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ACHALASIA OF THE ESOPHAGUS IN A SMALL URBAN COMMUNITY*

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Achalasia of the esophagus has been considered a rare disease of unknown cause. In 1674, Willis[1] described the first patient who had a dilated esophagus without an organic esophageal stricture. In 1900, Neumann assessed the number of cases in the medical literature as 70. Since then, many thousands of cases have been reported, but the true incidence of the disease is not known. Current estimates have been derived from hospital series or other selected statistics. The lack of satisfactory population denominators for most of these studies has impaired their value for comparison with other populations where incidence is considered unusual, for example, in Brazil where the increased frequency of a dilated esophagus is allegedly due to Chagas' disease.

The purpose of this study is to provide a useful set of base-line statistics for achalasia of the esophagus in a population in which there is no preconceived notion of any unusual frequency of this disorder and one that is subject to a unique medical and record retrieval system.

METHODS

The records were reviewed of all patients who were residents of Rochester, Minn., and who had a diagnosis of achalasia of the esophagus, mega-esophagus, esophageal spasm, cardiospasm, cardiac spasm, or pylorospasm. These were records of the Mayo Clinic and of other medical institutions in the community serving local residents during the past 30 years. Details of the medical resources and a newly established record retrieval system for this community have been described.[2] Any person known to have moved to Rochester merely to facilitate the diagnosis or treatment of the disease was excluded. The one absolute criterion for the diagnosis of achalasia was roentgenographic evidence of a dilated esophagus with no evidence of any organic esophageal stricture. Treatment by dilation or by esophagomyotomy was only an ancillary factor in the selection of cases.

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Table 1.—**Residents of Rochester, Minn., With Achalasia During the Period 1935 Through 1964**

Sex	Age at onset, yr	Year of onset	Age at diagnosis, yr	Year of diagnosis	Age at last observation
Residents at onset of achalasia					
F	48	1926	58	1936	77*
F	69	1940	71	1942	77*
F	63	1946	63	1946	84
F	71	1955	72	1956	83
F	68	1956	70	1958	75*
M	28	1958	29	1959	37
F	62	1954	72	1964	75
Nonresidents at onset of achalasia					
F	26	1907	49	1930	65*
M	24	1939	36	1951	45
F	24	1952	24	1952	28
M	34	1960	37	1963	41

*Patient has died.

RESULTS

Eleven patients (three men and eight women) who were residents of Rochester, Minn., at the time of diagnosis (1935 through 1964) were found to have esophageal achalasia. Seven were residents of Rochester at the time of onset of symptoms; in six, symptoms occurred during the period 1935 through 1964, whereas in one, symptoms had developed in the previous decade (1925 through 1934) (Table 1). Follow-up in all cases has been either to death of the patient or to Jan. 1, 1968. The ages at onset for all patients ranged from 24 to 71 years (mean 47). Five of the eight women were more than 60 years of age, whereas none of the three men were more than 40 years of age. For the period of study, the average annual incidence rate, based on those who were residents at the time of onset of symptoms, was 0.6 per 100,000 persons per year* (Table 2). No appreciable change in annual incidence of achalasia occurred during this period of time. All patients were treated; in five patients, forceful dilation of the esophagus

Table 2.—**Number and Average Annual Incidence of Achalasia Among Residents of Rochester, Minn., by Decade of Onset**

Decade of onset	No. of patients	Average annual incidence, per 100,000
1925-1934	1	0.5
1935-1944	1	0.4
1945-1954	2	0.7
1955-1964	3	0.7
Overall (1925-1964)	7	0.6

*The 95% confidence interval for the annual incidence rate is $0.3/100{,}000 \leq R \leq 1.3/100{,}000$.

was the only form of therapy, whereas in the remaining six patients a modified Heller esophagomyotomy was performed.[3]

COMMENT

A review of the literature reveals few data relative to the incidence of esophageal achalasia in the general population. The disease as it affects Caucasians has been well documented in European and North American literature, but has been described more rarely as it affects other races. However, achalasia has been found in Negroes of the United States[4] and of Africa.[5,6] Most of the Negroes affected were adults, but one report was of a Negro infant.[7] The disease also has been noted in Indians[8] and Indian children,[9] and has been seen among the Japanese.[10] Thus, although achalasia does not appear to be limited to any one race, the extent to which any race is affected has not been determined. The geographic distribution cannot be assessed from isolated case reports. Most of the earlier series included only surgical patients or patients treated by dilation and thus represent a minimal incidence figure.

At the University Hospitals of Lund and Malmo in Sweden, 26 patients were treated by Heller's operation during a 7-year period (1943 to 1950).[11] In this study, the incidence was difficult to ascertain because the residence status of the 26 patients was not provided and there was no population denominator available on which an accurate estimate of the incidence could be calculated. In Liverpool, England, which has a population of about 750,000 persons but which also serves a wider area, 74 patients were treated by Heller's operation at the Liverpool Regional Thoracic Surgical Center from 1951 to 1961.[12] Thus, the average minimal annual incidence for surgically treated patients with achalasia was about 1 per 100,000 persons. But whether all 74 patients were bona fide residents of Liverpool at the time of diagnosis was not stated. From evidence at autopsy, Köberle[13] noted that the incidence in Vienna, Austria, was 2 in the 60,000 autopsies done during a 25-year period.

Males and females are generally found to be affected in equal proportions, although in one large adult series, males predominated by 327 to 274,[14] and, in one group of children, males predominated by 16 to 11.[15] At St. Thomas' Hospital, London, there were more males than females less than 50 years old but above this age the reverse was true.[16] The figures usually given are not necessarily representative of the age and sex distribution in the general population and are not comparable. As a result, no definite conclusions can be drawn

from the figures on the relative frequency in men or women. Although achalasia is present in neonates, the relative incidence of the congenital or infantile type is probably less than 5% of the total.[12,15]

The high incidence of mega-esophagus in South America is probably due to a chronic endemic infection by a trypanosome that causes Chagas' disease.[17-19] Most patients have other stigmata of chronic Chagas' disease, such as a dilated heart, megacolon, positive complement-fixation test for trypanosomiasis, and occasional histologic evidence of the parasite in the esophagus. Although isolated cases of acute trypanosomiasis have been described in the southern states of the United States, Chagas' disease in the North American continent is an unlikely cause of achalasia because the other more frequent manifestations of the disease such as megacolon, an enlarged heart, and a positive complement-fixation test are not present.

The present report comprises an epidemiologic study designed to identify all residents in a defined population suffering from esophageal achalasia. Case findings through individual examination of the population are of course impractical so the completeness of such a survey is limited by diagnostic facilities and the presence of patients with asymptomatic disease. Thus, rates derived from this survey must be considered minimal, because only those patients whose symptoms were severe enough to require them to seek medical attention and receive a diagnostic work-up necessary to confirm the diagnosis were included. The diagnosis in the present series was made by roentgenography in all cases and was confirmed in four of them by esophageal motility studies.

The incidence of achalasia in the community was studied by using only those records of persons who lived within the geographic limits of Rochester, Minn. It must be emphasized that these figures apply only to a small midwestern community and although an extrapolation can be made for the whole of the United States, the figures obtained are not necessarily valid because of population variations.

The average annual incidence rate was 0.6 per 100,000 persons. Based on the date of onset of symptoms, this rate has not changed appreciably during the past 30 years in Rochester. Although this rate is somewhat lower than the figures assessed from the hospital data in Liverpool, it is probably more accurate, because the population at risk in the other communities, as well as the residency status of the patients, was not reported. Actually based on the confidence

interval it can be said that the Rochester rate and the Liverpool rate are in fair agreeemnt.

Follow-up studies of larger series of achalasia support the findings that the disease does not necessarily shorten life, although there may be an increased risk of carcinoma of the esophagus.[20] The suggested relationship between achalasia and carcinoma has the limitation of being based on restrospective rather than prospective analysis.

The large number of elderly women in this study indicates that the disease probably does not shorten life expectancy; in 5 of the 11 patients, the onset of symptoms occurred in women more than 60 years of age. In none of the 11 patients did carcinoma of the esophagus subsequently develop. This finding may be due to the small numbers involved and the fact that few patients either had achalasia for more than 20 years or went without treatment, both of which may be factors in the suggested causal relationship.

The present study has not identified any etiologic factors nor supplied evidence of any real trend in the average annual incidence rate during the period of study. The importance of racial and geographic factors cannot be assessed, but it is anticipated that these figures will provide a basis for future comparative epidemiologic work on achalasia in other communities.

SUMMARY

An epidemiologic study of achalasia of the esophagus was made in Rochester, Minn. Eleven resident patients were found to have this condition diagnosed during the 30-year period 1935 through 1964. The overall incidence rate by decade of onset was 0.6 per 100,000 persons per year. This rate did not change appreciably during the years studied, and no etiologic factors could be identified. Although achalasia occurred in young patients of both sexes, there was some selection in our series for females in the older age groups. Among those patients, there appeared to be no effect on life expectancy.

REFERENCES

1. Willis, T.: Pharmaceuticae rationalis: Sive diatraba de medicamentorum, operationibus in humano corpore. London, Hagae-Comitis, 1674, 426 pp.
2. Kyle, R. A., Nobrega, F. T., Kurland, L. T., and Elveback, Lila R.: The 30-Year Trend of Leukemia in Olmsted County, Minnesota, 1935 Through 1964. Mayo Clin Proc *43*:342-353 (May) 1968.
3. Ellis, F. H., Jr., Kiser, J. C., Schlegel, J. F., Earlam, R. J., McVey, J. L., and Olsen, A. M.: Esophagomyotomy for Esophageal Achalasia: Experimental, Clinical, and Manometric Aspects. Ann Surg *166*:640-655 (Oct.) 1967.
4. Kurlander, D. J., Raskin, H. F., Kirsner, J. B., and Palmer, W. L.: Therapeutic Value of the Pneumatic Dilator in Achalasia of the Esophagus: Long-Term Results in Sixty-two Living Patients. Gastroenterology *45*:604-613 (Nov.) 1963.
5. Carayon, A., Perquis, P., and Courbil, L. J.: Résultats de six opérations d'Heller pour méga-oesophage chez l'Africain. Bull Soc Med Afr Noire Lang Franc *10*:699-701, 1965.

6. Chabel, J., and Mensah, A.: Le traitement chirurgical du méga-oesophage idiopathique en Afrique. Bull Soc Med Afr Noire Lang Franc *10*:163-167, 1965.
7. Redo, S. F., and Bauer, C. H.: Management of Achalasia in Infancy and Childhood. Surgery *53*:263-269 (Feb.) 1963.
8. Ahmed, H. U.: Carcinoma Oesophagus as a Complication of Achalasia Cardia (Report of a Case). Indian J Surg *26*:559-562 (July) 1964.
9. Gupta, R. S., and Rao, Rama: Achalasia of the Oesophagus in Children (A Case Report). Indian J Surg *25*:869-872 (Dec.) 1963.
10. Okamoto, E., Iwasaki, T., Ueda, T., and Takeda, Y.: Pathogenesis of Cardiospasm: Clinical and Experimental Studies. Med J Osaka Univ *14*:245-261 (Mar.) 1964.
11. Malm, A.: Cardioplasty in the Surgical Treatment of Achalasia of the Esophagus: A Critical Review Founded on Experimental Studies. Scand J Clin Lab Invest 3(Suppl 1):5-89, 1951.
12. Rickham, P. P., and Boeckman, C. R.: Achalasia of the Esophagus in Young Children. Clin Pediat (Phila) *2*:676-681 (Dec.) 1963.
13. Köberle, F.: Enteromegaly and Cardiomegaly in Chagas Disease. Gut *4*:399-405 (Dec.) 1963.
14. Olsen, A. M., Holman, C. B., and Andersen, H. A.: The Diagnosis of Cardiospasm. Dis Chest *23*:477-497 (May) 1953.
15. Payne, W. S., Ellis, F. H., Jr., and Olsen, A. M.: Treatment of Cardiospasm (Achalasia of the Esophagus) in Children. Surgery *50*:731-735 (Nov.) 1961.
16. Barrett, N. R.: Achalasia of the Cardia: Reflections Upon a Clinical Study of Over 100 Cases. Brit Med J *1*:1135-1140 (May 2) 1964.
17. Köberle, F., and Nador, E.: Etiologia e patogenia do megaesôfago no Brazil. Rev Paul Med *47*:643-661 (Dec.) 1955.
18. Ferreira-Santos, R.: Aperistalsis of the Esophagus and Colon (Megaesophagus and Megacolon) Etiologically Related to Chagas' Disease. Amer J Dig Dis *6*:700-726 (Aug.) 1961.
19. Atías, A., Neghme, A., Aguirre MacKay, I., and Jarpa, S.: Megaesophagus, Megacolon, and Chagas' Disease in Chile. Gastroenterology *44*:433-437 (Apr.) 1963.
20. Ellis, F. G.: The Natural History of Achalasia of the Cardia. [Abridged.] Proc Roy Soc Med *53*:663-666 (Aug.) 1960.