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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: Although hiatus hernia frequently occurs in adults, it is rare in children; congenital hiatus hernia is even rarer. We describe a series of infants with congenital hiatus hernia and discuss the management problems.

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

RESULTS: Among the patients, four were females and three were males. One was prenatally diagnosed, whereas the mean age at the time of diagnosis for others was 18.6 months. Four patients had type IV hiatus hernia, two had type III, and one had type I. The diagnoses were made using chest X-ray, computed tomography, and/or upper gastrointestinal series. The hiatal repair was done in all, in two with laparoscopy. Two patients underwent Nissen fundoplication and three underwent Thal fundoplication during hiatal repair. Recurrence occurred in two patients who had undergone Thal fundoplication.

CONCLUSION: Recurrence with sliding hernias and Thal fundoplication seem more frequent in the series. If the esophagogastric junction is in the thorax, mediastinal dissection of the esophagus can be considered to have a good abdominal esophagus to prevent recurrence.

Keywords: Congenital; hiatus hernia, nissen fundoplication; short esophagus; thal fundoplication.

Hiatus hernia (HH) is a rare entity in children, although it is common in adults. 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. Although it might be secondary to anti-reflux surgery due to extensive dissection of the hiatus as is in adults, it is usually primary or congenital in children, which itself can cause gastroesophageal reflux [1, 2]. Failure to thrive, coughing, and vomiting are the most frequent complaints of patients with HH. Because of its rarity, not many series have been published regarding the pathology [1, 3, 4].

Here we describe a case series of seven infants with congenital HH and discuss the presenting symptoms, diagnosis, and treatment.

MATERIALS AND METHODS

After approval of the ethics committee for clinical research, records of patients who were diagnosed with congenital HH in two different medical facilities in neighboring cities between 2010 and 2016 were extracted from hospital records. Demographic data, presentation symptoms, diagnostic investigations, operative details, postoperative follow-up, and early and late postoperative complications were retrieved, and retrospective evaluation was performed. Late complications were stated as recurrence, continuation of reflux symptoms, and continuation of pulmonary symptoms.

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Correspondence: Dr. Ahmet Ali Tuncer. Department of Pediatric Surgery, Afyon Kocatepe University Faculty of Medicine, Afyonkarahisar, Turkey.
Tel: +90 505 948 93 71 e-mail: drtaali@yahoo.com

RESULTS

In total, four females and three males with congenital HH were included in the study from pediatric surgical departments of two different university hospitals during 6 years (Table 1) . All patients except one were diagnosed after delivery at a mean age of 18.6 months (range, 4–41 months). Coughing was the most frequent complaint, followed by vomiting and failure to thrive. However, HH was incidentally detected following trauma in one patient.

All patients were diagnosed following anteroposterior (A-P) and lateral X-rays (Figs. 1, 2), and diagnosis was confirmed with upper gastrointestinal (GI) series (Fig. 3) and/or computed tomography (CT) (Fig. 3). A thoracic cystic mass was prenatally diagnosed in one patient during the third trimester by prenatal ultrasonography. The mother was being followed up for diabetes mellitus and hypertension. Postnatal evaluation of the newborn with chest X-rays and upper GI series revealed HH.

Four patients had type IV HH, two had type III, and one had type I. In type IV HH, small bowel and colon accompanied the stomach in the hernial sac. No additional congenital anomalies were detected in any of the patients.

Laparotomy was performed in five patients and la-

Table 1. Characteristics of patients

Patients	Age at diagnosis (mo)	Gender	Presenting symptom	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 antireflux 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	?

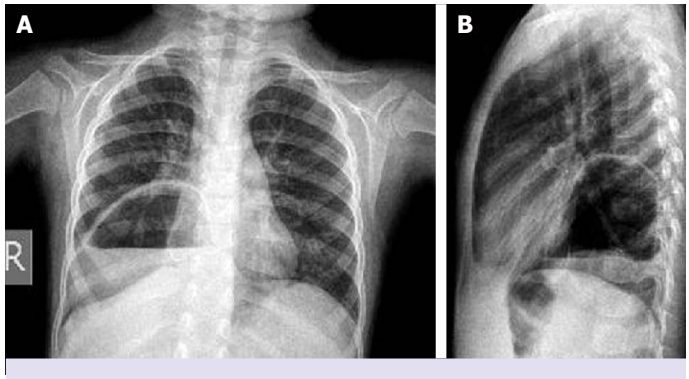

FIGURE 1. (A) A-P chest X-ray of patient 4 showing a slightly median air–fluid level on the right hemithorax. Gastric air is absent in its usual location. (B) Lateral chest X-ray of the same patient showing that the air–fluid level is placed posteriorly. This patient had type IV HH.

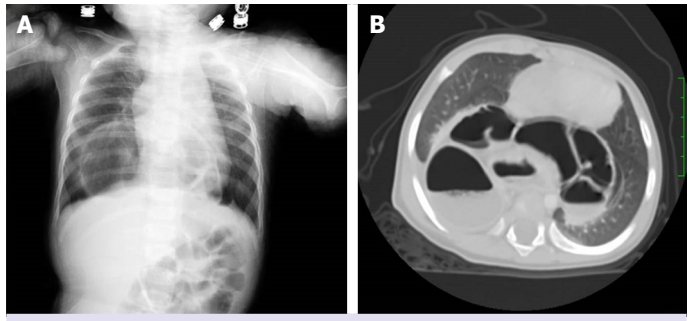

FIGURE 2. (A) A-P chest X-ray of patient 1 showing mid-thoracic air loculations resembling intestinal gas. The lesion looks like a Morgagni hernia. Gastric air is not seen at its usual location. (B) Computed tomography of the same patient shows posteriorly placed diaphragmatic defect that includes intestinal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

paroscopy in two. Laparoscopy was switched to laparotomy because of perforation of the stomach in one patient in the laparotomy group. Reduction of the viscera, excision of the hernial sac, and mobilization of the esophagus for an appropriate intraabdominal length was 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 procedure, Nissen fundoplication was preferred in two patients and Thal fundoplication was preferred in three. The mean hospitalization duration of the patients was 5.5 days postoperatively. There were no early postoperative complications.

Late postoperative complications were recurrence of hernia in two patients and continuation of reflux symptoms in two. Three patients had no complaints. One of the two patients who underwent laparoscopic repair and one patient in the open approach group had recurrence of HH (28.5%). One of the patients had an unsatisfactory intraabdominal esophagus who had undergone Thal fundoplication with hiatal repair. She was admitted with vomiting and coughing in the first postoperative month. Collis gastroplasty was not preferred because of the patient's poor nutritional status, and Nissen fundoplication and gastrostomy were added for the repair of the recurrent HH. Reflux symptoms continued for a while in this patient. The other recurrence was in a patient with type III HH in whom Thal fundoplication was added to the hiatal repair. The parents refused re-operation, and the patient was lost for follow-up. One patient had transient slow transit time probably due to vagal injury. He had motility-enhancing treatment about a year to control the reflux symptoms. Another patient had transient vomiting attacks that lasted for a few months.

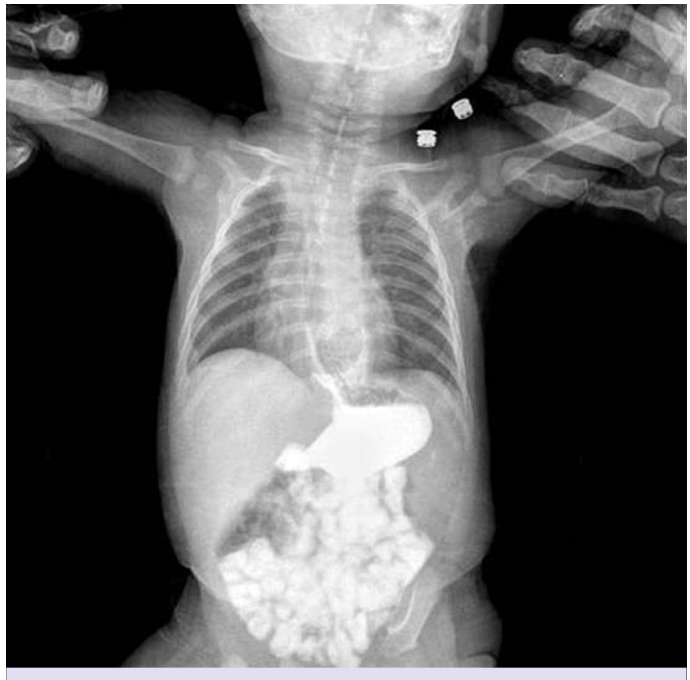

FIGURE 3. Upper gastrointestinal series of patient 2 showing thoracic replacement of gastric fundus from a medial diaphragmatic defect. This patient had type I HH.

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

DISCUSSION

HH are extremely rare pathologies in children in which the underlying reason may be congenital or acquired.

Karpelowsky et al. [1] stated in their study that HH is frequent following anti-reflux surgery in children. In a recent study by Akbulut et al., it was mentioned that HH was detected in only 0.26% (n=8) among 3081 patients who underwent esophagogastroduodenoscopy [5]. Although we have a limited number of patients in our study group, none of the patients had previously undergone anti-reflux procedure; therefore, they were considered to have congenital HH. An incidentally diagnosed trauma patient did not have a diaphragmatic trauma that could cause HH.

Four types of HH have been defined in the 2013 SAGES guidelines: type I, sliding hernia in which the gastroesophageal junction migrates above the diaphragm; type II, pure rolling or paraesophageal hernia (PEH) in which 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 the presence of other structures such as omentum, colon, or small bowel within the hernial sac [6]. Types II–IV HH are also named as PEH [1, 3]. Type I HH comprises more than 95% of cases [6]. In contrast, 57% of the patients in this series had type IV HH.

Presenting findings including coughing, vomiting, failure to thrive, and pneumonia were consistent with those reported in the literature. Particularly in type IV HH, 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 seen in one of our patients. Failure to thrive was also seen in another patient with type I HH due to frequent vomiting. HH 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 such complications in our series.

Chest X-rays that were taken during the evaluation of cough or gastroesophageal symptoms led to the suspicion of HH. Simply A-P and lateral chest X-rays may show HH; however, because large and small bowel will be protruded through the thorax as well as the stomach, type IV HH may easily be confused with posterolateral diaphragmatic hernias. If the hernia is on the medial side in 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 seen in one of the initial patients in this series. An upper GI series or contrast-enhanced CT of the thorax can confirm the diagnosis. In our series, although five patients in the study group had CT scans, all patients had a fluoroscopic investigation. Therefore, we believe that to corroborate HH suspicion in A-P and lateral chest X-rays, fluoroscopic GI series is satisfactory for the diagnosis. With our current experience, CT alone is also satisfactory for diagnosis because it shows the anatomy in both sagittal and coronary views and additionally shows the mediastinum and lung lesions in the patient. Considering the radiation burden on children, only one of these radiological investigations should be preferred, depending on the experience of the radiologist and surgeon [4].

Prenatal diagnosis of 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 in as early as the second trimester [4, 8, 9]. Diagnosis was established prenatally in one of the patients in this series. Preliminary ultrasonography diagnosis was thoracic cyst in the third trimester. Postnatal chest X-rays and GI series confirmed the diagnosis of HH.

Surgical repair of HH has been recommended in children in the recent SAGES guidelines [6]. Laparotomy or laparoscopy can be preferred depending on the size of the patient, size of the hernia, and experience of the surgical team. In our series, there were four repairs with laparotomy, one conversion to laparotomy, and two laparoscopic repairs. The number of patients is not enough for an objection ; however, we believe that laparoscopic repair is also successful for exhibiting the real anatomy and also further dissection in the mediastinum. Moreover, although there are several approaches for additional anti-reflux procedures, anti-reflux operation is a part of HH repair [1, 4]. The anti-reflux operation was performed in our series except for two early patients . These both had type IV HH with intraabdominal gastroesophageal junction, and only the hiatal defect was repaired. We did not see recurrence of HH or persistent 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 an abdominal esophagus length shorter than 2–3 cm in adults of different series [6]. This description is not clear in children. It may be

associated with HH as seen in one of our patients. This patient had a recurrence and underwent a re-operation with extensive mobilization of the esophagus, hiatal 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 because of 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 hernial sac led to gastric perforation during laparoscopy while pulling the stomach intraabdominally. This experience led to a meticulous dissection of the hernial sac with extensive mediastinal dissection of the esophagus by laparoscopy, which resulted in a good intraabdominal esophagus length in our last patient. If the gastroesophageal junction is placed in the thorax, this step seems to be important to prevent recurrence and gastroesophageal reflux symptoms.

Recurrence rate was high in our series (28.5%) compared with that in the Montreal series (7%) and South African series (7%) [1, 4]. Recurrences were in type I and type III HH with Thal fundoplication. Presence of short esophagus probably led to recurrence in patients with type I HH. This series is small to comment on the effect of the type of the fundoplication. In a series of 136 children operated for esophageal HH, the recurrence rate was higher for Thal fundoplication than for Nissen–Rosetti fundoplication [10].

HH should be kept in mind for children who present with symptoms of coughing, 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 intraabdominal esophagus, Nissen fundoplication should be preferred as the anti-reflux procedure because of its lower recurrence rates.

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