Training and Self Assessment Sets for

Detection/Rating of White Matter Disease, Lacunes, and EPVS

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Training Set for

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The author has nothing to disclose financially. Please review the information and disclaimers below before starting the program

• If more than one answer can be true, this would be specifically stated in the question. Otherwise, just select one answer.

• Both the training and self assessment sets are for personal training purposes. Separate test questions for CME are provided. The current set will improve the understanding of the topic and make it easier for the participant to apply the information presented to real world patients.

• Medicine is an ever-changing science. As new research and clinical experience broaden our knowledge, changes in treatment and drug therapy are required. The authors and the publisher of this work have checked with sources believed to be reliable in their efforts to provide information that is complete and generally in accord with the standards accepted at the time of publication. Readers are encouraged to confirm the information contained herein with other sources and obtain detailed historical and exam data from individual patients in order to accurately diagnose and treat the individual patient. The authors, publishers and all individuals involved in the preparation of this course disclaim all responsibility for any errors or omissions or for the results obtained from use of the information contained in this work.
Definition of Centrum Semiovale (CS)

- Centrum semiovale represents the mass of white matter superior to the lateral ventricles and corpus callosum, present in each cerebral hemisphere, subjacent to the cerebral cortex.

- The centrum semiovale (CS) levels that can be used to assess enlarged perivascular spaces (CS-EPVS) are shown on the left, including the axial T2 slices where the actual CS-EPVS counts are obtained and also the corresponding sagittal T1 slices to show the levels of these axial T2s. One axial T2 slice is used to count EPVS.

- Note the appearance of the roof of lateral ventricles in the bottom panel (highlighted in blue ovals), this is acceptable.
Clinical Vignette #1

76 year old man with history of diabetes, hypertension, early dementia, presented with 3 days of lethargy, presented with large left temporal intracerebral hemorrhage (red arrow)
An ovoid small lesion is observed in the right basal ganglia, FLAIR sequence (red circle). Its maximum axial diameter is 5.1mm. On a GRE sequence it is isointense.

This is:
a) A microbleed
b) A lacune
c) An enlarged perivascular space
d) An acute small subcortical ischemic infarct
• Correct answer: b
• This is a lacune: it has ovoid/circular shape and it is dark on the FLAIR sequence (i.e: CSF-isointense)
• What differentiates it from EPVS: its size (>3mm) and a rim of hyperintensity (representing reactive gliosis). EPVS are smaller than 3mm and typically do not have a hyperintense rim.
• A microbleed would be GRE hypointense
• An acute small subcortical infarct would not have already cavitated.
Clinical Vignette #2

• 60-year old man with history of hypertension, presented with acute dysarthria and was found to have a left internal capsule hemorrhage (partially captured, red circle).

• Are there any small vessel disease markers visualized in this Axial T2 section? If yes, which ones and what grade?
• This patient has multiple enlarged perivascular spaces in the centrum semiovale.
• Observe the multiple linear, T2 hyperintense lesions following the track of penetrating small vessels.
• Based on the extent (21-40 in this section), this would be assigned a grade 3.
*Remember that the EPVS count used for the grading refers to one side of the brain.
• If there is discrepancy between the two sides on the same axial slice, then the side with the higher count is used.
• In patients with focal lesions such as ischemic strokes or hemorrhages, the contralateral side is used for grading.
Clinical Vignette #3

• 63 year old man with history of liver failure and thrombocytopenia, presented with a right putaminal hemorrhage (highlighted in the red oval).

• What Fazekas scale score WMH is visualized in this FLAIR image?
    a) Periventricular WMH Fazekas 1
    b) Deep WMH Fazekas 2
    c) Deep WMH Fazekas 1
    d) Periventricular WMH Fazekas 3
• Correct answer: a
• This patient has periventricular WMH Fazekas scale 1. Observe the pencil-thin hyperintense rim (caps) on the edge of the frontal horns (orange circles), and the even fainter signal on the posterior (occipital) horn (green circles)
• There are no Deep WMH visualized in this image
• For Periventricular Fazekas scale 3, much more conspicuous, confluent WMH should be present
Clinical Vignette #4

- 71 year old man with long standing hypertension, hypercholesterolemia, with a right thalamic lacunar infarct (not visualized in this image.
- What Fazekas scale score WMH is visualized in this FLAIR image?
  a) Deep WMH Fazekas 3
  b) Deep WMH Fazekas 2
  c) Deep WMH Fazekas 1
  d) Deep WMH Fazekas 0
• Correct answer: b
• This patient has Deep WMH Fazekas scale 2. (red arrows)
• These are more conspicuous than small punctate lesions that would be graded as Fazekas 1 and not confluent yet to be considered Fazekas 3
Clinical Vignette #5

- 70 year old woman with dementia, coronary artery disease, hypertension
- What EPVS grade would be assigned?
  a) Centrum Semiovale grade 1
  b) Basal Ganglia grade 2
  c) Basal Ganglia grade 1
  d) Centrum Semiovale grade 2
• Correct answer: C
• This patient has basal ganglia EPVS grade 1
• T2 hyperintense (CSF isointense), with round shape at the basal ganglia level, <3mm in size (differentiation from lacunes)
• Remember that the grading is done taking into account one side of the brain, the one with the highest lesion count (if an acute/chronic ischemic or hemorrhagic stroke, always use the uninvolved side)
• In this case the left side has slightly higher count. The total count is >1, but <10, thus the grade assigned is 1
Clinical Vignette #6

- 65 year old woman with atrial fibrillation on warfarin, hypertension, diabetes, presented with severe headache. She was found to have a right temporal hemorrhage, not visualized here.

- What Fazekas scale score WMH is visualized in this FLAIR image?
  a) Periventricular WMH Fazekas 1
  b) Deep WMH Fazekas 0
  c) Deep WMH Fazekas 1
  d) Periventricular WMH Fazekas 3
• Correct answer: d
• This is an example of Periventricular WMH Fazekas 3
• These are large confluent periventricular hyperintensities, more pronounced on the occipital horns (green circles).
• Slightly less pronounced but also confluent WMH can be seen surrounding the frontal horns (orange circles)
• Deep WMH Fazekas is 2, not 0 or 1
Clinical Vignette #6 continued

What Fazekas scale score WMH is visualized in this FLAIR image?

a) Periventricular WMH Fazekas 1
b) Deep WMH Fazekas 0
c) Deep WMH Fazekas 3
d) Periventricular WMH Fazekas 3
• Correct answer: c
• This is an example of Deep WMH Fazekas 3
• Firstly, this is beyond the periventricular level, therefore periventricular WMH grading is precluded on this slice
• Confluent symmetric WMH on both sides, much more conspicuous than punctate or larger solitary lesions
Clinical vignette #6 continued

- Besides WMH are there any other visible small vessel disease MRI markers on this image? If yes, which one(s)?
In addition to the Deep WMH Fazekas 3, there is also a lacune seen on this FLAIR image (red circle). This is a “lobar lacune” as it is situated in subcortical white matter outside of the classical deep locations such as thalamus, internal capsule, basal ganglia or pons. “Lobar lacunes” are found more commonly in cerebral amyloid angiopathy, as opposed to the “deep lacunes” seen in hypertensive cerebral small vessel disease.

Notice the spherical/ovoid cavitating lesion, FLAIR hypointense (i.e. CSF isointense) with a rim of FLAIR hyperintensity which on close inspection is visible even within the surrounding WMH FLAIR hyperintensity (see zoomed-in image, red arrows).
Clinical Vignette #7

• 74 year old woman with recurrent lobar ICH (left parieto-occipital, red circle).

• Which small vessel disease marker is visualized in this Axial T2 image and what is its severity?
• This patient has Centrum Semiovale EPVS, grade 3 (21-40). Notice a mix of linear (green arrow) and round (orange arrows) T2 hyperintense (i.e. CSF isointense) lesions.

• Notice the discrepancy between Centrum Semiovale and Basal Ganglia EPVS on the same patient. Very few scattered lesions seen in basal ganglia (EPVS Grade 1, yellow arrows).

• Topographical distribution patterns of EPVS might signify different underlying pathophysiologic processes. CSO-predominant EPVS might be associated with cerebral amyloid angiopathy, whereas basal ganglia-predominant is thought to signify hypertensive arteriopathy.

• This patient has recurrent lobar ICH, raising concern for cerebral amyloid angiopathy. The EPVS pattern is CSO-predominant.
Self assessment Q/A for:

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Question 1

Which of the following cerebral small vessel disease pathologies are seen in this axial FLAIR image? (more than one might apply)

a) Deep lacune  
b) Enlarged Perivascular spaces  
c) White matter hyperintensities  
d) Cortical microinfarcts
Question 1 answer

• Correct answer(s): (a) Deep lacune and (c) White matter hyperintensities
Question 2

Which of the following cerebral small vessel disease pathologies are seen in these axial MR images from the same patient?

a) Lobar lacune
b) Severe white matter hyperintensities
c) Cortical microbleeds
d) All of the above
Question 2 answer

• Correct answer: (d) All of the above
For the patient in question 2, what would be the most likely etiology of the cerebral small vessel disease based on distribution of lesions?

a) Hypertensive cerebral small vessel disease
b) Cerebral amyloid angiopathy
c) Cerebellar stroke
d) None of the above
Question 3 answer

• Correct answer: (b) Cerebral amyloid angiopathy
Question 4

What is the grade of EPVS seen in this image?

a) The are no EPVS
b) Basal Ganglia, Grade 1
c) Centrum Semiovale Grade 2
d) Basal Ganglia Grade 2
e) Centrum Semiovale Grade 1
Question 4 answer

• Correct answer: (b) Basal Ganglia Grade 1
Question 5

What is the Fazekas scale score seen in this image?

a) Periventricular WMH Fazekas 1
b) Deep WMH Fazekas 1
c) Deep WMH Fazekas 2
d) Periventricular WMH Fazekas 2
Question 5 answer

• Correct answer: (d) Periventricular WMH Fazekas 2
Question 6

What is the grade of EPVS seen in this image?

a) The are no EPVS
b) Basal Ganglia, Grade 1
c) Centrum Semiovale Grade 3
d) Basal Ganglia Grade 3
e) Centrum Semiovale Grade 1
Question 6 answer

• Correct answer: (c) Centrum Semiovale Grade 3
Question 7

What is the Periventricular WMH Fazekas scale score seen in this image?

a) 0  
b) 1  
c) 2  
d) 3
Question 7 answer

• Correct answer: (b) Periventricular WMH Fazekas Scale 1
Question 8

Which of the following small vessel disease markers are visualized on this image? (more than one might apply)

a) Lacunes
b) Cerebral microbleeds
c) White matter hyperintensities
d) Enlarged perivascular spaces
Question 8 answer

• Correct answer: (c) White matter hyperintensities (d) Enlarged perivascular spaces
What is the grade of EPVS on this image?

a) Basal ganglia Grade 1
b) Centrum semiovale Grade 2
c) Basal ganglia Grade 2
d) Basal Ganglia Grade 3
Question 9 answer

• Correct answer: (c) Basal Ganglia EPVS Grade 2 (11-20)