PREVALENCE AND DISTRIBUTION OF CONGENITAL DIAPHRAGMATIC HERNIA WITH SPECIAL REFERENCE TO TIME OF ONSET OF SYMPTOMS AND PROGNOSIS-A RETROSPECTIVE STUDY FROM A NEUROSURGICAL PERSPECTIVE

Anand Prabhavathy Raghavan[1], Gejoe George[2]

[1]Assistant Professor, Department of Neurosurgery, Government Medical College, Thrissur.
[2]Additional Professor, Department of General Surgery, Government Medical College, Parippally.

ABSTRACT

BACKGROUND

Congenital diaphragmatic hernia (hereby referred to as CDH) is a relatively rare anomaly with a prevalence of 1 in 3000 livebirths. Time of onset of symptoms has much significance as far as the prognosis of CDH is concerned. CNS anomalies co-exist in up to 10% of non-syndromic CDH cases; hence, the diagnosis, treatment and prognosis of CHD is of interest to paediatric neurosurgeons also.

MATERIALS AND METHODS

Retrospective study-descriptive, based on case records. Study subjects were all in patients 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

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.

CONCLUSION

The prevalence of CDH in our institute is around 1 in 2288, which is slightly higher than the prevalence worldwide. CDH is more commonly seen in male population. Younger the age of onset of symptoms (<24hrs.), worse the prognosis ($p<0.05$).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 foetuses associated with CNS anomalies (which were not included), in our study group, during the given study period.

KEYWORDS

Congenital Diaphragmatic Hernia, Prevalence and Distribution, CNS Anomalies, Age of Onset and Prognosis.

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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]

CDH has been described as early as 1679 by Bonnet. In 1848, Bochdalek described CDH as occurring through a posterolateral defect, which has from that time borne his name.[1,3] 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).[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]

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

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Corresponding Author:
Dr. Anand Prabhavathy Raghavan,
No. 8/609, Athira, P.O. Kalapathy,
Palakkad- 678003.
E-mail: anandpraghavan@gmail.com
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(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 leads 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.[4]

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.[2] 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 schisis-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.[4]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.[4]
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.[4,12]

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 2400cycles per second. The volume used is less than the anatomical dead pace 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 ribcage 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]

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 (Hendrick 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 tracheloplasty).[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.

CDH is a relatively rare entity. "Earlier the symptom, early the diagnosis. Earlier the diagnosis, better the prognosis." This dictum holds true in most medical conditions, except CDH. This information is vital in the treatment and prognostication of CDH and hence this study.

Aims and Objectives

1. To assess the prevalence of CDH in a tertiary care institute in the state of Kerala.
2. To find out the association between age of onset of symptoms and prognosis in CDH.

3. To assess the prevalence of CNS anomalies among CDH patients.

Study Design- Retrospective study descriptive based on case records.
Study Setting- Department of Paediatric Surgery, SAT Hospital, Trivandrum.
Study Subjects- All in patients 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 12years 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.

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

$\alpha = 1.96$

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

Age is the main explanatory variable in the study.

Association of CDH with CNS anomalies was also studied.

Age groups of <6hrs., >6hrs. to <24hrs., >24hrs. to <1 month,>1month to <1year and>1year are taken for the study. Almost, 50% of study subjects come in the first group.

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 non-parametric test. For all statistical evaluations, a two-tailed probability of value, <0.05 was considered significant.

RESULTS

Year	Birth	IBN	
		Count	%
August 2004-July 2005	12,500	4	0.032
August 2005-July 2006	12,000	4	0.033
August 2006-July 2007	12,000	4	0.033
August 2007-July 2008	11,000	6	0.055
August 2008-July 2009	9,700	7	0.072
Total	**57,200**	**25**	**0.044**

Table 1. Livebirths in IBN During 2005-2009

1 in 2,288 births.

Age of Onset	Count	Percentage
<6hrs.	12	21.4
6-24hrs.	12	21.4
24hrs. to <1 month	21	37.5
1 month to 1year	8	14.3
>1year	3	5.4

Table 2. Percentage Distribution of the Sample According to Age of Onset

Sex	Count	Percentage
Female	17	30.4
Male	39	69.6

Table 3. Percentage Distribution of Sample According to Sex

Year	Count	Percentage
August 2004-July 2005	11	19.6
August 2005-July 2006	10	17.9
August 2006-July 2007	11	19.6
August 2007-July 2008	12	21.4
August 2008-July 2009	12	21.4

Table 4. Percentage Distribution of Sample According to Year

Prenatal Diagnosis	Count	Percentage
No	34	60.7
Yes	22	39.3

Table 5. Percentage Distribution of Sample According to Prenatal Diagnosis

Born	Count	Percentage
IBN	25	44.6
OBN	21	37.5
Others	10	17.9

Table 6. Percentage Distribution of Sample According to Birth

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

Table 7. Percentage Distribution of Sample According to CNS Anomalies

Type of Delivery	Count	Percentage
Normal	46	82.1
Cesarean section	10	17.9

Table 8. Percentage Distribution of the Sample According to Type of Delivery

Outcome	Count	Percentage
Preop death	8	14.3
Postop death	12	21.4
Relieved	36	64.3

Table 9. Percentage Distribution of the Sample According to Outcome

Age	Death		Relieved		X^2	P
	Count	%	Count	%		
<24 hrs.	12	50	12	50	3.73	0.049
>24 hrs.	8	25	24	75		

Table 10. Association of Outcome and Age of Onset

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.

Total number of births in the institution during the period August 2004-July 2005, August 2005-July 2006, August 2006-July 2007, August 2007-July 2008 and August 2008-July 2009 were 12500, 12000, 12000, 11000, 9700, respectively. Prevalence of CDH in the institution is 1 in 2288.

There were 39 (69.6%) male and 17(30.4%) female patients. Age of presentation were 12 patients less than 6hrs.; 12 patients >6 hrs. and <24 hrs.; 21 patients >24 hrs. and <1 month; 8 patients >1 month and <1 year; and 3 patients >1 year. Mortality rate was 50% for patients who presented before 24 hrs. after birth and 25% for those who presented later. The result was found to be statistically significant. The overall survival rate is 64.3%.

29 (39.3%) out of the 56 were prenatally diagnosed to have CDH. The radiologist and USG showed an accuracy of 100%.

CNS anomalies usually coexist in upto 10% of non-syndromic CDH cases (David and Illingworth, 1976; Dillon et al, 2000; Dott et al, 2003).In our study, none of the cases had coexisting CNS anomalies.

Hernia and mediastinal shift were seen in all patients who were radiographed.

Surgery was performed in 48 out of 56 children. Others could not be adequately resuscitated.

Closure of the defect was done with 3-0 Prolene or 2-0 silk. Prosthesis were not used in any of the cases.

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

According to the study conducted by Barbara Pober, Mehan Russel and Kate Ackerman of Department of Paediatric Surgery, Mass General Hospital, Boston, (the study followed up 76 cases of CDH from presentation to 5 years after surgery) the prevalence was 1 in 3200.[2]The prevalence in this study is a little higher at 1 in 2288, which comes within the limit expected for CDH. The higher prevalence may also be due to the tertiary status of the institution, racial differences and the large population it drains.

Male-to-female ratio (65.8% and 34.2% vs. 69.6% and 30.4% in our study) was comparable in both studies.

Percentage of cases, which presented before 24 hrs. was 69.2% (Boston) vs. 43% (our study).[2] The higher number of cases in Boston study may be attributed to the better antenatal care in that country. The survival rate for these children were 69.4% vs. 50% (our study) respectively showing the efficiency of sophisticated antenatal, postnatal and postoperative care available in developed economies. Overall survival rate is 85.2% vs. 64.3% (our study). Prenatal diagnosis was 78.9% vs. 49.6% (our study).

Association of age of onset with outcome shows poorer prognosis (50% vs. 75% with P value of 0.049) in patients presenting <24 hrs. after birth. Results are comparable to studies conducted by Akber Sheriff et al and Jain et al.[3,4]

CNS anomalies co-exist in upto 10% of non-syndromic CDH cases (David and Illingworth, 1976; Dillon et al, 2000; Dott et al, 2003).[7,8,9] In our study, 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 foetuses associated with CNS anomalies (which were not included) in our study group during the given study period.

CONCLUSION

1. The prevalence of CDH in our institute is around 1 in 2288, which is slightly higher than the prevalence worldwide. This may be due to the tertiary care status of the institution, larger drainage area as well as racial and social differences.

2. CDH is more commonly seen in male population.
3. Younger the age of onset of symptoms (<24hrs.), worse the prognosis ($p<0.05$).
4. 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 foetuses associated with CNS anomalies (which were not included), in our study group, during the given study period.
5. Surgically treated patients do well if they survive the operation.

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