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Spectrum of Esophageal Disorders in Children with Chest Pain

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The charts of 83 children with chest pain who underwent esophageal manometry followed by esophagogastroscopy were reviewed. Forty-seven (57%) had normal esophageal histology and normal motility (group I). Esophagitis and normal motility were demonstrated in 15 children (group II), normal esophageal histology and esophageal dysmotility in 13 (group III), and both esophagitis and abnormal motility in 8 (group IV). Diffuse esophageal spasm and achalasia were the most common motility disorders identified (in seven and four patients, respectively). The presence and duration of symptoms, the age, and the gender were not different among the four patient groups. After six months of H_2-receptor blockade, 12 of 15 group II patients were asymptomatic, whereas a significantly smaller percentage (five of 18) of patients with abnormal esophageal motility responded to esophageal dilation or treatment with calcium channel blockade, H_2-receptor antagonist, and/or prokinetic agents ($P < 0.01$). These data suggest that the evaluation of children with chest pain should include esophageal motility testing and esophagoscopy, even in the absence of other gastrointestinal-associated symptoms, and that while treatment of esophagitis results in resolution of symptoms, motility disorders were relatively refractory to therapy.

KEY WORDS: chest pain; esophagitis; esophageal dysmotility.

Chest pain in adults often indicates the presence of atherosclerotic heart disease. Approximately 50% of adult patients with chest pain will have coronary artery disease found at angiography. In patients with angiographically normal coronary arteries, 30–40% will have esophagitis and/or abnormal esophageal motility subsequently identified as the cause of their symptoms (1–6). In contrast, the incidence of cardiac disease in children with chest pain is low, ranging from 0.4% to 4% (7, 8), and the majority of pediatric patients are diagnosed as having either idiopathic or functional chest pain. Selbst et al (7) reported that the incidence of gastrointestinal disease in children with chest pain is approximately 4% compared to about 30% as found in adults (1–6).

Recently, Berezin et al reported that in 17 of 27 adolescents with "idiopathic" chest pain, esophagitis or abnormal esophageal motility was the cause of their symptoms (9). In this report, we extend these observations concerning the relationships between abnormal esophageal motility, esophagitis, and chest pain in pediatric patients and describe both the spectrum of motility disorders found in these patients and the efficacy of treatment.

MATERIALS AND METHODS

We reviewed the charts of 83 patients between 1 and 20 years of age with chest pain and dysphagia or vomiting who were referred to the Division of Pediatric Gastroen-

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TABLE 1. CHARACTERISTICS OF 83 ADOLESCENTS AND CHILDREN WITH CHEST PAIN AND/OR DYSPHAGIA

	Group I (N = *47*)	*Group II* (N = *15*)	*Group III* (N = *13*)	*Group IV* (N = *8*)
Age (years) (mean ± SEM)	12.7 ± 0.76	12.7 ± 1.1	11.3 ± 1.4	12.2 ± 2.0
Male/Female	23/24	6/7	9/4	4/4
Duration of Symptoms (months) (mean ± SEM)	20.6 ± 3.6	21.1 ± 4.1	34.9 ± 8.6	39.2 ± 20
Symptoms at presentation [number of patients (%)]				
Chest pain	39 (83)	13 (87)	11 (84)	7 (87)
Dysphagia	10 (21)	7 (47)	6 (46)	3 (37)
Vomiting	18 (38)	10 (67)	9 (69)	3 (37)

terology at New York Medical College between July 1987 and June 1991 for evaluation, which included cardiorespiratory, neurologic and abdominal physical examinations, electrocardiogram, chest x-ray, esophagogastroduodenoscopy, and esophageal motility testing.

Esophageal manometry was performed using a four-lumen polyvinyl catheter assembly (individual outer diameter 1.2 mm) with three pressure-sensing orifices separated by 5 cm. The catheter was perfused with sterile water at a rate of 0.5 ml/min by a low-compliance pneumatic hydraulic capillary infusion system, with a pressure response greater than 800 mm Hg/sec. Pressures were transmitted via the fluid-filled catheters to transducers connected to a three-channel polygraph recording system. The station pull-through technique was used to determine the lower esophageal sphincter pressure and location (10). For evaluation of esophageal peristalsis, 10 wet swallows (5 cc water each) were given, separated by 30-sec intervals. Esophageal motility abnormalities were classified according to the criteria of Richter et al (10) for adult patients. Nutcracker esophagus was defined by average peristaltic pressures (mean of 10 wet swallows) >180 mm Hg and of prolonged duration (>6 sec) with normal peristaltic progression. Lower esophageal pressures <10 mm Hg or >45 mm Hg were considered outside the normal range (11).

Esophagogastroduodenoscopy was subsequently performed using an Olympus GIF-XP10 gastroscope (Olympus Co., Lake Success, New York) after informed consent was obtained, and intravenous sedation with meperidine and diazepam was administered. Esophagitis was endoscopically defined by friability, erythema, ulceration, or granularity of the lower esophagus. In all patients, an esophageal biopsy specimen was taken at least 7–10 cm above the gastroesophageal junction (*Z* line) under direct visualization. Esophagitis was confirmed histologically according to established criteria (12).

Statistical significance was determined using analysis of variance, comparing all four patient groups and chi-squared analysis comparing response to treatment in groups 2, 3, and 4. A P value <0.05 was considered statistically significant.

RESULTS

In 47 of 83 patients (56.6%), esophagogastroduodenoscopy and esophageal manometry testing were normal (group I). Histologic evidence of esophagitis was found in 15 children (18%) with normal esophageal motility (group II), and an additional 13 patients (15.6%) had abnormal esophageal motility and normal distal esophageal histology (group III). Eight patients (9.6%) had both distal esophagitis and esophageal dysmotility (group IV). There were no significant differences in age, gender, or the location, character, or duration of symptoms among the four patient groups (Table 1).

The manometric diagnoses of the 21 patients (25.3%) with abnormal esophageal motility are shown in Figure 1. Diffuse esophageal spasm was diagnosed in seven patients; achalasia in four; hypotensive lower esophageal sphincter in three; aperistalsis of the distal esophagus in three; nutcracker esophagus in two; and hypertensive lower esophageal sphincter in two.

Twelve of 15 (80%) children with esophagitis and normal motility had complete resolution of chest pain and were able to discontinue therapy after six months of treatment with an H_2-receptor antagonist. The three additional group II patients required continued treatment for one year and remained

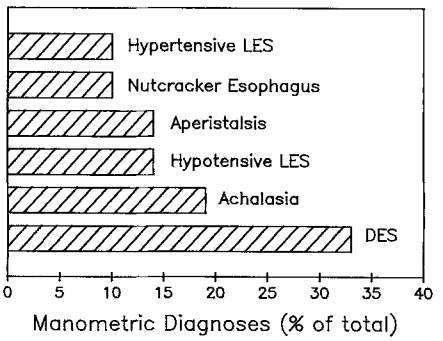

Fig 1. Distribution of esophageal motility disorders in pediatric patients with chest pain. LES = lower esophageal sphincter; DES = diffuse esophageal spasm. All data are shown as percentage of the total number of patients (21).

TABLE 2. RESPONSE TO THERAPY OF CHILDREN WITH ABNORMAL ESOPHAGEAL MOTILITY*

Patient	*Age*	*Diagnosis*	*Therapy*	*Outcome*
1	13	DES	CCB	Asymptomatic
2	20	DES	CCB	Symptomatic
3	13	DES	CCB	Symptomatic
4	15	DES	CCB	Symptomatic
5	10	DES/E	R	Symptomatic
6	15	DES/E	R	Symptomatic
7	13	DES/E	R/CCB	Symptomatic
8	10	ACH/E	DIL/R	Asymptomatic
9	4	ACH	DIL	Asymptomatic
10	12	ACH	DIL	Asymptomatic
11	12	ACH/E	CCB/DIL/R	Symptomatic
12	9	HYPO	R/MET	Symptomatic
13	5	HYPO	R/MET	Symptomatic
14	20	HYPO/E	R/MET	Symptomatic
15	13	APER	None	Symptomatic
16	11	APER/E	R	Symptomatic
17	7	APER/E	R	Symptomatic
18	18	NUT	None	Symptomatic
19	12	NUT	None	Symptomatic
20	3	HYPER	DIL	Symptomatic
21	10	HYPER	DIL	Asymptomatic

*Abbreviations: DES, diffuse esophageal spasm; ACH, achalasia; HYPO, hypotensive lower esophageal sphincter; APER, aperistalsis; NUT, nutcracker esophagus; HYPER, hypertensive lower esophageal sphincter; E, esophagitis; CBB, calcium-channel blocker (10 mg 30 min before meals); R, ranitidine (150 mg twice daily); DIL, esophageal dilatation; MET, metaclopramide (0.15 mg/kg 30 min before meals and at bedtime).

asymptomatic while on therapy. In contrast, a significantly smaller (five of 18, $P < 0.01$) number of patients with abnormal esophageal motility (groups III and IV) had resolution of chest pain after therapy (Table 2).

DISCUSSION

Recent studies in adults with noncardiac chest pain show that 30–40% have gastroesophageal reflux and/or abnormal esophageal motility as a cause of their symptoms (1–6). In a study of adolescents with "idiopathic" chest pain, Berezin et al (9) reported that 17 of 27 (63%) adolescents who underwent gastrointestinal evaluation had esophagitis and showed that, in 16, resolution of symptoms was temporally associated with histological normalization of the esophageal lesion.

Our experience in children with mitral value prolapse (13) or asthma (14) with chest pain indicates that 75% have esophagitis and that resolution of symptoms will follow healing of esophageal inflammation. Therefore, it appears that esophageal disorders often cause chest pain in children with primary cardiac and pulmonary disease. The data presented in this report extend these observations in an additional 83 patients and show that 44% of children referred for evaluation of chest pain have an esophageal etiology of their symptoms. To our knowledge, this is the first report to describe the presence of nutcracker esophagus in pediatric patients, the spectrum of esophageal motility disorders causing chest pain in children, and to evaluate therapeutic efficacy in children with esophageal dysmotility.

Our data in children with noncardiac chest pain are similar to studies in adult patients (1–6, 15, 16). Janssens et al (16) identified an esophageal etiology in 48% of their adult patients with noncardiac chest pain and found abnormal motility in 58%, gastroesophageal reflux in 30%, and both esophageal dysmotility and gastroesophageal reflux in 11%. Nutcracker esophagus was the predominant motor disorder, found in 48% of patients with esophageal dysmotility, and diffuse esophageal spasm was identified in only 10% of their adult population (15, 16). In contrast to these findings, diffuse esophageal spasm was the most common dysmotility identified in our patient population, accounting for 33% of the children with abnormal esophageal motility, and nutcracker esophagus was found in 10%. These differences are may represent different stages in the development of smooth muscle dysfunction.

In the present study, 80% of patients with chest pain and esophageal inflammation had resolution of their symptoms after treatment with H_2 antagonists, whereas 73% of children with primary esophageal motility disorders continued to have chest pain and/or dysphagia despite treatment of their esophagitis and dysmotility. The response to esophageal dilation and H_2 antagonists seen in three of our four patients with achalasia is similar to that described by Boyle et al (17) and Berquist et al (18), who successfully treated 80% of children with achalasia by pneumatic dilation of the lower esophageal sphincter. Successful treatment of children with achalasia using calcium-channel blockers has been reported by Maksimak et al (19). In contrast, only 14% of our patients with other esophageal motor disorders responded to therapy with calcium-channel antagonists or prokinetic agents. In a report describing the efficacy of treatment in children with diffuse esophageal spasm, Milov et al (20) found that sublingual administration of nitrates controlled symptoms in three of five patients but was discontinued because of drug-associated postural hypotension. In all five patients, symptoms resolved over two years without specific therapy. Because of

the limited experience in treating children with esophageal dysmotility, additional studies are needed.

Chest pain is a frequent concern among adolescents (21), and accounts for approximately 5% of outpatient clinical visits in this age group (22). Unlike adult patients, the majority of children with chest pain are found to be free of cardiac disease (23). In adolescents with chest pain, Selbst (24) and Pantell and Goodman (25) reported that 48% and 59%, respectively, had either functional or idiopathic pain. Patients under 12 years of age were more likely to have an organic etiology of their complaint, whereas adolescents were more likely to have a psychologic cause of their symptoms. However, both studies were performed in pediatric emergency departments and did not include children who were referred for gastrointestinal evaluation. Because clinical presentation appears to be of little predictive value in identifying children likely to have esophageal disease, the true incidence of gastrointestinal disorders causing chest pain may be higher than 4% as previously reported by Selbst et al (7).

In summary, we have shown that in a significant percentage of children with chest pain, an esophageal etiology can be identified using esophageal motility testing and esophagogastroduodenoscopy. Further, while treatment of esophagitis results in resolution of chest pain in a large percentage of children, esophageal motility disorders remain relatively refractory to treatment.

REFERENCES

1. McCallum RW: The spectrum of esophageal motility disorders. Hosp Pract 22:71–83, 1987
2. DeMeester TR, O'Sullivan GC, Bermudez G, Midell AI, Cimochowski G, O'Drobinak J: Esophageal function in patients with angina-type chest pain and normal coronary angiograms. Ann Surg 196:488–498, 1982
3. Richter JE, Bradley LA, Castell DO: Esophageal chest pain: Current controversies in pathogenesis, diagnosis and therapy. Ann Intern Med 110:66–78, 1989
4. Peters L, Maas L, Petty D, Dalton C, Penner D, Wu WC, Castell DO, Richter J: Spontaneous noncardiac chest pain: Evaluation by 24-hour ambulatory esophageal motility and pH monitoring. Gastroenterology 94:878–886, 1988
5. Breumelhof R, Nadorp JHSM, Akkermans LAM, Smout AJP: Analysis of 24-hour esophageal pressure and pH data in unselected patients with noncardiac chest pain. Gastroenterology 99:1257–1264, 1990
6. Benjamin SB, Richter JE, Cordova CM, Knuff TE, Castell DO: Prospective manometric evaluation with pharmacologic provocation of patients with suspected esophageal motility dysfunction. Gastroenterology 84:893–901, 1983
7. Selbst SM, Ruddy RM, Clark BJ, Henredig FM, Santulli T Jr: Pediatric chest pain: A prospective study. Pediatrics 82:319–323, 1988
8. Brenner JI, Ringel RE, Berman MA: Cardiologic perspectives of chest pain in childhood: a referral Problem? Pediatr Clin North Am 31:1241–1258, 1984
9. Berezin S, Medow MS, Glassman MS, Newman LJ: Chest pain of esophageal origin. Arch Dis Child 63:1457–1460, 1988
10. Richter JE, Wu WC, Johns DN, Blackwell JN, Newson JL, Castell JA, Castell DO: Esophageal manometry in 95 healthy adult volunteers: Variability of pressure with age and frequency of abnormal contractions. Dig Dis Sci 32:583–592, 1987
11. Castell DO, Richter JE, Dalton CB, (eds). Esophageal Motility Testing. New York, Elsevier, 1987
12. Behar J, Sheahan DC: Histologic abnormalities in reflux esophagitis. Arch Pathol 99:387–391, 1975
13. Woolf PK, Gewitz MH, Berezin S, Medow MS, Stewart JM, Fish BG, Glassman MS, Newman LJ: Noncardiac chest pain in adolescents and children with mitral valve prolapse. J Adolesc Health Care 12:1–4, 1991
14. Berezin S, Medow MS, Glassman MS, Newman LJ: Esophageal chest pain in children with asthma. J Pediatr Gastroenterol Nutr 12:52–55, 1991
15. Katz PO, Dalton CB, Richter JE, Wu WC, Castell DO: Esophageal testing of patients with noncardiac chest pain or dysphagia: Results of three years' experience with 1161 patients. Ann Intern Med 106:593–597, 1987
16. Janssens J, Vantrappen G, Ghillebert G: 24-hour recording of esophageal pressure and pH in patients with noncardiac chest pain. Gastroenterology 90:1978–1984, 1986
17. Boyle JT, Cohen S, Watkins JB: Successful treatment of achalasia in childhood by pneumatic dilation. J Pediatr 99:35–40, 1981
18. Berquist WE, Byrne WJ, Ament ME, Fonkalsrud EW, Euler AR: Achalasia: Diagnosis, management and clinical course in 16 children. Pediatrics 71:798–805, 1983
19. Maksimak M, Perlmutter DH, Winter HS: The use of nifedipine for the treatment of achalasia in children. J Pediatr Gastroenterol Nutr 5:883–886, 1986
20. Milov DE, Cynamon HA, Andres JM: Chest pain and dysphagia in adolescents caused by diffuse esophageal spasm. J Pediatr Gastroenterol Nutr 9:450–453, 1989
21. Brunswick AP, Bovie JM, Tanca C: Who sees the doctor? A study of urban black adolescents. Soc Sci Med 13A:45–46, 1979
22. Brown RT: Chostochondritis in adolescents. J Adolesc Health Care 1:198–201, 1981
23. Fife DA, Moodie DS: Chest pain in pediatric patients presenting to the cardiac clinic. Clin Pediatr 23:321–324, 1984
24. Selbst SM: Chest pain in children. Pediatrics 75:1068–1070, 1985
25. Pantell RH, Goodman BW Jr: Adolescent chest pain: A prospective study. Pediatrics 71:881–887, 1983