ROYAL AUSTRALASIAN COLLEGE OF SURGEONS

Notes to Candidates
Otolaryngology Head & Neck Surgery Fellowship Examination
September 2016

These notes are to familiarise you all with the format of the examination that you are presenting for shortly.

It is expected that higher level thinking is displayed throughout the exam. The exam is designed to test the application of knowledge and clinical judgement.

THE STRUCTURE OF THE EXAMINATION

There are seven components consisting of two written and five clinical/viva examinations.

Exams are held twice a year with the viva segments being held in May and September.

The May exam alternates between Melbourne (odd years) and Brisbane (even years). The September exam alternates between Adelaide (odd years) and Sydney (even years).

The two Written Exams occur 5-6 weeks before the viva segments of the exam.

Clinical Scenarios are generally held on the Friday, and the Clinical Case and Surgical Anatomy on the Saturday. Surgical Pathology and Operative Surgery take place on the Sunday.

Examiners are paired for each examination. A pair of examiners will mark each candidate in each segment of the exam. Each examiner will score the candidate individually but the pair of examiners will reach an overall consensus mark for each candidate in each segment of the exam.

THE MARKING SYSTEM

Over the last few years there has been an evolution of what was previously termed the Close Marking System to what is now called an Expanded Close Marking System.

Each of the Exam Segments now has a number of defined Marking Points. Each of those Marking Points is scored according to the Close Marking system grades (4 = well above the required standard; 3 = at or above the required standard, 2 = below the required standard, 1 = well below the required standard).

Every candidate’s performance is assessed by 2 examiners in each exam segment. Both examiners score the Marking Points individually for each candidate but will reach an overall consensus grade of 4; 3; 2 or 1 for the candidate in each exam segment. Although each exam segment contains different numbers of Marking Points, the 7 exam segments are equally weighted when determining if overall performance has been satisfactory.

At the end of the Fellowship exam, the Specialty Court in Otolaryngology Head and Neck Surgery (comprising all examiners participating in that exam and the Senior Examiner) meets to discuss all the candidates’ results. Candidates who have been successful in all segments of the exam will pass the Examination. Candidates who have not passed all 7 segments of the exam may still pass the Examination if the Specialty Court considers that their overall performance throughout the exam was satisfactory. The overall performance is based on consideration of the distribution of all the Marking Point grades through all 7 segments of the Examination.
WRITTEN EXAMINATION

Transition to Electronic Delivery

The September 2016 Otolaryngology, Head & Neck Surgery written papers will be delivered electronically, meaning that all answers will be typed on computer (no paper used). However during the transition period, candidates will be given the opportunity to choose a paper based delivery of this examination paper if they prefer.

Due to the change to electronic delivery all candidates, regardless of examination delivery method, will no longer have a specified “reading time” period at the start of the examination. The ten minutes reading time will be added on to the two hours examination time for candidates to use as they see fit, meaning a total examination time of 2 hours 10 minutes (130 minutes).

IMPORTANT INFORMATION (for candidates sitting the computer based version):

1. Answers are typed in the text box provided for each question. The amount of space provided for essay questions is unlimited.
2. Answers are auto-saved every 60 seconds and whenever the 'Next' button is clicked.
3. If a candidate runs out of time, all answers will be submitted automatically and the examination will close.

IMPORTANT INFORMATION (for candidates sitting paper based version):

1. The papers are identified only by your examination number.
2. The written papers are photocopied and sent to the examiners once you have completed your examination.
3. It is important to note that if you use highlighters or different colours in diagrams, or headings, that this does not photocopy well and the point of your diagram may be lost.
4. Please write clearly and use either a black/blue pen. Write only on the lined side of the paper.

Written Paper One

- 2 hours 10 minutes duration
- 1 extended response question (1 hour)
- 4 short response questions (1 hour)

Written Paper Two

- 2 hours 10 minutes duration
- 1 extended response question (1 hour)
- 4 short response questions (1 hour)

Example Answers

A satisfactory written response should include:

- information relevant to the question
- a good understanding of the topic
- most of the important issues regarding the topic
- a good understanding of the relationship of the condition to other disorders in the specialty
- a good idea development supported by facts
- clarity with reasonable detail
- organisation of the major concepts and principles

It’s important to note that future exam questions will be designed to move away from simple “knowledge” based questions (define, list, recall, or name) to questions where analysis, synthesis of knowledge and evaluation is required. This will be tested especially in the long (1 hour) questions.

The specialty court believes that an advanced surgical trainee should begin practising written technique early in training. We would recommend that trial questions be prepared and discussed with your surgical supervisor throughout training, not just in the months leading up to the final fellowship exam.
In order to help improve the quality of the written responses a number of example answers have been provided. These can be found in the Appendix at the end of this document. As a specialty court, we hope that these examples will guide you towards satisfactory completion of the written papers.

**CLINICAL/VIVA EXAMINATION**

The order in which the five clinical/viva components are examined may vary from the order listed below. You will receive a timetable from the Examinations Department closer to the examination date which will outline the order.

**Clinical Scenarios**

- 60 minutes duration

No patients are used in this exam.

This examination consists of 5 clinical protocols. Each protocol consists of a clinical scenario where the candidate obtains a history from the examiners, describes examination techniques, requests and interprets investigations, presents a differential diagnosis, and discusses treatment options.

**Clinical Case**

- 40 minutes duration

Candidates are escorted by the examiners to a number of patients and requested to exam specified regions to elicit clinical signs. Each candidate will see 8 patients, for 5 minutes each.

The exam may include interpretation of investigations relevant to the patients, including imaging, audiograms and other material.

**Surgical Anatomy**

- 30 minutes duration

This examination may be conducted in an anatomy laboratory facility.

You may be examined using wet specimens, computer images, CT and other imaging modalities.

The anatomy examination emphasises the clinical and surgical implications of anatomy.

**Surgical Pathology**

- 30 minutes duration.

You may be shown computer images of pathology specimens, histological slides or clinical photographs and asked to answer related questions.

The surgical pathology examination emphasises the clinical and surgical implications of pathology.

**Operative Surgery**

- 30 minutes duration

You will be examined on aspects of operative surgery which may include pre-operative decision making and workup, operative technique and strategies and management of operative and post-operative complications. Computer images may be shown.

I look forward to meeting you during the exam and at the announcement ceremony at the conclusion of the Examination. For any queries prior to the examination or to request copies of past written papers, please contact the Examinations Department by email on examinations@surgeons.org.

Mr Scott Stevenson
Senior Examiner in Otolaryngology, Head & Neck Surgery
Write short notes on

Primary hemi-facial spasm

- Definition hemi-facial spasm is a type of hyperkinetic disorder involving the facial muscles.
- Hyperkinetic disorders include hemi-facial spasm, bilateral blepharospasm, facial dystonia including Meig’s syndrome and hyperkinesia associated with synkinetic regeneration.
- Hemi-facial spasm is a unilateral disorder

Incidence - Uncommon

Aetiology - Primary hemi-facial spasm is caused by a vascular loop irritating the nerve root entry zone of the facial nerve at the brain stem. The vascular loop is commonly the anterior inferior cerebellar artery.

Other lesions which irritate the facial nerve can masquerade as primary hemi-facial spasm. These lesions include cerebellar pontine angle tumours, intra-temporal lesions as well as intra-parotid lesions.

Pathology

Macroscopically, AICA or a branch thereof is found at the nerve root entry zone of the facial nerve which is the ponto-medullary junction of the brain stem.

From an electro-physiological viewpoint, this causes very high frequency motor unit action potentials resulting in the hemi-facial spasm.

Clinical Findings

A progressive unilateral facial spasm which usually originates in the eye, and gradually becomes progressively severe ultimately causing periods of unilateral functional blindness and severe unilateral facial distortion. There can often be some degree of facial nerve weakness at rest.

Investigations

1. MRI/MRA looking specifically at the vasculature of the brain stem.
2. Angiography should further visualization of the brain stem vasculature be required.
3. EMG showing pathognomonic high firing rate of the motor unit action potentials up to 350 spikes per second (maximum normal during voluntary contraction is 50-70 spikes per second)

Treatment

Neurovascular decompression as described by Janetta accomplished through a posterior fossa approach inserting a Teflon pad between the nerve root entry zone and the vascular loop.

Pros – Up to 90% of these procedures are successful, with complete abolition of hemi-facial spasm in the majority of cases.
Less invasive treatment has been recommended historically. These include:

1. Carbamazapine an anti-epileptic
2. Botulinum toxin injections to the affected muscles
3. Selective neurolosis for selective myectomy
Part 1

a. Describe the epidemiology, natural history, pathophysiology and clinical presentation of glomus jugulare tumours.

b. Define the relevant investigations which determine the extent and classification of glomus jugulare tumours.

Part 2

Discuss the treatment options for glomus jugulare tumours. Include in your answer the outcomes of surgical therapy, and the management of possible complications.

Part 1 and Part 2 of equal value

Note to examiners: This question is not aimed to determine a detailed knowledge of lateral skull base surgery. Rather, the question explores a more general knowledge of the natural history and pathophysiology of glomus tumours, relevant investigations which then determine how the tumour is classified, and then finally a balanced discussion about the realities of treatment which include simple observation, consideration of focused irradiation, and surgical resection focusing on the post-operative complications and their management.

Overview Statement

Glomus Jugulare tumours are rare, slow growing, highly vascular tumours in an area of the skull base which is difficult to access. The tumours produce non-life threatening symptoms in the vast majority of patients. The natural history is that of slow growth producing pulsatile tinnitus, conductive hearing loss and gradually evolving lower cranial neuropathy. Treatment is controversial. Observation is justifiable. Irradiation commonly stabilises these tumours with little additional morbidity. Surgical approaches to glomus jugulare tumours have been developed, but may be associated with life changing morbidity and mortality which must be considered in advising treatment for these patients.

Part 1.

a. The natural history, pathophysiology and clinical presentation of glomus jugulare tumours

Epidemiology

• Chemodectoma or paraganglioma
• The cells of origin are the neural crest cells
• There is a familial tendency probably related to an autosomal dominant genes 11q13 and 11q23 which shows incomplete penetrance.
• Female to male preponderance 2 to 1 (some say up to 6:1)
• Mean age at presentation 50
• Multi centricity of these tumours may occur involving chemoreceptor tissue at the carotid bifurcation, aortic bodies, pulmonary bodies and the vagus nerve.

Natural history

• Rare
• Slow growing
• Locally invasive
• Vascular tumours

Directions of growth from the jugular bulb include:

1. Superior into the floor of the middle ear
2. Inferior into the lumen of the internal jugular vein
3. Medial into the petrous temporal bone to encase the carotid artery
4. Lateral into the middle ear and the external auditory canal
5. Posterior into the occipital and mastoid bone
6. Intracranial into the posterior fossa
Pathophysiology

- Intense vascularity of the tumour produces pulsatile tinnitus
- Progressive enlargement and erosion of the jugular foramen involves the pars nervosum of the jugular foramen resulting in progressive cranial neuropathy involving IX, X, and XI cranial nerves.
- Progressive growth into the middle ear results in a conductive hearing loss.
- Growth in and around the internal carotid artery rarely causes symptoms, but may account for Horner’s syndrome.
- Medial growth into the posterior fossa can produce symptoms and signs of space occupying lesion in the posterior fossa.
- Involvement of the facial nerve within the mastoid may produce a gradual evolving facial neuropathy.
- Posterior growth and erosion of the occipital bone may produce hypoglossal neuropathy.
- Metastasis is said to occur in 4% Clinical Presentation
  The most common presentation is unilateral hearing loss (73%), pulsatile tinnitus (62%), otalgia (36%), facial nerve weakness (24%), otorrhoea (18%), lower cranial neuropathy (18%). Mild hoarseness may be present. Swallowing disorder in the non operated patient is rare. A vascular mass behind an intact drum with a bruit may be present.

b. Define the relevant investigations which determine the classification of glomus jugulare tumours

Investigations:

- High resolution CT scan with intravenous contrast
- Erosion of the jugular foramen. Permeative bone margins, and aircell opacity related to direct extension or obstruction.
- Erosion of the vaginal process separating the carotid canal from the jugular foramen
- Vascular mass centred on the jugular foramen with contrast. MRI
- MRI with gadolinium demonstrates a vascular tumour centred on the jugular foramen involving the intrapetrous carotid +/- the posterior fossa. T1 salt and pepper (flow voids)
- MRA/MRV to demonstrate the blood supply of the tumour. Usually involves the ascending pharyngeal artery. High velocity flow void. Rapid tumour blush and early venous egress.
- Jugular venous occlusion. An intra-luminal mass may be demonstrated on MRV

Angiography

Angiography is usually performed with embolisation of the feeding vessels if surgical resection is anticipated. Angiography is also used to assess carotid artery involvement. If carotid artery damage or resection is anticipated, balloon occlusion may be considered after appropriate trial of occlusion with EEG or Xenon perfusion studies.

Audiometry: Air conduction and bone conduction thresholds. Some describe a saw tooth pattern on tympanometry

Serum and urinary catecholamine estimations do not help with classification, but are required to identify secreting tumours (up to 4%)

Genetic testing for succinate dehydrogenase holding genes SDHB and SDHD are available, and may demonstrate a predisposition to head and neck paragangliomas, phaeochromocytomas, and renal cell carcinomas.

Classification

The FISCH classification is used

a. glomus tympanicum tumours confined to the middle ear space
b. glomus tympanicum tumours confined to the tympanomastoid cleft
c. 1. glomus jugulare tumours with limited involvement of the vertical segment of the carotid artery
  2. glomus jugulare tumours with significant involvement of the vertical section of the carotid artery
  3. glomus jugulare tumours with involvement of the vertical and horizontal section of the carotid artery
d. 1. glomus jugulare tumours with intracranial extension of less than 2cm
  2. glomus jugulare tumours with intra-cranial extension of greater than 2cm.
Management Options

Overview
Following Fisch’s description of infratemporal fossa surgery in the 1970’s, surgical therapy for glomus jugulare tumours was embraced worldwide. During the late 80’s and 90’s, an increased interest in quality of life outcomes has brought about a more reflective approach to glomus jugulare treatment. A rediscovery of the relatively benign natural history of these lesions, as well as an honest appraisal of the post-operative morbidity associated with the often incomplete resection of these tumours now dictates a careful and probably less aggressive approach.

Treatment Options

Treatment options include:

1. Observation
2. Stereotactic radiation therapy
3. Surgical resection

1. Observation
Consists of periodic evaluation of the patient’s symptoms, signs, and imaging. Most authors would recommend observation for all patients over the age of 60, and many would consider observation for patients over 50.

Advantages:
• Avoid surgical procedure

Disadvantages:
• The possibility of increasing growth of the glomus tumour making resection impossible.
• Metastasis - said to be up to 4% over a 30 year period, to bone, lung, liver
• Failure to relieve symptoms

2. Stereotactic radiation therapy or external beam stereotactic radiation
Glomus jugulare tumours are at the skull base, and are therefore accessible with gamma knife collimated radiation therapy. Alternatively, external beam stereotactic radiation therapy may be given.

Advantages:
• Up to 92% cessation of growth over a ten year period claimed.
• Avoids surgical intervention
• Does not add to tumour morbidity

Disadvantages:
• The tumour remains
• Tumour may progressively increase in size despite the radiation therapy (up to 10% of cases)
• If surgery is required, morbidity may be increased due to surgery in an irradiated field
• Malignant tumour development in the radiated field i.e., PTB

3. Surgery
The classic surgical approach to glomus jugulare tumours is the FISCH type A infratemporal fossa approach. More conservative jugulo-petrosectomies have been described avoiding permanent transposition of the facial nerve. This is not expected knowledge.

Elements of the FISCH type A Surgical approach:
1. Excision of the skin of the external auditory canal, drum head, lateral ossicles, and permanent blind sac closure of the external auditory canal with permanent obliteration of the eustachian tube.
2. Permanent anterior transposition of the facial nerve from the geniculate ganglion to the stylomastoid foramen via extended mastoidectomy.
3. Ligation of the sigmoid sinus in the mastoid.
4. Ligation of the internal jugular vein in the neck and surgical access to the internal carotid artery in the neck.
5. Exposure of the jugular bulb by infralabyrinthine temporal bone resection
6. Exposure of the intrapetrous internal carotid artery
7. Opening of the jugular bulb with packing of the inferior petrosal vein, associated with tumour removal from the bulb and jugular fossa and attempted preservation of the lower cranial nerves.
8. Removal of the intra petrous segment of the tumour related to the internal carotid artery
9. Removal of any intra cranial component of the tumour, often performed at a second sitting.

Complications of surgical intervention. Intraoperative

- Bleeding from the tumour, the jugular fossa, and the inferior petrosal vein.
- Injury to the facial nerve at mobilisation
- Inner ear injury
- Injury to CN IX, X, XI and XII
- Injury of intrapetrous carotid artery either inadvertently or planned

Short term complications

- Acute lower cranial nerve neuropathies including cranial nerves IX, X, XI with secondary laryngeal incompetence, aspiration, and inability to swallow requiring cuffed tube intubation, cuffed tracheostomy intubation, and peg feeding. This may be ameliorated by preoperative speech therapy
- Intrapetrous carotid injury resulting in thrombosis, propagating thrombosis and cerebro vascular accident. When combined with ipsilateral multiple LMN cranial neuropathy, devastating.
- Facial nerve injury associated with anterior transposition commonly resulting in incomplete recovery and House Grade III outcome.

Long term complications

- Hoarseness which may require vocal cord medialisation.
- Swallowing abnormalities. May benefit from vocal cord medialisation, cricopharyngeal myotomy, nasogastric feeding, or permanent PEG.
- Facial nerve incomplete recovery. 85% of patients have a House Grade III recovery or better
- Persistent disease, associated with diffuse bone infiltration of the glomus jugulare tumour and incomplete petrous bone resection particularly around the carotid artery requiring radiation therapy.

Prognosis

- Left untreated, glomus jugulare tumours have between a 77% & 94% twenty year survival rate.
- Surgical resection is often incomplete, may be associated with significant mortality and definite ongoing life changing morbidity.

Future direction in glomus jugulare treatment

- Genetic manipulation or family screening to prevent glomus prone patients
- Advances in radiation therapy seem to be promising
Write short notes on:

Vestibular (sound) evoked myogenic potentials.

**Definition:** Vestibular or sound evoked myogenic potentials are inhibitory neural impulses detected in the sternocleidomastoid muscle on presentation of loud sound in the ipsilateral ear.

**Proposed pathway:**
- Sound enters the inner ear by the external canal, tympanic membrane and ossicular chain.
- Sound stimulates the saccule.
- Afferent signal via the inferior vestibular nerve to the vestibular nucleus.
- Internuclear stimulation of the nucleus ambiguous (accessory nerve nucleus).
- Efferent inhibition of the ipsilateral sternocleidomastoid muscle via accessory nerve.
- VEMPS are therefore a test of saccular (static vestibular) and inferior vestibular nerve function.

**How are VEMPS evoked?**
- Headphones supply 110dB click to the ipsilateral test ear.
- Surface measuring electrodes applied to the sternocleidomastoid muscle.
- Both sternocleidomastoid muscles kept under tension by lying patient supine and elevating head from the pillow.
- Inhibitory nerve impulse detected in the ipsilateral sternocleidomastoid.

**Clinical Utility**
1. The detection of superior semicircular canal fistula. VEMPS in this condition show an enlarged amplitude and can be evoked at pathologically low volume.
2. The prediction of benign paroxysmal positional vertigo after vestibular neuronitis. Vestibular neuronitis may involve the superior vestibular nerve, the inferior vestibular nerve or both. The inferior vestibular nerve supplies not only the saccule but the ampulla of the posterior semicircular canal. If the inferior vestibular nerve is destroyed by vestibular neuronitis, then the patient will not suffer from benign paroxysmal positional vertigo after vestibular neuronitis despite canalolithiasis as the PSCC ampulla is denervated.
3. The assessment of endolymphatic hydrops in the contralateral ear in Menieres disease. Saccular dilatation is a hallmark of endolymphatic hydrops. It is proposed that VEMPS may be of use in assessing the contralateral ear in Menieres disease to assess whether it is likely that the contralateral ear will become involved in endolymphatic hydrops. Asymptomatic dilatation has been found in up to 50% of apparently unilateral Menieres disease. This would have therapeutic implications particularly if a destructive procedure was contemplated in the symptomatic ear.

**Marking Guideline**
9 Points should be awarded if the candidate defines VEMPS correctly is aware of the proposed neural pathway and is aware of the utility of VEMPS in the diagnosis of superior semicircular canal fistula. 9½ points should be awarded for knowledge of how VEMPS are evoked, and the utility of VEMPS in BPPV and endolymphatic hydrops.