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Incidence of Achalasia in South Australia Based on Esophageal Manometry Findings

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BACKGROUND & AIMS: **Achalasia is a disorder of esophageal motility with a reported incidence of 0.5 to 1.6 per 100,000 persons per year in Europe, Asia, Canada, and America. However, estimates of incidence values have been derived predominantly from retrospective searches of databases of hospital discharge codes and personal communications with gastroenterologists, and are likely to be incorrect. We performed a cohort study based on esophageal manometry findings to determine the incidence of achalasia in South Australia.**

METHODS: **We collected data from the Australian Bureau of Statistics on the South Australian population. Cases of achalasia diagnosed by esophageal manometry were identified from the 3 adult manometry laboratory databases in South Australia. Endoscopy reports and case notes were reviewed for correlations with diagnoses. The annual incidence of achalasia in the South Australian population was calculated for the decade 2004 to 2013. Findings were standardized to those of the European Standard Population based on age.**

RESULTS: **The annual incidence of achalasia in South Australia ranged from 2.3 to 2.8 per 100,000 persons. The mean age at diagnosis was 62.1 ± 18.1 years. The incidence of achalasia increased with age (Spearman rho, 0.95; $P < .01$). The age-standardized incidence ranged from 2.1 (95% CI, 1.8–2.3) to 2.5 (95% CI, 2.2–2.7).**

CONCLUSIONS: **Based on a cohort study of esophageal manometry, we determined the incidence of achalasia in South Australia to be 2.3 to 2.8 per 100,000 persons and to increase with age. South Australia's relative geographic isolation and the population's access to manometry allowed for more accurate identification of cases than hospital code analyses, with a low probability of missed cases.**

Keywords: Achalasia; Incidence; Manometry; Esophagus.

Achalasia is a disorder of esophageal motility defined by absent peristalsis and impaired relaxation of the lower esophageal sphincter in the absence of lower esophageal sphincter obstruction. The pathophysiology involves degeneration of the inhibitory neurons of the myenteric plexus, however, the etiology remains unknown. Achalasia becomes more common with age, and has no sex predilection.[1–7] Historically, reported annual incidence rates have been 0.5 to 1.2 per 100,000 population (Table 1), and achalasia accounts for 5% of presentations with dysphagia.[8] These studies of incidence are derived predominantly from retrospective searches of databases of hospital discharge codes and personal communications with gastroenterologists and are likely to be inaccurate.[3–7,9–16] Although some studies quote suspected case detection error rates of 5% to 10%,[6] when case notes have been cross-checked directly with database search results, the error rate is 50% to 66%.[9,10]

More recently, where significant measures have been taken to reduce the case identification error rate, the reported incidence of achalasia has been as high as 1.6 per 100,000 population in both Canadian and Italian populations.[5,10] Of interest, these reports coincided with the introduction of high-resolution manometry and pressure topography plotting in 2000.[17] This has become the gold standard for diagnosing and classifying achalasia,[18]

Abbreviations used in this paper: CI, confidence interval; DES, diffuse esophageal spasm; HRM, high-resolution manometry; LES, lower esophageal sphincter; NSMD, nonspecific motility disorder.

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Table 1. All Publications Reporting an Incidence of Achalasia

Study	Cohort location	Years	Method of case identification	Number of diagnoses	Incidence M/F (per 100,000)
Earlam et al[11]	Rochester, Minnesota	1925–1964	Gastroenterologist estimations of disease frequency ICD code	11	0.6
Mayberry and Rhodes[12]	Cardiff, Wales	1926–1977	Not identified	48	0.4
Galen et al[13]	Virginia	1975–1980	Not identified	31	0.6
Mayberry and Atkinson[14]	Nottingham, England	1966–1983	ICD code, case notes	53	0.51
Arber et al[4]	Israel	1973–1978	Direct communication with gastroenterologists, ICD code, case notes	Overall 162	0.8
		1979–1983		Overall 162	1.1
Mayberry and Atkinson[7]	Scotland	1972–1983	Direct communication with specialists	583	1.12
	Wales	1970–1973	ICD code	197	0.71
	Northern Ireland	1970–1973		153	0.98
	Eire	1976–1982		453	1.34
	England	1972–1983		4920	1.08
Stein et al[15]	Zimbabwean natives	1974–1988	Not identified	25	0.03
Mayberry and Atkinson[6]	New Zealand	1980–1984	ICD code	152	0.95
Howard et al[3]	Lothian Region, Edinburgh	1986–1991	Registry search–confirmed by endoscopy and manometry	38	0.80/0.83
Ho et al[16]	Singapore	1989–1996	Manometry, case notes	49	0.3/0.27
Birgisson and Richter[9]	Iceland	1952–2002	ICD code, case notes	62	0.55
Farrukh et al[2]	South Asians in Leicester, England	1986–2005	ICD code, endoscopy, manometry, botulinum toxin stock transaction registries, case notes	13	0.89
Gennaro et al[5]	Veneto Region, Italy	2001–2005	ICD code, case notes	365	1.59/1.58
Sadowski et al[10]	Alberta, Canada	2007	ICD or procedure code (balloon dilation or esophagomyotomy)	463	1.85/1.43
Kim et al[1]	Republic of Korea	2011	ICD code	191	0.33/0.44

ICD, International Classification of Diseases; M/F, male/female.

raising the possibility that, with the introduction of this new technology, there is an improved diagnostic capacity for the identification of patients with achalasia.

In South Australia, during the past decade, the adult South Australian population has had routine esophageal manometry performed at 1 of 3 manometry laboratories for investigation of dysphagia not associated with structural abnormalities. Each laboratory has maintained prospective databases of patient demographics and manometry results. Therefore, in South Australia, it seems possible to identify and validate all diagnosed cases of achalasia by a search of these 3 prospective adult manometry databases. This study was performed to provide information regarding local burden of disease.

Methods

This study was approved by the Human Research Ethics Committee of the Royal Adelaide Hospital in Adelaide, South Australia, Australia (HREC/14/RAH/326, protocol 140803).

Cohort

Data for the South Australian adult population for the decade 2004 to 2013 were obtained from the Australian Bureau of Statistics. Adults were defined as 18 years and older. Geographic population data were available in 2008 and 2013. Residence was defined as Capital City if living in the Greater Capital City Statistical Area, Greater Adelaide (4GADE), and as regional/remote if living in the Greater Capital City Statistical Area Rest of South Australia (4RSAU) as per the Australian Statistical Geography Standard 2011.

Esophageal Manometry

During 2004 to 2013, esophageal manometry was performed at 1 of 3 adult laboratories and the patient details and corresponding manometry report were recorded in a prospective database. One additional motility laboratory exists in Adelaide that services the pediatric population, but access to these data proved too difficult for inclusion in our study. To the authors' knowledge, no one in Adelaide offers manometry in the private sector because the Medicare Benefits Schedule fee fails to cover the costs of the procedure. Esophageal pressures were measured using low- and high-resolution manometry (HRM), with motility shown as a continuum of pressure and time using a color display.

Manometric data from the Gut Function Laboratory, Department of Gastroenterology and Hepatology, Royal Adelaide Hospital, were acquired between 2004 and 2012 using a Dentsleeve 10-channel water perfused system

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