Achalasia cardia: A five-year review in Hospital Tuanku Ja'afar, Seremban

Daniel Hong Jeng Chin, MBBS[1], Kean Ghee Lim, FRCS (Edin)[1], Hong Shawn Peh, MBBS[1], Ismail Abdul Sattar Burud, MSURG[1], Mahadevan Deva Tata, MSURG[2], Jasiah Zakaria, MSURG[2]

[1]Department of Surgery, Clinical School, International Medical University, Jalan Rasah, Seremban, Negeri Sembilan, Malaysia,
[2]Department of Surgery, Hospital Tuanku Ja'afar Seremban, Ministry of Health, Malaysia

ABSTRACT

Introduction: Achalasia cardia is an oesophageal motility disorder that affects various age groups. This study focused on the epidemiological features of achalasia, its risk factors, treatment modalities offered and the clinical outcomes in a tertiary hospital.

Materials and Methods: A retrospective search was carried out on all patients with a diagnosis of achalasia cardia in Hospital Tuanku Ja'afar (HTJ), Seremban, Malaysia between 2014 and 2018. Demographic data, patient symptomatology, and definitive management options were determined from the records. Telephone interviews were conducted to evaluate patient satisfaction with the outcome of treatment.

Results: There were 30 patients with a newly diagnosed achalasia cardia in that 5-year period, with an equal incidence among men and women. The mean age of presentation was 44.63 ± 18.21 years. Malays formed the largest group. The mean weight and body mass index were 46.8 ± 10.4 kg and 18.0 ± 4.4 kg/m2 respectively. There was a wide range of duration of symptoms at presentation with a mean of 30.11 ± 35.29 months. Almost all patients presented with dysphagia (96.7%) while 70% also noted loss of weight. All patients underwent oesophagogastroduodenoscopy (OGDS) and 26 patients (86.7%) had barium swallow as part of diagnostic workup. A total of 18 patients underwent a laparoscopic Heller myotomy with or without Dor Fundoplication and/or cruroplasty while two patients (6.7%) underwent pneumatic dilatation as first treatment. Iatrogenic mucosal perforations were detected in 8 patients who underwent myotomy and fundoplication and were repaired intraoperatively. Of the patients who underwent myotomy and fundoplication, the mean weight increase was 15.6kg, increasing from 43.0 ± 8.4 kg to 58.6 ± 13.7 kg. All the patients who underwent treatment were satisfied with their treatment outcomes.

Conclusion: Most patients with achalasia cardia deemed suitable for surgery and counselled accordingly accept surgery resulting in high levels of satisfaction and weight gain in almost all these patients. A small minority who opt for pneumatic dilatation may also achieve satisfactory outcomes comparable to surgery in the short term. Although rare, clinicians should be able to recognise this disease early as early intervention often leads to satisfactory long-term outcomes.

KEYWORDS:
Achalasia Cardia, Dysphagia, Hellers myotomy, Dor Fundoplication

INTRODUCTION

Achalasia cardia is a form of motor dysphagia which results from progressive degeneration of ganglion cells in the myenteric plexus, leading to failure of relaxation of the lower oesophageal sphincter, accompanied by a loss of peristalsis in the distal oesophagus.[1] First described over 300 years ago, there has been much debate over the aetiology of achalasia with several potential triggers being implicated as the cause for the inflammatory destruction of inhibitory neurons in the oesophageal myenteric plexus. These include autoimmune responses, infectious agents and genetic factors.[2] Although there have been strong associations between these triggers and primary achalasia, none have been proven conclusively. It is likely to involve a multifactorial aetiology following an initiating event.[1,2]

Achalasia is considered a relatively uncommon disorder with incidence rates between 0.5-1.2/100000 per year.[1] Incidence rates have been reported to be as low as 0.03/100000 per year in Zimbabwe to as high as 1.63/100000 per year in Canada and the incidence rates of achalasia appear to be rising over time.[1,2,3] Whether this geographical variation and rise in incidence reflects a true rise in incidence or greater awareness and improved diagnosis remains uncertain.[1] There have been no distinct patterns of achalasia incidence based of sociodemographic factors with the disease affecting both genders, all races and all ages.[4] A few studies have demonstrated a bimodal distribution of incidence by age with peaks at around age 30 and 60 years while others have shown a generally increased risk with increased age.[5-7] Several studies show that females have a higher incidence of achalasia while others have shown equal incidence among genders.[5,6,7,8]

In Malaysia, Ganesananthan et al described a 6-year case series of achalasia cardia in Hospital Kuala Lumpur from year 2000-2005 which had a sample size of 61 patients.[9] His series noted a trend towards a younger age group with a mean age of 48 years, a female predilection (M:F = 1:1.4) and a slight Malay majority (56%).[9] Fifty patients from this series underwent pneumatic dilatation without complications and had excellent symptomatic relief with an average post-

This article was accepted: 18 November 2021
Corresponding Author: Daniel Hong Jeng Chin
Email: danielchin.hj@gmail.com

procedural weight gain of 4kg over 3-24 months.[9] No patients from this series underwent a Heller myotomy.[9] More recently, Siow SL et al described a series of 55 cases enrolled in a study of clinical outcomes of laparoscopic Heller myotomy and anterior Dor fundoplication between 2010-2019.[10] His series showcased a similar mean age of 49 years but noted a male predilection instead (53%).[10] Patients in this series reported a 100% satisfaction rate with no mortalities.[10] The patients were from 4 institutions with consultant level expertise in Sarawak, Johore, Penang and Kedah.[10]

We undertook a retrospective study of patients seen at Hospital Tuanku Ja'afar (HTJ), Seremban to describe the epidemiological features of achalasia in this area, its risk factors, treatment modalities offered to them and the clinical outcomes in a tertiary upper gastrointestinal surgery hospital. We also aimed to raise awareness among clinical practitioners on the recognition of this enigmatic disease as early diagnosis with prompt treatment often yields good long-term clinical outcomes for most patients across the entire spectrum of the disease.

MATERIALS AND METHODS

A retrospective search of all patients with a discharge diagnosis of achalasia cardia (ICD 10 code of K22.0) in HTJ between 2014 and 2018 was conducted. Medical records of patients were procured from the hospital records and were retrospectively reviewed and analysed. A parallel search of records of all patients who underwent oesophageal manometry in HTJ from 2014 onwards was also undertaken to augment and complement the search. Through this parallel search, we found an additional 8 patients with achalasia cardia and 2 further patients with diagnoses of primary motility disorder. These patients were initially missed by the preliminary search based on the discharge diagnosis of achalasia cardia as they were neither admitted nor underwent definitive treatment in HTJ throughout the duration of our study.

Demographic data, patient symptomatology, data required to confirm diagnoses and definitive management options were determined from these records. Patients' regions of residence were categorized into either urban or rural based on postcodes.

Patients with errors in coding who were diagnosed with pseudoachalasia due to other causes of dysphagia such as gastroesophageal reflux disease, corrosive ingestion, carcinoma of the oesophagus or scleroderma were excluded. Those with established diagnoses before the study period or were no longer treatment naïve were also excluded from this study.

Ethical clearance was obtained from the International Medical University Joint Committee as well as the National Medical Research and Ethics Committee.

Follow-up data was obtained via both surgical outpatient department records and telephone interviews where appropriate. The outcome status of patients, including their satisfaction was determined in June and July 2020 via contact tracing by phone. Patient satisfaction was determined using a questionnaire asking patients to self-report their change in quality of life. Their satisfaction status was then classified into "Satisfied", "Dissatisfied", or "Neither".

Data was collected and presented using a combination of Google Drive, Microsoft Word and Excel. The data was then analysed using IBM SPSS version 23.0 for Windows (SPSS Inc., Chicago, IL, USA).

RESULTS

A total of 30 patients were diagnosed with achalasia of the cardia. The mean age of presentation was 44.63 years ± 18.21 years, with a range from 16 years to 80 years (Table I). There was equal incidence of achalasia among men and women (1:1). Malays formed the largest group (n = 24, 80%). Almost all patients originated from the Southern half of Peninsular Malaysia, namely Negeri Sembilan, Melaka and Johor. Only one patient came from Selangor, a close neighbour to Seremban. Conspicuously, 63% (19/30) of the patients resided in urban areas. The Chinese were all from urban areas while the sole Indian patient had a rural address.

The mean weight and body mass index (BMI) of the cohort was 46.8 ± 10.4 kg and 18.0 ± 4.4 kg/m^2 respectively. Half the patients in this series presented with no medical comorbidities. Only two patients (6.7%) had diabetes, two (6.7%) had hypertension while only one patient was found to have cerebrovascular disease, liver disease or peptic ulcer disease. 23% (7) were smokers and five (16.7%) had pulmonary diseases.

The mean duration of symptoms at presentation in this series was 30.11 months ± 35.29 months (Table II). Almost all patients presented with dysphagia (n = 29, 96.7%) while loss of weight (n = 21, 70.0%) and vomiting (n = 18, 60.0%) were the next most common symptoms. (Table II) All other symptoms such as heartburn (13.3%), abdominal pain (16.7%), regurgitation (13.3%), choking sensation (16.7%) and chest pain (6.7%) were uncommon.

All patients in this series were treatment naïve upon presentation at this centre. All patients underwent oesophagogastroduodenoscopy (OGDS) during diagnostic workup. In addition 26 patients (86.7%) were investigated with barium swallow and 16 (53.3%) oesophageal manometry.

A total of 18 patients (n = 18, 60%) underwent a laparoscopic Heller myotomy with or without Dor Fundoplication and/or cruroplasty (Table III). Two patients (6.7%) underwent pneumatic dilatation as first treatment. One patient with a sigmoid oesophagus as seen on barium swallow underwent a thoracoscopic assisted oesophagectomy which was converted to a transhiatal 3-stage oesophagectomy plus gastric pull-up. One refused a referral for treatment and opted for conservative observation. The remaining 8 patients (26.7%) were also not treated, of which half were lost to follow up while the other half were referred to other centres.

Table I: Demographic characteristics of Achalasia patients in Hospital Tuanku Ja'afar, Seremban

Sociodemographic Characteristics		N (%)	
Gender	Male	15	(50.0)
	Female	15	(50.0)
Race	Malay	24	(80.0)
	Chinese	5	(16.7)
	Indian	1	(3.3)
State	Negeri Sembilan (urban)	7	(23.3)
	Negeri Sembilan (rural)	3	(10.0)
	Melaka (urban)	5	(16.7)
	Melaka (rural)	4	(13.3)
	Johor (urban)	7	(23.3)
	Johor (rural)	3	(10.0)
	Selangor (urban)	1	(3.3)
BMI (kg/m^2)	>15	4	(13.3)
	15 – 19.9	14	(46.7)
	20 – 24.9	4	(13.3)
	25 – 29.9	1	(3.3)
	>30	1	(3.3)
	No record	6	(20.0)
Weight Category (kg)	31.0 – 40.9	11	(36.7)
	41.0 – 50.9	8	(26.7)
	51.0 – 60.9	2	(6.7)
	61.0 – 70.9	2	(6.7)
	71.0 – 80.9	1	(3.3)
	No record	6	(20.0)
Age Group (age at diagnosis)	11 – 20	4	(13.3)
	21 – 30	3	(10.0)
	31 – 40	6	(20.0)
	41 – 50	4	(13.3)
	51 – 60	9	(30.0)
	61 – 70	1	(3.3)
	71 – 80	3	(10.0)

Table II: Clinical characteristics of patients with achalasia of the cardia

Clinical Characteristics	N(%)	
Dysphagia	29	(96.7)
Heartburn	4	(13.3)
Abdominal Pain	5	(16.7)
Loss of Weight	21	(70.0)
Loss of Appetite	3	(10.0)
Vomiting	18	(60.0)
Regurgitation	4	(13.3)
Choking Sensation	5	(16.7)
Lethargy	2	(6.7)
Chest Pain	2	(6.7)
Dyspnoea	1	(3.3)
Odynophagia	1	(3.3)
Cough	1	(3.3)
Duration of Symptoms		
0 – 6 months	11	(36.7)
7 – 12 months	6	(20.0)
13 – 18 months	0	(0.0)
19 – 24 months	2	(6.7)
More than 24 months	11	(36.7)
ASA score		
I	15	(50.0)
II	15	(50.0)
III	0	(0.0)
IV	0	(0.0)

Table III: Interventions and outcomes of achalasia patients in Hospital Tuanku Jaafar

Interventions and Outcomes		Count	Column N %
OGDS		30	100.0%
Barium		26	86.7%
Manometry		16	53.3%
Chest X-ray		3	10.0%
CT		10	33.3%
Treatment	Laparoscopic Heller Myotomy with Dor Fundoplication	14	46.7%
	Laparoscopic Heller Myotomy with Dor Fundoplication and Cruroplasty	1	3.3%
	Laparoscopic Heller Myotomy with Cruroplasty only	3	10.0%
	Pneumatic Dilatation	2	6.7%
	Oesophagectomy	1	3.3%
	Not treated (Under follow up)	1	3.3%
	Referred out	4	13.3%
	Not treated (Lost to follow up)	4	13.3%
Complications	Iatrogenic mucosal perforation	8	26.7%
	Poor motility	1	3.3%
	Stricture	2	6.7%
	No complications	21	70.0%
Satisfaction (N=21)	Satisfied	21	100.0%
	Dissatisfied	0	0%
	Neither	0	0%

Patients with surgery had a follow up period ranging from 9-62 months (median 45.5 months, mean 40.4 months). Of the patients who underwent myotomy and fundoplication, their mean weight increase was 15.6kg, increasing from 43.0 ± 8.4 kg to 58.6 ± 13.7 kg (n-17, no record for 1 patient). One patient reportedly lost weight from 50kg (BMI 20.8) to 48kg. The greatest weight gain was observed in a patient of 39kg (BMI15.2) who increased to 80kg (BMI 31.3). The patient with oesophagectomy also had a weight gain of 4kg after one year. The outcome status of patients as determined in June and July 2020 revealed no deaths.

One patient who had eleven pneumatic dilatations over 15 months gained 7kg in weight. Another who had 3 dilatations and subsequently discontinued treatment reported a stagnant weight (40kg).

Iatrogenic mucosal perforations were detected in 8 patients who underwent myotomy and fundoplication (44.4%) intraoperatively and repaired immediately. Two patients (11.1%) sustained oesophageal strictures post myotomy for which they underwent successful dilatations. One patient with associated mega-oesophagus and hiatal hernia developed poor motility post myotomy for which subsequent dilatation was also done. Four patients were not treated because they were either unfit or refused any intervention. The outcome of these four patients is not known as they were lost to follow up.

At follow up tracing, patients who underwent intervention were asked to rate their subjective assessment as "satisfied", "dissatisfied", or "neither". All of the 21 patients who underwent surgical treatment were satisfied with the treatment received and overall clinical outcomes.

DISCUSSION

This is a review of all treatment naïve patients diagnosed at HTJ, a hospital with tertiary upper gastrointestinal surgical services up until 2018. The majority of the cohort included in this series had surgical treatment. However, as with any service some patients may decline any form of treatment, be lost to follow up or receive treatment finally in a different hospital. There were 10 (33.3%) such patients in this series. Four of these were patients from Johor. This affirms the mobility of Malaysians have in seeking treatment in the network of public hospitals. A check with the authors of Siow et.al [10] confirm that there are no duplicate patients in this series and theirs.

In the presenting cohort, we noted, similar to other Malaysian series [9,10] that there is no gender predilection for achalasia in Malaysia. Similarly, most do not have many co-morbidities and have good ASA scores. Malays form a large majority in our sample as they form the largest majority of the population and are the population that depends most heavily on our public hospitals. Our data does not suggest the disease is more common in either the urban or rural setting.

Only 53.3% of patients in HTJ underwent manometry prior to initiation of treatment. OGDS and barium swallow were deemed adequate for evaluation in many cases. This rate was similar compared to Siow et al's [10] 45.5% despite manometry being the gold standard for diagnosis of achalasia due to its high sensitivity.[11] Manometry was used to rule out other motility disorders. Although the use of manometry enables the classification of achalasia into subtypes with subsequent therapeutic considerations[12], it is not considered routinely essential for satisfactory surgical outcomes.[10]

The preferred surgical procedure offered and deemed suitable for most patients remains a myotomy with an antireflux procedure.[10,13,14] Follow-up shows that most patients gained weight and achieved good long-term outcomes. Peroral endoscopic myotomy, with promising early results, has gained popularity in the last decade as the definitive procedure for achalasia.[10,15] However, the issue with troublesome postprocedural reflux, the lack of long-term data and its relatively long learning curve means that the former

remains the preferred procedure at the present time.[16,17,18]

Pneumatic dilatation was offered and accepted by two patients. One patient was judged unsuitable for Heller's myotomy and the other declined surgery. Another four patients were referred out in 2018 due to the lack of an upper gastrointestinal surgeon servicing the hospital. The surgeons servicing the hospital during the duration of study affirmed that most patients were in fact referred for surgical intervention after conservative measures had failed. Options of pneumatic dilatation versus surgical myotomy were discussed with patients with patient preferences also taken into account.

The rate of perforation is high compared to other studies. Nevertheless, they were all detected and managed intraoperatively and did not result in early post-operative morbidity. A similar recent study in Malaysia reported a perforation rate of 7.3%[10,19] and an acceptable rate of intraoperative perforation is considered to be below 10%.[20] The use of different energy devices in the form of hook cautery, harmonic scalpel in performing cardiomyotomy could be a reason.[21] Another reason for high perforation rate could be attributed to the fact that HTJ is a training centre and the operations were performed by surgeons with varying levels of experience, an independent risk factor as demonstrated by Tsuboi K et al.[22]

Although achalasia remains a disease with relatively low incidence in most communities, the prospect of a good outcome makes it important that clinicians recognise the disease early in order for patients to seek the required surgical expertise.

This study had a few limitations. Due to the retrospective nature of this review, the medical records of patients were dependent on manual documentation, thus variable and had some inconsistency. Subjective data, such as patient satisfaction was open to interviewer and patient recall bias because of the time gap and was dependent on the patients' memory. Our sample size was dependent on the numbers available and was not as large as we had hoped.

The main strength of this study was the robust search because of three parallel search avenues. The records of patient were adequate for complete data extraction and all patients were contactable via telecommunication. Patients' data was collected from the study subject and/ or their legal representative. One investigator was assigned to collect the data through telemedicine to ensure consistent bidirectional communication.

CONCLUSIONS

Most patients with achalasia cardia deemed suitable for surgery and counselled accordingly readily accept surgery. Laparoscopic Heller Cardiomyotomy and fundoplication results in high levels of patient satisfaction and weight gain in almost all these patients with minimal incidence of postoperative complications. A small minority who opt for pneumatic dilatation may also achieve satisfactory short-term outcomes comparable to surgery. Additional studies with long-term outcomes are required to further determine treatment durability as well as patient satisfaction in the long term.

ACKNOWLEDGEMENTS

The authors thank the Director General of Health, Malaysia for his permission to publish this article. We acknowledge no conflict of interests. An internal funding grant was received for this research project from the International Medical University

REFERENCES

1. O'Neill OM, Johnston BT, Coleman HG. Achalasia: a review of clinical diagnosis, epidemiology, treatment and outcomes. World J Gastroenterol 2013; 19(35): 5806-12.
2. Sadowski DC, Ackah F, Jiang B, Svenson LW. Achalasia: incidence, prevalence and survival. A population-based study. Neurogastroenterol Motil 2010; 22: e256-61.
3. Stein CM, Gelfand M, Taylor HG. Achalasia in Zimbabwean blacks. S Afr Med J 1985; 67: 261–2
4. Podas T, Eaden J, Mayberry M, Mayberry J. Achalasia: a critical review of epidemiological studies. Am J Gastroenterol 1998; 93: 2345–7.
5. Howard PJ, Maher L, Pryde A, Cameron EW, Heading RC. Five year prospective study of the incidence, clinical features, and diagnosis of achalasia in Edinburgh. Gut 1992; 33: 1011–5.
6. Mayberry JF, Atkinson M. Variations in the prevalence of achalasia in Great Britain and Ireland: an epidemiological study based on hospital admissions. Q J Med 1987; 62: 67–74.
7. Ho KY, Tay HH, Kang JY. A prospective study of the clinical features, manometric findings, incidence and prevalence of achalasia in Singapore. J Gastroenterol Hepatol 1999; 14: 791-5.
8. Gennaro N, Portale G, Gallo C, Rocchietto S, Caruso V, Costantini M, et al. Esophageal achalasia in the Veneto region: epidemiology and treatment. Epidemiology and treatment of achalasia. J Gastrointest Surg 2011; 15: 423–8.
9. Ganesananthan S, Melvin R, Anil R, Gan IE, Khor BP. The 2006 Kuala Lumpur Hospital Achalasia Cardia Registry: Lessons to learn from a comprehensive six years review. Annual Scientific Meeting of the Malaysian Society of Gastroenterology and Hepatology 2006; 31.
10. Siow SL, Mahendran HA, Najmi WD, Lim SY, Hashimah AR, Voon K, et al. Laparoscopic Heller myotomy and anterior Dor fundoplication for achalasia cardia in Malaysia: Clinical outcomes and satisfaction from four tertiary centers. Asian J Surg 2021; 44(1): 158-63.
11. von Rahden BH, Filser J, Seyfried F, Veldhoen S, Reimer S, Germer CT. Diagnostik und Therapie der Achalasie [Diagnostics and therapy of achalasia]. Chirurg 2014; 85(12): 1055-63.
12. Pripisi L, Hoara P, Constantin A, Predescu D, Birla R, Gindea C, et al. The Value of Intraoperative Endoscopy and Manometry in the Surgical Treatment of Achalasia. Chirurgia (Bucur) 2018; 113(4): 469-77.
13. Patti MG, Andolfi C, Bowers SP, Soper NJ. POEM vs Laparoscopic Heller Myotomy and Fundoplication: Which Is Now the Gold Standard for Treatment of Achalasia? J Gastrointest Surg 2017; 21(2): 207-14.
14. Patti MG, Allaix ME. Recurrent symptoms after Heller myotomy for achalasia: evaluation and treatment. World J Surg 2015; 39(7): 1625-30.
15. Inoue H, Minami H, Kobayashi Y, Sato Y, Kaga M, Suzuki M, et al. Peroral endoscopic myotomy (POEM) for esophageal achalasia. Endoscopy 2010; 42(4): 265-71.

16. Schlottmann F, Luckett DJ, Fine J, Shaheen NJ, Patti MG. Laparoscopic Heller Myotomy Versus Peroral Endoscopic Myotomy (POEM) for Achalasia: A Systematic Review and Meta-analysis. Ann Surg 2018; 267(3): 451-60.
17. El Zein M, Kumbhari V, Ngamruengphong S, Carson A. K, Stein E, Tieu A, et al. Learning curve for peroral endoscopic myotomy. Endosc Int Open 2016; 4(5): E577–82.
18. Liu Z, Zhang X, Zhang W, Zhang Y, Chen W, Qin W, et al. Comprehensive Evaluation of the Learning Curve for Peroral Endoscopic Myotomy. Clin Gastroenterol Hepatol 2018; 16(9): 1420-6.
19. Campos GM, Vittinghoff E, Rabl C, Takata M, Gadenstätter M, Lin F, et al. Endoscopic and surgical treatments for achalasia: a systematic review and meta-analysis. Ann Surg 2009 Jan; 249(1): 45-57.
20. Smith CD, Stival A, Howell DL, Swafford V. Endoscopic therapy for achalasia before Heller myotomy results in worse outcomes than heller myotomy alone. Ann Surg 2006; 243(5): 579-86.
21. Kathy S, Hajdu Z, Bokor L, Bagi R. Use of harmonic scalpel at laparoscopic cardiomyotomy. A new method. Acta Chir Hung 1997; 36(1-4): 154-5.
22. Tsuboi K, Omura N, Yano F, Hoshino M, Yamamoto SR, Akimoto S, et al. Identification of risk factors for mucosal injury during laparoscopic Heller myotomy for achalasia. Surg Endosc 2016; 30(2): 706-14.