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SCENARIO 17. MEDIAN NERVE PALSY

Identifying clinical signs

The median nerve serves the intrinsic small muscles of the hand not served by the ulnar nerve. The median nerve also serves muscles of the volar surface of the forearm, which includes the main wrist flexors.

The most common median nerve palsy appearing in the PACES examination is a carpal tunnel syndrome. Indeed this is the most common peripheral mononeuropathy. However, there can be more proximal lesions of the median nerve that offer more objective clinical signs.

Distal median nerve lesions (carpal tunnel syndrome)

**Inspection:** the hand shows thenar wasting (a late sign) with hypothenar and first dorsal interosseous sparing. The thumb is externally rotated and adducted into the plain of the palm (Figure 3.18) due to weakness of opponens pollicis and abductor pollicis, and some refer to this as ‘ape-hand’ deformity (but it is best to describe the position of the thumb). The index (first) and middle (second) fingers may also be held in extension due to weakness of flexor digitorum profundus I and II and flexor digitorum superficialis, and unopposed action of the radial nerve-innervated finger extensors. Some call this the ‘papal sign’. Look for surgical or traumatic scars at the wrist, palmar crease, forearm or elbow.

**Motor function:** there is weak thumb abduction (abductor pollicis brevis), tested for by asking the patient to place the hand palm up and point the thumb up to the sky perpendicular to the plane of the palm and test against resistance of your thumb. There is weakness of thumb opposition, tested for by asking the patient to oppose the thumb with the little finger and stop you pulling them apart. There may be normal flexion at the thumb MCP joint due to intact ulnar nerve innervation of flexor pollicis brevis. There is normal flexion of the thumb at the IP joint due to intact innervation of flexor pollicis longus in the forearm. Arm pronation and wrist flexion are also normal if the median nerve lesion is distal.

**Sensory loss:** there is sensory loss in the palmar aspect of the thumb and lateral two and a half fingers but normal sensation over the thenar eminence (supplied by the palmar cutaneous branch of the median nerve which does not pass through the carpal tunnel).

**Additional tests:** you can ask to perform Tinel’s test – tapping over the median nerve at the wrist may cause paraesthesia in the distal median distribution. Phalen’s test involves flexing the wrist to 90° for at least 60 seconds, which may cause paraesthesia in the distal median distribution. The median nerve compression test involves pressing on the palmar aspect of the wrist for up to 60 seconds, which may cause paraesthesia in the distal median distribution.

**Additional signs**

Examine for signs of other diseases associated with carpal tunnel syndrome such as:
• Hypothyroidism: goitre, slowly relaxing reflexes, pretibial myxoedema
• Acromegaly: supraorbital ridge, prognathism, interdental separation, bitemporal hemianopia, large doughy hands, hypertension
• Rheumatoid arthritis: deforming arthropathy, elbow nodules, steroid skin changes, episcleritis/scleritis, interstitial lung disease
• Diabetes mellitus: finger-prick testing, peripheral neuropathy, retinopathy
• Gout: deforming arthropathy and gouty tophi on hands, ears and feet.

Proximal median nerve lesions

**Pronator syndrome:** compression of the median nerve can occur as it passes between the two heads of the pronator teres, high in the volar aspect of the forearm. It can present with **purely sensory symptoms** of pain over the volar surface of the forearm at rest or with forearm pronation. There will be sensory loss within the median distribution including the **thenar eminence** (unlike carpal tunnel syndrome).

**Anterior interosseous nerve palsy**

This nerve has branches to flexor digitorum profundus I and II, flexor pollicis longus and pronator quadratus. It is typically affected by a midshaft fracture of the radius, excessive exercise or penetrating injuries of the forearm. There is **weakness of the thumb and index finger flexion**, best shown with the ‘okay’ sign, ie flattened due to failure of distal flexion (see Figure 3.17). The thenar eminence muscles are spared. There is **no sensory loss**.

**Differential diagnosis**

- Elbow lesions:
  - Fracture: supracondylar fractures are most common
  - Dislocation
  - Compression: ligament of Struthers
- Forearm lesions:
  - Fracture: midshaft radial fracture causing anterior osseous nerve palsy
  - Injury: penetrating injuries of the forearm
Compression: pronator teres syndrome

Wrist lesions:
- Fracture/trauma
- Carpal tunnel syndrome (CTS)

Clinical judgement and maintaining patient welfare

CTS is caused by compression of the median nerve within the carpal tunnel, which is bound by the carpal bones below, flexor retinaculum above, radially by the scaphoid and trapezium, and medially by the pisiform and hamate.

It is most commonly idiopathic, but associated conditions include: pregnancy, menopause, hypothyroidism, diabetes, acromegaly, rheumatoid arthritis, gout, renal failure, multiple myeloma and amyloidosis.

Investigations

Electrophysiology: nerve conduction studies and EMG.

Imaging: seldom needed, but MRI might be considered.

Management

- Treat any underlying associated conditions
- Physiotherapy and splint the wrists with a degree of dorsiflexion
- Steroid injections into the carpal tunnel area
- Surgical decompression of the carpal tunnel.
Identifying clinical signs

The ulnar nerve serves most of the intrinsic small muscles of the hand. However, the ulnar nerve also supplies two important extrinsic muscles of the hand: flexor carpi ulnaris and flexor digitorum profundus 4 and 5; it is important to remember this when considering a claw hand deformity. Clues pointing towards an ulnar nerve lesion include unilateral signs, clawing of the hand and wasting of the hypothenar eminence with sparing of the thenar eminence.

Distal ulnar nerve lesion (distal to the elbow)

**Inspection:** with the hands held prone there is **dorsal guttering** and marked **wasting of the first interosseous** between the thumb and index finger. The hand shows a claw deformity (you might want to say a **clawed appearance**) with hyperextension at the fourth and fifth MCP joints and flexion at the fourth and fifthPIP and DIP joints, due to paralysis of the medial lumbricals. There is slight ulnar deviation of the fifth finger (known as **Wartenburg’s sign**) from unopposed action of extensor digit minimi (which inserts into the ulnar side of the little finger and is innervated by the radial nerve). With the hands held supine, **marked hypothenar wasting** with sparing of the thenar eminence can be seen, and no wasting of the forearm muscles. Look for scars or deformity from trauma, surgery or arthritis around the forearm and wrist.

**Motor function:** there is **weak abduction** (dorsal interossei) and **weak adduction** (palmar interossei) of the fingers. There is **weak thumb adduction** (adductor pollicis) as demonstrated by attempting to grip a piece of paper held between the borders of the index finger and extended thumb. Grip can be maintained only using thumb flexion (intact flexor pollicis longus served by the median nerve), known as **Froment’s sign**. There is **intact flexion of the fourth and fifth DIP joints**, as shown by the marked ulnar claw appearance (it is difficult to test actively when the fingers are held in fixed flexion). There is **intact medial/ulnar flexion of the wrist**.

**Sensory loss:** in the **ulnar distribution** over the fifth finger (see Figure 3.19), adjacent ulnar side of the fourth finger, ulnar side of the palm and dorsal aspect of the hand. (In distal ulnar nerve lesions this sensory loss may be variable or present only in the fifth finger.) Test along the radial side of the fourth finger and up the ulnar border of the wrist and forearm to check that this does not represent a C8–T1 lesion instead (muscle wasting would also be different, see Scenario 16).

**Additional tests:** you can ask to perform **Tinel’s test** tapping along the course of the ulnar nerve from the wrist along the ulnar border of the forearm and up to the elbow, which may elicit tingling from irritation of the nerve.

Proximal ulnar nerve lesion (at the elbow)

**Inspection:** with the hands held prone there is **dorsal guttering** and **marked wasting of the first interosseous** between the thumb and index finger. The hand shows a claw deformity with hyperextension at the fourth and fifth MCP joints and flexion at the fourth and fifth PIP and DIP joints, due to paralysis of the medial lumbricals. There is slight ulnar deviation of the fifth finger (known as **Wartenburg’s sign**) from unopposed action of extensor digit minimi (which inserts into the ulnar side of the little finger and is innervated by the radial nerve). With the hands held supine, **marked hypothenar wasting** with sparing of the thenar eminence can be seen, and no wasting of the forearm muscles. Look for scars or deformity from trauma, surgery or arthritis around the forearm and wrist.

**Motor function:** there is **weak abduction** (dorsal interossei) and **weak adduction** (palmar interossei) of the fingers. There is **weak thumb adduction** (adductor pollicis) as demonstrated by attempting to grip a piece of paper held between the borders of the index finger and extended thumb. Grip can be maintained only using thumb flexion (intact flexor pollicis longus served by the median nerve), known as **Froment’s sign**. There is **intact flexion of the fourth and fifth DIP joints**, as shown by the marked ulnar claw appearance (it is difficult to test actively when the fingers are held in fixed flexion). There is **intact medial/ulnar flexion of the wrist**.

**Sensory loss:** in the **ulnar distribution** over the fifth finger (see Figure 3.19), adjacent ulnar side of the fourth finger, ulnar side of the palm and dorsal aspect of the hand. (In distal ulnar nerve lesions this sensory loss may be variable or present only in the fifth finger.) Test along the radial side of the fourth finger and up the ulnar border of the wrist and forearm to check that this does not represent a C8–T1 lesion instead (muscle wasting would also be different, see Scenario 16).

**Additional tests:** you can ask to perform **Tinel’s test** tapping along the course of the ulnar nerve from the wrist along the ulnar border of the forearm and up to the elbow, which may elicit tingling from irritation of the nerve.
interosseous between the thumb and index finger. The hand shows a mild/no clawed appearance with hyperextension at the fourth and fifth MCP joints but only mild flexion at the fourth and fifth PIP joints and no flexion at the DIP joints. This is known as the ulnar paradox, with proximal lesions resulting in less clawing. There is slight ulnar deviation of the fifth finger known as Wartenburg's sign (see above). With the palms facing up, there is marked hypothenar wasting with sparing of the thenar eminence. In addition, there is wasting of the ulnar border of the forearm. Look for scars or deformity from trauma, surgery or arthritis around the elbow.

Motor function: there is weak abduction (dorsal interossei) and weak adduction (palmar interossei) of the fingers. There is weak thumb adduction (adductor pollicis) with Froment's sign (see above). There is loss of flexion of the fourth and fifth DIP joints (loss of flexor digitorum profundus with high lesions) as shown by holding the fourth and fifth MCP and PIP joints extended and asking the patient to try to flex the fourth and fifth DIP joints (this takes practice to demonstrate effectively!). There is weakness of medial/ulnar flexion of the wrist (flexor carpi ulnaris) not found in distal lesions (see above).

Sensory loss: this occurs in the ulnar distribution over the fifth finger, ulnar side of the fourth finger, ulnar side of the palm and dorsal aspect of the hand (see Figure 3.19).

Additional tests: you can ask to perform Tinel's test, tapping along the course of the ulnar nerve from the wrist up the ulnar border of the forearm towards the elbow, which may elicit tingling from nerve irritation.

The elbow flexion test can be used to test for ulnar nerve compression at the elbow, particularly in cubital tunnel syndrome. The elbow is flexed fully with the forearm supinated, and within 60 seconds the patient starts to feel pain or tingling in the fourth and fifth fingers.

**Differential diagnosis**

**Elbow lesions**
- Fractures:
  - Supracondylar fractures are most common and late complications of fracture/surgery can include cubitus valgus deformity of the elbow joint.
- Dislocation:
  - Arthritis: bony spur and narrowing of the ulnar groove
  - Compression: cubital tunnel syndrome describes constriction under the fibrous arch of the two points of insertion of flexor carpi ulnaris. Certain occupations are more at wrist such as secretaries (from leaning on elbows) and decorators (from repeated elbow flexion and extension).

**Wrist lesions**
- Fractures
- Ganglion
- Tumour
- Mononeuritis multiplex.

**Clinical judgement and maintaining patient welfare**

**Investigations**

**Imaging:** plain radiograph of elbow joint, ultrasoundography of the cubital tunnel or MRI.

**Nerve conduction studies** localise the site of the lesion

**Blood tests:** FBC, CRP, ESR, anti-nuclear antibodies (ANAs), anti-DNA, pANCA (perinuclear anti-neutrophil cytoplasmic antibody or ANCA), cANCA (cytoplasmic ANCA), rheumatoid factor, hepatitis B and C serology (mononeuritis multiplex).

**Management**

**Conservative:** avoidance of aggravating factors, physiotherapy, splinting, NSAIDs.

**Surgical:** transposition of the ulnar nerve and/or decompression of the cubital tunnel.
**Identifying clinical signs**

The radial nerve serves the extensors of the elbow, wrist and fingers. With knowledge of the anatomy of radial nerve innervation it is possible to describe the level of the lesion giving rise to clinical signs (see Figure 3.20).

**Radial nerve lesion at the axilla**

**Inspection**: there is wrist drop and slight finger flexion but with no wasting of the hand muscles. Look for surgical or traumatic scars anywhere along the route of the radial nerve from the axilla to the wrist.

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**Figure 3.20** Route and muscles supplied by the radial nerve
**Motor function:** there is weakness of all radially innervated muscles:

- **Weakness of elbow extension and flexion** (midway between supination and pronation)
- **Forearm supination** (tested with arm by the side and attempted supination of forearm against resistance applied to the patient’s hand)
- **Wrist extension and finger extension at the MCP joints.**

Both triceps and brachioradialis (biceps) deep tendon reflexes are absent.

**Sensory loss:** this occurs over the triceps, posterior forearm and first dorsal interosseous.

**Radial nerve lesion in the spiral groove of the humerus**

**Inspection:** there is wrist drop and slight finger flexion but with no wasting of the hand muscles. Look for surgical or traumatic scars.

**Motor function:** there is weakness of all radially innervated muscles below the triceps. There is weakness of elbow flexion (midway between supination and pronation) but elbow extension is intact (triceps innervation above the spiral groove). There is weakness of forearm supination, wrist extension and finger extension at the MCP joints. The triceps reflex is preserved but the brachioradialis (biceps) deep tendon reflex is absent.

**Sensory loss:** there is sensory loss over the posterior forearm and first dorsal interosseous. There may be variable loss of sensation over the triceps.

**Radial nerve lesion confined to the posterior interosseous nerve**

**Inspection:** there is wrist drop and slight finger flexion but with no wasting of the hand muscles. Look for surgical or traumatic scars.

**Motor function:** there is weakness of radially innervated muscles below the supinator (the supinator nerve comes off before the posterior interosseous nerve dips beneath the fibrous arcade of Frohse (‘supinator arch’) which is the common entrapment area). There is intact elbow flexion (midway between supination and pronation) and intact elbow extension and forearm supination. There is weakness of wrist extension and finger extension at the MCP joints. Triceps and brachioradialis deep tendon reflexes are intact.

**Sensory loss:** there is no sensory loss because the posterior interosseous branch is a purely motor nerve.

**Radial nerve lesion at the wrist**

**Inspection:** the arm and hand appear normal. Look for surgical or traumatic scars around the wrist.

**Motor function:** there is no motor weakness (because the radial nerve serves only extrinsic extensor muscles of the hand from above the wrist).

**Sensory loss:** there is sensory loss in the first dorsal interosseous only (as the sensory branch becomes superficial at the wrist).

**Differential diagnosis**

- **Axillary lesions:**
  - Fracture/dislocation of humeral head
  - Compression: use of shoulder crutch or ‘Saturday night palsy’ from prolonged hanging of the arm over the back of a chair (when intoxicated)

- **Spiral groove lesions:**
  - Fracture: mid-shaft fracture of the humerus
  - Compression: wheelchair users resting back of their arm against the chair

- **Posterior lesions:**
  - Compression: from the arcade of Frohse/supinator arch

- **Interosseous lesions:**
  - Tumours/lipomas or ganglia near the elbow
Wrist lesions:
- Fracture: at the distal radius
- Compression: tight bracelets/handcuffs/plaster casts.

Clinical judgement and maintaining patient welfare

Investigations
Electrophysiology: nerve conduction studies and EMG.

Imaging: ultrasonography/MRI may rarely be considered.

Management
- Conservative management of ‘Saturday night palsy’ with spontaneous improvement
- Physiotherapy and splinting for mild compressive lesions
- Surgical correction of fracture/dislocations.
Identifying clinical signs

Common peroneal nerve palsy is the most common cause of foot drop. However, a lesion in any of the areas, including the motor cortex, spinal cord, lumbar nerve roots L4–5, lumbar sacral plexus and sciatic nerve, and peripheral neuropathies or myopathies, can cause foot drop with different associated clinical signs. Knowledge of the anatomy of the common peroneal nerve will help determine the level of the lesion.

The sciatic nerve (L4, L5, S1–3) divides into its terminal branches, the tibial nerve and the common peroneal nerve, two-thirds down the posterior thigh. The tibial nerve serves the posterior compartment of the lower leg, producing plantar-flexion and inversion. The common peroneal nerve (Figure 3.21) serves the anterior part of the lower leg, winding around the neck of the fibula and dividing into the superficial peroneal nerve (foot eversion and sensation to lateral lower leg and dorsum of foot) and the deep peroneal nerve (foot and toe dorsiflexion and sensation to the dorsal web space between the hallux and second toe).

Common peroneal nerve palsy

Inspection: there is foot drop with a high-stepping gait. There is wasting of the anterolateral compartment of the lower leg. Look for surgical or traumatic scars near the knee and neck of the fibula. Look for any ankle supports/adapted footwear.

Figure 3.21 Route and muscles supplied by the common peroneal nerve and cutaneous distribution
Motor function: there is weakness of ankle dorsiflexion, and hallux extension (deep peroneal nerve) and eversion (superficial peroneal nerve). Test for eversion in a passively dorsiflexed foot because the everters cannot exert their action if the foot is in plantarflexion. The ankle jerk is intact and plantar reflex is downwards. There is normal plantarflexion and inversion. In mild cases, weakness may be seen only when asking the patient to walk on the heels.

Sensory loss: over the lateral calf and dorsum of the foot, but sparing the little toe. The little toe has sensation from the sural nerve, a branch of the tibial nerve.

Superficial peroneal nerve palsy
Inspection: there is wasting of the lateral compartment of the lower leg but no obvious high-stepping gait. Look for surgical or traumatic scars near the knee and neck of the fibula. Look for any ankle supports/adapted footwear.

Motor function: there is slight weakness of ankle dorsiflexion which might be seen only when the patient is asked to walk on the heels. There is no weakness in hallux extension, but there is weakness of eversion. There is normal plantarflexion and inversion.

Sensory loss: over the lower lateral calf and dorsum of the foot, but sparing the little toe.

Deep peroneal nerve palsy
Inspection: there is foot drop with a high-stepping gait. There is wasting of the anterior compartment of the lower leg. Look for surgical or traumatic scars near the knee and neck of the fibula, anterior lower leg. Look for any ankle supports/adapted footwear.

Motor function: there is weakness of ankle dorsiflexion (deep peroneal nerve) and hallux extension but intact eversion (superficial peroneal nerve).

Sensory loss: only in the dorsal web space between the hallux and second toe.

Differential diagnosis
Myopathy: tends to give rise to proximal weakness but there are distal variants. Signs will be bilateral. All foot movements will be weak including plantarflexion and hallux flexion. The ankle jerk may be reduced. There will be no sensory loss. There will be myopathic signs in the upper limbs and possibly the face.

Sensorimotor peripheral neuropathy: there is also weak plantarflexion and a stocking pattern of sensory loss including the little toe. Signs will be bilateral. There may be dorsal column signs and a positive Romberg’s test and sensorimotor signs in the upper limbs.

Neuromuscular junction disorder: signs tend to be bilateral with upper limb and facial involvement. Weakness will affect plantarflexion and be fatigable. Reflexes and sensation will be normal.

MND: signs are bilateral, although in early disease there can be asymmetry. There will be a mixture of UMN and LMN signs with increased tone, brisk ankle jerks, possibly upgoing plantar reflexes, marked wasting and fasciculations. There tend to be signs in the upper limbs and face. There are no sensory findings.

Sciatic nerve lesion: a peripheral nerve lesion affecting the sciatic nerve can cause foot drop, but also weak plantarflexion and inversion, weak knee flexion (with preserved knee extension) and hip extension. The ankle jerk is absent with a preserved knee jerk. Sensory loss will be along the posterior thigh, lower leg and foot, sparing the medial side of the lower leg. Injury to the sciatic nerve can occur through hip surgery, misplaced gluteal injections and pelvic pathology, such as trauma, haematoma, abscess or tumours.

Lumbosacral plexus lesion: presents similarly to a sciatic nerve lesion but with femoral nerve involvement. This will cause additional weakness in
hip flexion, abduction and adduction, and knee extension. The knee and ankle jerks are lost. There will be more extensive sensory loss including the anterior thigh and medial lower leg.

**L4–5 radiculopathy**: leads to similar findings as a common peroneal nerve lesion. In addition there will be weak hip abduction and adduction and variable effects on knee flexion and extension. The knee jerk (L3–4) may be lost but ankle jerks (S1–2) preserved. Sensory loss will include the medial part of the lower leg (L5). The patient often has back pain because nerve root compression usually arises from lumbar disc herniation (less commonly a tumour). Straight leg raise, stimulating nerve root irritation from stretching, will reproduce symptoms.

**Spinal cord lesion**: this can cause weakness in foot dorsiflexion but will also cause weakness in other muscle groups of the leg depending on the level of the lesion. Foot drop is not so obvious due to the UMN spasticity. Lesions can be unilateral or bilateral. Sensory signs will depend on the location of the lesion in the cord and there may be dissociated sensory loss, giving rise to the Brown–Séquard syndrome if the lesion is unilateral.

**Cortical lesion**: a tumour or stroke affecting the motor cortex, such as a lacunar infarct or parasagittal meningioma, can rarely cause localised lower limb weakness and foot drop. There will be UMN signs.

**Causes of common peroneal nerve palsy**
- Trauma
- Fibular fracture
- Knee surgery
- Compression: plaster cast, leg crossing, weight loss.

**Clinical judgement and maintaining patient welfare**

**Investigations**
- **Electrophysiology**: nerve conduction studies and EMG.
- **Imaging**: MRI might be considered.

**Management**
- Avoidance of aggravating factors such as leg crossing or squatting
- Physiotherapy and splinting of the ankle/foot
- Surgical repair or release if there has been transection or tethering.
Nystagmus will usually be a sign as part of another disorder such as cerebellar syndrome or MS, but rarely it can form the full case in the PACES neurology station.

Nystagmus is an involuntary rhythmic oscillation of the eye(s) that may be physiological, congenital or acquired. It represents a problem with the neural mechanisms/centres involved in maintaining image fixation on the fovea for optimal visual acuity.

**Identifying clinical signs**

Nystagmus is usually described with reference to:

- Monocular or binocular/conjugate
- Position: primary (looking forward) or only gaze related
- Type: pendular (equal velocity in either direction) or jerk: a slow drift then fast corrective phase – the direction of nystagmus refers to the fast phase
- Plane: horizontal, vertical or rotatory/torsional (sometimes it is easier to tell by looking at the pull on the conjunctival vessels).

**Cerebellar nystagmus**

This is a binocular/conjugate, primary and gaze-related jerk nystagmus which is in the horizontal plane, and the direction of the nystagmus (fast phase) is towards the same side as the cerebellar lesion and maximal on looking towards this side. The nystagmus does not fatigue with continued gaze to the affected side. There is also loss of smooth saccades.

Further cerebellar testing will show homolateral poor finger–nose pointing with dysmetria and intention tremor, dysdiadochokinesis, poor heel–shin coordination and an ataxic gait falling towards the side of the lesion. There may be dysarthric speech.

**Additional**

Finish your exam by asking to test the cranial nerves, particularly looking for any abnormality of cranial nerve V (corneal and facial sensation) or cranial nerve VIII which may suggest a lesion at the cerebellopontine angle (see Scenario 27). Ask to examine the fundus for optic atrophy which may be present in MS.

Ask to examine the upper and lower limbs, which may show pyramidal weakness consistent with MS, or dorsal column signs in alcohol misuse or vitamin B₁₂ deficiency.

Romberg’s test is not a test of cerebellar function, but rather of dorsal column/proprioceptive function. A patient with a cerebellar lesion cannot usually stand steady, feet together with arms by the sides even with the eyes open, so Romberg’s test cannot be performed.

**Differential diagnosis**

Unilateral cerebellar pathology and nystagmus will tend to be caused by structural lesions:

- Cerebrovascular events, eg lateral medullary syndrome
- Demyelination, eg MS
- Cerebellar/posterior fossa tumours, eg astrocytomas, haemangioblastomas, medulloblastomas and metastatic disease (breast, lung, skin, kidney).

Bilateral cerebellar nystagmus will be caused by systemic pathology:

- Toxins (alcohol, chemotherapy and anticonvulsants)
- Autoimmune and paraneoplastic processes
- Inherited disorders involving cerebellar degeneration, eg olivopontocerebellar
degeneration and Friedreich’s ataxia (see Scenario 12).

**Vestibular nystagmus**

**Peripheral vestibular nystagmus**: this is a binocular/conjugate horizontal or rotatory/torsional nystagmus which is a primary and gaze-related unidirectional jerk nystagmus with the fast component maximal to the opposite side of the vestibular lesion. With upward or downward gaze the nystagmus remains horizontal and in the same direction. With continued gaze away from the side of the lesion the nystagmus fatigues. There are no cerebellar signs. The gait may be unsteady, falling towards the side of the lesion, and the patient may describe vertigo symptoms. These are often worse when testing gait. Tinnitus and deafness may be found on the side of the lesion.

**Differential diagnosis**
The peripheral vestibular system includes the semicircular canals, otoliths and the vestibular portion of cranial nerve VIII.

The causes of peripheral vestibular nystagmus include labyrinthitis, acoustic neuroma, Ménière’s disease, benign paroxysmal positional vertigo (BPPV), autoimmune inner ear disease (AIED) and degenerative middle-ear disease such as otosclerosis.

**Central vestibular nystagmus**

This is a binocular/conjugate horizontal/vertical/rotatory or mixed (may appear chaotic) nystagmus, which is a primary and gaze-related nystagmus. It is multidirectional so that on looking to the left it is leftward (fast component to the left), looking to the right it is rightward and looking up it is upward. There is no fatigue of the nystagmus on sustained gaze in any direction. The patient may have a tendency to fall in any direction on testing gait; vertigo is unusual. There are no peripheral symptoms such as tinnitus or deafness and no cerebellar signs.

**Differential diagnosis**
The central vestibular system includes the vestibular nerve nuclei and their projections to the cerebellum, extraocular nuclei via the medial longitudinal fasciculus, the spinal cord via the vestibulospinal tract and projections to the cortex.

Causes of central vestibular nystagmus include brain-stem stroke and vertebrobasilar insufficiency, brain-stem tumours, demyelination such as MS, syringobulbia (see Scenario 7) and basilar-type migraine (some classify this as a cause of peripheral vestibular nystagmus when it is associated with benign paroxysmal positional vertigo).

**Pendular nystagmus**

This is a conjugate or monocular multidirectional nystagmus that can appear chaotic. The oscillation has equal velocity in all directions. It is present in all positions including the primary position.

**Additional**: look around the bed for clues of visual aids used by the blind patient and a general appearance that might suggest albinism. Ask to perform fundoscopy to look for optic atrophy, signs of retinitis pigmentosa and cataracts with disruption of the red reflex.

**Internuclear ophthalmoplegia (INO or ataxic nystagmus)**

There is failure of conjugate eye movements on lateral gaze. In the primary position there may be a divergent strabismus. In a left-sided INO (lesion...
in the ipsilateral MLF) there is **partial or total failure of adduction** of the left eye on looking right, but normal abduction of the right eye, with **jerk horizontal nystagmus in the abducting eye** with fast corrective phase towards the right side (opposite side to the lesion). The nystagmus of the abducting eye is not necessary for the diagnosis of INO. The patient may describe diplopia when looking to the right (contralateral) side. It can be shown that this is not a left medial rectus palsy by the fact that, by covering the right abducting eye, the left eye adducts normally with convergence. Sometimes convergence can be affected if the lesion extends into the midbrain. In a bilateral INO there will be failure of adduction of either eye with lateral gaze in the opposite direction.

**Additional**

State that you would like to look for additional eye signs supportive of MS such as optic atrophy, visual field defects, optic disc pallor on fundoscopy and/or a relative afferent pupillary defect (RAPD). Look for cerebellar signs in support of MS or brain-stem infarction.

**Differential diagnosis**

INO is due to a lesion in the MLF within the pons and midbrain, which connects the contralateral nerve VI nucleus to the ipsilateral oculomotor (nerve III) nucleus.

MS is the most common cause in a younger person, and brain-stem infarction causing unilateral INO is the most common cause in an older person. Other causes include brain-stem tumours, viral infection, syphilis infection, Lyme disease, trauma, Arnold–Chiari malformation, syringobulbia, and drug (phenothiazines, phenytoin, tricyclic antidepressants) and alcohol intoxication.

**Downbeat nystagmus**

There is a **bilateral downbeat nystagmus in the primary gaze**, which remains downbeat in all directions of gaze. Lateral gaze may accentuate the nystagmus.

**Additional**

Look for signs of syringobulbia and syringomyelia which may occur together with an Arnold–Chiari malformation, eg bulbar palsy, INO, balaclava-helmet loss of facial sensation, a dissociated sensory loss usually in a cape distribution, LMN signs in the upper limbs and UMN signs in the lower limbs. Look for cerebellar signs that may also suggest syringobulbia or a posterior fossa tumour.

**Differential diagnosis**

Downbeat nystagmus usually signifies pathology at the craniocervical junction. Arnold–Chiari malformation is the most common cause. Downbeat nystagmus may also be seen with brain-stem stroke, syringobulbia, spinocerebellar degeneration, MS and drug (phenytoin, lithium) and alcohol toxicity.

**Upbeat nystagmus**

There is a **bilateral vertical nystagmus** in the primary position with the fast phase beating in the upward position.

If the nystagmus increases on upward gaze, this suggests pathology in the anterior vermis of the cerebellum. There may be other cerebellar signs such as loss of smooth saccades, slurred speech and truncal ataxia.

If the nystagmus increases on downward gaze, this suggests pathology in the medulla. There may be associated brain-stem signs such as palatal weakness with nasal speech.

**Physiological nystagmus**

This is a gaze-evoked jerk nystagmus occurring at the extremes of gaze and absent in the primary position. The elastic pull of the extraocular mus-
cles and tendons exerts a force that tends to bring the eye back to the midline, but the neural integrator tries to overcome this with corrective quick movement (jerk nystagmus) to the desired extreme of gaze.

Clinical judgement and maintaining patient welfare

Investigations
These will be determined by the type of nystagmus and the associated differential diagnosis.

Management
- Cessation of causative medications
- Correction of refractive errors with contact lenses
- Downbeat nystagmus may be treated with base-out prisms, which induce convergence
- Botulinum toxin injection into rectus muscles can ameliorate acquired nystagmus. However, this diminishes all types of eye movement and can cause diplopia, ptosis and increased nystagmus in the uninjected eye.
SCENARIO 22. OPHTHALMOPLEGIA

The most common gaze palsies seen in the PACES examination are cranial nerve III and VI palsies. Remember to look for associated cranial nerve abnormalities or peripheral examination signs that will point towards a specific disease or help further localise the lesion.

It is worth revisiting the anatomy and function of the extraocular nerves III, IV and VI together with their muscles so that their examination becomes straightforward (see Figures 3.22 and 3.23).

Nerve III (oculomotor) palsy

See Figure 3.24. Nerve III supplies most of the extraocular muscles including superior rectus (elevation), medial rectus (adduction) and inferior rectus (adduction and intorsion).
oblique (extorsion, elevation and abduction). It also elevates the eyelid through its innervation of levator palpebrae superioris and carries preganglionic parasympathetic innervation which causes pupil constriction via the sphincter (constrictor) pupillae.

The nerve III nucleus in the midbrain is divided into subnuclei to innervate the separate muscles, and it is partnered with the nearby Edinger–Westphal nucleus which gives rise to the preganglionic parasympathetic fibres; these lie superficially within the nerve and are therefore easily compromised by compressive lesions.

The subnuclei serve the IPSILATERAL extraocular muscles (medial rectus, inferior rectus and inferior oblique) with two important exceptions:

1. The nucleus sends fibres across to the opposite oculomotor nucleus, which then innervates the CONTRALATERAL superior rectus.
2. The nucleus supplies BOTH levator palpebrae with crossed and uncrossed fibres.

The nerve III fascicles/trunks leave the midbrain passing ventrally. Nerve III then passes in the subarachnoid space and runs close to the posterior communicating artery. It enters the lateral side of the cavernous sinus, crosses over the trochlear nerve, and exits medially as superior and inferior branches through the superior orbital fissure. The superior branch serves levator palpebrae superioris and superior rectus, which is also joined by sympathetic supply from the internal carotid artery. The inferior branch serves the other oculomotor muscles and carries the parasympathetic axons to the constrictor pupillae.

**Identifying clinical signs**

There is a left-sided ptosis (see Figure 3.24) and when the eyelid is raised the eye is in a **down and out** position (due to the unopposed combined action of lateral rectus and superior oblique), giving a **divergent strabismus/squint**. The patient cannot move the affected eye across the midline. Diplopia is maximal on trying to look away from the affected side and up. There is a **dilated and non-reactive pupil** (to light or attempted accommodation) from parasympathetic fibre involvement.

With a nuclear lesion there will be contralateral eye signs in addition to the ipsilateral signs. There is a contralateral partial ptosis (bilateral innervation of levator palpebrae superioris) but ptosis will be more pronounced on the ipsilateral side. There will also be a contralateral elevation palsy (the nucleus innervates the contralateral superior rectus). Remember, there is an ipsilateral elevation palsy because fibres from the contralateral subnucleus

![Figure 3.24](left-sided-III-nerve-palsy.png)
pass through the ipsilateral subnucleus before innervating the superior rectus.

**Differential diagnosis**

**Nuclear lesion:** infarction, haemorrhage, tumour, abscess, demyelination.

**Midbrain lesion:** herniation, infarction, haemorrhage, tumour, abscess, demyelination (MS).

**Subarachnoid lesion:** aneurysm, haemorrhage, meningitis, inflammation including vasculitides (giving rise to mononeuritis multiplex), tumour, migraine.

**Cavernous sinus lesion:** tumour (pituitary, cranio-pharyngioma), thrombosis, aneurysm, fistula, infection, inflammatory.

**Orbital lesion:** trauma, tumour.

**Small vessel disease:** diabetes, hypertension, atherosclerosis.

**Infection:** Lyme disease, syphilis, basilar meningitis (bacterial, mycobacterial, fungal, parasitic).

**Nerve IV (trochlear) palsy**

Nerve IV supplies the superior oblique muscle which intorts, depresses and abducts the globe. This combined action allows the eye to look down and in. The nerve IV nuclei lie in the midbrain where the nerves decussate and then exit dorsally. It is the only cranial nerve to exit the brain dorsally and it has a long course, making it susceptible to trauma.

A nerve IV palsy can be very subtle so a candidate must actively look for it.

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**Figure 3.25** Right-sided IV nerve palsy. Reproduced with the kind permission of Professor Chua Chung Nen
Identifying clinical signs
The right eye appears slightly elevated/normal and the patient has a head tilt away from the side of the lesion, tucking the chin in slightly to bring the visual axis of the affected eye central again (see Figure 3.25). The affected eye cannot look down in adduction (towards the nose). It is in this position that the patient experiences most vertical diplopia (giving rise to the classic history of difficulty reading books or climbing stairs when this direction of gaze is needed). Remember that diplopia is always worse in the direction of gaze of the paretic muscle. The false outer/upper image disappears when the affected eye is covered.

Differential diagnosis
- Congenital
- Trauma (most common cause)
- Small vessel disease: diabetes, hypertension, atherosclerosis
- Inflammatory: mononeuritis multiplex, peripheral neuropathy
- Infection: Lyme disease, syphilis, basal meningitis (bacterial, mycobacterial, fungal, parasitic)
- Midbrain/nuclear lesion: infarct, haemorrhage, tumour, abscess, demyelination (MS)
- Cavernous sinus lesion: tumour (pituitary, craniopharyngioma), thrombosis, aneurysm, fistula, haemorrhage, infection, inflammatory.

Nerve VI (abducens) palsy
Nerve VI innervates the ipsilateral lateral rectus, which abducts the eye. The nerve VI nucleus is in the caudal part of the pons. Approximately 40% of its neurons pass to the nearby MLF, to then cross over to the contralateral nerve III nucleus to innervate the contralateral medial rectus and produce conjugate lateral gaze.

Figure 3.26  Left sided VI nerve palsy. Reproduced with the kind permission of Professor Chua Chung Nen
Identifying clinical signs
There is a convergent strabismus in the primary position due to the unopposed action of the intact medial rectus (see Figure 3.26). There is horizontal diplopia maximal on attempted gaze in the direction of the paretic muscle. The outer image disappears on covering the affected eye.

On testing the remaining cranial nerves, pay particular attention to nerves VII and VIII, and check for nystagmus and other cerebellar signs that would indicate a cerebellopontine angle lesion. There may be signs of bilateral papilloedema on fundoscopy from a space-occupying lesion or idiopathic intracranial hypertension. Here, the nerve VI palsy acts as a ‘false localising sign’ due to downward displacement of the brain stem causing stretching of the abducens nerve.

Differential diagnosis
- Congenital: congenital absence of nerve VI – Duane’s syndrome
- Trauma
- Raised ICP: space-occupying lesion or idiopathic intracranial hypertension
- Small vessel disease: diabetes, hypertension, atherosclerosis
- Inflammatory: mononeuritis multiplex, postviral, peripheral neuropathy
- Infection: Lyme disease, syphilis, basilar meningitis (bacterial, mycobacterial, fungal, parasitic)
- Pontine/nuclear lesion: infarct, haemorrhage, tumour, abscess, demyelination (MS)
- Petro-sous bone pathology: in severe ongoing otitis media there can be infiltrative osteomyelitis involving the petrous temporal bone
- Cavernous sinus lesion: tumour (pituitary, craniopharyngioma), thrombosis, aneurysm, fistula, haemorrhage, infection, inflammatory.

Complex ophthalmoplegia

Identifying clinical signs
Thyroid ophthalmopathy will usually present as a complex ophthalmoplegia not attributable to any single nerve lesion. This does not represent a true ophthalmoplegia. It is due to soft-tissue inflammation and swelling within the orbit causing restriction of eye movements. There is usually proptosis, chemosis, lid lag and other thyroid signs.

Myasthenia gravis may present as a complex ophthalmoplegia not attributable to any single nerve lesion. Eye movements are fatigable and there are no pupillary signs.

The Miller–Fisher variant of Guillain–Barré syndrome may present initially with an ophthalmoplegia, with the descending paralysis from peripheral demyelination giving the classic triad of ophthalmoplegia, ataxia and areflexia.

Chronic progressive external ophthalmoplegia (CPEO) is the most common manifestation of mitochondrial myopathy, in itself very rare, and usually presents as a bilateral progressive ptosis, which proceeds to a bilateral ophthalmoplegia without pupillary changes. Kearns–Sayre syndrome is a mitochondrial myopathy that presents with the triad of age <20 years, CPEO and retinitis pigmentosa. Other associated features include cerebellar syndrome, cognitive impairment, Babinski’s sign, hearing loss, seizures, short stature and delayed puberty, with other endocrine abnormalities and cardiac conduction defects.

Oculopharyngeal dystrophy (see Scenario 10) is an autosomal dominant trinucleotide repeat disease occurring in 60–70 year olds, with progressive ptosis and ophthalmoplegia without pupillary changes, leading to dysphagia and
facial weakness, and in the latter stages of disease to proximal muscle weakness.

Cavernous sinus syndrome

Structures contained within the cavernous sinus:

- Internal carotid artery
- Sympathetic carotid plexus
- Cranial nerves: III, IV, VI and V1 and V2 branches (V3 lies outside the sinus).

Cavernous sinus syndrome signs:

- Painful ophthalmoplegia (unilateral single or usually combined nerve III, IV and VI palsies)
- Horner’s syndrome (with no associated anhidrosis because the lesion occurs after the superior cervical ganglion and the pupil may be mid-position and fixed with both parasympathetic and sympathetic disruption)
- Anaesthesia of forehead, maxilla and conjunctiva (V1 and V2 branches)
- Proptosis (if pulsating suggests carotid–cavernous fistula)
- Conjunctival injection with chemosis
- Papilloedema ± visual loss
- Orbital bruit.

Differential diagnosis of cavernous sinus syndrome

- Tumours: meningiomas, extension of pituitary or craniopharyngiomas, metastatic disease
- Vascular: cavernous sinus aneurysms or fistulae
- Thrombosis: usually complicating infection of the ethmoid, frontal and sphenoid sinuses or extension of dental or orbital infection
- Inflammatory: herpes zoster, sarcoidosis and Wegener’s granulomatosis
- Idiopathic: Tolosa–Hunt syndrome is a rare granulomatous inflammation of the cavernous sinus and superior orbital fissure. It causes a painful ophthalmoplegia ± pupillary effects.

Clinical judgement and maintaining patient welfare

Investigations

- These are urgent if an aneurysm, subarachnoid haemorrhage (SAH), uncal herniation, meningitis, stroke or trauma is suspected.
- Imaging: CT or MRI of the brain is indicated if suspecting aneurysmal, SAH, stroke, space-occupying lesion ± herniation, or traumatic cause. Cerebral angiography may be needed to investigate aneurysmal disease and arteriovenous (AV) malformations including fistulae.
- Blood tests: investigations for small-vessel disease will include fasting blood glucose/HbA1c, autoimmune profile, pANCA and cANCA, ESR and CRP if suspecting giant cell arteritis (GCA).
- Lumbar puncture: indicated if suspecting meningitis, and a space-occupying lesion has been ruled out.

Management

This is directed by the underlying cause.

Nerve III (oculomotor) palsies may resolve spontaneously over months if the underlying cause is ischaemia (typically in hypertensive or diabetic patients) of the vasa nervosa. This typically gives relative sparing of the pupil and is often painful for unknown reasons. NSAIDs may ameliorate this. Patching of the deviated eye can be a useful short-term measure. In the long term, surgical correction may be indicated for a non-resolving stable angle.

Nerve IV (trochlear) palsies have been treated successfully with botox injection (of other muscles), prisms and surgical correction.
Nerve VI (abducens) palsies, when isolated in children and young patients, are often benign and resolve spontaneously within 6 months. The cause is unclear. Alternate patching may be useful to prevent amblyopia. In older patients GCA should be considered and treated with steroids if appropriate.
STATION 5

Integrated Clinical Assessment
STRUCTURE OF THE ICA

The older Station 5 (until 2009) involved a series of three short cases based around examination of the eyes, an endocrine disorder or rheumatological condition. Like the other examination stations, this involved a ritualised examination followed by presentation of findings to the examiners. There are still ‘echoes’ of this old format in many textbooks and courses; skin, locomotor and endocrine scenarios have been shoe-horned into an ‘integrated clinical assessment’ (ICA) format. This slightly misrepresents the content of the exam, which is highly varied and demands far more than an ability to elicit clinical signs. Indeed, this is the only station of the PACES exam that assesses candidates in all categories of the marking scheme.

Station 5 is organised as follows:

• Two 10-minute cases known as ‘brief clinical consultations’.
• The candidate has 8 minutes with the patient to take a focused history, carry out a relevant examination, reach a diagnosis or identify a clinical problem and then communicate this to the patient.
• The remaining 2-minute discussion with the examiners will not begin until these 8 minutes have elapsed.
• The examiner will ask the candidate to state the positive physical findings, their concluding diagnosis and differential diagnoses (if appropriate) based on their assessment.

CONTENT AND MARKING OF THE ICA

The cases found in Station 5 typically involve a presenting complaint that guides you to the relevant system for a targeted examination. Alternatively, there may be obvious clinical signs (e.g., thyroid eye disease) and a rather vague history.

The seven areas in which candidates are scored are summarised below, with candidates being scored on a three-point scale: satisfactory, borderline or unsatisfactory.

Clinical communication skills
• Eliciting a history relevant to the complaint
• Explaining information to the patient in a focused, fluent and professional manner

Physical examination
Performing an examination in a correct, appropriate, practised and professional manner

Clinical judgement
Selecting a sensible and appropriate investigation and treatment plan

Managing patient’s concerns
• Detecting, acknowledging and attempting to address patient’s concerns
• Listening
• Demonstrating empathy

Identifying physical signs
• Identifying the correct physical signs
• Not finding signs that are not present

Differential diagnosis
Constructing a sensible differential diagnosis, including the correct diagnosis

Maintaining patient welfare
Treating the patient respectfully and sensitively, ensuring comfort, safety and dignity

A formal marksheet is shown on page can be found on the MRCP(UK) website.
INFORMATION GIVEN TO CANDIDATES, PATIENTS AND EXAMINERS

Before each Station 5 examination, each candidate, patient and examiner will be given some information.

Below is a worked example of the information given to each person based on guidance on the MRCP website (www.mrcpuk.org).

Information to candidate

You will be asked to see two patients at this station. The clinical information about one of these patients is given in the box below. You should have a second sheet giving you information about the other patient.

- You have 10 minutes with each patient. The examiners will alert you when 6 minutes have elapsed and will stop you after 8 minutes.
- In the remaining 2 minutes, one examiner will ask you to report abnormal physical signs (if any), your diagnosis or differential diagnosis, and your plan for management (if not already clear from your discussion with the patient).

Your role: You are the medical doctor on call

Patient name: Mrs Beverley Gordon – age 39 years

This lady was admitted to the orthopaedic ward for a carpal tunnel release operation.

She mentioned to the orthopaedic doctors that she has swelling in her neck and they have asked for the opinion of a physician. You were asked by your consultant to see the patient and to assess the suspected swelling in her neck.

Your task is to assess the patient’s problems and address any questions or concerns raised by the patient.

- You should assess the problem by means of a relevant clinical history and a relevant physical examination. You do not need to complete the history before carrying out appropriate examination.
- You should respond to any questions the patient may have, advise the patient of your probable diagnosis (or differential diagnoses) and your plan for investigation and treatment where appropriate.
- You have 8 minutes to complete the task.

Accompanying notes are given to each patient and may take the following format.
Information for patients

The doctors sitting the examination have been asked to assess your problem. They will have 8 minutes to ask you about the problem and any other relevant issues. They will also examine you. They should explain to you what they think is wrong and what action should be taken and answer any questions you have, for example about the diagnosis, tests that may be needed, or treatment. One of the examiners will ask them to describe any abnormal examination findings and give their diagnosis.

Your history is described below.

You are: Mrs Beverley Gordon – age 39 years

Your problem: A swelling in your neck

You are in the orthopaedic ward and you mentioned to the admitting doctor that your neck seemed swollen. One of the medical doctors has been asked to see you about this.

You have been suffering from pain in your right forearm and a numb or tingly feeling in your right hand – affecting your third, fourth and fifth fingers. The aching in your arm is worse at night and you have found it more and more difficult to get comfortable. The problem has been present for several years and was worse when you were pregnant with your daughter, 3 years ago, but improved for a time after that.

A trapped nerve in the wrist has been diagnosed. You are having a short admission to have the nerve released.

You mentioned that you thought your neck was swollen when the admitting doctor was examining you. You have not mentioned this to a doctor before, but your sister has been commenting on it for a few years. Your sister is 36 and has an underactive thyroid.

You do not have any of the symptoms your sister had when her thyroid was underactive.

Indeed you feel well. Your weight is stable. Your skin and hair are normal. You do not seem to feel the heat or the cold any more than anyone else. Your bowel works normally. You do not have any problem with swallowing.

You should ask why your neck seems swollen and whether there is something wrong with your thyroid gland. You should ask if any tests are required and, if so, what these tests will be.

Finally, for the same worked example, the examiners have the following information, which would typically list the findings the candidate would be expected to observe or elicit.
**Information for examiners**

**Patient:** Mrs Beverley Gordon – age 39 years

Examiners should discuss and agree the criteria for pass and for fail in the competencies being assessed.

As a general guide, candidates would be expected to:

- Note the history of neck swelling and the family history of thyroid disease
- Enquire about symptoms of disturbed thyroid function
- Examine the neck and identify the smooth and symmetrical thyroid enlargement. Note absence of bruit
- Examine for signs of overactive and underactive thyroid gland and confirm clinical euthyroid state
- Confirm to the patient that her thyroid gland does seem enlarged but reassure her that the gland seems to be working normally judging from clinical examination. Advise on appropriate further investigations.

The lead examiner should:

- Advise the candidate after 6 minutes have elapsed that ‘You have two minutes remaining with your patient’
- Ask the candidate to describe any abnormal physical findings that have been identified
- Ask the candidate to give the preferred diagnosis and any differential diagnosis that is being considered
- Ensure any remaining areas of uncertainty, e.g., regarding the plan for investigation or management of the problem, are addressed in any time that remains.

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**THE APPROACH TO STATION 5**

It is very easy to fall into the familiar modes of the history-taking station or the examination stations, then find, with a minute remaining, that you have either almost wordlessly examined your patient without garnering any information about their symptoms or that you have taken a thorough history but examined nothing. You may choose to attempt to divide each consultation between history and examination, but we strongly recommend taking a history and examination in parallel, just as you would during a busy on-call or clinic.

It is also crucial to remember that the examiners are not looking for the ritualised, detailed and complete examination of systems as in other stations, or for a full history. They are testing your ability to perform a targeted examination and elicit the most salient symptoms. This is not solely for the purpose of forming a differential diagnosis – don’t forget that you have been specifically asked to address the patient’s principal concerns and must therefore explain to them what you think the underlying problem is and how you are going to investigate and manage it.

Each of the Station 5 cases in this chapter is based around a flow chart that gives a worked example of how you might perform an ‘integrated clinical assessment’ (ICA) for a given presenting condition.

All cases can be divided into a new condition or an exacerbation of an existing chronic illness. These require slightly different emphasis in the history taking and examination, and this is reflected in each flow chart.

An important feature of this approach is the ‘call-back’, wherein towards the end of the consultation candidates re-state the presenting complaint of the patient and any other specific concerns they raised during the consultation. This is an easy way of demonstrating to the examiners that you have registered and understood the patient’s main concern (which is not necessarily the main clinical priority). It also gives the patient an opportunity to mention other salient points which the candidate missed. If you have built a good rapport with the patients/actor, they may help
you out at this stage by volunteering important information.

The consultation should end with you describing:

- How you will address the patient’s main concern (e.g., analgesia)
- What you think the diagnosis may be
- What investigations and initial treatments are necessary.

This is an opportunity to demonstrate to the examiner that you have come to a reasonable differential diagnosis and formed a safe and efficient investigation and management plan.

**EXAMINATION TECHNIQUES FOR STATION 5**

Station 5 often requires examination routines not encountered elsewhere in PACES, for example examination of a goitre. These are described below.

**Examination of the hands**

It is important to ensure the patient is comfortable and to rest the hands on a pillow. Expose the hands and forearms up to and including the elbows. A large proportion of the patients may have painful and tender joints and it is imperative therefore to ask about this before palpating the joints, which should be done after careful inspection.

A rheumatological examination typically involves inspection, palpation, neurological assessment, functional assessment and examination for extra-articular signs. However, you may not have time to do all of these completely.

1. **Inspection**
   The bulk of the available information will be gathered by inspection rather than palpation or active movement.

   - Peripheral accessories, e.g., walking stick
   - Peripheral arthropathies, e.g., knees, ankles
   - Systemic sclerosis – tight, shiny, stretched skin with beaked nose +/- telangectasia
   - Cushingoid appearance – steroid treatment
   - Horner’s syndrome – T1 lesion (see page...)
   - Ears for evidence of psoriasis, or gouty tophi in helix of ear.

   Then make an assessment of:

   - **Nails** – pitting, onycholysis, clubbing, nail fold infarcts, Beau’s lines
   - **Skin** – tight shiny skin over dorsum of hand or fingers (scleroderma); tissue paper thin +/- purpura (steroid therapy); surgical scars (joint replacement); tar staining
   - **Muscles**
     - bilateral wasting of the small muscles with dorsal guttering (rheumatoid arthritis, syringomyelia, motor neurone disease)
     - unilateral wasting of the small muscles of the hand (C8/T1 root lesion, e.g., cervical rib, Pancoast tumour)
     - unilateral wasting involving thenar eminence (median nerve, e.g., carpal tunnel syndrome)
     - unilateral wasting sparing thenar eminence (ulnar nerve, e.g., elbow trauma)
   - **Joints** (in order to describe the location of the abnormality accurately, candidates should know the names of the bones and joints)
   - **Distribution** of any abnormalities – symmetrical (e.g., rheumatoid arthritis) or asymmetrical (e.g., seronegative arthritides); proximal or distal joints
   - **Specific deformities** e.g., ‘swan neck’, ‘Boutonniere’, Z-shaped thumb, subluxation, ulnar deviation, Heberden’s nodes, gouty tophi
   - **Inflammation** – calor, rubor, dolor, tumor and loss of function
     - NB rubor is replaced with shininess of the skin in those with dark skin

Once the hands have been inspected, an assessment of the elbows is essential, as this can reveal
psoriatic plaques or rheumatoid nodules (indicating sero-positive rheumatoid arthritis).

The whole examination can be achieved by three movements – assessment of the dorsum of the hand, then plantar aspect, followed by crossing the arms over and exposing the extensors surface of the elbows.

2. Palpation (with caution)
This must only be assessed with extreme caution on the examination and only where there is an absolute requirement to do so in the time given, such as if there is a specific complaint by the patient (ie painful joints) or inspection has demonstrated a specific abnormality.

- Palm – Dupuytren’s contracture
- Elbow nodules
- Joints – palpate any swelling to determine whether it is soft and boggy (rheumatoid arthritis) or hard and bony (Heberden’s nodes or gouty tophi)
- Skin – tightness or calcinosis in finger pulps (scleroderma/CREST).

This does not mean that you need to examine every joint in the hands, but merely the joints involved, eg MCP joints.

3. Functional assessment of the hands
You will be expected to perform this assessment if the patient reports reduced function.

- First, you can ask the patient to make a fist, followed by the prayer sign, followed by the reverse prayer sign.
- Finally, you could ask the patient to do up a button on a shirt or hold a pen.

These manoeuvres have been validated against more complex assessments of function and provide a quick means of assessing the likelihood of impaired function in daily activities.

Neurological assessment – if relevant to the presenting complaint (eg symptoms of carpal tunnel syndrome), one should proceed to perform a neurological assessment focusing particularly on the median and ulnar nerve. Single nerve lesions are occasionally encountered in this station.

4. Extra-articular signs
Having screened for symptoms suggestive of systemic manifestations of joint disease, examination should then focus on the affected system.

Examination of the axial spine
A patient may present with stiffness of the axial spine and restricted movements (eg as in ankylosing spondylitis and other spondylarthropathies).

Inspection is crucial again. For example, the ‘question mark posture’ (loss of lumbar lordosis, fixed kyphoscoliosis of the thoracic spine with extensive of the cervical spine) of ankylosing spondylitis can be a ‘spot diagnosis’. The characteristic posture is usually immediately evident unless the patient is lying down with the head supported by pillows.

The following examination routine should stage disease and detect associated signs:

1. Establish restricted spinal movement
Two quick tests can be performed:

   Lumbar spine: modified Shober’s index:
   With the patient standing upright, place two marks 10 cm apart on the lumbar spine in the midline. The lower mark is at the level of the posterior superior iliac spines. The patient then flexes forward (ask them to touch their toes) and at maximal flexion the distance is re-measured. In normal subjects there is an expansion of at least 5 cm between the two marks. Lower values indicate decreased mobility of the lumbar spine.

   Thoracic spine: Occiput-to-wall distance:
   The subject stands with their back against a wall (both heels and buttocks must be touching the wall) with a horizontal gaze. In normal subjects
the occiput will touch the wall. Any wall-to-
occiput gap is a measure of restriction of the
thoracic and cervical spines.

2. Examine for sacroiliitis/enthesitis
Examine (carefully) for tenderness over the sa-
croiliac joints. Palpate for evidence of other en-
thesitides over the heels, costochondral joints
and iliac crest.

Tell the examiner you would like to perform the
FABERE (flexion, abduction, external rotation and
extension) test. The patient places one ankle on
the opposite knee and allows the ipsilateral knee
to fall outwards (external rotation at the hip) to
form a figure ‘4’. If this causes pain over the sacroiliac joint, sacroiliitis should be suspected.

3. Exclude extra-articular manifestations from
head to toe:
If a spondylarthropathy is suspected (eg ankylos-
ing spondylitis), a focused history should include
questions related to the extra-articular manifesta-
tions associated with these conditions.

- **Eyes**: acute uveitis
- **Mouth**: mucosal inflammation manifesting as
  oral ulceration is common
- **Chest**: apical fibrobulous disease (1%)
- **Cardiac**: aortic root dilatation and associated
  aortic valve incompetence
- **Abdomen**: 15–20% will develop symptomatic
  Crohn’s disease (stoma present?). Look for
evidence of amyloidosis (hepatomegaly,
evidence of renal failure or replacement
  therapy)
- **Nervous system**: paraesthesia, signs of cord
  compression
- **Feet**: Achilles tendonitis and plantar fasciitis

**Examination of thyroid status**

The assessment of the thyroid status of a patient
is a fundamental clinical skill that should not
present difficulties provided that the following
scheme is followed. For obvious reasons, patients
with severe hyper- or hypothyroidism are unlikely
to appear, but over/under-replacement of thyrox-
ine is quite a common scenario.

**General observation**
- **Hypothyroidism**
  - Pale dry skin
  - ‘Peaches and cream’ complexio
  - Dry hair
    - Note: Loss of the outer one-third of the
eyebrows is unreliable and non-specific
- **Hyperthyroidism**
  - Anxious, fidgety patient
  - Staring eyes (lid retraction)
  - Sweating

**Hands**
Shake their hands
- Warm and sweaty or cool and dry?
- Fine tremor – hands outstretched with a piece
  of paper resting on fingers
- **Pulse**
  - Rate
  - Rhythm (AF often occurs in thyrotoxicosis)
  - Volume (typically large volume and
collapsing in hyperthyroidism)
- **Thyroid acropachy** – a rare feature of Graves’
disease
- **Tar staining** – Graves’ ophthalmopathy is
  worse in smokers

**Neuromuscular manifestations**
- Reflexes: Slow relaxing in hypothyroidism; brisk in thyrotoxicosis
- Proximal myopathy: Thyrotoxicosis – ask the
  patient to stand from a chair unaided

**Dermatological manifestations of
thyroid disease**
- **Graves’ dermopathy**
  - Sheet-like myxoedema – coarse diffuse skin with
    non-pitting oedema
  - Nodular localised – violaceous infiltrative waxy
    area on the shin, resembling erythema nodosum
  - Horny – papilliform irregular firm red dermopathy
    on shin/upper foot
Examination of the thyroid gland

Follow the sequence of inspection, palpation and percussion. If you find a goitre, it would be prudent to comment on the thyroid status, combining reported symptoms and a formal examination of thyroid status as above. You must also comment on the most likely aetiology.

Inspection
Ask the patient to swallow a sip of water and look for upward movement of the thyroid gland. NB a thyroglossal cyst will move upwards both on swallowing and protrusion of the tongue, and can be trans-illuminated.

Is there any evidence from the history or examination that the thyroid is compressing any of the following:

- Trachea
  - monophasic syncope (rare), but the history may suggest some dyspnoea, particularly on lying flat
- Recurrent laryngeal nerve
  - hoarseness
- Oesophagus
  - dysphagia, very rarely odynophagia
- Venous return from the head
  - superior vena cava obstruction (very rare)

Look carefully for a scar – previous hemi/total thyroidectomy

Palpation
Stand behind the patient and gently palpate the gland, located two finger widths below the thyroid cartilage, with one hand on each side and the neck gently flexed.

If a goitre is present, comment on its:

- Size
- Consistency
  - soft
  - firm
  - hard

Note: Soft – ‘like lips’, firm – ‘like the tip of the nose’ and hard ‘like the forehead’ is a good aide memoir to remember (but not repeat in the exam) when thinking about how to measure the consistency of the goitre.

- Diffuse or nodular
  - If nodular – multinodular or a single nodule
- Tender – suggests thyroiditis
- Lymphadenopathy
- Tethered – this suggests cancer.

Percussion
Percuss gently for retro-sternal extension.

Auscultate
Bruit – classically occurs in Graves’ thyrotoxicosis.

Examination of thyroid eye disease

This may not be necessary in the exam unless the patient is complaining of eye symptoms or if on inspection the patient has obvious thyroid eye disease.

Note that lid lag and lid retraction are signs of hyperthyroidism rather than Graves’ disease, although the two may clearly co-exist.

Lid retraction
Indicated by visible sclera above the superior limbus of the cornea

This results from sympathetic stimulation of levator palpabrae superioris of any aetiology, eg thyrotoxicosis, anxiety, β-agonists.

Lid lag
Ask the patient to follow the slow downward movement of your finger at a distance of about 50 cm. The upper lid lags behind the descending eyeball.
Look for clinical features of Graves’s affecting the eyes

Exophthalmos
Sclera visible below the inferior limbus of the cornea with the patient sitting at the same level as you and looking straight ahead.

This sign only occurs in Graves’ disease (cv), the term is synonymous with proptosis; it can be unilateral, although a retro-orbital tumour should always be excluded.

Other features of Graves’
- Periorbital oedema
- Chemosis
- Conjunctival injection
- Ophthalmoplegia

If Graves’ ophthalmopathy is present on inspection it is necessary to perform a more detailed examination and take further history.
- Are the eyes painful in any way? Are they gritty or dry? Also ask if any part of the subsequent exam causes pain or discomfort.
- Is eyelid closure adequate?
  - With exophthalmos there is a greater volume of eye to be covered with each blink, the frequency of blinking is reduced and the time of each blink is increased.
- Ask patient to follow your finger (and to say if they experience diplopia) as you test all directions of gaze.
  - Limitation of upward gaze is the most common abnormality in Graves’ ophthalmopathy.
  - However, the combination of enlarged ocular muscles +/- subsequent fibrosis may lead to complex ophthalmoplegia that is not explained by either single nerve or muscle disease.
- Ptosis – a very rare occurrence in either Graves’ disease or hyperthyroidism. Its presence should raise the possibility of co-existent myasthenia gravis.
- Acuity (see page 405) – full assessment (with ophthalmoscopy) rather than gross assessment of acuity may be unnecessary, unless there is a high suspicion this has been compromised.
  - Since the most important concern of Graves’ ophthalmopathy is a threat to the sight, it is critical to assess vision in the following fashion.
  - Acuity using a Jaeger chart or Snellen chart (+ pinhole)
  - Colour vision – either using Ishihara plates or a red pin to look for desaturation
  - Fields – compression of the nerve head at the orbital apex can cause constriction of the fields
  - Ophthalmoscopy
    - Is there papilloedema or consecutive optic atrophy?
    - If closure is poor or you have concerns about the cornea you may want an ophthalmologist to perform slit lamp examination to look for corneal scars or ulcers – you should state this to the examiner. One can get a reasonable view with a direct ophthalmoscope, but not good enough to preclude a proper examination.

Examination of a skin lesion/rash

The dermatology cases in the Station 5 ICA require a special mention. Classically, the history associated with the condition may not be long or detailed, and most of the differentiating information is gleaned from examination. Furthermore, the presentation of the examination is essential too. Remember that many dermatological conditions reflect underlying medical conditions that should not be overlooked.

It is important to ensure you are fluent with the common terminology:

Macule  Circumscribed area of erythematous change without elevation
Papule  Solid raised lesion < 1 cm in size
Nodule  Solid raised lesion ≥ 1 cm in size
Plaque  Circumscribed elevated confluence of papules $\geq 1$ cm in size
Pustule Circumscribed area containing pus
Vesicle Circumscribed fluid-filled area $< 1$ cm in size
Bulla Circumscribed fluid-filled area $\geq 1$ cm in size

When examining the patient, you need to expose the patient as much as the situation will allow (which the examiner should guide) and inspect the patient as a whole, considering:

- Is there a rash and what is its distribution?
- Is there any hair growth or loss?
- Are there any features of systemic disease?

And more specifically:

- If there is a rash – is it red or not?
- Is the rash macular, papular, in patches or plaques?
- Are there scales or evidence of excoriation?
- Are there fluid-filled lesions?
- Are these vesicles, pustules or blisters?
- Are there signs of systemic disease giving clues as to the aetiology of the skin lesion, eg a colostomy bag suggesting ulcerative colitis?

On presenting your findings, use the correct terminology and be succinct. If there is a suggestion from the history that the dermatological condition is associated with an underlying condition (eg erythema nodosum and inflammatory bowel disease), then an examination of the relevant system may be necessary.

For example:

‘There is a vesicular rash bilaterally on the extensor aspects of the elbow, with associated evidence of excoriation, on a background of a patient with symptoms consistent with coeliac disease, suggesting that this is dermatitis herpetiformis.’

This gives a clear and precise description of the rash, its location, associated excoriation and links it to the most likely underlying aetiological cause.

**Examination of the eyes**

As with the dermatology ICA cases, the history associated with an ophthalmological condition may be brief, but the examination is crucial. Taking a history and examining simultaneously can be easily achieved. Be cautious, however, since red flag symptoms must not be missed (such as in papilloedema or diabetic retinopathy), as some conditions reflect very serious underlying medical conditions or even malignancy.

This does not mean, however, that all aspects of the eye examination must be completed in the examination. For example:

In a patient with a **painful red eye** you may consider examining:

- Visual acuity – may be reduced
- Fundoscopy – may show anterior uveitis or corneal scarring
- Pupil – may be irregular, small, or unreactive

However, you might expect visual fields and eye movements to be normal and tell the examiner you would include the examination of these as part of your further investigations.

In a patient with a history of **progressive deterioration in vision** you may consider examining:

- Visual acuity
- Fundoscopy – retinitis pigmentosa, diabetic retinopathy
- Eye movements
- Visual fields – peripheral loss first

It is important to **tailor** your examination according to the presenting complaint.

However, it is still important to know how to examine the eye in a systematic way. Fundoscopy is a core skill for a practising clinician and MRCP candidate, but one with which many have considerable difficulty.
**General appearance of the eye**
Look carefully for symmetry of the eyelids, pupils and general eye movements. In particular, check for signs such as ptosis, lid retraction or irregular pupils.

Look for clues suggesting decreased visual acuity, eg white stick, adjacent Braille books, or for evidence of other systemic diseases such as diabetes, eg glucose testing sticks, diabetic drinks, foot ulcers or evidence of co-existent dialysis.

**Visual acuity**
Assess acuity with the patient wearing their normal glasses.

Ask the patient if they can see from each eye, and then assess individually by use of a Snellen chart. Visual acuity is defined as \( V = \frac{d}{D} \) where \( d \) = distance at which numbers are read and \( D \) = distance at which they should be read.

**Pupillary reflexes**
Each eye should be tested in turn for direct and consensual pupillary reflexes with a pen torch while the patient faces forward with eyes focused directly ahead.

**Visual fields**
Visual fields should be assessed by confrontation with you sitting approximately 1 metre from the patient and with your eyes at the same level. When assessing the patient's right eye, ask them to cover up their left eye, and after covering your right eye, slowly bring an object in from the periphery in a plane equal between you and the patient.

What should the object be? A moving finger is commonly used and may be fine for a screening test if examining the cranial nerves. The top of a pen, placed in the quadrants in turn, is better, but do not bring it inwards too quickly, as the patient must have the time to say when they first see it.

A red pin can be used for assessing central colour vision that is located around the macula.

Where it is first visible gives a good indication of peripheral vision.

**Eye movements (see p 225)**

**Fundoscopy**
This should be performed in a dark room – if it is not dark ask to turn the lights off.

A mydriatic (dilating the pupil) such as tropicamide may have been used. It is essential that you examine the patient’s right eye with your right eye and their left eye with your left eye. The latter is a routine many candidates have difficulty with, but failure to do this, for whatever reason, is frowned upon.

Find the red reflex, keep it in view and get in close!

Although the temptation and tendency is to go straight to the retina, you should get into the routine of gradually racking down through the lens strengths, examining first the front of the eye and gradually, by reducing the strength of the lens, moving to the retina, which is usually observed with a zero or a slightly negative lens, depending on whether or not the patient is myopic.

The disc and each quadrant of the retina should be identified and studied in turn.

**Disc** – note its shape, colour (pale in optic atrophy), margins and if there is papilloedema.

**Retinal blood vessels** – examine the vessels, noting their diameter (arteries are narrower than veins) and the point at which they cross. Is there A–V nipping?

Look for arterial emboli (arteries become much thinner or thread like) or venous thrombosis (veins become engorged with surrounding haemorrhages).
Each retinal quadrant should be examined for the presence of:

- Haemorrhages
  - dot, blot or flame shaped
- Microaneurysms
- Pigmentation
  - Exudates, distinguish between:
    - hard – white/yellow and shiny with well-defined edges
    - soft (‘cotton wool spots’)
- The presence of new vessels should be identified and any previous photocoagulation scars.

**The macula** – It should be specifically examined as the surrounding area is, as the name suggests, the site of diabetic maculopathy.

Examine the peripheries of the retina, looking particularly for evidence of retinitis pigmentosa.

Considering the cases you might be asked to see; look around the bedside for clues.

**FINAL THOUGHTS**

Remember that Station 5 is very different from the other stations. You are somewhat liberated from the regimented examination routines of the other stations. However, you need to gather a great deal of information and communicate a plan to the patient in a very short space of time. It is perhaps the most accurate and global reflection of how you will perform as a registrar in clinic or on-call. This should not be intimidating – if in doubt just resort to what you would normally do in exactly that scenario at work!
Integrated Clinical Assessment

1. Acromegaly
2. Addison's disease
3. Ankylosing spondylitis
4. Anterior uveitis
5. Asthma
6. Atrial fibrillation
7. Coeliac disease
8. Congestive cardiac failure
9. Cranial nerve III palsy
10. Crohn's disease/bloody diarrhoea
11. Cushing's disease
12. Dermatomyositis
13. Diabetic retinopathy
14. Eczema
15. Erythema nodosum
16. Gout
17. Graves' disease
18. Henoch–Schönlein purpura
19. Hereditary haemorrhagic telangiectasia
20. Herpes zoster
21. Horner syndrome
22. Hypothyroidism
23. Impetigo
24. Marfan syndrome
25. Osteogenesis imperfecta
26. Paget's disease
27. Papilloedema
28. Pemphigus
29. Pleural effusion
30. Psoriasis
31. Psoriatic arthropathy
32. Pyoderma gangrenosum
33. Retinitis pigmentosa
34. Rheumatoid arthritis
35. Scleroderma
36. Systemic lupus erythematosus
37. Tremor
38. Turner syndrome
39. Vitiligo
**SCENARIO 1. ACROMEGALY**

**Introduce yourself**

Confirm the information you’ve been given and patient details: this 36-year-old man has presented with headaches. Please assess him.

‘What is the main problem from your point of view?’ ‘For the last 3 months I have been having headaches, and I am worried about my vision as I have recently crashed my car into a parked car when driving along the road. A friend has commented that my appearance has changed.’

**New condition**

**Focused history** of the presenting complaint and relevant systems enquiry. This should include screening for endocrine disturbance (any aspect of the hypothalamic–pituitary axis can be disrupted) and for localising neurology (eg diplopia, visual field defect, carpal tunnel syndrome)

**Identify alarm symptoms**: sudden headache, diplopia, ptosis (may imply pituitary apoplexy). Polyuria/polydipsia may imply either diabetes insipidus or diabetes mellitus

**Pre-existing conditions**: hypertension, diabetes, colonic polyps, sleep apnoea

**Social/occupational/family circumstances**: especially if driving professionally (eg taxi driver, group 1 driver)

**Drug history** – antihypertensives, oral hypoglycaemics

**DRUG ALLERGIES**

**Other medical conditions**: any co-morbidity that might preclude hypophysectomy (eg bleeding diathesis)?

**Begin a targeted** examination of relevant area/system and continue while taking the history

**General appearance (see Figure 5.1)**

- Prominent supraorbital ridges, nose and lips
- Pronounced jawline (prognathism)
- Macroglossia and increased interdental separation
- Enlarged sweaty hands with thickened skin

In addition

- Examine visual fields for bitemporal hemianopia. There may be optic atrophy if the adenoma extends into the parasellar region
- Assess for cranial nerves palsies, especially nerves III, IV and VI (if raised intracranial pressure)
- Examine for carpal tunnel syndrome/scars from previous decompression
- Myopathy (especially proximal) and arthropathy is common, but no synovitis will be found
- Examine for associated endocrine features, including goitre, gynaecomastia and/or hirsutism
- Perform a cardiac and abdominal examination if has symptoms, looking for associated cardiomegaly and hepatomegaly
- Measure blood pressure, assess for glycosuria (may reflect associated impaired glucose tolerance/diabetes mellitus)

**Any old photographs of the patient available?**

**CALLBACK**: ‘You said at the beginning that the main problem was headaches, change in vision and appearance. Is there anything else troubling you that we haven’t covered?’

**RESPOND TO PATIENT’S CONCERNS (1–2 min)**

1. I think that your headaches, change in vision and appearance are due to a condition resulting from acromegaly, which is caused by a benign growth in the pituitary gland that produces too much growth hormone
2. For your headaches, I can give you some painkillers as a temporary measure. However, it is very important to establish the diagnosis by doing some urgent investigations. As your vision is impaired, I am afraid that you will have to stop driving immediately because of the risk to yourself and other road users
3. I am going to organise some urgent blood tests (glucose tolerance tests, IGF-1, other pituitary hormones). We will need to arrange an urgent MRI of your pituitary gland and ask the eye specialists to see you to assess to what extent this growth has affected your vision

Thank the patient
Acromegalic facies

Increased interdental spacing

Figure 5.1 Acromegaly
<table>
<thead>
<tr>
<th>Differential diagnosis</th>
<th>Clinical communication skills and managing patient concerns</th>
<th>Physical examination and identifying physical signs</th>
<th>Clinical judgement and maintaining patient welfare</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Craniopharyngioma</strong></td>
<td>Symptoms of raised intracranial pressure (headache, nausea and vomiting), visual field defects and endocrine deficiencies (e.g., pubertal delay from gonadotrophin deficiency)</td>
<td>Visual defects: In contrast to other pituitary adenomas, this starts by causing a bitemporal inferior quadrantanopia, because the suprasellar lesion initially compresses the optic chiasma from above. Over time, this may progress to bitemporal hemianopia</td>
<td>Diagnosed by characteristic appearances on pituitary imaging. Transcranial frontal surgery is the major modality of treatment, either with or without radiotherapy. A large proportion of patients will require hormone replacement</td>
</tr>
<tr>
<td><strong>Pituitary adenoma</strong></td>
<td>Symptoms of mass effects, visual field defects and endocrine disorders</td>
<td>Mass effects: May first present with headache, nausea and vomiting. Visual defects: Initially bitemporal superior quadrantanopia, as outlined above. Endocrine disorders: In hormone-secreting adenomas, clinical features reflect the particular hormone being secreted excessively (e.g., Cushing's disease from a corticotroph adenoma). As tumours expand, they may disrupt other hormone axes and cause symptoms and signs of hormone deficiency.</td>
<td>Tumours are often diagnosed on pituitary imaging, with MRI being the preferred modality. Assessment of pituitary function to assess for hormone hypersecretion and/or any associated hormone deficiencies. Visual field testing should be performed at the start of treatment and thereafter to assess efficacy of treatment. Transphenoidal surgery, either with or without radiotherapy. Careful ongoing pituitary function assessment will be needed to assess if long-term hormone replacement is indicated</td>
</tr>
<tr>
<td><strong>Pseudoacromegaly</strong></td>
<td>Symptoms very similar to true acromegaly as described below. No visual field defects</td>
<td>Clinical phenotype similar to acromegaly without visual field defects. Disturbance of other hormone axes rare</td>
<td>Investigation focuses on exclusion of growth hormone-secreting adenoma</td>
</tr>
</tbody>
</table>
Clinical judgement and maintaining patient welfare

Acromegaly is a clinical syndrome resulting from the hypersecretion of growth hormone (GH). Most cases are caused by GH-secreting adenomas situated in the anterior pituitary. Approximately three-quarters of these are macroadenomas (ie >10 mm diameter) at the time of diagnosis. These adenomas account for approximately a third of all hormonally secreting pituitary adenomas. Rare causes of acromegaly include growth hormone-releasing hormone (GHRH) secretion from hypothalamic tumours, or ectopic GHRH or GH secretion (eg from neuroendocrine tumours).

Adenomas tend to grow very slowly; it is estimated that there is an average delay of more than 10 years between initial symptoms and diagnosis. The most consistent early symptoms (and those most likely to form the stem of a Station 5 question on the condition) include headaches and visual disturbance. Other relatively early features are those of endocrine disturbance, particular gonadotrophin deficiency, ie oligo-/amenorrhoea in women, or erectile dysfunction, loss of libido and testicular atrophy in men. As the condition progresses, a large number of organ systems may be affected, and a number of physical signs may develop, as described in the differential diagnosis table. There may also be signs of treatment of the condition (eg scars around the nose reflecting previous transsphenoidal surgery).

Acromegaly incurs a significantly increased mortality rate (estimated as at least twice as high) as the healthy population, with those with the highest GH levels at the greatest risk of premature death. This is mostly attributable to increased cardiovascular disease.

Investigations

1. Establish the presence of GH excess
   - Random GH plasma levels in themselves are not diagnostic, because these fluctuate greatly between individuals and at different times of the day. However, serum insulin-like growth factor 1 (IGF-1) levels are elevated in almost all patients with acromegaly.
   - The most effective means of diagnosing GH excess is with an oral glucose tolerance test; although GH levels suppress to very low levels after a glucose load in healthy individuals, GH levels will fail to suppress or paradoxically rise in patients with acromegaly.

2. Establish the aetiology (Table 5.1)
   - Pituitary imaging should be performed; MRI is the most sensitive modality.
   - Establish whether a pituitary adenoma is functional.

3. Look for complications
   - Visual perimetry: to establish visual fields.
   - ECG, echocardiogram and chest radiograph: if cardiomyopathy is suspected.

Table 5.1 Differentiation between anterior and posterior pituitary causes

<table>
<thead>
<tr>
<th>Anterior pituitary</th>
<th>Posterior pituitary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corticotroph: 9 am cortisol. If &lt;500 nmol/L, perform a dynamic test, ie insulin or glucose tolerance test</td>
<td>ADH secretion – plasma and urinary sodium and osmolality. A water deprivation test may also be required</td>
</tr>
<tr>
<td>Thyrotroph: T₄, T₃ and TSH</td>
<td></td>
</tr>
<tr>
<td>Gonadotroph: LH, FSH, testosterone and SHBG or estradiol</td>
<td></td>
</tr>
<tr>
<td>Lactotroph: prolactin</td>
<td></td>
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</tbody>
</table>
• Oral glucose tolerance test: used to make the diagnosis, but also to establish the presence of associated impaired glucose tolerance/diabetes mellitus.
• Nerve conduction studies: if an associated carpal tunnel syndrome is suspected.
• Sleep studies: if obstructive sleep apnoea is suspected.
• Colonoscopy: surveillance for polyps and colorectal malignancies, a recognised complication of acromegaly.
• Bone profile and parathyroid hormone: the combination of acromegaly and hyperparathyroidism raises the possibility of multiple endocrine neoplasia type 1 (MEN-1) syndrome; there should be a low threshold to investigate for a possible pancreatic tumour.

Treatment

Surgical
• Transsphenoidal hypophysectomy is successful in up to 90% of patients with microadenomas and 50% of those with macroadenomas in the hands of the most experienced neurosurgeons.
• Long-term deficiency in one or more of the pituitary hormones will develop in approximately 70% of patients.

Radiotherapy
• Pituitary external irradiation is indicated for patients unfit for or not cured by surgery. It results in a 50% decline in GH levels by 2 years, but with a continuing exponential decline thereafter.
• There is a significant risk of late hypopituitarism, with other complications including cranial neuropathies and visual field impairment.

Medical
• Somatostatin analogues (eg octreotide or lanreotide) are administered subcutaneously. They reduce serum GH in 90% of patients, and to a level of GH that will prevent adenoma growth in approximately 50% of patients. Side effects are nausea and gallstones (approximately 50% by 5 years).
• Dopamine agonists (eg bromocriptine, cabergoline) are effective in only 10% of patients.
• Pegvisomant is a GH receptor antagonist, administered subcutaneously daily, which normalises serum IGF-1 in >90% of patients, helping to reduce many of the clinical features. However, it does not reduce GH secretion or constrain tumour growth, so it is currently reserved for patients with ongoing disease despite surgery, radiotherapy and other medical therapies.

Maintaining patient welfare
It is important in managing acromegalic patients to think not only about the immediate treatment for the condition, but also about the means of minimising long-term complications. Candidates can show that they are aware of these by suggesting colonoscopic screening for colonic malignancy (this is currently recommended every 3 years), and minimising the patient’s risk of cardiovascular disease by optimising their blood pressure, lipid profile and smoking cessation advice.