

**On-line Table 1: Comparison of the neuropsychological test scores between the two patient subgroups (DMD proximal and distal), and healthy controls**

SN/Test	Subset	Group (I)	Group (J)	Difference (I-J)	Significance	95% CI, Lower-to-Upper Bound
1/Wechsler-Intelligence Scale for Children	Picture Completion	Control	Proximal	3.216 <sup>a</sup>	.000	1.45–4.98
			Distal	5.474 <sup>a</sup>	.000	3.93–7.02
	Information	Control	Proximal	2.257 <sup>a</sup>	.009	0.45–4.06
			Distal	2.475 <sup>a</sup>	.002	0.74–4.21
	Coding	Control	Proximal	5.337 <sup>a</sup>	.000	3.82–6.86
			Distal	2.862 <sup>a</sup>	.000	1.09–4.64
	Similarities	Control	Proximal	1.343	.468	–0.95–3.63
			Distal	3.201 <sup>a</sup>	.001	1.20–5.21
	Picture Arrangement	Control	Proximal	1.859	.169	–0.49–4.20
			Distal	2.775 <sup>a</sup>	.000	1.23–4.32
	Arithmetic	Control	Proximal	4.315 <sup>a</sup>	.000	2.96–5.67
			Distal	1.540	.059	–0.04–3.12
	Block Design	Control	Proximal	3.043 <sup>a</sup>	.000	1.24–4.85
			Distal	4.667 <sup>a</sup>	.000	3.08–6.25
	Vocabulary	Control	Proximal	1.623	.105	–0.23–3.47
			Distal	1.409	.289	–0.64–3.45
	Object Assembly	Control	Proximal	4.338 <sup>a</sup>	.000	2.54–6.13
			Distal	2.929 <sup>a</sup>	.003	0.83–5.02
	Comprehension	Control	Proximal	1.497	.054	–0.02–3.01
			Distal	3.087 <sup>a</sup>	.000	1.76–4.41
	Digit Span	Control	Proximal	1.589 <sup>a</sup>	.043	0.04–3.14
			Distal	3.541 <sup>a</sup>	.000	1.70–5.38
	Mazes	Control	Proximal	5.142 <sup>a</sup>	.000	3.53–6.75
			Distal	1.601	.123	–0.28–3.48
	AVLT1	Control	Proximal	1.771 <sup>a</sup>	.012	0.31–3.24
			Distal	1.515 <sup>a</sup>	.015	0.23–2.80
	AVLT2	Control	Proximal	–0.256	1.000	–1.76–1.24
			Distal	4.514 <sup>a</sup>	.000	2.55–6.48
	AVLT3	Control	Proximal	6.259 <sup>a</sup>	.000	4.54–7.98
			Distal	1.745	.112	–0.27–3.76
AVLT4	Control	Proximal	1.483	.099	–0.19–3.15	
		Distal	3.745 <sup>a</sup>	.000	2.28–5.21	
AVLT5	Control	Proximal	2.262 <sup>a</sup>	.005	0.55–3.97	
		Distal	5.566 <sup>a</sup>	.000	3.39–7.74	
AVLT_TO	Control	Proximal	6.082 <sup>a</sup>	.000	4.18–7.99	
		Distal	0.516	1.000	–1.71–2.74	
AVLT_IR	Control	Proximal	1.091	.194	–0.33–2.51	
		Distal	3.245 <sup>a</sup>	.000	2.00–4.49	
AVLT_DR	Control	Proximal	2.153 <sup>a</sup>	.002	0.70–3.61	
		Distal	1.727 <sup>a</sup>	.014	0.28–3.18	
AVLT_DR	Control	Proximal	3.793 <sup>a</sup>	.000	2.52–5.06	
		Distal	2.065 <sup>a</sup>	.003	0.58–3.55	
AVLT_DR	Control	Proximal	1.925 <sup>a</sup>	.014	0.31–3.54	
		Distal	4.041 <sup>a</sup>	.000	2.63–5.45	
AVLT_DR	Control	Proximal	2.116 <sup>a</sup>	.007	0.46–3.77	
		Distal	1.683 <sup>a</sup>	.031	0.11–3.25	
AVLT_DR	Control	Proximal	4.183 <sup>a</sup>	.000	2.81–5.56	
		Distal	2.500 <sup>a</sup>	.001	0.89–4.11	
AVLT_DR	Control	Proximal	1.352	.062	–0.05–2.75	
		Distal	3.522 <sup>a</sup>	.000	2.29–4.75	
AVLT_DR	Control	Proximal	2.170 <sup>a</sup>	.001	0.73–3.61	
		Distal	7.778 <sup>a</sup>	.018	1.04–14.51	
AVLT_DR	Control	Proximal	18.783 <sup>a</sup>	.000	12.88–24.69	
		Distal	11.005 <sup>a</sup>	.001	4.10–17.91	
AVLT_DR	Control	Proximal	2.475 <sup>a</sup>	.006	0.57–4.38	
		Distal	4.865 <sup>a</sup>	.000	3.20–6.53	
AVLT_DR	Control	Proximal	2.390 <sup>a</sup>	.011	0.44–4.34	
		Distal	2.720 <sup>a</sup>	.001	1.00–4.44	
AVLT_DR	Control	Proximal	4.969 <sup>a</sup>	.000	3.46–6.47	
		Distal	2.249 <sup>a</sup>	.007	0.49–4.01	

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**On-line Table 1: Continued**

SN/Test	Subset	Group (I)	Group (J)	Difference (I-J)	Significance	95% CI, Lower-to-Upper Bound
3/Memory for Designs Test	AVLT_%	Control	Proximal	13.5751682 <sup>a</sup>	.002	4.082150–23.068186
			Distal	19.6638323 <sup>a</sup>	.000	11.340977–27.986688
		Proximal	Distal	6.0886641	.392	–3.638383–15.815711
	AVLT_REC	Control	Proximal	0.492	.545	–0.40–1.38
			Distal	1.843 <sup>a</sup>	.000	1.06–2.62
	AVLT_ERR	Proximal	Distal	1.351 <sup>a</sup>	.001	0.44–2.26
		Control	Proximal	–1.195	.340	–3.02–0.63
			Distal	–4.251 <sup>a</sup>	.000	–5.85 to –2.65
		Proximal	Distal	–3.056 <sup>a</sup>	.000	–4.92 to –1.19
	MFDT1/10	Control	Proximal	0.847	.470	–0.61–2.30
			Distal	1.838 <sup>a</sup>	.004	0.49–3.19
	MFDT2/10	Proximal	Distal	0.991	.305	–0.48–2.46
		Control	Proximal	1.080	.419	–0.70–2.86
			Distal	3.169 <sup>a</sup>	.000	1.52–4.82
	MFDT3/10	Proximal	Distal	2.089 <sup>a</sup>	.017	0.29–3.89
		Control	Proximal	0.534	1.000	–1.14–2.21
			Distal	3.076 <sup>a</sup>	.000	1.52–4.63
	MFDT4/10	Proximal	Distal	2.542 <sup>a</sup>	.001	0.85–4.24
		Control	Proximal	0.216	1.000	–1.49–1.93
			Distal	2.496 <sup>a</sup>	.001	0.91–4.08
	MFDT5/10	Proximal	Distal	2.280 <sup>a</sup>	.006	0.55–4.01
		Control	Proximal	0.443	1.000	–1.00–1.89
			Distal	2.556 <sup>a</sup>	.000	1.21–3.90
	MFDT_TO/50	Proximal	Distal	2.113 <sup>a</sup>	.002	0.65–3.57
		Control	Proximal	3.119	.886	–4.17–10.41
			Distal	13.134 <sup>a</sup>	.000	6.37–19.90
	MFDT_DR/10	Proximal	Distal	10.015 <sup>a</sup>	.004	2.66–17.37
		Control	Proximal	1.028	.356	–0.57–2.63
			Distal	4.115 <sup>a</sup>	.000	2.63–5.60
	MFDT_%/10	Proximal	Distal	3.086 <sup>a</sup>	.000	1.47–4.70
		Control	Proximal	9.8528439	.157	–2.415305–22.120993
			Distal	30.5033541 <sup>a</sup>	.000	19.145255–41.861453
MFDT1/14	Proximal	Distal	20.6505102 <sup>a</sup>	.000	48.036712–33.264308	
	Control	Proximal	1.817	.165	–0.48–4.12	
		Distal	1.656	.082	–0.15–3.46	
MFDT2/14	Proximal	Distal	–0.162	1.000	–2.53–2.20	
	Control	Proximal	2.611	.065	–0.12–5.34	
		Distal	3.944 <sup>a</sup>	.000	1.80–6.09	
MFDT3/14	Proximal	Distal	1.333	.725	–1.48–4.14	
	Control	Proximal	3.048 <sup>a</sup>	.005	0.77–5.32	
		Distal	4.200 <sup>a</sup>	.000	2.42–5.98	
MFDT4/14	Proximal	Distal	1.152	.671	–1.18–3.49	
	Control	Proximal	1.976	.056	–0.04–3.99	
		Distal	4.100 <sup>a</sup>	.000	2.52–5.68	
MFDT5/14	Proximal	Distal	2.124 <sup>a</sup>	.043	0.05–4.19	
	Control	Proximal	2.198 <sup>a</sup>	.035	0.12–4.27	
		Distal	3.189 <sup>a</sup>	.000	1.56–4.82	
MFDT_TO/70	Proximal	Distal	0.990	.754	–1.14–3.12	
	Control	Proximal	11.651 <sup>a</sup>	.015	1.87–21.43	
		Distal	17.089 <sup>a</sup>	.000	9.41–24.77	
MFDT_DR/14	Proximal	Distal	5.438	.549	–4.61–15.49	
	Control	Proximal	2.024	.224	–0.74–4.79	
		Distal	4.701 <sup>a</sup>	.000	2.53–6.87	
MFDT_%/14	Proximal	Distal	2.676	.071	–0.17–5.52	
	Control	Proximal	–1.9302787	1.000	–20.703920–16.843363	
		Distal	18.9113109 <sup>a</sup>	.008	4.176693–33.645929	
	Proximal	Distal	20.8415896 <sup>a</sup>	.030	1.549447–40.133733	

**Note:**—“MFDT10 Total- TO” or “MFDT14 Total- TO” indicates the total number of pictures learnt after 5 trial; MFDT, Memory for Designs Test; TO, cumulative sum of the scores in all 5 trials; DR, delayed recall; AVLT, Auditory Verbal Learning Test; REC, Recall- Total number of correct hits is calculated under “Recall”; ERR, ERROR - Total number of commissions and omissions; IR, immediate recall; SN, serial number; MFDT10, children from 6-8 years of age where 10 pictures are presented for testing and 5 trials given similar to AVLT; MFDT14, 8-10 years old children where 14 pictures are presented.

<sup>a</sup> Level of significance.

**On-line Table 2: Quantitative values of DTI derived metrics (FA and MD) from different regions of the brain and comparison of FA and MD between healthy controls and DMD subgroup patients (DMD proximal and distal mutation)**

SN	Brain Regions	Control	Proximal Dp140+	Distal Dp140-	ANOVA (p-value)	Paired t-test (p-value)		
						Proximal vs. control	Distal vs. control	
			Fractional Anisotropy (FA)				Proximal vs. distal	
1	Anterior thalamic radiation-L (ATR)	0.45±0.01	0.44±0.01	0.44±0.02	0.001 <sup>a</sup>	0.001 <sup>a</sup>	0.01	1.00
2	Anterior thalamic radiation-R (ATR)	0.45±0.02	0.44±0.01	0.44±0.01	0.01 <sup>a</sup>	0.01 <sup>a</sup>	0.22	0.36
3	Corticospinal tract-L (CST)	0.54±0.01	0.53±0.01	0.53±0.02	0.02	0.02	0.47	0.42
4	Corticospinal tract-R (CST)	0.54±0.01	0.53±0.01	0.53±0.02	0.03	0.05	0.16	1.00
5	Cingulum (cingulate gyrus)-L (Cg)	0.42±0.02	0.40±0.01	0.41±0.02	0.001 <sup>a</sup>	0.001 <sup>a</sup>	0.01	0.90
6	Cingulum (cingulate gyrus)-R (Cg)	0.39±0.02	0.38±0.01	0.39±0.02	0.07	0.07	0.52	0.86
7	Inferior fronto-occipital fasciculus-L (ILOF)	0.44±0.02	0.43±0.01	0.44±0.02	0.08	0.10	0.34	1.00
8	Inferior fronto-occipital fasciculus-R (ILOF)	0.44±0.02	0.43±0.02	0.43±0.02	0.12	0.14	0.54	1.00
9	Inferior longitudinal fasciculus-L (ILF)	0.39±0.02	0.38±0.02	0.39±0.02	0.09	0.12	0.40	1.00
10	Inferior longitudinal fasciculus-R (ILF)	0.39±0.02	0.36±0.02	0.37±0.02	0.03	0.04	0.29	0.94
11	Superior longitudinal fasciculus-L (SLF)	0.42±0.02	0.04±0.02	0.40±0.02	0.001 <sup>a</sup>	0.001 <sup>a</sup>	0.001	1.00
12	Superior longitudinal fasciculus-R (SLF)	0.41±0.02	0.40±0.01	0.40±0.02	0.02	0.02	0.12	1.00
13	Uncinate fasciculus-L (Unc)	0.39±0.02	0.37±0.02	0.38±0.02	0.07	0.07	0.68	0.69
14	Uncinate fasciculus-R (Unc)	0.39±0.02	0.39±0.02	0.38±0.04	0.05	0.04	1.00	0.29
15	Superior longitudinal fasciculus (temporal part)-L (SLFt)	0.40±0.02	0.38±0.02	0.39±0.02	0.02	0.02	0.25	0.80
16	Superior longitudinal fasciculus (temporal part)-R (SLFt)	0.47±0.02	0.43±0.02	0.44±0.02	0.04	0.08	0.10	1.00
17	Body of corpus callosum (bCC)	0.60±0.03	0.68±0.03	0.68±0.04	0.01	0.01	0.01	1.00
<b>Mean Diffusivity (MD)</b>								
1	Anterior thalamic radiation-L (ATR)	0.82±0.03	0.83±0.03	0.84±0.03	0.001 <sup>a</sup>	0.08	0.001 <sup>a</sup>	1.00
2	Anterior thalamic radiation-R (ATR)	0.83±0.04	0.84±0.04	0.84±0.02	0.23	0.72	0.32	1.00
3	Corticospinal tract-L (CST)	0.76±0.03	0.76±0.02	0.77±0.02	0.01 <sup>a</sup>	0.95	0.001 <sup>a</sup>	1.00
4	Corticospinal tract-R (CST)	0.76±0.03	0.77±0.02	0.78±0.02	0.01 <sup>a</sup>	0.79	0.01 <sup>a</sup>	1.00
5	Cingulum (cingulate gyrus)-L (Cg)	0.78±0.04	0.80±0.03	0.81±0.02	0.001 <sup>a</sup>	0.06	0.001 <sup>a</sup>	1.00
6	Cingulum (cingulate gyrus)-R (Cg)	0.80±0.04	0.81±0.02	0.82±0.01	0.01	0.62	0.01	1.00
7	Forceps major (FMa)	0.77±0.04	0.79±0.03	0.82±0.02	0.001	0.06	0.001 <sup>a</sup>	1.00
8	Forceps minor (FMi)	0.80±0.04	0.82±0.03	0.82±0.02	0.07	0.18	0.14	1.00
9	Inferior fronto-occipital fasciculus-L (ILOF)	0.80±0.04	0.81±0.02	0.82±0.02	0.08	0.10	0.08	1.00
10	Inferior fronto-occipital fasciculus-R (ILOF)	0.80±0.04	0.81±0.02	0.82±0.02	0.07	1.00	0.06	1.00
11	Inferior longitudinal fasciculus-L (ILF)	0.81±0.04	0.83±0.03	0.84±0.02	0.01 <sup>a</sup>	0.13	0.001 <sup>a</sup>	1.00
12	Inferior longitudinal fasciculus-R (ILF)	0.82±0.03	0.80±0.02	0.84±0.02	0.03	0.81	0.02	1.00
13	Superior longitudinal fasciculus-L (SLF)	0.78±0.04	0.80±0.02	0.81±0.02	0.001 <sup>a</sup>	0.04	0.001 <sup>a</sup>	1.00
14	Superior longitudinal fasciculus-R (SLF)	0.78±0.03	0.79±0.03	0.81±0.03	0.001 <sup>a</sup>	0.05	0.001 <sup>a</sup>	1.00
15	Uncinate fasciculus-L (Unc)	0.89±0.05	0.90±0.03	0.90±0.03	0.17	1.00	0.18	1.00
16	Uncinate fasciculus-R (Unc)	0.97±0.06	0.97±0.04	0.95±0.03	0.20	1.00	0.24	0.23
17	Superior longitudinal fasciculus (temporal part)-L (SLFt)	0.79±0.04	0.81±0.02	0.82±0.02	0.001 <sup>a</sup>	0.15	0.001 <sup>a</sup>	1.00
18	Superior longitudinal fasciculus (temporal part)-R (SLFt)	0.80±0.03	0.82±0.02	0.82±0.02	0.001 <sup>a</sup>	0.07	0.001 <sup>a</sup>	1.00
19	Body of corpus callosum (bCC)	0.81±0.04	0.83±0.04	0.85±0.04	0.001 <sup>a</sup>	0.37	0.01 <sup>a</sup>	0.63

**Note:**—L indicates left; R, right; SN, serial number.  
<sup>a</sup> statistically significant values.