The Role of Saccades in Monitoring Progression of Huntington’s Disease
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Prevalence & Genetics
Huntington’s disease (HD) is an autosomal dominant neurodegenerative disease. Genetically, HD patients have an extended CAG repeat sequence in the Huntington gene.

Table 1. Worldwide Prevalence of HD.

<table>
<thead>
<tr>
<th>Area</th>
<th>Cases per 100,000</th>
</tr>
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<tbody>
<tr>
<td>Worldwide</td>
<td>2.71</td>
</tr>
<tr>
<td>Asia</td>
<td>0.40</td>
</tr>
<tr>
<td>North America, Europe, Australia</td>
<td>5.70</td>
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<tr>
<td>Lake Maracaibo</td>
<td>700</td>
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Saccadic Deficiencies & Disease Severity
Individuals with HD have slower and more variable saccadic reaction times and higher incidence of movement errors than age and gender matched controls. As seen in Figure 3, HD patients make more errors in anti-saccadic movements and with increasing delays than controls do. Combined timing and direction errors in saccadic movements in delayed anti-saccadic movement tasks are highly correlated with disease severity (P<0.01).

Conclusions
Huntington’s disease is a lethal, progressive disease for which there is no cure. Pharmacological symptomatic treatments are common, but newer research focuses on boosting mitochondrial function supplements like coenzymes, creatine, and even medical marijuana. Electrococulography, as seen in Figure 5, may offer an inexpensive, portable means to track disease progression in studies directed towards delaying HD onset or slowing progression.

References

Mechanisms of Neurodegeneration
Overstimulation of glutamate receptors, especially the NMDA receptors may cause HD neurodegeneration. In HD patients, wild type astrocytes increase glutamate production and release, activating NMDA receptors. NMDA receptor overstimulation allows higher influx of Ca2+ ions.

Saccadic Deficiencies & Neurodegeneration
Two types of neurons were identified when neuronal activity was measured in the caudate nucleus of rhesus monkeys: one reacted to pro and another to anti-saccades (as seen in Figure 4).

Figure 2. Illustartion of possible contributing points of excitotoxicity.
Mitochondria in HD positive individuals cannot maintain homeostasis of cytosolic Ca2+ and release apoptotic factors into the cytosol. Interestingly, this mitochondrial malfunction is also seen in peripheral tissues. In theory, these cellular malfunctions occur throughout life but appear as symptoms only when self-defence mechanisms can no longer keep up with damage.

Figure 3. Errors made in delayed pro and anti-saccadic tasks.
These measures are sensitive enough to detect deficiencies in pre-symptomatic and pre-diagnosed HD gene carriers and may be more effective than the currently employed motor test.

Figure 4. Two types of CN neurons respond to either pro or anti-saccades.
The neurons activated in anti-saccades act through the indirect basal ganglia pathway to inhibit pro-saccade neurons in the superior colliculus. The degeneration of these caudate nucleus neurons seems to be responsible for the inability of HD patients to inhibit pro-saccades.

Figure 5. Electrode placement for electrooculography measurement.