

CHARACTERISTICS OF CONGENITAL DIAPHRAGMATIC HERNIA AND ITS RELATIONSHIP TO PROGNOSIS WITH SPECIAL MENTION ON SHORT-TERM COMPLICATIONS- A RETROSPECTIVE STUDY FROM A NEUROSURGICAL PERSPECTIVE

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ABSTRACT

BACKGROUND

CNS anomalies coexist in up to 10% of non-syndrome CDH cases. Hence, the diagnosis, treatment and prognosis of CHD is of interest to paediatric neurosurgeons. Congenital diaphragmatic hernia (hereby referred to as CDH) is a relatively rare anomaly with a prevalence of 1 in 3000 livebirths. CDH present with varying characteristics (side, contents, presence of sac, morgagnian hernia), which in turn influence its outcome. Study details the characteristics of CDH, their impact on outcome, as well as short-term complications of CDH in general.

MATERIALS AND METHODS

Retrospective study- Descriptive, based on case records. Study subjects were all inpatients admitted with CDH during the study period in Paediatric Surgery Department, SAT Hospital, Trivandrum. No definite sampling method has been adopted for the study. Consecutive cases of CDH have been recruited. Statistical analysis is done by Chi-square test, proportion and percentage analysis. Data were analysed using computer software, Statistical Package for Social Sciences (SPSS) version 10.

RESULTS

Cases which contained stomach as a content (18/56 or 32.14%) had a poor survival (8/18 or 44.44%) and this result was found to be statistically significant (P value 0.033). Sac was seen in 15 (26.7%) cases and the survival was poor in these cases (7/15 or 46.7%, P value 0.096). None of the cases had any CNS anomalies. Outcome was bad for right-sided hernias. Short-term complications other than postoperative death were few.

CONCLUSION

Presence of sac as well as stomach as a content of sac in CDH leads to poor survival. None of the cases had coexisting CNS anomalies, which may not be statistically significant. It may be due to the greater number of stillbirths in fetuses associated with CNS anomalies (which were not included) in our study group during the given study period. Mortality of right-sided hernias exceeded that of left-sided defects, which is against the commonly held belief (our study is supported by Touloukian-Gibson report and Skari, et al study). Surgical complications are few and mainly due to respiratory infection and adhesive obstruction. Surgically-treated patients do well if they survive the operation.

KEYWORDS

Congenital Diaphragmatic Hernia, CNS Anomalies, Characteristics of CDH, Prognosis, Short-Term Complications.

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BACKGROUND

Congenital diaphragmatic hernia is defined as the herniation of abdominal contents through a congenital defect in the diaphragm.^{1,2} The prevalence of CDH is around 1 in 3000 livebirths, the left-sided hernia being more common accounting for 80-85% of cases.³

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CDH has been described as early as 1679 by Bonet. In 1848, Bochdalek described CDH as occurring through a posterolateral defect, which has from that time borne his name.^{1,2,3} In 1901, Aue carried out the first surgery, which was unsuccessful. The first patient to survive operative correction of a Bochdalek hernia in the newborn period was operated on by Gross in 1946.⁴ The herniation in CDH occurs usually through a posterolateral defect (Bochdalek's hernia), but can also occur through a defect in the parasternal region (foramen of Morgagni hernia) or through the oesophageal hiatus (hiatus hernia).^{5,6} Hiatus hernia is, however, considered separately; although, called a hernia, it is usually not a true hernia. Foramen of Morgagni hernia accounts for 2% of diaphragmatic defects and is found in older children and adults. The prevalence of true hernia (hernia with a sac) is between 10 and 38%. The contents of the hernia vary a

great deal, which also has a prognostic significance. On the left side, the hernia usually contains small intestine, spleen, stomach, left lobe of liver and most of the colon. Hernia on the right side usually contains the liver and variable amounts of small and large intestine.^{1,2}

Congenital anomalies of the diaphragm are either due to fusion defects (commonest is posterolateral hernia) or due to defect in the formation of diaphragmatic muscle (eventration of diaphragm). Despite, the apparent simplicity of the anatomic defect, the pathophysiology is complex. In most of the cases, herniation of abdominal viscera into the thoracic cavity during gestation prevents normal growth and development of ipsilateral lung. The mediastinal shift contributes to hypoplasia of contralateral lung. There is a decrease in both the number and size of the respiratory units and a corresponding decrease in pulmonary vascular bed. This leads to increased pulmonary vascular resistance. Increase in the muscle content of the media of pulmonary vessels lead to development of persistent pulmonary hypertension. This leads to persistent foetal circulation with shunting of blood from R-L through PDA and foramen ovale, which results in hypoxia, hypercarbia and acidosis.⁶

Currently, about 20% of individuals with CDH have an identifiable cause for their diaphragm defect, which are classified as syndromic CDH. In the remaining 80-85% of cases with CDH, the aetiology is not known and likely caused by small genomic microdeletion or microduplication, a mutation in a major gene important for diaphragm development, polygenic inheritance and multifactorial gene-environment interactions.

At least, one third of infants with CDH have additional major malformations that do not occur as part of currently recognised monogenic syndrome or chromosome abnormality.⁴ The most common associated malformations are cardiovascular (10-15%), central nervous (10%), musculoskeletal and genitourinary. CNS anomalies coexist in up to 10% of non-syndromic CDH cases; the most common diagnosis are neural tube defects and hydrocephalus (David and Illingworth, 1976; Dillon et al, 2000; Dott et al, 2003). The reason for the common association with a neural tube defect is not known, but has been postulated to be a problem of schisms-fusion or midline instability (Czeizel et al, 1981; Opitz, 1982).^{7,8,9,10} Hence, the diagnosis, treatment and prognosis of CHD is of interest to paediatric neurosurgeons also.

Increasing use of prenatal ultrasound has led to the discovery of diaphragmatic hernia in the foetus.¹¹ Newborns present with respiratory distress, absent breath sounds on ipsilateral side and occasionally bowel sounds in the chest. The apical heart is placed to the side opposite the diaphragmatic defect.⁵ The infant has increasing cyanosis, tachypnoea and retraction and may rapidly succumb from respiratory failure.

The key determinants of mortality are-

1. Whether the CDH is isolated or complex.⁵
2. The degree of pulmonary hypoplasia.

3. The severity of pulmonary hypertension in the perinatal period.
4. Whether the hernia is right-sided, left-sided or bilateral; bilateral CDH always confers a very high mortality.
5. Other predictors of high mortality include very high alveolar-arterial oxygen difference postoperatively, low PO₂ and high PCO₂ not corrected with mechanical ventilation, over distension of contralateral lung with shift of mediastinum to side of lesion postoperatively and evidence of pneumothorax.^{5,12}

Since the availability of ECMO (extracorporeal membrane oxygenation) and HFV (high-frequency ventilation), the management and survival of patients with diaphragmatic hernia has drastically changed.

ECMO basically is a cardiopulmonary bypass used for long-term support ranging from 3-10 days. The purpose of ECMO is to allow time for intrinsic recovery of lungs and heart.^{13,14} ECMO has brought down the mortality of infants with CDH and secondary pulmonary hypertension from close to 90% to lower levels.

HFV is oxygen delivery by ventilation at rates of up to 2400 cycles per second. The volume used is less than the anatomical dead space and the gas exchange appears to occur by a diffusion process.

In places where such facilities are not available, it would be wiser to operate on the patient as soon as the patient is considered fit for surgery, after resuscitation. Generally, a subcostal approach is made on the affected side. Retraction of the rib cage exposes the diaphragm. The hernia is reduced gently by traction. If there is a sac, it is resected. The collapsed lung is not forcibly expanded. A chest tube is inserted and sutured into position. The anterior and posterior rims of the defect are identified. The defect is sutured with 2-0 or 3-0 Prolene mattress sutures or Prolene mesh. Following repair, abdomen is closed with no attempt made at this juncture to correct malrotation. A transthoracic approach may also be carried out especially for right-sided hernias.^{1,2}

Intensive postoperative care is needed in such patients. Ventilator support is instituted. In patients who have had surgery under ECMO, this facility is continued until the infants can safely be weaned. Weaning from ventilator should be meticulous and slow.

A few words about foetal surgery. Foetal surgery can be either tracheal ligation (Hendricks, et al) or repair of hernia in the foetus (Harrison, et al). Tracheal ligation technique is also called PLUG (plug the lung until it grows). At term, tracheal occlusion is removed by the technique known as EXIT (Ex Utero intrapartum tracheoplasty).¹⁵ EXIT is done when the baby is being delivered through a caesarean section. However, the several complications encountered during surgery and during EXIT result in limited survival rate.

It is interesting to note that, the prognosis of CDH, apart from age of onset and respiratory complications, also depends heavily on its characteristics, i.e. side and type, contents of hernia as well as presence or absence of a true

hernial sac. Further, short-term complications in CDH has remained a less studied topic. The association of CNS anomalies with a small, but fixed number of CDH cases has been known for some time. Hence, this study is intended to fulfil the following objectives.

Aims and Objectives

1. To find out the association between characteristics of CDH (side, contents, presence of sac, morgagnian hernia) and prognosis.
2. To study the short-term complications due to CDH. (Complications which are considered are wound infection, pneumothorax, adhesive obstruction, recurrent RTI and recurrence of hernia. The cases are followed up for 6 months).
3. To assess the types of CNS anomalies among CDH patients.

MATERIALS AND METHODS

Study Design- Retrospective study- Descriptive, based on case records.

Study Setting- Department of Paediatric Surgery, SAT Hospital, Trivandrum.

Study Subjects- All inpatients admitted with CDH during the study period in Paediatric Surgery Department, SAT Hospital, Trivandrum.

Study Period- August 2004 to July 2009.

Inclusion Criteria

1. All patients admitted in Paediatric Surgery Units, SAT, Trivandrum, with CDH during the study period.
2. All patients readmitted in Paediatric Surgery Units, SAT, Trivandrum, following complications after surgery.
3. All patients below 12 years are considered in paediatric age group.

Exclusion Criteria

1. Patients admitted in Paediatric Medicine.
2. Patients who lost follow up.

Sampling

No definite sampling method has been adopted for the study. Consecutive cases of CDH have been recruited.

$$N = \frac{Z\alpha^2PQ}{L^2}$$

$\alpha = 1.96$

P is the proportion of successful surgeries, which is taken as 65% under Indian setting. Success rate in Western settings maybe as high as 70-90%.

Q= 100-P.

L= 20% of P.

Substituting values, N=52. However, 56 cases have been taken for this study.

Data Collection

1. Data retrieved from operative registry.
2. Data retrieved from case sheets in records library.

3. Data regarding complications are collected from case sheets.

Study Variable- The study variable (outcome) is the number of successful surgeries undertaken by Paediatric Surgery Department, SAT Hospital, Trivandrum, for CDH in specified age group and based on specific characteristic of hernia, which is expressed in percentage. The term prognosis represents outcome variable in the study.

The prognosis in the study is based on the end result after undergoing surgery for CDH. Those patients who are discharged alive are considered as successful cases and others as failures. The end result is expressed as percentage of successful surgeries in each age group and also based on characteristics of hernia.

Hernia characteristics is the main explanatory variable in the study.

Association of CDH with CNS anomalies was also studied.

Hernia characteristics like presence or absence of stomach as content, presence or absence of sac, side of hernia and morgagnian hernia are also taken for study.

Other variables, which can affect the outcome of study are presence and severity of respiratory tract infections, haematemesis, jaundice, antenatal events like polyhydramnios, respiratory arrest before referral, muscle tone, etc.

Statistical Analysis

Statistical analysis is done by Chi-square test, proportion and percentage analysis. Correlation between explanatory variable and outcome variable is assessed and expressed in Chi-square statistics. Data were analysed using computer software, Statistical Package for Social Sciences (SPSS) version 10. Data are expressed in its frequency and percentage as well as its mean and standard deviation. To elucidate the associations and comparisons between different parameters, Chi-square test was used as nonparametric test. For all statistical evaluations, a two-tailed probability of value, <0.05 was considered significant.

RESULTS

Sac	Count	Percentage
No	41	73.2
Yes	15	26.8

Table 1. Percentage Distribution of Sample According to Presence of Sac

Side	Count	Percentage
Left	45	80.4
Right	11	19.6

Table 2. Percentage Distribution of the Sample According to Side

Stomach Inside the Hernial Sac	Count	%
No	38	67.9
Yes	18	32.1

Table 3. Percentage Distribution of the Sample According to Stomach Inside the Hernial Sac

Postoperative Complications	Count	Percent
Adhesive obstruction	3	5.4
Recurrent RTI	5	8.9
Unrelated a/c scrotum	1	1.8
Wound infection	2	3.6
Pneumothorax	2	3.6

Table 4. Percentage Distribution of the Sample According to Postoperative Complications

Morgagnian Hernia	Count	Percent
No	52	92.9
Yes	4	7.1

Table 5. Percentage Distribution of the Morgagnian Hernia

Side	Death		Relieved		x ²	P
	Count	Percent	Count	Percent		
Left	14	31.1	31	68.9	2.11	0.146
Right	6	54.5	5	45.5		

Table 6. Association of Outcome and Side

Sac	Death		Relieved		x ²	P
	Count	Percent	Count	Percent		
No	12	29.3	29	70.7	2.77	0.096
Yes	8	53.3	7	46.7		

Table 7. Association of Outcome and Sac

Stomach Inside the Hernia Sac	Death		Relieved		x ²	P
	Count	Percent	Count	Percent		
No	10	26.3	28	73.7	4.55	0.033
Yes	10	55.6	8	44.4		

Table 8. Association of Outcome and Stomach Inside the Hernia Sac

Morgagnian Hernia	Death		Relieved		x ²	P
	Count	Percent	Count	Percent		
No	20	38.5	32	61.5	2.39	0.122
Yes	0	0	4	100		

Table 9. Association of Outcome and Morgagnian Hernia

CNS Anomaly	Count	Percentage
No	56	100%
Yes	0	0%

Table 10. Percentage Distribution of Sample According to CNS Anomalies

DISCUSSION

56 cases were admitted in paediatric surgical wards with a suspected diagnosis of CDH during the time period from August 2004 to July 2009. Of these, there were 25 in-house and 31 referred cases. Three patients had history of respiratory arrest prior to referral and none of them could be adequately resuscitated and expired soon after admission. Surgery was performed in 48 out of 56 children. Others could not be adequately resuscitated.

The result of this study could be analysed considering various studies published before on CDH.

At surgery, 45 (80.3%) out of 56 had left-sided defect, whereas, in the Mayo series, the value was still higher at 90.7%.¹⁶ Outcome was bad for right-sided hernias (death rate 54.5% vs. 31.1%) as in study conducted by Touloukian and Gibson.^{17,18}

Sac was seen in 15 (26.7%) cases and the survival was poor in these cases (46.7%).

The main content in the sac was colon followed closely by small intestine (as in Mayo series).

The presence of stomach as a content of hernial sac lead to poor prognosis. This result was found to be statistically significant (P value 0.033). Stomach accounted for 32.2% in our study vs. 24.7% in the Mayo series. In both the studies, prognosis turned out to be poor when stomach was one of the contents.¹⁸

4 out of 56 (7.1%) had herniation through the foramen of Morgagni, while the rest were Bochdalek hernias. The prevalence of morgagnian hernia was a litter higher in our study (7.1% vs. 4.3%).

Short-term complications other than postoperative death were few with 5 cases of recurrent RTI, 3 cases of adhesive intestinal obstruction, 2 cases of wound infection, 2 cases of pneumothorax and a case of unrelated acute scrotum. The complication rates in our study were comparable to the study conducted by Bhat, Kumar and Rao, Department of Paediatric Surgery, Manipal, except in that, the number of recurrences we had were nil compared to their score of 4 out of 50 cases.^{19,20} This may be due to the higher number of more complicated cases (and hence the use of artificial prosthesis), which is being done in that institute.

Unable to assess the types of CNS anomalies associated with CDH in this series since none of the cases had coexisting CNS anomaly. It may be due to the greater number of stillbirths in fetuses associated with CNS anomalies during the given study period.

CONCLUSION

1. CDH is more commonly seen on the left side.
2. Presence of stomach in the sac signifies a poorer prognosis ($p < 0.05$).
3. Sac was seen in 15 (26.7%) cases and the survival was poor in these cases (46.7%).¹²
4. Mortality of right-sided hernias exceeded that of left-sided defects, which is against the commonly held belief (our study is supported by Touloukian-Gibson report and Skari et al study).^{18,19,20}
5. None of the cases had coexisting CNS anomalies, which may not be statistically significant. It may be due to the greater number of stillbirths in fetuses associated with CNS anomalies (which were not included) in our study group during the given study period.
6. Surgically-treated patients do well, if they survive the operation.
7. Surgical complications are few and mainly due to respiratory infection and adhesive obstruction.

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