

## OPEN

# Emergency Management of Acute Late-Presenting Congenital Diaphragmatic Hernia in Infants and Children

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**Objectives:** Acute late-presenting congenital diaphragmatic hernia (CDH) might result in mediastinal shift away from the lesion and even sudden cardiopulmonary arrest. This study aimed to discuss the prompt and effective emergency management of acute late-presenting CDH.

**Methods:** A retrospective review of acute late-presenting CDH cases in West China Hospital of Sichuan University and Guizhou Provincial People's Hospital from October 2010 to June 2016 was conducted.

**Results:** A total of 22 patients were included in this study. All the patients presented with respiratory symptoms. Chest x-ray revealed swollen stomach and mediastinal shift. After nasogastric tube placement, fluid infusion, and nasal oxygen breathing, the symptoms in 8 patients ameliorated, and 14 patients had no signs of obvious relief. Three patients underwent the bedside percutaneous puncture of distensible stomach, and 1 patient died in the process of emergent management for critical condition. The remaining 21 patients underwent emergency surgery. Five thoracotomies and 16 thoracoscopies were performed. Five thoracoscopies that were converted to thoracotomies were required for the difficult reduction of herniated stomach. At follow-up, all patients improved their condition.

**Conclusions:** Acute late-presenting CDH is a clinical emergency that can be fatal. The sudden and progressive expansion of the stomach is mainly responsible for this emergent condition. The prompt and effective management is key to decrease the mortality and achieve favorable prognosis.

**Key Words:** late-presenting, congenital diaphragmatic hernia, management

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Patients with congenital diaphragmatic hernia (CDH) commonly present with symptoms in the neonatal period. Pulmonary hypoplasia and pulmonary hypertension in late-presenting CDH are slightly better compared with neonatal CDH. Therefore, most of the late-presenting CDH patients have no symptoms and are diagnosed incidentally.<sup>1</sup> However, in some conditions, acute symptoms, which are life-threatening, may arise as part of the late-presenting CDH cases.

Acute late-presenting CDH may occur as a result of the herniated and swollen stomach and strangulation and incarceration of the abdominal viscera.<sup>2</sup> Currently, the herniated and swollen stomach as a result of the mediastinal shift is considered to be

the most common reason.<sup>3,4</sup> If the first aid treatments are not managed promptly and effectively, the results are very serious. Preoperative mortality rate of late-presenting CDH is as high as 3.9%, and many patients have presented acute symptoms before the occurrence of sudden cardiorespiratory arrest.<sup>5,6</sup> The prompt and effective management is key to cure the disease. Unfortunately, the reports about the emergent management, operative time, and method of acute late-presenting CDH are still rare, and most are isolated case reports.<sup>6–8</sup>

In this study, we retrospectively reviewed all acute late-presenting CDH cases and tried to summarize the symptoms of this condition to manage this serious condition, decrease the mortality, and achieve favorable prognosis.

## METHODS

We retrospectively reviewed all the patients who presented with acute late-presenting CDH between October 2010 and June 2016 at West China Hospital of Sichuan University and Guizhou Provincial People's Hospital. The study was approved by the Institutional Review Board of West China Hospital of Sichuan University and Guizhou Provincial People's Hospital.

All patients were not diagnosed by antenatal ultrasound screening and had no history of surgery. Neonatal CDH that was diagnosed within 1 month after birth and asymptomatic late-presenting CDH were excluded. All patients were sent to the emergency department because of the manifestations of respiratory symptoms. On examination, instead of decreased or absent breath sounds, bowel sounds were heard with dullness to percussion noted within the left chest. An emergent chest x-ray confirmed the diagnosis (Fig. 1).

## RESULTS

A total of 22 patients were involved in this study. The ages ranged from 2 months to 10 years, with a mean age of 22 months. All chest x-rays revealed the left CDH and a compelling herniated swollen stomach as a result of the mediastinal shift. Immediate fluid infusion, nasal oxygen breathing, and gastrointestinal decompression were performed in all patients. Eight patients were relieved after extracting an ample amount of gas and fluid using the nasogastric tube (NGT). Shortness of breath was noted in 14 patients who were not obviously relieved owing to the ineffective placement of the NGT. Five patients underwent conventional ventilation with tracheal intubation. None of the patients underwent high-frequency oscillating ventilation, extracorporeal membrane oxygenation, and inhaled nitric oxide. Three patients underwent the bedside percutaneous puncture of distensible stomach because of shortness of breath, and oxygen saturation continued to decrease. Two patients improved their condition, and 1 patient died because of the failure to extract the contents in his/her stomach. Five thoracotomies and 16 thoracoscopies were performed in the remaining 21 patients. Five thoracoscopies were converted into thoracotomies because of the difficulty in decreasing herniated swollen stomach. After the operations, all patients were relieved and were able to recover 4 to 12 days postoperative stay. All patients,

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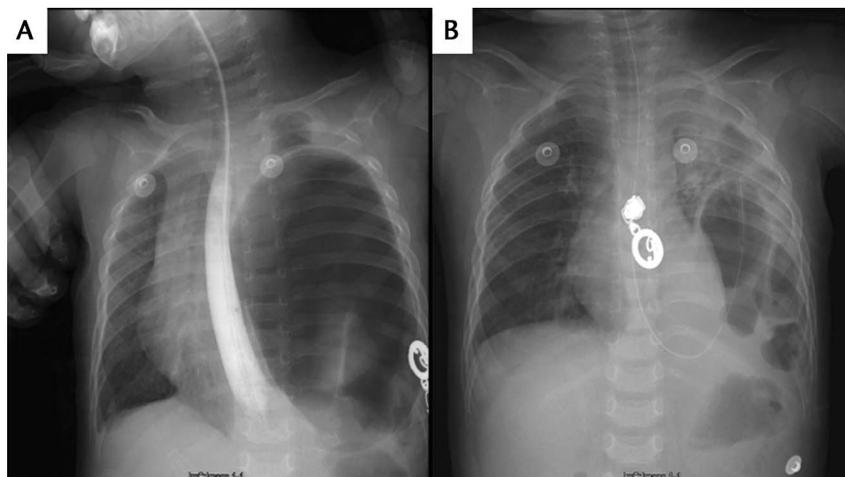
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**FIGURE 1.** Preoperative chest x-ray. A, Radiograph of a child with a left-sided late-presenting CDH showing herniation of stomach into the thoracic cavity and severe mediastinal shift away from the lesion. B, Radiograph of a child with a left-sided late-presenting CDH showing herniation of bowel into the thoracic cavity and slight mediastinal shift.

including the 2 puncturing patients, did not report any intrathoracic infection. Patients had normal diet on postoperative 3.2 days (range, 1–10 days). All patients underwent follow-up at least 3 months. The average period was 22 months (range, 3 months to 5 years). All patients improved their condition without recurrence during the follow-up.

## DISCUSSION

Most CDH cases are diagnosed during antenatal ultrasound screening, but inaccurate diagnoses are still not infrequent, especially in poor areas. Pulmonary hypoplasia and pulmonary hypertension of late-presenting CDH are slightly better compared with neonatal CDH.<sup>9</sup> Most of the late-presenting CDH patients were asymptomatic.<sup>1,10</sup> However, because of the existence of diaphragmatic defect, late-presenting CDH might lead to herniated and swollen stomach and strangulation and incarceration of the abdominal viscera. In our study, all of the patients developed herniated and swollen stomach that resulted in mediastinal shift away from the lesion. It develops when an intrathoracic stomach was largely expanded by inhaling air and/or fluid. Meanwhile, because of esophageal peristalsis and twisted doorway, it formed a 1-way transmission and was difficult to discharge. In addition, the hernia ring was smaller, which presented acute manifestations in some cases. The compression of the hernia ring was more serious for the herniated and swollen stomach. Then, the herniated stomach enlarged progressively and resulted in tension pneumothorax causing the mediastinal shift away from the lesion. Therefore, the main treatment of the disease aimed to decrease the intrathoracic pressure, and the placement of effective NGT was the most direct and the simplest way. However, the NGT might kink at the level of the hernia ring and might work ineffectively. In the group, the symptoms in 8 patients were alleviated, and the condition of the 14 patients did not change.

As to the patients whose symptoms could not be alleviated by the NGT, other methods should be considered such as transthoracic open surgery as soon as possible and even the bedside percutaneous puncture of distensible stomach for the critically ill patients.<sup>11</sup> In our study, 3 patients underwent the bedside percutaneous puncture, and 2 patients were relieved and 1 patient died because of the failure to extract the contents in his/her stomach. In view of our finding, as for critical patients who were on decompensatory stage, the prompt bedside percutaneous puncture of distensible

stomach might save their lives. The thick needle and multiple punctures could be managed to prevent the blocking of the food debris in the stomach. Moreover, during critical times, bedside general anesthesia and emergent thoracotomy might be the life-saving methods. Unfortunately, 1 patient in our series died before we performed these procedures.

After the emergent management, surgery should be performed as quickly as possible. Operative approaches of late-presenting CDH could be performed by transabdominal and transthoracic open techniques and laparoscopic or thoracoscopic minimally invasive surgery. The main objective of the operative techniques to repair acute late-presenting CDH was to reduce the herniated swollen stomach and decrease the compression to enhance cardiopulmonary circulation.<sup>11</sup> Koop and Johnson<sup>12</sup> reported that the transthoracic approach had the following advantage: it empties the thoracic cavity rapidly, especially in the respiratory-distressed child. In view of our experience, direct thoracotomy was more appropriate to prevent death, especially for the critically ill patients. Thoracoscopy was more suitable for patients whose general situations were good and whose symptoms could be alleviated by NGT. It was necessary to closely monitor the patients' intraoperative vital signs and blood gases. When the herniated stomach was difficult to reduce, thoracoscopic operation should be converted to thoracotomy immediately. Thoracoscopy had the following advantage: it has less postoperative pain, scar, wound infection, and deformation of the thorax. However, it also had the following disadvantages: long learning curve, additional performance of intraoperative artificial pneumothorax, and invisible view by the shelter of the herniated stomach that was also obvious.<sup>13–15</sup>

In conclusion, the core of acute late-presenting CDH was the compression of herniated stomach that expanded dramatically. The objective of the pivotal emergent management was to decrease the intrathoracic pressure. Effective NGT placement was the most direct and simplest method. The surgery should be performed quickly. Once prompt and effective treatments were managed, the prognosis could be favorable.

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