

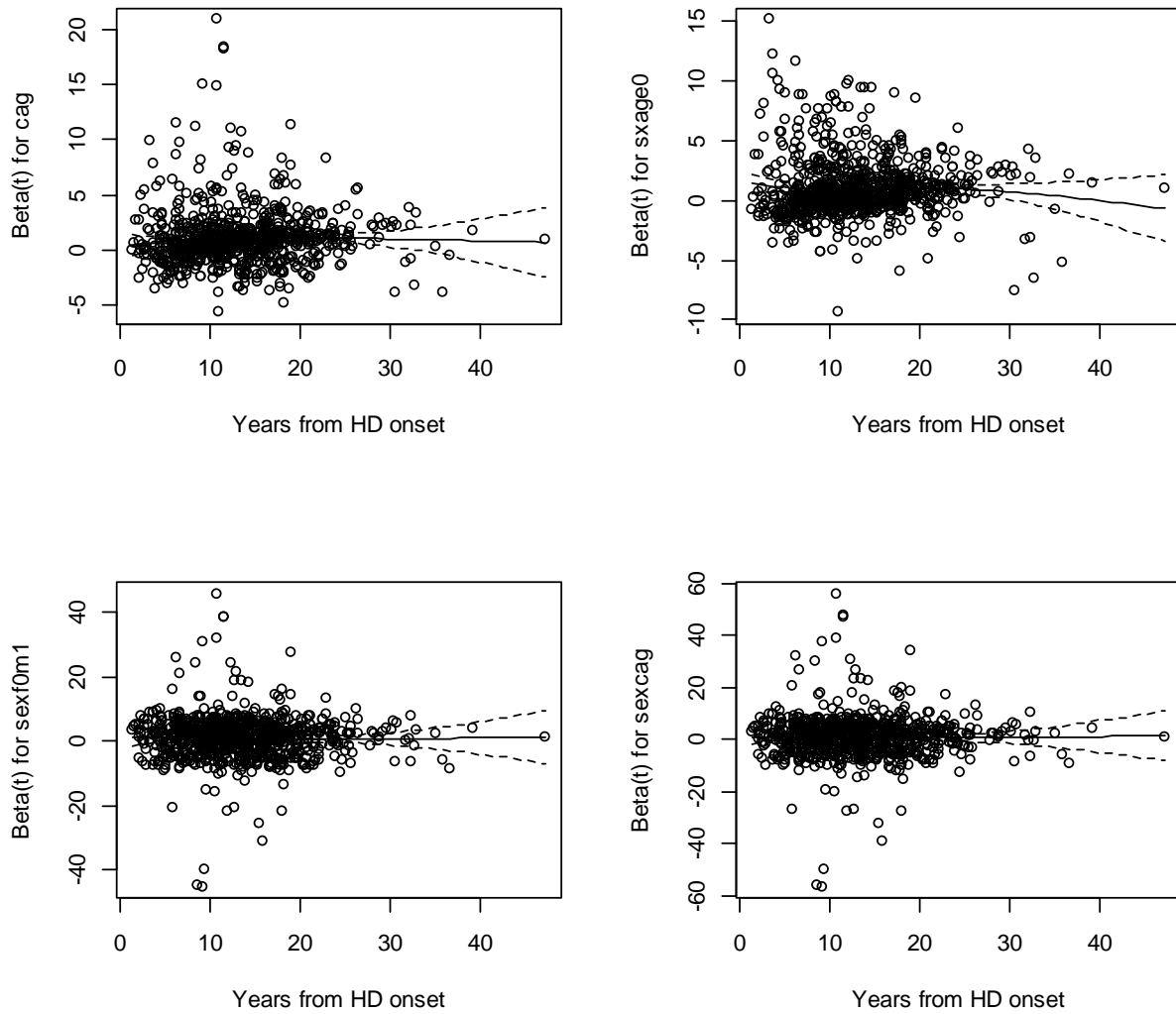
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Supplemental information

**Longer CAG repeat length is associated
with shorter survival after disease onset
in Huntington disease**

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Disease Network**

Figure S1. ZPH proportional hazard goodness of fit test results



Abbreviations: *cag* = main effect of CAG length, *sxage0* = Joint effect of 3 degree-of-freedom restricted cubic spline fit of age-at-onset; *sexf0m1* = main effect of men vs women; *sexcag* = interaction of CAG and men vs women.

The near linearity of the Beta(t) coefficients, especially up to 30 years from HD onset, indicate good fit of the proportional hazards assumptions in the main model (Table 2 of the paper).

Supplemental Table 1. ZPH Proportional Hazard Goodness-of-fit for the final model

Variables	rho	Chi-square	p
CAG Main Effect	0.0355	0.883	0.347
Age of HD Onset (3 df)	-0.0235	0.442	0.506
Men vs Women	-0.0191	0.278	0.598
CAG by (M vs W)	-0.0112	0.096	0.757

p = p value. (Also see Supplemental Figure 1.)

Supplemental Table 2. Frequency of CAG Lengths in 5-Year Windows Surrounding Plotted Ages in Figures 1 and 2 (Main Text)

Age 23-27

	39	40	41	42	43	44	45	46	47	48	49	50	51	52	53	54	55	56	57	58	59	60	62	63	63	tot
F	1	0	3	2	5	7	8	15	17	12	20	25	15	15	22	18	12	8	6	3	5	2	2	0	0	223
M	0	1	3	3	6	5	10	8	11	15	18	25	15	13	15	8	7	3	3	1	2	1	3	1	1	178
tot	1	1	6	5	11	12	18	23	28	27	38	50	30	28	37	26	19	11	9	4	7	3	5	1	1	401

Age 33-37

	36	38	39	40	41	42	43	44	45	46	47	48	49	50	51	52	53	54	55	56	58	68	tot
F	2	1	2	5	7	22	38	44	69	100	82	43	37	22	10	3	0	3	0	0	0	0	490
M	0	0	2	5	8	22	35	63	64	75	79	48	38	22	11	2	1	0	2	1	1	1	480
tot	2	1	4	10	15	44	73	107	133	175	161	91	75	44	21	5	1	3	2	1	1	1	970

Age 43-47

	37	38	39	40	41	42	43	44	45	46	47	48	49	50	51	57	tot
F	3	4	5	11	40	106	118	166	101	44	27	11	4	0	2	0	642
M	1	2	8	11	28	108	154	142	80	34	18	7	5	3	0	1	602
tot	4	6	13	22	68	214	272	308	181	78	45	18	9	3	2	1	1244

Age 53-57

	36	37	38	39	40	41	42	43	44	45	46	47	48	49	50	52	54	tot
F	0	3	5	12	35	113	162	109	34	16	2	3	1	0	0	0	1	496
M	1	0	4	6	43	127	171	83	30	11	3	2	3	1	1	1	0	487
tot	1	3	9	18	78	240	333	192	64	27	5	5	4	1	1	1	1	983

Supplemental Table 3. Survival curve estimates by CAG length for women with HD onset at age 25

CAG	39*			40*			41			42			43			44		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.991	0.986	0.995	0.989	0.985	0.994	0.988	0.983	0.993	0.987	0.981	0.992	0.985	0.979	0.991	0.983	0.976	0.989
10	0.960	0.944	0.976	0.955	0.938	0.971	0.949	0.932	0.967	0.943	0.924	0.961	0.935	0.916	0.955	0.927	0.907	0.948
15	0.889	0.850	0.929	0.875	0.835	0.918	0.860	0.818	0.905	0.844	0.800	0.890	0.825	0.780	0.873	0.805	0.758	0.854
20	0.756	0.683	0.837	0.729	0.654	0.812	0.699	0.623	0.785	0.667	0.591	0.754	0.633	0.557	0.720	0.596	0.520	0.684
25	0.613	0.514	0.732	0.575	0.477	0.695	0.535	0.438	0.654	0.493	0.399	0.610	0.450	0.359	0.564	0.405	0.319	0.515
CAG	45			46			47			48			49			50		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.981	0.974	0.988	0.978	0.970	0.986	0.975	0.967	0.984	0.972	0.963	0.981	0.968	0.958	0.979	0.964	0.953	0.976
10	0.918	0.896	0.940	0.908	0.885	0.931	0.897	0.872	0.921	0.884	0.858	0.910	0.870	0.842	0.898	0.854	0.824	0.885
15	0.782	0.734	0.833	0.757	0.708	0.810	0.730	0.680	0.785	0.701	0.649	0.757	0.669	0.616	0.728	0.635	0.579	0.697
20	0.558	0.483	0.644	0.517	0.443	0.602	0.474	0.402	0.558	0.430	0.360	0.513	0.385	0.318	0.467	0.340	0.275	0.421
25	0.360	0.279	0.465	0.315	0.240	0.414	0.271	0.202	0.364	0.229	0.167	0.315	0.189	0.133	0.268	0.152	0.103	0.225
CAG	51			52			53			54			55			56		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>
5	0.947	0.973	0.955	0.941	0.969	0.949	0.933	0.965	0.943	0.925	0.961	0.935	0.915	0.956	0.927	0.904	0.951	0.947
10	0.804	0.870	0.817	0.782	0.855	0.796	0.756	0.838	0.773	0.728	0.820	0.747	0.697	0.801	0.719	0.662	0.781	0.804
15	0.540	0.664	0.560	0.498	0.630	0.519	0.453	0.595	0.477	0.406	0.559	0.433	0.358	0.523	0.388	0.309	0.487	0.540
20	0.232	0.376	0.252	0.191	0.332	0.210	0.152	0.290	0.172	0.117	0.251	0.136	0.087	0.215	0.105	0.061	0.182	0.232
25	0.076	0.185	0.090	0.054	0.150	0.066	0.036	0.119	0.046	0.023	0.093	0.031	0.013	0.071	0.020	0.007	0.054	0.076
CAG	57			58			59											
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>									
5	0.918	0.891	0.946	0.908	0.877	0.940	0.897	0.861	0.934									
10	0.689	0.625	0.760	0.656	0.584	0.738	0.621	0.540	0.715									
15	0.343	0.261	0.450	0.298	0.215	0.414	0.254	0.171	0.378									
20	0.078	0.041	0.151	0.056	0.025	0.125	0.039	0.015	0.101									
25	0.012	0.003	0.039	0.007	0.002	0.028	0.003	0.001	0.020									

Data is plotted in Figure 1 of the main article. *surv* = survival rate, *lower* = lower 95% confidence bound, *upper* = upper 95% confidence bound.

* CAG length represents less than 1% of those onset HD onset in this age range. Estimate for this CAG length is an extrapolation from the survival model.

Supplemental Table 4. Survival curve estimates by CAG length for women with HD onset at age 35

CAG	39*			40*			41			42			43			44		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.983	0.977	0.989	0.981	0.975	0.987	0.979	0.972	0.986	0.976	0.968	0.983	0.973	0.964	0.981	0.969	0.960	0.978
10	0.929	0.910	0.948	0.920	0.901	0.940	0.910	0.890	0.930	0.899	0.878	0.920	0.886	0.865	0.909	0.873	0.850	0.896
15	0.809	0.767	0.853	0.787	0.745	0.831	0.763	0.720	0.807	0.736	0.694	0.781	0.707	0.665	0.753	0.676	0.633	0.722
20	0.604	0.535	0.682	0.566	0.498	0.642	0.525	0.460	0.599	0.483	0.421	0.554	0.439	0.380	0.507	0.394	0.338	0.459
25	0.414	0.334	0.514	0.369	0.295	0.463	0.324	0.256	0.411	0.280	0.218	0.359	0.237	0.182	0.309	0.197	0.148	0.262
CAG	45			46			47			48			49			50		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.965	0.955	0.975	0.961	0.950	0.972	0.956	0.943	0.968	0.950	0.936	0.964	0.944	0.928	0.960	0.937	0.919	0.955
10	0.857	0.833	0.882	0.840	0.814	0.867	0.821	0.793	0.851	0.801	0.769	0.833	0.778	0.742	0.815	0.753	0.712	0.796
15	0.642	0.598	0.689	0.606	0.561	0.656	0.568	0.520	0.621	0.527	0.476	0.585	0.485	0.429	0.549	0.442	0.380	0.512
20	0.349	0.296	0.412	0.304	0.253	0.366	0.261	0.211	0.322	0.219	0.170	0.281	0.179	0.133	0.242	0.143	0.099	0.207
25	0.159	0.116	0.218	0.125	0.088	0.178	0.095	0.063	0.144	0.070	0.043	0.114	0.050	0.028	0.088	0.034	0.017	0.067
CAG	51			52			53*			54*			55*			56*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>
5	0.929	0.908	0.950	0.920	0.896	0.945	0.910	0.882	0.939	0.899	0.867	0.933	0.887	0.849	0.926	0.873	0.829	0.918
10	0.725	0.678	0.775	0.695	0.641	0.754	0.663	0.601	0.732	0.628	0.557	0.709	0.592	0.511	0.685	0.552	0.462	0.660
15	0.397	0.331	0.476	0.352	0.281	0.440	0.307	0.233	0.404	0.263	0.188	0.369	0.221	0.146	0.335	0.182	0.109	0.301
20	0.111	0.071	0.174	0.083	0.048	0.145	0.060	0.031	0.119	0.042	0.018	0.096	0.028	0.010	0.077	0.017	0.005	0.060
25	0.022	0.009	0.050	0.013	0.005	0.037	0.007	0.002	0.026	0.004	0.001	0.018	0.002	0.000	0.012	0.001	0.000	0.008
CAG	57*			58*			59*											
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>									
5	0.857	0.807	0.911	0.840	0.782	0.903	0.821	0.755	0.894									
10	0.511	0.412	0.634	0.468	0.361	0.608	0.424	0.310	0.581									
15	0.145	0.078	0.269	0.113	0.054	0.238	0.085	0.034	0.210									
20	0.010	0.002	0.046	0.006	0.001	0.035	0.003	0.000	0.026									
25	0.000	0.000	0.005	0.000	0.000	0.003	0.000	0.000	0.002									

Data is plotted in Figure 1 of the main article. *surv* = survival rate, *lower* = lower 95% confidence bound, *upper* = upper 95% confidence bound.

* CAG length represents less than 1% of those onset HD onset in this age range. Estimate for this CAG length is an extrapolation from the survival model.

Supplemental Table 5. Survival curve estimates by CAG length for women with HD onset at age 45

CAG	39*			40			41			42			43			44		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.981	0.974	0.987	0.978	0.971	0.985	0.975	0.968	0.983	0.972	0.964	0.980	0.969	0.960	0.978	0.965	0.955	0.975
10	0.918	0.901	0.936	0.908	0.890	0.927	0.897	0.878	0.916	0.884	0.864	0.904	0.870	0.849	0.891	0.854	0.832	0.877
15	0.783	0.745	0.822	0.758	0.720	0.798	0.731	0.694	0.771	0.702	0.664	0.742	0.670	0.632	0.711	0.636	0.596	0.679
20	0.559	0.499	0.626	0.518	0.460	0.582	0.475	0.420	0.537	0.431	0.379	0.490	0.386	0.336	0.444	0.341	0.292	0.398
25	0.362	0.294	0.444	0.317	0.255	0.393	0.272	0.217	0.343	0.230	0.180	0.294	0.190	0.145	0.249	0.153	0.113	0.207
CAG	45			46			47			48			49*			50*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.960	0.949	0.971	0.955	0.942	0.968	0.949	0.935	0.964	0.943	0.926	0.959	0.936	0.917	0.955	0.927	0.906	0.950
10	0.837	0.813	0.862	0.818	0.790	0.846	0.797	0.765	0.829	0.773	0.737	0.811	0.748	0.706	0.793	0.720	0.671	0.773
15	0.600	0.557	0.645	0.561	0.515	0.611	0.520	0.469	0.577	0.478	0.421	0.542	0.434	0.371	0.507	0.389	0.321	0.472
20	0.297	0.248	0.354	0.253	0.205	0.312	0.211	0.164	0.273	0.173	0.126	0.236	0.137	0.093	0.202	0.106	0.066	0.171
25	0.120	0.084	0.170	0.091	0.060	0.138	0.066	0.040	0.109	0.047	0.025	0.086	0.031	0.015	0.066	0.020	0.008	0.049
CAG	51*			52*			53*			54*			55*			56*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>
5	0.918	0.893	0.944	0.908	0.879	0.939	0.897	0.863	0.932	0.884	0.845	0.926	0.870	0.824	0.919	0.854	0.801	0.911
10	0.690	0.633	0.752	0.657	0.591	0.730	0.622	0.547	0.708	0.585	0.500	0.684	0.545	0.450	0.660	0.504	0.400	0.635
15	0.344	0.271	0.437	0.299	0.223	0.402	0.256	0.178	0.368	0.214	0.137	0.334	0.175	0.101	0.302	0.139	0.072	0.270
20	0.079	0.044	0.143	0.057	0.027	0.118	0.039	0.016	0.096	0.026	0.009	0.077	0.016	0.004	0.060	0.009	0.002	0.047
25	0.012	0.004	0.036	0.007	0.002	0.026	0.003	0.001	0.018	0.002	0.000	0.013	0.001	0.000	0.008	0.000	0.000	0.005
CAG	57*			58*			59*											
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>									
5	0.837	0.776	0.903	0.818	0.748	0.895	0.797	0.716	0.886									
10	0.461	0.348	0.609	0.416	0.297	0.583	0.371	0.248	0.556									
15	0.108	0.048	0.240	0.081	0.031	0.212	0.058	0.018	0.186									
20	0.005	0.001	0.036	0.003	0.000	0.027	0.001	0.000	0.020									
25	0.000	0.000	0.003	0.000	0.000	0.002	0.000	0.000	0.001									

Data is plotted in Figure 1 of the main article. *surv* = survival rate, *lower* = lower 95% confidence bound, *upper* = upper 95% confidence bound.

* CAG length represents less than 1% of those onset HD onset in this age range. Estimate for this CAG length is an extrapolation from the survival model.

Supplemental Table 6. Survival curve estimates by CAG length for women with HD onset at age 55

CAG	39			40			41			42			43			44		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.976	0.968	0.984	0.973	0.964	0.981	0.969	0.960	0.979	0.965	0.955	0.976	0.961	0.949	0.972	0.956	0.943	0.969
10	0.898	0.877	0.921	0.886	0.863	0.909	0.872	0.848	0.896	0.857	0.832	0.882	0.839	0.813	0.867	0.820	0.791	0.850
15	0.735	0.690	0.783	0.706	0.661	0.754	0.674	0.629	0.723	0.641	0.595	0.690	0.605	0.557	0.656	0.566	0.517	0.620
20	0.481	0.415	0.557	0.437	0.374	0.510	0.392	0.333	0.462	0.347	0.290	0.415	0.302	0.248	0.368	0.259	0.207	0.323
25	0.278	0.212	0.365	0.235	0.176	0.314	0.195	0.143	0.266	0.157	0.112	0.222	0.124	0.084	0.181	0.094	0.061	0.146
CAG	45			46*			47*			48*			49*			50*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.950	0.936	0.965	0.944	0.927	0.960	0.937	0.918	0.955	0.929	0.907	0.950	0.920	0.895	0.945	0.910	0.881	0.939
10	0.800	0.768	0.833	0.777	0.741	0.814	0.751	0.711	0.794	0.724	0.677	0.774	0.694	0.640	0.752	0.662	0.600	0.729
15	0.526	0.473	0.584	0.483	0.427	0.547	0.440	0.379	0.510	0.395	0.330	0.473	0.350	0.281	0.436	0.305	0.233	0.400
20	0.217	0.167	0.281	0.178	0.131	0.241	0.142	0.098	0.205	0.110	0.070	0.172	0.082	0.047	0.143	0.059	0.030	0.117
25	0.069	0.042	0.115	0.049	0.027	0.089	0.033	0.016	0.068	0.021	0.009	0.050	0.013	0.004	0.037	0.007	0.002	0.026
CAG	51*			52*			53*			54*			55*			56*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>
5	0.898	0.866	0.932	0.886	0.848	0.925	0.872	0.829	0.918	0.857	0.806	0.910	0.839	0.782	0.901	0.820	0.754	0.893
10	0.627	0.557	0.706	0.590	0.511	0.681	0.551	0.462	0.656	0.509	0.412	0.630	0.466	0.361	0.603	0.422	0.310	0.575
15	0.261	0.187	0.364	0.219	0.146	0.329	0.180	0.109	0.295	0.144	0.079	0.263	0.112	0.054	0.232	0.084	0.035	0.203
20	0.041	0.018	0.094	0.027	0.010	0.074	0.017	0.005	0.058	0.010	0.002	0.044	0.005	0.001	0.033	0.003	0.000	0.024
25	0.004	0.001	0.018	0.002	0.000	0.012	0.001	0.000	0.008	0.000	0.000	0.005	0.000	0.000	0.003	0.000	0.000	0.002
CAG	57*			58*			59*											
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>									
5	0.800	0.724	0.883	0.777	0.690	0.873	0.751	0.654	0.863									
10	0.377	0.260	0.547	0.332	0.213	0.519	0.288	0.169	0.491									
15	0.061	0.021	0.177	0.042	0.012	0.152	0.028	0.006	0.130									
20	0.001	0.000	0.018	0.001	0.000	0.012	0.000	0.000	0.009									
25	0.000	0.000	0.001	0.000	0.000	0.001	0.000	0.000	0.000									

Data is plotted in Figure 1 of the main article. *surv* = survival rate, *lower* = lower 95% confidence bound, *upper* = upper 95% confidence bound.

* CAG length represents less than 1% of those onset HD onset in this age range. Estimate for this CAG length is an extrapolation from the survival model.

Supplemental Table 7. Survival curve estimates by CAG length for men with HD onset at age 25

CAG	39*			40*			41			42			43			44		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.982	0.974	0.990	0.981	0.973	0.989	0.979	0.971	0.988	0.977	0.969	0.986	0.975	0.966	0.985	0.973	0.964	0.983
10	0.925	0.898	0.952	0.919	0.891	0.947	0.912	0.884	0.941	0.905	0.877	0.934	0.897	0.869	0.927	0.889	0.860	0.920
15	0.798	0.737	0.865	0.783	0.721	0.850	0.767	0.706	0.835	0.751	0.689	0.818	0.733	0.671	0.800	0.714	0.653	0.780
20	0.585	0.487	0.704	0.559	0.463	0.676	0.533	0.440	0.646	0.506	0.416	0.615	0.477	0.391	0.583	0.449	0.366	0.550
25	0.392	0.285	0.540	0.363	0.261	0.503	0.333	0.238	0.465	0.304	0.216	0.427	0.275	0.194	0.390	0.247	0.173	0.352
CAG	45			46			47			48			49			50		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.971	0.961	0.981	0.969	0.958	0.980	0.966	0.955	0.978	0.964	0.952	0.975	0.961	0.948	0.973	0.957	0.944	0.971
10	0.881	0.851	0.911	0.871	0.841	0.903	0.861	0.831	0.893	0.851	0.819	0.883	0.839	0.807	0.873	0.827	0.794	0.862
15	0.694	0.634	0.760	0.673	0.614	0.738	0.651	0.592	0.716	0.628	0.570	0.692	0.604	0.546	0.668	0.579	0.521	0.644
20	0.420	0.341	0.516	0.390	0.316	0.482	0.361	0.290	0.448	0.331	0.264	0.415	0.302	0.239	0.382	0.273	0.213	0.350
25	0.219	0.152	0.316	0.193	0.133	0.281	0.168	0.114	0.248	0.145	0.097	0.217	0.123	0.081	0.189	0.104	0.066	0.163
CAG	51			52			53			54			55			56		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>
5	0.954	0.940	0.968	0.950	0.935	0.965	0.946	0.930	0.963	0.942	0.924	0.960	0.937	0.918	0.957	0.932	0.911	0.954
10	0.814	0.779	0.850	0.800	0.763	0.838	0.785	0.746	0.826	0.769	0.727	0.814	0.753	0.707	0.801	0.735	0.685	0.789
15	0.553	0.494	0.619	0.527	0.466	0.595	0.499	0.437	0.570	0.471	0.406	0.546	0.442	0.374	0.522	0.413	0.342	0.499
20	0.245	0.188	0.320	0.218	0.163	0.291	0.192	0.139	0.264	0.167	0.116	0.239	0.144	0.096	0.216	0.122	0.077	0.194
25	0.086	0.052	0.140	0.070	0.041	0.119	0.056	0.031	0.101	0.044	0.022	0.086	0.034	0.016	0.072	0.025	0.011	0.060
CAG	57			58			59											
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>									
5	0.926	0.903	0.950	0.920	0.895	0.947	0.914	0.885	0.944									
10	0.716	0.661	0.776	0.697	0.636	0.763	0.676	0.609	0.750									
15	0.383	0.309	0.476	0.354	0.276	0.454	0.324	0.243	0.432									
20	0.102	0.060	0.175	0.085	0.046	0.156	0.069	0.034	0.140									
25	0.019	0.007	0.050	0.013	0.004	0.042	0.009	0.003	0.034									

Data is plotted in Figure 2 of the main article. *surv* = survival rate, *lower* = lower 95% confidence bound, *upper* = upper 95% confidence bound.

* CAG length represents less than 1% of those onset HD onset in this age range. Estimate for this CAG length is an extrapolation from the survival model.

Supplemental Table 8. Survival curve estimates by CAG length for men with HD onset at age 35

CAG	39*			40*			41			42			43			44		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.968	0.958	0.979	0.965	0.955	0.977	0.963	0.951	0.974	0.960	0.948	0.972	0.956	0.944	0.969	0.953	0.939	0.966
10	0.868	0.838	0.899	0.858	0.828	0.889	0.847	0.817	0.878	0.835	0.806	0.866	0.823	0.793	0.854	0.810	0.779	0.841
15	0.666	0.608	0.731	0.644	0.587	0.706	0.621	0.566	0.681	0.596	0.543	0.654	0.571	0.520	0.627	0.545	0.495	0.600
20	0.381	0.309	0.470	0.351	0.285	0.433	0.322	0.260	0.397	0.293	0.236	0.362	0.264	0.212	0.328	0.236	0.189	0.296
25	0.185	0.128	0.268	0.161	0.111	0.234	0.138	0.094	0.202	0.117	0.079	0.172	0.098	0.065	0.146	0.080	0.053	0.122
CAG	45			46			47			48			49			50		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.949	0.935	0.963	0.945	0.929	0.960	0.940	0.924	0.957	0.935	0.918	0.953	0.930	0.911	0.950	0.924	0.903	0.949
10	0.795	0.764	0.828	0.780	0.748	0.814	0.764	0.730	0.800	0.747	0.710	0.786	0.729	0.689	0.772	0.710	0.666	0.795
15	0.518	0.469	0.572	0.490	0.441	0.545	0.462	0.411	0.518	0.433	0.380	0.492	0.403	0.348	0.467	0.374	0.315	0.518
20	0.209	0.165	0.265	0.184	0.142	0.237	0.159	0.120	0.211	0.137	0.099	0.188	0.116	0.080	0.167	0.096	0.063	0.209
25	0.065	0.041	0.102	0.052	0.032	0.084	0.040	0.023	0.070	0.031	0.017	0.057	0.023	0.011	0.047	0.017	0.007	0.065
CAG	51			52			53*			54*			55*			56*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>
5	0.946	0.918	0.895	0.942	0.912	0.886	0.939	0.905	0.876	0.935	0.897	0.865	0.930	0.889	0.854	0.926	0.881	0.841
10	0.757	0.690	0.641	0.743	0.669	0.614	0.729	0.647	0.586	0.714	0.624	0.556	0.700	0.600	0.525	0.685	0.574	0.492
15	0.443	0.344	0.282	0.420	0.315	0.250	0.397	0.286	0.218	0.375	0.257	0.187	0.354	0.230	0.158	0.334	0.203	0.131
20	0.148	0.079	0.048	0.131	0.064	0.036	0.115	0.051	0.026	0.101	0.040	0.018	0.089	0.030	0.012	0.077	0.023	0.008
25	0.038	0.012	0.005	0.031	0.008	0.003	0.025	0.006	0.002	0.020	0.004	0.001	0.016	0.002	0.000	0.013	0.001	0.000
CAG	57*			58*			59*											
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>									
5	0.871	0.827	0.918	0.861	0.812	0.913	0.850	0.796	0.909									
10	0.548	0.458	0.656	0.521	0.424	0.641	0.494	0.389	0.627									
15	0.178	0.107	0.295	0.154	0.085	0.277	0.131	0.067	0.260									
20	0.016	0.005	0.058	0.012	0.003	0.050	0.008	0.001	0.043									
25	0.001	0.000	0.008	0.000	0.000	0.006	0.000	0.000	0.005									

Data is plotted in Figure 2 of the main article. *surv* = survival rate, *lower* = lower 95% confidence bound, *upper* = upper 95% confidence bound.

* CAG length represents less than 1% of those onset HD onset in this age range. Estimate for this CAG length is an extrapolation from the survival model.

Supplemental Table 9. Survival curve estimates by CAG length for men with HD onset at age 45

CAG	39*			40			41			42			43			44		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.963	0.952	0.974	0.960	0.949	0.972	0.957	0.945	0.969	0.953	0.941	0.966	0.950	0.936	0.963	0.946	0.931	0.960
10	0.849	0.823	0.877	0.838	0.812	0.865	0.826	0.799	0.853	0.812	0.786	0.840	0.798	0.771	0.827	0.784	0.755	0.813
15	0.626	0.578	0.677	0.601	0.556	0.650	0.576	0.533	0.623	0.550	0.509	0.596	0.524	0.482	0.568	0.496	0.454	0.541
20	0.328	0.274	0.392	0.299	0.250	0.357	0.270	0.226	0.323	0.242	0.201	0.291	0.215	0.177	0.261	0.189	0.153	0.234
25	0.143	0.102	0.198	0.121	0.087	0.169	0.101	0.072	0.143	0.084	0.058	0.120	0.068	0.046	0.100	0.054	0.035	0.084
CAG	45			46			47			48			49*			50*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.941	0.925	0.957	0.936	0.919	0.954	0.931	0.913	0.950	0.926	0.905	0.947	0.920	0.897	0.943	0.913	0.888	0.939
10	0.768	0.737	0.799	0.751	0.718	0.785	0.733	0.696	0.772	0.714	0.673	0.758	0.694	0.648	0.744	0.673	0.621	0.730
15	0.468	0.424	0.515	0.439	0.393	0.491	0.410	0.359	0.467	0.380	0.326	0.443	0.350	0.292	0.421	0.321	0.258	0.399
20	0.164	0.129	0.209	0.141	0.107	0.187	0.120	0.086	0.167	0.100	0.068	0.148	0.083	0.052	0.132	0.067	0.039	0.117
25	0.043	0.026	0.069	0.033	0.019	0.057	0.025	0.013	0.047	0.018	0.008	0.039	0.013	0.005	0.032	0.009	0.003	0.026
CAG	51*			52*			53*			54*			55*			56*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>
5	0.906	0.878	0.935	0.899	0.868	0.931	0.891	0.856	0.927	0.882	0.843	0.923	0.873	0.830	0.919	0.863	0.815	0.915
10	0.652	0.593	0.716	0.629	0.563	0.702	0.605	0.531	0.688	0.580	0.498	0.674	0.554	0.465	0.660	0.527	0.430	0.646
15	0.292	0.225	0.378	0.263	0.194	0.358	0.235	0.164	0.338	0.209	0.136	0.319	0.183	0.111	0.301	0.159	0.089	0.283
20	0.054	0.028	0.103	0.042	0.019	0.091	0.032	0.013	0.080	0.024	0.008	0.070	0.018	0.005	0.061	0.013	0.003	0.053
25	0.006	0.002	0.021	0.004	0.001	0.017	0.002	0.000	0.013	0.001	0.000	0.011	0.001	0.000	0.009	0.000	0.000	0.007
CAG	57*			58*			59*											
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>									
5	0.853	0.799	0.910	0.841	0.782	0.906	0.829	0.763	0.902									
10	0.500	0.395	0.632	0.471	0.360	0.618	0.443	0.324	0.604									
15	0.136	0.070	0.266	0.115	0.053	0.249	0.096	0.039	0.234									
20	0.009	0.002	0.046	0.006	0.001	0.040	0.004	0.000	0.034									
25	0.000	0.000	0.005	0.000	0.000	0.004	0.000	0.000	0.003									

Data is plotted in Figure 2 of the main article. *surv* = survival rate, *lower* = lower 95% confidence bound, *upper* = upper 95% confidence bound.

* CAG length represents less than 1% of those onset HD onset in this age range. Estimate for this CAG length is an extrapolation from the survival model.

Supplemental Table 10. Survival curve estimates by CAG length for men with HD onset at age 55

CAG	39			40			41			42			43			44		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.954	0.940	0.968	0.950	0.936	0.965	0.946	0.931	0.962	0.942	0.926	0.958	0.937	0.920	0.955	0.932	0.913	0.951
10	0.814	0.782	0.848	0.801	0.769	0.834	0.786	0.753	0.819	0.770	0.737	0.805	0.753	0.719	0.789	0.736	0.700	0.774
15	0.554	0.501	0.613	0.528	0.476	0.584	0.500	0.451	0.555	0.472	0.424	0.526	0.443	0.395	0.497	0.414	0.365	0.469
20	0.246	0.195	0.311	0.219	0.172	0.278	0.193	0.150	0.246	0.168	0.129	0.218	0.144	0.109	0.192	0.123	0.090	0.168
25	0.086	0.056	0.134	0.070	0.045	0.111	0.056	0.035	0.091	0.044	0.026	0.074	0.034	0.019	0.060	0.026	0.014	0.048
CAG	45			46*			47*			48*			49*			50*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>
5	0.927	0.906	0.947	0.921	0.899	0.943	0.914	0.890	0.939	0.907	0.881	0.935	0.900	0.871	0.930	0.892	0.860	0.926
10	0.717	0.678	0.758	0.697	0.655	0.743	0.677	0.630	0.727	0.655	0.603	0.711	0.632	0.574	0.696	0.608	0.544	0.680
15	0.384	0.334	0.443	0.355	0.302	0.417	0.325	0.269	0.393	0.296	0.237	0.370	0.267	0.206	0.347	0.239	0.176	0.326
20	0.103	0.072	0.147	0.085	0.056	0.129	0.069	0.043	0.112	0.055	0.031	0.097	0.043	0.022	0.084	0.033	0.015	0.073
25	0.019	0.009	0.038	0.014	0.006	0.031	0.009	0.004	0.024	0.006	0.002	0.019	0.004	0.001	0.015	0.003	0.001	0.012
CAG	51*			52*			53*			54*			55*			56*		
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>	<i>surv</i>	<i>upper</i>	<i>upper</i>
5	0.884	0.848	0.921	0.875	0.835	0.916	0.865	0.820	0.912	0.854	0.805	0.907	0.843	0.788	0.902	0.831	0.770	0.897
10	0.583	0.512	0.665	0.558	0.479	0.649	0.531	0.445	0.633	0.504	0.411	0.618	0.476	0.375	0.602	0.447	0.340	0.587
15	0.212	0.148	0.305	0.187	0.122	0.285	0.162	0.099	0.266	0.139	0.078	0.248	0.118	0.060	0.231	0.099	0.045	0.215
20	0.025	0.010	0.063	0.018	0.006	0.054	0.013	0.004	0.046	0.009	0.002	0.039	0.006	0.001	0.033	0.004	0.001	0.028
25	0.002	0.000	0.009	0.001	0.000	0.007	0.001	0.000	0.005	0.000	0.000	0.004	0.000	0.000	0.003	0.000	0.000	0.002
CAG	57*			58*			59*											
years	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>	<i>surv</i>	<i>lower</i>	<i>upper</i>									
5	0.853	0.799	0.910	0.841	0.782	0.906	0.829	0.763	0.882									
10	0.500	0.395	0.632	0.471	0.360	0.618	0.443	0.324	0.541									
15	0.136	0.070	0.266	0.115	0.053	0.249	0.096	0.039	0.171									
20	0.009	0.002	0.046	0.006	0.001	0.040	0.004	0.000	0.017									
25	0.000	0.000	0.005	0.000	0.000	0.004	0.000	0.000	0.001									

Data is plotted in Figure 2 of the main article. *surv* = survival rate, *lower* = lower 95% confidence bound, *upper* = upper 95% confidence bound.

* CAG length represents less than 1% of those onset HD onset in this age range. Estimate for this CAG length is an extrapolation from the survival model.

Supplemental Table 11. Final Model in main paper

Variable	log HR	S.E.	z	p val	HR	95% CI	
CAG (per repeat)	0.1227	0.0134	9.157	5.34E-20	1.131	1.101	1.161
Age of HD Onset	0.0704	0.0124	5.677	1.37E-08	1.073	1.047	1.099
Age of HD Onset'	-0.1191	0.0265	-4.494	6.99E-06	0.888	0.843	0.935
Age of HD Onset''	0.5751	0.1113	5.167	2.38E-07	1.777	1.429	2.211
Men vs Women	2.2965	0.5782	3.972	7.13E-05	9.939	3.200	30.868
CAG by (M vs W)	-0.0422	0.0127	-3.323	8.91E-04	0.959	0.935	0.983

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. In p values, *XE-Y* indicates *X* times 10 to the *-Y* power (scientific notation). For age of HD onset, restricted cubic spline knots were placed at 23.05, 40.00, 50.00 and 65.00 years of age. *Age of HD Onset'* and *Age of HD Onset''* are the nonlinear restricted cubic spline coefficients for age of HD onset.

Supplemental Table 12. Model with 105 questionable cases of HD onset removed

Variable	log HR	S.E.	z	p val	HR	95% CI	
CAG (per repeat)	0.124	0.0134	9.254	<0.0001	1.132	1.103	1.162
Age of HD Onset	0.0712	0.0122	5.836	<0.0001	1.074	1.048	1.100
Age of HD Onset'	-0.1276	0.0278	-4.590	<0.0001	0.880	0.834	0.929
Age of HD Onset''	0.5777	0.1107	5.219	<0.0001	1.782	1.434	2.214
Men vs Women	2.2935	0.5775	3.971	<0.0001	9.910	3.195	30.733
CAG by (M vs W)	-0.0421	0.0127	-3.315	0.0009	0.959	0.935	0.983

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. For age of HD onset, restricted cubic spline knots were placed at 23.05, 40.00, 50.00 and 65.00 years of age. *Age of HD Onset'* and *Age of HD Onset''* are the nonlinear restricted cubic spline coefficients for age of HD onset.

Supplemental Table 13, Model with data restricted to CAG lengths 41 through 56

Variable	log HR	S.E.	z	p val	HR	95% CI	
CAG (per repeat)	0.1292	0.0218	5.927	<0.0001	1.138	1.090	1.188
Age of HD Onset	0.0648	0.0109	5.945	<0.0001	1.067	1.044	1.090
Age of HD Onset'	-0.1011	0.0457	-2.212	0.0268	0.904	0.826	0.989
Age of HD Onset''	0.395	0.1695	2.330	0.0198	1.484	1.065	2.069
Men vs Women	2.1869	0.9854	2.219	0.0268	8.908	1.291	61.448
CAG by (M vs W)	-0.0391	0.0218	-1.794	0.0733	0.962	0.921	1.004

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. In p values, *XE-Y* indicates *X* times 10 to the *-Y* power (scientific notation). For age of HD onset, restricted cubic spline knots were placed at 23.05, 40.00, 50.00 and 65.00 years of age. *Age of HD Onset'* and *Age of HD Onset''* are the nonlinear restricted cubic spline coefficients for age of HD onset.

Supplemental Table 14. Model controlling for (non-significant) effect of CAG length on the short allele

Variable	log HR	S.E.	z	p val	HR	95% CI	
CAG (per repeat)	0.1272	0.0142	8.958	<0.0001	1.136	1.104	1.168
Age of HD Onset	0.0764	0.0135	5.659	<0.0001	1.079	1.051	1.108
Age of HD Onset'	-0.1261	0.0288	-4.378	<0.0001	0.882	0.833	0.933
Age of HD Onset''	0.5912	0.1205	4.906	<0.0001	1.806	1.426	2.287
Men vs Women	2.3517	0.6148	3.825	1.00E-04	10.503	3.148	35.045
CAG by (M vs W)	-0.0426	0.0135	-3.156	1.60E-03	0.958	0.933	0.984
CAG short allele (per repeat)	0.0091	0.0104	0.875	3.78E-01	1.009	0.989	1.030

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. In p values, *XE-Y* indicates *X* times 10 to the *-Y* power (scientific notation). For age of HD onset, restricted cubic spline knots were placed at 23.05, 40.00, 50.00 and 65.00 years of age. *Age of HD Onset'* and *Age of HD Onset''* are the nonlinear restricted cubic spline coefficients for age of HD onset.

Supplemental Table 15. Model controlling for data subset within Registry HD

Variable	log HR	S.E.	z	p val	HR	95% CI	
CAG (per repeat)	0.1078	0.0137	7.869	<0.0001	1.114	1.084	1.144
Age of HD Onset	0.0639	0.0124	5.153	<0.0001	1.066	1.040	1.092
Age of HD Onset'	-0.0954	0.0263	-3.627	0.0003	0.909	0.863	0.957
Age of HD Onset''	0.4791	0.1114	4.301	<0.0001	1.615	1.298	2.009
Men vs Women	1.9171	0.5836	3.285	0.001	6.801	2.167	21.347
CAG-by-(M vs W)	-0.0348	0.0129	-2.698	0.0068	0.966	0.942	0.991
R3 vs R2 data source	-0.9545	0.2566	-3.720	0.0002	0.385	0.233	0.637
Onset Age * (R3 vs R2)	-0.0041	0.0054	-0.759	0.4496	0.996	0.985	1.007

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. In p values, *XE-Y* indicates *X* times 10 to the *-Y* power (scientific notation). For age of HD onset, restricted cubic spline knots were placed at 23.05, 40.00, 50.00 and 65.00 years of age. *Age of HD Onset'* and *Age of HD Onset''* are the nonlinear restricted cubic spline coefficients for age of HD onset. The exploratory variable of interest is the interaction between Onset Age and R3 vs R2 because the concern is that different methods for recording age of onset were used in these two subsets of registry-HD. In an additional model (not shown in a table) we tested the possible significance of an interaction of data source and all three onset age spline terms used to define the nonlinear details of the effect of onset age on survival. This interaction was not significant (Chi-Square = 1.31, 3df, p = .726.)

Supplemental Table 16. Model with Rater Estimate of Onset Age Used Throughout (Rather than Rater Age for Subset R2 and Motor Onset Age For R3)

Variable	log HR	S.E.	z	p val	HR	95% CI	
CAG (per repeat)	0.0948	0.0143	6.629	<0.0001	1.099	1.069	1.131
Age of HD Onset	0.0478	0.013	3.677	0.0002	1.049	1.023	1.076
Age of HD Onset'	-0.0978	0.0293	-3.338	0.0009	0.907	0.856	0.960
Age of HD Onset''	0.4401	0.1075	4.094	<0.0001	1.553	1.258	1.917
Men vs Women	2.2104	0.6101	3.623	0.0003	9.119	2.758	30.148
CAG-by-(M vs W)	-0.0405	0.0134	-3.022	0.0026	0.960	0.935	0.986

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. In p values, *XE-Y* indicates *X* times 10 to the *-Y* power (scientific notation). For age of HD onset, restricted cubic spline knots were placed at 23.05, 40.00, 50.00 and 65.00 years of age. Because the data were less complete, use of rater onset age rather than motor onset age reduced the sample to 804 deaths out of 7904 observations rather than 826 deaths in 8422 observations.

The two onset ages were are highly correlated. For the 6582 participants from R3 for whom both rater and motor ages were recorded, the Pearson correlation between the two was 0.949 and the standard deviation of the age differences was 3.976 years. The mean motor onset age was 1.17 years older that the rater-reported onset. The correlation between R2 rater onset and R3 motor onset ages was 0.962 among 807 overlapping participants with ages recorded in both data subsets. The standard deviation of the difference was 3.302 years.

Supplemental Table 17 Model with cluster effect for country in which data was collected

Variable	log HR	S.E.	z	p val	HR	95% CI	
CAG (per repeat)	0.1247	0.0129	9.667	<0.0001	1.133	1.105	1.162
Age of HD Onset	0.0702	0.0071	9.887	<0.0001	1.073	1.058	1.088
Age of HD Onset'	-0.1169	0.0211	-5.540	<0.0001	0.890	0.854	0.927
Age of HD Onset''	0.5636	0.0952	5.920	<0.0001	1.757	1.458	2.117
Men vs Women	2.3912	0.4178	5.723	<0.0001	10.927	4.818	24.780
CAG by (M vs W)	-0.0441	0.0088	-5.011	<0.0001	0.957	0.940	0.974

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. In p values, *XE-Y* indicates *X* times 10 to the *-Y* power (scientific notation). For age of HD onset, restricted cubic spline knots were placed at 23.05, 40.00, 50.00 and 65.00 years of age. *Age of HD Onset'* and *Age of HD Onset''* are the nonlinear restricted cubic spline coefficients for age of HD onset.

Supplemental Table 18. Model with cluster effect for site at which data was collected

Variable	log HR	S.E.	z	p val	HR	95% CI	
CAG (per repeat)	0.1247	0.0109	11.440	<0.0001	1.133	1.109	1.157
Age of HD Onset	0.0702	0.0112	6.268	<0.0001	1.073	1.049	1.097
Age of HD Onset'	-0.1168	0.0285	-4.098	<0.0001	0.890	0.841	0.941
Age of HD Onset''	0.5634	0.1292	4.361	<0.0001	1.757	1.364	2.263
Men vs Women	2.3905	0.4638	5.154	<0.0001	10.919	4.400	27.099
CAG by (M vs W)	-0.0441	0.0098	-4.500	<0.0001	0.957	0.939	0.975

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. In p values, *XE-Y* indicates *X* times 10 to the *-Y* power (scientific notation). For age of HD onset, restricted cubic spline knots were placed at 23.05, 40.00, 50.00 and 65.00 years of age. *Age of HD Onset'* and *Age of HD Onset''* are the nonlinear restricted cubic spline coefficients for age of HD onset.

Supplemental Table 19. Consistency of the effect of the estimated effect of CAG-length on Survival, Regardless of Model Adjustments.

Table	Model	Variable	log HR	S.E.	z	p val	HR	95% CI	
2	Final Model	CAG (per repeat)	0.1227	0.0134	9.157	<0.0001	1.131	1.101	1.161
3	105 questionable cases removed	CAG (per repeat)	0.1240	0.0134	9.254	<0.0001	1.132	1.103	1.162
4	CAG restricted to 41 to 56 range	CAG (per repeat)	0.1292	0.0218	5.927	<0.0001	1.138	1.090	1.188
5	Adjust for short allele CAG length	CAG (per repeat)	0.1272	0.0142	8.958	<0.0001	1.136	1.104	1.168
6	Adjust for Registry-HD data subset	CAG (per repeat)	0.1078	0.0137	7.869	<0.0001	1.114	1.084	1.144
7	Rater's onset age used in both data subsets	CAG (per repeat)	0.0948	0.0143	6.629	<0.0001	1.099	1.069	1.131
8	Adjust significance for clustering within countries	CAG (per repeat)	0.1247	0.0129	9.667	<0.0001	1.133	1.105	1.162
9	Adjust significance for clustering by study site	CAG (per repeat)	0.1247	0.0109	11.44	<0.0001	1.133	1.109	1.157

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women.

Supplemental Table 20. Consistency of the Interaction Effect of CAG Length and Sex on Survival, Regardless of Model Adjustments

Table	Model	Variable	log HR	S.E.	z	p val	HR	95% CI	
2	Final Model	CAG by (M vs W)	-0.0422	0.0127	-3.323	0.000891	0.959	0.935	0.983
3	105 questionable cases removed	CAG by (M vs W)	-0.0421	0.0127	-3.315	0.0009	0.959	0.935	0.983
4	CAG restricted to 41 to 56 range	CAG by (M vs W)	-0.0391	0.0218	-1.794	0.0733	0.962	0.921	1.004
5	Adjust for short allele CAG length	CAG by (M vs W)	-0.0426	0.0135	-3.156	0.0016	0.958	0.933	0.984
6	Adjust for Registry-HD data subset	CAG by (M vs W)	-0.0348	0.0129	-2.698	0.0068	0.966	0.942	0.991
7	Rater's onset age used in both data subsets	CAG by (M vs W)	-0.0405	0.0134	-3.022	0.0026	0.96	0.935	0.986
8	Adjust significance for clustering within countries	CAG by (M vs W)	-0.0441	0.0088	-5.011	<0.0001	0.957	0.940	0.974
9	Adjust significance for clustering by study site	CAG by (M vs W)	-0.0441	0.0098	-4.5	<0.0001	0.957	0.939	0.975

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women.

Methods for simulating the effects of imprecise age of onset upon survival from onset.

The age of “onset” of Huntington’s disease, is vaguely defined. In many HD studies, including the R3 portion of Registry, estimates of onset age from more than one source are recorded. An implicit assumption in current and many other HD analyses is that this age of onset corresponds to a critical change in biological disease state that occurs at a more precise age. (Although progressive changes in many biological measures of HD are documented, the measures corresponding to this hypothetical biological state change have not been identified.)

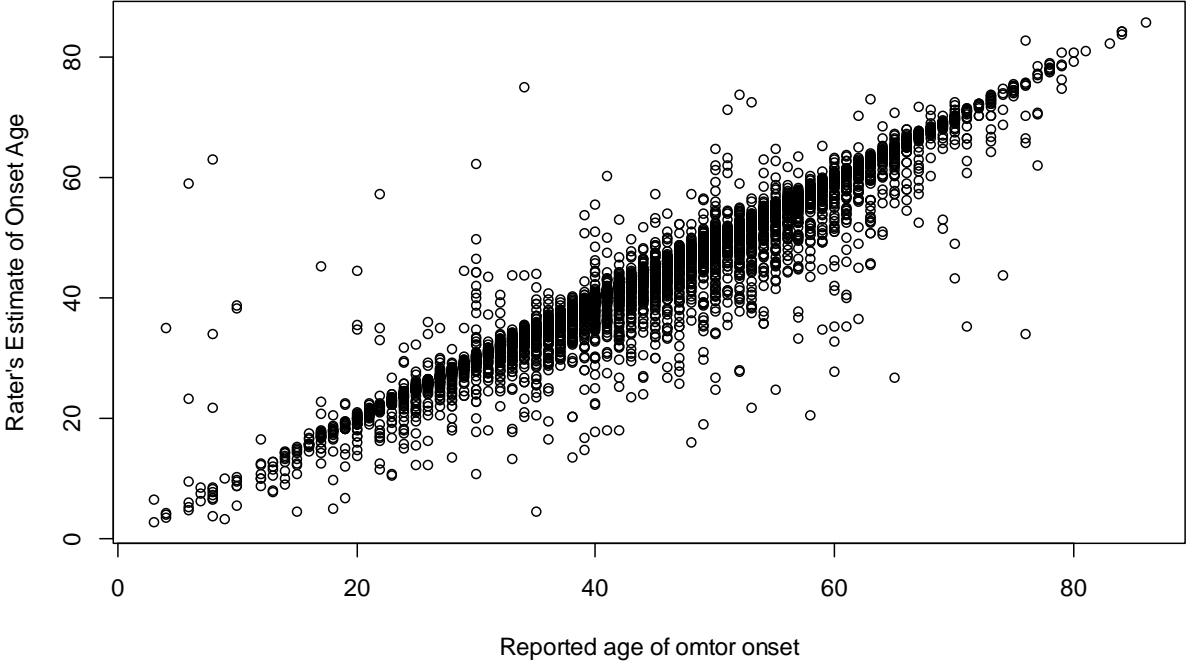
Such measurement error effects are especially relevant to the current analysis because they represent more than a loss of precision in a predictor variable. There is a perfect negative correlation between error in the age of onset and error in the length of survival after onset, which is the outcome measure of interest. In the context of Cox proportional hazard modelling, this greatly complicates any attempt to assess measurement error impact by incorporating it into the underlying mathematical equations. We have thus turned to a simulation study to gain insight.

If we assume that the assumption of an underlying critical biological change is correct, then we can assess the effects of errors in the reported ages of onset relative to the age at which the biological change occurs. We do this by assuming that the ages of onset in our data are indeed the correct underlying ages. We then add random error to these ages repeatedly and record the consequent change in analysis results.

We estimate the standard deviation of reported age around true age using the differences between the variables “age of motor onset” and “rater estimate of onset age” within the R3 portion of the Registry data (Supplemental figure 2). Although the correlation between the two ages is 0.949, the standard deviation of the age differences is 3.98 years. If measurement error in the two reported ages were statistically independent and thus uncorrelated, then the standard deviation of reported ages around the true age would be $3.98/\text{square-root}(2) = 2.81$ years. We used this as the standard deviation of normally distributed, measurement errors in the current simulation. We repeated the simulation 1000 times.

The simulation provides estimates of expected bias in the original log hazard ratio and statistical significance estimates. The ratio of the mean simulation parameters to those obtained from the original data is an estimate of the analogous ratio between the parameters from original data and the hypothetical underlying true onset ages. The adjusted parameter estimates are obtained by dividing the original log hazard and z values by these ratios. Results are summarized in supplemental table 20. In model 1, before including age of onset in the model, the estimated strength and significance of the main effect of CAG-Length increases slightly. Adding age-of-onset as a predictor (model 2), the simulation suggests that the effects of both CAG repeat length and age-of-onset as survival predictors are slight overestimates. However, the adjusted log hazard ratio CAG estimate (0.076 per CAG repeat) retains overwhelming statistical significance ($p = 6.27 * 10^{-17}$). The same is true for the CAG estimate in model 3, where the influence of age of onset is modelled as a nonlinear spline and an interaction between CAG length and sex is also fit.

Figure S2. Correlation between motor onset age and rater’s estimate of onset age within the R3 Registry HD data.



Supplemental Table 21. Comparison of measurement error simulation estimates to original model estimates.

Model 1	Estimates			Simulation Ratios		Adjusted Estimates		
Variable	log HR	z	p val	log HR	Z	log HR	z	p val
CAG (per repeat)	0.022	3.195	1.40E-03	0.932	0.934	0.024	3.417	6.34E-04
Men vs Women	0.359	5.066	4.07E-07	0.998	0.996	0.359	5.090	3.58E-07
Model 2								
Variable	log HR	z	p val	log HR	Z	log HR	z	p val
CAG (per repeat)	0.083	9.604	7.69E-22	1.099	1.148	0.076	8.363	6.27E-17
Men vs Women	0.369	5.201	1.98E-07	0.991	0.989	0.372	5.256	1.47E-07
Age of HD Onset	0.036	8.980	2.71E-19	1.168	1.205	0.031	7.455	8.99E-14
Model 3								
Variable	log HR	z	p val	log HR	Z	log HR	z	p val
CAG (per repeat)	0.123	9.157	5.34E-20	1.069	1.136	0.115	8.043	8.88E-16
Age of HD Onset	0.070	5.677	1.37E-08	1.080	1.116	0.065	5.088	3.61E-07
Age of HD Onset'	-0.119	-4.494	6.99E-06	1.016	0.981	-0.117	-4.585	4.53E-06
Age of HD Onset''	0.575	5.167	2.38E-07	0.934	0.959	0.616	5.389	7.07E-08
Men vs Women	2.297	3.972	7.13E-05	0.983	1.026	2.335	3.869	1.09E-04
CAG by (M vs W)	-0.042	-3.323	8.91E-04	0.984	1.025	-0.043	-3.240	1.19E-03

Simulation ratios are the mean parameters over 1000 simulations divided by the parameter estimate obtained from the original data.

Abbreviations: *HR* = hazard ratio, *S.E* = standard error, *CI* = confidence interval, *p val* = p value, *M* = men, *W* = women. In p values, *XE-Y* indicates *X* times 10 to the *-Y* power (scientific notation). For age of HD onset in Model 3, restricted cubic spline knots were placed at the quantiles (.05, .35, .65, .95) of age of onset. In the original data these quantiles correspond to 23.05, 40.00, 50.00 and 65.00 years of age. These values varied among the simulations, due to adding random noise to the ages of onset. *Age of HD Onset'* and *Age of HD Onset''* are the nonlinear restricted cubic spline coefficients for age of HD onset.

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