



# Two Infant Cases Admitted with Atypical Presentation and Diagnosed as Type IV Hiatal Hernia

## Atipik Prezantasyonla Tip IV Dev Paraözofageal Hiatal Herni Tanısı Alan İki Infant Olgu

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### Abstract

Type IV paraesophageal hiatal hernias are diaphragmatic hernias that are extremely rare in the pediatric age group in which the stomach and other intra-abdominal organs herniate from the esophageal hiatus of the diaphragm into the mediastinum. Since the defect in the hiatus is large, serious complications such as gastric volvulus may develop in these giant hernias as a result of the passage of a large part of the stomach into the thoracic cavity. Patients may present with atypical clinical manifestations such as growth retardation, not gaining weight, and recurrent pulmonary infections. In this report, two cases in the pediatric age group who were admitted with different clinical course and were diagnosed as type IV paraesophageal hiatal hernia who were successfully treated with surgical intervention are presented.

**Keywords:** Hiatal hernia, paraesophageal hernia, pediatrics, surgery, treatment, type IV

### Öz

Tip IV paraözofageal hiatal herniler; mide ve diğer intraabdominal organların diyafragmanın özofageal hiatusundan mediastene herniye olduğu, çocuk yaş grubunda oldukça nadir görülen diyafragmatik hernilerdir. Hiatastaki defekt büyük olduğundan midenin büyük bir kısmının torasik kaviteye geçmesi sonucu gelişen bu dev hernilerde, mide volvulusu gibi ciddi komplikasyonlar gelişebilir. Hastalar büyüme geriliği, kilo alamama, tekrarlayan akciğer enfeksiyonu gibi atipik kliniklerle karşımıza çıkabilir. Bu yazıda çocuk yaş grubunda farklı klinik tablolar ve semptomlarla başvuran ve tip IV paraözofageal hiatal herni tanısı alıp cerrahi müdahale ile başarılı şekilde tedavi edilen iki olgu sunulmuştur.

**Anahtar Kelimeler:** Hiatal herni, paraözofageal herni, pediatri, cerrahi, tedavi, tip IV

### Introduction

Paraesophageal hiatal hernias are rarely seen in childhood. The Society of American Gastrointestinal Endoscopic Surgeons has defined 4 types of hiatal hernias.<sup>1</sup> Type I hiatal hernias are characterized by the herniation of the gastroesophageal junction into the thoracic cavity only, called a sliding hernia. Type II-III-IV hiatal hernias are true paraesophageal hernias. They are classified according to the location of the

gastroesophageal junction and the organ herniated into the thoracic cavity. Type II paraesophageal hiatal hernia has a normal anatomically located gastroesophageal junction; however, a part of the stomach is herniated, mostly from the fundus hiatus. Type III paraesophageal hiatal hernia is the type in which a part of the stomach herniates from the hiatus and the gastroesophageal junction has an abnormal anatomical location. Type IV paraesophageal hiatal hernia is the type in which abnormally located gastroesophageal junction and

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another intra-abdominal organ, mostly the colon, herniates into the thoracic cavity.<sup>2</sup> If more than half of the stomach is in the thorax, it is called a giant paraesophageal hernia.

These hernias may be detected incidentally or may present as recurrent lung infections and gastrointestinal symptoms. Due to their rarity and non-specific clinical features, diagnosis may be delayed until adulthood.<sup>3</sup>

In this study, we presented the clinical and radiological findings of two cases with type IV giant paraesophageal hiatal hernia, who applied with different clinical symptoms and were diagnosed and treated by chance.

## Case Reports

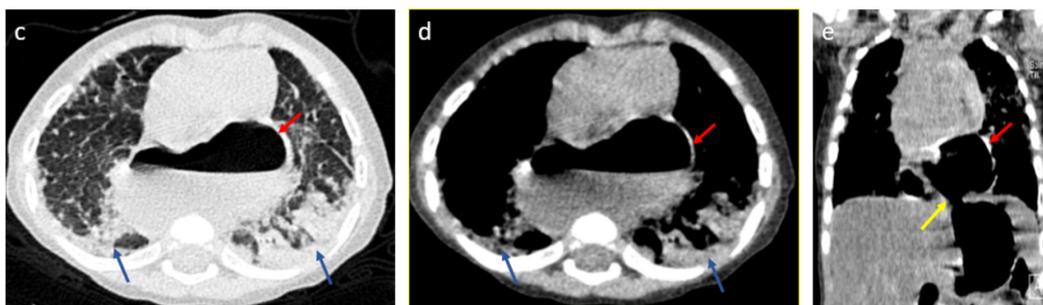
### Case 1

A 10-month-old baby girl was brought to the outpatient clinic with the complaints of weight gain, diarrhea and rash. The patient's history, who was born at term, was unremarkable. Her body weight was 5850 g (<3 percentile), her height was 65 cm (5 percentile), and her head circumference was 41.5 cm (5 percentile). On physical examination, there was a maculopapular rash on the arms and the liver was palpable 4 cm on the right midclavicular line. Respiratory system and cardiovascular system examination was normal. In laboratory examinations, white blood cell (WBC) was  $22.85 \times 10^3/\mu\text{L}$ , NE was  $6.35 \times 10^3/\mu\text{L}$ , LY was  $15.45 \times 10^3/\mu\text{L}$ , platelet count (PLT) was  $383 \times 10^3/\mu\text{L}$ , Hgb was 13.2 g/dL, and C-reactive protein (CRP) was 8.4 mg/L. Biochemical test results were

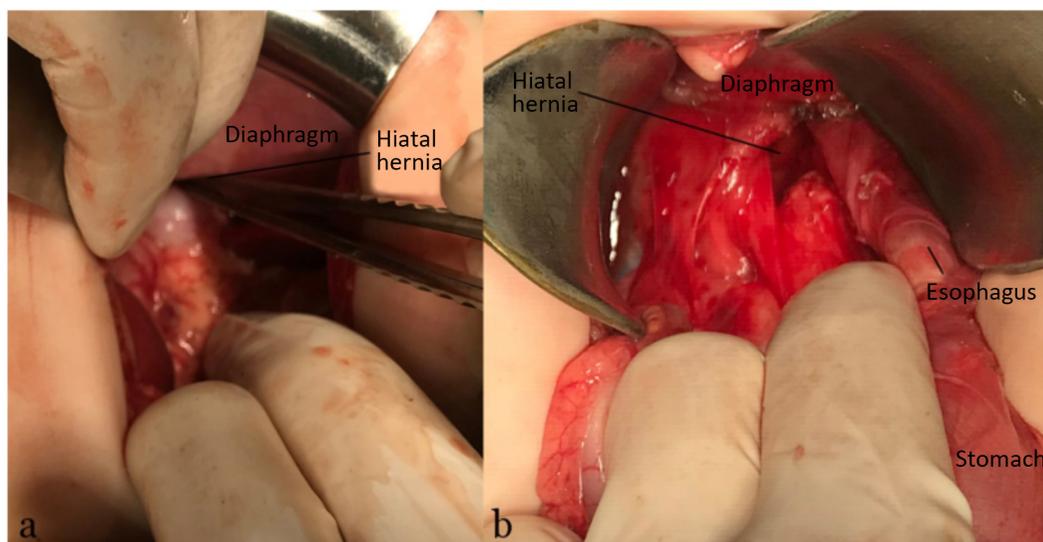
within normal limits. The patient was investigated for the differential diagnosis of rash, hepatomegaly, and growth retardation. Allergic and metabolic tests were sent. Posterior-anterior chest radiography (PACR) revealed gas shadow consistent with a hiatal hernia superposed to the heart and diffuse infiltration in both lungs (Figure 1a). In the esophagus-stomach-duodenum radiograph (ESD), it was observed that the esophagogastric junction and gastric fundus were located supradiaphragmatically in the retrocardiac area (Figure 1b). In non-contrast thorax computed tomography (CT), it was observed that most of the esophagogastric junction, stomach fundus and corpus were located in the mediastinum with a retrocardiac location (Figure 1c-e). A type IV paraesophageal hiatal hernia was diagnosed because there was a hiatus hernia with a diameter of 4 cm and the cardia, fundus and corpus part of the stomach from the hiatal defect, as well as the transverse colon and pancreas were herniated into the thorax (Figure 2a, b). Diffuse fibroatelectasis changes were also detected in both lungs on tomography and it was evaluated as the development of pulmonary fibrosis. The patient was operated by a pediatric surgeon. In the operation, herniated organs to the thorax were reduced into the abdomen, the hernia sac was removed, and the hiatus defect was repaired and the Nissen fundoplication was performed. The patient, who was followed up in the pediatric intensive care unit due to postoperative lung problems, was extubated after being followed up on a mechanical ventilator for 10 days. Oral feeding was started on the 4<sup>th</sup> postoperative day, and there was significant weight gain after the operation,



**Figure 1a.** A gas shadow superposed to the retrocardiac area is observed on the posterior-anterior chest radiograph (red arrow). In addition, areas of infiltration are seen in both lungs, more prominently on the left (asterisk). **b.** The esophagus-stomach-duodenal radiograph shows abnormal location of the esophagogastric junction (yellow arrow) and herniation of the gastric fundus into the thorax (red arrow)



**Figure 1 c-e.** Herniated stomach (red arrows) in the retrocardiac area is observed in the axial lung (c) and mediastinal (d) windows in uncontrasted thorax computed tomography examination. In addition, diffuse fibroatelectasis densities are seen in the lower lobes of both lungs (blue arrows). Coronal reformat image (e) shows the presence of hiatal defect (yellow arrow) and herniation of the gastric fundus (red arrow)



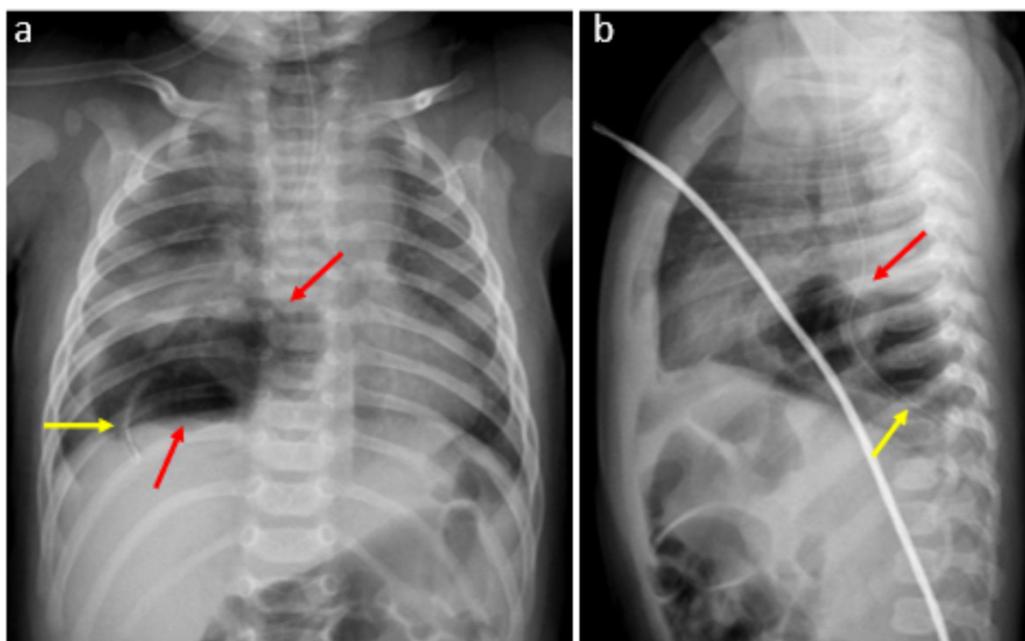
**Figure 2 a, b.** Intraoperative images of hiatal hernia: (a) The entire stomach, which should be on the left of the midline in laparotomy, below the left lobe of the liver, herniated into the thorax in a hiatal hernia, the tip of the forceps entered into the hiatal hernia opening, and transverse colon is observed just below the hernia sac. (b) Large hiatal hernia defect in the esophageal hiatus of the diaphragm with the stomach and esophagus reduced into the abdomen

respiratory complaints regressed, and diarrhea decreased. It was observed that there was a significant improvement in all symptoms of the patient. The general condition of the patient after discharge is good and her follow-up continues without any symptoms.

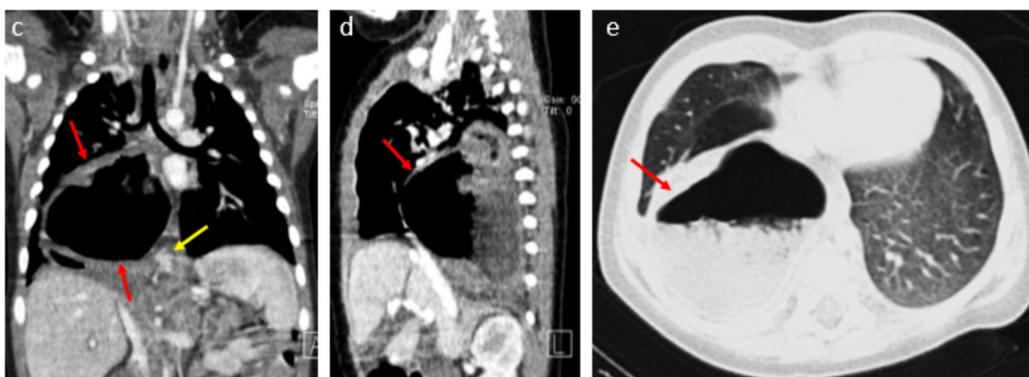
## Case 2

A 2-month and 27-day-old boy was brought to the emergency department with the complaints of fever, vomiting, restlessness, and decreased sucking. It was learned that the patient, who was born at term, used medication due to gastroesophageal reflux disease. On physical examination, it was found that there was no desaturation, but respiratory sounds were decreased in the right lung base. In laboratory examinations, WBC was  $12.63 \times 10^3/\mu\text{L}$ , PLT was  $733 \times 10^3/\mu\text{L}$ , Hgb was 12.8 g/dL, NE was  $6.86 \times 10^3/\mu\text{L}$ , LY was  $5.03 \times 10^3/\mu\text{L}$ , and CRP was  $<3.11$  mg/L. No pathology was detected in blood gas and biochemical tests. In PACR and right lateral chest X-ray, gas shadow with the termination of the nasogastric tube was observed in the lower zone of the right hemithorax

(Figure 3 a, b). In contrast-enhanced thorax CT, it was observed that the head of the pancreas and the total stomach were herniated from the midline behind the heart, from a paraesophageal defect, to the level of the right hemithorax carina; the esophagus was shifted to the right, enlarged and its contents were leveled; the stomach was folded on itself and was consistent with organoaxial volvulus (Figure 3 c-e). The findings were evaluated as consistent with type IV hiatal hernia and gastric volvulus. The patient underwent a corrective operation by the pediatric surgeon. A type IV paraesophageal hiatal hernia was detected in the operation. It was found that there was a hiatus hernia with a diameter of 6 cm and the entire stomach and pancreas tail herniated from the hiatal defect into the thorax. The herniated organs were reduced into the abdomen, the hernia sac was excised, the hiatus defect was repaired, and the Nissen fundoplication was performed. After the operation, the patient, whose pulmonary infection and respiratory symptoms improved and the complaint of vomiting regressed, was discharged. It is followed up without any problems in outpatient controls.



**Figure 3 a, b.** Posterior-anterior lung (a) and lateral lung (b) radiographs show that the stomach gas is not in its normal position and the air-containing gastric structure (red arrows) in which the nasogastric tube extends (yellow arrow) herniates into the right hemithorax



**Figure 3 c-e.** In contrast-enhanced thorax computed tomography examination, coronal (c) and sagittal (d) reformatted, axial plane lung window (e) images show the entire stomach (red arrows) and a part of the pancreas (yellow arrow) is herniated into the right hemithorax

## Discussion

Paraesophageal hiatal hernias may present with acute or chronic findings. Acute presentation can be in the form of life-threatening complications such as incarceration, obstruction, perforation, bleeding, and anemia, or it can be in the form of respiratory system symptoms.<sup>1,3,4</sup>

In chronic presentation, they may present with recurrent lung infections that occur with fever and cough attacks. There may be intermittent episodes of vomiting and it can usually be attributed to gastroesophageal reflux disease, which is common in this age group.<sup>5</sup> Restlessness and refusal to be fed may be other symptoms. Widespread infiltrates in the lungs in the first case and recurrent vomiting attacks in the second case were associated with gastroesophageal reflux disease, and a significant improvement was observed in symptoms after surgical correction in both cases.

Especially in type IV paraesophageal hiatal hernia, respiratory symptoms, which can be seen secondary to the compression of the lung by herniated organs and accompanying gastroesophageal reflux, are more pronounced. As in our first case, microaspiration findings secondary to gastroesophageal reflux and growth retardation due to recurrent pneumonia can be seen in the lung.

As in our second case, in conditions in which most or whole of the stomach herniates into the thorax, patients can also apply in an emergency due to gastric volvulus. Since the circulation of the stomach is disrupted and perforation may develop in gastric volvulus, it may cause morbidity and mortality.<sup>6-9</sup>

Hiatal hernia should be considered when air densities superposed with the shadow of the heart are seen in the thoracic cavity in the chest X-ray. In suspected cases, lateral chest X-ray and visualization of the nasogastric tube in the

thorax also support the diagnosis. Opacity or air fluid level at the cardiophrenic angle may be the only clue in many cases. A definitive diagnosis is made with ESD examination and/or thorax CT and herniated organs are defined more accurately. In our cases, hernia was suspected on chest X-rays and the diagnosis was confirmed with thorax CT.<sup>3,10</sup>

Although type IV paraesophageal hiatal hernia is rare in the pediatric age group, it should be kept in mind in the differential diagnosis in children who have growth retardation, who cannot gain weight, who have intermittent vomiting attacks, and who have recurrent pulmonary infections.

### Ethics

**Informed Consent:** Informed consent was obtained from the families of the patients.

**Peer-review:** Externally peer-reviewed.

### Authorship Contributions

Concept: E.Ö., M.U.Y., Design: E.Ö., Ö.B., S.K.Ş., Analysis or Interpretation: E.Ö., M.U.Y., Ö.B., A.K., S.K.Ş., Literature Search: E.Ö., M.U.Y., Ö.B., Writing: E.Ö.

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

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