Eur J Pediatr (1993) 152:418–420

European Journal of
Pediatrics

Association of hiatus hernia with asplenia syndrome

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Received: 26 February 1992 / Accepted: 9 September 1992

Abstract. During a 13-month period, 13 patients with asplenia syndrome were evaluated with MRI for cardiovascular and visceral anomalies. The MR images were reviewed for the presence of hiatus hernia which was found in three patients. One of the remaining ten patients with no MRI evidence of hiatus hernia was diagnosed as having gastro-oesophageal reflux and hiatus hernia by an oesophagogram and 24-h pH monitoring. This patient had undergone fundoplication prior to MRI. Out of the 13 patients (31%) with asplenia syndrome, 4 had hiatus hernia. It appears that among patients with the asplenia syndrome, hiatus hernia is a frequent finding. Recurrent pneumonia or bronchiolitis in patients with asplenia syndrome requires evaluation for the presence of hiatus hernia and gastro-oesophageal reflux.

Key words: Asplenia syndrome – Hiatus hernia – Magnetic resonance imaging

Introduction

Hiatus hernia in children is a congenital defect in which part of the stomach protrudes into the thoracic cavity through the oesophageal hiatus of the diaphragm and has a prevalence of 0.1% [6]. Although most patients are asymptomatic, some manifest during infancy and childhood [3]. The symptoms of hiatus hernia are related to the associated gastro-oesophageal reflux [4, 5, 7, 11, 12, 15].

We observed a female infant with asplenia syndrome who had recurrent bronchiolitis for 10 months. After gastro-intestinal bleeding occurred at 11 months of age, she underwent an oesophagogram and 24-h pH monitoring. Hiatus hernia with gastro-oesophageal reflux were diagnosed. Her condition improved following fundoplication. This observation prompted a prospective study to evaluate a possible association between asplenia syndrome and hiatus hernia.

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Patients and methods

From June 1990 to June 1991, 13 patients with asplenia syndrome were admitted to our institution for cardiovascular evaluation. Of these, 9 were boys and 4 were girls whose ages ranged from 7 days to 15 years (median 6 months). All the 13 patients were submitted to MRI to study the cardiovascular and visceral anomalies.

The diagnosis of the asplenia syndrome was made based on juxtaposition of abdominal great vessels on echocardiography [13], Howell-Jolly bodies on a peripheral blood smear, bilateral morphological right bronchi on high kilovoltage filtered chest films [9], and bilateral right atrial appendages on the angiogram.

The scanner used was a General Electric imaging system (1.5 Tesla). Spin-echo technique was applied. The echo delay time (TE) used ranged from 20 to 38 ms. A combination of coronal, sagittal and axial sections was routinely performed. In reviewing the MR images special attention was paid to the detection of a hiatus hernia. In two patients, both barium meal study and 24-h pH monitoring were performed.

Results

The clinical information of all 13 patients is summarised in Table 1. MR images demonstrating protrusion of part of the stomach into the thoracic cavity through the oesophageal hiatus indicated the presence of a hiatus hernia in three patients (Fig. 1). Another patient (case 1), who was diagnosed as having hiatus hernia and gastro-oesophageal reflux by oesophagogram and 24-h oesophageal pH monitoring and underwent fundoplication prior to the MR imaging study, displayed no evidence of hiatus hernia on MRI. Including this patient, 4 out of 13 patients (31%) with asplenia syndrome had hiatus hernia.

On plain X-ray films none of the patients had evidence of intestinal obstruction. Barium meal studies confirmed the presence of hiatus hernia in two symptomatic patients who had gastro-oesophageal reflux documented by 24-h pH monitoring (cases 1, 2). Congenital absence of the spleen could be demonstrated by MRI in all patients. Transverse liver was present in 12 patients. Two patients (cases 1, 2) underwent fundoplication with marked improvement. Of the remaining two patients, one (case 4) who had past history of frequent bronchio-

Table 1. A summary of clinical information of 13 patients studied with MRI

Case	Age	Sex	Cardiovascular anomalies	HH	GER	FP
1.	1 year	F	Common atrium, AVC, DORV, PS, Bilateral SVC	+	+	+
2.	1 month	M	Common atrium, AVC, DORV, PS	+	+	+
3.	1 week	M	Common atrium, AVC, TGA PS, Bilateral SVC, TAPVC	+	?	–
4.	4 years	F	Common atrium, AVC, DORV, PS, TAPVC	+	?	–
5.	4 months	M	Common atrium, DIRV DORV, PS, TAPVC	–	–	–
6.	1 month	F	Common atrium, AVC, DORV, PS, TAPVC	–	–	–
7.	1 month	F	Common atrium, DIRV, DORV PS, TAPVC, Bilateral SVC	–	–	–
8	15 years	M	ASD, AVC, DORV, PS, Bilateral SVC	–	–	–
9.	5 years	M	AVC, TGA, PS, post Fontan operation	–	–	–
10.	5 years	M	Common atrium, DIRV, DORV, PS	–	–	–
11.	1 month	M	Common atrium, AVC, DORV, PS	–	–	–
12.	1 month	M	AVC, DORV, PS, TAPVC Bilateral SVC	–	–	–
13.	10 years	M	AVC, DORV, PS, TAPVC, Bilateral SVC	–	–	–

AVC, Atrioventricular canal; DIRV, double inlet right ventricle; DORV, double outlet right ventricle; FP, fundoplication; GER, gastro-oesophageal reflux; HH, hiatus hernia; PS, pulmonary stenosis; SVC, superior vena cava; TAPVC, total anomalous pulmonary venous connection; ?, not confirmed

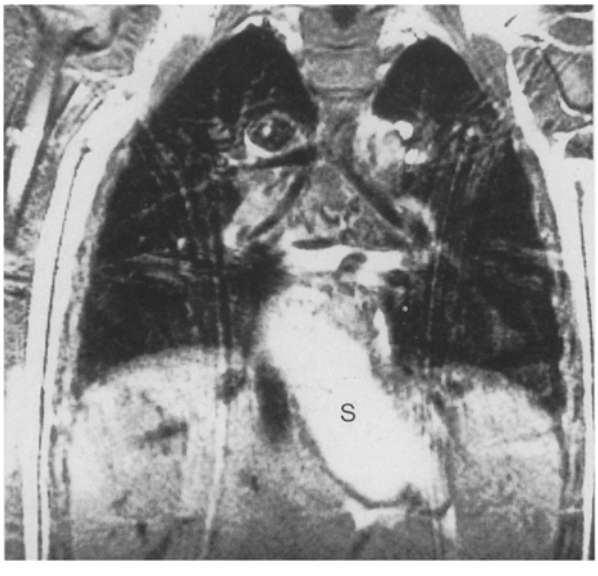

Fig. 1. A coronal section of a patient shows protrusion of part of the stomach *(S)* into the thorax. The stomach was located at the midline

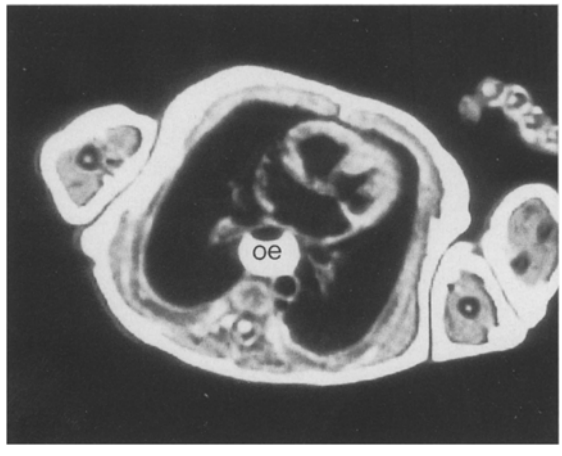

Fig. 2. An axial section of a 1-month-old male baby reveals a markedly dilated oesophagus *(oe)* mimicking achalasia. There is gastric content in the oesophagus indicating the presence of gastro-oesophageal reflux. An ambiguous atrioventricular connection via a common atrioventricular valve and a common atrium are also demonstrated

litis in infancy, died at age 4 during cardiac surgery for total anomalous pulmonary venous connection. The other (case 3) died at 1 month of age from respiratory failure with an obstructed total anomalous pulmonary venous connection. The gastro-oesophageal reflux may have been responsible for, or contributed to, the chronic respiratory problem in both of these patients, however, the potential relationship could not clearly be established.

Discussion

Asplenia syndrome is usually associated with complex cardiovascular and visceral anomalies [14, 20, 23]. The prognosis of affected patients has primarily been determined by the complexity of these anomalies. With the advent of improved cardiac surgery, most cardiac defects associated with the asplenia syndrome can now be palliated or corrected [12]. As the life span of these patients has increased, other associated anomalies and their complications have been increasingly recognised such as absence of the spleen, genito-urinary anomalies, malrotation of the gut, tubular stomach and gastric volvulus [1, 2, 4, 10, 17,19, 21, 24].

The incidence of hiatus hernia was underestimated in patients with asplenia syndrome, since the clinical course of most patients is dominated by cardiovascular anomalies or infection, and less attention may have been paid to the presence of hiatus hernia. Hiatus hernia has been sporadically reported to be associated with the asplenia syndrome in pathological studies [8, 19, 20], however, this association has never been reported in living asplenia syndrome patients.

Opitz and Gilbert [18] suggested that the midline of the embryo is an important developmental field and its disturbance may result in the defect of laterality of midline organs. Asplenia and polysplenia syndrome are midline developmental field defects. Heterotaxy syndrome has been used to refer this spectrum of anomalies with a lateralisation defect [23]. The hiatus hernia may be part of the asplenia syndrome/malformation spectrum.

Diagnosis of hiatus hernia in infants has generally been dependent upon obtaining a positive barium meal radiogram [22]. Recently, ultrasonography and computerised tomogoraphy were applied to assess the presence of hiatus hernia [16, 25]. In our study, MRI was used to document the presence of hiatus hernia.

The incidence of hiatus hernia in the asplenia syndrome appears to be higher than that in the normal population (31% vs. 0.1%) [6]. Recurrent pneumonia, bronchiolitis, and feeding difficulties were usually attributed to the cardiac defects or compromised immunological function due to asplenia syndrome, however, the presence of hiatus hernia with gastro-oesophageal reflux may also contribute to these problems. We recommend that patients with asplenia syndrome undergo a thorough evaluation for the presence of gastro-oesophageal reflux and hiatus hernia [4].

Acknowledgement. We express our thanks to Dr. Debra Kearney for reviewing this manuscript.

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