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CONGENITAL DIAPHRAGMATIC HERNIA: OUR INSTITUTIONAL EXPERIENCE

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ABSTRACT **Background:** Congenital diaphragmatic hernia (CDH) is a developmental defect, in which there is herniation of abdominal contents into the thoracic cavity. This causes pulmonary hypoplasia and pulmonary hypertension to varying degrees. The clinical profile of CDH neonates at a tertiary care institute were studied over a period of 22 months between October 2018 to June 2020.

Aims and objectives: To study the demography, including Age, Sex, birth weight, types, associated anomalies, surgery, complications and survival in CDH patients at a tertiary care referral centre in Rajasthan, India.

Materials and methods: This was a prospective observational study between October 2018 and June 2020, over 1 year and 9 months, at tertiary referral centre in North-Western India. Clinical data, radiological findings, pre-operative conditions were studied, intra-operative findings were noted and post-operative course was studied. Intravenous Sildenafil was administered and their effects on clinical improvement was noted.

Results: 102 cases of CDH were admitted, of which 62 were male and 40 were female. Of these cases, 78 underwent surgical repair, while the rest died prior to surgery. Majority (99) were left sided CDH. And majority (59) were without sac. Contents which were commonly encountered included Stomach, small and large intestine, spleen, liver. Patients who were administered with intravenous sildenafil showed clinical improvement in the pulmonary hypertension. Patients with CDH require urgent management of the respiratory distress and pulmonary hypertension, in order to improve their survival.

KEYWORDS : Congenital Diaphragmatic Hernia, Respiratory distress, Neonates, Management, CDH

INTRODUCTION

Congenital Diaphragmatic Hernia, which is the herniation of abdominal contents into the thoracic cavity, usually presents with respiratory distress. The severity of the distress varies with the degree of the pulmonary hypoplasia and pulmonary hypertension that is associated with the Hernia.

The reported incidence of CDH is estimated to be between 1 in 2000 to 5000 births (1).

Successful repair of CDH remained rare until 1940, when Ladd and Gross reported 9 of 16 patients surviving operative repair, the youngest being 40 hours old (2).

It has been classified into:

(a) Bochdalek hernia in which there is posterolateral diaphragmatic defect;
(b) Morgagni hernia where the diaphragmatic defect is located anteromedially on either side of the septum transversum and the thoracic wall (1).

Until the 1980s, the standard of care remained immediate neonatal surgery followed by postoperative resuscitative therapy, however, CDH is a physiologic emergency and not a surgical emergency (1). It is presently accepted that surgery should be undertaken only after cardio-respiratory functions are stable (3).

Factors important in recognition and salvage of infants with CDH are - (a) Prompt diagnosis depends on early suspicion and chest x-ray. (b) Successful resuscitation and transport require gastrointestinal decompression with a nasogastric tube and skillful respiratory management. (c) avoidance of positive pressure ventilation without an endotracheal tube or too-vigorous positive ventilation through an endotracheal tube causing pneumothorax. (d) Survival after repair of the hernia from either above or below the diaphragm depends on adequate pulmonary function (4).

CDH may be diagnosed antenatally during maternal ultrasound, either showing evidence of herniation of bowel loops in the chest of the foetus, or as part of a differential diagnosis of Polyhydramnios, which occurs due to kinking of gastroesophageal junction may help to antenatal diagnosis in some severe cases (5).

Post-natally, the patients usually present with respiratory distress, cyanosis, or chest retraction.

The objectives of this study were to explore the clinical characteristics, including age, sex, birth weight, types, surgery, complications and survival in CDH patients in a tertiary care centre in Rajasthan, India.

MATERIALS AND METHODS

This was a prospective observational study, conducted between October 2018 and June 2020, in the department of Paediatric Surgery, at a tertiary care centre in Rajasthan, India. The study included all diagnosed cases of Congenital Diaphragmatic Hernia, between the ages of 1 day and 5 months, that presented at our hospital. Patients presented either with respiratory distress, cyanosis or Chest retraction. Few patients were diagnosed antenatally. Clinically, most of the patients had a scaphoid abdomen, the heart-beat was displaced, bowel sounds were present in the chest. Diagnosis was confirmed by performing a chest X Ray which showed evidence of bowel loops in the chest. In cases in which there was a doubt about the diagnosis of CDH, an ultrasound of the chest and abdomen was done which showed evidence of bowel loops or stomach or liver in the chest.

On receiving the patient, temperature was noted and measures to ensure thermoregulation were taken. The patients were resuscitated. Oral and nasal suctioning were done.

Ambu mask ventilation was avoided as this results in worsening respiratory distress due to distension of the stomach.

A naso-gastric tube was inserted, contents were aspirated, and kept on free drainage. This was done for decompression of the stomach and the small bowel.

Blood sugar levels were checked and blood samples taken for investigations.

Intravenous fluids according to body weight and as per the age of presentation, were administered.

Prophylactic intravenous antibiotics were given.
In most of the patients, intravenous sildenafil was initiated to treat the pulmonary hypertension associated with CDH.

If there was severe respiratory distress, an endo tracheal tube was inserted, and the patient put on mechanical ventilation.

Packed cells were arranged prior to surgery.
After explaining the risk of surgery and obtaining a proper consent, the patient was posted for surgery.

The surgery was carried out under general anesthesia with the patient in supine position and the abdomen slightly turned to the opposite side by means of a bolster.

Nearly all patients underwent CDH repair through transabdominal route. One patient underwent transthoracic repair.

In the transabdominal route, the incision was a subcostal incision, providing adequate exposure. the herniated contents were inspected and reduced manually. The presence of sac was noted. After placing an intercostal drain, the defect in the diaphragm was repaired with non-absorbable polypropylene interrupted sutures, after identifying the anterior and posterior lips of the diaphragm. In most of the cases small bowel was present along with large bowel, stomach and occasionally spleen and liver. Patients who had associated malrotation of the gut underwent Ladd's procedure with appedicectomy additionally. Intercostal drain was placed in all the patients that underwent surgical repair.

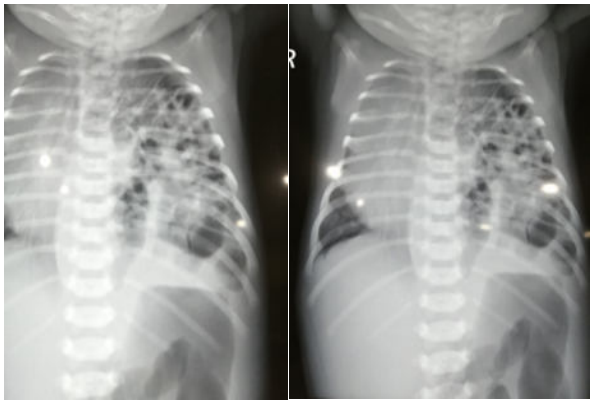

FIGURE 1 AND 2 – CHEST X RAY SHOWING AIR FLUID LEVELS IN THE LEFT HEMI-THORAX AND MEDIASTINAL SHIFT TO THE RIGHT SIDE – SUGGESTIVE OF LEFT CDH.

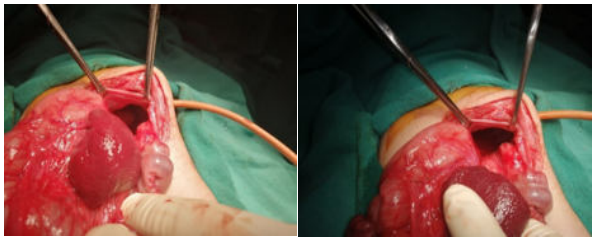

FIGURE 3 AND 4 – INTRA-OPERATIVE PHOTOGRAPH SHOWING THE DEFECT IN THE LEFT HEMI-DIAPHRAGM AND THE CONTENTS WHICH HAD HERNIATED INTO THE LEFT HEMI-THORAX – SMALL BOWEL, LARGE BOWEL AND SPLEEN.

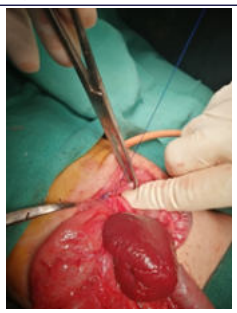

FIGURE 5 – INTRA-OPERATIVE PHOTOGRAPH DURING THE REPAIR OF THE DIAPHRAGMATIC DEFECT.

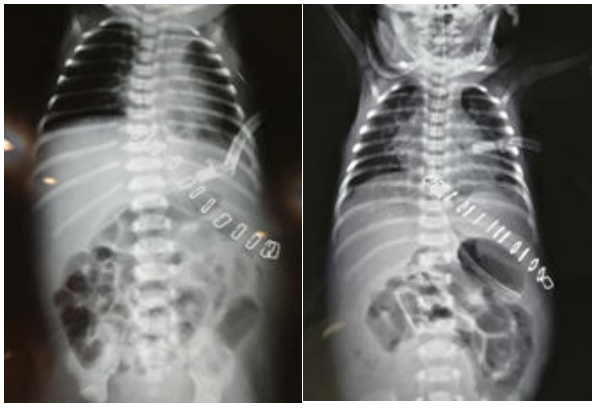

FIGURE 6 AND 7 – CHEST X-RAY POST OPERATIVELY SHOWING WELL EXPANDED LUNG ON THE LEFT SIDE.

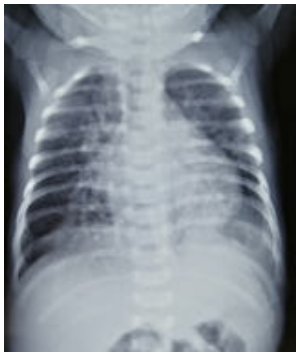

FIGURE 8 – CHEST X RAY TAKEN 2 WEEKS AFTER SURGERY SHOWING CLEAR WELL EXPANDED LUNG IN AN OPERATED CASE OF LEFT CDH.

RESULTS

There were 102 cases of Congenital Diaphragmatic hernia that presented between 1 day and 5 months, between October 2018 and June 2020.

Of the cases, 62 were male and 40 were female – 1.55:1.
The average age of presentation among neonates was 3.7 days.
41 out of 102 patients presented on live day one.
Average weight of the patients was 2.8 Kgs.

Majority of the patients had a left sided CDH – 97%, and 3% had right sided CDH.

Chest x ray was diagnostic in 98 (96%) patients. Ultrasound of the thorax was required in only 4 (4%) patients.

Intravenous sildenafil was administered in 76 of 102 cases, as a continuous infusion, majority of whom showed clinical improvement in the respiratory distress. 40 out of these 76 patients were discharged, 36 had expired. Out of 26 patients who did not receive intravenous sildenafil, 13 had been discharged, 13 had expired.

Out of the 102, 78 underwent surgical repair whereas 24 expired before surgery.

7 out of the 78 patients required pre-operative ventilatory support.

Repair was done through laparotomy in 77 out of 78 cases, whereas one case of Right CDH underwent repair through right thoracotomy. Sac was present in 19 out of the 78 operated cases (24.3%), whereas 59 out of the 78 cases (75.7%) had CDH without sac. The most common content was Small and large bowel and stomach - 53 out of 78 cases – 68%.

One patient had an associated Low anorectal malformation for which anoplasty was done.

Gastric perforation in the herniated stomach was present in one patient, which was noted at the time of surgical correction and successfully repaired.

An associated enteric duplication cyst was encountered in one of the cases and the cyst was excised.

Associated malrotation of the gut was seen in one patient, for which Ladd's procedure and appendicectomy was done along with the CDH repair.

One of the patients had an associated gastric volvulus, which was successfully de-rotated.

30 out of the 78 patients required post-operative mechanical ventilation.

Out of the 102 patients, 53 survived and 49 had expired with a survival of 52% and mortality rate of 48%.

Table 1 – Sex distribution of the patients

MALE	62
FEMALE	40
RATIO	1.55:1

Table 2 – Table showing distribution of the side of CDH

LEFT CDH	RIGHT CDH
99	3

Table 3 – Table showing requirement of Ventilatory support

PRE-OP VETILATORY SUPPORT	7
NO PRE-OP VENTILATORY SUPPORT	71

Table 4 – Number of patients operated and those that expired prior to surgery

OPERATED	EXPIRED BEFORE SURGERY
78	24

Table 5 – Presence of sac and associated survival noted

	SAC	NO SAC
	19 OF 78	59 OF 78
PERCENTAGE	24.3%	75.7%
SURVIVAL	16 (84%)	37 (62.7%)

Table 6 – Table showing various contents encountered

CONTENT	NUMBER OF CASES	PERCENTAGE
SMALL AND LARGE BOWEL, STOMACH	53	68%
SMALL AND LARGE BOWEL, SPLEEN	5	6.5%
SMALL AND LARGE BOWEL	16	20%
SMALL AND LARGE BOWEL, SPLEEN, LIVER	2	2.6%
SMALL AND LARGE BOWEL, LIVER	2	2.6%

Table 7 – Administration of intravenous sildenafil

	NUMBER OF CASES	PERCENTAGE	SURVIVAL	PERCENTAGE
INTRAVENOUS SILDENAFIL GIVEN	76	74.5%	40	52.6%
INTRAVENOUS SILDENAFIL NOT GIVEN	26	25.5%	13	50%

DISCUSSION

This study was a prospective observational study.

Earlier, CDH was considered to be a surgical emergency, with a focus on operating the CDH patients, rather than the management of the associated pulmonary hypertension and lung hypoplasia. However, the initial stabilization of the patient is extremely important prior to surgery.

The male to female ratio in this study was 1.55:1 which is comparable to a study by Crankson et al which had a ratio of 2:1 (6), which is in turn similar to the ratio found by Baglaj (7) in a review.

Left sided CDH was more common 96%, which is in accordance with those reported (1,7, 8).

CDH usually presents with respiratory distress after birth (6), which was the commonest presentation encountered in this study. Respiratory distress was the commonest presenting complaint – 95% of the cases, as is usually the case in CDH (6).

Even the patients presenting late had respiratory distress as the common complaint, which was also observed in study by Kitano et al (9).

Chest X-rays were used to diagnose majority of the patients. In those with a doubt in diagnosis, ultrasound of the chest confirmed the diagnosis of CDH, which has been described in literature as other modalities that can be used to diagnose this condition (10).

The patients presenting early had more severe respiratory distress and contributed to higher mortality than those that presented late – average age among non-survivors was 1.3 days, 6.1 among non-survivors in the neonatal age group. This is in accordance with the results observed by Aihole JS et al (11).

Small and large bowel were present in nearly all cases of CDH. Stomach was present in 53 (68%) cases. The position of the stomach is considered to be one of the prognostic factors, with studies showing survival of 30% when stomach had herniated into the thorax.

Various other studies did not show any predictive value (1, 12, 13). In our study, out of the 53 patients with stomach herniating into the thorax, 34 survived (64%). Hence, there was no correlation between position of the stomach and mortality, as opposed to other studies (13, 14). Right sided CDH was only found in 3 patients, of whom 2 survived (67%), one (23%) did not survive. 51 out of 99 (51%) Left sided CDH patients survived, whereas 48 did not (49%).

Certain studies show that right sided CDH patients have a higher mortality rate than left sided CDH patients (13, 15, 16). However, in our study, survival rate of right sided CDH patients was found to be higher compared to left sided CDH patients.

The presence of sac is considered to be a good prognostic indicator.

Sac was present in 19 out of 78 patients who underwent surgery (24.3%), out of which 16 survived (84%). 59 out of 78 patients who were operated had no sac. Of these, 37 survived (62.7%). This higher survival rate of CDH with sac has been observed in literature (13, 16).

Intravenous sildenafil was administered as continuous infusion in 76 patients, out of whom 40 (52.6%) survived. 26 patients did not receive sildenafil, out of whom 13 (50%) survived.

Clinical improvement in respiratory distress was noted in the patients who received sildenafil and is usually associated with a better outcome (12, 17).

Overall survival in this study was 52%, while mortality was 48%. In recent studies, the survival rate of CDH patients has been found to be between 21% and 83% (18, 19). According to a study by Wilkinson D et al in 2009 (20), the worldwide mortality rate was noted to be between 20% and 60%. A large number of patients die before the referral to a tertiary centre. Thus, the number of patients reaching tertiary care centres, may represent only 40-50% of the total number of CDH patients. This reflects the “hidden mortality” as described by Harrison et al (4).

A significant proportion of the patients have to travel a great distance

before they can reach tertiary care centres, such as is the case with our institute. Hence, the condition of the patients during travel often worsens owing to the need of NICU care that they require. This may also result in the significant number of patients presenting with respiratory distress and their high mortality rate encountered.

High frequency oscillatory ventilation (HFOV) has been used in many centres before extracorporeal membrane oxygenation (ECMO) as a "rescue therapy" to reduce barotrauma (20, 21). However, at our centre, HFOV and ECMO are not available.

In recent times, it has been shown that CDH is a medical emergency rather than a surgical emergency, with appropriate management of the pulmonary hypertension and medical therapy being essential to stabilize the patient, prior to surgery (5, 22).

CONCLUSION

Majority of the CDH patients present with respiratory distress. A significant proportion of the patients die before they reach tertiary care centres. Those who present early have a poorer prognosis than those that present late. Left sided CDH is much more common than right sided CDH. CDH with sac has a better prognosis than CDH without sac.

Patients with CDH require urgent management of their respiratory distress and need to be adequately stabilized prior to surgical correction.

Conflicts of interest

None

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