Editorial

Achalasia—a not so common yet fascinating esophageal motility disorder

Sarah K. Thompson

College of Medicine & Public Health, Flinders University, Bedford Park, South Australia, Australia

Correspondence to: Sarah Thompson, MD. Rm 6E112.5, Flinders Medical Centre, Bedford Park, South Australia, Australia.
Email: sarah.thompson@flinders.edu.au.

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In 2014, I was asked to present Grand Rounds by Professor John Hunter at the Oregon Health and Science University. I had completed a minimally invasive surgery fellowship under his (and Dr. Brett Sheppard's) tutelage in 2005, and it was a great honour to return as a visiting professor. The topic assigned to me was 'Achalasia'. Unfortunately, not the more comfortable topics of gastro-esophageal reflux or upper gastrointestinal cancer!

I set about preparing my talk and, in doing so, many questions were raised. Why did it seem as if we were operating on more cases of achalasia per year than the expected number based on an incidence of 0.5 to 1.6 per 100,000 (1,2)? Was this an anomaly or had the incidence thus far been under-reported? Is type III achalasia a negative predictor of outcome following a laparoscopic cardiomyotomy compared to type I or II achalasia? And is a laparoscopic stapled cardioplasty a good option for patients with end-stage achalasia?

Upon returning to Adelaide, these questions were examined in further detail. Investigating the incidence of achalasia in South Australia was an enticing study given the state's relative geographic isolation and the population's ready access to manometry. Historical studies had been derived from retrospective searches of databases of hospital discharge codes along with personal communications with gastroenterologists. In contrast, we collected data from the Australian Bureau of Statistics on the South Australian population and identified all cases of achalasia from the only three adult manometry laboratory databases in South Australia.

We found that indeed the incidence of achalasia was higher than that reported in the literature: 2.3 to 2.8 per 100,000 people (3). Our study was supported by a simultaneous study published in the same issue of *Clinical Gastroenterology and Hepatology* (4). The world-leading group in Central Chicago, led by Kahrilas and Pandolfino, also found an increased incidence of 2.92 per 100,000 using an integrated database of Northwestern Medicine electronic health records over roughly the same time span.

To answer the second question raised in preparation of the Grand Round, all original manometry tracings were re-reported to classify laparoscopic cardiomyotomy patients in our prospective database as type I, II or III achalasia. One hundred and ninety-five patients were subtyped to type I (n=60), type II (n=111), and type III (n=24) achalasia. And yes, in our patient cohort, patients with type III achalasia did worse after laparoscopic cardiomyotomy than those with types I and II. Of interest, dysphagia scores did not differ between subtypes, suggesting that the difference observed was due to persistent chest pain and/or regurgitation (5).

And finally, we re-visited our longer-term results for five patients who had undergone a laparoscopic stapled cardioplasty for end-stage achalasia (6). We realised that, despite the initial promising results, four out of five patients had progressed to, or were considering, esophagectomy (7). This is discussed in more detail in the attached article in this Special Series on end-stage achalasia.

When approached by Professor David Watson to oversee a special edition on Achalasia for the *Annals of Esophagus*, I saw the perfect opportunity to approach physicians in the field to write a paper on their area of expertise. I hope you will not be disappointed. Topics include an update on the captivating pathophysiology of achalasia by Prof. Prakash Gyawali and associates, strategies to overcome limitations

in clinical application for the Chicago Classification by Dr. Jenny Myers, the nuances of pseudoachalasia by A/Prof. Peter Devitt, a review of current guidelines for achalasia by Mr. Andy Cockbain, and an examination of the overall risk of cancer in patients with achalasia by Mr. Ronan Gray.

There are also updates on the various interventions for achalasia: botox injection by Drs. Dick Heddle and Charles Cock, pneumatic balloon dilatation by Dr. Tim Bright, laparoscopic cardiomyotomy by Prof. John Hunter, and a review of 100 consecutive POEM (per oral endoscopic myotomy) cases performed by Dr. Gary Crosthwaite in Melbourne, Australia. My colleague Mr. Jon Shenfine has written a paper on the impact of obesity on achalasia management, and A/Prof. Ahmad Aly and Dr. David Liu from the Austin Hospital in Melbourne have developed a decision matrix for the management of achalasia depending on subtype and co-morbidities. Finally, Professor Watson has outlined his approach to a patient with recurrent symptoms, and I have described our local experience with esophagectomy for end-stage achalasia.

The view from contributing authors has been overwhelmingly positive. It is often necessary to take a step back from our busy lives to reflect upon current results and outcomes, and to evaluate whether improvements are needed in our clinical decision-making and management of a particular problem. Perhaps this special edition will inspire you to investigate an aspect of achalasia in your patient population—certainly the Grand Round in 2014 did just that for me!

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Footnote

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