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Congenital hiatus hernia: A case series

Didem Baskin Embleton,[1] Ahmet Ali Tuncer,[1] Mehmet Surhan Arda,[2] Huseyin Ilhan,[2] Salih Cetinkursun[1]

[1]Department of Pediatric Surgery, Afyon Kocatepe University Faculty of Medicine, Afyonkarahisar, Turkey
[2]Department of Pediatric Surgery, Osmangazi University Faculty of Medicine, Eskisehir, Turkey

ABSTRACT

OBJECTIVE: Hiatus hernia is frequent in adults and rare in children; congenital hiatus hernia is even rarer. In this study, we describe a group of infants with congenital hiatus hernia and discuss its management.

METHODS: Records of patients (male: 3, female: 4) who were diagnosed with congenital hiatus hernia between 2010 and 2016 were extracted. Demographic data, presenting symptoms, diagnostic investigations, operative details, postoperative follow-up, and early and late postoperative complications were evaluated retrospectively.

RESULTS: Four patients were female and three were male. One patient was diagnosed prenatally while the mean age at diagnosis for others was 18.6 months. Four patients had type IV hernia, 2 had type III hernia, and one had type I hernia. The diagnosis was confirmed by chest X-ray, computerized tomography (CT) and/or upper gastrointestinal series. The hiatal repair was done in all patients either by laparotomy or laparoscopy. During the procedure, 2 patients had Nissen fundoplication and 3 patients had Thal fundoplication. Recurrence of hernia occurred in the 2 patients who had Thal fundoplication.

CONCLUSION: Recurrence of sliding hernias with Thal fundoplication seem more frequent in the series. If the esophagogastric junction is present in the thorax, mediastinal dissection of the esophagus may be required to achieve a good abdominal esophagus structure, which will prevent a recurrence.

Keywords: Congenital; hiatus hernia; Nissen fundoplication; short esophagus; Thal fundoplication.

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Hiatus hernia (HH) frequently occurs in adults but is a rare entity in children. It is characterized by the displacement of any abdominal structure other than the esophagus into the thoracic cavity through a widening of the hiatus of the diaphragm. It usually occurs secondary to anti-reflux surgery due to the extensive dissection of the hiatus in adults; whereas, in children, it is usually primary or congenital and can cause gastroesophageal reflux [1, 2]. Failure to thrive, coughing, and vomiting are the most frequent patient complaints. Due to its rarity, not very many series have been published regarding the pathology of HH [1, 3, 4].

In the present study, a case series of 7 infants with congenital HH was described, and the presenting symptoms, diagnosis, and treatment were discussed.

MATERIALS AND METHODS

After approval by the ethics committee for clinical research, records of patients who were diagnosed with congenital HH in two different medical faculties in neighboring cities, between 2010 and 2016 , were investigated. Demographic data, presenting symptoms, diagnostic investigations, operative details, postoperative follow-

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Correspondence: Dr. Ahmet Ali TUNCER. Afyon Kocatepe Universitesi Tip Fakultesi, Cocuk Cerrahisi Anabilim Dali, 03200 Afyonkarahisar, Turkey.
Tel: +90 505 948 93 71 e-mail: drtaali@yahoo.com

up, and early and late postoperative complications were retrieved, and a retrospective evaluation was done. Late complications included recurrence of hernia, a continuation of reflux symptoms, and a continuation of pulmonary symptoms.

RESULTS

A total of 7 children (4 girls and 3 boys) were diagnosed with congenital HH in the pediatric surgical departments of 2 university hospitals over the course of 6 years (Table 1). One patient was diagnosed prenatally and the mean age of diagnosis for the rest was 18.6 months (4–41 months). Coughing was the most frequent complaint, followed by vomiting and failure to thrive, respectively. HH was detected incidentally in 1 patient following trauma.

All patients underwent anteroposterior (A-P) and lateral X-rays (Fig. 1, Fig. 2), and the diagnosis was confirmed with upper gastrointestinal (GI) series (Fig. 3) and/or computerized tomography (CT) (Fig. 3). Prenatal diagnosis of a thoracic cystic mass was made in 1 patient during the third trimester with prenatal ultrasonography. The mother was under supervision for diabetes mellitus and hypertension. Postnatal evaluation of the newborn with chest X-rays and gastrointestinal series revealed a HH.

Table 1. Characteristics of patients

Patients	Age at diagnosis (mo)	Gender	Presenting symptoms	Diagnosis	HH type	Additional pathology	Approach	Complication	Late results
1	8	F	Cough	Chest x-ray, thorax CT	IV	Compressive atelectasis	Open, hiatal repair, esophageal fixation		Well after 3 yrs.
2	4	F	Vomiting, failure to thrive	Upper GI series	I		Open, hiatal repair, Thal fundoplicatin	Recurrence, hiatal repair, Nissen fundoplication	Occasional vomiting
3	5	M	Cough, vomiting	Chest x-ray, upper GI series,	IV		Open, hiatal repair		Occasional vomiting, received anti-reflux medication
4	41	F	Cough	Chest x-ray, thorax CT	IV	Compressive atelectasis	Open, hiatal repair, Thal fundoplication		well
5	18	F	Cough	Upper GI series, thorax CT	IV		Lap. hiatal repair, Nissen fundoplication		well
6	prenatal	M	Prenatal	Chest x-ray, upper GI series	III		Open hiatal repair, Nissen fundoplication		Well after 7 mo.
7	36	M	Incidental after trauma	Upper GI series	III		Lap. hiatal repair, Thal fundoplication	Recurrence	?

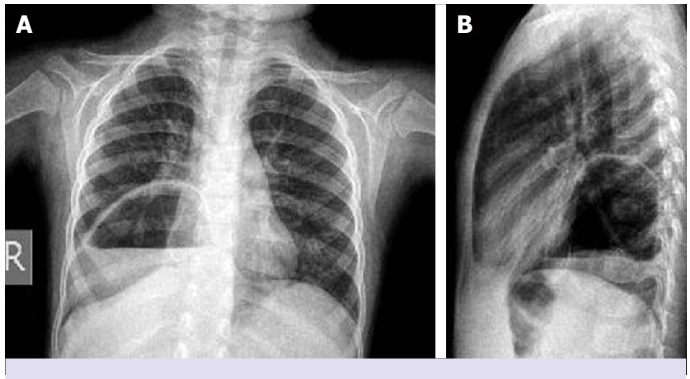

FIGURE 1. **(A)** A-P chest X-ray of patient 4 showing a slightly median air-fluid level on the right hemithorax. Gastric air is absent in its usual location. **(B)** Lateral chest X-ray of the same patient showing that the air-fluid level is placed posteriorly. This patient had a type IV hernia.

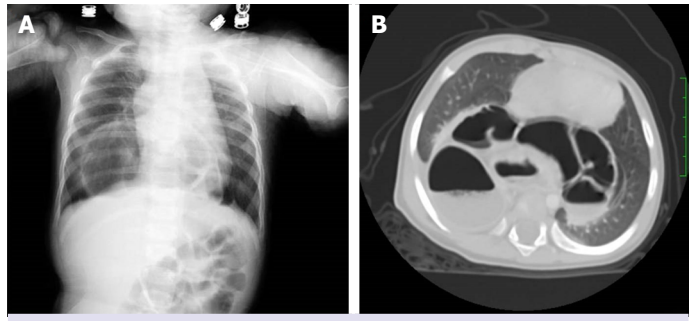

FIGURE 2. **(A)** A-P chest X-ray of patient 1 showing mid-thoracic air locules resembling intestinal gas. Gastric air is not seen at its usual location. The lesion looks like a Morgagni hernia. **(B)** Computerized tomography of the same patient shows posteriorly placed diaphragmatic defect which includes intestinal structures. This patient had a type IV hernia.

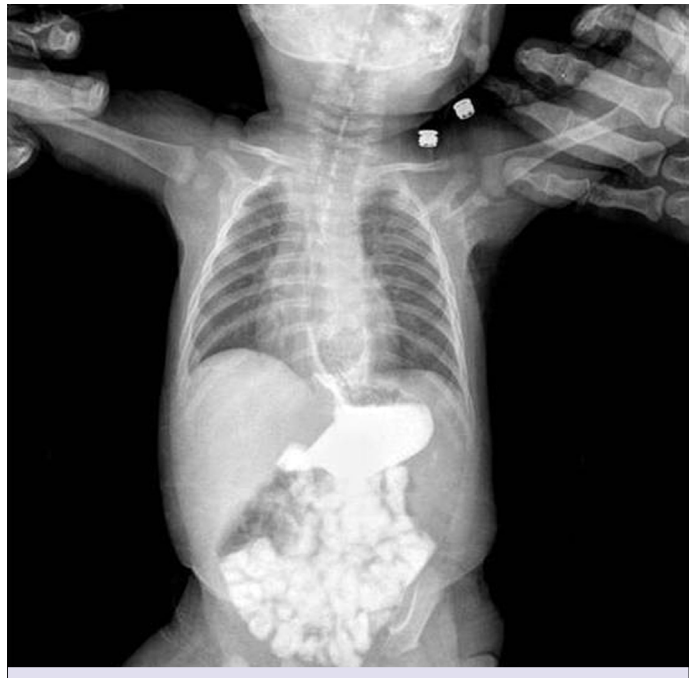

FIGURE 3. Upper gastrointestinal series of patient 2 showing thoracic replacement of gastric fundus from a medial diaphragmatic defect. This patient had a type I hernia.

Four patients had Type IV, 2 patients had Type III and one patient had Type I HH. In type IV hernias, the stomach was accompanied by a small bowel and colon projection in the hernia sac. No additional congenital anomalies were detected in any of the patients.

Laparotomy was performed in 5 patients and laparoscopy in 2 patients. The laparoscopy had been switched to laparotomy due to perforation of the stomach in 1 patient in the laparotomy group. Reduction of the viscera, excision of the hernia sac, and mobilization of the esophagus for an appropriate intra-abdominal length were performed in all patients independent of the surgical approach. Two hiatal stitches were applied with 2/0 round silk sutures in all patients. As an anti-reflux measure, Nissen fundoplication was performed in 2 patients, and Thal fundoplication was performed in 3 patients. Mean hospitalization duration of the patients was 5.5 days after the operation. There were no early postoperative complications.

Late postoperative complications included a recurrence of hernia in 2 patients and continuation of reflux symptoms in 2 patients. Three patients had no complaints. One of the two laparoscopic repair patients and 1 in the open approach group complained of hernial recurrence (28.5%). One of them was the patient with the unsatisfactory intra-abdominal esophagus who had undergone Thal fundoplication with the hiatus repair. She was admitted with vomiting and cough in the first month postoperatively. Collis gastroplasty was not performed due to the poor nutritional status, and Nissen fundoplication and gastrostomy were added to the treatment to repair the recurrent HH. Reflux symptoms continued for a while in this patient. The other recurrence was in a type III patient who had Thal fundoplication added to his hiatus repair. The parents refused re-operation and the patient did not attend the follow-up. One patient had transient slow transit time probably due to vagal injury. He underwent gastric motility enhancing treatment for about a year to control his reflux symptoms. Another patient had transient vomiting attacks that lasted for a few months.

Mortality was not observed at a follow-up of 1 month to 5 years in the study group.

DISCUSSION

HHs are extremely rare pathologies in children in which the underlying reason may be congenital or acquired. Karpelowsky et al. have stated that HH is frequent following anti-reflux surgery in children [1]. In a recent study by Akbulut et al., it was mentioned that HH was detected in only 0.26% (n=8) of 3081 patients who underwent esophagogastroduodenoscopy [5]. Although we had a limited number of patients in our study group, none of the patients underwent a previous anti-reflux procedure, so they were considered as having congenital HHs. The incidentally diagnosed trauma patient did not suffer a diaphragmatic trauma that could cause a HH.

Four types of HH have been defined in the SAGES guidelines (2013); Type I: sliding hernia where the gastroesophageal junction migrates above the diaphragm, Type II: pure rolling or paraesophageal hernia (PEH) where the gastroesophageal junction remains in place but a portion of the fundus herniates through the hiatus adjacent to the esophagus, Type III: combined sliding-rolling hernia, and Type IV: HH with presence of other structures such as omentum, colon or small bowel within the hernial sac [6]. Type II and type IV HH are also named as PEH [1, 3]. More than 95% of cases are classified as type I HH [6]. On the contrary, 57% of the patients in the presented series had type IV HH.

Presenting findings included cough, vomiting, failure to thrive, and pneumonia, which are consistent with classical knowledge. Especially in type IV hernias, respiratory symptoms were more prominent, probably due to the compression of the lungs by the herniated organs, which augmented the effect of gastroesophageal reflux. Recurrent pneumonia may also lead to failure to thrive as we observed in one of our patients. Failure to thrive was seen in another patient with type I HH, due to frequent vomiting. HHs in children may present with additional complications such as volvulus of the stomach that may lead to morbidity or mortality [3, 7]. We did not see any such complications in our series.

HH was suspected based on the findings of the chest X-rays taken during the evaluation of cough and the gastroesophageal symptoms. Anteroposterior (A-P) and lateral chest X-rays showed the presence of HH because the large and small bowel was observed to be protruded into the thorax as well as the stomach, due to which type IV hernias may easily be confused with posterolateral diaphragmatic hernias. If the hernia is on the medial side in the plain chest X-ray and lies posterior in the lateral chest X-ray, HH may be strongly suspected. Sometimes, the stomach may slide to the right side of the hiatus and the appearance may resemble a right-sided diaphragmatic hernia, as was observed in one of the initial patients in the series. An upper GI series or a contrast-enhanced CT of the thorax will provide the correct diagnosis in such a case. In our series, 5 patients in the study group underwent a CT scan and all cases underwent a fluoroscopic investigation. Therefore, we believe that to corroborate the HH suspicion in A-P and lateral chest X-rays, a fluoroscopic GI series is essential to achieve the correct diagnosis. With our experience, CT alone is also satisfactory for diagnosis, because it shows the anatomy of both sagittal and coronary views and additionally shows the mediastinum and lung lesions in the patient. Considering the burden radiation places on children, only one of these radiological investigations should be performed, depending on the experience of the radiologist and surgeon [4].

Prenatal diagnosis of a HH is possible and usually made in the third trimester according to the literature, but abnormal ultrasonography findings such as mediastinal mass or tubular structure can be detected as early as second trimester [4, 8, 9]. The diagnosis was established prenatally in one of our patients. Preliminary ultrasonography diagnosis was a thoracic cyst in the third trimester. Postnatal chest X-rays and GI series confirmed the diagnosis of a HH.

Surgical repair of HH has been recommended in children according to recent SAGES guidelines [6]. Either laparotomy or laparoscopy could be preferred depending on the size of the patient, size of the hernia, and experience of the surgical team. In our series, 4 repairs were done with laparotomy, 1 patient converted to laparotomy from laparoscopy, and 2 repairs were done with laparoscopy. Although the number of patients studied is not enough for a conclusive opinion, we believe that laparoscopic repair is successful in exhibiting the real anatomy and also allows for further dissection in the mediastinum. There are several approaches for additional anti-reflux procedures, all of which are a part of hiatal hernia repair [1, 4]. The anti-reflux operation was performed on all patients in our series except in 2 early patients. Both of these patients had type IV HH with intra-abdominal gastroesophageal junction, but only the

hiatal defect was repaired. We did not see a recurrence of the HH or persistence of gastroesophageal reflux in these patients.

Short esophagus and insufficient mobilization of the gastroesophageal junction may lead to recurrence or continuation of the gastroesophageal symptoms. A short esophagus is described as having an abdominal esophagus length shorter than 2–3 cm in adults in different series [6]. The description is not clear in children. It may be associated with a HH as seen in one of our patients, who had a recurrence and underwent a re-operation with extensive mobilization of the esophagus, hiatus repair, Nissen fundoplication, and gastrostomy to fixate the stomach and also to feed the patient. Collis gastroplasty or vagotomy is being offered for the management of short esophagus in adults [6]. It was not preferred in our patient due to the poor nutritional status of the child.

SAGES guidelines advocate the dissection and excision of the hernia sac to release the hiatus and gastroesophageal junction [6]. In one of our earlier patients, insufficient dissection of the hernia sac led to gastric perforation during laparoscopy while pulling the stomach intra-abdominally. This experience led to a meticulous dissection of the hernial sac with extensive mediastinal dissection of the esophagus under laparoscopy, which resulted in a good intra-abdominal esophagus length in our last patient. In cases where the gastroesophageal junction is placed in the thorax, this step is important to prevent recurrence and gastroesophageal reflux symptoms.

Recurrence rate was high in our series (28.5%) compared to the Montreal series (7%) and the South Africa series (7%) [1, 4]. Recurrences were seen in type I and type III hernias with Thal fundoplication. Presence of short esophagus probably led to recurrence in type I hernia. The series is not large enough to comment on the effect of the type of fundoplication. In a series of 136 children operated for esophageal hiatal hernia, the recurrence rate was higher in Thal fundoplication as compared to Nissen-Rosetti fundoplication [10].

HH should be considered as a possibility when evaluating children with symptoms of cough, vomiting, and failure to thrive. The investigation of congenital mediastinal cysts should also include HH. In spite of the limitations of our study, after achieving an adequate intra-abdominal esophagus length, we found that the Nissen fundoplication should be preferred as the anti-reflux procedure due to its lower recurrence rates.

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Authorship Contributions: Concept – DBE; Design – AAT; Supervision – SC; Materials – MSA; Data collection and/or processing – AAT, MSA, DBE, HI; Analysis and/or interpretation – HI, SC; Literature search – DBE, MSA; Writing – DBE, AAT; Critical review – SC, HI.

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