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Down's syndrome as a factor in the diagnosis, management, and outcome in patients of Morgagni hernia

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Abstract

Purpose: The purpose was to study the impact of Down's syndrome (DS) in the diagnosis, management, and outcomes of patients with Morgagni hernia (MH).

Methods: Twenty-two (22) patients with MH treated at a tertiary center were retrospectively studied for history, findings, associated anomalies, referral diagnoses, hospital admissions, radiological procedures for diagnosis, age at which operated on, operative procedure, complications, and recurrences. Eleven did not have DS (group 1); 11 others had associated DS (group 2). The ages at operation were compared in the 2 groups using the Mann-Whitney test. More than 3 hospital admissions for symptoms and signs relevant to MH before a diagnosis of MH were considered a "delayed diagnosis."

Results: Twenty-two patients (7 females, 15 males) aged 3 months to 10 years were seen. They presented with respiratory distress (n = 16), with vomiting (n = 5), with intestinal obstruction (n = 1), by serendipity (n = 2), and with recurrence from another hospital (n = 1). The mean age of group 1 was 14.5 months, and that of group 2 was 29.18 months; the difference was not significant ($P = .621$). Nine patients of group 2 were "delayed diagnosis" compared with 2 in group 1. Both delays from group 1 had severe associated anomalies. All patients underwent operative correction (17 open and 5 laparoscopic repairs). Two had recurrences, one operated on by the open method by us and another laparoscopically by the Lima technique at another center. Both had DS. Both were reoperated on by the open method.

Conclusions: The diagnosis of MH may be strikingly delayed when associated with DS or other severe congenital anomalies. Morgagni hernia should be strongly considered in patients with DS admitted repeatedly for chest infections. Chest x-rays in 2 planes may avoid misdiagnosis of MH. Both open and laparoscopic methods have proven satisfactory as operative treatment of MH. Recurrences were seen in patients with DS, which may be corrected by laparotomy or laparoscopically. We feel that resecting the sac and approximating the posterior lip of the defect to the anterior abdominal wall, whether in open or laparoscopic methods, may give stronger repairs, which may avoid recurrence.

Morgagni hernias (MHs) occur as a result of a defect between the xiphisternal and costal margin fibers of the diaphragm, both of which insert into the central tendon of the diaphragm. Morgagni hernias are uncommon in children [1].

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Their relative rarity and presentation with nonspecific symptoms and signs lead to delays in diagnosis and treatment [2]. Morgagni hernia is frequently associated with other congenital anomalies, chief among them is Down's syndrome (DS) [3,4]. It is unclear if patients with DS and MH behave differently from those who have no DS and if DS contributes to delayed diagnosis of MH. Studies suggest that both DS and MH are more common in Saudi Arabia [2,5,6]. This was borne out in the evaluation of our material, where half the patients of MH seen by us at a tertiary center in southwestern Saudi Arabia in the last 15 years had associated DS.

This article studies the impact of DS and other associated anomalies on the diagnosis, management, and outcomes of MH. It also addresses the operative treatment of MH and the impact of laparoscopic repairs in the operative treatment. Management in the event of a recurrence, which does not find much mention in the literature, is also addressed.

1. Patients and methods

All patients treated with a diagnosis of MH at Asir Central Hospital between January 1994 and December 2008 were studied. Asir Central Hospital serves the southern region of Saudi Arabia and is a tertiary care center and teaching hospital. Patients were divided into 2 groups: those with (group 2) and those without DS (group 1). Their records were retrospectively analyzed for demographics, presenting features, referral diagnoses, other associated congenital anomalies, investigations, and management including operative procedures and outcome of management.

The age at which the patients came to operation was compared in the 2 groups using the Mann-Whitney test (nonparametric test of significance) at 5% level of significance.

Delay in diagnosis was defined by ascertaining the number of admissions a patient underwent for symptoms pertinent to MH before being diagnosed as having MH. If a patient was admitted, say, for an injury, this was excluded from the number of admissions. Patients with more than 3 such hospital admissions were deemed to have a delayed diagnosis.

Open repairs were carried out by reduction of contents; resection of sac; and approximation of the edges with nonabsorbable, interrupted mattress sutures. Laparoscopic repairs were done by the technique described by Azzie et al [7]. The contents of the hernial sac were reduced, and the sac was resected. The approximating U-shaped sutures were

Table 1 The demographic features, referring diagnoses, associated anomalies, and delays in treatment

SN	Age	Sex	Presentation	Referring diagnoses	Associated anomalies	Delay
1	3 mo	F	RCI, poor feeding, failure to thrive, constipation	Eventration diaphragm, GER	Dysmorphic, CDH, arthrogryposis multiplex congenita, WHD	+
2	14 mo	F	Rt inguinal hernia; RCI (in past)	Nil	Rt inguinal hernia	−
3	19 mo	F	B/L inguinal hernia; RCI (in past)	Nil	B/L inguinal hernia	−
4	3 y 8 mo	M	Intestinal obstruction	Intestinal obstruction	Short stature, plagiocephaly, atrophied left frontal lobe, hypertelorism, low-set ears	−
5	12 mo	M	RCI	Dyspnea, RCI	Rt inguinal hernia	−
6	7 mo	M	RCI, poor feeding	Cardiomegaly	Nil	−
7	9 mo	F	RCI	Diaphragmatic hernia	Nil	+
8	2 y	F	Dyspnea, cough	Chest infection	Nil	−
9	12 mo	M	Respiratory distress	Diaphragmatic hernia	Nil	−
10	9 mo	M	RCI	Bronchiolitis	Nil	−
11	9 mo	F	RCI	-do-	Nil	−
12	4 y 6 mo	F	RCI	Abnormal cXR	DS	+
13	8 mo	M	RCI, intermittent vomiting	–	DS	−
14	8 mo	F	RCI	–	DS	+
15	10 mo	M	RCI	Pneumopericardium	DS	+
16	4 mo	M	RCI	RCI	DS, patent urachus	+
17	1 y 8 mo	M	Vomiting, aspiration after recurrence of MH	Diaphragmatic paralysis after cardiac operation	DS, ASD, congenital dysplastic hip, CTEV	+
18	10 y 2 mo	M	RCI	RCI, cardiomegaly	DS	+
19	5 y	M	GERD, failure to thrive	GERD	DS, duodenal web	+
20	1 y 3 mo	M	RCI	RCI	DS, pectus carinatum	+
21	1 y	M	RCI	RCI	DS, ASD	+
22	8 mo	M	RCI	Diaphragmatic hernia	DS,	−

Patient numbers 1 through 11 do not have DS (group 1). Patients 12 through 22 have MH and DS (group 2). RCI indicates recurrent chest infection; SN, serial number; B/L, bilateral; DS, Down's syndrome; cXR, chest x-ray; MH, Morgagni hernia; ASD, atrial septal defect; GER, gastroesophageal reflux; GERD, gastroesophageal reflux disease; CDH, congenital dysplasia of hip; WHD, Werdnig-Hoffman disease; CTEV, congenital talipes equinovarus.