EANS/UEMS European examination in neurosurgery

Part I (written)

Variants of questions with answers (compilation - Vyacheslav S. Botev, Department of Neurosurgery, M.Gorky Donetsk National Medical University)

Neuroophthalmology and otoneurology

1. Which of the following is shown on this fundoscopy?



- A. Papilledema
- B. Optic neuritis
- C. Optic atrophy
- D. Normal fundus
- E. Retinal artery occlusion

2. This woman had an uncomplicated pregnancy but developed severe headache 1 week after delivery. What can you see on fundoscopy?



- A.PapilledemaB.Optic neuritisC.Acute retinal necrosisD.Central retinal vein occlusionE.Central retinal artery occlusion
- **3**. Papilledema is seen in: (True or False):



- A. Constrictive pericarditis
- B. Cavernous sinus thrombosis
- C. Carbon monoxide poisoning
- D. Cor pulmonale with hypoxia
- E. Benign intracranial hypertension

4. The man to whom this fundus belongs has always had difficulty finding shoes to fit, and is wheelchair-bound by the age of 25 years. There is a no family history. What would you expect to find on examination and investigation?



A.Spastic limbsB.Optic atrophyC.Abnormal ECGD.Brisk tendon reflexesE.Peripheral neuropathy

5. Which of the following is shown on this fundoscopy?



- A. Papilledema
- B. Optic neuritis
- C. Optic atrophy
- D. Acute retinal necrosis
- E. Retinal artery occlusion

6. This man has had an episode of left arm weakness in the past. What is the diagnosis?



A.Optic neuritisB.Optic atrophyC.Angioid streaksD.Friedreich's ataxiaE. Retinal artery occlusion

7. Fundus photograph of the 70-year-old man with acute painless visual loss in his left eye three days previously is shown below. What is the diagnosis?



- A. Papilledema
- B. Optic neuritis
- C. Optic atrophy
- D. Normal fundus
- E. Retinal artery occlusion

8. A patient presents with the visual field illustrated below. Where is the lesion located?



- A. Optic chiasm B. Optic radiation
- C. Left occipital lobe
- D. Left temporal lobe
- E. Right occipital lobe

9. A 50-year-old man presents with the incongruous left homonymous hemianopsia. Which of the following structures is damaged?



- A. Optic chiasm
- B. Optic radiation
- C. Left optic tract
- D. Right optic tract
- E. Left occipital lobe

10. What type of hemianopsia is shown below?



- A. Binasal
- B. Bitemporal
- C. Homonymous, total
- D. Homonymous, partial
- E. Homonymous, contralateral

11. A patient has a bilateral, right-sided superior field defect. Where do you suspect the lesion is located?



- A. Optic chiasm
- B. Optic radiation
- C. Left occipital lobe
- D. Left temporal lobe
- E. Right occipital lobe

12. A 60-year-old retired bus driver has a history of hypertension. He complains of visual loss. Assessment shows the presence of a right inferior homonymous hemianopsia. Which of the following structures is damaged?



- A. Optic chiasm
- B. Optic radiation
- C. Left parietal lobe
- D. Left temporal lobe
- E. Right occipital lobe

13. A 50-year-old man presents with the left superior homonymous hemianopsia. Which of the following structures is damaged?



- A. Optic chiasm
- B. Optic radiation
- C. Left occipital lobe
- D. Right occipital lobe
- E. Right temporal lobe

14. A patient presents with the visual field illustrated below. Where is the lesion located?



- A. Optic chiasm
- B. Optic radiation
- C. Left occipital lobe
- D. Right parietal lobe
- E. Left temporal lobe

15. This is an audiogram of a left-handed gamekeeper who complained of hearing loss. There is no air–bone gap. What is the likely etiology?



- A. Ear wax
- B. Ménière's disease
- C. Age-related hearing loss
- D. Noise-induced hearing loss
- E. Cerebellopontine angle tumor

16. A 40-year-old air traffic controller complains of fluctuating tinnitus, vertigo and a feeling of fullness in his right ear. There is no air–bone gap. What is the likely diagnosis?



- A. Ear wax
- B. Ménière's disease
- C. Age-related hearing loss
- D. Noise-induced hearing loss
- E. Cerebellopontine angle tumor

17. Which of the following would be expected in a patient with congenital Horner's syndrome?



- A. Diplopia
- B. Iris rubeosis
- C. Darker colored iris in the affected eye
- D. Lighter colored iris in the affected eye
- E. Vermiform motion of the iris on pupillary constriction

Answers

1. D. Normal fundus.



Normal fundus of the right eye.

The disc has sharp margins and is normal in color, with a small central cup. Arterioles and venules have normal color, sheen, and course. Background is in normal color. The macula is enclosed by arching temporal vessels. The fovea is located by a central pit.

2. A. Papilledema.

As the pregnancy was uncomplicated and the headache occurred 7 days postdelivery, amniotic/ fluid embolus is excluded (and does not usually present with headache), and eclampsia is less likely but not impossible. Cerebral venous thrombosis is the commonest cause of Cerebrovascular disease in the puerperium and presents with worsening headache, seizures, weakness or numbness.



Note blurred disc margins and congested disc.

Pearls

The top of a swollen disc and the surrounding unaffected retina will not both be in focus on the same setting on direct ophthalmoscopy.

Papilledema is a bilateral process, though it may be slightly asymmetric. A unilateral swollen disc suggests a localized ocular or orbital process.
 Vision is usually normal acutely, though the patient may complain of transient visual changes. The blind spot is usually enlarged.

3. Diplopia from VI cranial nerve palsy can be associated with increased intracranial pressure and papilledema.

3. All are true.

Papilledema involves swelling of the optic nerve head, usually in association with elevated intracranial pressure. The optic discs are hyperemic with blurred disc margins; the venules are dilated and tortuous. The optic cup may be obscured by the swollen disc. There may be flame hemorrhages and infarctions (white, indistinct cotton wool spots) in the nerve fiber layer and edema in the surrounding retina. Ocular inflammation (e g, papillitis), tumors or trauma, central retinal artery or vein occlusion, optic nerve drusen, and marked hyperopia may present with similar findings.



Papilledema: disc is hyperemic and swollen with loss of sharp margins. The venules are dilated and tortuous. The cup is obscured. A small flame hemorrhage is seen at 12 to 1 o'clock on the disc margin.

- 4. B. Optic atrophy.
- 5. C. Optic atrophy.

Optic atrophy is the final common morphologic endpoint of any disease process that causes axon degeneration in the retinogeniculate pathway. Clinically, optic atrophy manifests as changes in the color and the structure of the optic disc associated with variable degrees of visual dysfunction. Optic atrophy is actually a misnomer; in the strict histologic definition, atrophy refers to involution of a structure resulting from prolonged disuse.

Ophthalmoscopic classification:

Primary optic atrophy



In conditions with primary optic atrophy (e g, pituitary tumor, optic nerve tumor, traumatic optic neuropathy, multiple sclerosis), optic nerve fibers degenerate in an orderly manner and are replaced by columns of glial cells without alteration in the architecture of the optic nerve head. The disc is chalky white and sharply demarcated, and the retinal vessels are normal. Lamina cribrosa is well defined.

Secondary optic atrophy



In conditions with secondary optic atrophy (e g, papilledema, papillitis), the atrophy is secondary to papilledema. Optic nerve fibers exhibit marked degeneration, with excessive proliferation of glial tissue. The architecture is lost, resulting in indistinct margins. The disc is grey or dirty grey, the margins are poorly defined, and the lamina cribrosa is obscured due to proliferating fibroglial

tissue. Hyaline bodies (corpora amylacea) or drusen may be observed. Peripapillary sheathing of arteries as well as tortuous veins may be observed. Progressive contraction of visual fields may be seen as well.

6. B. Optic atrophy.

There should not be much difficulty with this. The danger is in over diagnosing optic atrophy, so ensure you have looked thoroughly around the field and excluded other pathology. In this case, there is none.

7. E. Retinal artery occlusion.

There is diffuse retinal whitening and a cherry-red spot in the macula, indicating central retinal artery occlusion.

8. E. Right occipital lobe.

Left homonymous hemianopsia with temporal crescent in left eye is shown. The more congruous the defects, the more posterior their location. In addition, the nasal retina is larger and allows a temporal crescent in the visual field in the contralateral eye. Macular sparing or splitting also may occur.

9. D. Right optic tract.

Mass lesions of the optic tract are usually large enough to compromise the optic nerve and chiasm as well. Patients have an incongruous homonymous hemianopsia, bilateral optic disc atrophy, often in a "bow-tie" pattern, and a relative afferent defect on the side opposite the lesion (i.e., the eye with temporal field loss).

10. B. This is a bitemporal hemianopsia.

Lesions of the chiasm cause bitemporal hemianopsia because they damage the crossing nasal nerve fibers. Masses in this area include pituitary tumors, pituitary apoplexy, meningiomas, aneurysms, infection, craniopharyngiomas, gliomas, and other less common tumors. In addition, the chiasm may be damaged by trauma (typically causing a complete bitemporal hemianopsia), demyelinating disease, and inflammatory diseases such as sarcoidosis, and rarely, ischemia.

How do you describe a visual-field defect?

Position: Central (defined as the central 30 degrees), peripheral, or a combination of both. Note if the defects are unilateral or bilateral.
 Shape: Very helpful diagnostically. Visual-field defects can be monocular or binocular. The most common form of monocular sector defect is found in glaucoma. The shape is determined by physiologic interruption of nerve fiber bundles. The typical binocular sector defect is a hemianopsia, which can be subdivided as follows:

Homonymous, total: Loss of temporal field in one eye and nasal field of the other eye. The vertical midline is respected. The fixation point may be included or spared. This defect implies total destruction of the visual pathway beyond the chiasm unilaterally, anywhere from the optic tract to the occipital lobe.
Homonymous, partial: The most common visual-field defect. It may be caused

by injury to postchiasmal pathways. Again, it can result from damage at any point from the optic tract to the occipital lobe.

- Homonymous quadrantanopsia: This is a form of partial homonymous hemianopsia.

- Bitemporal: May vary from a loss of a small amount of the temporal field to complete temporal hemifield loss. This defect signifies damage in the optic chiasm.

- Binasal: This defect signifies an interruption of the uncrossed fibers in both lateral aspects of the chiasm, both optic nerves, or both retinas.

- Crossed quadrantanopsia: A rare defect in which the upper quadrant of one field is lost along with the lower quadrant of the opposite visual field. It can occur as part of the chiasmal compression syndrome where the chiasm is compressed from beneath against a contiguous arterial structure. This produces pressure simultaneously from above and below.

- Altitudinal: This defect can be unilateral or bilateral. A unilateral defect is prechiasmal.

Bilateral lesions may be produced by lesions that press the chiasm up, wedging the optic nerve, such as an olfactory groove meningioma.

- Double homonymous hemianopsia: A result of lesions of the occipital area. There is a loss of all peripheral vision with a remaining small area of central vision representing the spared macula of both eyes. Most are vascular in origin, but they can result from trauma, anoxia, carbon monoxide poisoning, cardiac arrest, and exsanguination. - Macula sparing: This is the rule in occipital damage. The central visual acuity can remain normal.

(Ophthalmology secrets in color / James F. Vander, Janice A. Gault. – 3rd ed., 2007, by Elsevier Inc.)

11. D. Left temporal lobe.

A "pie-in-the-sky" lesion is a homonymous quadrantanopsia involving the superior quadrant. The term indicates a lesion in the optic radiations through the temporal lobe, but similar defects can be seen with occipital lobe lesions as well. These patients often have coexistent seizures, de ja` vu experiences, or uncinate fits.

- 12. C. Left parietal lobe.
- **13**. E. Right temporal lobe.
- **14.** D. Right parietal lobe.

A "pie-on-the-floor" lesion is a homonymous quadrantanopsia involving the inferior quadrant. The term indicates a lesion in the parietal lobe. These patients often have coexistent spasticity of conjugate gaze (tonic deviation of eyes opposite to the side of the lesion when attempting Bell's phenomenon) and optokinetic asymmetry (diminished or absent response with rotation of optokinetic objects toward the side of the lesion).

15. D. Noise-induced hearing loss.

Unfamiliar tests send some candidates into a panic and therefore allow differences to emerge. The usual rule is that unusual tests are usually easy to interpret. Audiograms will be unfamiliar to many candidates, so it is probable they will be fairly easy to interpret. There will be a maximum of four lines plotted on the graph, representing air conduction and bone conduction for each ear. You are less likely to be shown cases of conductive deafness (air conduction lost compared to bone conduction) than sensorineural loss (air and bone conduction both lost), since the former are more in the domain of the ENT surgeon than the physician. The general format of an audiogram is to place increasing sound frequency on the horizontal axis. The vertical axis shows hearing level in decibels, decreasing towards the top of the axis (in fact, to be accurate, the vertical axis really represents hearing level compared to a representative control group against which the audiometer has been calibrated). In other words, the higher the plot, the better the hearing, as the patient is detecting a given frequency at a lower hearing level. In this question, you have been told that there is no air-bone gap, i.e. bone conduction is no greater than

air conduction. As this would not be the case if the question illustrated a conductive hearing loss, the defect here is sensorineural.

Deterioration in auditory acuity with age is normal. It is usually symmetrical and affects high tone acuity more severely. Asymmetrical high-tone sensorineural deafness suggests a cerebellopontine angle lesion such as acoustic neuroma or meningioma. Low-tone fluctuating sensorineural hearing loss is suggestive of Ménière's disease. Noise-induced sensorineural hearing loss secondary to the use of a shotgun only affects the contralateral ear, as the ipsilateral ear is protected by the butt of the gun. The gamekeeper in this case is left handed, so his right ear has been affected by the noise of the gun. By contrast, an explosion next to an individual will affect the ipsilateral ear more. Industrial noise injury is often bilateral, resulting in symmetrical 4 kHz dips. (AN Warrens. A Guide to the MRCP part 2 written paper. 2nd Ed., 2005, Edward Arnold (Publishers) Ltd, London, p 124).

16. B. Ménière's disease.

The diagnosis of Ménière's disease is based on a classical history of fluctuation of the characteristic features: sensorineural deafness, tinnitus, vertigo (typically lasting hours) and a feeling of fullness in the affected ear.

Sounds must be conducted through the middle ear and sensed by the cochlea and CN VIII; then they are processed by the cochlear nuclei and CNS pathways. Conductive deafness is hearing loss due to external auditory canal or middle ear disease. Sensorineural deafness is a perceptive loss of hearing due to disease of the inner ear or eighth nerve. In conductive deafness, bone conduction is better than air conduction, and the reverse is the case for sensorineural deafness. Central deafness is caused by disease affecting the central auditory pathways. Tinnitus an annoying noise in the ear that is usually benign, often is caused by cochlear or eighth nerve disorders. Hearing may be diminished, but the patient is not rendered fully deaf. Presbycusis is hearing loss due to advanced age.

17. D. The iris in the affected eye lacks sympathetic innervation and will be lighter in color.