CHALLENGES WITH RECERTIFICATION (OF EAGLE’S SYNDROME) – WHO HAS THE TIME?

Ian Cheng
BMed BE DipOEH DipAvMed MPH FACAsM FAFOEM

INTRODUCTION
A 50 year old male airline transport pilot licence (ATPL) pilot was diagnosed in December 2010 with Stage 4A non-Hodgkin’s follicular lymphoma following presentation to an Emergency Department with abdominal pain, diarrhoea, back pain, drenching sweats, nausea and vomiting. He was managed with six cycles of R-CHOP/Rituximab, Cyclophosphamide, Doxorubicin, Vincristine and Prednisone) plus two extra doses of Rituximab. By April 2011 the pilot was declared by his haematologist to be in remission and CASA issued him a Class 1 medical certificate with a multi-crew restriction together with an audit requirement for regular specialist reports. The multi-crew restriction was removed in 2012. His only ongoing prescribed medication was Rabeprazole 20mg daily

EAGLE’S SYNDROME
As part of the ongoing periodical surveillance arranged by his haematologist, the pilot underwent a neck, chest and abdomen CT scan on 6 May 2014 prior to his annual medical. The pilot subsequently telephoned to inform me that the radiologist had diagnosed ‘Eagle’s syndrome’. The CT scan reported: “There is an incidental finding of an elongated partially calcified left stylohyoid ligament (known as eagle syndrome)”. The pilot added that his haematologist had conducted a preliminary search about Eagle’s syndrome, which raised concerns about potential complications and as a result his haematologist referred him to a vascular surgeon.

The first three webpages generated by a Google™ search of ‘Eagle’s syndrome’ indicated that potential complications could include transient ischaemic attacks, carotid artery dissection, syncope and sudden death.2,4 Eagle’s syndrome (or Eagle syndrome) was named after an American otolaryngologist, Watt Eagle who wrote a number of papers linking various symptoms with an elongated or calcified styloid process.5-7 Interestingly, Watt Eagle did not suggest or coin the eponymous term ‘Eagle’s syndrome’ in his three articles. Indeed he noted that several others before him had noted an association between an elongated styloid process and clinical symptoms.5,6

The styloid process arises from the inferior surface of the temporal bone and passes downwards, forwards and medially. The stylohyoid ligament connects the styloid process to the (lesser cornu) of the hyoid bone.8,13,15 A number of papers state that styloid process lengths greater than 3 cm are considered elongated.8-10,13,14,17,18,21,25

The prevalence of elongated styloid process is thought to be 3 to 4% of the population8-11,13,15,18,22, although, a range of 1.4 to 30% has been described in some studies.23 However, only a small percentage (between 4% and 10.3%) of this group is thought to actually be symptomatic.8-11,14,23 A large range of styloid ligament calcification prevalence has been quoted, ranging from 18%13 to 68%.16

The first three webpages generated by a Google™ search of ‘Eagle’s syndrome’ indicated that potential complications could include transient ischaemic attacks, carotid artery dissection, syncope and sudden death.2,4 Eagle’s syndrome (or Eagle syndrome) was named after an American otolaryngologist, Watt Eagle who wrote a number of papers linking various symptoms with an elongated or calcified styloid process.5-7 Interestingly, Watt Eagle did not suggest or coin the eponymous term ‘Eagle’s syndrome’ in his three articles. Indeed he noted that several others before him had noted an association between an elongated styloid process and clinical symptoms.5,6

The styloid process arises from the inferior surface of the temporal bone and passes downwards, forwards and medially. The stylohyoid ligament connects the styloid process to the (lesser cornu) of the hyoid bone.8,13,15 A number of papers state that styloid process lengths greater than 3 cm are considered elongated.8-10,13,14,17,18,21,25

The prevalence of elongated styloid process is thought to be 3 to 4% of the population8-11,13,15,18,22, although, a range of 1.4 to 30% has been described in some studies.23 However, only a small percentage (between 4% and 10.3%) of this group is thought to actually be symptomatic.8-11,14,23 A large range of styloid ligament calcification prevalence has been quoted, ranging from 18%13 to 68%.16

ABSTRACT
A 50 year old male airline transport pilot licence (ATPL) pilot had been on a CASA audit requirement (CAR) for Non-Hodgkin’s Lymphoma since 2010. As part of his ongoing haematological medical surveillance a neck, chest, abdomen and pelvis CT scan in 2014 reported an “incidental” abnormality in his neck. Perusal of the first several articles raised from a “Google” search of the abnormality linked it with possible stroke, carotid dissection and death. Would CASA now cancel or suspend this pilot’s medical? Could this pilot become a casualty of VOMIT (victim of modern imaging technology)? If DAMEs were delegated the responsibility to be able to issue Class 1 certificates, how would many spend the time (and charge commensurately) to perform a more detailed literature search and critical appraisal to support an aero-medical decision one way or the other? This paper discusses the process and time taken to aero-medically assess a pilot who had an incidental radiological diagnosis of a rare condition.

CLINICAL PRESENTATION
Eagle’s syndrome has been described as presenting in one of two ways,

• Classic styloid syndrome that can occur post-tonsillectomy or post-neck trauma. It is thought that scarring can compress cranial nerves V (trigeminal), VII (facial), IX (glossopharyngeal) or X (vagus) manifesting as a foreign body sensation or pharyngeal pain, neck pain, dysphagia, odynophagia, cervico-facial pain, hyper salivation and more rarely temporary voice changes.8-15,18,19

• Stylo-carotid syndrome that is not associated with trauma. In this condition, the stylohyoid apparatus compresses or impinges the internal and/or the external carotid arteries or their perivascular sympathetic fibres, resulting in persistent pain along the carotid artery distribution or ischaemic symptoms associated with head movement.8-11,13,15,18,19,22

Although an initial internet search suggested serious adverse events could arise from Eagle’s syndrome, a Medline search (accessed 28 July 2015) only elicited 136 articles for ‘Eagle’s syndrome’, 113 articles for ‘elongated styloid process’, and only 4 articles on ‘styloid ligament calcification’. Many of the articles described just a single case report of Eagle’s syndrome.

Only three case reports of carotid artery dissection associated with an elongated styloid process were located. These included a previously healthy 43 year old male psychiatrist who sustained an internal carotid artery dissection after a prolonged telephone call while holding the telephone handset between his left ear and shoulder. He was treated with anticoagulation for three months.20 Another case was that of a 41 year old woman who experienced a right internal carotid artery dissection who constantly tilted her head to the right at night when nursing her newborn baby. She was also treated with anticoagulation for three months.21 A further case involved a 60 year old man who developed bilateral internal carotid artery dissection following an “intense shaking” dance. He was initially managed with anticoagulation then subsequent surgical resection of both styloid processes.19

Only two case reports associating sudden death with an elongated styloid process were discovered. Both cases centred on findings at post-mortem and with other causes excluded.18,24

ABOUT THE AUTHOR
Dr Ian Cheng is Staff Specialist – Occupational Physician at Royal North Shore Hospital. He is a consultant in Aviation Medicine, both for the Civil Aviation Safety Authority (CASA) as well as in private practice. He also holds an Adjunct Associate Professor position at James Cook University, College of Public Health, Medical and Veterinary Sciences.

CORRESPONDENCE
Dr Ian Cheng
icheng@asam.org.au

DISCUSSION
Clinical assessment by a vascular surgeon in conjunction with a normal carotid duplex ultrasound scan concluded that this asymptomatic pilot’s carotid circulation was not compromised.

The pilot’s annual CASA medical renewal was submitted together with a covering letter explaining why the ‘diagnosis’ of Eagle’s syndrome, as stated in the CT scan report of the time, was not supported. There are several reasons why this diagnosis was not supported. Firstly, a syndrome can be defined as a group of symptoms and signs that occur together and characterize a particular abnormality or condition. However, as this pilot was asymptomatic then by definition he did not have Eagle’s syndrome. Secondly, his vascular investigations were normal.

Although a general internet-based search yielded information stating that potential dramatic complications from Eagle’s syndrome can occur, a Medline-based search demonstrated that the syndrome itself and risk of fatal or permanent complications was rare. In addition, the published literature was mostly single case reports or case series with small numbers generally describing how surgery relieved patients of their symptoms. A more accurate prevalence or incident rate of Eagle’s syndrome was lacking, owing to publication bias whereby cases that did not have symptom-relief from surgery would unlikely be submitted for publication. Also case reports do not allow one to establish a cause-effect relationship. Finally, an elongated styloid process had not been noted in the pilot’s previous or subsequent neck CT scans.

Fortunately, this pilot did not become a victim of VOMIT (victim of modern imaging technology). The outcome was that CASA issued a renewed Class 1 and 2 medical certificate with his usual audit requirement for ongoing surveillance by his haematologist for his non-Hodgkin’s follicular lymphoma in remission, but without any additional requirements for what was radiologically reported (on one occasion only) as an elongated styloid process.

USE OF EVIDENCE-BASED MEDICAL RESOURCES BY DAMES
Conference delegates responded to a series of interactive questions based around this presentation. When asked about the aeromedical disposition at the time of the initial consultation, 46% of respondents asserted the need for an appropriate understanding of the relevant literature prior to giving any advice regarding aeromedical certification. Another 36% responded that they did not have the time to do this literature search, and would refer the case to CASA for an aeromedical decision. Delegates were subsequently asked if they would charge the pilot for this extra time and effort in researching his rare condition and submitting a report to CASA based on the research and specialist findings. Interestingly, 58% of respondents indicated they would not charge extra for this additional work.

The final question enquired of delegates as to their accessibility to certain medical reference databases: 70% of respondents indicated that they had access to resources such as Up-to-Date® and BMJ Best Practices®, leaving the remaining 30% to operate without ready access to these resources.

CONCLUSION
This case study raises an interesting case of a pilot with a rare, unusual clinical condition for which there is ample ‘evidence’ readily available on the open-source internet. A search of peer-reviewed, scientific journals and contemporary clinical resources can be time-consuming, but is an essential part of the process to provide pilots with appropriate, evidence-based opinion about risk and safety.

Based on the delegates’ responses to these questions, it would appear that most DAMEs would be appropriately cautious making aeromedical decisions in the absence of good clinical evidence. On the other hand, once reasonable scientific literature was available most respondents would be prepared to certify this pilot if they had delegation to do so. It is somewhat interesting to note that a slight majority of respondents would generously give their time freely to research a pilot’s health condition without charging for the time to do so. It is reassuring to note that most DAMEs who attended the conference have access to medical reference databases.

DECLARATION OF INTERESTS
The author is the pilot’s CASA Designated Aviation Medical Examiner. Consent was obtained to present and publish this case.

REFERENCES

Disclaimer: The views expressed in this article are those of the author, and do not reflect a position of the Editor or Committee of the Australasian Society of Aerospace Medicine.