

Supporting Information

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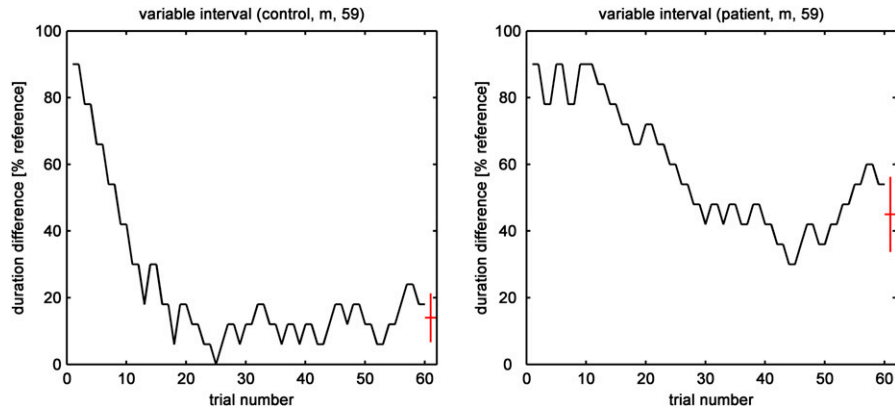


Fig. S1. Adaptive threshold tracking shown for variable single interval discrimination in (A) one control subject (male, 59 y) and (B) one SCA6 patient (male, 59 y). Both tracks demonstrate the typical course of the adaptive tracking procedure: start at suprathreshold levels, followed by adaptive tracking based on a two-down, one-up procedure, switching from a larger to a smaller step size after four reversals, leveling off around threshold, which is estimated as the arithmetic mean of the final six reversals (red cross, mean \pm SD).

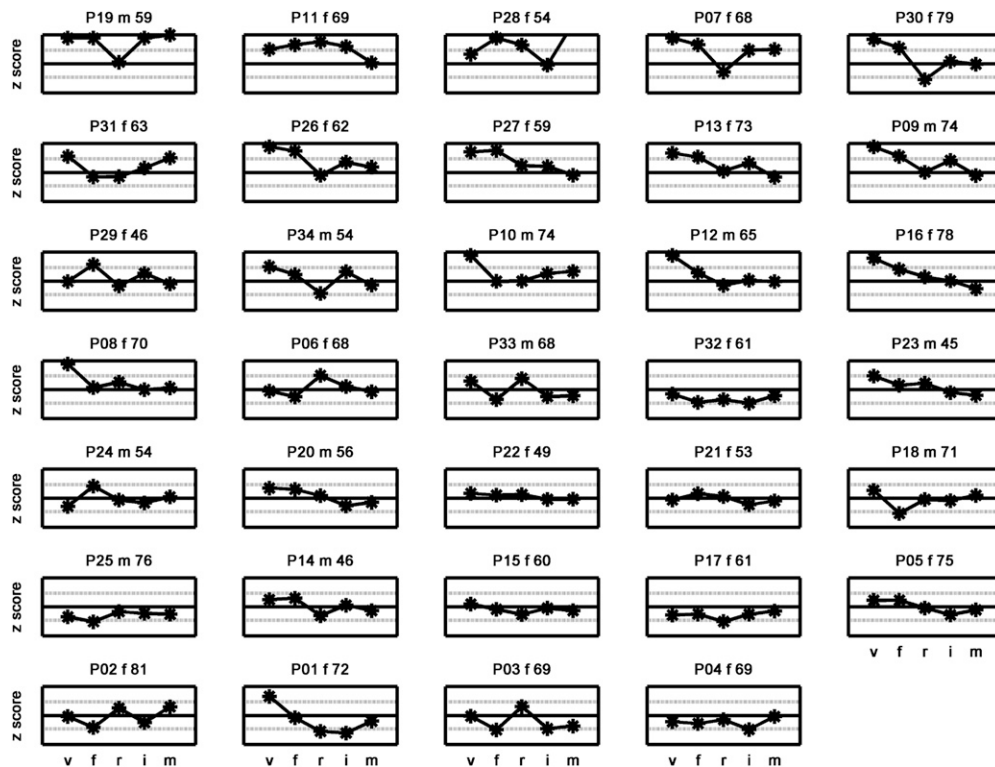


Fig. S2. Individual patients' z score profiles. Each subplot depicts one patient's z scores for the five tasks in the order of the timing of variable interval duration (v), fixed interval duration (f), regular beat detection (r), isochronous beat violation (i), and metrical beat distortion (m). The z scores were calculated based on an age-dependent regression of thresholds (*Materials and Methods*). Significance level was $z > 1.65$ ($P < 0.05$), indicated by gray line. The upper limit for display was set to $z = 3.09$ ($P = 0.001$). The order of individual subplots is based on the order of severity of deficits, based first on the number of significantly elevated z scores and second on mean z scores. Also note the frequent occurrence of significantly elevated z scores for tasks of timing of single intervals (v, f) in contrast to the rare occurrence of deficits in the timing of a regular beat (r, i, m).

Table S1. Patient characteristics

ID	Sex	Age, y	Education, y	Nonverbal IQ	Est verbal IQ	Onset age, y	Duration, y	Motor severity
1	M	72			105			5
2	M	81	10		106	67	15	3
3	F	69	12		105	59	10	3
4	M	69	11	107		55	14	3
5	F	75	9	101	102	65	10	3
6	M	68	10	107	93	58	10	4
7	F	68	10	91	94	58	10	3
8	M	70	10	102	100	53	17	2
9	F	74				70	4	2
10	M	74	9	91		62	12	2
11	F	69	10		106	49	20	4
12	F	65			85			2
13	M	73	12	111		59	20	4
14	M	46	17	116	114	35	11	3
15	F	60	10	95	108	58	2	2
16	F	78	9	102	108	75	3	3
17	M	61	10	101	117			3
18	F	71	12	95	100			3
19	F	59	12	104	108			1
20	F	56	11	101	121	0	0	0
21	F	53	13	110	109	0	0	0
22	F	49	13	104	111	0	0	0
23	M	45	11	101	110	0	0	0
24	F	54	12	103	123	51	5	2
25	F	76	9	87	92	73	3	3
26	F	62	10	90	94	62	1	2
27	M	59	11	97	92	56	3	3
28	F	54	11	101	92	50	4	2
29	F	46	13	103				2
30	F	79	10	99	91	76	1	3
31	F	63	12	105	118	50	13	3
32	F	61	12	105	118	55	6	2
33	F	68	12	106	119	50	18	3
34	M	54	13			41	13	3

Nonverbal and verbal IQ were estimated based on demographic variables (1) and the National Adult Reading Test (2), respectively. Onset age of symptoms was self-reported. Motor impairment was measured on a locally-developed scale of 1–5, based on four subscores of the severity of gait ataxia (0 = not present, 1 = mild, 2 = severe), limb ataxia (0 = not present, 1 = present), dysarthria (0 = not present, 1 = present) and nystagmus (0 = not present, 1 = present). Est, estimated; IQ, intelligence quotient; F, female; M, male.

1. Crawford JR, et al. (1989) Estimating premorbid IQ from demographic variables: Regression equations derived from a UK sample. *Br J Clin Psychol* 28:275–278.
2. Nelson HE, Willison JR (1991) *National Adult Reading Test Manual* (NFER-Nelson, Windsor, UK) 2 Ed.

Table S2. Age dependency of timing thresholds in controls and patients

	Controls (n = 40)	Patients (n = 34)
Variable	$r = 0.23, P = 0.031^*$	$r = 0.41, P = 0.008^*$
Fixed	$r = 0.42, P = 0.003^*$	$r = 0.01, P = 0.473^*$
Regular	$r = -0.10, P = 0.267^*$	$r = -0.16, P = 0.186$
Isochronous	$r = 0.22, P = 0.082$	$r = 0.07, P = 0.351$
Metrical	$r = 0.17, P = 0.153$	$r = 0.27, P = 0.062$

Correlation coefficients and *P* values were obtained by using Pearson's product-moment and Spearman's rho for normally and non-normally (*) distributed samples (tested by the Lilliefors modification of Kolmogorov–Smirnov test for composite normality), respectively. Significant effects (at level of $P < 0.05$; in boldface type) were found for variable and fixed single-interval timing in controls, as well as variable interval timing in patients. The other tasks showed trends that were not significant.