CNS – SPEECH & HIGHER CORTICAL FUNCTIONS

STEPS OF EXAMINATION

Step 1: Approach the patient

- Read the instructions carefully for clues
- Shake hands, introduce yourself
- Ask permission to examine him "I am just going to ask you some questions; is that alright?"

Step 2: General inspection:

- **Bedside:** walking stick, shoes-callipers, built-up heels
- **General appearance:** scan the patient quickly looking for:
  - Nutritional status (under/average built or overweight)
  - Abnormal movement or posture (rest or intention tremors, dystonia, choreoathetosis, hemiballismus, Myoclonic jerks, tics, pyramidal posture)
  - Abnormal facial movements (hemifacial spasm, facial myokymia, blepharospasm, oro-facial dyskinesia)
  - Facial asymmetry (hemiplegia)
  - Nystagmus (cerebellar syndrome)
  - Facial wasting (muscular dystrophy)
  - Sad, immobile, unblinking facies (Parkinson's disease)
  - Ptosis, lack of facial expression, slack mouth (myasthenia gravis)
  - Myxoedematous facies (variegated dysarthria)
  - Horner's syndrome (syringomyelia, Pancoast's syndrome)
  - Peroneal wasting (Charcot-Marie-Tooth disease)
  - Pes cavus (Friedreich's ataxia, Charcot-Marie-Tooth disease)
- **Face (mouth)**:
  - Ask the patient to open his mouth: shine your pen torch into the opened mouth; look for poorly-fitting or absent denture, painful ulcer
  - Look at the tongue as it lies in the floor of the mouth for wasting or fasciculation
  - Ask the patient to protrude his tongue out, then to move it from side to side (inspect the posterolateral edge of the tongue)
- **Hands**: tell the patient and demonstrate "outstretch your hands like this (palms facing downwards)"… then "like this (palms facing upwards)"
  - Check for wasted hands (MND, Charcot-Marie-Tooth disease, syringomyelia)
  - Feel the radial pulse (AF → thromboembolism)

Step 3: Speech assessment: ask the patient some general questions to get him talking: "Please could you tell me your name? Your age? Are you right handed or left handed? What is your first language? Where do you live?" If he does not appear to understand, repeat louder. Note the volume of sound, rhythm of speech and clarity of enunciation. You should be able to quickly distinguish between dysphonia (disorders of phonation), dysarthria (disorders of articulation), and dysphasia (disorders of Speech content)

Step 4: Dysphonia: the patient is able to give his name and address but is unable to produce normal volume of sound, or speaks in a whisper (indicates impairment of voice production from the larynx). Ask the patient to:

- "Cough". Listen to the quality of the cough:
  - A bovine cough (cough lacks explosive start) → vocal cord palsy (innervated by the recurrent laryngeal from the vagus)
  - Dysphonia with normal cough → local laryngeal cause e.g. laryngitis (most commonly due to common cold) or hypothyroidism (thickening of the vocal cords from amyloid deposits), or psychological disturbance (hysteria)
- "Say a sustained 'eeeeee'". If the note cannot be sustained and fatigues, consider myasthenia

Step 5: Dysarthria: the patient is able to give his name and address but the words are not formed properly (indicates inability to articulate properly because of local lesion in the mouth or disorder of speech muscles or their connections).

- **First ask about** any poorly-fitting or absent denture, or painful mouth
- **Then ask the patient to repeat difficult phrases (tongue twisters):** e.g. "rhinoceros", "Bri**t**ish cons**tit**u**t**ion", "Baby hi**pp**o**p**otamus", "egg", "rub", "ye**ll**ow **l**orry". Listen carefully for the rhythm of the speech, clarity of enunciation (slurred words) and which sounds cause the greatest difficulty. You must be able to recognize the various types of dysarthria and look for **additional signs** to support your diagnosis (see theoretical notes)

Step 6: Dysphasia: If the patient has no dysarthria or dysphonia but the speech is meaningless (receptive aphasia) or non fluent (expressive aphasia), assess the following:

- **Comprehension (understanding):** ask the patient (without gesturing) to perform a few simple commands, e.g. "please put your tongue out, shut your eyes, touch your left ear with your right hand".
- **Speech fluency**: test the patient's ability to form sentence by asking the patient to describe something in more detail, e.g. "what is your job? What you actually do when at work? How you would get home from here?" Listen carefully for fluency of speech, word-finding pauses, jargon speech, paraphasias, neologisms
- **Naming**: hold up your keys and ask the patient "what is this?" If no answer, then ask, "Is this a spoon? Is it a pen? Is it keys?"
- **Repetition**: ask the patient to repeat simple sentence, e.g. "today is Tuesday"
- **Orofacial dyspraxia**: if there are expressive problem, check to see if the problem is true expressive dysphasia or whether there is orofacial dyspraxia (difficulty in volitional movements of the lip and tongue → hesitancy in word production). Ask the patient (without gestures) to perform various orofacial movements (provided that there is no receptive dysphasia), e.g. "please show me you teeth. Put out your tongue and move it from side to side" subsequently ask the patient to obey the same commands but with gesture, i.e. so the patient can mimic. This should distinguish between **ideational** dyspraxia and **ideomotor** dyspraxia, so that the type of speech therapy is tailored accordingly
- **Ask if you may test reading** (ask the patient to read a sentence and to obey a written command) **and writing** (ask the patient to write a sentence): impaired reading (dyslexia) and impaired writing (dysgraphia) indicate extension of damage to parieto-occipital region, and may be part of Gerstmann's syndrome. Now you must be able to recognize the various types of dysphasia and look for **additional signs** to support your diagnosis (see theoretical notes)

Step 7: Higher mental functions: use the following "abbreviated mental test" (AMT) to test attention and concentration, orientation, memory, general knowledge and intelligence. Each item scores 1 point (8-10 → Normal, 4-7 → mild-moderate dementia, less than 4 → moderate-severe dementia)

- Age "How old are you?"
- Address "Where do you live (town or road)?"
- Address for recall at end of test "I'd like to test your memory. Please say this address '42 west street'. Now remember it and repeat it when asked". Then at end of test ask ""Can you name the address I named before?"
- Time (the nearest hour) "What time is it?"
- Year "What year is it?"
- Place "Where are we now?"
- Recognition of two persons" Who am I? Who is this (doctor, nurse, etc.)?"
- Name of national leader "Who is our president?"
- Year of World War 2 "When did the second world war start?"
- Count backwards from 20 to 1 "Begin with 20 and count backwards till one"

Step 8: Thank the patient and cover him (her)

THEORETICAL NOTES

Process and abnormalities of speech

Process	abnormality
Hearing	Deafness
Understanding (comprehension)	Dysphasia
Thought and word finding (expression)	Dysphasia
Voice production	Dysphonia
Articulation	Dysarthria

Types of dysarthria:

1. Ataxic (cerebellar) dysarthria	
Features: slurred, slow, jerky and explosive (lalling, staccato, scanning) speech, with irregular breaks in articulation and equal emphasis on each syllable, thus "rhinoceros" is pronounced "rhi-noc-er-os". There may be inspiratory whoops indicating the lack of coordination between respiration and phonation.	**Causes:** cerebellar lesions **Additional signs:** other cerebellar signs, e.g. nystagmus, impaired finger-nose test with past pointing and intention tremor, impaired heel-shin test, dysdiadochokinesia, ataxic gait with tendency to fall to side of the lesion

2. Spastic (UMN) dysarthria	
Features: laboured speech, patient hardly opens mouth as if trying to speak from the back of the mouth, slurred, monotonus, high-pitched, 'hot potato', Donald Duck' speech due to a tight, immobile tongue. The precision of consonant pronunciation is lost, thus "British constitution" becomes "Brizh conshishushon" as the crispness of the 'ta' sound depends on adequate power in the tip of the tongue.	**Causes:** bilateral UMNL of the corticobulbar tract (pseudobulbar palsy) due to bilateral CVA of the internal capsule and less commonly due to MS, MND, high brainstem tumours, head injury **Additional signs:** other signs of pseudobulbar palsy, e.g. emotional lability, cannot protrude his tongue which lies on the floor of the mouth and is small and tight, dysphagia (particular to liquids, including saliva → dribbling from the mouth), nasal regurgitation, absent palatal movement, positive jaw jerk (bilateral V lesion), evidence of associated bilateral corticospinal tract (pyramidal) lesions, e.g. bilateral spasticity and extensor plantars

3. Flaccid (LMN) dysarthria	
Bulbar palsy Features: nasal twang, indistinct, decreased modulation, slurring of labial and lingual consonants.	**Causes:** disease affecting bulbar cranial nerves, e.g. MND, syringobulbia, Guillan-Barre syndrome, poliomyelitis, subacute meningitis, neurosyphilis **Additional signs:** other signs of bulbar palsy: • **IX, X** lesion, e.g. dysphagia (particular to liquids, including saliva → dribbling from the mouth), nasal regurgitation, absent palatal movement, palatal air escape may be audible during phonation, • **XII** lesions (tongue is wasted, wrinkled, flaccid and fasciculating)
Paralysis of VII (facial palsy) Features: distorted speech especially labial consonants (B, M, P) – test with phrase "baby hippopotamus"	**Additional signs: VII** lesion (unilateral facial droop, absent nasolabial fold and forehead creases)
Paralysis of IX, X (palatal dysarthria) Features: nasal speech as with a bad cold. The patient is unable to pronounce words that require complete closure of the nasopharynx. Thus "egg" is sounded as "eng" and "rub" becomes "rum".	**Causes:** vagal damage at or above the level of the pharyngeal branches. **Additional signs:** other signs of **IX, X** lesion, e.g. dysphagia (particular to liquids, including saliva → dribbling from the mouth), nasal regurgitation, absent palatal movement, palatal air escape may be audible during phonation
Paralysis of XII (tongue palsy) Features: distorted speech especially lingual consonants (L, R) – test with phrase "yellow lorry"	**Additional signs:** signs of **XII** lesions (tongue is wasted, wrinkled, flaccid and fasciculating)

4. Hypokinetic (extrapyramidal) dysarthria	
Features: monotonus without accents or emphasis, low pitched somewhat slurred speech.	**Causes:** Parkinson's disease **Additional signs:** other extrapyramidal signs, e.g. expressionless unblinking face, drooling (due to excessive salivation and some dysphagia), pill rolling tremor, blepharo-clonus (tremor of the eyelids when the eyes are gently closed), titubation, cogwheel rigidity at the wrist and lead-pipe rigidity at the elbow, bradykinesia (demonstrated by asking the patient to touch his thumb successively with each finger), glabellar tap sign, Parkinsonian gait (stooped posture, reduced arm swing, delayed initiation of walking, short shuffling steps, occasional festinant gait)

5. Hyperkinetic (choreiform/dystonic) dysarthria	
Features: Variable speaking rate (slower in dystonia), improper stress, inappropriate silences. This gives the speech a bursting quality.	**Causes:** Choreiform disorders (Huntington's disease), myoclonic disorders, dystonia musculorum deformans **Additional signs:** other signs of Huntington's in choreiform dysarthria (chorea, dementia)

6. Myopathic/Myasthenic dysarthria	
Features: weak hoarse voice with a nasal quality, soft accents. Muscle fatigability in myasthenia gravis (demonstrated by making the patient count).	**Causes:** myopathy, myositis, myasthenia gravis **Additional signs:** other signs of myasthenia gravis in myasthenic dysarthria**:** • Ptosis (accentuated by upward gaze) • Variable strabismus (with diplopia) • Facial and proximal muscle weakness all of which worsen with repetition (lack of facial expression, mouth is slack, snarls when tries to smile, cannot whistle, demonstrate proximal muscle weakness and fatigability in the upper limbs)

7. Myotonic dysarthria	
Features: slurred, suppressed speech due to myotonia of tongue and pharynx.	**Causes:** myotonic dystrophy **Additional signs:** other sings of myotonic dystrophy, e.g. myopathic facies (long, thin and expressionless), wasting of facial muscles, sternocleidomastoid, shoulder girdle and quadriceps, bilateral ptosis, frontal baldness, grip myotonia, percussion myotonia, difficulty opening his eye after firm closure, dysphagia, wasting and weakness of the distal muscles (forearm and legs) with areflexia

8. Variegated dysarthria	
Hypothyroidism Features: low pitched, catarrhal, hoarse, croaking, guttural voice as if the tongue is too large for the mouth	**Additional signs:** other signs of hypothyroidism, e.g. myxoedematous facies (thickened and coarse facial features, periorbital puffiness and pallor), skin is rough, dry , cold and inelastic, "peaches and cream" complexion (anaemia and carotenaemia), hair is thin, dry and brittle with loss of outer third of eyebrows, xanthelasma, goitre or thyroidectomy scar, generalized non pitting swelling of subcutaneous tissue, bradycardia, slow relaxing ankle jerks
Amyloidosis Features: rolling and hollow, hardly modulated speech due to large tongue	
Multiple ulcers or thrush in the mouth Features: some parts of the speech indistinct	
Parotitis or temporomandibular arthritis Features: monotonous, suppressed, badly modulated	
Generalized paresis of the insane Features: slurred, hesitant or feeble voice	**Additional signs:** other signs of GPI, e.g. dementia, vacant expression, tremor of the hands, lips and tongue (trombone tremor – the tongue darts in and out of the mouth involuntarily), spastic paraparesis, brisk reflexes, extensor plantars.

Model of speech understanding and output

- Recognition of the sounds as language in **Wernicke's area**
- Understanding of the meaning of the words in the **concept area** (temporo-parietal association areas)
- Generation of the speech output in **Broca's area**
- Wernicke's area is also connected directly to Broca's area by the **arcuate fasciculus**
- All these areas are in the dominant hemisphere. The left hemisphere is dominant in all right-handed patients and in 50% of left-handed patients

Types of dysphasia:

1. Wernicke's (receptive) (sensory) (fluent) (Jargon) dysphasia	
Features: • **Impaired comprehension** • **Fluent (jargon) speech:** the patient replies fluently (often rapidly), but often unintelligible. He puts words together in the wrong order and mixes them with paraphasias (syllabic substitution with some phonemic relationship to the original word, e.g. "toothspooth" for "toothbrush" or word substitution with a semantic relationship to the correct word, e.g. hand" for "foot") and neologisms (syllabic or word substitution with no relationship to the original syllable/word) • **Impaired repetition, naming, reading, and writing**. Attempts to repeat result in paraphasic distortions and irrelevant insertions. • Loss of insight into his dysphasia • The prognosis for recovery is poor	**Lesion** of Wernicke's area**:** posterior part of superior temporal gyrus plus adjacent parts of parietal and occipital cortex of dominant hemisphere **Causes:** embolus to the inferior division of the middle cerebral **Additional signs:** right homonymous superior quadrantanopia, right cortical sensory loss

2. Transcortical receptive (sensory) (fluent) dysphasia	
Features: similar to Wernicke's aphasia but with **preserved repetition**	**Lesion:** Disruption of the connection between Wernicke's area and concept area (posterior parieto-occipital region) **Causes:** Infarctions in the posterior watershed zone or neoplasms that involve the temporo-parietal cortex posterior to Wernicke's area

3. Broca's (expressive) (motor) (non-fluent) (telegraphic) dysphasia	
• **Features:** preserved comprehension • **Non-fluent speech:** the content is good but the patient says little**.** Speech is interrupted by many word-finding pauses and lacks the filler words (to, but, so, the…). For example, "phone daughter evening" • **Impaired repetition and naming and writing** (reading is comparatively preserved) • preserved Insight into his condition (so patient could be tearful and depressed) • The prognosis for eventual adaptation of the patient to his disability is good	**Lesion** of Broca's area: inferior frontal gyrus of dominant hemisphere **Causes:** occlusion of the superior division of the middle cerebral artery **Additional signs:** right hemiparesis**,** right facial weakness, orofacial ideomotor dyspraxia

4. Transcortical expressive (motor) dysphasia	
Features: similar to Broca's aphasia, but with **preserved repetition**	**Lesion:** disruption of the connection between the concept area and Broca's area **Causes:** • Lesion of anterior watershed zone between anterior and middle cerebral artery territories • Lesion of supplementary motor cortex in the territory of the anterior cerebral artery

5. Global dysphasia	
Features: • **Impaired comprehension.** • **Non-fluent speech**. • **Impaired repetition, naming, reading, and writing**.	**Lesion:** Combined dysfunction of Broca's and Wernicke's areas **Causes:** strokes that involve the entire left middle cerebral artery. **additional signs:** right hemiplegia, hemisensory loss, homonymous hemianopia and general intellectual deterioration

6. Conductive dysphasia	
Features: • Preserved comprehension • Fluent speech output, but **paraphasic** • **Impaired repetition**, **naming and writing** (reading loudly may be similarly compromised; while reading silently for comprehension is intact)	**Lesion** of the arcuate fasciculus in the dominant hemisphere **Causes:** occlusion of a portion of the angular branch of the middle cerebral artery **Additional signs:** vary according to the primary lesion site.

7. Nominal (anomic) dysphasia	
Features: • Preserved comprehension. • Fluent speech output, but paraphasic • preserved repetition • **Impaired naming and word finding** (excessive word-finding pauses)	**Lesion:** • Nominal aphasia is found in both motor and receptive dysphasia • Lesions of angular gyrus may produce a syndrome in which nominal dysphasia is associated with dyslexia plus Gerstmann's syndrome (dysgraphia, right-left disorientation, acalculia, and finger agnosia) • Nominal aphasia is also seen in degenerative disorders such as Alzheimer's disease.

N.B. Dyspraxia:

- Inability to perform purposeful volitional familiar movements in the absence of motor weakness, sensory deficit or severe incoordination.
- The defect is often in the dominant parietal lobe, with disruption of connections to the motor cortex (frontal lobe) and to the opposite hemisphere (corpus callosum).
- It is classified into
  - **ideational** dyspraxia: the patient is unable to perform movement though understanding the command (due to loss of the concept of performing the movement)
  - **ideomotor** apraxia: patient is unable to perform movement on command (or do it in odd or bizarre fashion) , although he may do this automatically or when asked to mimic (as the concept is present but the motor programme for the usage is not accessible)
- Common types are dressing, gait, limb, truncal, constructional and orofacial dyspraxia

N.B. apraxia and agnosia are complex disorders and should not usually form part of the examination – RCP UK statement