



REGISTRY V3
UHDRS FOR ADVANCED HD PATIENTS (UHDRS-FAP)

Study Site:

Examiner:

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Subject:

Date info obtained:

D	D

 .

M	M

 .

Y	Y	Y	Y

All items must be completed. Use U if information is unavailable. Use N if information is not applicable.

Information

Context:

- 0 = patient seen alone
- 1 = accompanying person

Relationship of the person accompanying the patient:

Living Environment

Institution:

- 0 = at home
- 1 = at home and day care
- 2 = temporary residential care (respite care)
- 3 = permanent residential care

Speech therapy:

- 1 = yes
- 0 = no

sessions/week:

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time/session:

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Physiotherapy:

- 1 = yes
- 0 = no

sessions/week:

--	--

time/session:

--	--

Occupational therapy:

- 1 = yes
- 0 = no

sessions/week:

--	--

time/session:

--	--

Psychologist:

- 1 = yes
- 0 = no

sessions/month:

--	--

time/session:

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Non-medical carer:

1 = yes
0 = no

sessions/week:

time/session:

Orderly/Nurse Assistant:

1 = yes
0 = no

sessions/week:

time/session:

Nurse:

1 = yes
0 = no

sessions/week:

time/session:

Family caregiver:

1 = yes
0 = no

sessions/week:

time/session:

Other:

1 = yes
0 = no

Specify:

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sessions/week:

time/session:

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Comments for the visit:

I Motor Section

Walk around (10m):

- 0 = possible without help
- 1 = possible with walking assistance device (rollator/cane)
- 2 = with aid of person or own handed wheelchair
- 3 = wheelchair only with help
- 4 = impossible (bedridden patient, unable to sit)

Capacity to transfer:

- 0 = normal
- 1 = correct but slow
- 2 = difficult and needs help
- 3 = difficult, only possible with help
- 4 = impossible

Falls:

- 0 = absent
- 1 = less than 1/month
- 2 = between 1/week and 1/month
- 3 = almost every day
- 4 = daily

Capacity to eat:

- 0 = normal
- 1 = slow but correct
- 2 = possible but unable to cut meat
- 3 = can eat only with spoon or fingers
- 4 = complete help
- 5 = gastrostomy but oral food still possible
- 6 = gastrostomy, and oral food impossible

Choking (or coughing) on food or liquid during swallowing:

- 0 = less than once a month
- 1 = between once a week and once a month
- 2 = almost every day (drinking and eating)
- 3 = almost every day even outside of any food or drink intake (saliva)
- 4 = several times each day, whatever the type
- 5 = permanent gastric tube

Dysphagia:

- 0 = absent
- 1 = rare < 1/month
- 2 = medium, between 1/week and 1/month
- 3 = frequently, several times a week
- 4 = daily

Dysarthria:

- 0 = normal
- 1 = questionable no need to repeat
- 2 = necessary to repeat to understand
- 3 = almost incomprehensible
- 4 = impossible articulation



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Capacity to dress:

- 0 = normal
- 1 = normal with external encouragement
- 2 = normal when clothing is prepared and external encouragement
- 3 = external help (with participation of patient)
- 4 = external help (without participation of patient)

Capacity to wash:

- 0 = normal
- 1 = normal with encouragement
- 2 = assistance required for certain tasks but other tasks are spared
- 3 = assistance required for all tasks (with help of patient)
- 4 = complete support (without help of patient)

Other Neurological Signs

Pyramidal signs:

- 0 = no
- 1 = yes - unilateral
- 2 = yes - bilateral

Cerebellar syndrome:

- 0 = absent
- 1 = present

Imitation synkinesias:

- 0 = absent
- 1 = present (contralateral hand or foot)
- 2 = present, bilateral and/or recruitment in other limbs

Muscle contractures:

Tendon contractures (intensity):

- 0 = none (only incomplete extension)
- 1 = mild (normal joint)
- 2 = moderate (visibly malformed joint)
- 3 = major (extremely deformed)

Tendon contractures (number of joints):

- 0 = no contracture
- 1 = one site
- 2 = two sites
- 3 = three sites
- 4 = four sites
- 5 = > four sites

II Cognitive Section

PEGV:

Functional matching:

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Categorical matching:

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Y	Y	Y	Y

All items must be completed. Use U if information is unavailable. Use N if information is not applicable.

Total correct (max. =10):

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,

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Stroop test:

Has the Stroop test already been completed during this visit?

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1 = yes
0 = no

Colour naming (correct in 45 sec):

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Word reading (correct in 45 sec):

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Interference (correct in 45 sec):

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Orientation in time:

For the next 2 questions, please use:

0 = incorrect
1 = correct

Day of the month:

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Year:

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For the next 4 questions, please use:

0 = incorrect
1 = correct (cued)
2 = correct (spontaneous)

Month:

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Season:

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Day of the week:

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Part of the day:

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Participation in activities:

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0 = not at all, despite encouragement/prompting
1 = sometimes refuses, despite encouragement/prompting
2 = agrees when encouraged
3 = agrees spontaneously when an activity is proposed
4 = agrees spontaneously, finds something to do

Imitation (apraxia):

For the next 5 questions, please use:

0 = severe
1 = moderate
2 = questionable
3 = normal

Bucco-facial:

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Right arm:

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Y	Y	Y	Y

All items must be completed. Use U if information is unavailable. Use N if information is not applicable.

Left arm:

Right leg:

Left leg:

Automatic series:

For the next 4 questions, please use:

- 0 = incorrect
- 1 = correct

a, b, c, d, ...

Monday, Tuesday, Wednesday, Thursday ...

January, February, March, April ...

1, 2, 3, 4 ...

III Somatic Section

Sweating:

- 0 = not present
- 1 = present in emotional situation
- 2 = permanently present, requiring no further measures
- 3 = permanently leading to frequent change on clothing

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Hypersalivation:

- 0 = absent
- 1 = sometimes, not daily
- 2 = intermittent, daily
- 3 = permanent

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Urinary incontinence by night:

- 0 = absent
- 1 = rare < 1/month
- 2 = medium, between 1/week and 1/month
- 3 = frequently, several times a week
- 4 = daily

Urinary incontinence by day:

- 0 = absent
- 1 = rare < 1/month
- 2 = medium, between 1/week and 1/month
- 3 = frequently, several times a week
- 4 = daily

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Y	Y	Y	Y

All items must be completed. Use U if information is unavailable. Use N if information is not applicable.

Digestion disorders:

For the next 3 questions, please use:

- 1 = yes
- 0 = no

Constipation:

Diarrhoea:

Vomiting:

Faecal incontinence:

- 0 = absent
- 1 = rare < 1/month
- 2 = medium, between 1/week and 1/month
- 3 = frequently, several times a week
- 4 = daily

Daytime sleeping:

- 0 = absent
- 1 = planned midday sleep
- 2 = sleep periods not planned
- 3 = falls asleep inappropriately (e.g. during conversations)

Pressure ulcers:

- 0 = absent
- 1 = redness
- 2 = erosion
- 3 = superficial lesion
- 4 = deep lesion

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IV Behaviour Section

For the next 8 questions, please use:

- 1 = yes
- 0 = no

The patient seems depressed:

The patient seems apathetic:

The patient seems agitated (without recognisable cause):

The patient seems anxious:

The patient seems irritable (easily agitated):

The patient seems aggressive (tense, agitated):

The patient seems to hallucinate or has a delirium:

The patient seems obsessed or to have fixed ideas:



Hints:

Information

The 'UHDRS-FAP' complements the original UHDRS and does not replace it. Thus, until validation on a large cohort of patients, both scales should be administered during the same clinical assessment. The 'UHDRS-FAP' aims to evaluate the needs of patients in the advanced stage of the disease, even when no other information is available. Ideally, a caregiver should be available to provide information.

Institution

If patient is in permanent residential care, number of hours for nurses and orderly is not required. The family caregiver might be acknowledged if s/he assumes a responsibility in care (e.g. providing lunch).

I Motor Section

Capacity to transfer

If no carer is present, the examiner should ask the patient to transfer from a bed to a chair

Falls

Rate "daily" if walking without help is impossible

Capacity to dress

Ask the patient to put on a jacket or sweater if no carer is available

Cerebellar syndrome

Avoid the finger-to-nose if the patient is too choreic, instead ask the patient to take the examiner's hand or a pen.

Imitation synkinesias

To test imitation synkinesias:

1. Finger tapping with right hand, then left.
2. Foot tapping with right foot, then left.

If there are synkinesias of the contralateral hand during finger tapping/contralateral foot during foot tapping, score 1; if controlateral synkinesias of the upper limbs appear during right and left finger tapping (and/or controlateral synkinesias of the lower limbs appear during right and left foot tapping), score 2; if there is a recruitment of another part of the body (e.g. mouth/leg during finger tapping), also score 2. If the patient fails to follow the instructions of the examiner (if s/he does not understand the task or has a motor incapacity), score 2 and precise it in comment.

If synkinesias have already been observed during the apraxia test, (cognitive section UHDRS-FAP) please enter the synkinesia score based on this assessment (only enter if the same rater completes the motor and the cognitive sections).

Muscle contractures

The following joints must be examined bilaterally: ankles, knees, wrists.

For intensity, score:

- mild when the joint is retracted without evident deformation
- moderate when the joint is visibly deformed
- major when the joint is severely deformed

II Cognitive Section

Aim to obtain the best performance in the patient. If there is a hint of an answer, the examiner should wait for the patient to provide the full answer. There is no time limit but the examiner should limit the duration of this section in respect of his/her own constraints in order to be able to complete the assessment.

PEGV

Protocole d'Evaluation des Gnosies Visuelles (visual agnosia test). The examiner may point to each picture to help the patient provide a yes/no response to each of the targets. Record as failure if the patient says "yes" to an incorrect picture even if s/he goes on to say "yes" to the correct one (it may be response bias towards "yes"). For more detailed hints, see specific instructions.

Pointing

The examiner should ask the patient to point to each item. Say, "Point to..."

Scoring:

Score 1 if patient looks at, touches, or indicates towards the target object. What counts is task comprehension. If the patient cannot move, answers provided by gaze direction (to the window, the door, etc.) are accepted if the patient clearly looks at the target
Score 0.5 if the patient makes left/right errors
Score 0 if the patient mistakes the target

Simple commands

The examiner reads aloud each simple command.

Scoring

Score 1 if correct
Score 0.5 for cued (correct) if an example is given or if there is confusion over left/right
Score 0 for incorrect if cue leads to an incorrect response

Has the Stroop test already been completed during this visit (from the cognitive section of the standard UHDRS)?

If yes, Stroop scores will be populated from the Brief Cognitive eCRF.

Orientation in time

Ask the patient to give the date spontaneously (day of the month, month, year, day of the week, season, part of the day such as morning, afternoon, etc).
If not successful at giving the date spontaneously, offer multiple choices using the cards.

Card A = part of the day

Card B = seasons

Cards C1, C2, C3 each show 4 months (present card containing actual month)

Card D = days of the week.

When the examiner feels more comfortable with auditory multiple choices rather than cards, s/he might propose to do this instead. Note, the examiner must provide the same number of items orally as given on the cards.



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Imitation (apraxia)

Imitation test for apraxia

- Face: clicking of the tongue, the teeth, showing teeth, coughing, stick out the tongue.
- Arms: show a fist with thumb up, turn it down, open 3 fingers, side of hand against cheek palm backwards, fist under chin, and flat palm.
- Legs: foot tapping and rotating the foot to the right and to the left.

Scoring:

- Severe = severe apraxia, no gestures
- Moderate = moderate apraxia, vague hint of a gesture
- Questionable = questionable apraxia
- Normal = normal imitation of gestures

Automatic series

The examiner says "Please complete the next item in this series...". Score as correct when the patient correctly produces the fifth item.

III Somatic Section

Vegetative signs should be evaluated over the past month. The examiner should ask a carer about bodily functions, or if no carer is present, try to find cues from the patient. For a mute patient, the examiner should check for use of diapers. In the same way, the examiner observes sweating, hypersalivation, slavering, and sleeping by day.

Urinary incontinence by night

If the patient is incontinent, score frequency as daily.

Urinary incontinence by day

If the patient is incontinent, score frequency as daily.

Faecal incontinence

If the patient is incontinent, score frequency as daily.

IV Behaviour Section

The following items are judged based on observation during the consultation and the impression of the informant (if present) over the last month. All sources of information are allowed (caregivers, family, examiner's observation).