



EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

REGISTRY 2 – data collection / CRF completion guidelines

Index

Guidelines of Form Demographics	2
Guidelines of Form Medical History	3
Guidelines of Form CAG Analysis	3
Guidelines of Form General Variable Items	3
Guidelines of Form Medication Log.....	4
Guidelines of Form Comorbid Conditions	5
Guidelines of Form Huntington's Disease Rating Scale '99 - Motor Assessment.....	5
Guidelines of Form Huntington's Disease Rating Scale '99 - Behavioral Assessment.....	7
Guidelines of Form Huntington's Disease Rating Scale '99 - Functional Assessment	9
Guidelines of Form Huntington's Disease Rating Scale '99 - Functional Capacity.....	11
Guidelines of Form Huntington's Disease Rating Scale '99 - Cognitive Assessment.....	12
Guidelines of Form Huntington's Disease Rating Scale '99 - Clinical Summary	13



Guidelines of Form Demographics

Label	Instructions
Date of enrolment	When was the participant enrolled into Registry?
Compute next visit	Select this field to automatically suggest a date for the next visit. Visits should be annual.
Date of next visit	Automatically calculated.
Comments	Add a comment or notes here. The content is for your personal use only.
Gender	Refers to self-reported gender. Please comment if genetic and phenotypic/behavioural gender dissociate.
Ethnicity	<p>The self assigned ethnicity - here operationalised by the area of origin - should be reported. Please ask: 'How would you describe your ethnic background?' and write down the answer of your participants in the field provided.</p> <p>It is understood that 'ethnicity' is not precisely defined. Ethnicity is used here to indicate shared origins, culture and traditions but not in an attempt to propose a taxonomic division of humankind by physical/genetic characteristics as implied by the term 'race'.</p> <p>Examples:</p> <ul style="list-style-type: none"> • Caucasian: synonymous to 'white' (e.g. British, French, German, Irish, Italian, Swedish etc.) • African-Black: area of origin south of the Sahara • African-North: area of origin Sahara and north of the Sahara (e.g. Algeria, Egypt, Morocco, Tunisia etc.) • American-Black: people of African descent whose area of origin is within the Americas (e.g. Canada, Caribbean, Brazil, US) • American-Latin: people sharing the latino culture whose area of origin is within the Americas (e.g. Mexico, South-America, US etc.) • Asian-West: area of origin e.g. Bangladesh, India, Iran, Iraq, Pakistan etc. • Asian-East: area of origin e.g. China, Japan, Korea etc. • mixed: please indicate as precisely as possible in form of a comment (e.g. asian-white, black-white, mestizo etc.) • other: please indicate as precisely as possible in form of a comment (e.g. aboriginal North-America, aboriginal Australia, semitic etc.)
Professional qualification	<p>The highest qualification/degree obtained by attending school/university/any other formal training institution should be marked (irrespective of the length of time required/the ways chosen to achieve the respective professional qualification).</p> <p>Note: yos = years of schooling (regular duration of schooling regardless of actual years spent in school by the respective individual).</p>
Years of education	<p>Will be filled in automatically based on the entries in the field 'professional qualification'.</p> <p>Note: if 'other degree' is checked, the years of education will be set to '99' - please fill in a realistic, estimated number of years on the</p>



EUROPEAN HUNTINGTON'S DISEASE NETWORK

	comment page.
Handedness	Relies on the self-report of your participant. Note: if a more reliable assessment is desired, please use the Edinburgh-Inventory (supplied as pdf). If your assessment is based on information derived from the Edinburgh Inventory (R.C. Oldfield, 1970), please indicate this fact as a comment .

Guidelines of Form Medical History

Label	Instructions
Date data obtained	Enter the date you obtained the pertinent information . The date you enter data onto the eCRF is stored automatically - you therefore do not need to record it.
Is the participant naturally or surgically sterile?	male or female
HD diagnosed	Please enter date of clinical diagnose, not of the genetical.

Guidelines of Form CAG Analysis

Label	Instructions
Date data obtained	Enter the date of the Genetics Laboratory report on the repeat sizes. The date you enter data onto the eCRF is stored automatically.

Guidelines of Form General Variable Items

Label	Instructions
Weight	Please measure (if possible) the actual weight rather than rely on the self report of the participant. Note: weight is recorded in kg (not pounds!).
Height	Please measure (if possible) the actual height rather than rely on the self report of the participant. Note: height is recorded in cm (e.g. 174 cm - not in feet or inches!).
BMI	Calculated automatically
Occupation	Please indicate as precisely as possible using the self-report of your participant (i.e. in her/his native language). Note: refers to the type of occupation held during most of her/his professional career.
Employment	Note: <ul style="list-style-type: none">• 'not employed' refers to people who do not hold a gainful employment at the time of interview for reasons other than being laid off, e.g. househusband/housewife.• 'unemployed' refers to people who were laid off and who are seeking gainful employment.• 'partially unemployed' refers to people whose



EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

	employers temporarily reduce working hours and consequently pay less due to shortage of orders instead of laying people off.
Residence	Self-reported categories of residence. Rough guidelines: - village : 0 - 5.000 residents - town : 5.000 - 50.000 residents - city : 50.000 - 10.000.0000

Guidelines of Form Medication Log

Label	Instructions
Drug name	You can either enter the proprietary name (= trade name) used in your respective country or the generic name. Periodically, the drug name will be coded in WHO-DD terminology by language or central coordination.
Indication1	The disorder representing the indication for use of a compound can be given in English or in your native language . Please make sure that you describe the indication as precisely as possible , by making use of the 'comment' field. Periodically, your entries will be coded using the ICD10 terminology by language/central coordination.
Dose/Unit	Enter dose and unit separately, e.g. 100 and mg in the fields provided.
Frequency	Enter 102 to indicate that a drug is prescribed e.g. as 1 tablet in the morning, 0 tablets at noon and 2 tablets in the evening. Use more than 3 numbers if a compound is given more than 3 times a day. To indicate a prescription of 0.5 tablets in the morning, at noon and in the evening, the dosage needs to be adopted accordingly e.g. 0.5 tablets in the morning, 0.5 tablets at noon and 0.5 tablets in the evening of 100 mg should be entered as 111 of 50 mg (dosage/unit).
Route	p.o. - per os p.r. – rectal s.c. – subcutan i.m. – intramuscular i.v. – intravenous td - transderma/topical sl – sublingual inh - inhaled
Stop date	No entry in this field implies that this medication is ongoing. Therefore please review all entries at each visit and enter end dates if appropriate.



Guidelines of Form Comorbid Conditions

Label	Instructions
Concomitant disorders	Concomitant disorders can be given in English or in your native language. Please make sure that you describe the concomitant disorder as precisely as possible, e.g. by making use of the comment field in addition. Periodically, your entries will be coded using the ICD10 terminology by language/central coordination.
End date	No entry in this field implies that the condition is ongoing. Therefore please review all entries at each visit and enter end dates if appropriate.

Guidelines of Form Huntington's Disease Rating Scale '99 - Motor Assessment

Label	Instructions
Date data obtained	Enter the date you perform the examination. The date you document the examination (=enter information in the eCRF) is stored automatically - you therefore need not to take it down/record it.
Motor score	Will be calculated automatically.
Ocular pursuit	Should be assessed over a range of approximately 20° with a target passing slowly at $\leq 10^\circ$ per second, which corresponds to about 2 seconds for moving an object from one shoulder to the other.
Saccade initiation	Should be tested over a 20° range, as for ocular pursuits. Saccade movement should be elicited by a sound (snapping fingers) or movement (wiggle fingers), but not by a verbal command to look to the right or left.
Saccade velocity	Should be tested at a larger range of approximately 30° so as to be able to detect incomplete range.
Tongue protrusion	Suggestion: Please ask your participants to open their mouthwide while you inspect it using a torch. Then ask your participants to protrude their tongue well beyond their front teeth while keeping their mouth wide open and to keep it out as long as it takes you (as the examiner) to count aloud from 1 to 10. Participants should be made aware that they are not allowed to prevent their tongue from slipping back into the mouth by biting on it.
Finger taps	Participant taps thumb with index finger in rapid succession with widest amplitude possible, each hand separately.
Pronate/supinate-hands	Requires the participant to alternately hit the palmar and dorsal surface of one hand against the palm of the opposite hand. Use the palm of the opposite hand as a target instead of some other surface such as the participant's leg or the table



EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

	surface. The participant should do this task as quickly as possible over a five-second interval. The task is graded according to the degree of slowing and irregularity.
Luria	Fist-hand-palm sequencing - Say 'Can you do this?' Examiner puts hand into fist on flat surface (or in lap) and sequences as follows: fist, side, flat (DO NOT REPEAT THIS OUT LOUD). Watch to make sure that participant can mimic each step. Continue to practice Luria 3-step for 1 - 2 minutes. When participant is able to join you then say 'Very good, now keep going, I am going to stop.' Rest hand and start timing participant's sequences. A sequence is considered correct only if it is unaided by examiner model and in the correct order. Count completed sequences and score. If participant was unable to complete any sequences over a 10-second period, then continue as follows. Say 'Now let's try it again. Put your hands like this. FIST; SIDE; FLAT'. Watch to make sure the participant can mimic each step. Using the verbal labels, begin the sequences again and ask the participant to 'Do as I do, Fist, Side, Flat' (repeat this as you continue). Continue to perform Luria 3-step. When participant is able to join you say 'Very good, now keep going, I am going to stop'. Rest hand and start timing participant's sequences. A sequence is considered correct if it is unaided by examiner model and in the correct order. Count completed sequences and score as above.
Rigidity-arms	Rigidity is judged on passive movement of the arms with the participant relaxed in the sitting position.
Bradykinesia-body	Observe the participant during spontaneous motion such as walking, sitting down, arising from a chair, and executing the tasks required during the examination. This rating reflects the examiner's overall impression of bradykinesia.
Maximal dystonia	Maximal dystonia is defined here as a tendency toward a posture, posturing along an axis. Observe the participant during the examination; i.e., no particular maneuvers are required to illicit these features. Maximal dystonia are typically observed during demanding motor tasks such as tandem gait. When rating dystonia facial dystonia (blepharospasm, jaw opening and closing) should be included in your assessment of the truncal region. Please indicate in a comment what subtypes of dystonia (blepharospasm, torticollis) you included in your rating of truncal dystonia. RUE refers to right, LUE to left upper extremity, RLE to right, LLE to left lower extremity.
Maximal chorea	Maximal Chorea is defined here as movement, not posture. Observe the participant during the examination; i.e., no particular maneuvers are required to illicit these features. Maximal chorea is typically observed during demanding motor tasks such as tandem gait. Chorea is rated by specific regions. BOL refers to buccal-oral-lingual, RUE to right, LUE to left upper extremity, RLE to right, LLE to left lower extremity. Please comment whether the chorea you observe is more distal or more proximal (e.g. distal much more than proximal).
Gait	Observe the participant walking approximately 9 meters (10 yards) as briskly as they can, then turning and returning to the starting point.
Tandem walking	The participant is requested to walk ten steps in a straight line



EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

	with the foot placed (accurately but not quickly) such that the heel touches the toe of the other foot. Deviations from a straight line are counted.
Retropulsion pull test	The participant's response to a sudden posterior displacement produced by a pull on the shoulder while the participant is standing with eyes open and feet slightly apart is assessed. The shoulder pull test must be done with a quick firm tug after warning the subject. The participant should be relaxed with feet apart and should not be leaning forward. If the examiner feels pressure against his/her hands when placed on the participant's shoulders, the examiner should instruct the participant to stand up straight and not lean forward. The examiner should instruct the participant to take a step backward to avoid falling. Examiners must catch subjects who begin to fall.

Guidelines of Form Huntington's Disease Rating Scale '99 - Behavioral Assessment

Label	Instructions
Date data obtained	Enter the date you obtained the pertinent information. The date you enter data on the eCRF is stored automatically - you therefore do not need to record it.
Behavioural score	Will be calculated automatically.
Behavioral Assessment	Instructions: Please rate the frequency and severity of the behavior for the past month. Frequency and severity should be uniquely assessed as independent qualifiers of positively affirmed behavioral symptoms. Frequently occurring behaviors do not indicate a high severity rating (e.g., presence of anxiety may be constant but mild in impact). Severity is indicative of the behavior's impact on the individual's ability to carry out daily activities. ["I will now ask you about experiences that most individuals have at some point in life. I will ask you to help me gauge how frequently and severely these experiences occur."]
Depressed mood	Please ask: In the past month have you been feeling sad (or down or blue)? Has your mood affected your daily activities? Have you found yourself doing something you would ordinarily enjoy and realized you were not having any fun? Evidence of sad mood from behavioral observation includes sad voice or expression, tearfulness.
Low self-esteem/guilt	Please ask: In the past month have you been feeling badly about yourself? Have you found yourself thinking or saying that you are a failure, or blaming yourself for things? Evidence of low self-esteem/guilt includes self-blame without justification, self-deprecation including feelings of being a bad or unworthy person, feeling like a failure.
Anxiety	Please ask: In the past month have you found yourself feeling worried about things? Evidence of anxiety includes worrying, panic, feeling frightened or fearful for no apparent reason.
Suicidal thoughts	Please ask: Since your last visit, have you found yourself thinking that life is not worth living or that you would be better off dead? Have you thought about hurting yourself or killing yourself? Are you planning to hurt yourself or kill yourself?



EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

	Have you taken any steps toward carrying out your plan?
Disruptive or aggressive behavior	Please ask: Since the last visit, have you had any emotional or temper outbursts? Have you had times when you lost control of yourself? Have you hit or shoved or thrown things or expressed your temper in a physical way? Have you used threats or hostile words? This item is used to rate loss of temper and impaired self-restraint. Threatening behavior includes physical violence or aggression, verbal outbursts, threatening, foul or abusive language.
Irritable behavior	Please ask: In the past month, have you felt impatient? Do you behave in a demanding way? Do others say you behave in a demanding way or have a short fuse or are overly sensitive? Note that this item is used to rate the ease with which the subject loses his/her temper rather than how extreme the behavior is once self-control is lost.
Perseverative/obsessional thinking	Please ask: Within the past month, have you found yourself getting stuck on certain ideas? Within the past month, have you been bothered by thoughts, images or fears that keep coming back even if you try not to have them? This item is used to rate inflexibility or perseveration of thinking.
Compulsive behaviour	Please ask: In the past month, have you found yourself doing certain things over and over again? Are you unable to resist doing some of these things? For example, do you wash your hands again and again, or count up to a certain number, or check that the door is locked over and over to make sure that you have done it correctly? This item is used to rate repetitive, purposeful, and intentional behaviors.
Delusions	Please ask: I'm going to ask you about unusual experiences that people sometimes have. Since the last visit, has it ever seemed like people are out to get you or perhaps are controlling you? Has it seemed like you have any special powers or importance, or that books, TV, and radio statements are referring to you? Are there any other unusual things you experience that I have not asked about? Delusions are fixed false beliefs that are not culturally shared.
Hallucinations	Please ask: Since the last visit, have you heard things that other people could not hear, such as noises or the voices of people whispering or talking? Did you ever have vision or see things that other people could not see? How about any other strange sensations in your body: skin, smell, or taste? Hallucinations are perceptions without a physical stimulus (e.g., hearing voices when no one is in the room).
Apathy	Please ask: Within the past month, have you found that you have lost interest in things that used to be important to you? Do you sit around a lot doing nothing? Are you just as interested as always in trying new things, starting new projects? Apathy is a lack of interest or emotional involvement in things, and can be distinguished from anhedonia which refers to inability to experience pleasure. Apathy is reflected behaviorally by neglecting hygiene, being inactive, sitting around doing nothing, doing nothing unless told to do it by someone else, saying little in conversation, failing to initiate conversation. This question should definitely be addressed to an informant if possible.
Behavioral Milestones	These items assess whether the participant has reached certain behavioral milestones.



EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

Does the examiner believe the participant is confused?	Confusion is defined as intermittent or persistent disorganized thinking, perceptual disturbances or disorientation to time, place, or person.
Does the examiner believe the participant is demented?	Dementia is defined as progressive impairment in memory, abstract thinking or judgement that interferes with work or usual social activities and relationships.
Does the examiner believe the participant is depressed?	Depression is defined as persistent depressed mood, anhedonia, or vegetative signs sufficient to interfere with daily functioning.

Guidelines of Form Huntington's Disease Rating Scale '99 - Functional Assessment

Label	Instructions
Date data obtained	Enter the date you obtained the pertinent information. The date you enter data on the eCRF is stored automatically - you therefore do not need to record it.
Functional Assessment Score	Will be calculated automatically
Could subject engage in gainful employment in his/her accustomed work	If the subject is no longer able to work at the job he/she had for the majority of his/her life, answer 'no'. For example, if the person worked in a fast food chain as a cashier and after developing HD was forced to leave that job and worked in a less demanding job, the answer would be 'no' to gainful employment in accustomed work. If the subject is a homemaker who never worked for pay, the probe for this person might be: 'Can the subject manage the household today as well as he/she always has or must they have assistance to do so?' If assistance is now required, the answer would be 'no'.
Could subject engage in any kind of gainful employment?	Gainful employment means that the person is paid for their services. This is judged as potential capacity, not whether the person is actually working.
Could subject engage in any kind of volunteer or non-gainful work?	Volunteer or non-gainful work means the person is not paid for their services.
Could subject manage his/her finances (monthly) without any help?	An informant or a subject may report that the subject has always had difficulty managing monthly finances without any help. To help to determine whether the subject could perform this activity unassisted, the probe might be: 'Compared to today, do you think he/she could have managed the monthly finances better a year ago?' Alternatively, the probe could be 'Do you think he/she could have managed the monthly finances better before he/she had some of the symptoms/signs of HD?' these probes which highlight change in function may help to determine the subject's capacity to perform at the present time.
Could subject shop for groceries without help?	Shopping for groceries without help means going into the store obtaining groceries without assistance. If the subject requires help carrying bundles, but can otherwise handle the task, the answer is 'yes'.
Could subject handle money as a purchaser in a simple cash (shop) transaction?	The person should be able to go to a shop and come back with the correct change.
Could subject supervise children without help?	Supervising children means physically as well as cognitively caring for children who could not otherwise be left alone. This



EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

	does not mean infants.
Could subject operate an automobile safely and independently?	Operating an automobile safely and independently means the subject can drive without others feeling afraid to drive with the subject and showing good judgment. If the person has never learned how to drive, please file a comment indicating 'Not applicable'.
Could subject do his/her own housework without help?	Housework activities might include cooking, vacuuming, dusting, taking out the rubbish, and doing dishes. If a subject never did any housework, ask about picking up after themselves (e.g., doing light dusting or making the bed) and hanging up his/her clothes. Housework might also extend to light gardening if that was the subject's responsibility.
Could subject do his/her own laundry (wash/dry) without help?	If the subject only folds laundry and does nothing else, the answer is 'no'.
Could subject prepare his/her own meals without help?	Preparing meals can include making a sandwich, heating up soup or using the microwave, as long as the person does it himself/herself. A probe might be 'if the subject were left alone, would he/she able to prepare his/her own meals?'
Could subject use the telephone without help?	Using a telephone without help means the ability to make outgoing calls and answer the telephone
Could subject take his/her own medications without help?	If the subject has the pills in a dispenser but he/she is able to remember to take them by himself/herself, then the answer is 'yes'. If the subject cannot physically handle medications without assistance, the answer is 'no'.
Could subject feed himself/herself without help?	If the subject cannot cut his/her own food without assistance, then the answer to ability to feed himself/herself without help is 'no'.
Could subject dress himself/herself without help?	If the subject must have clothes laid out, but he/she can dress properly (i.e., enough to be presentable), the answer is 'yes'.
Could subject bathe himself/herself without help?	If the subject requires assistance getting into the shower/tub, but then bathes himself/herself, the answer is 'yes'.
Could subject use public transportation to get places without help?	Public transportation includes bus and train. If there is no public transportation the question should be; 'If public transportation were available, could he/she use it without assistance?' en-us Public transportation includes bus and train. If there is no public transport, the question should be; 'If public transportation were available, could he/she use it without assistance?'
Could subject walk to places in his/her neighbourhood without help?	Walking to places in the neighborhood without help implies not getting lost. A probe might be 'would he/she be able to find his/her way home if he/she was out on one of the streets in the neighborhood?'
Could subject walk without falling?	Falling should occur at least once a week for a 'no' answer. A one-time fall does not indicate a 'no' answer.
Could subject walk without help?	Required use of a walker or a cane is 'help'. In other words, if the subject cannot walk without an assistive device, the answer is 'no'.
Could subject's care still be provided at home?	Care at home implies only whether the person is capable of living at home, rather than in the equivalent of institutional care.
Subject's independence in %	Independence is given as percentage of normal in five percent graduations; each bullet indicates a five percent increment. If you select a bullet, the percentage will appear in the field Independence Scale.



Guidelines of Form Huntington's Disease Rating Scale '99 - Functional Capacity

Label	Instructions
Date data obtained	Enter the date you obtained the pertinent information. The date you enter data on the eCRF is stored automatically - therefore you do not need to record it.
Functional score	Will be calculated automatically.
Occupation	The participant's capacity to engage satisfactorily in gainful or voluntary works is assessed regardless of whether or not the participant is actually working. Normal refers to gainful employment, actual or potential, with usual work expectations. Reduced capacity refers to full or part-time gainful employment with lower than usual work expectations (relative to the participant's training and education), but with satisfactory performance. Marginal refers to a capacity only for part-time employment, actual or potential with low work expectations. Unable refers to a participant who would be unable to work, even with considerable assistance and oversight.
Finances	Assessed by surveying the participant's involvement in personal and family finances including balancing a checkbook, paying bills, budgeting, shopping, etc. Normal capacity refers to satisfactory handling of these basic financial tasks. Requires slight assistance refers to mild difficulties which would require the assistance/supervision of a family member or financial advisor. Requires major assistance refers to a participant who would require extensive supervision in handling routine financial tasks. Unable refers to a participant who would be unable to carry out these financial tasks, even with considerable assistance and supervision.
Domestic chores	Refers to the participant's capacity to carry out routine domestic tasks such as cleaning, laundering, dishwashing, table setting, cooking, lawn care, answering mail, maintaining a calendar, etc. Normal capacity refers to a full capacity without assistance. Impaired refers to impaired capacity requiring only slight assistance or supervision. Unable refers to marked incapacity requiring major assistance.
ADL	Refers to the traditional areas of 'activities of daily living' (ADL) including eating, dressing and bathing. Normal refers to full capacity. Minimal impairment refers to impaired capacity requiring only slight assistance. Gross tasks only refers to impaired capacity requiring moderate assistance and supervision. Total care refers to major incapacity requiring total assistance and supervision.
Care level	Refers to the most appropriate care environment to meet the participant's capacity, whether at home , at home or chronic care facility or full skilled nursing care (24 hours a day supervision).



Guidelines of Form Huntington's Disease Rating Scale '99 - Cognitive Assessment

Label	Instructions
Date data obtained	Do not enter! Will be calculated automatically
Verbal fluency test	Participants are asked to produce as many words as possible beginning with a specified letter . Please download and print a pdf-file with stimuli appropriate for your language and write down all words given by the participant in the order they are produced, even if they are incorrect or repetitions. Time: one minute
Symbol digit modality test	Please download and print a pdf-file with stimuli and hand the sheets to the participants. Participants are given 90 seconds to work on the task. Instructions: <i>'Please look at the boxes at the top of the page. Each box in the upper row has a symbol in it, and each box below it has a number. Now look at the next line of boxes (point to the first line of boxes without numbers). Notice that the boxes on the top have symbols, but boxes beneath are empty. You are to fill in each empty box with the number that goes with each symbol according to the way they are paired at the top of the page. For example, if you look at the first symbol (point to the first symbol in the row beneath the key), and then look up at the key, you see that his symbol is paired with number 'one' (show the pairing). So you would write a 'one' in this box (write a '1' in the first box). This next symbol (point to the next symbol) is paired with five. So you would put a 'five' in this box (write '5' in the second box). Now what number goes in this box (point to third box)?'</i> When the participant appears to comprehend the task, say, <i>'Good, now for practice, fill in the boxes up to this double line, and then stop'</i> Correct immediately any errors made during the practice period, explaining the participant's error. Repeat the instructions and review the correct coding of the practice boxes as necessary until the participant understands the task. Do not administer the remainder of the test if a participant can not complete any of the practice items. Continue with the test by saying, <i>'When I say 'go' write in the numbers just like you have been doing as fast as you can until I say 'stop'. Work as quickly as you can moving from one line to the next without skipping any boxes. If you make a mistake, don't erase it. Just write the correct answer over the mistake. Remember to work as quickly as you can. Ready? Go!'</i> Start timing. At the end of 90 seconds, say <i>'Stop!'</i> Be sure that the participant does not continue working after the time limit is reached. Do not allow the participant to skip any boxes. Scoring: The score is the number of correct responses in 90 seconds. Do not include the practice sample in the total score.
Colour naming	Time: 45 seconds Instruction: <i>'Please read across the top line, naming the colors you see, either red, green, or blue'. Occasionally a participant</i>



EUROPEAN **HUNTINGTON'S DISEASE** NETWORK

	<p>will incorrectly identify a color (e.g., call a blue spot 'purple'). Indicate to the participant that the three colors used in the test are red, green and blue. If the participant cannot discriminate the colors, terminate this test. Continue by pointing to the second line and say, <i>'Begin here, and go across the rows from left to right without skipping any. Tell me the colors as quickly as you can. Go!'</i> Begin timing, stop after 45 seconds.</p> <p>Scoring: Count the total number of words correctly read.</p>
Word reading	<p>Time: 45 seconds</p> <p>Instruction: <i>'Please read across the top line, reading the names of the colors ('red', 'green', 'blue') that appear in black print'. If a participant cannot read, terminate this test. Continue by pointing to the second line and say, 'Begin here, and go across the rows from left to right without skipping any. Read the names of the colors as quickly as you can. Go!'</i> Begin timing, stop after 45 seconds.</p> <p>Scoring: Count the total number of words correctly read.</p>
Interference	<p>Time: 45 seconds</p> <p>Instructions: <i>'This card has words written in colored ink, but you can see that each word is in the wrong color of ink. For example, here the word 'red' is written in blue ink (point to the first word of the top line). Please read across the top line, telling me the color of ink that each word is written in. Ignore the words, just tell me the color of ink you see'. Additional review of the instructions to name ink colors and not read the words may be necessary. When it is clear that the participant understands, continue by pointing to the second line and say, 'Begin here, and go across the rows from left to right without skipping any. Remember to ignore the words, and simply tell me the colors of ink that you see. Go!'</i> Begin timing, stop after 45 seconds.</p> <p>Scoring: Count the total number of words correctly read.</p>

Guidelines of Form Huntington's Disease Rating Scale '99 - Clinical Summary

Label	Instructions
Date data obtained	Enter the date you obtained the pertinent information. The date you enter data on the eCRF is stored automatically - therefore you do not need to record it.
Do you believe that this participant has manifest HD?	With a confidence level $\geq 99\%$ and based on UHDRS (Motor, Cognitive, Behavioral, Functional components)