Original Paper

CONGENITAL DIAPHRAGMATIC HERNIA IN NEONATE EXPERIENCE IN A TEACHING HOSPITAL

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Abstract

Introduction: **Congenital diaphragmatic hernia (CDH) is one of the major surgical causes of respiratory distress in neonates. Reported survival averages 60% but may be significantly lower. Pulmonary hypertension and pulmonary hypoplasia are recognised as two corner stones of the pathophysiology of CDH.**

Objective: **Objective of the study was to evaluate the outcome of this birth defect in Bangladesh situation.**

Method: **This retrospective study was carried out at the Department of Paediatric Surgery of Combined Military Hospital, Dhaka over a period of five years. During this period a total of 8 neonates of CDH were admitted in this hospital. All the data were collected from record sheet and were compiled.**

Result: **Age of patients ranged from 1 day to 20 days. Out of 8 neonates 5 (62.50%) were male and 3 (37.50%) were female. All patients were diagnosed postnatally. All the 8 neonates had Bochdalek type of CDH. Seven patients (87.50%) had left sided hernia. Two patients (25%) died before operation in the stabilization phase while on ventilator and 6 (75%) were operated. Out of these 6 patients, 5 (left sided) were operated through abdominal route and 1 (right sided) was approached through thorax. Overall outcome was satisfactory in 5 neonates and one died.**

Conclusion: **Early intervention can result good prognosis in CDH.**

Key words: **Congenital diaphragmatic hernia, neonate, Bochdalek type**

Introduction

A congenital diaphragmatic hernia (CDH) is a birth defect or abnormality that occurs before birth as a foetus is forming in the mother's uterus[1]. It should be diagnosed in prenatal period and promptly referred to a tertiary centre for imaging, genetic testing and multidisciplinary counselling. It is one of the major surgical causes of respiratory distress in neonates. CDH occurs in approximately 1 in every 2000-4000 babies and accounts for 8% of all major congenital anomalies[2-4]. The diagnosis of CDH is often made on a prenatal ultrasound (USG) examination and is accurate in 40% to 90% of cases[5]. The 3 basic types of CDH are the posterolateral Bochdalek hernia, the anterior Morgagni hernia and the hiatus hernia[6]. Affected neonates are born with a complex interface of pulmonary hypoplasia and pulmonary hypertension[7]. The incidence of associated anomalies in neonates with CDH ranges from 10% to 50% and the final outcome depends much on it[8,9].

The left sided Bochdalek hernia having a male preponderance (1.5:1) occurs in approximately 80-90% of cases[6]. Morgagni hernia are rare and usually not diagnosed in neonatal period. The emphasis in postnatal management has shifted from the neonatal surgical emergency approach to a delayed procedure designed to deal with pulmonary hypoplasia and the pulmonary vascular abnormalities[10,11]. Despite advances in postnatal care, patients with CDH suffer substantial morbidity and mortality[12]. Reported survival averages 60% but may be significantly lower. Neonates with CDH are critically ill, but can expect better survival if other serious anomalies are not present, however, significant morbidities can exist in the survivors[13-15]. Young CDH survivors continue to have ongoing medical problems and a high incidence of motor and language problems[16]. Neonates with CDH show a wide range of anatomic and physiologic abnormalities, making it difficult to compare the efficacy of management protocol between institutions[8].

This study was done to share experience with CDH in neonate and the early results of operative treatment.

Material and Method

This retrospective study was carried out at the Department of Paediatric Surgery, Combined Military Hospital, Dhaka from July 2004 to June 2009. During this study period a total of 8 neonates of CDH were admitted in the mentioned hospital. Particulars of all those patients were recorded which included age, sex, weight etc. Detailed history was taken and thorough clinical examination done in all the patients. Relevant investigations (plain x-ray,

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contrast x-ray, USG, blood chemistry, arterial blood gas etc) were done as required (Fig1 & 2). All the patients with CDH were operated by open method after initial stabilization. Postoperatively the patients were followed up at the end of 1st month and 3rd month. Relevant clinical information, investigation results, operative procedures, post operative sequelae and follow up results were documented.

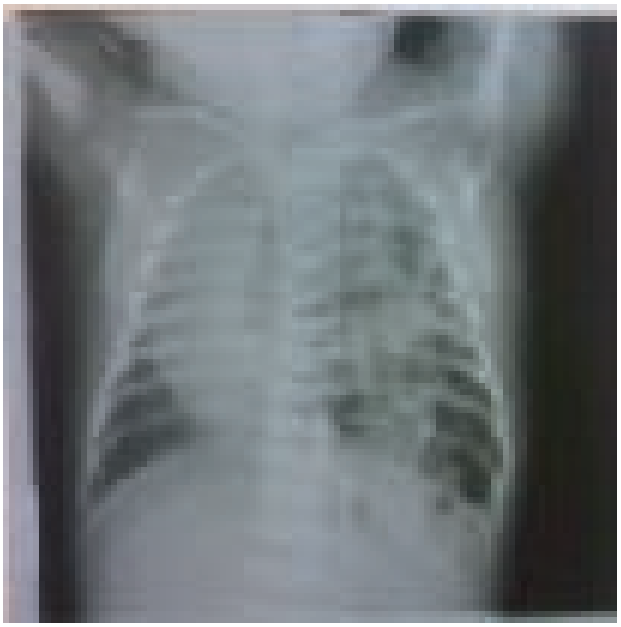

Fig-1: Plain x-ray chest showing left sided Bochdalek hernia

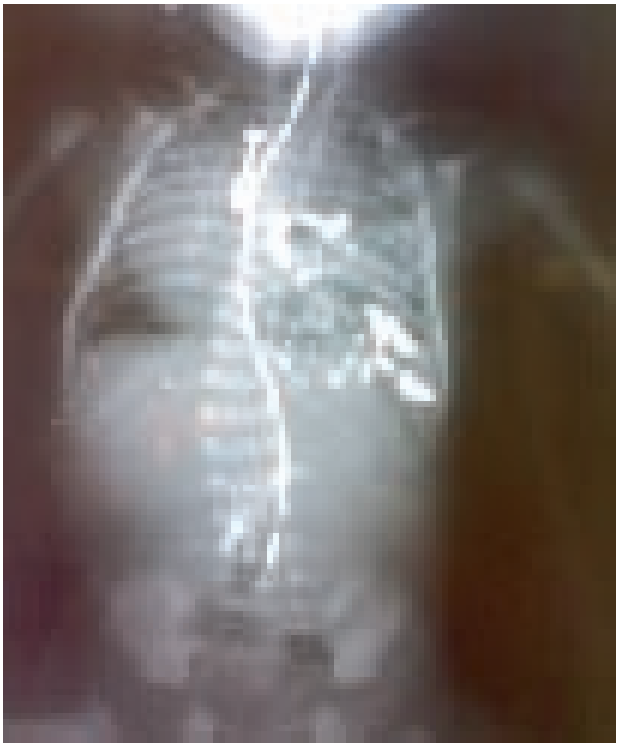

Fig-2: Contrast x-ray of upper GITshowing left sided Bochdalek hernia

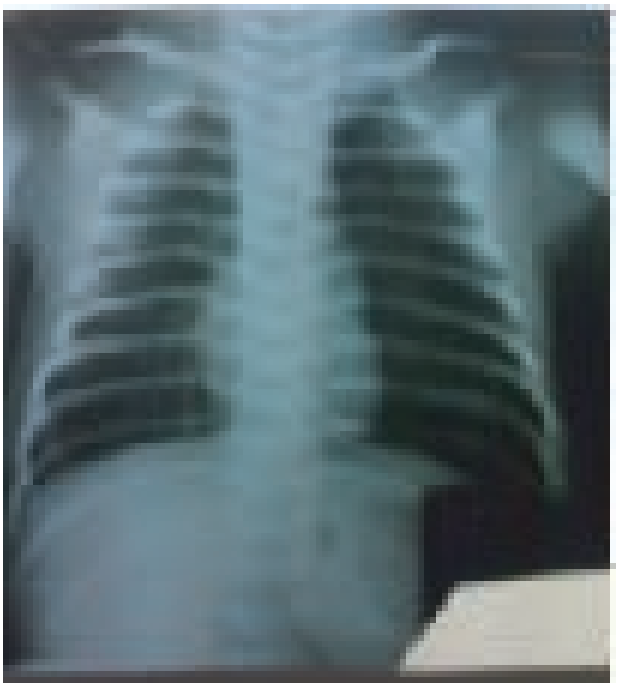

Fig-3: Plain x-ray chest of a case of left sided Bochdalek hernia after surgery

Result

Out of total 8 neonates of CDH 6 neonates (75%) had age less than 1 week and 2 (25%) had 1 week to 1 month. Age ranged from 1 day to 20 days. Five neonates (62.50%) were male and 3 (37.50%) were female. All the 8 neonates had Bochdalek type of CDH and were diagnosed postnatally. Seven patients (87.50%) had left sided hernia and 1 (12.50%) had right sided lesion. Two neonates (25%) died before operation in the stabilization phase while on ventilator and 6 (75%) were operated. Out of these 6 patients, 5 (left sided) were operated through abdominal route and 1 (right sided) was approached through thorax. Synthetic mesh was not required in any of the cases. One patient died on 2nd post operative day. All other five recovered satisfactorily and on regular follow up.

Discussion

CDH is a relatively common birth defect. Fetuses with CDH who have a "poor prognosis" with postnatal treatment now can be identified early (before 25 weeks' gestation) on the basis of liver herniation, and a low lung to head ratio (LHR)[17]. The use of a protcolized management that included antenatal assessment, antenatal steroid, planned delivery, prophylactic surfactant, pressure limited gentle ventilation, permissive hypercarbia and hypoxia and extracorporeal membrane oxygenation (ECMO), if indicated, has improved the outcome in high risk population[18].

In this study, all the 8 neonates had Bochdalek type of CDH. Incidence of Bochdalek hernia ranges from 90-95% among all CDH and mostly this type present in neonatal period[19]. None of them had antenatal diagnosis although antenatal diagnosis is possible in 40-90% of the cases[5]. This may be because of the fact that maternal USG is not routinely and meticulously done throughout the country. Out of 8 neonates with CDH, 6 (75%) patients presented to hospital in 1st week of their life. Neonates most commonly present with the stigmata of cyanosis and respiratory distress in the first few days of life, although a later presentation is possible[2,5,7]. Among 8 neonates, 5 were male and 3 were female making a male to female ratio of 1.7:1. Reported male-female ratio is 1.5:16. Seven neonates (87.50%) had left sided Bochdalek hernia. Left sided hernias are more commonly (80-90%) observed type reported in various studies[3,5,6]. Associated anomalies in neonates with CDH is 10-50%[8,9]. But in this series, none of the 6 operated patients had associated anomalies. The 2 neonates that died before operation might have hidden anomalies. Autopsy was not done. All patients were operated after initial resuscitation and stabilization. ECMO, high frequency oscillatory ventilation, dedicated neonatal surgical intensive care unit (ICU) were not available in the centre. Outcome was satisfactory in 5 cases. Survival rate of patients with CDH is reported 60-80%[2,6,19]. CDH may result in severe respiratory insufficiency with a high morbidity[20]. A poor prognosis, however, is associated with polyhydramnios, presence of foetal stomach in the chest and an early presentation (i.e., distress in the first few hours of life).

Conclusion

CDH represents a treatable cause of perinatal morbidity and mortality. Outcome of treatment often depends on associated anomalies. Significant long-term morbidities like developmental delay, poor growth, gastro oesophageal reflux disease, hearing loss and musculoskeletal abnormalities can exist in the survivors. As such they require regular follow-up for long time.

Refernces

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