

Supplementary Table 3. Cognitive Status

Test	Case 3	Case 4
IQ Estimate (1)	SS / Pr	SS / Pr
Raven's Progressive Matrices	75 / 5 th (Borderline)	78 / 7 th (Borderline)
Memory and Learning (2)	ScS / Pr	ScS / Pr
WRAML2 Picture Memory	--	8 / 25 th (Average)
WRAML2 Picture Memory Delayed Recognition	--	5 / 5 th (Borderline)
WRAML2 Design Memory	6 / 9 th (Borderline)	5 / 5 th (Borderline)
WRAML2 Design Memory Delayed Recognition	6 / 9 th (Borderline)	9 / 37 th (Average)
WRAML2 Verbal Learning	9 / 37 th (Average)	8 / 25 th (Low Average)
WRAML2 Verbal Learning Delayed Recall	7 / 16 th (Low Average)	8 / 25 th (Low Average)
WRAML2 Verbal Learning Delayed Recognition	10 / 50 th (Average)	6 / 9 th (Below Average)
Psychomotor Speed and Attention	T or Z score / Pr	T or Z score / Pr
Trail Making Test Part A (3) *	31 / 3 rd (Mild-Moderate Impairment)	40 / 16 th (Below Average)
SDMT-Written (4)	--	-3.00 / 1 st (Very Low)

SDMT-Oral (4)	--	-2.70 / 1 st (Very Low)
Executive Functioning	T score / Pr	T score / Pr
Trail Making Test Part B (3) *	--	35 / 7 th (Mild Impairment)
COWA Semantic Verbal Fluency (5) *	35 / 7 th (Mild Impairment)	28 / 2 nd (Moderate Impairment)
COWA Linguistic Verbal Fluency (5) *	37 / 9 th (Mild Impairment)	28 / 2 nd (Moderate Impairment)

Pr = Percentile rank

SS = Standard Score (mean = 100, standard deviation = 15)

ScS = Scaled Score (mean = 10, standard deviation = 3)

T score (mean = 50, standard deviation = 10)

Z score (mean = 0, standard deviation = 1)

WRAML2 = Wide Range Assessment of Memory and Learning, Second Edition

SDMT = Symbol Digit Modalities Test

COWA = Controlled Oral Word Association

* T scores derived from demographically adjusted neuropsychological normative data (6)

Article title: Rapid-onset dystonia-parkinsonism associated with the I758S mutation of the ATP1A3 gene: A neuropathologic and neuroanatomical study of four siblings

Journal name: Acta Neuropathologica

Authors: Adrian L. Oblak, Ph.D., Matthew C. Hagen, M.D., Ph.D., Kathleen J Sweadner, Ph.D., Ihtsham Haq, M.D., Christopher T. Whitlow, M.D., Ph.D., Joseph A. Maldjian, M.D., Francine Epperson, Jared F. Cook, M.A., Mark Stacy, M.D., Jill R. Murrell, Ph.D., Laurie J Ozelius, Ph.D., Allison Brashear, M.D., Bernardino Ghetti, M.D.

Corresponding author:

Bernardino Ghetti, MD

Indiana University School of Medicine

Department of Pathology and Laboratory Medicine

bghetti@iupui.edu

Reference List

- (1) Raven JC. Raven's Progressive Matrices and Coloured Progressive Matrices with Research Supplement. (2000) Harcourt Assessment, San Antonio, USA
- (2) Sheslow D, Adams W. Wide Range Assessment of Memory and Learning, Second Edition. ed. (2007) Wide Range Inc., Florida, USA
- (3) Washington DC- Adjutant General's Office WD. Army Individual Test Battery. Manual of directions and scoring. (1944)
- (4) Smith A. Symbol Digit Modalities Test: Manual. (2007) Western Psychological Services, Los Angeles, USA
- (5) Spreen O, Strauss E. A Compendium of Neuropsychological Tests. (1998) Oxford University Press, New York, USA
- (6) Heaton RK, Miller W, Taylor MJ, Grant I. (2004) Revised Comprehensive Norms for an Expanded Halstead-Reitan Battery: Demographically Adjusted Neuropsychological Norms for African American and Caucasian Adults. Psychological Assessment Resources, Inc. USA