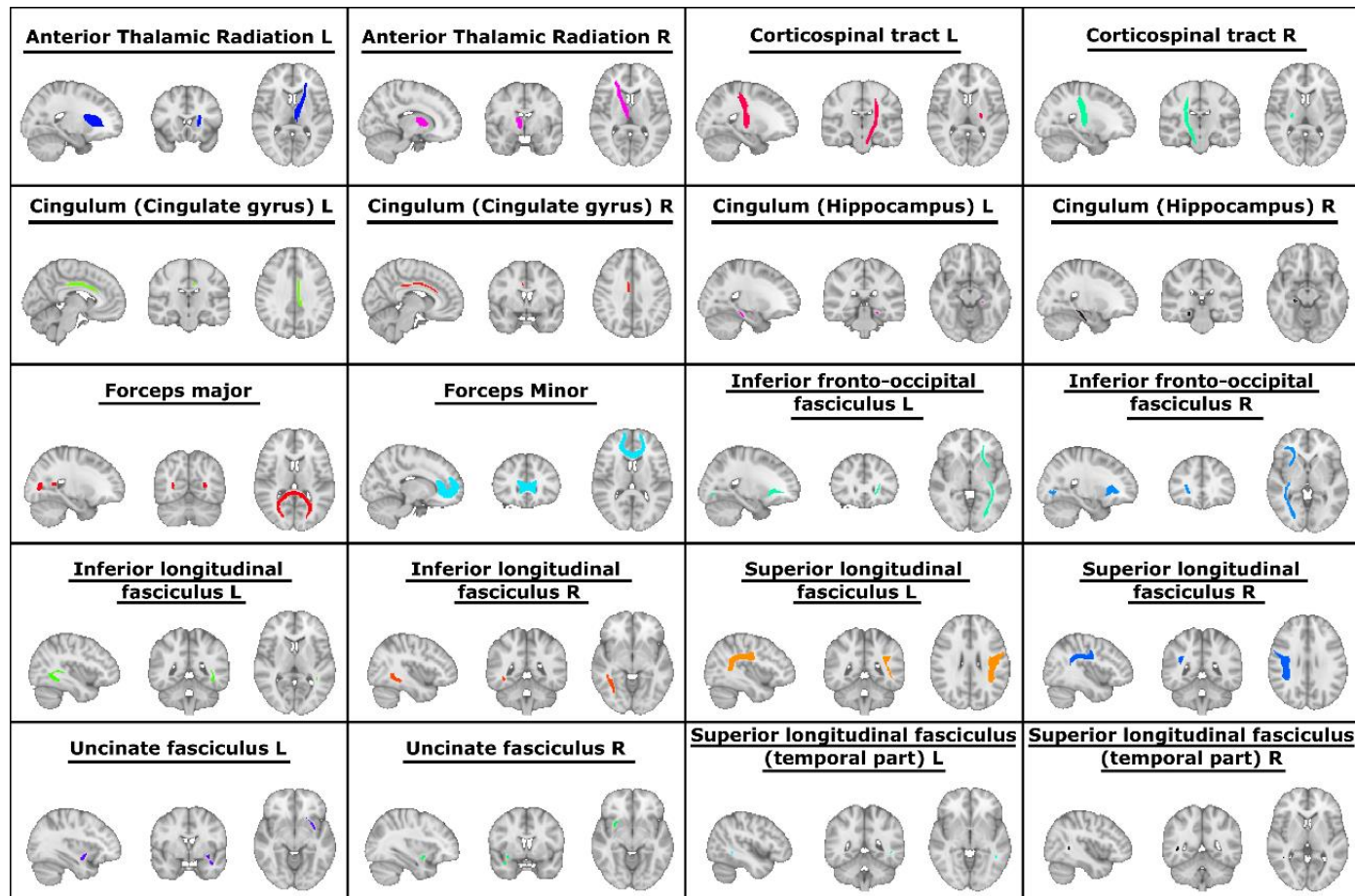


Supplementary material

White matter integrity changes and neurocognitive functioning in adult-late onset DM1: a follow-up DTI study

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Supplementary Figure 1: Major tracts used for assessing white-matter alterations in DM1.

Supplementary Table 1. Analysis of neuropsychological performance from baseline to follow-up in DM1 patients.

Domain	Baseline		Follow-up		<i>T</i>	<i>p</i>	<i>d</i>
	Mean	(SD)	Mean	(SD)			
Attention/Processing speed	45.13	(5.05)	39.61	(4.26)	2.74	.029	1.37
Verbal memory	50.69	(11.14)	45.64	(8.98)	1.15	.289	0.57
Visual memory	47.50	(9.75)	46.75	(8.38)	0.19	.857	0.09
Visuoconstruction	49.81	(9.45)	47.75	(8.25)	0.69	.513	0.34
Executive functioning	45.45	(7.75)	42.06	(9.64)	1.42	.198	0.71
Language	48.81	(11.04)	50.88	(3.37)	-1.11	.302	.557
Intellectual Functioning	97.63	(11.54)	100.00	(10.53)	-0.79	.458	0.39

Note. SD: Standard Deviation

Supplementary Table 2. Group differences in WM integrity FA values.

	Intergroup differences		Post hoc analyses							
	χ^2	p	DM1 baseline vs HC baseline		DM1 follow-up vs HC follow-up		HC baseline vs HC follow-up		DM1 baseline vs DM1 follow-up	
			W	P	W	P	W	p	W	p
Anterior Thalamic Radiation	192.67	0.000	64	0.034	77	.000	--	--	36	.008
Corticospinal Tract	--	--	--	--	71	.004	--	--	33	.039
Cingulum (Cingulate Gyrus)	111.60	0.010	--	--	67	.016	47	.049	35	.016
Cingulum (Hippocampus)	100.98	0.020	--	--	69	.009	--	--	36	.008
Forceps Major	93.07	0.030	--	--	68	.012	--	--	35	.016
Forceps Minor	127.54	0.005	--	--	70	.006	--	--	36	.008
Inferior Fronto-occipital Fasciculus	18.70	0.000	67	0.016	77	.000	--	--	36	.008
Inferior Longitudinal Fasciculus	161.14	0.001	69	0.009	72	.003	--	--	36	.008
Superior Longitudinal Fasciculus	174.02	0.001	63	0.043	74	.001	49	.027	36	.008
Uncinate Fasciculus	159.39	0.001	73	0.002	69	.009	--	--	--	--
Superior Longitudinal Fasciculus (Temporal Part)	95.79	0.020	--	--	--	--	--	--	36	.008
Global FA	173.28	0.001	--	--	74	.001	47	.049	36	.008

Note. Only results where statistically significant differences were found are shown (remaining cells are left blank). DM1: Myotonic Dystrophy Type 1; HC: Healthy controls; FA: Fractional Anisotropy. P values > 0.01563 did not survive to FDR multiple-comparison correction.

Supplementary Table 3. Group differences in WM integrity MD values.

	Intergroup differences		Post hoc analyses							
	χ^2	p	DM1 baseline vs HC baseline		DM1 follow-up vs HC follow-up		HC baseline vs HC follow-up		DM1 baseline vs DM1 follow-up	
			W	P	W	p	W	p	W	p
Anterior Thalamic Radiation	17.2	.001	13	.016	2	.000	48	.037	--	--
Corticospinal Tract	8.8	.033	--	--	11	.009	--	--	--	--
Cingulum (Cingulate Gyrus)	9.1	.027	17	.043	15	.027	--	--	--	--
Cingulum (Hippocampus)	11.8	.008	--	--	16	.034	49	.027	--	--
Forceps Major	11.6	.009	--	--	10	.006	49	.027	--	--
Forceps Minor	13.5	.004	14	.021	8	.003	--	--	3	.039
Inferior Fronto-occipital Fasciculus	22.2	.000	14	.021	1	.000	55	.002	--	--
Inferior Longitudinal Fasciculus	21.7	.000	14	.021	1	.000	55	.002	--	--
Superior Longitudinal Fasciculus	14.4	.002	14	.021	7	.002	--	--	--	--
Uncinate Fasciculus	15.1	.002	12	.012	7	.002	--	--	--	--
Superior Longitudinal Fasciculus (Temporal Part)	13.7	.003	12	.012	12	.012	--	--	--	--
Global MD	15.4	.002	--	--	5	.001	--	--	--	--

Note. Only results where statistically significant differences were found are shown (remaining cells are left blank). DM1: Myotonic Dystrophy Type 1; HC: Healthy controls; MD: Mean Diffusivity. P values > 0.01554 did not survive to FDR multiple-comparison correction.

Supplementary Table 4. Significant regions resulting from group comparisons (DM1 < HC) at baseline.

Tract (JHU ICBM)	Peak X (MNI)	Peak Y (MNI)	Peak Z (MNI)	Peak p	Peak t	Mean t	N Voxels	Vol (mm ³)
Inferior Fronto-occipital Fasciculus R	28	16	-4	.044	4.8354	2.9123	20	28
Inferior Longitudinal Fasciculus R	42	-10	-20	.046	4.6402	3.249	14	22
Uncinate Fasciculus R	32	2	-10	.049	3.5133	2.6954	10	18

Note. Only tracts with N voxels greater than 10 are shown. L: left; R: right

Supplementary Table 5. Significant regions resulting from group comparisons (DM1 < HC) at follow-up.

Tract	Peak X (MNI)	Peak Y (MNI)	Peak Z (MNI)	Peak p	Peak t	Mean t	N Voxels	Vol (mm ³)
Anterior Thalamic Radiation L	-10	0	-2	.003	4.3372	1.9391	63	504
Anterior Thalamic Radiation R	18	18	0	.002	3.979	1.89	51	408
Corticospinal Tract L	-24	-16	6	.004	3.1946	1.7549	46	368
Corticospinal Tract R	22	-16	-2	.002	3.1426	1.9909	47	376
Cingulum (Cingulate Gyrus) L	-8	-24	32	.004	3.04	2.0445	15	120
Cingulum (Hippocampus) R	24	-24	-18	.008	4.0946	2.0253	25	200
Forceps Major	22	-80	8	.043	3.5965	1.8643	135	1080
Forceps Minor	-14	44	-10	.002	5.9171	2.1132	528	4224
Inferior Fronto-occipital Fasciculus L	-34	2	-6	.002	5.7826	2.0435	202	1616
Inferior Fronto-occipital Fasciculus R	40	-22	-10	.002	4.7534	2.0815	243	1944
Inferior Longitudinal Fasciculus L	-46	0	-16	.002	5.3999	2.183	142	1136
Inferior Longitudinal Fasciculus R	44	-8	-14	.005	5.2797	2.3102	82	656
Superior Longitudinal Fasciculus L	-38	-44	16	.002	4.9712	1.9591	237	1896
Superior Longitudinal Fasciculus R	36	-10	24	.004	4.6874	2.1655	195	1560
Uncinate Fasciculus L	-40	4	-26	.002	2.907	1.7682	38	304

Note. Only tracts with N voxels greater than 10 are shown. L: left; R: right.

Supplementary Table 6. Correlation analyses (Pearson *r*) between clinical and neuropsychological data and regional and global FA at follow-up.

Domain	Anterior Thalamic Radiation	Corticospinal Tract	Cingulum (Cingulate Gyrus)	Cingulum (Hippocampus)	Forceps Major	Forceps Minor	Inferior Fronto- occipital Fasciculus	Inferior Longitudinal Fasciculus	Superior Longitudinal Fasciculus	Uncinate Fasciculus	Superior Longitudinal Fasciculus (Temporal Part)	Global FA
Attention/ Processing speed	-0.33	-0.059	0.02	0.146	-0.44	-0.022	-0.33	-0.346	-0.182	-0.615	-0.206	-0.252
Verbal memory	-0.26	0.062	0.277	0.35	0.112	0.005	-0.126	-0.09	-0.22	-0.146	0.153	0.075
Visual memory	-0.213	0.506	0.341	0.154	0.205	-0.051	-0.076	0.107	0.157	0.191	0.029	0.269
Visuo- construction	0.733	0.581	0.663	0.692	0.772*	0.778*	0.798*	0.7	0.614	0.739	0.695	0.788*
Executive functioning	-0.638	-0.138	0.109	-0.017	-0.367	-0.252	-0.555	-0.519	-0.418	-0.564	-0.19	-0.382
Intellectual Functioning	0.032	0.45	0.839*	0.511	0.543	0.472	0.245	0.213	0.228	0.387	0.614	0.37
CTG	-0.511	-0.239	-0.18	-0.463	-0.651	-0.461	-0.705	-0.727	-0.479	-0.581	-0.67	-0.52
MIRS	-0.34	-0.475	-0.337	-0.744	-0.442	-0.442	-0.476	-0.533	-0.368	-0.237	-0.41	-0.618
WM lesion load	-0.64	-0.64	-0.28	-0.56	-0.64	-0.57	-0.79*	-0.88**	-0.8*	-0.66	-0.55	-0.76*

Note. * $p < 0.05$; ** $p < 0.01$. FA: Fractional Anisotropy;