.

Methods: Medical records of all newborn infants of Saudi nationality admitted and treated for congenital diaphragmatic hernia in the neonatal intensive care unit of Assir Central Hospital, Abha were reviewed. The diagnosis of congenital diaphragmatic hernia was based on clinical, radiological and surgical findings.

Results: Thirty-six cases of congenital diaphragmatic hernia referred from health institutions in Assir Province were seen over a period of 2.5 years (January 1996 - June 1998). Congenital diaphragmatic hernia constituted 7% of total neonatal admissions during the period. Sex incidence was equal and all cases were of the Bochdalek type; 34 were sited on the left and 2 on the right. All the patients were asphyxiated and required ventilatory support at birth. Eight percent of patients had other congenital anomalies. The overall mortality rate was 36%; 14% of the patients died preoperatively, and postoperative death was largely related with persistent respiratory insufficiency and septicemia.

Conclusion: The pattern of congenital diaphragmatic hernia in Assir Province is closely at par with global experience but the incidence of associated congenital anomalies and fatality rates appear to be comparatively low. To improve the outcome, medical institutions in the Assir Province need better facilities to optimize neonatal care. There is a need to provide facilities for extracorporeal membrane oxygenation.

Keywords: Congenital diaphragmatic hernia, pattern.


There is paucity of information regarding the pattern of congenital diaphragmatic hernia among infants in the Kingdom of Saudi Arabia. To our knowledge, there is only one report from Al Khobar in the Eastern Province of the country. In that study, which covered the period 1983-1989, all the 34 patients presented with left-sided defects, there was a male preponderance (4:1), 41% of patients had associated congenital anomalies and the mortality rate was 56%. A similar and recent survey has not been conducted in other parts of the Kingdom; there may be some regional variation in the pattern of this condition. It will also be of interest to compare our findings with the global experience. This communication attempts to evaluate the clinical pattern of congenital diaphragmatic hernia (CDH) seen in the neonatal intensive care unit (NICU) of Assir Central Hospital (ACH), Abha, Saudi Arabia during a period of 2.5 years. For genetic and racial homogeneity, the study was confined to infants of Saudi nationality.

Methods. All neonates treated in ACH for CDH during a 2.5 year period (January 1996-June 1998) constitute the subject of this survey. Charts of patients were reviewed for date of admission, sex, age at diagnosis, mode of presentation, operative findings, management and outcome (dead or discharge). Assir Central Hospital serves as the only tertiary and referral institution for all the hospitals and clinics in the Assir Province of Saudi Arabia. The institution has no obstetric service of its own; therefore the NICU of the hospital is entirely a

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referral unit which serves the whole Province (land
area 80,000 km²; population 2 million). Assir Central
Hospital is the only medical facility, which can
provide Level 3 neonatal service in the Province
which has 19 hospitals and 238 primary health care
centres (PHCC). Indeed, it is the only hospital in the
Province which provides pediatric and neonatal
surgical services. The NICU has a bed capacity of
20, and admits an average of 250 infants per year.

All the patients in this study were referred from
other hospitals in the Province with a diagnosis of
diaphragmatic hernia based on both the clinical and
radiological findings at birth. Three of the cases
were diagnosed in utero by ultrasonographic
scanning. As a policy, we insisted by fax or phone
communication that all CDH patients referred to our
institution should have a nasogastric tube inserted
and an endotracheal intubation for the purpose of
adequate ventilation during transportation. Face-
mask oxygenation with Ambu-bagging was strictly
forbidden.

On arrival in the ACH, a detailed history and a
thorough physical examination were carried out.
Mandatory investigations on admission included
blood gases, complete blood count, urea, electrolytes
and repeat radiographs of the chest and abdomen.
Also, as a routine, septic work-up (blood and urine
cultures) was conducted on every infant upon
admission. Spinal tap was performed on the infants
only if they demonstrated clinical signs of sepsis.
The diagnosis of CDH was confirmed by radiological
and surgical findings. No autopsy was performed on
any of the deceased infants.

Pre-operative treatment of the patients consists of the
administration of oxygen, intravenous infusion,
and gastric decompression with nasogastric tube and
prophylactic antibiotics. As a policy, surgery was
undertaken only when the infant was stabilized and
serial blood gas results found satisfactory.

Pancuronium, inotropics and tolazol were administered
pre and postoperatively, when indicated. Facilities for extracorporeal membrane
oxygenation (ECMO) are not available in our centre.

Post-operatively, ventilatory and other intensive
care supports were continued in the NICU. As a
routine, fentanyl was added to the drug regimen for
every infant post-operatively. Serial arterial blood
gases and chest radiographs were further conducted
to monitor the patients’ condition and the progress of
lung expansion.

Statistical test of significance was used as and
when necessary.

Results. During the period of this survey, there
were 38 consecutive neonatal admissions with
diaphragmatic hernia in the unit. Two of these were
of non-Saudi nationality and were therefore excluded
leaving 36 for further analysis. During the period,
there were 549 neonatal admissions in the unit; CDH
therefore constituted 6.6% of admissions. The sex
distribution of the infants was equal (18 males and 18
females). One infant had a positive history of a
similar defect in a sibling who died.

Thirty-three patients were full-term with birth
weight ranging from 2.15 to 4.42kg; 3 were
premature (30, 32 and 33 weeks gestation; birth
weight, 1.4, 1.65 and 2.03kg). All the infants
presented with respiratory distress and cyanosis from
birth. Apgar scores were available in only 22 of the
36 infants; the averages of the scores were 3.5 in one
minute and 5.6 in 5 minutes. From the history, all
the patients required respiratory support at birth
which involved endotracheal intubation with manual
ventilation in 26 cases. Face mask ventilation with
Ambu-bagging had been applied on 10 infants before
referral.

All the defects were of the Bochdalek type; 34 on
the left and 2 on the right side of the hemithoraces.
Apart from the detection of bowel sounds on the right
hemithorax, the clinical signs of the 2 infants with
right-sided hernia were not different from those with
the left-sided lesion. Fourteen (39%) of the 36
infants presented with scaphoid abdomen. Serial
post-operative chest radiographs demonstrated small
lung volume with slow expansion on the ipsilateral
side thus suggesting lung hypoplasia which was a
constant feature in all the cases. Three (8.3%) of the
infants had other congenital anomalies, which
included a true dextrocardia (situs solitus),
ventricular septal defect and Turner syndrome.

Operative treatment. Five of the patients
succumbed without surgical intervention because
their condition was persistently too unstable to permit
surgery. The remaining 31 were operated upon at
varying ages: 16 in less than 24 hours; 7 on the 2nd
day of life; 2 each on the 3rd and 4th day and 4 on
days 5, 7, 9 and 10.

Surgery involved a subcostal abdominal approach to
reduce the hernia and close the defect. The size of
the defect was not indicated in any of the operative
notes and no prosthesis was used in the repair of the
defects. Twenty-six of the patients had under-water
seal tube thoracostomy drainage on the operated side;
5 had no chest tubes. None of the patients with
scaphoid abdomen had a staged reduction of the
abdominal contents.

Hernia contents. Of the 31 patients who
underwent surgery, 4 (13%) had one abdominal
organ while a majority (27 or 87%) had 2 to a
maximum of 5 organs in the thorax. The liver had
hemiated in the 2 infants with a right-sided and in
one of the infants with a left-sided defect.

Mortality rate. There was a total of 13 deaths in
the series, 5 preoperatively and 8 postoperatively,
thus giving an overall mortality rate of 36%. Of the
5 who died before surgical intervention, 4 succumbed
before the age of 24 hours and one at the age of 4
days. The main cause of death in all the 5 was severe
Table 1 - Outcome related to the number of abdominal organs in the thorax of 31 operated patients.

<table>
<thead>
<tr>
<th>Number of organs</th>
<th>Died</th>
<th>Alive</th>
<th>Total</th>
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<tbody>
<tr>
<td>≤ 3 organs</td>
<td>4</td>
<td>13</td>
<td>17</td>
</tr>
<tr>
<td>&gt; 3 organs</td>
<td>4</td>
<td>10</td>
<td>14</td>
</tr>
<tr>
<td><strong>TOTAL</strong></td>
<td><strong>8</strong></td>
<td><strong>23</strong></td>
<td><strong>31</strong></td>
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respiratory failure associated with pulmonary hypertension, and pneumothorax in 3 of them. Seven or 87.5% of the 8 patients who died postoperatively had their course complicated with septicemia; 3 of the 8 cases (1 of them without septicemia) also developed respiratory failure with persistent fetal circulation after surgery. All the 13 infants who died had Apgar scores below 3 in one minute and were intubated in the delivery room. Of the 14 infants with scaphoid abdomen, 5 (36%) died; 9 survived. Fisher's exact test was applied which was significant (P = 0.005), hence it can be predicted that the absence of scaphoid abdomen is associated with increase in survival rate and vice versa. We attempted to relate the mortality to the number of abdominal viscera in the chest but when the Fisher's exact test was applied, the P value was not found significant (P = 0.534) (Table 1). Therefore, the number of abdominal organs in the chest does not appear to influence the prognosis of patients with CDH.

**Discussion.** As a policy, it is mandatory that any hospital patient in the Assir Province who requires a transfer for the purpose of specialized investigations and treatment must first be referred to the ACH. We therefore assume that ACH had pulled all the diagnosed cases of CDH in the Province. Nevertheless, the true incidence of CDH in our localities cannot be accurately determined as some cases with this anomaly die in utero and some live borns with this defect may not be diagnosed before death. This limitation applies generously to our environment where autopsy examination is not possible. This study therefore can only provide a clue regarding the incidence of the defect among the Saudi population in the Assir Province.

The study has revealed that diaphragmatic hernia, as a single entity, constituted up to 7% of neonatal admissions in the unit during the period, and we regard this as substantial. Our admission rate for CDH is at the average of 15 cases per year which is more than double the frequency in the Eastern Province (6 cases per year). A previous study on congenital malformation among Saudi infants in the same environment, revealed that CDH alone constituted 8% of all major birth defects encountered in the Assir Province. All these suggest that this birth defect may be contributing significantly to the perinatal and infant morbidity in the Assir Province. However, the true incidence of CDH in the Assir Province can only be determined based on the total live births in the environment. Such a figure on live births is not yet available.

The sex incidence of infants with CDH in our environment is equal. This is at variance with the observation in the Eastern Province of Saudi Arabia where there is a male preponderance of 4:1. The explanation for this regional variation is not obvious. Perhaps a longer period of study with a larger patient population could provide a more reliable sex differentiation.

In the present study there was a predominance of the defect (94%) on the left side, which is in par with global experience. The literature variously reports 70-85% on the left; 5% can be bilateral but no patients in our series had a bilateral defect. One hundred percent of our cases presented with the Bochdalek type of hernia. A majority (87%) had multiple abdominal organs as contents of the hernia but the number of abdominal viscera incarcerated in the thoracic cavity did not appear to affect the prognosis. Scaphoid abdomen seemed to be associated with a poor outcome and this may be associated with the reduction of the chest contents into the small capacity abdomen which could have splinted the diaphragm thus further compromising the respiration.

The survey in the Eastern Province of Saudi Arabia identified a 41% incidence of associated congenital anomalies among infants with diaphragmatic hernia. Generally, the frequency of major congenital malformation is increased in infants with diaphragmatic hernia and the global average figure is as high as 20 - 30% of cases. In the present survey, only 8% of the patients had additional birth defects which were on their own, single and isolated, apart from the case with Turner syndrome. Our figure of 8% suggests that associated congenital malformation is comparatively rare among Saudi infants with CDH in the Assir Province.

Congenital diaphragmatic hernia is not known to be genetic or familial; nevertheless, an autosomal recessive inheritance mode has been suggested in families with complete agenesis of the diaphragm. In one patient who succumbed due to a left-sided defect, there was a positive family history of CDH in one of the older siblings who had died with a similar defect but none of this pair had any other associated congenital anomalies. A study in the Al Qassim area of Saudi Arabia has revealed that a maternal age above 25 years, chronic maternal disease such as diabetes, and consanguineous marriage (54%) were associated with a high incidence of birth defects. Consanguineous marriage is an important correlate of congenital malformation though we are unable to
establish this regarding CDH due to the limitation of this study which was undertaken in a purely referral unit. For instance, some of the referral notes were too sketchy and the parents were rarely able to provide the sociodemographic aspects of the history.

The mortality rate in our centre was 36%, the Al Khobar figure was 56%; and the global figure is currently 40-50%. We attribute this comparatively better outcome in our centre to the caution and care observed before and during transportation whereby the stomach was continuously decompressed and the airway was well secured. Also, only the infants who were hemodynamically stabilized with satisfactory blood gas were taken to surgery. It has been identified that the outcome is poor if a very ill infant is rushed to surgery; currently, attention has shifted to extending the period of stabilization to correct hypoxia, acid base aberration and hemodynamic status before surgery. In our experience, the use of muscle paralytics, fentanyl and tolanzine to combat persistent pulmonary hypertension of the newborn with persistent fetal circulation is not yielding good results in patients with congenital diaphragmatic defect. This also appears to be the experience of other authors. The use of ECMO could have salvaged some of these infants.

Most authors have identified associated major anomalies, symptoms before 24 hours of age, and delivery in non-tertiary centre as factors that convey poor prognosis. In our series, all the infants were born in non-tertiary centres, all those who perished had required intubation in the delivery room due to severe birth asphyxia, and septicemia played a major role in postoperative mortality. These poor prognostic factors which involved patients with CDH in our environment need to be addressed, especially the issue of post-surgical infection which is highly preventable.

We recommend that all the hospitals in the Province with obstetric units should update their facilities for neonatal intensive care so as to improve the outcome of neonates at risk. The provision of ECMO facilities in our unit is necessary.

References