

Table e-1. Schedule of procedures for diagnosis and research outcomes

		Baseline	6 mo	12 mo	18 mo	24 mo	30 mo	36 mo
Diagnostic Tests	History	X	X		X		X	
	Family History	X						
	Caregiver interview	X	X		X		X	
	Neurological Exam	X	X		X		X	
	EMG	X	X*		X*		X*	
	MiniMental State Exam ¹	X	X		X		X	
	Frontal Assessment Battery ²	X	X		X		X	
	Mattis Dementia Rating Scale -2 ^{3, 4}	X	X		X		X	
	Research Outcome Measures							
Motor	ALSFRS-R⁵	X	X	X	X	X	X	X
	Timed gait, timed reading, finger and foot tapping, 9-hole peg test	X	X		X		X	
	Physiology: TMS, EIM	X	X		X		X	
Cognitive	Letter Fluency Score	X	X	X	X	X	X	X
	DRS, FAB, MMSE, D-KEFS Trails, Sorting, Fluency, 20 Questions ⁶	X	X		X		X	
	Language: ADRC FTLD module ⁷	X	X		X		X	
Behavioral	Frontal Behavioral Inventory⁸	X	X	X	X	X	X	X
Mood, Pseudo-bulbar affect	ALS depression Inventory, ⁹ CNS lability ¹⁰	X	X		X		X	
Quality of Life	SF-8 Health Survey ¹¹	X	X		X		X	
Imaging	MRI							
Biospecimens	CSF, blood	X	X		X		X	
	Skin biopsy	X	X**					

*EMG repeated when EI Escorial certainty less than definite

** Skin Biopsy performed only once, usually first, occasionally second visit

References

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