INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a well-known cause of severe respiratory distress in the newborn and is associated with high mortality rates (1). Although congenital diaphragmatic hernias become symptomatic immediately after birth or in the first days of life, 5-20% of these cases are diagnosed after the newborn period (1-3). The symptoms, diagnosis, management, and complications of late presenting CDH differ considerably from neonatal CDH. Late-presenting CDH might be a significant diagnostic challenge since its clinical presentation is not specific, and therefore the diagnosis is often overlooked (4, 5).

Here, we report three cases of CDH with late presenting symptoms and discuss the difficulty of the diagnosis. The asymptomatic period in the three patients terminated with acute respiratory symptoms forcing the children and the parents to seek medical advice Table 1.

Case Reports

Case 1
A nine year-old boy was referred to our clinic with suspected diaphragmatic rupture after a minor bi-
cycle accident. Previously he had never had any respiratory or gastrointestinal symptoms, apart from the abdominal pain following the accident. The patient was admitted in good condition in order to perform his diagnostic routine. Although at the examination increased respiratory effort and decreased breath sounds on the left side of his chest were found, he was still not complaining of chest pain, dyspnea or cyanosis. His chest X-ray showed air-fluid levels in the middle and lower parts of his left hemithorax with an indistinct left diaphragm. CT scan could not be done due to ongoing technical problems at the emergency department at that time.

Since the findings of the patient strongly supported the diagnosis of diaphragmatic rupture, he was operated on for diaphragmatic rupture on the day of admission. At operation, the stomach, spleen and a part of the small bowel were found to be herniated into the pleural cavity. There was no hernial sac and there were no findings suggesting a diaphragmatic rupture (diaphragmatic discontinuity, thickening and hematoma were not found at laparotomy). Following this, a small congenital diaphragmatic defect was closed with 2-0 non-absorbable monofilament sutures. The postoperative course of the patient was uneventful.

Case 2
A 53-day-old girl was admitted to the ward with the complaint of dyspnea and cyanosis after vomiting. Her past medical history did not reveal any previous respiratory problems or fever. On examination, she had cyanosis and decreased breath sounds over the left lower hemithorax. As her initial chest radiograph was misinterpreted as a pneumothorax, a chest tube was inserted. However, no air drained through it. Thankfully, the stomach was not perforated. After insertion of a nasogastric tube, a chest x-ray was obtained. It revealed a hugely-dilated intrathoracic stomach and herniated intestinal loops, which were missed initially. When the x-ray at admission was re-evaluated, no gastric gas shadow in the abdomen was noted. An emergency laparotomy was performed to repair the defect in the diaphragm. The stomach, transverse colon and spleen were found herniated into the left hemithorax. Following reduction of the intraabdominal contents, the small diaphragmatic defect was repaired by using interrupted 4-0 propylene sutures. The postoperative course was uneventful, and the patient was discharged on the third postoperative day. The patient has been free of signs and symptoms during a 9-month follow-up period.

Case 3
A sixteen-month-old girl was referred to our clinic with suspected diaphragmatic hernia. She had no complaints 15 days before the admission. She was taken to the pediatric emergency department due to a high body temperature rising to over 39.0°C and a cough. In her chest radiogram intestine-like images were noticed in her left hemithorax, suggesting a congenital diaphragmatic hernia. Then Computerized Tomography was performed and a Bochdalek Hernia was identified (Figure 1). The patient had no previous respiratory complaints. In her medical history she had undergone an operation at the age of two months for lypomeningomyelocele with an uneventful postoperative course, which was quite interesting. In addition, the diaphragmatic hernia, which was recognized incidentally during the evaluation of the high body temperature, was not identified at the time of the operation. An elective surgery was performed at the time of the operation. An elective surgery was performed during the evaluation of the high body temperature, was not identified.

Table 1. Clinical data of the patients with late presentation congenital diaphragmatic hernia

<table>
<thead>
<tr>
<th>Year/sex</th>
<th>Symptom</th>
<th>First diagnosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case-1</td>
<td>9y/M Abdominal and chest pain after accident</td>
<td>Diaphragmatic rupture</td>
<td>Laparotomy and primary repair</td>
</tr>
<tr>
<td>Case-2</td>
<td>2m/F Dyspnea, cyanosis, vomiting</td>
<td>Pneumothorax</td>
<td>Laparotomy and primary repair</td>
</tr>
<tr>
<td>Case-3</td>
<td>16m/F Fever, cough</td>
<td>Diaphragm hernia</td>
<td>Laparotomy and primary repair</td>
</tr>
</tbody>
</table>

Figure 1. Gastric gas was shown in the thorax on Tomography and spleen and stomach were found in the thorax. After reduction of the contents, the diaphragmatic defect was repaired with 4-0 propylene sutures. The patient was discharged on postoperative day five and the follow up period was uneventful.

DISCUSSION

Congenital diaphragmatic hernias are known as a neonatal disease. Late presentation is not a common feature for diaphragmatic hernias. Five to 20% of Bochdalek hernias have been reported to appear beyond the neonatal period (2, 5). Although presumably the congenital diaphragmatic defects of these delayed CDH are anatomically similar to the defects of neonatal diaphragmatic hernias, the clinical presentation, operative management and complications differ considerably from those of neonatal CDH (2, 3, 5).

Late-presenting CDH is characterized by a variety of clinical features. The patients may be symptomatic or completely asymptomatic. The symptomatic patients may present with a wide range of acute or chronic, respiratory or gastrointestinal complaints. Most asymptomatic Bochdalek hernias are right sided and the diaphragmatic defect is covered by the liver but it seems possible for left-sided hernias to be asymptomatic (1, 2, 6). Usually there is no foreign content in the right pleural cavity, so maybe the asymptomatic period of the left Bochdalek hernia could be the period of the absence of foreign content.
content in the left pleural cavity. This suggests that the asymptomatic period finishes with the entrance of abdominal contents through the left diaphragmatic defect (3-6). Thus, the symptoms of the patient in case one could have started with the acute entrance of the abdominal contents into the left pleural cavity due to the sudden increase in the abdominal pressure following the abdominal trauma.

The symptoms and signs are far from specific and overlap considerably with other pathologies. Our second case of misdiagnosed late-presenting CDH, which was thought to be a pneumothorax or pleural effusion with subsequent insertion of a chest drain, has been reported in the literature (3, 7, 8). All of these publications discussed the difficulties in establishing the diagnosis of the late onset congenital diaphragmatic hernias. In such cases CDH presents a far more complex diagnostic problem. Various diagnostic modalities such as plain chest radiographs, real-time ultrasound and contrast meal and enema have been suggested to evaluate the integrity of the diaphragm (8-10). Confirmative barium studies have been regarded as unnecessary in neonates but have been advocated in older children.

Although the herniation in our cases appeared later, the question of whether the diaphragmatic defect itself is congenital or acquired remains unsolved. It has been assumed that a diaphragmatic defect was present in the prenatal period, but the defect is small and pulmonary hypoplasia was insignificant. The size itself of the hernia foramen is unlikely to be a determining factor for the time of clinical presentation of CDH (8, 9).

CONCLUSION

Clinical late presentation of left Bochdalek hernia is not specific and the diagnosis is often overlooked. Therefore, late presenting left CDH should be suspected in every patient whose diaphragm is not clearly visible on radiography.

Acknowledgment: The authors are thankful to Dr. İ. Onur Ozen, University of Gazi Medical School for reviewing this manuscript.

Conflict of Interest
No conflict of interest was declared by the authors.

REFERENCES