check

Independent learning program for GPs

Unit 508 August 2014

Head pain

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The five domains of general practice

- Communication skills and the patient-doctor relationship
- Applied professional knowledge and skills
- Population health and the context of general practice
- Professional and ethical role
- Organisational and legal dimensions

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ABOUT THIS ACTIVITY

Headache and head pain are common medical conditions and about 15% of Australians use pain-relieving medication for a headache at any given time.[1] Headaches are classified as being primary or secondary in nature. Primary headaches include migraine and cluster and tension headaches, and are of unknown aetiology, whereas secondary headaches are attributed to underlying problems, such as infection, injury or a tumour. Secondary headaches are very important clinically as some of these, for example thunderclap headache due to a carotid dissection, require urgent medical care.[2–4] In many cases, headaches are benign, but they may decrease overall quality of life.

This edition of *check* considers scenarios of headache and head pain of relevance to general practice.

LEARNING OUTCOMES

At the end of this activity, participants will be able to:

- describe red flags that are relevant in the management of head pain and headaches
- outline current treatment and management options for cluster headache
- explain the risks of combined oral contraceptives (COCs) use for women with migraine and COC options for those who wish to use them
- summarise the defining traits of thunderclap headaches and their management
- specify secondary prevention strategies for transient ischaemic attacks/stroke.

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GUIDE TO ABBREVIATIONS AND ACRONYMS IN THIS UNIT OF *CHECK*

ASA	atrial septal aneurysm
BMI	body mass index
COC	combined oral contraceptives
CrCl	creatinine clearance
CSF	cerebrospinal fluid
CT	computed tomography
ICHD-3	International Classification of Headache Disorders 3rd edition
IIH	idiopathic intracranial hypertension
MRA	magnetic resonance angiogram
MRI	magnetic resonance imaging
NICE	National Institute for Health and Care Excellence
NOAC	non-vitamin K oral anticoagulant
NSAIDs	nonsteroidal anti-inflammatory drugs
PBS	Pharmaceutical Benefits Scheme
PFO	patent foramen ovale
RCVS	reversible cerebral vasoconstriction syndrome
SAH	subarachnoid haemorrhage
SUNCT	short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing
TAC	trigeminal autonomic cephalalgia
TIA	transient ischaemic attack
WHO	World Health Organization

CASE 1

DEBBIE HAS HAD HEADACHES MOST OF HER LIFE

Debbie is 43 years of age and has four children. During her appointment, she comments that she is sick of having frequent headaches. She can recall having headaches for most of her life but the frequency of her headaches has increased over the past 5 years.

QUESTION 1

How would you differentiate the type of headache Debbie has from other types of headaches?

FURTHER INFORMATION

While taking a more detailed history, Debbie describes throbbing headaches on either side of the head lasting for hours to days. Debbie's headaches are associated with photophobia, phonophobia and nausea on at least 4 days a week and last for up to 6 hours per day. She has never had any visual or other disturbance prior to the headaches. The headaches prevent her from fully participating in various activities and, as she finds that they are triggered by heat and exertion, she avoids exercise. The headaches have not changed in character recently but have increased in frequency over the past 5 years.

QUESTION 2

Does it matter which type of headache Debbie has? What is the classification of Debbie's headaches?

QUESTION 3

What questions would you want to ask about Debbie's lifestyle with regard to her increased headache frequency?

FURTHER INFORMATION

Debbie has become overweight and has avoided exercise for years because of her frequent migraines. Since having her four children (the youngest child is now seven), she has had markedly disrupted sleep and always feels tired, so she drinks two cans of caffeinated soft drink and several cups of tea daily. Her worst migraines are often at the time of her menstrual periods and these don't respond to her usual nonsteroidal anti-inflammatory drugs (NSAIDs). Although she avoids many activities due to frequent disruption by her headaches, she denies any symptoms of depression or anxiety.

QUESTION 4

What management strategies could be used to target these comorbidities?

QUESTION 5

What pharmacological management would be most appropriate for this patient?

CONCLUSION

Debbie agrees to start topiramate as migraine prophylaxis, as her blood pressure was too low to tolerate propranolol previously, and she refuses pizotifen because of the unacceptable risk of further weight gain. Over the next 3 months she tolerates topiramate without significant side effects. She gradually has a marked improvement in her migraines, with predominantly menstrually related migraines remaining, which can be effectively managed with her triptan. She has been able to return to regular exercise and has a part-time job in the school canteen.

CASE 1 ANSWERS

ANSWER 1

Clinical features define primary headache disorders so a careful history is useful in differentiating between headache types. Salient questions for Debbie would include asking about the characteristics and duration of her headaches, preference for rest or ability to perform activities, and associated features such as sensitivity to light or noise, and nausea or vomiting. *Table 1* summarises information

Table 1. Diagnosis of tension-type headache, migraine and cluster headache[1]

Headache feature	Tension-type headache		Migraine (with or without aura)		Cluster headache	
Pain location*	Bilateral		Unilateral or bilateral		Unilateral (around the eye, above the eye and along the side of the head/face)	
Pain quality	Pressing/tightening (non-pulsating)		Pulsating (throbbing or banging in young people aged 12–17 years)		Variable (can be sharp, boring, burning, throbbing or tightening)	
Pain intensity	Mild or moderate		Moderate or severe		Severe or very severe	
Effect on activities	Not aggravated by routine activities of daily living		Aggravated by, or causes avoidance of, routine activities of daily living		Restlessness or agitation	
Other symptoms	None		Unusual sensitivity to light and/or sound or nausea and/or vomiting Aura† Symptoms can occur with or without headache and: • are fully reversible • develop over at least 5 minutes • last 5–60 minutes. Typical aura symptoms include visual symptoms such as flickering lights, spots or lines and/or partial loss of vision; sensory symptoms such as numbness and/or pins and needles; and/or speech disturbance		On the same side as the headache: • red and/or watery eye • nasal congestion and/or runny nose • swollen eyelid • forehead and facial sweating • constricted pupil and/or drooping eyelid	
Duration of headache	30 minutes–continuous		4–72 hours in adults 1–72 hours in young people aged 12–17 years		15–180 minutes	
Frequency of headache	<15 days per month	≥15 days per month for more than 3 months	<15 days per month	≥15 days per month for more than 3 months	1 every other day to 8 per day‡, with remission§ >1 month	1 every other day to 8 per day‡, with a continuous remission§ <1 month in a 12-month period
Diagnosis	**Episodic tension-type headache**	**Chronic tension-type headache¶**	**Episodic migraine (with or without aura)**	**Chronic migraine# (with or without aura)**	**Episodic cluster headache**	**Chronic cluster headache**

*Headache pain can be felt in the head, face or neck.

†See recommendations 1.2.2, 1.2.3 and 1.2.4 in the NICE guidelines[1] for further information on diagnosis of migraine with aura.

‡The frequency of recurrent headaches during a cluster headache bout.

§The pain-free period between cluster headache bouts.

¶Chronic migraine and chronic tension-type headache commonly overlap. If there are any features of migraine, diagnose chronic migraine.

#NICE has developed technology appraisal guidance on botulinum toxin type A for the prevention of headaches in adults with chronic migraine (headaches on at least 15 days per month of which at least 8 days are with migraine).

Reproduced with permission from the National Institute for Health and Clinical Excellence (2012) CG150 Headaches: diagnosis and management of headaches in young people and adults.[1]

from the National Institute for Health and Care Excellence (NICE) 2012 guidelines,[1] which can be used to differentiate between migraine, tension type headaches and cluster headaches. These guidelines caution against routinely referring people with migraine for neuroimaging in the absence of red flags solely for the purpose of providing reassurance.[1] The Australian Therapeutic Guidelines red flags for prompting further investigation are listed in *Table 2*.[2]

Table 2. Warning signs in the diagnosis of headache[2]

Type of headache	Possible organic causes
Sudden onset, particularly with confusion, drowsiness or vomiting, or with mild stroke-like symptoms or signs (eg mild hemiparesis, ataxia, Horner syndrome, diplopia including sixth nerve palsy)	Subarachnoid or intracranial haemorrhage, carotid or vertebral artery dissection, cerebral venous thrombosis, thunderclap headache, reversible cerebral vasoconstriction syndrome
Recent onset with confusion, drowsiness or fever	Meningitis, encephalitis, intracranial abscess, severe hypertension (hypertensive encephalopathy)
Recent onset in a young obese patient	Idiopathic (benign) intracranial hypertension (look for papilloedema)
Recent onset in a patient over 50 years of age	Brain tumour, giant cell arteritis (temporal arteritis), cervicogenic, medication overuse, subdural collection, herpes zoster, sinusitis
Recent onset with cough, exertion or sexual activity	Subarachnoid haemorrhage, brain tumour
After head injury, particularly with loss of consciousness, or if severe or prolonged	Intracranial haemorrhage

ANSWER 2

According to the International Classification of Headache Disorders, 3rd edition (ICHD-3), beta version classification, Debbie has chronic migraine.[3]

Chronic migraine occurs in approximately 2% of the adult population in Western countries.[4] It is defined as headache that occurs on 15 or more days per month for more than 3 months and has the features of migraine headache on at least 8 days per month.[3] Chronic migraine is considered a complication of episodic migraine; 2.5% of people progress yearly from episodic to chronic migraine.[5] It is the most disabling of the different types of chronic daily headaches, yet is often underdiagnosed and undertreated.[4] In an Australian cohort, only 20% of patients with more than three headaches per month were receiving prophylaxis, while the remaining 80% had either never received such treatment (66%) or had tried but stopped migraine prophylaxis (33%).[6] Correct diagnosis is important as it determines prognosis and the best ongoing management options.

Debbie has never had aura. Migraine without aura is defined as headaches lasting 4–72 hours (when untreated or unsuccessfully treated) with at least two of the following four characteristics: unilateral location, pulsating quality, moderate or severe pain intensity, and aggravation by or causing avoidance of routine physical activity (eg walking or climbing stairs), and at least one of the following: nausea and/or vomiting, and photophobia and phonophobia.[1,3]

ANSWER 3

In patients with episodic migraine, modifiable risk factors for development of chronic migraine include increasing attack frequency, obesity (body mass index >30 kg/m^2), life stressors, snoring/sleep apnoea/sleep disturbance and caffeine consumption. Age, gender, low socioeconomic status, head injury and allodynia are also associated with chronic migraine but are less modifiable risk factors.[7–9] Anxiety and depression are common comorbidities with chronic migraine and may affect central antinociceptive networks.[7]

ANSWER 4

Educating the patient about reducing triggers by improving sleep hygiene, having regular meals and caffeine cessation may be useful.[10,11] Relaxation training, biofeedback, stress management and cognitive behavioural therapy are recommended.[2,12] Limited studies suggest a role for weight loss and exercise in the prevention of migraine.[13,14] In this case, by recognising the more difficult-to-treat menstrually related migraines, the patient could be taught to target those specific migraines earlier and more effectively by taking a triptan, without overusing triptans at other times of the month. The lowest dose of a triptan is recommended, followed by the higher dose if the low dose is ineffective.[2]

ANSWER 5

An effective acute headache management plan is limited in chronic migraine because of the high frequency of headaches. There is a risk of medication overuse headaches if analgesics are taken on more than 15 days per month (only 9 days per month are required if strong analgesics such as codeine or triptans are used frequently).[15] Migraine prophylaxis is therefore important. The goals of migraine prophylaxis are to reduce attack severity, frequency and duration, to improve responsiveness to acute treatments and to reduce disability.[12] The Australian Therapeutic Guidelines recommend choosing the prophylactic medication on the basis of potential adverse side effects. According to these guidelines, amitriptyline, pizotifen and propranolol are recommended as first-line prophylaxis, while sodium valproate, topiramate and verapamil are second-line options.[2] The American Academy of Neurology and the European Federation of Neurological Societies have published evidence-based recommendations regarding migraine prophylaxis, based on the quality of evidence.[12,16] Many of the migraine prophylactic medications, such as candesartan and gabapentin are not indicated for migraine on the Pharmaceutical Benefits Scheme (PBS).[17] Other agents, such as vitamin B2, feverfew and magnesium, could also be offered.[18]

When starting migraine prophylaxis, the patient should be given realistic goals over several months (such as a 50% reduction in headache days), with education about correct dosage and possible side effects. The patient should be reviewed after 2 months to adequately assess effectiveness.[12,19] An objective headache diary is

useful for the doctor and patient's conviction in identifying a gradual response to management over the subsequent months. If headaches are resistant to treatment, referral to a specialist in headache for further migraine prophylaxis options, including onabotulinum toxin A may be appropriate in selected cases.[20] Onabotulinum toxin A (or botulinum toxin) is marketed in Australia for migraine prevention in headache occurring on at least 15 days/month.[21]The overall aim should be to reduce the migraine days from chronic migraine to episodic migraine by a combination of lifestyle and prophylactic management.

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RESOURCES FOR PATIENTS

- Information and a register for migraine patients is available at http://headacheaustralia.org.au/migraine
- An easy-to-use and informative headache diary for patients is available at www.aspenpharma.com.au/patRes/2014_Headache_Diary.pdf

RESOURCES FOR DOCTORS

- American Headache Consortium Guidelines, http://eguideline.guidelinecentral.com/i/76857/5
- The International Classification of Headache Disorders, 3rd edn (beta version), www.ihs-classification.org/_downloads/mixed/International-Headache-Classification-III-ICHD-III-2013-Beta.pdf

CASE 2

LISA HAS THROBBING HEADACHES

Lisa, 21 years of age, smokes and binge drinks on weekends. She has a 2-month history of twice weekly episodes of throbbing headaches lasting less than 12 hours and requiring rest in a dark room. Over-the-counter pain relievers, such as non-steroidal anti-inflammatory drugs (NSAIDs) and paracetamol, have been only marginally effective. You take a history and enquire about headache red flags, and perform a general physical and neurological examination. Vital signs and fundoscopy are normal. You diagnose migraines and provide Lisa with some sample packs of an oral triptan medication. A follow-up appointment in a fortnight is scheduled.

QUESTION 1

What type of historical findings in a headache history would be considered red flags for serious problems?

FURTHER INFORMATION

Lisa arrives for her follow-up appointment accompanied by her mother. Lisa cannot cope with her university studies. Over the past fortnight she used the triptan samples and had to resort to codeine-containing analgesics for six episodes of migraines. Lisa had two episodes of transient visual symptoms in her right eye, manifesting with blurred and shimmering central vision, which lasted for 15 minutes; peripheral vision was unaffected and there was no headache in the aftermath.

After collecting additional history with her mother's help, you establish that Lisa has had infrequent migraines since the age of 14 years, but had never experienced visual disturbances or neurological symptoms. There is a family history of migraines and her brother had a structural heart defect. You learn that 3 months ago Lisa started using a combined oral contraceptive (COC) pill. You instruct Lisa to stop taking the COC pill and start low-dose aspirin. Recently you learned that GPs can request a rebatable magnetic resonance imaging (MRI) scan of the brain for unexplained headaches with suspected intracranial pathology and you arrange a semi-urgent MRI brain scan to be carried out within 1 week.

QUESTION 2

What are common triggers that can elicit a migraine attack or increase the frequency and severity of migraine attacks in a person who has a history of migraines?

QUESTION 3

What is your diagnosis for Lisa's headaches? What is your diagnosis for Lisa's new onset visual symptoms? How are you going to evaluate Lisa's new visual symptoms?

QUESTION 4

What structural cardiac condition is common in patients with migraine that may confer an increased risk of ischaemic stroke?

CONCLUSION

A week later you receive an urgent call from a radiologist. Lisa's MRI revealed evidence of acute embolic stroke mainly in the right anterior cerebral artery territory, as well as evidence of a small ischaemic infarct in the right middle cerebral artery

territory. After discussing the results with Lisa and her mother, you establish that 3 days before the MRI scan, during a family reunion, Lisa had a witnessed episode of sudden-onset left leg weakness and speech arrest, which resolved within 10–15 minutes. She had some mild headaches in the aftermath, but no residual neurological deficit. Lisa is admitted to a tertiary care hospital where she is found to have PFO with ASA. An interventional cardiologist carried out percutaneous closure of PFO using a small umbrella-like device.

CASE 2 ANSWERS

ANSWER 1

As most patients with headaches have normal general physical and neurological and examinations, a thorough history is crucial to screen for headache red flags, which suggest underlying systemic or intracranial pathology. *Table 1* lists red flags that might be uncovered during history taking for headache presentations and possible differential diagnoses for red flag findings.[1]

Table 1. Headache red flags and possible differential diagnoses[1]

Red flags	Differential diagnoses
• Sudden onset of headaches • Thunderclap headaches • Precipitation of headache with Valsalva manoeuvres • Persistent strictly unilateral headaches	Subarachnoid haemorrhage Pituitary apoplexy Haemorrhage into a mass lesion or vascular malformation Mass lesion (especially posterior fossa mass) Upper cervical spine pathology
• Headache beginning after 50 years of age	Temporal arteritis Mass lesion
• A change in frequency and severity of headaches and emergence of new neurological symptoms	Vascular pathology Mass lesion Subdural haematoma Medication overuse
• Headache in young overweight people	Idiopathic intracranial hypertension Cerebral sinus thrombosis
• Headaches subsequent to trauma	Intracranial haemorrhage Subdural haematoma Epidural haematoma Post-traumatic headache
New onset headache in: • a patient with systemic illness, fever, cancer or human immunodeficiency virus • pregnancy or the postpartum period • patients on anticoagulants and immunosuppressant medications	Meningitis Encephalitis Metastasis Vasculitis Any of the differential diagnoses in the above categories

ANSWER 2

Common triggers that may elicit a migraine attack or increase the frequency and severity of migraine attacks include:[2]

- stress or relaxation after periods of stress (stress can include bright lights, loud noise, long-distance travel and extremes of weather)
- anxiety and depression
- dehydration
- sleep deprivation or excessive sleep
- missed meals
- trauma to the head or neck
- oral contraceptives and vasodilators such as glyceryl trinitrate.

Dietary factors include cheese, chocolate, alcohol and citrus fruits (these are only occasionally important in management and too much effort in identifying them may be counterproductive).

ANSWER 3

According to the International Classification of Headache Disorders, 3rd edition (ICHD-3), beta version diagnostic criteria, Lisa's headaches are consistent with the diagnosis of migraine without aura.[3] According to these criteria, her visual symptoms are consistent with the diagnosis of migraine manifesting with typical aura without headache.[3] An older term that describes these headaches is 'acephalgic migraine'.

If a patient has a long history of stereotypical migraine auras with and without headaches, but has no deficits found on the physical or neurological examination, a complete work-up with laboratory and imaging tests is probably not mandatory. However, a complete evaluation should be undertaken if a patient presents with new-onset migraine aura without headaches.[4] An MRI scan of the brain should be carried out within 10–14 days of having such an episode of migraine to rule out an ischaemic stroke. A cerebral MR angiography and carotid Doppler are used to assess for intra- and extracranial stenosis, due either to arteriosclerosis or a vasculitis. Other ancillary investigations may include serological testing for thrombophilia and systemic vasculitis. Practitioners are reminded that Medicare rebates apply for thrombophilia screening or testing for specific factor deficiencies only if the patient has a personal proven history of venous thromboembolism or has a first-degree relative with a proven factor defect.[4] Epidemiological studies have suggested that the risk of ischaemic stroke is slightly increased in women with migraines, with the risk reported to be substantially higher in certain sub groups of women.[5,6]

ANSWER 4

Over the last decade, patent foramen ovale (PFO) associated with atrial septal aneurysm (ASA) has been identified as an independent risk factor for ischaemic stroke, particularly in young adults with cryptogenic stroke.[7] Recently, migraine was found to be significantly associated with PFO when compared with controls. At least 50% of patients with migraine with aura were found to have a PFO, and the size of the intracardiac shunts was larger in patients with migraines

than in controls.[8] In susceptible women, the association between migraines with aura and PFO leads to an increased risk of ischaemic stroke.[7] In terms of pathophysiology, it has been proposed that in this population of women, COCs may trigger venous embolism with subsequent paradoxical embolism through PFO or formation of a mural thrombus locally in the atrial septum within the conduit of PFO.[9]

Importantly, an isolated finding of PFO without any other high-risk features in an asymptomatic individual is of equivocal clinical significance. Routine screening for PFO is not recommended.[7–9]

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CASE 3

STEVEN HAS AN ONGOING HEADACHE

Steven, 57 years of age, comes to see you today with his wife to discuss a 3-week history of a constant unilateral retro-orbital headache. His wife states that he has looked more tired than usual since the onset of the headache.

QUESTION 1

What are the red flags in this history? What would point to a secondary cause of headache during history taking?

FURTHER INFORMATION

Steven has been healthy without any significant past medical history and he does not take any regular medication. He denies significant headaches in the past. One night 3 weeks ago, Steven was woken up by a sudden-onset, severe headache. He took paracetamol and the headache improved slightly. The pain was over his left eye and it was constant. He denies having had any head or neck trauma before the symptoms started. The pain over the left eye continued for several weeks and has gradually improved.

QUESTION 2

What are your differential diagnoses for thunderclap headache?

FURTHER INFORMATION

On examination, Steven has left partial ptosis and miosis (*Figure 1*). The difference in pupil size is enhanced in the dark and pupillary responses are normal. The remainder of the visual and neurological examinations are normal. There is no lack of sweating on the left side versus the right. Left carotid artery is palpable without any bruits.

Figure 1. Eye examination

QUESTION 3

What is his diagnosis? How would you investigate further?

FURTHER INFORMATION

You order a CT scan of Steven's orbit, cavernous sinus and pituitary fossa as part of the initial workup investigation and it is unremarkable. A magnetic resonance imaging scan (MRI) with a magnetic resonance angiogram (MRA) of the aortic arch to the Circle of Willis, which is now covered by Medicare when ordered by a GP, is performed and reveals a left internal carotid artery dissection (*Figure 2*).

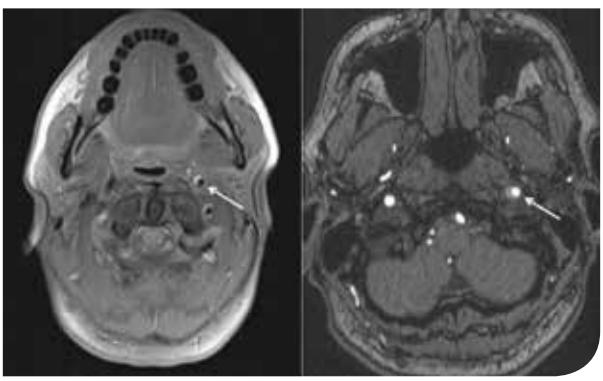

Figure 2. Axial T1 MRI and time-of-flight MRA showing a crescent sign in the left internal carotid artery

QUESTION 4

How would you manage Steven?

FURTHER INFORMATION

Steven was started on aspirin and comes back for follow up 3 months later. He asks 'will it happen again?'.

QUESTION 5

How would you respond to Steven's question?

CASE 3 ANSWERS

ANSWER 1

Given the very short history in this presentation, it is difficult to determine the underlying cause of the headache and whether it is likely to have a serious pathophysiology. Primary headache presentations (ie migraine, tension, cluster headache) most often have a history of similar attacks in the past.[1] Steven's age (57 years) is another factor that may indicate that this headache may not be benign in aetiology.[2] Patients over the age of 50 years who present with a new headache should always be investigated and their concerns taken seriously.[3] A good rule of thumb is that the 'first or worst' headache should prompt investigation. However, severity is not always a reliable guide as primary headaches are some of the most severe on the pain spectrum.[1]

Other important points on history that may indicate a secondary cause include information regarding headache onset (ie gradual versus thunderclap presentations; *Table 1*).[4–6] The latter tend to indicate a vascular presentation.[7]

Table 1. Warning signs for secondary headache[4–6]

Thunderclap presentation
• Usually indicative of vascular presentations
Age >50 years
• Unusual for primary headaches to present for the first time in this age group
New headache
• Sinister aetiologies usually become apparent within 6 months
Systemic symptoms
• Infectious or inflammatory disorders may have fever or other organ involvement
Focal signs
• Persistent signs such as cranial nerve dysfunction, diplopia and papilloedema should always suggest secondary cause and warrants careful investigation
Confusion/agitation
• Can be the end result of several secondary causes
Cough-only headache
• Indicative of foramen magnum lesions
Postural headache
• May suggest low or high pressure headache
Medical comorbidities
• Cancer history • Immunosuppression/HIV • Pregnancy • Anticoagulants

Focal neurological signs are also paramount in a headache presentation.[8–10] Patients with cranial nerve symptoms or signs require a thorough investigation of the base of the skull, cavernous sinus or subarachnoid space. Aphasia, hemiparesis, hemisensory loss or visual loss are also important to note as they may indicate a space-occupying lesion.[11]

Systemic symptoms such as fever, myalgia, anorexia and weight loss should be sought and may suggest underlying infectious, inflammatory or malignant processes.[3,12] Confusion or agitation may occur with a number of different processes and is a potentially serious finding.[13]

Several medical conditions should lower the threshold for further investigation. A prior significant cancer history should prompt concern for intracerebral metastasis. Immunosuppression should raise concern about a meningitic process or opportunistic infection.[3] Anticoagulant use and a history of mild trauma should prompt investigation for a subdural haemorrhage.[14] Pregnancy and the postpartum period is a risk factor for cerebral venous sinus thrombosis.[15] Previous neurosurgery or ventriculoperitoneal shunting is a risk factor for intracranial infection or cerebrospinal fluid (CSF) pressure-related headache.[16] Obesity or use of retinoids or tetracycline should raise concern about idiopathic intracranial hypertension (IIH).[17–19]

Other characteristics of Steven's headache may provide further clues. A cough-only headache can suggest a foramen magnum lesion, particularly a Chiari malformation.[20] A headache that changes significantly with posture (ie lying versus standing) tends to suggest a low or high CSF pressure headache.

A few other rules of thumb apply with headache presentations:

- Headache severity is not always a reliable sign as migraine presentations can be associated with severe pain and frank neurological signs.[21]
- Space-occupying lesions and brain tumours rarely present with headache in isolation and almost always have focal signs or symptoms.[22]
- Fundoscopy should be performed in all patients as IIH is easily missed if papilloedema is not looked for specifically.

ANSWER 2

Thunderclap headache is used to describe a headache that reaches its peak intensity within 60 seconds.[4,6] The most feared condition with this presentation is subarachnoid haemorrhage.[6] Urgent assessment is usually indicated with an emergency department admission and computed tomography (CT) imaging.[6] Most of the other important differential diagnoses are vascular in origin and are listed in *Table 2*.[4,6] If there has been a delay of several weeks with the presentation and the patient is otherwise well, a less urgent approach can be undertaken. This is because the danger period is fairly early with most of the differential diagnoses.[23,24] Primary thunderclap headache is a diagnosis of exclusion,[6] and vascular causes require exclusion first.

Table 2. Differential diagnoses for thunderclap headache presentations[4,6]

- Subarachnoid haemorrhage
- Acute subdural haemorrhage
- Dissection of cervical arteries
- Reversible cerebral vasoconstriction syndrome
- Cerebral venous thrombosis
- Pituitary apoplexy
- Intracranial hypotension
- Hypertensive crisis/phaeochromocytoma
- Primary thunderclap headache/migraine

ANSWER 3

Steven has Horner's syndrome, affecting the left eye. Horner's syndrome is classified into first-, second- and third-order neuron lesions (*Table 3*). The different degrees of neuron lesions may be associated with other clinical signs.[25] The sympathetic nerve supply to the eye takes a long course from the hypothalamus, down the brainstem and cervical cord, and exits at the lower cervical level near the brachial plexus. It then ascends in the neck and lies in close proximity to the internal carotid artery all the way into the orbit. Given the long course of this nerve supply, various differential diagnoses are possible, but can be narrowed by looking for associated features. *Table 3* summarises the location, aetiology and suggested investigations for Horner's syndrome.[25] More sophisticated pharmacological tests can also be used to determine the site of Horner's syndrome.[25] This is usually only performed in difficult cases and by those with experience. If the diagnosis and underlying aetiology are unclear, referral to a neurologist may be indicated.

Table 3. Horner's syndrome[25]

Neurone	Location	Aetiology	Investigations
First-order	Brainstem C-spine C8–T2 spine	Brainstem stroke Arnold-Chiari Trauma, syringomyelia	Brain MRI and cervicomedullary junction
Second-order	C8–T2 nerve root Lung apex Sympathetic chain	Cervical rib Lower brachial plexus injury Tumours Subclavian artery aneurysm	MRI of the cervical spine, chest CT +/– MRI brachial plexus
Third-order	Carotid artery Cavernous sinus orbit	Carotid dissection, aneurysm, arteritis Base of skull tumours or mass *Herpes zoster*	Vascular imaging of the carotid artery via CTA/MRA MRI of base of skull

The sudden onset of symptoms in Steven's case suggest a vascular cause is highly likely and further vascular imaging of the carotid arteries should be obtained.

ANSWER 4

Involvement of a stroke neurologist/physician may be indicated. The priority in carotid artery dissection is stroke prevention. The true incidence of stroke following dissection is unknown.[26] It is likely that many patients have minimal symptoms and do not present at all. Carotid artery dissection is a common cause of stroke in the younger population (<45 years of age) and may be responsible for up to 20% of strokes in this subgroup.[27]

Antithrombotic therapy has a theoretical advantage in that it may prevent thrombus formation at the site of occlusion and also distal embolisation.[26] There are no randomised controlled trials that compare the effectiveness of antiplatelet therapy or anticoagulation with placebo or with each other.[28] A 2010 Cochrane review found no evidence to support either strategy.[28] Several observational studies have been conducted and a meta-analysis published in 2012 showed that there is no difference in outcomes with either treatment in terms of mortality or risk of ischaemic stroke.[29]

Notwithstanding the above, a minimum of 3–6 months of antithrombotic treatment is usually necessary as this is the time frame in which recanalisation and vessel healing occur.[30] Most experts tend to use anticoagulants for dissections associated with ischaemic symptoms or stroke, in line with 2011 American Heart Association/American Stroke Association (AHA/ASA) guidelines, which recommend use of antithrombotic therapy for 3–6 months for patients with ischaemic stroke or transient ischaemic attack, and arterial dissection, even in light of the uncertainties about the relative efficacy of antiplatelet agents versus anticoagulation therapy.[31] Those without ischaemic symptoms are managed with antiplatelet therapy.[26]

ANSWER 5

The risk of recurrent dissection is 2% in the first 3 months and the risk of recurrent stroke or ischaemia is 2%.[32,33] The risk of stroke is highest within the first few weeks following a dissection.[33–35] Steven can be reassured, therefore, that his risk of another episode is very low.

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CASE 4

EMMA HAS HEADACHES WITH ARM TINGLING AND NUMBNESS

Emma, aged 20 years, is a university student who presents with a history of headaches for the past 3 years. The first episode occurred during Year 12, shortly after exams. At the time she experienced tingling in her left hand that gradually progressed up to the shoulder and then spread to her lips and tongue. Symptoms lasted for about 20 minutes and were followed by numbness in the affected area for an additional 10–15 minutes before normal sensation returned. However, 30 minutes later, a moderately severe headache started in the left retro-orbital and temporal regions. It progressed to a severe level over 30 minutes, at which point she also experienced nausea and vomiting and was compelled to lie down in a dark, quiet room. The headache waxed and waned over 2 days, after which she felt tired and 'washed out'. Over the past 3 years, similar episodes have occurred sporadically, every 1–3 months and usually in the setting of stress. She finds that paracetamol and ibuprofen sometimes decrease the pain but generally she needs to 'wait it out'. Recently, she was unable to do an exam due to a headache, which prompted a visit to another GP at your practice.

QUESTION 1

What is Emma's headache diagnosis?

QUESTION 2

What are the stages of migraine?

QUESTION 3

What are the typical features of migraine aura?

QUESTION 4

Are any investigations required?

QUESTION 5

How should Emma treat her migraine attacks?

QUESTION 6

Is preventive treatment indicated? What considerations should be made when choosing a treatment?

QUESTION 7

What lifestyle modifications would you recommend?

FURTHER INFORMATION

Six months later Emma returns for a follow-up consultation. She has had three attacks of migraine with aura, which responded within 30 minutes to a combination of sumatriptan nasal spray and naproxen. She is here today to speak to you about starting the birth control pill.

QUESTION 8

What do you advise her about oral contraceptives?

CASE 4 ANSWERS

ANSWER 1

Emma's headaches are most consistent with migraine with aura.

Migraine is a recurrent headache disorder characterised by attacks lasting 4–72 hours. Typical features of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity, and association with nausea and/or photophobia and phonophobia.[1] Paraesthesia and numbness are characteristic of migraine aura and are discussed in detail in the following questions and answers.

The 3-item ID Migraine screener[2] is a validated and reliable screening instrument for migraine headaches and can improve migraine recognition in primary care settings. It consists of three questions:

- Are you nauseated or sick to your stomach when you have a headache?
- Does light bother you when you have a headache?
- Has a headache limited your activities for a day or more in the last 3 months?

A positive answer to two or more questions translates into a 93% positive predictive value for migraine.

The differential diagnoses for Emma's presentation include:

- Hemiplegic migraine, which is a form of migraine with aura in which the aura includes motor weakness.[1]
- Chronic migraine, defined as a headache occurring on 15 or more days per month for more than 3 months, which has the features of migraine headache on at least 8 days per month.[1]
- Tension-type headache, which is usually bilateral and mild or moderate in severity and lasts from minutes to days. It is not associated with nausea but photophobia or phonophobia may be present.[1]

- Cluster headaches, which are characterised by attacks of severe, unilateral pain in the orbital, supraorbital and/or temporal region, and has ipsilateral autonomic features (ie conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, facial sweating, miosis, ptosis and/or eyelid oedema). Attacks last for 15–180 minutes and occur from once every other day to eight times per day during 'cluster periods', which may last for weeks or months.[1]

ANSWER 2

Migraine comprises several stages:

- prodrome
- aura
- headache
- postdrome.

Patients may experience some or all of the stages. Most migraineurs have a prodrome phase but it is often unrecognised, as symptoms may occur hours to days before the headache onset. Prodromal or premonitory symptoms are often vague or non-specific but tend to be consistent for the individual. Typical prodromal symptoms include euphoria, depression, difficulty concentrating, hyperactivity, repetitive yawning, excessive thirst, anorexia or cravings for sweet or salty foods.[3]

Postdromal symptoms are more readily recognisable and may persist for 24 hours after the headaches. Typical postdromal symptoms include irritability, fatigue, euphoria, muscle weakness or myalgias, anorexia or food cravings.[3]

ANSWER 3

Typical aura may be visual, sensory and/or dysphasic in nature. Visual aura is the most common type of aura and can present as positive features (flickering lights, spots or lines) and/or negative features (loss of vision). It often presents as a 'fortification spectrum', a zigzag figure that may gradually spread right or left, developing into a convex shape with a scintillating edge and leaving absolute or variable degrees of scotoma after it fades.[1] Sensory auras are less common but often follow a typical pattern of positive features ('pins and needles') in the point or origin with gradual involvement of all or part of one side of the body, face and/or tongue. Numbness may occur after paraesthesia fades, but numbness may be the only symptom. Dysphasic speech disturbances are least common. Aura symptoms may occur in succession. Although the sequence usually progresses from visual to sensory to aphasic symptoms, other sequences may occur.[1]

Symptoms of migraine aura are characterised by several features:[1]

- complete reversibility
- gradual development, either alone or in succession, over at least 5 minutes
- duration of each symptom is 5–60 minutes
- at least one aura symptoms is unilateral
- the aura is accompanied, or followed within 60 minutes, by headache.

Although weakness occurs during the aura in patients with familial or sporadic hemiplegic migraine, motor weakness is not a typical aura symptom and warrants further evaluation. Of note, patients with aura characterised by numbness may describe 'heaviness' in the affected limb, which can be mistaken for weakness.

ANSWER 4

Emma's aura symptoms are stereotypical of migraine with aura and therefore imaging and investigations are not required. Patients should not be referred for neuroimaging solely for the purposes of providing reassurance.[4] Investigations are indicated for patients who present with or without migraine headache and any of the following atypical aura symptoms:[4]

- motor weakness
- double vision
- visual symptoms affecting only one eye
- poor balance
- decreased level of consciousness.

Other headache red flags that warrant further evaluation include:[5]

- first or worst severe headache
- change in the pattern of previous migraine
- abnormal neurologic examination
- recent head trauma
- headache triggered by cough or Valsalva
- onset of migraine after age 50 years
- new onset of headache in an immuncompromised patient
- headache with fever.

ANSWER 5

In patients with mild or moderate migraine pain, a combination of antiemetics, simple analgesics and non-steroidal anti-inflammatory drugs (NSAIDs) can be used as an initial step. Triptans should be used if pain progresses to a moderate-to-severe intensity.[6]

Moderate-to-severe migraines should be treated early in the attack with a triptan. The evidence for combining triptans with aspirin or other NSAIDs and metoclopramide is limited but these combinations could be considered if a triptan alone is not beneficial.[6] Triptans are efficacious in about two-thirds of people and some benefit is usually experienced within 30–60 minutes of oral ingestion.[6] Antiemetics (metoclopramide or procholeperazine) may be added as well.[6] Given the occurrence of nausea and vomiting early in Emma's migraines, non-oral formulations (eg sumatriptan nasal spray, NSAID suppositories, rizatriptan wafers) are likely to be most effective. If the migraine improves initially but subsequently recurs, the triptan dose may be repeated in 2–4 hours. If, however, there is no initial response, a second dose is unlikely to be effective. If the triptan is consistently ineffective in treating migraines other triptans should be trialled.[6]

Opioids, including codeine preparations with paracetamol or aspirin, should be avoided in acute migraine management[7] as opioids have

little evidence for efficacy and they can predispose patients to the development medication-overuse headache.[6] As codeine-containing medications are easily accessible in Australia, patients should be asked directly about their intake of these medications and should be educated early about the risks of transformation of their problem to a chronic daily headache pattern.

ANSWER 6

In general, preventive treatment should be initiated in patients with migraines with 2–3 or more attacks per month.[6] However, preventive medications may be considered for migraines occurring at any frequency that interferes with the patient's quality of life. In Emma's case, as the migraines occur every 1–3 months, the optimisation of acute treatment and implementation of lifestyle modifications are appropriate first steps in management.

The goals of migraine prevention are to decrease the frequency and severity of the attacks, reduce headache-associated disability and improve the response to acute treatment when breakthrough migraines occur.[6] An adequate trial of a preventive medication is usually at least 2 months (as it may take 1–3 months for the full effect to be experienced), during which time the medication is slowly titrated to an effective or maximally tolerated dose.[6] During this time headache frequency should be monitored with a calendar or diary.[4,6]

When choosing preventive treatments, the patient's comorbidities and the risk of adverse effects should be taken into account. First-line preventive agents for migraines include tricyclic antidepressants (amitriptyline, nortriptyline), beta-blockers (propranolol, metoprolol, atenolol) and pizotifen.[6,8] Of the beta-blockers, propranolol has the most evidence for efficacy and is approved for use in migraine, as is metoprolol, whereas atenolol is not. Although data are lacking, other tricyclic antidepressants such as nortriptyline and dothiepin may be effective if side effects limit the use of amitriptyline.[8] Pizotifen may be effective but its use is often limited by side effects including drowsiness and weight gain.[8]

If the first-line agents are ineffective or associated with unacceptable side effects, second-line options include sodium valproate, topiramate or verapamil sustained release.[6,8] Sodium valproate should be avoided in women of childbearing potential.[6] According to the Australian Medicines Handbook 2014, topiramate is as effective as valproate and probably as effective as propranolol. Topiramate is associated with a risk of fetal malformations and can impair the effectiveness of hormonal contraceptives.[9] Sodium valproate and verapamil sustained release are not marketed for migraine. Other drugs, such as gabapentin, are also used but often have limited evidence or efficacy.

If headaches are well controlled for 3–6 months, slow tapering of the preventive drug may be feasible, particularly in patients with less frequent migraine attacks, fewer years of migraine and fewer comorbid conditions such as depression, anxiety and fibromyalgia.[10]

ANSWER 7

Lifestyle modifications can be very effective in reducing migraines and should be incorporated into Emma's daily routine. These include:

- hydration – at least 8 glasses of non-caffeinated drinks daily. Although caffeine is often beneficial in the acute treatment of migraines, daily consumption of more than 1 or 2 caffeinated beverages can result in the development of chronic daily headache
- sleep hygiene – recommending 8 hours of sleep at night. Bedtime and awakening should be at consistent times each evening and morning throughout the week, as lack of sleep and too much sleep can both trigger headaches
- exercise – incorporating 30–60 minutes of aerobic exercise at least 3 times per week
- meals – making sure Emma consumes three daily meals, including mid-morning and mid-afternoon snacks if hunger triggers headaches. Meals should be high in protein, vegetables and fibre, and low in fat and sugar. Highly processed foods or those with additives and preservatives are best avoided.

ANSWER 8

Women with migraine with aura should be advised to avoid combined oral contraceptive (COC) pills.[11,12] A 2009 meta-analysis found that the risk of stroke was doubled in persons with migraine with aura, and a 3-fold increase was observed in the women with migraine with aura. The risk was increased further in those aged >45 years, smokers and women who use COCs.[13]

COCs may be used in women with migraine without aura who need or want contraception.[11] Although the use of COCs in migraine prevention is not clearly supported by the medical literature, the use of a monophasic low-dose (35 µg ethinyl oestradiol or less) COC may be beneficial in reducing menstrually related migraines by keeping oestradiol levels steady. For those women who continue to have menstrually related migraines, a continuous-dose regimen (skipping placebo pills) may be considered.[14]

Women with migraine without aura who are prescribed COCs should be counselled to report new-onset aura symptoms, and cardiovascular risk factors should be monitored.[14] If migraines worsen after starting the COC or if the patient develops an aura, the COC should be discontinued.[14] Note, guidelines generally do not recommend the use of COCs for women >35 years of age who have migraine without aura.[15]

Progestin-only contraceptives (oral or depot forms) have not been associated with an increased risk of stroke. The etonogestrel implant, the levonorgestrel-releasing intrauterine device and copper-containing intrauterine devices may be safer contraceptive options than the COC in women with migraine.[16]

The World Health Organization (WHO) has also set out medical criteria for the continuation of COCs when a woman develops migraine during their use. If migraine without aura is present and the woman is >35 years old, she should preferably stop using COCs (WHO Category 3). If she develops migraine without aura at an older age, or migraines with aura at any age, she must stop them (WHO Category 4).[11] If migraines with aura start during the use of progestin-only contraceptives, they should generally be suspended unless other more appropriate methods of contraception are not available (WHO Category 3).[11,17]

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CASE 5

MICHAEL COMPLAINS OF A SEVERE HEADACHE

Michael is a fit 30-year-old who has come to see you today about a recent severe headache that had an instantaneous onset.

QUESTION 1

Does this presentation warrant investigation? What questions do you need to ask about this headache?

FURTHER INFORMATION

Michael says that this was the worst headache he has ever had and that it came on abruptly, as if 'someone had hit him across the back of his head with a block of wood'. There were no accompanying symptoms. He has not had headaches in the past, apart from occasional hangover headaches.

QUESTION 2

What is your differential diagnosis? What investigations, if any, are needed and how quickly?

FURTHER INFORMATION

Michael now reveals that he has had several thunderclap headaches over the past few weeks.

QUESTION 3

What further questions do you need to ask in addition to the above questions?

QUESTION 4

What are primary headaches? What primary headaches can be associated with thunderclap headaches?

CASE 5 ANSWERS

ANSWER 1

Headache is one of the most common neurological problems that presents to a GP or a neurologist. It is also one of the most common acute and chronic pain conditions. Tension-type headache and migraine are the second and third most prevalent medical disorders.[1] Migraine accounts for 30% of the global burden and more than 50% of the disability burden attributable to all neurological diseases worldwide. Overall, migraine is the fourth ranking cause among women and the seventh ranking cause of all disease-associated disability worldwide.[1] Thus headache and migraine are major problems worldwide. The *Atlas of Headache Disorders and Resources in the World 2011*[2] states that 'Headache disorders are ubiquitous, prevalent and disabling. Yet they are under-recognized, under-diagnosed and under-treated'.

Taking a detailed history is very important in a presentation such as this. It is important to establish:

- how quickly the headache came on
- how severe it was (was it 10/10?)
- what was Michael doing at the time
- how long did the headache last, including the severe component
- whether there was any residual pain
- whether there were any associated features.

A thunderclap headache is a severe headache, often the worst headache the patient has experienced, that comes on usually within a matter of seconds and which reaches its maximum within seconds to about a minute. A headache that reaches 7/10 or more in terms of pain within less than a minute is a thunderclap headache.[3] It is one of the red flags of headache.[3] The initial severe headache can persist for a variable period of time and there is often a less severe headache lasting for a variable amount of time thereafter. Some thunderclap headaches may be associated with loss of consciousness, nausea and vomiting, neck stiffness and other neurological symptoms. They may occur only once or be recurrent, depending on the cause.[4]

ANSWER 2

Irrespective of whether there are accompanying symptoms and even if the patient has a past history of headache, the first diagnosis to consider as the cause for a thunderclap headache is a subarachnoid haemorrhage (SAH). Seventy percent of patients with a SAH present with a headache, 50% of which are thunderclap headache.[4] Up to 25% of patients with a thunderclap headache have a subarachnoid haemorrhage, usually due to a ruptured aneurysm, but other serious conditions that should be considered are cervical artery dissection, intracerebral haemorrhage, cerebral vein thrombosis and the reversible cerebral vasoconstriction syndrome (RCVS).[4]

The patient needs urgent referral to an emergency department for an initial non-contrast computed tomography (CT) brain scan and lumbar puncture, if the CT shows no blood. A brain CT without contrast, performed in the first 12–24 hours, has been shown to be highly sensitive and specific for the diagnosis of subarachnoid haemorrhage.[5] The sensitivity of CT decreases in the ensuing days from 86% on day 2, to 58% after 5 days.[6] Other investigations, such as magnetic resonance imaging (MRI) of the brain, may also be needed.[4,5]

ANSWER 3

Additional questions that Michael could be asked include:

- Did the attacks occur randomly or were they associated with certain activities and what was the temporal pattern, if any?
- Has he been taking any new medications or illicit drugs recently?

Some thunderclap headaches occur repeatedly with coughing, sneezing or other Valsalva manoeuvre, or with sexual activity.[4] These headaches need to be thoroughly investigated for an underlying structural cause such as an Arnold-Chiari malformation, hydrocephalus, a colloid cyst of the third ventricle or intracranial hypotension.[7] In the RCVS, thunderclap headaches occur repeatedly over a short period of time (several weeks to a month) and are self-limiting.[7] They can occur with or without neurological symptoms and severe hypertension may be present.

In addition to the above investigations, imaging of the cerebral vasculature with magnetic resonance angiography (MRA) and CT or digital subtraction angiography of the cerebral arteries is mandatory. The characteristic radiological findings are of alternating segments of cerebral arterial constriction and dilatation (so called beading). This may need to be repeated over time as the changes may be present only intermittently despite recurrent attacks.[7] RCVS is usually benign, but ischaemic infarction and intracerebral or subarachnoid haemorrhage can occur and will be detected with CT and MRI. The RCVS can be triggered by sexual activity, exertion, emotion or by certain drugs such as sympathomimetic or illicit drugs that can cause vasoconstriction.[7] Women in the postpartum period are particularly at risk of the RCVS but can also have thunderclap headaches due to eclampsia or cerebral vein thrombosis.[8]

ANSWER 4

Primary headaches are those where, at present, there is no known underlying structural pathology.[7] Migraine and tension type headache are the most frequently occurring primary headaches. Primary cough headache, primary headache associated with sexual activity and primary thunderclap headaches can all present with thunderclap headaches. Primary cough headache is more common in older men and responds to indomethacin.[7] Sixty percent of patients with isolated sexual headaches have RCVS.[4] Thus a thunderclap headache due to a primary headache is a diagnosis of last resort and can only be made after extensive investigations of the patient to rule out an underlying cause.[9]

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CASE 6

DAVID HAS SPEECH DIFFICULTIES

David is 65 years of age and presents to your clinic at 1 pm with a history of speech difficulties, which he describes as 'talking gibberish', that lasted about 10 minutes when he was out shopping at 11 am. He has a history of hypertension treated with perindopril 2.5 mg daily and smokes 5 cigarettes a day. His body mass index (BMI) is 32 kg/m^2 and his blood pressure today is 145/85 mmHg.

QUESTION 1

What clinical features will help localise the pathology in the nervous system?

QUESTION 2

What is the differential diagnosis?

QUESTION 3

What investigations need to be performed and with what urgency?

FURTHER INFORMATION

You now suspect that David had a TIA.

QUESTION 4

What management should be initiated and with what urgency?

FURTHER INFORMATION

You find an irregular pulse on examination and ECG confirms atrial fibrillation.

QUESTION 5

What management should be initiated and with what urgency?

FURTHER INFORMATION

Left internal carotid artery stenosis of 70% and right internal carotid artery stenosis of 60% was found on Doppler ultrasound.

QUESTION 6

How is this best managed?

FURTHER INFORMATION

On more detailed examination you find that David still has difficulty with high-level naming and there is a loss of the right nasolabial fold.

QUESTION 7

Would the diagnosis still be TIA?

QUESTION 8

What management should occur?

CASE 6 ANSWERS

ANSWER 1

Speech disturbance can be due to dysarthria (poor articulation) or dysphasia (language disturbance – comprehension, fluency, naming). In general, patients can distinguish between slurring the correct words (dysarthria) and using the wrong words or 'talking gibberish' (dysphasia). Dysphasia localises to the left hemisphere in right-handed patients and most left-handed patients. Dysarthria can involve either hemisphere or brainstem (non-localising). The presence of associated symptoms, including facial droop, sensory loss, 'heaviness'/weakness', is very helpful to confirm the affected hemisphere. Diplopia or vertigo localise to the posterior circulation (brainstem/cerebellum).

ANSWER 2

The following conditions could be considered in the differential diagnoses for this presentation.

Transient ischemic attack (TIA) or stroke

TIA occurs when there is a temporary occlusion of a cerebral artery that resolves without causing brain infarction. It is the most important possible diagnosis. The definition of TIA has evolved with the removal of the old 'resolves within 24-hour' criterion. This caused confusion when a patient presented with symptoms of a few hours' duration, leading to a frequent question 'how do you know it's not a TIA as it could still resolve by 24 hours?'. The average duration of a TIA is about 10 minutes.[1] If symptoms are still present when the patient is seen then the diagnosis is stroke not TIA and needs to be treated differently. The new definition is based on the 'absence of infarction on brain imaging'.[1] However, this depends on the type of imaging modality used, either computerised tomography (CT) or the much more sensitive diffusion magnetic resonance imaging (MRI). Up to 40% of fully resolved events that are regarded as TIA on the basis of clinical presentation have small diffusion lesions on MRI (technically a stroke) and have a much higher risk of recurrent stroke.[1]

Migraine

Migraine can cause transient dysphasia, dysarthria, sensory symptoms and weakness. The headache can follow or be mild or absent in some cases. It is unusual to have a first episode of migraine at the age of 65 years so the patient's history is important. A history of symptom migration (eg paraesthesia spreading from face to hand over about 15 minutes) is suggestive of migraine as cortical spreading depression (a wave of electrophysiological hyperactivity followed by a wave of inhibition) propagates across the cortex at about 2–5 mm/min.

Seizure

Seizure usually causes positive phenomena (paraesthesia or jerking) rather than loss of function.

Metabolic disturbance

A metabolic disturbance, such as hypoglycaemia, often causes generalised symptoms and can lead to a focal neurological deficit in some cases.

Intercurrent illness

Intercurrent illness with worsening of an old deficit can occur (eg facial droop re-emerging in the context of a urinary tract infection).

Functional illness

Functional illness is surprisingly common in neurological practice. There may be features on examination (eg Hoover's sign: first test hip extension in the 'weak' leg; leave your hand under the ankle while testing the contralateral hip flexion. Normal co-activation of the contralateral 'weak' hip extensor muscles indicates functional weakness) but this can be difficult to identify even for neurologists.

ANSWER 3

The following investigations need to be performed, preferably on the same day.[2,3] The ability to arrange these investigations will depend on the location and the resources available. The local hospital may have a rapid access TIA clinic or a rapid assessment pathway through the emergency department.

- CT brain scan provides a relatively low yield but is important to exclude small bleeds, subdural haematoma, tumour, etc. MRI diffusion imaging has greater sensitivity for differentiating stroke (abnormal) from TIA (normal) but may not be readily available.
- Carotid imaging using carotid Doppler ultrasound or CT angiogram can reveal symptomatic carotid artery stenosis. When performed early, carotid endarterectomy has been found to significantly reduce subsequent stroke risk.[2]
- An electrocardiogram should be performed routinely to look primarily for atrial fibrillation (also old Q-waves may indicate an akinetic ventricular segment that occasionally can lead to mural thrombus).[2] Note, atrial fibrillation may be paroxysmal (and therefore not present at the time the patient is seen). Paroxysmal atrial fibrillation has the same stroke risk as permanent atrial fibrillation.[4] Holter monitor data may be useful in detecting some cases of atrial fibrillation but may still miss some paroxysmal atrial fibrillation so it is important to continue to monitor the patient clinically at every opportunity.

ANSWER 4

The current approach to stroke secondary prevention is based on the concept of absolute cardiovascular risk. By definition, a patient with a TIA is at high risk and should receive rapid treatment of all vascular risk factors, regardless of the individual risk factor level.[2]

The following should be considered:

- Antiplatelet treatment – aspirin is generally used (eg a loading dose of 300 mg followed by 100 mg daily).[5] A combination of aspirin plus dipyridamole is also available and slightly more effective but is generally commenced once daily in combination with 100 mg aspirin and then increased to twice daily after 1–2 weeks (with cessation of separate aspirin) to reduce the incidence of headache. Clopidogrel, which is slightly more effective than aspirin and equally effective as aspirin plus dipyridamole,[6] is an option for those intolerant of aspirin or who have had their TIA while already taking aspirin. Combined aspirin plus clopidogrel is not recommended long term for stroke prevention but a recent trial (CHANCE) has suggested some benefit of short-term combination therapy for about 1 month in high-risk patients.[7]
- Statins – on the basis of a randomised trial using 80 mg atorvastatin,[8] statins reduce recurrent stroke regardless of baseline cholesterol.
- Antihypertensive agents – hypertension is a particularly potent risk factor for stroke with no 'threshold' for benefit (ie the lower the blood pressure, the lower the stroke risk).[2] Angiotensin converting enzyme inhibitors, angiotensin 2 receptor antagonists and calcium antagonists are all reasonable first-line choices.[2] Given that David's blood pressure is 145/85 mmHg, increasing his perindopril dose to target his blood pressure to <135/80 mmHg would be a reasonable start, although, as mentioned, the target level is arbitrary since lower is better provided postural hypotension is not problematic.

With regards to urgency, management of risk factors should commence on the same day (after CT scan). A study in the UK (EXPRESS) showed that stroke risk could be reduced by 80% simply by starting these medications immediately rather than delaying by a couple of weeks.[9]

Additional factors that should be considered for risk factor reduction are listed below. Management should be individualised and delivered using behavioural techniques, such as motivational interviewing techniques:[2]

- smoking cessation – offer counselling and/or behavioural therapy and consider use of pharmacological therapies (eg nicotine replacement therapy, bupropion)
- screen for diabetes with fasting glucose (oral glucose tolerance test (OGTT) could also be performed and manage according to national diabetes guidelines
- increasing exercise – for example, recommend a 30 minute brisk walk every day
- diet – provide information/education on following a low-fat diet, high in fruit and vegetables, and advise to avoid fried and processed foods, not to add salt to food (following a Mediterranean diet and eating tree nuts may reduce risk[10]).

ANSWER 5

Atrial fibrillation is a major risk factor for stroke and anticoagulation is underused. Unless there is a very strong reason not to use anticoagulants (eg active gastrointestinal bleeding) David should start anticoagulation treatment immediately. He could be started on warfarin, using an appropriate protocol to initiate dosing, to a target INR of 2–3.[4] If the creatinine clearance is >50 ml/min, a non-vitamin K oral anticoagulant (NOAC; eg apixaban 5 mg BD, dabigatran 150 mg BD or rivaroxaban 20 mg daily) could be used.[4,11] If the creatinine clearance (CrCl) is 30–50 ml/min, dose modification is generally required and all patients on NOACs should have CrCl monitored periodically.[4,11,12]

Note, onset of anticoagulation is 2–3 hours for NOACs versus several days for warfarin, and there are no accepted antidotes for the NOACs at this time.[11]

ANSWER 6

It is important to localise the hemisphere affected, as an asymptomatic stenosis does not warrant surgical intervention. As David has dysphasia, the left internal carotid artery stenosis is symptomatic.

Endarterectomy reduces the rate of recurrent stroke for symptomatic stenosis by >70% with more marginal benefit for stenosis 50–70%. If there is uncertainty regarding the severity of stenosis on Doppler ultrasound, a CT angiogram could be performed. Surgery should be performed as soon as possible after a TIA and certainly within 2 weeks, as the highest risk of recurrent stroke is early. The asymptomatic right carotid stenosis will benefit from intensive medical management (statins, antiplatelet and antihypertensive agents) and there is no need for surgical intervention.[2]

ANSWER 7

The absence of residual symptoms at the time of examination indicates the diagnosis is probably stroke (in the left middle cerebral artery territory) not TIA.

ANSWER 8

You should call an ambulance. The patient will be taken to the nearest stroke-equipped hospital. As the patient is within 4.5 hours from the onset of symptoms, thrombolysis may be an option depending on severity, brain imaging findings and other clinical criteria. Thrombolysis with intravenous administration of tissue plasminogen activator within 4.5 hours of the 'last known well time' has level 1 evidence (meta-analysis of randomised trials) showing significant reductions in disability and a neutral effect on mortality.[13] Regardless of time window, all patients with stroke should be managed in a stroke unit, which reduces the risk of disability and death for all age groups, stroke subtypes and severities.[2]

KEY POINTS

- TIA is a medical emergency. It is characterised by a sudden onset of focal neurology with complete resolution and has an average duration of about 10 minutes.
- If symptoms/signs are still present when the patient is seen then the diagnosis is stroke not TIA; call 000 and send the patient to a stroke-equipped emergency department.
- The aim of TIA management is to prevent a subsequent stroke (the highest risk is in the subsequent 48 hours).
- A CT brain scan excludes diagnoses that mimic TIA (tumour, small bleed); carotid imaging (Doppler US or CT angiogram) can be used to investigate for symptomatic carotid stenosis.
- Detecting atrial fibrillation is critical but difficult as it can be paroxysmal so an electrocardiogram should be performed and use of a Holter monitor considered, but the patient should be monitored clinically at every opportunity.
- NOACs (subject to CrCl) or warfarin should be commenced immediately if a TIA occurs as a consequence of atrial fibrillation
- In the absence of atrial fibrillation, most patients should be immediately commenced on antiplatelet treatment (aspirin, aspirin plus dipyridamole or, if PBS criteria are met, clopidogrel), statin and antihypertensive agents.
- Carotid endarterectomy reduces the risk of recurrent stroke for symptomatic carotid stenosis by >70% (with a smaller benefit if 50–70% stenosis).
- Asymptomatic carotid stenosis should be managed with intensive medical therapy if symptoms/signs are still present (the diagnosis is stroke) the time window for standard thrombolysis is 4.5 hours from the time the patient was last seen well.
- Even if >4.5 hours have elapsed, every stroke patient should be assessed and managed in a stroke unit.

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RESOURCES FOR DOCTORS

- National Stroke Foundation Clinical Guidelines for Stroke Management 2010, http://strokefoundation.com.au/site/media/clinical_guidelines_stroke_managment_2010_interactive.pdf

CASE 7

ROB HAS A DISABLING HEADACHE

Rob is a labourer aged 30 years. He comes to your practice with his wife to discuss a 5-year history of headaches. These headaches are disabling, causing 1–2 months of sick leave each year when they occur. The pain is mainly behind the left eye and temple, and radiates to the vertex. He rarely experiences pain on the right side. The pain comes on rapidly (within minutes) and Rob describes it as '20/10' in intensity. It is so excruciating that it often causes him to double over and writhe around. It occurs, predictably, soon after falling asleep. His wife states that he may have multiple episodes of headache (sometimes up to 4) occurring daily. Each episode seems to last 0.5–2 hours. After 1–2 months of this pattern, the headaches subside and he feels well again 'for months until another attack'. Magnetic resonance imaging (MRI) of the brain was performed recently and was normal. His neurological examination is normal.

QUESTION 1

What are differential diagnoses for Rob's headache?

QUESTION 2

Of the short-lasting head pain syndromes, what headache types are consistent with the attack frequency described by Rob?

FURTHER INFORMATION

Rob experiences an attack in the examination room. During the attack, he has sudden onset of severe pain behind the left eye and grabs his left side with his hands. You notice that he is shielding his left eye and is pacing restlessly around the room. The pain is so severe that he indicates that he cannot converse or answer any further questions. He vomits. When questioned, his wife indicates that this is a typical event for Rob. The attack subsides after 30 minutes. On examination you notice that Rob's left eye is red and he has had tearing. He also has obvious nasal congestion.

QUESTION 3

What further information do you need to clarify the diagnosis? Specifically, what direct questions do you need to ask?

FURTHER INFORMATION

Rob's wife states that they have seen five different GPs so far over the years and the most common diagnoses have been sinusitis or migraine headache. He has been previously treated with muscle relaxants, gabapentin, beta-blockers and long- and short-acting nonsteroidal anti-inflammatory drugs (NSAIDs). Recently, a doctor started him on indomethacin, which 'did absolutely nothing' to relieve the headaches. He has now become reliant on opioids, often requiring intramuscular morphine for acute attacks.

QUESTION 4

How does the insensitivity to indomethacin help in further defining the diagnosis?

QUESTION 5

What is your approach to acute treatment now, during the attack?

QUESTION 6

What preventive treatment options are most likely to be effective in this patient?

FURTHER INFORMATION

The severity of his condition has led to Rob often losing his job as a laborer around the times of the cluster headache attacks every year. He has tried to decrease his opioid use but has had symptoms of palpitations, nausea and sweating when attempting to cut down. He also reports fatigue, frequent spells of crying, night sweats and generally feeling 'helpless'. He tells you that he 'cannot take this anymore' but denies suicidality.

QUESTION 7

What is the most appropriate course of action at this time?

CASE 7 ANSWERS

ANSWER 1

Possibilities for unilateral headache include migraine, hemicrania continua, and trigeminal autonomic cephalalgias (TAC). The latter include cluster headache, paroxysmal hemicranias and short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT). In addition to TAC, other short-lasting headaches, primary stabbing headache (ie icepick headache) and trigeminal neuralgia, should also be considered (*Table 1*).[1]

Table 1. Short-lasting headaches[1]

Trigeminal autonomic cephalalgias (TAC)	Cluster headache Paroxysmal hemicranias SUNCT
Other short-lasting headaches	Hemicrania continua Primary stabbing headache Trigeminal neuralgia Primary cough headache Primary exertional headache Primary sex headache

Although migraine is unilateral in approximately 60% of patients and can have a rapid/sudden onset, it is normally of a longer duration (by definition 4–72 hours). Also, this patient is not in a typical age group where new-onset migraine would be common.[1,2] Therefore, this is unlikely to be a primary migraine headache.

Hemicrania continua is a constant, continuous, strictly one-sided headache punctuated by severe pain exacerbations. It affects the temple or periorbital region and usually affects the same side of the head. It is routinely misdiagnosed because patients only report the exacerbations that occur, which often have varying frequency several times a week. These exacerbations can last from minutes to several days with an intensity ranging from mild to severe.[3] The underlying pain is constantly present, 24 hours a day, every day of the week.[1,4] However, the pain of hemicrania continua is reported to be not as excruciatingly severe as the pain of cluster headache.[3] Some patients have associated symptoms common to migraine, such as nausea, vomiting, photophobia and phonophobia. They may also have autonomic symptoms and similarities in clinical presentation, treatment and pathophysiology.[4] Thus, hemicranias continua is commonly regarded as the fourth TAC-type headache, but it differs from the TACs in having continuous underlying pain. For this reason they are not included as a TAC in the International Classification for Headache Disorders.[1]

Trigeminal neuralgia consists of severe, sharp, electric or knifelike repetitive attacks lasting seconds, usually in a V2 or V3 distribution, sometimes radiating into the teeth. It rarely occurs in a V1 distribution and thus is not consistent with this Rob's pain distribution.[5]

Primary stabbing headache is predominantly reported over a V1 distribution (orbit, temple, parietal areas) and is stabbing in character as the name suggests, so it should be considered in Rob's case. The

main feature that helps to distinguish primary stabbing headache from the TAC headaches is the presence of any associated autonomic features.[5,6]

ANSWER 2

Attack duration and frequency are key components in differentiating headache type and thus management (*Table 2*).[7]

Cluster headaches are typically 15–180 minutes in duration (average 45–90 minutes) and are more variable in their frequency (range 3–8 per day).[1] They also have a consistent circadian rhythm, with attacks occurring at a similar time each day and they commonly occur soon after falling asleep. There also tends to be a seasonal periodicity with cluster headaches.[8] Typically, the headache occurs daily for 8–10 weeks per year (*Table 3*).[9] Patients are generally asymptomatic between attacks. Rob's history is most consistent with this type of headache.

Attack duration in paroxysmal hemicranias last for 2–45 minutes and patients report 15 or more attacks per day (usual frequency being 1–40).[1,8] Therefore, the shorter duration and the greater frequency of paroxysmal hemicranias are inconsistent with Rob's headache syndrome.

In SUNCT, pain episodes last for 15–120 seconds and the frequency of attacks is about 3–100 episodes per day,[1,8] which clearly does not fit Rob's case.

Cluster headache can be further differentiated into episodic and chronic. Episodic cluster headache is defined by recurrent episodes of headache daily or every other day and lasting more than 1 week, separated by remissions lasting more than 1 month. This can occur once or twice a year, but can continue in this pattern for several years. Chronic cluster headache occurs in only 20% of those with diagnosed cluster headache and is defined as no remission within 1 year or a remission period that lasts <1 month.[5,10]

ANSWER 3

It would be important to clarify the presence or absence of associated features. A significant differentiating feature between the TAC group and other short-lasting head pain syndromes is the presence of associated features, particularly autonomic features in the former (*Table 4*).[7] Cranial autonomic symptoms are cardinal features of the TACs, especially cluster headache. This includes lacrimation, nasal congestion, conjunctival injection and rhinorrhoea, miosis, eyelid swelling or ptosis.[8] Because of this, misdiagnosis of sinus headache is common in these patients and in a clinical study, nearly 25% of these patients were being treated by an ear, nose and throat specialist for sinusitis.[11] The underlying pathology is thought to be cranial parasympathetic activation related to pain (rather than local inflammation).[12]

Other associated features can also include migraine-like features, such as nausea or vomiting, photophobia and phonophobia. The latter two symptoms are usually on the same side as the headache in TAC.[5,8] This lateralisation of symptoms and signs is an important feature of TAC attacks, particularly for cluster headache, and is a key differentiating feature of TAC headaches.[8] By contrast, in migraine the features are usually bilateral, less prominent and variable in presentation.

Other important information to ascertain about Rob's headaches is whether there are specific triggers for the headaches. Alcohol is often a strong precipitating factor of the pain in cluster headache,[7] differentiating it from SUNCT (which does not have an alcohol trigger)[7] and paroxysmal headache, where only one-fifth of patients have this trigger.[7] Cutaneous triggers, such as touching the skin, talking and chewing, do not trigger cluster headache, whereas they are significant triggers in SUNCT.[13,14]

The history should include detailed knowledge of the patient's past history, including headaches, and his family history. This information may affect treatment choices and further management.

Table 2. Differential diagnosis of short lasting headaches[7]

Feature	Cluster headache	Paroxysmal hemicrania	SUNCT*	Idiopathic stabbing headache	Trigeminal neuralgia	Hypnic headache
Sex (male:female)	5:1	1:2	2:1	F>M	F>M	5:3
Pain						
Type	Boring	Boring	Stabbing	Stabbing	Stabbing	Throbbing
Severity	Very severe	Very severe	Severe	Severe	Very severe	Moderate
Location	Orbital	Orbital	Orbital	Any	V2/V3>V1	Generalised
Duration	15–180 minutes	2–45 minutes	15–120 seconds	<30 seconds	<1 seconds	15–30 minutes
Frequency	1–8/day	1–40/day	1/day–30/hour	Any	Any	1–3/night
Autonomic	+	+	+	–	–†	–
Trigger	Alcohol, nitrates	Mechanical	Cutaneous	None	Cutaneous	Sleep
Indomethacin	?	+	–	+	–	+

*Short lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing

†Cranial autonomic activation may be seen in first division trigeminal neuralgia

Table 3. Diagnostic criteria for cluster headache[9]

3.1 Diagnostic criteria
A, At least five attacks fulfiling B–D
B. Severe or very severe unilateral orbital, suproorbital and/or temporal pain lasting 15–180 minutes if untreated
C. Headache is accompanied by at least one of the following
1. Ipsilateral conjunctival injection and/or lacrimation
2. Ipsilateral nasal congestion and/or rhinorrhoea
3. Forehead and facial sweating
4. Ipsilateral eyelid oedema
5 Ipsilateral forehead and facial sweating
6. Ipsilateral miosis and/or ptosis
7. A sense of restlessness or agitation
D. Attacks have a frequency from one every other day to eight per day
E. Not attributed to another disorder
3.1.1 Episodic cluster headache
Description: Occurs in periods lasting seven days to one year separated by pain free periods lasting one month or more
Diagnostic criteria:
a. All fulfilling criteria A–E of 3.1
b. At least two cluster periods lasting from 7 to 365 days and separated by pain free remissions of one month or more
3.1.2 Chronic cluster headache
Description: Attacks occur for more than one year without remission or with remissions lasting less than one month
Diagnostic criteria:
a. All alphabetical headings of 3.1
b. Attacks recur for more than one year without remission periods or with remission periods lasting less than one month

Table 4. Primary short-lasting headache[7]

Prominent autonomic features	Sparse or no autonomic features
Cluster headache	Trigeminal neuralgia
Paroxysmal hemicrania	*Idiopathic stabbing headache
SUNCT syndrome	Cough headache
	Benign exertional headache
	Headache associated with sexual activity
	Hypnic headache

*Likely to be renamed primary stabbling headache when the International Headache Society Classification[1] is revised.[9]

ANSWER 4

Most of the headache types mentioned thus far respond to indomethacin, except for cluster headache. Paroxysmal hemicranias and hemicranias continua, in particular, are responsive to indomethacin. Indeed, a response to indomethacin is required in these cases for diagnostic certainty.[15] Rob's lack of response to indomethacin is helpful in eliminating these forms of headache.[16]

ANSWER 5

Cluster headache is often considered one of the most painful of all headache types. During attacks, patients are unable to remain still and they move constantly, as was seen in Rob's case earlier. By contrast, patients with migraine prefer to remain still.

Acute treatment needs to reach the full therapeutic response rapidly and have a rapid onset. Many patients respond well to oxygen inhalation, which should be administered as at 10 L/min (range 7–12 L/min) for 15 minutes.[17,18] This is best given via a mask to enable the high-flow rate.[17–20] According to the *Australian Medicines Handbook*, inhaled oxygen relieves symptoms in 70–80% of people within 15 minutes.[18] Oxygen therapy should be stopped after 15 minutes if there is no improvement in status, as there is a risk of oxygen toxicity with longer treatment.[20] Of note, oxygen therapy does not alleviate the other forms of TAC headache.

Other acute treatment options include subcutaneous sumatriptan at a dose of 6 mg.[18,21] Sumatriptan has a rapid onset of action and is the only drug that has approval from the US Food and Drug Administration (FDA) for treatment for cluster headache. Nasal sprays of sumatriptan (20 mg) or zolmitriptan (5 mg) are also effective in acute cluster headache.[22] Note, zolmitriptan nasal spray is not currently registered in Australia. The *Australian Medicines Handbook* indicates that although there is less evidence for the efficacy of sumatriptan nasal spray, compared with subcutaneous sumatriptan, it may improve or eradicate a cluster headache within 30 minutes of use.[18] The bitter taste of the nasal spray may render this treatment unacceptable for some.[21] Conversely, treatment with oral sumatriptan (eg 100 mg tds) is ineffective and can be associated with medication-overuse headache.[23]

Table 5 outlines current treatment recommendations for cluster headache supported by Australian guidelines. The National Institute for Clinical Excellence 2012 headache guidelines do not recommend use of paracetamol, NSAIDs, opioids, ergotamine or oral triptans for the acute treatment of cluster headache.[17]

Table 5. Current Australian treatment recommendations for cluster headache

Type of treatment	Treatment
Acute	Oxygen inhalation for up to 15 minutes[18,20]
	Sumatriptan, subcutaneous (6 mg)[18,20]
	Sumatriptan nasal spray (20 mg) – authority streamlined[18,20,24]
	Lignocaine 4% solution instilled into the nose on the side of the pain[20]
	A short course of high-dose corticosteroids[18]
Prevention	Initially, verapamil 240 mg daily (usual range 240–960 mg daily in 1–4 doses depending on the formulation)[25]
	OR
	Verapamil sustained-release 160 or 180 mg orally, once daily, up to 360 mg daily[20]
	OR
	Lithium 250 mg orally, twice daily, titrate according to clinical response and tolerance, guided by serum concentration levels[20]

ANSWER 6

Verapamil is considered first-line for preventive treatment[18] and although it is not marketed for this indication in Australia, prophylaxis of cluster headache is an accepted indication.[25] For cluster headache, a patient should be started on 240 mg daily (usual dose range 240–960 mg daily in 1–4 doses depending on the particular formulation used).[25] Alternatively, the Therapeutic Guidelines recommend use of verapamil sustained-release at doses of 160 or 180 mg orally, once daily, with total daily doses up to 360 mg daily.[20] As patients often require high doses, side-effects must be adequately screened and monitored. Common side-effects (frequency >1%) include nausea, vasodilatory effects such as headache and flushing and peripheral oedema.[25] Infrequent side effects (0.1–1% incidence) include gingival hyperplasia and constipation.[25,26] An ECG must be done before initiation of treatment and monitoring for cardiac arrhythmias, particularly heart block, must be undertaken.[20] Effects of slowing conduction on the atrioventricular node can take up to 10 days to appear and therefore 2-weekly intervals are recommended between dose changes. ECGs should be done before each dose escalation and routinely every 6 months after the drug has been initiated.[27]

Other preventive options include lithium (which is comparable in efficacy to verapamil),[26] topiramate and gabapentin. However, there have been no controlled studies for the latter two medications, and recommendations have been based on case reports only.[8] For patients with shorter episodes of cluster headache, limited courses of oral corticosteroids can also be useful.[8,18,23]

Please refer to *Table 5* for a summary of preventive treatment options.

ANSWER 7

Although a correct diagnosis of cluster headache has been made and a treatment regimen has been planned, the chronicity of his symptoms has affected his life to the point that medications alone are unlikely to help his current mental state. His opioid use could also be contributing to a medication-induced depression and, ultimately, also contributing to his headache. The most efficacious course of action at this time would be referral for a period of inpatient treatment for help to wean him off the opioids and to allow him to receive ongoing psychological support during this time.

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Head pain

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FOR A FULL LIST OF ABBREVIATIONS AND ACRONYMS USED IN THESE QUESTIONS PLEASE GO TO PAGE 3.
FOR EACH QUESTION BELOW SELECT ONE OPTION ONLY.

QUESTION 1

Selena is a lawyer aged 26 years and was diagnosed with migraine without aura last year. She recently moved in with a new partner and has presented today requesting the pill. Which of the following statements regarding the use of oral contraceptives by Selena, given her migraine without aura diagnosis, is CORRECT?

A. Combined oral contraceptive pills (COCs) are contraindicated.
B. COCs are contraindicated in all women younger than 35 years of age.
C. If she smoked, she could still use COCs as COCs are not contraindicated in smokers.
D. If Selena were diagnosed with migraine with aura, COCs could be prescribed as they are not contraindicated.
E. On the basis of the information given, COCs are not contraindicated for Selena.

QUESTION 2

With regards to secondary prevention of stroke and current guideline recommendations, which of the following options is the most CORRECT?

A. Only patients deemed to be at high risk require management of their risk factors.
B. All patients who have had a stroke or TIA require management of all of their risk factors, irrespective of their risk factor level.
C. Antiplatelet agents, statins and antihypertensive agents use should be considered as well as lifestyle changes such as optimising diet, increasing physical activity and smoking cessation.
D. Answers A and C are correct.
E. Answers B and C are correct.

QUESTION 3

With regard to thunderclap headaches, which of the following statements is the most CORRECT?

A. An urgent referral to an emergency department is generally required and CT imaging needs to be performed; additional investigations may also be required.
B. Suspected recent thunderclap headache is not a medical emergency.
C. Subarachnoid haemorrhage, acute subdural haemorrhage and dissection of cervical arteries are not differential diagnoses for thunderclap headache presentations.
D. Up to 2.5% of patients with a thunderclap headache have a subarachnoid haemorrhage.
E. Thunderclap headaches do not present as primary headaches.

QUESTION 4

Which of the following statements is CORRECT with regard to the incidence and management of carotid artery dissection?

A. Carotid artery dissection is a common cause of stroke in older people.
B. The priority in carotid artery dissection is stroke prevention.
C. Randomised controlled trials report similar efficacy for antiplatelet and anticoagulation therapy for stroke prevention in carotid artery dissection.
D. Randomised controlled trials report superior efficacy for antiplatelet therapy over anticoagulation therapy for stroke prevention in carotid artery dissection.
E. Randomised controlled trials report superior efficacy for anticoagulation therapy over antiplatelet therapy for stroke prevention in carotid artery dissection.

QUESTION 5

Alice, aged 33 years, is a part-time childcare worker with two children of her own. She comes to see you wanting to discuss her frequent headaches. She has had regular headaches since her teenage years but they have increased in frequency in the past few years. After taking a detailed history, you diagnose chronic migraine without aura. Which of the following statements is the most CORRECT?

A. NICE guidelines recommend referral of people with migraine for neuroimaging.

B. Anxiety and depression are rare comorbidities with chronic migraine.

C. Chronic migraine is defined as headache occurring on 15 or more days per month for at least 3 months, which has the features of migraine headache on at least 8 days per month.

D. Migraine risk factors are all modifiable.

E. Onabotulinum toxin A (or botulinum toxin) is not marketed in Australia for migraine prevention.

QUESTION 6

Headaches may be due to primary or secondary causes. Which of the following statements is the most CORRECT with regard to causes of headache?

A. Thunderclap headaches should raise suspicion of underlying vascular cause(s) and patients with these presentations should be evaluated urgently.

B. It is unusual for primary headaches to present for the first time in people aged over 50 years.

C. Postural headaches may suggest low intracranial pressure.

D. Persistent focal signs presenting with headache suggest a secondary cause for the headache.

E. All of the above are correct.

QUESTION 7

With regard to the pharmacological management of migraine headache which of the following statements is the most CORRECT?

A. Opioids, including codeine preparations with paracetamol or aspirin, are recommended for the acute treatment of migraine.

B. The Australian Therapeutic Guidelines recommend medications such as amitriptyline, pizotifen, propranolol, sodium valproate, topiramate and verapamil as migraine prophylactic options.

C. Migraine prophylactic medications recommended by the Australian Therapeutic Guidelines, such as candesartan and gabapentin, are all licensed and listed on the Pharmaceutical Benefits Scheme for the indication of migraine.

D. Propranolol, sodium valproate and topiramate are all safe for use in women of childbearing potential.

E. Triptans should be used to treat all types of migraines.

QUESTION 8

Amanda, a speech therapist aged 27 years, has been diagnosed with chronic migraine with aura. With regard to management options, which of the following statements is the most CORRECT?

A. A headache diary is a useful way of documenting Amanda's response to therapy.

B. Development of medication overuse headaches is a risk if analgesics are taken on more than 15 days per month.

C. The goals of migraine prophylaxis are to reduce attack severity, frequency and duration, to improve responsiveness to acute treatments and to reduce disability.

D. When starting migraine prophylaxis, the patient should be given realistic goals, education about correct dosage and information about potential medication side effects.

E. All of the above are correct.

QUESTION 9

Which one of the following statements regarding cluster headaches is CORRECT?

A. Oxygen inhalation administered as 100% for 30 minutes is recommended for management of an acute attack of cluster headache.

B. Subcutaneous sumatriptan is recommended for prevention of cluster headaches.

C. Sumatriptan nasal spray is recommended for prevention of cluster headaches.

D. Verapamil is considered first-line for preventive treatment of cluster headache and although not marketed for this indication, prophylaxis of cluster headache is an accepted indication.

E. Lithium is useful for management of an acute attack of cluster headache.

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