NEUROSTATUS-EDSS FAQS

1. VISUAL (OPTIC) FUNCTIONS

1. If the patient has a visual acuity between 0 and 0.09 in the worse eye, does it lead to unconverted visual FSS 5 or 6?

If the worse eye has a visual acuity between 0.00 and 0.09, this can meet the definitions of unconverted visual FSS 5 or 6 depending on the acuity of the better eye:

- If the better eye's visual acuity is more than 0.33, it meets the criteria for unconverted visual FSS 5.

-If the better eye's visual acuity is 0.33 or less, it meets the criteria for unconverted visual FSS 6.

2. If the patient has a visual acuity between 0.10 and 0.20 in the worse eye, does it lead to unconverted visual FSS 4 or 5?

If the worse eye has a visual acuity between 0.10 and 0.20, this can meet the definitions of unconverted visual FSS 4 or unconverted visual FSS 5 depending on the acuity of the better eye:

- If the better eye's visual acuity is more than 0.33, it meets the criteria for unconverted visual FSS 4.

-If the better eye's visual acuity is 0.10-0.33, it meets the criteria for unconverted visual FSS 5.

3. If the patient has a visual acuity between 0.21 and 0.33 in the worse eye, does it lead to unconverted visual FSS 3 or 4?

If the worse eye has a visual acuity between 0.21 and 0.33, this can meet the definitions of unconverted visual FSS 3 or 4 depending on the visual acuity of the better eye:

- If the better eye's visual acuity is more than 0.33, it meets the criteria for unconverted visual FSS 3.

-If the better eye's visual acuity is 0.21-0.33, it meets the criteria for unconverted visual FSS 4.

4. ***What is the visual FSS if visual acuity in both eyes is 0.67?***

   If visual acuity in both eyes is 0.67, the FSS is 2 (“worse eye with maximal visual acuity of 0.34-0.67”).

5. ***What is the difference between unconverted visual FSS 4 and 5 in contrast to the other Visual FSS?***

   Both FSS 4 and 5 have two independent definitions each (as you can see on the Neurostatus-EDSS visual calculation table).

2. BRAINSTEM FUNCTIONS

1. What would constitute an isolated unilateral internuclear ophthalmoplegia (INO) score?

In the unilateral internuclear opthalmoplegia there is a lesion on the medial longitudinal fasciculus. There is an ipsilateral adduction deficit (partial or complete) with a contralateral, horizontal abducting nystagmus on attempted gaze to the contralesional side.

Firstly, a complete paralysis of the adduction in one eye meets the criteria for a moderate EOM impairment (subscore of 3: “complete loss of movement in one direction of gaze in either eye”).

Secondly, a sustained nystagmus of the abducting eye meets the criteria for a severe nystagmus (subscore of 3).

A severe nystagmus leads to brainstem FSS 3.

2. What would constitute an isolated bilateral internuclear ophthalmoplegia (INO) score?

This occurs if there is a bilateral damage to the medial longitudinal fasciculus. There is a bilateral adduction paralysis and a bilateral abduction nystagmus.

A bilateral internuclear ophthalmoplegia would still lead to brainstem FSS 3. A bilateral adduction paralysis still meets the criteria for moderate EOM impairment (subscore of 3). The sustained bilateral nystagmus – just as the unilateral sustained nystagmus – is defined as severe nystagmus (subscore of 3). Additionally, you cannot score a higher FSS than if there is only extraocular movement impairment or nystagmus and no other cranial nerve involvement, so both uni- and bilateral INO lead to an FSS of 3.

3. What would a one-and-a-half syndrome score?

In the one-and-a-half syndrome there is a lesion of the medial longitudinal fasciculus, and the paramedian pontine reticular formation and/or cranial nerve VI nucleus on the same side.

The ipsilateral eye has no horizontal movement, while the contralateral eye can abduct (there is also an abduction nystagmus) but cannot adduct. This is in line with a marked EOM impairment (subscore of 4: “complete loss of movement in more than one direction of gaze in either eye”) and a severe nystagmus (subscore of 3). They both lead to brainstem FSS 3.

4. ***If a patient had a trigeminal nerve ablation for symptomatic trigeminal neuralgia and currently has hypesthesia of the second branch of trigeminal, which would be the brainstem FSS?***

   If trigeminal neuralgia was MS related and the hypesthesia is a consequence of the treatment, you should score the deficit, as it is MS related. The score depends on the severity of the sensory deficit (for example, if sharp/dull discrimination were impaired, there would be a moderate trigeminal damage. If this was the only brainstem deficit, brainstem FSS would be 3).

5. ***Bearing in mind the rare association of clinically diagnosed Bell Palsy (Peripheral Seventh Cranial Nerve Palsy) and Multiple Sclerosis [A Diagnostic Dilemma - A Case Report. Saleh C, Patsi O, Mataigne F, Beyenburg S. Case Rep Neurol. 2016 Jan 23;8(1):27-33.] how would you score the Brainstem FS for a facial weakness ascribed to Bells palsy (LMN CNVII) in the context of known MS?***

   In this case the rater is the one that must decide if the symptoms are MS related. However, in case of doubt, we recommend assuming a relation to MS. If a relation to MS is assumed, the same should be considered in the follow-up examinations.

   As an example, if the patient has a mild facial weakness (subscore of 2) and the rest of the brainstem examination is normal:

   - If we consider it MS related, the brainstem FSS should be 2.

   -If we do not consider it MS related, the subscore in facial weakness would be 2 P or 2T (“P” for permanent, or “T” for temporary) and the brainstem FSS should be 0.

3. PYRAMIDAL FUNCTIONS

1. ***What are examples of fatigability in motor tasks?***
   - A patient is now able to jog a considerably shorter distance or for considerably shorter time than previously (not secondary to other conditions unrelated to MS).
   - A patient has difficulty with his daily walk, which he has done every day for the past year without problems.
   - A patient's ability to play football is worsening and he/she cannot keep up with his/her peers.
   - A patient previously exercised three times a week at the gym without problems. Now when the patient exercises, he/she is completely exhausted for the rest of the day**.**

2. ***A patient reports heavy legs while going/ running upstairs. He cannot run upstairs as fast as before after his children while playing. Assessment of muscle strength is BMRC grade 5 in all muscle groups. What pyramidal FSS should this patient get?***

   If there is no other reason for this problem (e.g arthrosis, knee pain…) you should assume that this is due to MS. This would be an example of reduced overall motor performance (OMP). In this case, OMP should be 1 and pyramidal FSS should be 2 (according to motor fatigability or reduced performance in strenuous motor tasks).

4. CEREBELLAR FUNCITONS

1. ***If a patient has a moderate hemiparesis (BMRC grade 3) and she is unable to walk in a straight line (tandem walking), but she does not appear ataxic. How should tandem walking and the EDSS Step be scored?***

   If there are no other signs of ataxia you should score a 2 for tandem walk and for the cerebellar FSS a 2X. X indicates problems with cerebellar scoring due to interfering paresis or sensory deficits and must be added next to the cerebellar FSS.

2. ***How can I score gait ataxia for a patient who walks with assistance by another person and cannot walk alone due to the ataxia?***

   The subscore for gait ataxia would be 4 (severe gait ataxia: unable to walk more than a few steps unassisted or requires a walking aid or assistance by another person because of ataxia). You may have to add an "X" to indicate an underlying weakness if this is also the case.

3. ***If a patient has a monoplegia of one upper limb and cannot be tested for limb ataxia, which subscore should be given for the limb ataxia?***

   The subscore for limb ataxia would be 4 (severe) for all subscores (tremor/dysmetria and rapid alternating movements). The cerebellar FSS depends on the rest of the subscores, but an "X" must be placed after the final FSS to indicate the role of weakness.

4. ***If a patient has severe gait ataxia and only mild limb ataxia, what is the Cerebellar FSS?***

   The Cerebellar FSS would be 3. The presence of severe gait ataxia alone results in a Cerebellar FSS of 3.

   For cerebellar FSS of 4, the patient should have a severe gait or truncal ataxia AND also a severe ataxia in three or four limbs.

5. ***Am I right to understand that the outcome for the cerebellar FS is either 0, 1, 2, 3, 4, 5 and that in addition, the rater can add an X to these scores?***

   The X should be placed after the FSS only if weakness (usually when muscle groups are BMRC grade 3 or less) or sensory deficits interfere with the testing of ataxia (for example cerebellar FSS 3X). Please note that the X should not be placed behind the EDSS Step. The X is only for documentation of the possible role of weakness/ sensory deficits when testing cerebellar functions.

6. ***A patient has non-MS-related findings (mild ataxia and dysmetria due to infantile cerebral paralysis). Should these findings be reflected in the cerebellar FSS (FSS 2), but not considered for EDSS Step calculation? Or should they neither be reflected in the cerebellar FSS (FSS 0) nor in the EDSS Step?***

   The FSS and EDSS Step should reflect MS related deficits only. Non-MS related findings should be marked (e.g. with a P) and will not be taken into consideration when assessing the FSS and EDSS Steps. So, in this case, if the cerebellar symptoms are clearly only apparent due to the non-MS related cerebral paralysis, the cerebellar FSS would be 0. In case of any doubt, the examining physician should assume a relation to MS.

5. SENSORY FUNCTIONS

1. ***What is the FSS of a patient with impairment of sharp and dull during testing, who is not aware of the deficit, and who does not have any other signs in superficial sensation, vibration sense or position sense?***

   An impaired discrimination is already a moderate grade in superficial sensation, which will result in an FSSof 3 or higher - depending on how many limbs are involved. However, it is rare that a patient has such severe sensory deficit without complaining about sensory disturbance at all. There might be an attention deficit or a problem in cooperation or not taking the examination seriously enough. Here a judgement of the investigator is required using experience and common sense.

2. ***If a patient has moderately decreased vibration sense in four limbs and no other findings in the sensory examination, what is the sensory FSS?***

   The Sensory FSS would be 3. Please note that not all possible combinations are described in the definitions. No score exists which corresponds to moderately decreased vibration sense in four limbs. However, the patient exceeds the limitations of sensory FSS 2 (moderate decrease in vibration in one or two limbs) and does not exceed the limitations of sensory FSS 3. For that reason, the appropriate score would be a sensory FSS of 3.

3. ***In the sensory examination, the patient is found to have reduced temperature sensation, but the patient is not aware of the deficit. Therefore, the patient receives a score of 1 (signs only) for superficial sensation. There are no other findings in the Sensory FS. What is the Sensory FSS?***

   - If the patient has reduced temperature sensation in one or two limbs but was not aware of the deficit before formal testing, then the sensory FSS is 1.
   - If the patient has reduced temperature sensation in three or four limbs but was not aware of the deficit before formal testing, then the Sensory FSS is 2.

6. BOWEL/BLADDER FUNCTIONS

1. ***If a patient needs suppositories to evacuate bowel due to constipation, what is the bowel/bladder FSS?***

   If the constipation is due to MS and he needs suppositories on a regular basis, this would be scored as a severe bowel dysfunction (a subscore of 3). If there are no other symptoms, this would lead unconverted bowel/bladder FSS 3 (converted also 3).

2. ***What is the difference between a subscore of 3 and a subscore of 4 in urinary hesitancy?***

   Subscore 3 (severe urinary retention): the patient can still feel the full bladder and the urge to urinate but requires intermittent use of catheterization.

   Subscore 4 (loss of function): the patient requires constant catheterization (=fixed system).

3. ***What is the difference between a subscore of 3 and a subscore of 4 in urinary urgency?***

   Is there any sign of remaining sphincter activity? If yes, we would recommend scoring a 3 (severe urinary urgency), otherwise a 4 (loss of function).

   Subscore 3 (severe urinary urgency): it is a severe problem but still manageable for the patient without constant catheterization. Patient wears incontinence pads regularly.

   Subscore 4 (loss of function): patient has constant catheterization or always wears incontinence pads.

4. ***What is the difference between a subscore of 3 and a subscore of 4 in bowel dysfunction?***

   Is there any sign of remaining sphincter activity? If yes, we would recommend scoring a 3 (severe bowel dysfunction), otherwise a 4 (complete loss of function).

   Subscore 3 (severe bowel dysfunction): the patient has a severe problem but still has some limited ability to steer the function of the bowel.

   Subscore 4 (complete loss of function): complete loss of sphincter function, meaning that the patient must wear incontinence pads or has a stoma.

5. ***If the patient is using drugs to improve bladder function, how should this chapter be scored?***

   You should score current patient's symptoms independently of the use of drugs.

7. CEREBRAL FUNCTIONS

1. ***How does the presence of depression or euphoria affect the Cerebral FSS?***

   Depression or euphoria must be documented in the scoring sheet, but neither is taken into consideration for the FSS and EDSS Step calculation. Therefore, the cerebral FSS is not affected by depression or euphoria alone.

   If a patient is taking antidepressants and reports no depressive symptoms, how should I score the depression subscore?

   If the patient has no depressive symptoms under medication, the depression subscore should be 0.

2. ***What would be an example of 'sign only' in mentation?***

   A score of 1 in mentation means that the neurologist notices cognitive changes, of which the patient and the family/partner are not aware.

8. AMBULATION

1. ***Is the use of a foot orthotic device considered unilateral assistance?***

   No, the use of foot orthotic devices is considered "no assistance".

2. ***Do we consider a functional electrical stimulator (FES) as unilateral assistance or as an ankle foot orthosis (no assistance)?***

   The use of a functional electrical stimulator (FES) is considered as “no assistance”.

3. ***What are some examples of bilateral assistance?***

   Examples of bilateral assistance are walker, assistance by another person, bilateral crutches, bilateral canes, bilateral sticks.

   Please note that assistance by another person is considered bilateral assistance even if they stabilize the patient only on one side.

4. ***How can I rate the AS if the patient refuses to measure the walking distance?***

   Sometimes it might not be possible to encourage a patient to measure the walking distance. In this case you must rely on the patient's statement and rate the AS per reported distance, your judgement from the physical examination as well other facts (e.g. how did the patient come to the clinical visit, walking ability in previous visits…).

5. ***How can I rate the AS if the walking ability is temporarily restricted due to a condition unrelated to MS?***

   If the distance measured and/or the assistance has changed due to a condition unrelated to MS (e.g surgery, pain), to the examining neurologist should add a T for ‘temporary’ next the corresponding subscore. The AS should be calculated using the walking ability prior to this temporary condition, that is, based only on the walking restriction that is related to MS.

6. ***How can I determine if ambulation is unrestricted depending on age and physical condition? For example, if the patient reports that she can walk 5 km, is that enough to be considered ‘unrestricted ambulation’?***

   It is useful to ask the patient if they themselves feel restricted in their ambulation. If the patient is reliable, he/she does not feel restricted in ambulation, and you consider 5 km to

be comparable to healthy individuals of similar age and physical condition you can rate ambulation as "unrestricted".

7. ***Is the patient allowed to rest during the measurement of the walking distance? If yes, how often and how long?.***

   The patient is allowed to rest only for a few seconds (2-3 seconds) without holding the wall or another person. He/she can rest a maximum of two times.

8. ***Is the patient allowed to touch the wall during the measurement?***

   The patient must be allowed to touch the wall to avoid falling. However, the measurement of the walking distance would end at that point.

9. ***What is the AS of a patient who can walk 5 to 10 metres without help and 200 metres with bilateral assistance?***

   This patient needs assistance in daily life, therefore a distance of 5 to 10 metres without help does not reflect the real patient's real-life walking "situation". We would therefore recommend rating the AS on the basis of a distance measured of 200 metres with bilateral assistance. This corresponds to an AS of 7.

10. ***Does the examining neurologist have to observe the distance measured or can this action be delegated to a study staff member?***

    In general, the examining neurologist should be the one observing the patient walking with the assistance needed and measure the time. However, this task can be delegated in individual cases to a well-trained health care professional (HCP). The HCP should be able to motivate the patient to give their best performance but at the same time take safety measures to prevent falls and know how to react to short stops or touching the wall or the accompanying person because of ataxia.