

Table S1. Clinical characteristics of subjects in each human biofluid study

Experiment	Disease stage	Number of subjects	Female/male	Mean age (range)
Cytokine multiplex assay Plasma	Control	69	43:26	42 (22–67)
	Premanifest	34	19:15	39 (23–54)
	Early	47	24:23	47 (25–80)
	Moderate	44	30:14	52 (38–76)
IgA, IgM assay Plasma	Control	26	18:8	46 (29–65)
	Premanifest	11	5:6	39 (27–49)
	Early	17	9:8	47 (31–61)
	Moderate	18	14:4	50 (26–76)
IgG assay Plasma	Control	19	11:8	40 (26–67)
	Premanifest	17	9:8	38 (27–47)
	Early	13	8:5	42 (25–60)
	Moderate	20	11:9	51 (38–74)
GM-CSF assay Plasma	Control	24	20:4	37 (22–67)
	Premanifest	24	15:9	38 (27–54)
	Early	24	14:10	48 (25–80)
	Moderate	28	17:11	51 (26–77)
IL-6 and IL-8 assay Matched CSF and plasma	Control	9	6:3	45 (25–66)
	Early	9	4:5	51 (38–64)
	Moderate	28	1:10	53 (38–72)

Table S2. Plasma cytokine levels by disease stage measured by multiplex ELISA assay

	Control Mean (SD)	n	Premanifest Mean (SD)	n	Early HD Mean (SD)	n	Moderate HD Mean (SD)	n
IL-6	3.06 (2)	69	5.26 (2.66)	34	5.3 (3.01)	47	6.65 (3.46)	46
IL-8	2.11 (1.03)	69	2.27 (0.97)	34	3.08 (1.47)	47	3.98 (2.67)	46
IL-4	0.32 (0.38)	68	0.58 (0.88)	34	0.63 (0.71)	45	0.84 (0.78)	45
IL-10	0.87 (0.87)	66	1.85 (2.75)	34	1.67 (1.68)	45	2.15 (3.06)	44
TNF- α	2.62 (1.25)	69	3.04 (1.51)	34	3.03 (1.27)	47	3.32 (1.05)	46
IL-5	0.34 (0.23)	66	0.38 (0.33)	34	0.49 (0.88)	45	0.53 (0.5)	46
IL-2	0.86 (0.99)	66	1.06 (0.79)	34	1.01 (0.96)	45	1.51 (4.63)	46
IFN- γ	0.54 (0.58)	66	0.38 (0.39)	34	0.44 (0.44)	45	0.77 (0.76)	46
IL-13	3.17 (2.62)	66	1.84 (1.46)	34	2.48 (2.23)	45	2.35 (12.49)	46
IL-12	1.38 (1.11)	66	1.31 (1.32)	34	1.97 (2.11)	45	0.85 (1.5)	45
IL-1 β	0.83 (0.82)	69	0.81 (0.87)	34	0.85 (0.89)	47	0.74 (0.77)	46
GM-CSF	0.19 (0.16)	23	0.17 (0.12)	24	0.39 (0.48)	24	0.08 (0.1)	36
IL-6	3.06 (2)	69	5.26 (2.66)	34	5.3 (3.01)	47	6.65 (3.46)	46
IL-8	2.11 (1.03)	69	2.27 (0.97)	34	3.08 (1.47)	47	3.98 (2.67)	46
IL-4	0.32 (0.38)	68	0.58 (0.88)	34	0.63 (0.71)	45	0.84 (0.78)	45
IL-10	0.87 (0.87)	66	1.85 (2.75)	34	1.67 (1.68)	45	2.15 (3.06)	44

All cytokine levels are in ng/L.

Table S3. Demographic, clinical, and pathological characteristics of subjects in postmortem striatal expression study

Group	<i>n</i>	Female/ male	Mean age at death (Range)	Mean CAG repeat length (SD)	Relative RNA quantification, % of controls (SD)		
					IL-6	IL-8	TNF- α
Control	6	0:6	59 (42–74)	45 (5) 41 (3)	100 (88.3)	100 (40.1)	100 (60.8)
	17	3:14	54 (40–74)		1520 (159)	1110 (125)	208 (110)
	4	0:4	50 (41–58)		4220 (147)	596 (108)	382 (235)
VS2	4	0:4	62 (47–74)	45 (4)	694 (122)	402 (73.6)	92.1 (153)
VS3	4	0:4	54 (40–64)	45 (4)	3210 (195)	1950 (152)	306 (75.8)
VS4	5	3:2	49 (45–53)	53 (4) ^a	775 (218)	2740 (36.7)	684 (57.7)

^aCAG repeat lengths available for two samples only.

VS, Vonsattel pathological grade (Vonsattel, J.P., R.H. Myers, T.J. Stevens, R.J. Ferrante, E.D. Bird, and E.P. Richardson Jr. 1985. *J. Neuropathol. Exp. Neurol.* 44:559–577).

Table S4. Details of animals used for murine experiments

Experiment	Mouse model	Age of animals	No. WT animals	No. disease animals
Serum cytokine multiplex assay	R6/2	12 wk	20	20
Macrophage stimulation study and serum IL-6 ELISA	<i>Hdh</i> ^{Q150Q/Q150}	22 mo	10	9
	YAC128	12 mo	3	4
Microglial stimulation study	YAC18	12 mo	4	4
	R6/2	Neonatal	4	4

Table S5. Details of subjects whose blood was used for human monocyte studies

Huntingtin expression study	Sex	Age	Status	Disease stage ^a	CAG sizes
C1	F	45.8	Control		
C2	M	43.8	Control		
HD1	M	41.9	HD	2	17/45
HD2	F	44.2	HD	2	25/43
HD3	F	61.4	HD	2	16/42
Monocyte functional study	functional n	Female/male		Mean age (SD)	Mean expanded CAG repeat length (range)
Controls	8	2:7		42.2 (10.3)	
Premanifest HD	9	4:4		38.8 (6.2)	43.5 (41–48)

^aShoulson, I., and S. Fahn. 1979. *Neurology*. 29:1–3.

Table S6. Primers used for human monocyte expression studies

Gene	Forward primer	Reverse primer	Probe (5'-FAM, 3'-TAMRA)
Abl	TGGAGATAACACTCT AAGCATAACTAAAG GT	GATGTAGTTGCTTGG GACCCA	CCATTTTGTTTGG GCTTCACACCATT
β-2-microglobulin	GAGTATGCCTGCCGT GTG	AATCCAAATGCGGCA TCT	CCTCCATGATGCTGC TTACATGTCTC
Huntingtin	GCTGCACCGACCGTG AGT	CGCAGGCTGCAGGGT TAC	CAGCTCCCTGTCCCCG GCGG
CAG repeat sizing	ATGAAGGCCTTCGAG TCCCTCAAGTCCTTC ^a	GGCGGCTGAGGAAG CTGAGGA	N/A

^aFAM-tagged primer.