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Congenital diaphragmatic eventration in children: 12 years' experience with 177 cases in a single institution☆

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ABSTRACT

Objective: This study sought to summarize the diagnostic and treatment aspects of congenital diaphragmatic eventration (CDE) in children by retrospectively analyzing their medical records to identify and understand the complications of CDE, its treatment, and to evaluate the long-term outcomes of diaphragmatic plication.
Methods: The medical records of children who received treatment for CDE from January 2000 to December 2011 at the Children's Hospital of Chongqing Medical University, China were analyzed. Data analyzed included the following: age, sex, symptom, location of eventration, associated anomalies, surgical procedures, complications, and survival and follow up details after diaphragmatic plication.
Results: The medical records of 177 children (boys: 128, girls: 49, mean age: 10.28 ± 2.35 months) with CDE were included in this study. Specific symptoms of eventration of the diaphragm were reported for 86 cases; and the typical symptoms included rapid breathing, vomiting, and recurrent respiratory infections. Except for a bilateral case, all the other patients had unilateral CDE. Associated malformations were observed in 31 cases (17.5%), hypoplastic lung (10 cases) was the most common followed by congenital heart disease (9 cases), and cryptorchidism (3 cases). Interestingly, 91 patients were asymptomatic. Diaphragmatic plication was performed in all symptomatic patients (86 cases, 48.5%) and none had recurrence.
Conclusions: Clinical symptoms of CDE varied in severity, ranging from asymptomatic conditions to life-threatening respiratory distress. Timely accurate diagnosis and treatment of symptomatic CDE could effectively resolve respiratory morbidity and reduce complications. The diaphragm plication surgery provided good results among the study population with no recurrence.

Eventration of the diaphragm was first described by Jean Louis Petit in 1774 [1]. The reported incidence is 1 per 1400 patients who had chest radiographs and it was more common in males than females [2]. Clinical manifestations of congenital diaphragmatic eventration (CDE) in infants and children are diverse, ranging from asymptomatic conditions to mild gastrointestinal disorders such as nausea and vomiting to sometimes life-threatening respiratory distress [3]. The CDE may easily be missed or misdiagnosed. Early diagnosis and repair of the diaphragm can prevent gastrointestinal disorders, reduce recurrent respiratory tract infections, and improve quality of life [4,5]. The purpose of this study was to summarize the clinical manifestations, physical examination findings, and radiological features of children with CDE in order to identify the opportunities to improve the diagnostic and treatment aspects and to evaluate the long-term outcomes of diaphragmatic plication.

1. Patients and methods

1.1. Study design

This retrospective analysis involved the review of medical records of children who received treatments for CDE from January 2000 to December 2011 at the Children's Hospital of Chongqing Medical University (Department of Cardiothoracic Surgery), the largest child-care hospitals in southwestern China.

The diagnosis of CDE was established in all cases by routine chest radiographs and fluoroscopy. Patients were excluded from the study if eventration was related to phrenic nerve injury at delivery or hypothermia for cardiac surgery. Initial treatment of patients included upright positioning, supplemental oxygen therapy, treatment of pneumonia if present, and nutritional support in symptomatic children.

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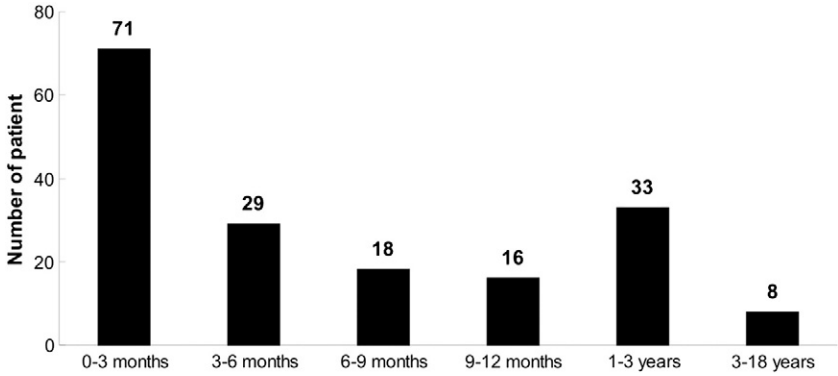

Fig. 1. Age distribution of children at the time of diagnosis of congenital diaphragmatic eventration.

Patients who failed to respond to conservative treatment were subjected to surgery. Diaphragmatic plication was performed through a posterolateral thoracotomy using a contralateral single lung ventilation in right CDE, and through an abdominal approach in left CDE. The hemidiaphragm was transected approximately 5 cm initially to avoid intra-abdominal organ injury and then plicated from medial to lateral with a series of six to eight parallel U sutures (2-0 polypropylene) until it became taut and flat.

The medical history, physical examination findings, chest X-ray, flouroscopy or ultrasonography and thorax spiral computed tomography (CT) or magnetic resonance imaging (MRI) in the preoperative period and during follow up period after the surgical plication were reviewed.

We also assessed with standard pulmonary function tests (PFT), including: percent predicted forced vital capacity (FVC%), percent predicted forced expiratory volume in 1 s (FEV1%) and percent predicted maximum forced inspiratory flow (FIFmax%). All PFT results were collected in the upright position at the following intervals: preoperatively, 1 month after diaphragmatic plication, and 2 years after diaphragmatic plication.

1.2. Ethics

The Human Research Ethics Committee of the hospital approved the study protocol.

1.3. Data collection

The details such as age, sex, symptoms, location of eventration, associated anomalies, surgical procedures, complications, survival and follow up details including improvement in symptoms and pulmonary function, complications of surgery, and imaging results of diaphragm were collected, analyzed, and compared.

1.4. Statistics

Statistical analyses were performed using SPSS software version 17.0 (SPSS Inc. Chicago, IL). Categorical data were compared using chi-square test. Follow-up data are compared with preoperative values using Student's t-test for paired and unpaired data as appropriate. A p value of <0.05 was considered statistically significant.

2. Results

2.1. Sex distribution and age at diagnosis

During the study period of 12 years, medical records of 182 children were reviewed. Two patients in whom eventration was related to phrenic nerve injury at delivery and 3 children in whom eventration was related to hypothermia for cardiac surgery were excluded from the study. The remaining 177 patients (boys: 128, girls: 49) who received treatment for CDE were analyzed. Patients' age ranged from 41 min to 12.7 years with a mean age of 10.28 ± 2.35 months and peak age was 0–3 months (71/177). The age distribution of children at the time of diagnosis of CDE is shown in Fig. 1.

2.2. Symptoms of eventration of the diaphragm

Specific symptoms of eventration of the diaphragm were reported for 86 of 177 cases. The main symptoms of CDE in infants included the following: rapid breathing, vomiting, difficult breathing, tachycardia or arrhythmia, and cyanosis. The main symptoms of CDE in children aged >12 months included the following: chest tightness, recurrent respiratory tract infections, and cough or expectoration. Few patients reportedly had two or three symptoms simultaneously. In the present study, the most common symptom of CDE across all age groups of children was rapid breathing (69 cases, 38.9%), which was followed by vomiting (42 cases, 23.7%), recurrent respiratory infections (39 cases, 22.0%), and difficulty breathing (21 cases, 11.8%). About 91 cases were found to asymptomatic or accidentally discovered on routine physical examination (51.4%, Table 1).

2.3. Imaging of eventration of the diaphragm

The chest x-ray with CDE showed unusual increase in basal segments on the lesion side of diaphragm (Fig. 2A). Defect in eventration, which was clearly seen on the lesion side of diaphragm with fluoroscopy, had resulted in the sudden changes in breathing. In addition, an upper gastrointestinal contrast, chest computed tomography scan, magnetic resonance imaging scan, ultrasound scan, or isotope scan could also help to identify diaphragmatic paralysis and diaphragmatic hernia (Fig. 3).

2.4. Localization of congenital diaphragmatic eventration

CDE was unilateral in 176 cases and bilateral in one case. Of the 176 unilateral CDE, 52 (29.55%) were found on the left side and 124

Table 1
Symptoms of congenital diaphragmatic eventration in children (n = 177).

Symptoms	No. of cases	Rate (%)
Rapid breathing	69	38.9
Vomiting	42	23.7
Recurrent respiratory infections	39	22.0
Difficulty breathing	21	11.8
Chest pain	10	5.6
Tachycardia	8	4.5
Arrhythmia	4	2.2
Cyanosis	3	1.6
Other non-specific symptoms	13	7.3
Asymptomatic	91	51.4

Rate meant the probability of one specific symptom appearing in 177 patients.