

Motor Neuron Disease Register for England, Wales and Northern Ireland: Comprehensive data set and definitions.

Free text and user defined fields will be added on request to facilitate local data collection of extra items such as detailed phenotype or medication information. Those highlighted in grey are included in the SOPHIA dataset and the definitions are compatible.

No	Data item	Options/ data definition
1	Unique ID number	Used to link information in patient identifiable database with information in clinical information database
2	First Name	
3	Surname	
4	NHS Number	
5	Patient/control	
6	Date of birth	
7	Date of death	
8	Notes	
9	Gender	Gender at birth
10	Ethnicity	<u>White</u> A British, B Irish, C Any other White background <u>Mixed</u> D White and Black Caribbean, E White and Black African, F White and Asian, G Any other mixed background <u>Asian or Asian British</u> H Indian, J Pakistani, K Bangladeshi, L Any other Asian background <u>Black or Black British</u> M Caribbean, N African, P Any other Black background <u>Other Ethnic Groups</u> R Chinese, S Any other ethnic group, Z Not stated National code Z - Not Stated should be used where the PERSON has been given the opportunity to state their ETHNIC CATEGORY but chose not to.
11	Post code of residence at diagnosis	If consent for the inclusion of identifiable data is not obtained, only the postcode sector will be included. Eg from the post code SE5 8AF we would record SE5 8

12	Source of ascertainment	Options include: MND Care and Research Centre, Specialist Neurology Outpatient Clinic, General Neurology Outpatient Clinic, Clinical Nurse Specialist, Community Multidisciplinary team, Hospice, Other
13	Date of diagnosis	Date that the diagnosis was first discussed with patient, by the ascertaining centre.
14	Diagnosis	Type of motor neuron disease
15	Co-morbidity	Available options in drop down menus: Cancer, Cardiovascular disease, Parkinson's disease, Psychiatric disorder(s), Auto-immune disorder(s), Absent/unknown Free text box available for: Co-morbidity, other Two co-morbidities can be indicated
16	Date of symptom onset	Reported by the patient when: – Limb symptoms: muscle weakness was noticed – Speech/swallowing difficulties – Respiratory symptoms: dyspnea was noticed – Generalized symptoms In most cases middle of period: – E.g. January 2014 -> 15 January 2014 – In 2014 -> 1 July 2014 – Winter (spring, summer, autumn) 2014 -> 1 February 2014 (May, August, November) Except for: – Beginning of 2014 -> 1 January 2014 – End of 2014 -> 31 December 2014 – Patient seen on 23 July 2014: "Five months ago..." -> 23 February 2014 – Patient knows exact date
17	Site of onset	<ul style="list-style-type: none"> • Site of onset: Bulbar, Spinal, Thoracic/respiratory, Generalized (e.g. both swallowing issues and weakness in leg reported at the same time) • If Spinal, then select: Arms, Legs (dropdown box); and select: left, right (dropdown box) • Other first symptoms: <ul style="list-style-type: none"> – Weight loss (checkbox) (>10% weight loss in 6 months prior to time of disease onset as defined by date of onset symptoms) – Cognitive or behavioural (checkbox)
18	Limb dominance	Leg (L/R), Arm (L/R)
19	Current number of regions affected	Speech, swallowing, Hand (L/R), arm (L/R), foot (L/R), leg (L/R) trunk, breathing.

20	Cognitive impairment	Yes/No (and details of formal assessment where applicable)
21	EI Escorial category	EI Escorial categories: Suspected ALS, Possible ALS, Probable ALS lab supported, Probable ALS, Definite ALS The dropdown options for EI Escorial contain both the old and the revised EI Escorial options. This way data of patients assessed with the old EI Escorial criteria can be added just like data of patients with the revised EI Escorial criteria, in one data field. This simplifies the database set-up and data analysis. If a case of suspected ALS (= PMA) progresses to ALS, the diagnostic pathway can be traced
22	EI Escorial date	
23	ALS-FRS	<ul style="list-style-type: none"> •Date of ALS-FRS-R (date field) •ALS-FRS-R 12 sub scores(number fields) •ALS-FRS-R total score (number field) •ALS-FRS-R total score (automatically calculated) (If no ALS-FRS-R sub scores are available, then fill in the total score at the end of the table only. The data field for ALS-FRS-R total score (automatically calculated) will fill automatically with either this manually input score or the score based on the sub scores that were put in. This is also the data fields which should be used when extracting the data/running queries.) <ul style="list-style-type: none"> •Rater name (free text field)
24	Forced vital capacity	<ul style="list-style-type: none"> • Forced Vital Capacity table (preferred): <ul style="list-style-type: none"> – Date FVC (date field) – FVC (litres) (open text field) – FVC (% of predicted) (open text field) – Testing not performed or not feasible: Not tested, Not feasible (dropdown box) Follow-up data possible by adding multiple rows
25	Sniff Nasal Inspiratory Pressure	<ul style="list-style-type: none"> • SNIP table (optional): <ul style="list-style-type: none"> – Date SNIP (date field) – SNIP (cm H2O) (open text field) – Testing not performed or not feasible: Not tested, Not feasible (dropdown box) Follow-up data possible by adding multiple rows
26	Family history	Family history – Number of affected family members with ALS/MND and FTD/Dementia <ul style="list-style-type: none"> • First degree (Parents, brothers, sister, sons, daughters): <ul style="list-style-type: none"> – ALS/MND (number field) – FTD/Dementia (number field) • Other family members: <ul style="list-style-type: none"> – ALS/MND (number field)

		<ul style="list-style-type: none"> - FTD/Dementia (number field) • Notes family history (free text field)
27	End point	<ul style="list-style-type: none"> Endpoints: • Date of death (date field) • If tracheostomy, date of tracheostomy (date field) • If >23h NIV/day, date of endpoint (date field) • Date of checking endpoint (if not deceased, no tracheostomy and no >23h NIV/day) (date field)
28	ECAS cognitive screen	
29	ECAS behavioral screen	
30	Height (cm)	
31	Weight (kg)	
32	Medications	
33	Gastrostomy	
34	Respiratory intervention	