Emotional Functioning in Huntington's Disease Patients and its Relation to Caregiver Experience

by

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Abstract

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Chairperson: Joan C. Borod, Ph.D.

Huntington's disease (HD) is an inherited, neurodegenerative disorder characterized by progressively worsening abnormalities of movement, cognition, and behavior. Although motor symptoms are the clinical hallmark of the disease, cognitive and psychiatric problems may have the greatest impact on quality of life, and yet, relatively little is known about these types of difficulties. Among the studies on emotional processing, all have considered the perceptual aspect, but most have only examined the facial channel. HD individuals have been found to be impaired in emotional perception, with a selective impairment in disgust and fear. Progression of the disease process ultimately places the person with HD in need of caregiving from family and friends, yet little is known about what factors might negatively impact the physical and mental well being of the caregiver. In the current study, an extensive examination of emotional perceptual functioning was carried out, wherein facial, prosodic, and lexical channels were investigated. Within each channel and across channels, the pattern of recognition of individual emotions was considered. Further, factors potentially related to caregiver burden were comprehensively explored.

Participants were 18 early- and middle-stage HD patients and 17 caregivers of HD patients. HD patients were compared to healthy control data on facial, prosodic, and

lexical perceptual identification tasks from the New York Emotion Battery (NYEB; Borod, Welkowitz, & Obler, 1992). Group comparisons revealed that HD subjects exhibited emotional perception deficits across all three emotional channels. After accounting for nonemotional perceptual functioning, impairments remained for the facial and lexical channels. HD tends to affect some basic emotions more than others, and does not seem to reflect more widespread deficits or cognitive decline. HD subjects recognized happiness the best, with impairment in disgust and fear recognition. These results are largely consistent across channels, with the exception of the prosodic channel, wherein happiness was also perceived poorly.

There was a significant positive association between the Zarit Burden Inventory (Zarit, Reever, & Bach-Peterson, 1980). and the Huntington's Disease Quality of Life Inventory for Carers (Aubeeluck & Buchanan, 2006). Through utilization of these two measures, various characteristics were found to be associated with increased burden. These were aggression, anger, behavioral disturbances, impaired cognition, impaired mood, lower self care abilities, younger caregiver age, financial strain, and limited social support.

Exploratory analyses revealed that within the HD group, the relationship between the facial and prosodic nonverbal channels was the strongest, with weaker relationships between the nonverbal and verbal channels, which is consistent with previous research for healthy adults and other neurological populations (e.g., Borod et al., 1998, 2000). Greater numbers of CAG repeats were associated with earlier age of onset, also consistent with the literature (The U.S.-Venezuela Collaborative Research Project & Wexler, 2004).

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Finally, I would like to express a sincere thanks to the Huntington's disease patients and their family members who participated in this project. I hope that their involvement serves to better our understanding of this devastating disease and to improve the quality of life of patients and their caregivers.

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INTRODUCTION

Defining Huntington's disease

Huntington's disease (HD) is an inherited, neurodegenerative disorder characterized by progressively worsening abnormalities of movement, cognition, and behavior. It is a fully penetrant autosomal dominant disorder. HD affects approximately 1 per 10,000 people in the Americas, Europe, Australia, and Asia (*Huntington's Disease*, 2002). Onset is typically in the prime of life; average age in Canadian, American, and Venezuelan populations is 40.36, 37.47, and 34.35 years, respectively (Rosenblatt et al., 2001). The latest known age of onset is 80. About 10% of cases start before the age of 20, with the earliest known onset at the age of 2. The disease progresses to death over an average of approximately 20 years (*Huntington's Disease*, 2002). Phenotypic expression, such as age of onset, disease duration, and progression of physical and mental symptoms vary considerably among individuals.

The HD Gene and the Huntingtin Protein

In 1993, the Huntington's Disease Collaborative Research Group, a team of researchers from around the world, isolated a gene IT-15 ("interesting transcript 15") on chromosome four (p16.3). The HD genetic defect is an unstable expanded polymorphic trinucleotide CAG (cytosine, adenosine, and guanine) repeat. CAG is the codon for the amino acid glutamine and the repeat produces polyglutamine, which encodes part of the huntingtin protein. Seventeen to 26 copies of the trinucleotide CAG is considered normal, and will not result in development of HD in the individual or their offspring. Twenty-seven to 35 copies is called the "intermediate range", as the allele is not associated with a HD phenotype in the individual who possesses it, but it is unstable and can expand during

transmission, resulting in repeat sizes of 36 or greater in the next or subsequent generations (the risk of which is greater in offspring of male carriers; Goldberg et al., 1995). Alleles with CAG repeats in the intermediate range are sometimes referred to as normal mutable alleles. Individuals with 36 to 39 repeats are in what is called the "reduced penetrance range"; these people do not always develop HD during their expected life span. More than 39 CAG repeats results in HD, and the greater the number of repeats, the earlier the age of onset (The U.S.-Venezuela Collaborative Research Project & Wexler, 2004). Paternal inheritance, in particular, tends to increase the number of repeats, such that the disease increases in severity and has an earlier onset in successive generations. This phenomenon is called "anticipation". In addition to age of onset, it is also thought that the CAG repeat number is correlated with neuropathologic severity.

In IT-15 heterozygous individuals, both the normal protein and a protein containing an expanded polyglutamine tract, transcribed by the CAG region, are expressed (The Huntington's Disease Collaborative Research Group, 1993). The normal function of huntingtin and the pathogenic mechanisms caused by the expanded polyglutamine of mutant huntingtin remain unknown. It is, however, known that huntingtin is required for normal early development, as mice with targeted knockout of the huntingtin gene are embryonic lethal (Nasir et al., 1995; J. K. White et al., 1997; N. M. White, 1988; Zeitlin, Liu, Chapman, Papaioannou, & Efstratiadis, 1995); although hemizygous knockout mice develop normally. Surprisingly, IT-15 homozygotes (i.e., a person with two proteins containing an expressed, expanded polyglutamine tract) do not manifest symptomatology any more severely than IT-15 heterozygotes.

It would seem intuitive that the neuropathological damage would occur in all of the structures that contained the huntingtin protein. However, this is not the case; huntingtin mRNA is expressed in almost all tissues of the body and homogenously throughout the brain in both people with and without HD; yet the neuropathology is limited to a discrete set of structures.

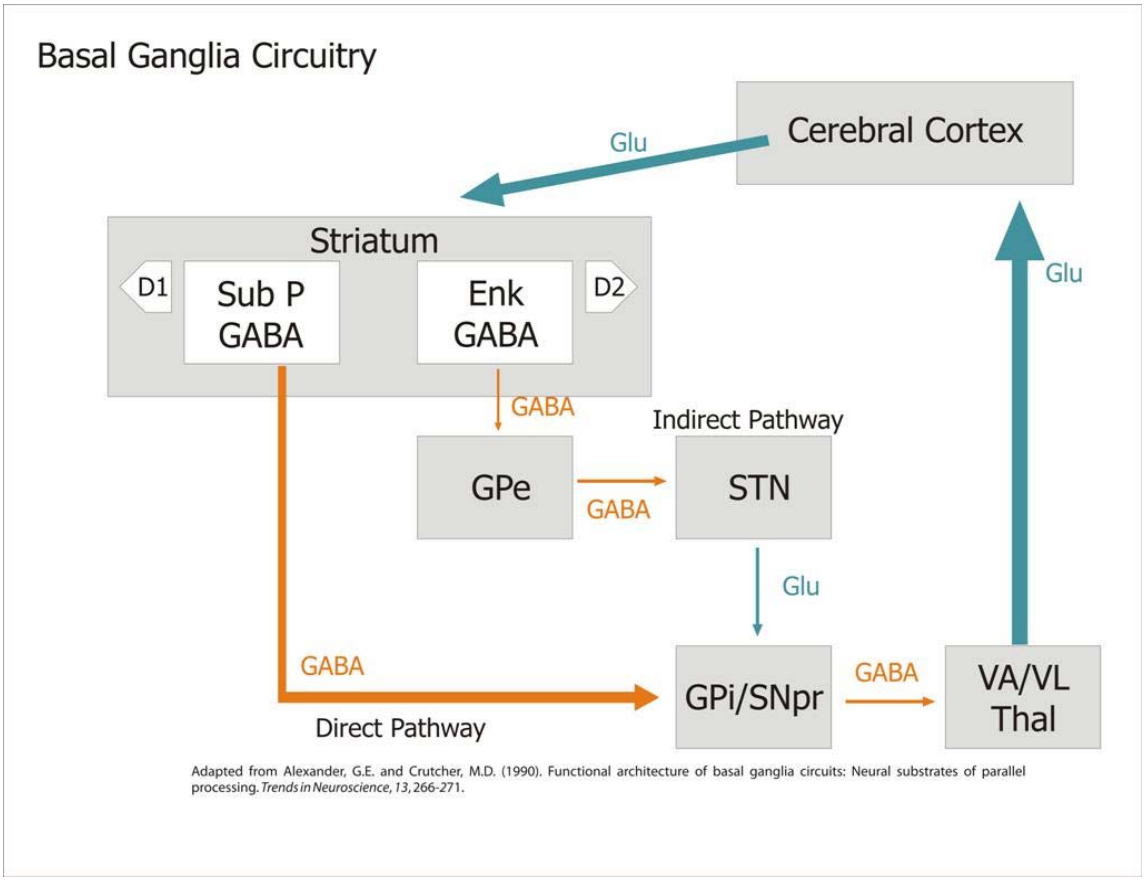
Pathology

A general loss of tissue occurs throughout the HD brain, with greatest atrophy in the basal ganglia. The pathology consists of a loss of gamma-aminobutyric acid (GABA)-containing medium-spiny neurons (comprising 90% of striatal neurons), progressing from dorsomedial caudate and dorsal putamen ventrally, with up to 60 percent loss of tissue in the striatum. The degeneration may also occur in the globus pallidus, cerebellum, thalamus, and cortex (Lezak, 1995; Myers, Schoenfeld, & Bird, 1985). In advanced cases, the whole brain weight may be decreased by 20-30 percent (Bird, 1980; Myers, Schoenfeld, & Bird, 1985).

There are two pathways that connect subcortical structures to regions of the frontal lobe that are impacted by HD pathology: the direct and the indirect. In the direct pathway, the substance P/dynorphin/GABA-containing neurons inhibit both neurons in the internal segment of the globus pallidus (GPi) and the substantia nigra pars reticulata (SNr), which in turn disinhibits (via GABA) the ventral anterior (VA) and ventral lateral (VL) thalamic nuclei, thereby exciting (via glutamate) the cortex. The functional result is increased movement. In the indirect pathway, the enkephalin/GABA-containing neurons inhibit the external segment of the globus pallidus (GPe), which in turn disinhibits (via GABA) the subthalamic nucleus (STN), leaving the STN free to excite (via glutamate)

the GPi/SNr, which then inhibits (via GABA) the VA/VL of the thalamus, thereby resulting in reduced cortical excitation. The functional result is decreased movement. The net result of increased movement from the direct pathway and decreased movement from the indirect pathway is a balanced system with no motor abnormalities (Cummings, 1993). Figure 1 provides a visual representation of the basal ganglia circuitry described above.

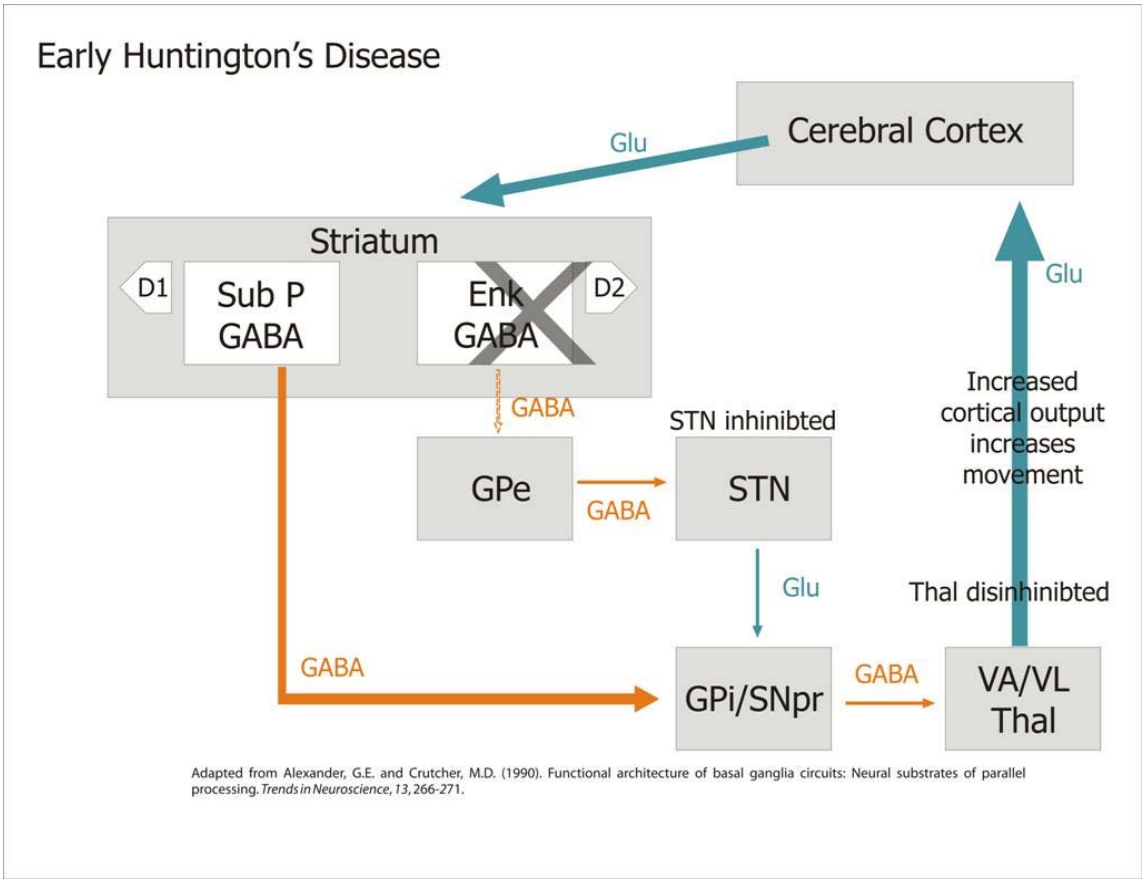
Figure 1. Healthy Basal Ganglia Circuitry



Within the striatum, HD affects projection neurons and spares interneurons. There are two classes of projection neurons, both of which contain GABA as a primary neurotransmitter: The substance P/dynorphin/GABA-containing D1 neurons, and the enkephalin/GABA-containing D2 neurons.

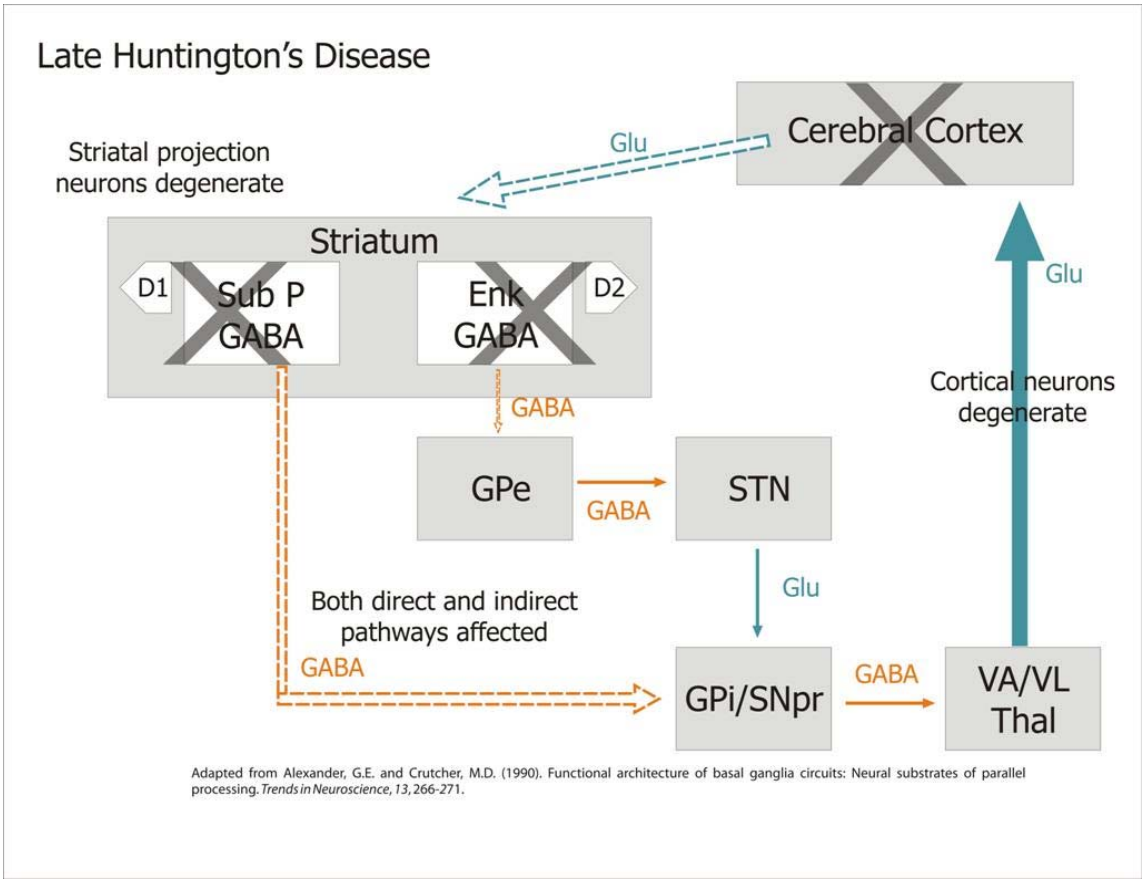
In HD, the neurons of the indirect pathway are lost first, causing thalamic disinhibition, resulting in increased choreiform movement. Figure 2 provides a visual representation of the basal ganglia circuitry in the early stages of HD.

Figure 2. Basal Ganglia Circuitry in Early Stage HD



In later stages of adult HD, the neurons of the direct pathway also become involved, causing thalamic inhibition, and resulting in rigidity and bradykinesia. Similar to late-stage adult-onset HD, degeneration of both direct and indirect pathway striatal neurons are seen in juvenile HD (Albin et al., 1988). Figure 3 provides a visual representation of the basal ganglia circuitry in the late stages of HD.

Figure 3. Basal Ganglia Circuitry in Late Stage HD



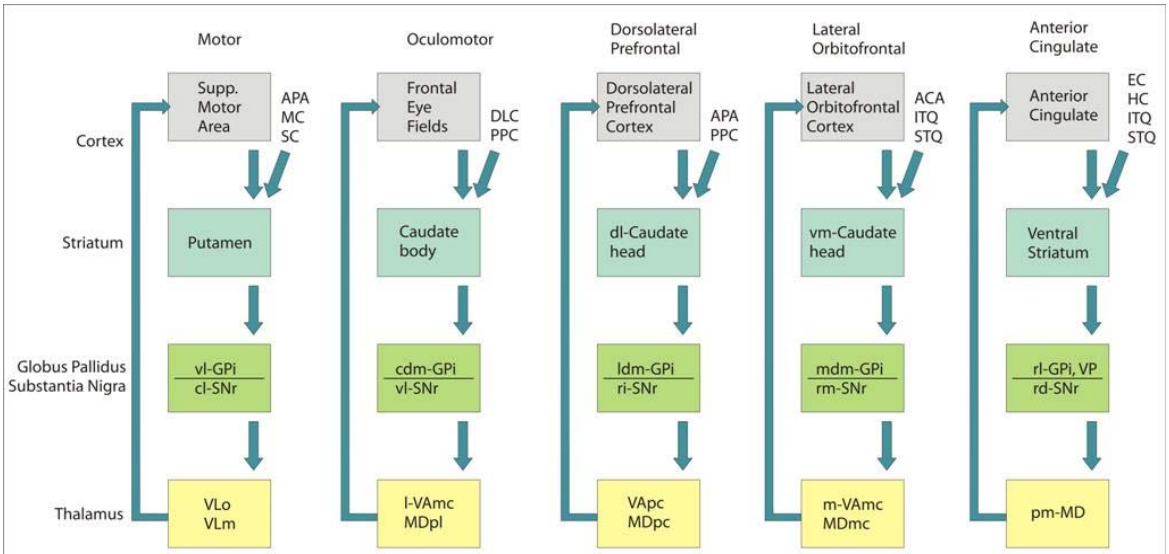
There are currently two main theories of pathogenesis in HD; the first implicates glutamate as the potential excitotoxic process, and the second implicates mitochondrial dysfunction. However, it is unclear if either of these theories is correct as neither excitotoxicity nor mitochondrial dysfunction has been shown to relate to the protein huntingtin.

HD Symptomatology and Neural Substrates

The main clinical features of HD are movement disorder, personality change, psychiatric disorder, and cognitive impairment. Five frontal-subcortical circuits have been identified (Alexander, DeLong, & Strick, 1986) that are important to understanding the various dysfunctions in Huntington's disease. (Although the *in vivo* frontal-subcortical

circuits are highly complex, the proposed circuits may be oversimplified in order to emphasize the relationships between structure and function of the circuits). Named according to their function or cortical site of origin, the circuits are: the motor circuit (dedicated to motor activity), the oculomotor circuit (dedicated to eye movements), the dorsolateral prefrontal circuit (dedicated to executive functions), the lateral orbital circuit (dedicated to social behavior), and the anterior cingulate circuit (dedicated to emotion). Each of these circuits shares the same member structures, which include the frontal lobe, striatum, substantia nigra (SN), globus pallidus (GP), and thalamus. Within each of the circuits, there are two pathways: a direct pathway and an indirect pathway (as described above in the Pathology section). These loops exist in parallel to each other, but may interact with each other, as well as with other regions of the brain that share functions with that circuit. Clinical symptomatology and its relevant neural correlates are detailed in the subsequent sections. The following diagram provides a visual representation of the five loops (direct only) proposed by Alexander, DeLong, and Strick (1986).

Figure 4. Parallel Organization of the Five Frontal Subcortical Circuits Proposed by Alexander, DeLong, and Strick



Abbreviations are as follows: APA = arcuate premotor area; EC = entorhinal cortex; GPi = globus pallidus, internal segment; HC = hippocampus; ITG = inferior temporal gyrus; MC = motor cortex; MDpl = dorsomedial nucleus pars paralamellaris; MDmc = dorsomedial nucleus pars magnocellularis; MDpc = dorsomedial nucleus pars parvocellularis; PPC = posterior parietal cortex; SC = somatosensory cortex; SNr = substantia nigra pars reticulata; STG = superior temporal gyrus; VAmc = ventral anterior nucleus pars magnocellularis; VApc = ventral anterior nucleus pars parvocellularis; VLm = ventral anterior nucleus pars medialis; VLo = ventral lateral nucleus pars oralis; VP = ventral pallidum; cl = caudolateral; cdm = caudal dorsomedial; dl = dorsolateral; l = lateral; ldm = lateral dorsomedial; m = medial; mdm = medial dorsomedial; pm = posteromedial; rd = rostrodorsal; rl = rostrolateral; rm = rostromedial; vm = ventromedial; vl = ventrolateral.
Adapted from Alexander, G. E., DeLong, M. R., & Strick, P. L. (1986). Parallel organization of functionally segregated circuits linking basal ganglia and *cortex. Annual Review of Neuroscience, 9,* 357-381.

Motor Dysfunction

Although HD is characterized by a triad of motor, cognitive, and behavioral symptoms, the most well-known of these is the physical, or motor, manifestation. Indeed, confirmation of the presence of the disease is made from the neurological examination, and not from psychiatric diagnosis or cognitive assessment. Chorea, the motor hallmark of the adult-onset form of the disorder, is defined as a "state of excessive, spontaneous movements, irregularly timed, randomly distributed and abrupt" (Barbeau et al., 1981). These uncontrollable movements are always present in states of arousal, increase with stress, and cease during sleep. Early in the disease, chorea may present as restlessness with mild, intermittent exaggeration of gesture and expression. As the disease progresses,

the fidgeting movements of the extremities incorporate the trunk in an unstable, dance-like gait until there is a continuous flow of disabling violent movements.

Although chorea is the most characteristic physical symptom of adult onset HD, other motor symptoms include athetosis, dystonia, bradykinesia, spasticity, akathisia, parkinsonism, postural reflex loss, ataxia, apraxia, clumsiness of voluntary movements, motor restlessness, and tremor. Difficulty initiating and executing movements as well as myoclonus may occur (Feigin, 1998; Folstein, Brandt, & Folstein, 1990; Rubin, 1995). Difficulty in speaking and swallowing secondary to the motor deficits interferes with communication and proper nutritional balance. Weight loss is common, partially due to the extra energy required for adventitious movements, but also due to increased resting basal energy expenditure. HD patients typically die of aspiration pneumonia, malnutrition, heart failure, or other complications (Myers, Schoenfeld, & Bird, 1985a). The juvenile form is more parkinsonian in nature, of which the prominent features are bradykinesia, rigidity, tremor, and seizures.

The circuit relevant to motor dysfunction is the *Motor Circuit*. It originates from neurons in the supplementary motor area (SMA), premotor cortex, motor cortex, and postcentral somatosensory cortex. Projection is largely to the putamen, where integration of movement and sensory information occurs. The putamen, in turn, projects to the ventrolateral GPi, GPe, and caudolateral SNr**.** The GPi/SNr connects to VA, VL, and centromedian (CM) thalamic nuclei, whose efferents are to the SMA, premotor cortex, and motor cortex (Cummings, 1993). The motor circuit is involved in preparatory pre-movement activity, serial processing of movements initiated in the cortex, and concurrent parallel processing in the structures of the circuit (Lichter, 1996).

Oculomotor Dysfunction

Abnormalities in ocular movement occur early in the disease in the majority of patients and are therefore one of the first noticeable clinical signs in a neurological examination. Abnormalities include slow saccades, impaired smooth pursuit, sluggish optokinetic nystagmus, and difficulty initiating saccades without moving the head or blinking.

The circuit relevant to oculomotor dysfunction is the *Oculomotor Circuit*. It originates in the frontal and supplementary eye fields and connects sequentially to the central body of the caudate. The caudate projects (via the direct and indirect pathways) to the dorsomedial GP and ventrolateral SN, which then projects to the VA and mediodorsal (MD) thalamic nuclei, and subsequently to the frontal eye fields and the superior colliculi (Cummings, 1993). The oculomotor circuit is involved in the control of saccadic eye movements (Lichter, 1996).

Cognitive Dysfunction

The gradual and progressive course of cognitive deficits tends to parallel the motor deterioration (Brandt, Strauss, Larus, Jensen, & Folstein, 1984). Longitudinal studies show a decline in intellectual function, although a relative conservation occurs with respect to other cognitive functions. Initially, language function is relatively preserved; however, as the disease progresses, HD patients show a significant decline in spontaneous speech production and in verbal fluency (Deus-Yela, Pujol, & Espert, 1997). Research shows (Podoll, Caspary, Lange, & Noth, 1988) that this can be attributed to both progressive neuropsychological (e.g., visuoperceptive impairment) and motor (e.g., dysarthria) changes. Deficits of memory and learning are the most distinguishing

cognitive characteristics. Present several years before the onset of motor symptoms, HD patients maintain the ability to store learned material, but have difficulty retrieving stored information (Butters, Sax, Mongtomery, & Tarlow, 1978; Fedio, Cox, Neophytides, Canal-Frederick, & Chase, 1979). Visuospatial processing and spatial manipulation are gravely altered in HD patients, but spatial judgment remains intact (Mohr et al., 1991). Executive dysfunctions in HD include an inability to generate strategies and to plan, organize, and program activities; rigidity in social conduct; a tendency to perseverate; and marked difficulty in maintaining or shifting sets (Deus-Yela, Pujol, & Espert, 1997). HD patients exhibit poor organizational and constructional strategies, poor memory search strategies, stimulus-bound behavior, mental rigidity and inflexibility, poor alternation between ideas, perseveration, poor inhibition of inappropriate responses, and difficulty focusing and sustaining attention (Deus-Yela, Pujol, & Espert, 1997; Lichter, 1996).

One cognitive deficit not commonly considered, but that deserves special attention, is impaired awareness. Potential explanations for illness or symptom unawareness in HD include a nonspecific reduction in insight due to dementia or frontal dysfunction, denial of illness as a psychological defense, and a physiological failure to experience abnormal movement. Snowden, Craufurd, Griffiths, and Neary (1998) investigated awareness of involuntary movements in a sample of 40 HD subjects. They ruled out the psychodynamic explanation for symptom unawareness because even though HD subjects failed to report experiencing choreiform movements, they accurately experienced other motor symptoms of their disorder (e.g., motor slowing). The authors also ruled out the cognitive hypothesis due to the absence of a relationship between

failure to report motor symptoms and degree of cognitive impairment. Snowden et al. reasoned that the physiological explanation was the most salient because the HD subjects were aware of the consequences of their movement disorders (e.g., dropping things) yet did not subjectively experience the chorea. In contrast to Snowden et al.'s study (1998), Deckel and Morrison (1996), Vitale et al. (2001), and Hoth (2006) favored a cognitive explanation, with unawareness directly related to disease duration and severity (Deckel & Morrison, 1996; Vitale et al., 2001). One study (Hoth, 2006) explored the relationship between awareness and cognitive dysfunction. It was found that patients' self ratings of their behavioral problems were not associated with their objective clinical performance. Patients overestimated their abilities across domains of fundamental behavioral control, social-emotional appropriateness, and activities of daily living. The authors suggested that an association exists between impaired self-awareness and deficits in executive functioning and memory.

The circuit relevant to cognitive dysfunction in HD is the *Dorsolateral Prefrontal Circuit* (DLPFC)**.** It originates in Brodmann's areas 9 and 10 and projects to the dorsolateral caudate, the lateral mediodorsal GPi, and the rostrolateral SNr via the direct pathway. The indirect pathway sends fibers to the dorsal GPe, the lateral STN, and the GPi/SNr. Output of the basal ganglia projects to parvocellular portions of VA and MD thalamic nuclei, and back to areas 9 and 10 (Cummings, 1993). The DLPF circuitry is involved in "executive functions": control, regulation, and integration of sensory information; organization of behavioral responses; use of verbal skills; anticipation; goal selection; motivation; attention; planning; monitoring; the use of feedback in task performance; and working memory (Lichter, 1996).

Apathy and Aggression

Alterations in behaviors such as irritability, social withdrawal, apathy, and aggression frequently occur in HD. These behavioral symptoms can have the greatest impact on caregivers and can occur many years before the onset of motor and cognitive symptoms. Apathy is the most frequent and characteristic personality change associated with HD. Patients experience loss of motivation, initiative, and spontaneous expression. It is a particularly troublesome symptom, as it is often misdiagnosed and mistreated as depression. Apathy, once present, will worsen over time.

Irritability and aggression are personality changes that severely impact relationships. The patient may become irritable for no reason and can easily be provoked into an outburst of angry or violent behavior (Burns, S.E. Folstein, Brandt, & M.F. Folstein, 1990). These episodes can last for days and be retriggered by the slightest provocation. This behavior tends to be directed toward one family member, usually the primary caregiver. Prevalence is difficult to determine, as operational definitions often differ from study to study. Folstein et al. found a prevalence of 31% of HD patients suffering from "intermittent explosive disorder" (S.E. Folstein, Chase, Wahl, McDonnell, & M.F. Folstein, 1987). Craufurd et al. (2001) reported finding clinically significant irritability in 57% of males and 49% of females. Forty percent of patients displayed verbal outbursts of temper and 22% displayed threatening behavior or actual violence in the 4 weeks prior to the interview (Craufurd, Thompson, & Snowden, 2001).

The circuit relevant to the manifestation of apathy and aggression is the *Anterior Cingulate Circuit* (AC). It originates in Brodmann's area 24 and projects to the ventromedial caudate, ventral putamen, nucleus accumbens, and olfactory tubercle. The

ventral striatum also receives limbic input from the hippocampus, amygdala, and entorhinal cortices. Projections leaving these areas innervate the rostromedial GPi, ventral pallidum, and rostrodorsal SNr. From here, the pathway continues to the MD thalamic nucleus and then back to anterior cingulate cortex (Cummings, 1993, 1996). The anterior cingulate circuit plays a role in motivated behavior, procedural learning, and personality (Lichter, 1996).

Personality Changes and Emotional Dysfunction

The early appearance of personality alterations in Huntington's disease includes depression, anxiety, obsessive-compulsive disorder (OCD), behavioral disinhibition, impulsive or destructive manic behavior, emotional lability, failure to respond to social cues, and lack of empathy (Berrios et al., 2002; S. E. Folstein, Chase, Wahl, McDonnell, & M.F. Folstein, 1987). These patients lack judgment and social tact, and may exhibit inappropriate jocularity. Decreased impulse inhibition may be associated with improper sexual remarks or gestures and other antisocial acts.

Reported psychiatric manifestations in HD are affective disorder, schizophrenia and schizophrenia-like disorders, OCD, altered sexual behavior (which, in turn, increases fecundity), and anxiety. The most universally cited psychiatric manifestation of HD, however, is depression (Caine & Shoulson, 1983; S.E. Folstein, Abbott, Chase, Jensen, & M.F. Folstein, 1983). Living with a debilitating disease can lead to feelings of demoralization, hopelessness, loss of self-worth, and expression of a wish to die. This reaction may seem quite normal given the tremendous losses the person has and will suffer. However, rationalizing the depressive symptoms as reactive can be potentially dangerous in that clinicians may fail to aggressively treat the symptoms. Although

depression may be reactive, it is known that depression is clearly an independent factor. In fact, depression predates the motor and cognitive symptoms of the disease by 2 to 20 years in two thirds of the cases (Cummings, 1995; S. E. Folstein & M.F. Folstein, 1983). Depression is generally more common among patients with later onset HD.

Reported rates of major affective disorders in the literature range from 1% to 55% (De Marchi & Mennella, 2000). Caine and Shoulson identified 11 patients (37%) who met criteria for either major depression or dysthymia (Caine & Shoulson, 1983). S.E. Folstein and M.F. Folstein (1983) investigated a group of HD patients and their relatives and found that 36 patients (41%) with HD were also suffering from major affective disorder (28 unipolar and eight bipolar). Within their cohort of patients, it was found that the affective disorder preceded the onset of chorea by an average of 5.1 years (S.E. Folstein & M.F. Folstein, 1983). One study (Mindham, Steele, C. Folstein, & Lucas, 1985) looked at the possibility that depression is a nonspecific feature of dementing illnesses. They used Alzheimer's disease (AD) patients as a comparison group, and found that in those with HD, there were twice as many cases of major affective disorder. Shiwach reported a lifetime prevalence of depression of 39% in 110 patients with HD. Among these patients, 33% showed depressive symptoms an average of 4.3 years in advance of motor symptoms (Shiwach, 1994).

Directly associated with the prevalence of depression in HD is the existence of suicide. It is assumed that the high rate of suicide is an understandable response to an almost intolerable situation. In a survey of 35 at-risk individuals, approximately half indicated they would commit suicide if they became ill (Wexler, 1979). However, patients who have expressed an intention to kill themselves when the condition becomes

more advanced rarely do so. It is thought that the cognitive and personality changes, such as apathy and affective blunting, become severe enough to make suicide unlikely. Paulsen and colleagues (2005) delineated the rates of suicidal ideation (SI) based on the stage of the illness (c.f., the Stages of Illness section): SI was present in 9.1% of at-risk persons with a normal neurological examination, 19.8% of at-risk persons with soft neurological signs, 23.5% of persons with probable HD, 16.7% of persons diagnosed and in stage 1, 21.6% of persons in stage 2, 19.5% of persons in stage 3, 14.1% of persons in stage 4, and 9.8% of persons in stage 5 (Paulsen, Hoth, Nehl, Stierman, & The Huntington Study Group, 2005). Therefore, two critical periods for increased risk of suicide occur at the first appearance of HD symptoms, and in stage 2, at a time when activities begin to be restricted. Suicide is the third most common cause of death in HD patients after pneumonia and cardiovascular disease. Schoenfeld et al. (1984) found that rates of suicide in HD patients living in New England was at a prevalence of 4% compared to the 1% occurrence of suicide in the general population; the increased suicide rate was particularly prevalent in the 50-69 year-old range. Additionally, the rate of suicide was four times greater in those with suspected HD than among the diagnosed cases (Schoenfeld et al., 1984).

Another psychiatric disorder commonly reported in HD is schizophrenia-like psychosis. Diagnosis is difficult as the disorder shares symptoms with those of HD, such as social withdrawal, emotional blunting, and loss of volition. Research indicates a 3-6% prevalence of actual schizophrenia in patients with HD. However, when considering psychotic features in a more general sense (e.g., hallucinations, delusions, or disorganized thinking), the prevalence increases to 30% (Naarding, Kremer, & Zitman, 2001). Positron

Emission Tomography (PET) studies of HD patients with schizophrenia-like symptoms exhibit the hypofrontality as seen in PET images of schizophrenic patients. Interestingly, though depression is reported to occur more commonly in those with later-onset HD, psychiatric symptoms in HD are reported to occur more often in persons with earlier onset HD. Psychotic features of the disease may be treated with neuroleptics, however there is the risk of worsening the choreic movements (Rosenblatt et al., 2001).

Reports of promiscuous behavior and sexual deviations in HD patients have been reported as early as George Huntington's original description in 1872. Studies in the 1960s and 1970s revealed sexual aberrations such as morbid sexual jealousy, indecent exposure, homosexual assault, incestuous sodomy, voyeurism, assault on females, and promiscuity. One such study that was frequently referenced found 19% of participants to display hypersexuality, and 11% to exhibit hyposexuality (Dewhurst, Oliver, & McKnight, 1970). A more recently published paper, which used semistructured interview methods and operationally defined diagnostic criteria, found that 63% of male patients and 75% of female patients had hypoactive sexual desire, whereas 30% of HD males and 25% of females experienced hypersexuality. This same study also found that 56% of males and 42% of females had inhibited orgasm, and 19% of males and 8% of females exhibited paraphilias such as sexual aggression, exhibitionism, or voyeurism (Federoff, Peyser, Franz, & S.E. Folstein, 1994).

Several other psychiatric disorders exist in patients with HD, but are not common; obsessive-compulsive disorder (OCD) is one example. True obsessive-compulsive disorder is rare, but patients may become obsessively preoccupied about cleanliness or the manner in which particular activities are performed. In some cases, it is possible that

the OCD-like behaviors may actually be the physical manifestation of the stereotyped repetitive choreic movements. When OCD is truly present, it is treated with serotonergic antidepressant agents. Mania is also observed at a slightly increased prevalence as compared to the general population. Brief episodes of hypomania, as manifested by increased levels of activity, pressured speech, uncharacteristic cheerfulness, and large and inappropriate purchases, were observed in 10% of the patients (S.E. Folstein, Chase, Wahl, McDonnell, & M.F. Folstein, 1987). Anxiety is also exhibited by some HD patients.

Although the literature regarding psychiatric symptoms in HD as inconsistent and sometimes confusing, it seems that psychotic disorders occur in a small minority of patients with HD (Naarding, Kremer, & Zitman, 2001). Prevalence rates in some studies were as low as 3-5%, and in other studies with higher rates of prevalence, the methodology was questionable. One thing that is certain, however, is that the psychiatric presentation within HD is quite variable and unstable over time. Careful consideration must be given to the diagnosis of psychiatric disorders, and constant review of symptomatology is necessary to track the evolution of the psychiatric symptoms.

The circuit relevant to personality change and emotional dysfunction, the neocortical representation of the limbic system, is the *Lateral Orbitofrontal Circuit* (LO). It originates in Brodmann's areas 10 and 11 and projects to the ventromedial caudate, the medial portion of the GPi, and the rostromedial SNr. The ventromedial caudate sends an indirect loop through the dorsal GPe to the lateral STN, the GPi, and the SNr. Projections are sent from the GPi and SNr to the medial magnocellular division of the VA thalamus, as well as the inferomedial sector of the magnocellular division of the MD thalamic

nucleus. The circuit then projects to the orbitofrontal cortex. The LO circuit communicates directly with the amygdala, hippocampus, and olfactory system (Cummings, 1993). This circuit appears to play a role in mediating empathetic and socially appropriate responses; specifically, it's involved in the determination of the appropriate time, place, and strategy for environmentally elicited behavioral responses, impulse control, maintenance of set, and maintenance of ongoing behaviors (Lichter, 1996).

Stages of Illness

Although the prior section outlines the myriad of deficits that occur throughout the course of the illness, some attention to the progression is warranted. Progression is complex due to the variability in the way that the disease affects individuals and the potential range of deficits that can occur. Despite these obstacles, neurological staging was established during development of the Unified Huntington's Disease Rating Scale (UHDRS), a tool that provides a uniform assessment of the clinical features and course of HD (UHDRS; Shoulson & Fahn, 1979). This staging is known as the Total Functional Capacity Scale (TFC) and begins with the onset of symptoms (Stage 1) to the end stages of life (Stage 5). Determination of stage is accomplished through assessment of capacity for maintaining employment, handling financial affairs, managing domestic responsibilities, performing activities of daily living, and self-care. The first stage is typified by ability to maintain independence both at home and at work. In the second stage, the individual remains employable, but has to work at a lower capacity due to the progression of physical and cognitive symptoms. Stage three's prominent features are inability to work and failure to manage household responsibilities. The fourth stage is

characterized by inability to carry out activities of daily living; individuals are generally placed in a skilled nursing facility during this stage. In the fifth stage, the individual requires complete care and support with all activities of daily living.

Emotional Functioning in HD

Although depression is the emotional disorder most frequently associated with HD, little research has been performed on specific deficits of emotional processing in HD. In the current paper, emotional processing refers to the expression, perception, and experience of emotions across facial, prosodic, and lexical channels (e.g., Borod, 1993). To date, there are 10 experimental studies in the literature that have attempted to delineate emotional functioning in Huntington's disease patients and 1 commentary article. The majority of research has investigated the perceptual aspect of emotion, most of which utilized facial stimuli (8 out of 10 studies), whereas a few of these included studies on other channels of emotional functioning. In general, the results of these studies have revealed impaired perception of disgust, with some discrete deficits seen in other emotions. Four of the ten studies also assessed HD individuals' experience of emotion through the use of questionnaires. One study investigated expression and perception of emotions depicted in images and scenes (Hayes, Stevenson, & Coltheart, 2007). In the following text, this literature will be described. Below is a summary of the various aspects of emotion investigated in these 10 existing experimental studies about emotional processing in HD.

Table 1. Aspects of Emotion Investigated in Existing HD Emotion Literature

Study	Perception	Experience	Expression	Other
Gray et al., 1997	Facial	-	-	-
Hayes et al., 2007	Prosodic & Lexical	Disgust	Lexical	Categorization of images and scenes
Jacobs et al., 1995	Facial			
Milders et al., 2003	Facial	-	-	-
Montagne et al., 2006	Facial	-	-	-
Speedie et al., 1990	Prosodic	-	-	-
Sprenglemeyer et al., (1996)	Facial & Prosodic	Anger, Fear, and Disgust	-	-
Sprenglemeyer et al., (1997)	Facial	Anger, Fear, and Disgust	-	-
Sprenglemeyer et al., (2006)	Facial & Prosodic	Anger, Fear, and Disgust	-	-
Wang et al., 2003	Facial	-	-	-

Linguistic prosody is the use of stress and rhythm to change the lexical meaning of speech (for instance, the pronunciation of "greenhouse" versus "green house" (Benke, Bosch, & Andree, 1998; Blonder, R.E. Gur, & R.C. Gur, 1989). Linguistic dysprosody is the inability to differentiate the communication of a statement versus an inquiry versus an exclamation. Emotional prosody is the use of intonation and stress to communicate emotion, to differentiate between serious and sarcastic statements, and to add emotional content to lexically neutral phrases (e.g. Ross, 2000; Ross & Monnot, 2008; Scott, Caird, & Williams, 1984). Emotional dysprosody is the inability to vocally express feelings (e.g., excitement, anger, sadness, etc.) or convey verbal humor (Scott, Caird, & Williams,

1984). Emotional dysprosody is often interpreted by others as emotional indifference (Benke, Bosch, & Andree, 1998).

Speedie, Brake, Folstein, Bowers, and Heilman (1990) investigated emotional and linguistic (as described in the previous paragraph) prosody in 6 early-stage HD patients, 8 right-hemisphere-damaged stroke patients, 9 left-hemisphere-damaged stroke patients and healthy controls (Speedie, et al., 1990). Subjects were presented with happy, sad, and angry intoned sentences that had been speech-filtered to prevent recognition of the words. Subjects were instructed to indicate which emotion was being represented in the sentence by pointing to a response card, which contained drawings of a man's face portraying each of the emotions, paired with the appropriate label for the emotion printed below each face. The linguistic prosody task was presented in the same fashion, and subjects had to decide if the sentences presented to them were portraying a question, command, or statement. HD and stroke patients were impaired in the recognition of both types of prosody compared to controls. The authors noted that HD patients also performed poorly on a task of musical tonal memory (Seashore Measures of Musical Talents), indicating a compromise in the ability to understand the subtleties of prosodic communication in general (Speedie, et al., 1990).

Jacobs, Shuren, and Heilman (1995) tested five patients with Huntington's disease on measures of neutral and emotional face discrimination and matching. Emotions used in this study were happiness, sadness, anger, and fear. Four patients were impaired on at least one task of face discrimination and all patients were impaired on at least one task of emotional facial discrimination (Jacobs, et al., 1995).

Sprengelmeyer, Rausch, Eysel, and Przuntek (1996) investigated the recognition of facial expressions in HD patients. Using computer-interpolated continua produced from six basic emotions, patients were asked to identify the emotions represented by black-and-white slides of eight female and six male individuals exhibiting facial expressions of emotion (Ekman & Friesen, 1976). Overall, it was found that HD patients performed significantly worse than normal controls on the facial emotion recognition task. Exploration of the pattern of individual emotion recognition accuracy revealed that HD subjects perceived happiness, sadness, and surprise as well as normal controls, but that they were significantly worse at perceiving anger, fear, and disgust (Sprengelmeyer et al., 1996), with poorest recognition for disgust.

In a follow-up study (Sprengelmeyer et al., 1997), the authors investigated two patients from the original study, who showed severe and selective deficits in the perception of facial emotion. The technique of morphing images was used to generate three separate experimental tasks. Morphing, a process by which a graphics program takes two prototypical images and creates a series of interpolated images along a continuum between the prototypes, was initially used to investigate emotion recognition by Etcoff and Magee (1992) and identity recognition by Beal and Keil (1995). In the Sprenglemeyer et al. study (1997), images began at one prototype and were morphed 10%, 30%, 50%, 70%, and 90% toward the other prototype to create a series of images ranging from predominantly one prototype to predominantly another prototype. The first task involved recognition of a continuum ranging from sadness to happiness and from fear to anger. The second task used an emotion “hexagon”, wherein a bipolar continuum was created for each of the six basic emotions from the Ekman and Friesen series (Ekman

& Friesen, 1976). Emotions were ordered by placing each one adjacent to the one it would most likely be confused with, creating the following continua: happiness to surprise, surprise to fear, fear to sadness, sadness to disgust, disgust to anger, and anger to happiness. In the final task, emotion "megamixes" were used to further explore impairments of disgust and fear found in the emotion hexagon task. The megamix is similar to the hexagon in that a series of bipolar continua are created. However, in this case, two megamixes are created in which each basic emotion plus neutral are morphed toward the prototype (e.g., in the disgust megamix, continua are neutral-disgust, happiness-disgust, surprise-disgust, etc.). Deficits in the recognition of fear and disgust were evident for one of the subjects, whereas the other subject demonstrated significant deficits in the recognition of disgust and mild impairment in the recognition of fear. Neither subject showed deficits in the recognition of happiness, sadness, surprise, or anger. Questionnaires examining self-assessed feelings of anger, disgust, and fear indicated that the two subjects' scores were almost identical to the normative mean, but scores on the disgust and fear questionnaires were approximately one standard deviation (SD) below that of the normative mean. Based on the relatively greater deficit in disgust relative to fear recognition in the HD subjects, paired with findings from the literature of relatively greater impairments in fear relative to disgust recognition in patients with bilateral amygdala damage (Adolphs, Tranel, H. Damasio, & A. Damasio, 1994), the authors postulated that fear and disgust may have separate neural substrates from each other (Sprengelmeyer et al., 1997).

Gray, Young, Barker, Curtis, and Gibson (1997) investigated the recognition of facial expression in 40 presymptomatic at-risk individuals (17 with the HD gene, at-risk

+; 23 without, at-risk -). Twenty-four photographs of faces representing the six basic facial expressions of emotion from the Ekman and Friesen series (Ekman & Friesen, 1976) were presented in random order. Subjects were asked to indicate which emotion was represented by each photograph. ANOVA results indicated a trend toward significance (p=.09) in recognition of emotions between at-risk + and at-risk – subjects, with at-risk + individuals showing a slight impairment. Further examination of the individual emotions revealed that the at-risk + individuals performed significantly more poorly than did the at-risk – individuals in the recognition of disgust. The groups did not differ in the recognition of any other emotion.

Halligan (1998) published a commentary on emotion research in HD and proposed that Huntington's disease findings on emotional perception support neuroanatomical localization of distinct emotions. Specifically, Halligan (1998) used Sprenglemeyer et al. (1996) and Gray et al.'s (1997) findings to illustrate that there can be a selective impairment in one emotion (i.e., disgust) within the context of other emotions being intact. In this vein, it is logical that there would be distinct neuroanatomical correlates for disgust as well as for other emotions. Support for this notion comes from a functional magnetic resonance imaging study in a healthy control population, which revealed predominant anterior insular cortex activation when participants were exposed to facial expressions of disgust (Philips et al., 1997). Additional substantiation for emotion-anatomic correlates is found in a study of patients with bilateral amygdala lesions, who showed selective loss of the ability to recognize fear and anger in the context of intact recognition of other facial emotional expressions (Adolphs et al., 1994). Halligan proposed that future studies should explore the

possibility of emotion-specific neuroanatomical structures in other disease processes and emotional modalities.

Wang, Hoosain, Yang, Meng, and Wang (2003) studied six symptomatic HD patients and 16 healthy controls. Modeled after Etcoff and Magee's (1992) original emotion study with healthy controls, a continuum of morphed photographs covering the emotions of happiness, surprise, fear, sadness, disgust, and anger was presented to subjects, who were then asked to identify the emotion in each photograph. HD patients showed poor recognition of disgust as compared to controls (HD patients only achieved 38% of the total correct achieved by that of controls). Accuracy of recognition of all other emotions was statistically similar to that of controls (happiness, 99%; surprise, 90%; fear, 77%; sadness, 85%; and anger 89%; Wang et al., 2003).

Milders, Crawford, Lamb, and Simpson (2003) designed a study which included 20 symptomatic HD individuals, 20 presymptomatic HD individuals, and 20 healthy controls. The authors administered one emotion task in which participants were shown 60 photographs of facial expressions and asked to select which of six emotions was being expressed. Milders et al. (2003) also administered a similar task in which participants were asked to match one of four facial emotional expressions to each other. Symptomatic individuals were impaired in recognizing anger, disgust, sadness, and fear, with the impairment for fear being greatest. The presymptomatic individuals evidenced a trend toward impaired recognition of fearful faces. On the matching task, HD individuals performed significantly worse than controls on all four expressions and significantly poorer than presymptomatic individuals on fear, disgust, and anger, but not sadness. The matching scores of the presymptomatic individuals and controls did not differ for any

expression. In summary, the authors concluded that the observed impairment for fear recognition was a product of the task difficulty and that the basal ganglia is not likely to be involved in disgust recognition.

Montagne et al. (2006) observed that previous studies (Lough et al., 2006; Milders, Crawford, Lamb, & Simpson, 2003) have reported ceiling effects for specific deficits in the perception of disgust, even when participants have emotion-perception deficits. Montagne et al. (2006) asserted that this is likely due to utilization of stimuli that are both static and represent full-blown emotions. The authors confronted these methodological drawbacks by investigating affective facial expressions at different intensities in early symptomatic HD. They presented video of persons expressing varying degrees of different facial emotions. Results revealed that a specific impairment in disgust and anger recognition was present even at low emotion intensity, with early-stage individuals, and with small sample sizes.

Sprenglemeyer, Schroeder, Young, and Epplen (2006) investigated whether or not there is a selective disgust-processing deficit in presymptomatic individuals by presenting emotionally laden stimuli in both facial and prosodic channels. Participants were 22 at-risk individuals tested at three different time points over the period of a year. Genetic testing performed between the first and second behavioral testing revealed that 14 of the at-risk individuals were HD positive and eight were HD negative. An additional control group of 37 was used as a basis for comparison. All participants were administered two facial expression recognition tests (assessing happiness, surprise, fear, sadness, disgust, and anger) adapted from the Ekman 60 Faces test and the Emotion Hexagon test from the FEEST (Young, Perrett, Calder, Sprengelmeyer, & Ekman, 2002), a verbal prosodic

emotional recognition task (assessing happy, surprised, fearful, sad, disgusted, or angry vocal intonation), a nonverbal emotional sounds recognition task (assessing sounds of happiness, sadness, anger, fear, disgust, and surprise vocalizations), and three self-administered questionnaires which ask the subject to rate things and experiences that may cause anger, fear, and disgust. Results revealed that recognition of facial expressions of disgust was significantly impaired on all three assessments in the HD-positive group, while recognition of facial emotions and the experience of emotions were largely unaffected. At-risk individuals, who tested negative for the gene, performed significantly worse than healthy controls on recognizing angry facial expressions. The authors inferred that deficits in the recognition of facial expressions of disgust are an early symptom of HD. Based on these results, there seems to be a unimodal process of disgust recognition, impacting only the facial channel. However, the prosodic tests employed may not have been sensitive enough to detect mild deficits, and convincing evidence of a unimodal hypothesis would require evidence of impaired auditory, but not facial, disgust recognition in a different patient population. Further research of auditory recognition of emotion in presymptomatic HD patients was suggested (Sprengelmeyer, Schroeder, Young, & Epplen, 2006).

Given the fairly consistent reports of impairment for the recognition of facial expressions of disgust, Hayes et al. (2007) investigated whether this disgust impairment extended into other areas of functioning. They assessed 14 HD patients and 14 healthy controls on seven tasks: 1) knowledge for situational determinants of emotion, 2) recognition of emotion expressed in nonverbal vocalizations, 3) recognition of emotional content of explicit lexical stimuli, 4) recognition of emotional content in emotionally

laden pictures, 5) a disgust experience questionnaire, 6) a measure of olfactory hedonic responsiveness, and 7) a measure of gustatory perception. No control tasks were administered for any of these experimental tasks, nor were there any neutral task stimuli. HD patients enrolled in the study were free of significant neurological history other than HD.

The first task assessed participants' declarative knowledge of emotions by having them describe prototypical situations of happiness, sadness, anger, fear, disgust, and surprise. Patients generated fewer examples of disgusting situations than any other emotion as compared to healthy controls. The examples that were generated were then rated for intensity of emotion. It was found that HD patients' examples of happy, surprising, frightening, sad, and angry situations were not significantly different in intensity than those of controls. However, HD patients' disgusting examples were rated as significantly less disgusting as compared to healthy controls (Hayes, Stevenson, & Coltheart, 2007).

The second task administered involved recognition of emotion in nonverbal vocalizations. Participants were presented with recordings of nonverbal vocalizations and asked to identify the emotion being represented as disgust (e.g., retching), sadness (e.g., crying), anger (e.g., growls), or fear (e.g., gasping). HD patients and controls did not differ in their ability to classify vocalizations of fear or sadness, but they did recognize significantly fewer vocal expressions of anger and disgust than did controls. In the third task that was administered, participants were asked to categorize emotionally laden words as representing disgust (e.g., maggot), sadness (e.g., orphan), fear (e.g., intruder), or happiness (e.g., sunshine). It was found that HD patients and healthy controls did not

differ in performance on classification of any category of emotion. Task four comprised presentation of emotionally laden images and scenes, which participants were asked to identify as representing disgust (e.g., cockroach), fear (e.g., image of war), sadness (e.g., mourning party), or happiness (e.g., baby animals). HD patients did not differ in classification of scenes conveying happiness, sadness, or fear, but they performed significantly worse in classification of disgusting scenes.

The fifth task involved filling out a questionnaire assessing sensitivity to disgusting scenarios (e.g., "If I see someone vomit, it makes me sick to my stomach"); patients and controls did not differ in their total scores on this measure. In task six, participants were presented with five different odorants and asked to compare odors for degree of unpleasantness, identify the smell, rate the intensity of the smell, and rate the smell on scales corresponding to social traits (e.g., sociable-unsociable, clean-dirty, intelligent-unintelligent, and attractive-unattractive). The results of this task revealed a significant abnormality of hedonic odor responsiveness. Specifically, HD patients rated bad smelling odorants as smelling "worse" less frequently than did controls, and they rated pleasant smelling odorants as smelling "better" less frequently than did controls. A restricted hedonic range was also found in HD patients. Patient ratings deviated significantly for social traits as compared to healthy controls. That is, HD subjects were less likely than controls to associate bad odorants (i.e., manure and fermented shrimp) with the social traits of unattractiveness and dirtiness. Further, odor identification was less accurate and intensity ratings were reduced. Finally, in task seven, participants were presented with sweet, sour, salty, and bitter stimuli and asked to rate the taste on hedonics and intensity. Performance of HD patients was significantly worse only for sweet and

salty tastes. Based on findings from the seven tasks, Hayes et al. (2007) concluded that the impairment of disgust across various input domains suggests a fundamental impairment in disgust processing.

As outlined in the prior text, previous research on emotional functioning in HD has focused primarily on facial perception. Many of these studies have found that a discrete deficit in the perception of disgust exists, so much so that Hayes, Stevenson, and Coltheart's (2007) study focused entirely on disgust functioning across various communication channels. The knowledge gained from the research to date is indeed a good start, but a systematic investigation is called for in order to better characterize emotional perception deficits in HD across other domains.

The current study aims to comprehensively examine emotional perceptual functioning of a range of positive and negative emotions across the facial, prosodic, and lexical channels. A novel aspect of this research is the use of the lexical/verbal channel, which has received relatively little attention in the literature (c.f., Blonder, R.E. Gur, & R.C. Gur, 1989; Hayes et al., 2007). Ability to recognize and distinguish emotional expressions in others is crucial to facilitating normal social interaction. It is hoped that a better understanding of HD patients' emotional perceptual capacity, which presumably affects their overall nonverbal communication skills, will lead to the development of therapies aimed at improving emotional health in persons with Huntington's disease and to improvement in communication patterns between Huntington's disease patients and their caregivers.

Caregiver Burden

Caregiver and Burden Defined

In the formal sense, a caregiver is a mental health professional. However, much more often than not, the caregiver is an informal one, a non-professional who provides partial or full assistance to a disabled or ill person. Caregivers can be friends or family members, but, most often they are spouses (Kessler, 1993). Caregiving activities range from assistance with instrumental activities of daily living (e.g., shopping and community mobility) to assistance with basic activities of daily living (e.g., feeding and bathing). Providing assistance has been found to have adverse psychological and long-term negative health effects. Caregivers are at substantially greater risk for physical illness and decreased psychological well-being (Whitlatch, Zarit, & von Eye, 1991). Caregivers have even been found to have increased rates of death compared to age- and sex-matched controls who are not serving as caregivers (Schulz & Beach, 1999). These specific physical, emotional, and financial strains are known as caregiver burden.

Burden and Other Diseases and Illnesses

Consensus about the importance of family caregivers has led to numerous research efforts to better understand the consequences of caregiving. There are a handful of existing studies which consider caregiver burden in the context of HD, whereas the bulk of research on caregiver burden has looked at AD and elderly populations. Other studies have focused on caregivers of cancer, stroke, traumatic brain injury, and multiple sclerosis patients. From this combined literature, it is evident that there are certain common contributory factors to the level of burden experienced by the caregiver. Factors

contributing to burden across a variety of different illnesses will be discussed first, followed by a focused discussion of burden as it specifically pertains to HD.

One significant contributor to caregiver burden is behavioral disturbances. A study of geriatric patients referred for memory problems showed that the rate of change in behavior problems, such as fear of being alone, anger, wandering, and aggression, was a significant predictor in whether the caregivers experienced role overload (Bedard, Molloy, Pedlar, Levar, & Stones, 1997). Hooker et al. (2002) looked at a group of AD patients and assessed the extent to which behavioral problems affected caregivers. They found two factors strongly associated with declining mental and physical health for the caregiver: increases in problematic behaviors of the patient and caring for someone in a long-term setting (Hooker et al., 2002). Similarly, Coen, Swanwick, O'Boyle, and Coakley (2007) found dysfunctional behaviors to be related to increased caregiver burden.

Another major factor which contributes to increased levels of caregiver burden is the ability of the patient to care for himself or herself. In a study assessing former rehabilitation patients, it was found that social cognition deficits, self-care deficits, and caregiver's age were significant predictors of level of caregiver burden. Results indicated that the more severe the deficits in patient cognition and self-care abilities, the higher the burden. Specific deficits were in the areas of social cognition, communication, locomotion, and transfer of the patient (Watson, Modeste, Catolico, & Crouch, 1998). A study of status post-stroke sufferers revealed that physical impairment and communication difficulties of the stroke survivor were associated with a lower caregiver

quality of life (C. L. White, Mayo, Hanley, & Wood-Dauphinee, 2003). Quality of life (QoL) decreased in those caring for a stroke survivor with communication difficulties.

Age of the caregiver and relation to the patient also play roles in the level of burden experienced. Younger caregivers reportedly experience higher levels of burden (Watson, Modeste, Catolico, & Crouch, 1998). Wives tend to be the caregiver most frequently; next are daughters and daughter-in-laws. Children bear significantly greater burden as they may view the caring role as an unanticipated obligation, whereas spouses may expect caregiving as part of their marital role (Llacer, Zunzunegui, Gutierez-Cuadra, Beland, & Zarit, 2002). A study of impaired Japanese elderly investigated success in adaptation over time after undertaking the caregiving role. They found that through time, spouses' level of burden decreased, representing an adaptation to their role as caregiver. In contrast, daughters-in-law caregivers' level of burden *increased* over time, suggesting that the role became increasingly intolerable (Arai, Zarit, Sugiura, & Washio, 2002).

Another factor related to increased burden is maintaining employment while providing care. For those who work, greater job conflict was associated with higher role overload, worry, and strain in the caregiver. In other words, while employment of caregivers does not directly contribute to increased burden, it may decrease the threshold for role strain and depressive symptoms (Edwards, Zarit, Stephens, & Townsend, 2002).

Perceived social support has been shown to have a beneficial impact on caregivers and vice versa. Coen et al. looked at behavioral disturbance in AD patients and found that increased burden was related to decreased levels of informal social support (Coen, Swanwick, O'Boyle, & Coakley, 1997). Interestingly, there is no connection between the

number of hours spent with the patient and the level of caregiver burden (Arai, Masui, Sugiura, & Washio, 2002).

Burden and HD

Although the caregiver burden literature in other disease processes provides a framework for anticipating which factors may impact caregiving in HD, one must consider that the complexity of symptoms, the onset of a neurodegenerative disease process in middle age, and the genetic nature of HD makes this caregiver role distinct from other types of caregiving. These and other themes will be reviewed in the following paragraphs.

The breadth of HD symptoms (physical, psychiatric, and neurologic), their chronic nature, and the fact that their presentation can metamorphose throughout the course of the disease, make caregiving for a family member with HD a considerable challenge. Of the triad of symptoms that could potentially present, impaired mood and behavior have been reported to be the most distressing to caregivers and can drastically alter family, and especially spousal, relationships (Hayden, Ehrlich, Parker, & Ferera, 1980; LoGiudice & Hassett, 2005; Semple, 1995). In a study by Tyler, Harper, Davies, and Newcome (1983), the relationship between HD disease state, family breakdown, and stress was examined in a sample of 92 patients. They found that violence, promiscuity, and bizarre and slovenly behavior were reported as the primary causes of marital breakdown. Behavioral outbursts were one of the main causes of stress within the family, with dangerous and aggressive behavior reported in nearly half of all patients (Tyler, Harper, Davies, & Newcome, 1983).

Another factor that uniquely affects caregiving in HD is that the onset of progressive cognitive impairment in middle age is dramatically out of sequence with the developmental trajectory. In middle age, energies are generally focused on work, marriage, childrearing, and tending to aging parents. Instead, the affected individual's insight becomes impaired, their sense of responsibility for family commitments declines, and they withdraw from the provider role. As the patient is faced with developmentally discordant changes, the caregiver is faced with emotionally and socially incongruent changes. The loss of support from the affected spouse, paired with their increasing dependency needs, places significant challenges on the partner and children (LoGiudice & Hassett, 2005). The caregiver is placed in a position of total responsibility, having to maintain their own role as well as assume the role of their spouse (Kessler, 1993). Additionally, the caregiver is confronted with mourning the loss of an intimate and loving relationship, all the while living with the spouse as a reminder of that loss (Korer & Fitzsimmons, 1985).

The presence of HD in a family usually suggests that the caregiver's responsibilities will not be limited to caring for just one family member. Instead, its genetic nature, more often than not, ensures the caregiver undertake this role for several loved ones. It is not uncommon for a person to nurse his or her parent, then an older sibling, and finally succumb to HD themselves (Kessler, 1993). A non-HD spouse may be faced with caring for more than one generation of sufferers, or to have to choose between caring for a spouse versus children (Tyler, Harper, Davies, & Newcome, 1983). Beyond the practical implications of genetics, there is a strong emotional repercussion. The worry of transmission to children can be overwhelming, and has been reported to

stimulate feelings of resentment and hostility toward the affected spouse (Hans & Koeppen, 1980).

A recurring theme in the HD caregiver literature is the family's complaints of deficient service provision for the patient (Hans & Gilmore, 1968; Skirton & Glendinning, 1997). There are two main reasons for this. First, the triad of symptoms that HD patients present with traverse health and social care domains, resulting in boundary line confusion as to who should care for these individuals (Aubeeluck, 2005; Lowit & van Teijlingen, 2005). This results in logistical problems coordinating the care between health professionals or, in the worst cases, simple neglect. The second problem that generally arises is the lack of experience and training that professionals have with HD. Many health workers don't have a real understanding of the disease or the needs of the patient and carers (Lowit & van Teijlingen, 2005). Families of the affected do not feel they receive consistent help from family doctors, primary health care teams, or social services (Korer & Fitzsimmons, 1985). While this problem occurs throughout all stages of the disease, it seems to be most poignant at the time of skilled nursing placement. Generally, facilities are equipped to handle aged care placement, which is inappropriate in view of the relative youth of those affected (LoGiudice & Hassett, 2005). Nursing home staff are often not aware of some of the fundamental requirements of an HD resident (e.g., that caloric requirement is in the realm of 5000 calories per day). Also, facilities tend to be inadequately staffed for the dependency levels of the HD patient.

Some research has been dedicated to palliative care in HD, which aims mainly to improve the quality of life of a patient. Although meeting patients' needs does not directly benefit caregivers, it does so *indirectly* by decreasing burden and stress that

would otherwise be placed on the caregiver. Palliative care studies have found certain issues as ranking high in importance to caregivers; these include financial assistance, equipment and home modification, respite and home support, transport, access to allied health services, regular contact with associations in rural areas, information about condition and treatment, education of service providers, improved public understanding and awareness, and dedicated programs for these conditions (Aoun, Kristjanson, & Oldham, 2006; LoGiudice & Hassett, 2005; Skirton & Glendinning, 1997). It was emphasized that in order to be useful, however, care must be delivered in a timely fashion, care delivery among different providers must be coordinated, and type of care delivered must be flexible based on the constantly changing needs of the HD patient (LoGiudice & Hassett, 2005; Lowit & van Teijlingen, 2005).

HD family caregivers experience a wide range of emotions as a result of their caregiving role, which consequently impacts well-being (Semple, 1995). Living with HD has been likened to "learning to live with stress, anxiety, fear, and loss" (Lowit & van Teijlingen, 2005). Many caregivers report being severely distressed and overburdened (Kessler, 1993; LoGiudice & Hassett, 2005). One study showed that 82% of caregivers reported feeling stress (Tyler, Harper, Davies, & Newcome, 1983). In another study, 45% of caregivers had clinical anxiety and depression (McGarva, 2001). Hans and Koeppen (1980) described the caregiver's situation as being one of continuous trauma (Hans & Koeppen, 1980). Wives described having ended up married to "a different person and perhaps not the sort of person they would have chosen". Feelings of regret, anger, and ambivalence are commonplace, and often marriages come under pressure (Hans & Koeppen, 1980).

Other negative consequences found to be associated with the caregiving role are health and financial problems (Hans & Gilmore, 1968). One study reported that 81% of caregivers in the sample endorsed poorer health, and 70% reported a poorer lifestyle than prior to taking on the caregiving role (McGarva, 2001). In another study, almost 20% of the caregivers reported suffering from stress-related illness (Skirton & Glendinning, 1997). Families frequently experience a drop in the standard of living after the patient develops the disease; this can be either a result of the affected individual or the caregiver relinquishing work. Bankruptcy is not uncommon (Korer & Fitzsimmons, 1985).

The coping style of HD caregivers has also been investigated, as it has been found to have a significant impact on quality of life. HD caregivers' coping styles tend to fall into one of two categories: those who accept the disease, and those who deny it. Caregivers who acknowledge the disease in their lives maintain their individual identities. They promote the same for family members and encourage establishment of relationships outside of the family. As a result, HD is manageable and quality of life is better. In contrast, caregivers who utilize denial as a coping mechanism tend to hide the existence of HD within the family by withdrawing from activities where a spouse's decline would be noticed and by avoiding affected family members. These caregivers find themselves unable to hold a discussion about the disease, preventing them from maintaining crucial ties with support networks (Lowit, 2005) and from providing accurate and complete information to children so that they can make appropriate plans for their future. It is as if these efforts will enable the family to avert the impending doom (Kessler, 1993). Whatever the reason, caregivers in denial report a lower quality of life (Helder et al., 2002). In fact, although avoidance and denial may provide short-term psychological

relief, the long-term consequences are serious. Planning can assist in provision of timely interventions (to support caregivers) that are vital to maintaining stability of the caregiving relationship and to avoid premature institutionalization (Dawson, Kristjanson, Toye, & Flett, 2004).

Development of the HDQoL-C Questionnaire

In an effort to delineate the factors that enhance and compromise quality of life in spousal caregivers of HD patients, a disease-specific assessment instrument was devised. In an initial study in 2001, HD caregivers and health professionals were recruited to evaluate a pre-existing generic quality of life measure (Comprehensive Quality of Life Questionnaire for Adults, CoMQoL-A5; Cummins, 1997) for relevance to the HD population and to generate a list of issues not addressed in the questionnaire that were pertinent to caregiving in HD. From the participants' feedback, it was evident that the generic measure was not entirely relevant. Analysis of the written statements revealed 21 sub-themes which clustered into four more general themes: level of support, dissatisfaction with caregiving role, practical aspects of caregiving, and feelings and emotional well-being. The authors utilized the newly identified themes as the basis for the development of a novel disease-specific quality of life scale for spousal caregivers (Aubeeluck & Buchanan, 2006b).

In a follow-up study, Aubeeluck and Buchanan (2006) aimed to further understand quality of life issues specific to spousal caregiving in HD. The authors utilized the "photovoice" method (C. C. Wang, 1999), a process in which photography is used as a vehicle to identify and represent issues important to a population of interest from their vantage point. Disposable cameras were provided to five HD caregivers. These

caregivers were asked to photograph subjects that either represented a compromise or enhancement of their quality of life, to label each photo as positive or negative, and then to briefly describe the reason they selected the subject matter. An analysis of the resulting 109 photos revealed 9 themes related to QoL in HD: care and security (e.g., making the house safe), small pleasures, loneliness, escape (i.e., no end to the caregiving role), sense of loss, neglected needs, support, lack of time, and daily hassles (e.g., shopping). Whereas a number of these themes were addressed on the CoMQoL-A5, some themes were specific to HD and did not appear on that measure, thereby reinforcing the need for creation of a disease-specific scale (Aubeeluck & Buchanan, 2006a).

In a third study (Aubeeluck & Buchanan 2006b), the knowledge gained from the "photovoice" study was utilized to help guide a more in-depth exploration of issues important to HD caregivers. They organized 47 spousal caregivers into six focus groups and asked each group to describe thoroughly the ways in which caring for a relative with HD affected their overall quality of life. Focus group sessions were audiotaped, transcribed, and analyzed for content; four main themes developed: levels of support, dissatisfaction with caregiving role, practical aspects of caregiving, and feelings of emotional wellbeing. Although some of the concerns mentioned in the focus groups echo those in the general caregiving literature, many topics indicated that the quality of life in HD caregivers is compromised in ways distinct from other dementias (e.g., genetic implications). Findings from this and the previous studies by these authors were utilized in the creation of the Huntington's Disease Quality of Life for Carers (HDQoL-C) questionnaire (Aubeeluck & Buchanan, 2006).

In the fourth study, the authors sought to validate the HDQoL-C. The newly devised measure addressed aspects of physical health, psychological state, level of independence, social relationships, personal beliefs, and relationships. Eighty-seven HD spousal caregivers were administered the questionnaire, and a factor analysis was performed on the resulting data. Although the authors did not provide specific factors which emerged from the analysis, they reported that the analysis established the HDQoL-C as a multidimensional and psychometrically sound, disease-specific, subjective QoL assessment tool. It was found to have good internal consistency, test-retest reliability, and congruent validity for use with spousal caregivers. The questionnaire may be used to establish a baseline quality of life measure for which follow-up assessments may reflect effectiveness of interventions. The questionnaire may also prove useful in the identification of important topics to target in individual therapeutic interventions. Finally, simply engaging in the activity of filling out the form was reported to have had therapeutic and cathartic benefits. The authors proposed that the measure may also prove useful in assessing other family caregivers, although they cautioned that this is yet to be determined (Aubeeluck & Buchanan, 2006b).

In summary, while there are some factors that seem to affect caregivers of many different disease processes, there seem to be a number of burdens unique to the HD population. And, although the qualitative exploration of these themes in early research has been extremely valuable in increasing understanding about this area, utilization of formal questionnaires in the current study allows for quantification and statistical analysis of burden levels, as well as for direct evaluation of how burden relates to other potentially relevant issues. Additionally, it is exciting to note that, to our knowledge, the current

study is the very first to implement the HD-specific caregiver burden measure designed and validated by Aubeeluck and Buchanan (2006). Increased knowledge about the psychological and emotional needs of HD caregivers enables mental health professionals to connect caregivers to the appropriate resources, assist them in coping with the demands of their roles, and, in turn, aid clinicians in improving their skills and tailoring treatment to address issues important to HD caregivers.

Research Aims and Hypotheses

Specific Aim 1: To investigate the emotional perceptual functioning of HD patients as compared to healthy controls. It was hypothesized that HD patients would demonstrate emotional perceptual deficits as compared to healthy controls.

Specific Aim 2: To investigate HD subjects' accuracy in identification of individual emotions. It was hypothesized that, as observed in the literature, HD subjects would be less accurate at identifying the emotions of disgust and anger.

Specific Aim 3: To explore the relationship between two different existing caregiver burden questionnaires. It was hypothesized that these two measures would be positively correlated.

Specific Aim 4: To investigate the relationship between emotional perceptual functioning and caregiver burden. It was hypothesized that emotional perception impairments would be positively correlated with elevated levels of caregiver burden on both measures.

Specific Aim 5: To investigate the relationship between psychiatric and personality functioning and caregiver burden. It was hypothesized that psychiatric

symptomatology and dysfunctional personality traits would be positively correlated with elevated levels of caregiver burden.

Specific Aim 6: To investigate the relationship between HD subject characteristics, HD subject cognitive profile, HD subject awareness functioning, caregiver characteristics and caregiver burden. It was hypothesized that the factors known to correlate positively in other disease processes with higher levels of burden (e.g., behavioral disturbances in patients, impaired cognition in patients, impaired patient communication, patients' impaired ability to carry out activities of daily living, younger caregiver age, and lower level of perceived social support by the caregiver) will also correlate positively with elevated levels of burden in HD caregivers.

METHODS

Subjects

Eighteen individuals diagnosed with HD participated in this study. Mean disease duration was 8.9 years. HD patients were native English-speakers or had learned English by the age of eight. They ranged in age from 39 to 74; nine were men, and nine were women. Thirteen were right-handers, four were left-handers, and one was ambidextrous. Ten had a significant psychiatric history, three had a history of substance abuse, and three had a history of learning disability. None had a history of mental retardation; neurological insult (i.e., acquired brain injury), disease (other than HD), or intervention; or major medical illness. Seven HD subjects were medicated with neuroleptics, 13 with antidepressants, 3 with mood stabilizers, and 6 with memory-enhancing medications (i.e., Namenda or Aricept) at the time of testing. Of the HD subjects, 7 were in Stage 1, 10 were in Stage 2, and 1 was in Stage 3 of the TFC scale (Shoulson & Fahn, 1979). Subjects in the earlier stages were preferred primarily to meet exclusionary criteria for dementia and secondarily to minimize the presence of motor impediments. However, results of a dementia screening measure (see below for details) revealed that 10 of the participants met criteria for dementia. It was felt that this cognitive impairment invalidated HD patients' performance on the experimental and control tasks. A meta-analysis on episodic memory impairments in HD (Montoya et al., 2006) demonstrated that the HD literature consistently divides symptomatic HD patients into those with mild dementia and those with moderate/severe dementia. The cutoff score on the DRS that differentiates these two groups is 128. Other HD and AD literature (Monsch et al., 1995; Paulsen et al., 1995) supports this cutoff. Therefore, a DRS score of 128 was utilized in

the current study to identify those subjects considered to have moderate to severe dementia. Of the 8 who did not meet criteria for dementia, 5 were in Stage 1 of the TFC scale and 3 were in Stage 2.

Seventeen caregivers participated. Caregivers were native English-speakers or had learned English by the age of eight. Although HD subjects and their caregivers were required to participate as a pair, one caregiver failed to attend the scheduled appointment and did not sign consent forms or participate. Therefore, one less caregiver than HD subjects participated in the study. Caregivers ranged in age from 38 to 73, included 8 men and 9 women, and 15 were spouse caregivers and 2 were parent caregivers.

HC comparison data were available in summary form (i.e., *M* and *SD*). These data were derived from a database of 103 HCs, ranging in age from 20 to 81, the individuals of which were administered the complete New York Emotion Battery (NYEB; Borod, Welkowitz, & Obler, 1992) at Mount Sinai School of Medicine as part of Joan Borod's NIH PSC-CUNY funded research. Summary data were available in one-way, two-way, three-way, and four-way tables regarding age, gender, ethnicity, and education. HD subjects' performance was compared to HC summary data.

Rationale for Number of Subjects

Effect Size

Effect Size Explained

The effect size may be defined as "the degree to which the phenomenon is present in the population" (Cohen, 1988). It can be expressed as the difference between two population parameters or the departure of a population parameter from a constant. Effect size, as it applies to a sample, is the standardized difference between group means (i.e.,

the difference between the experimental group's mean and the control group's mean, divided by some measure of dispersion). Glass' Δ, Hedges' *g*, and Cohen's *d* are examples of several such methods of calculation. Effect size estimates can vary in degree of accuracy based on which method of standardization (e.g., control group SD or pooled SD) is utilized. For example, when sample sizes are small or disparate, the effect size can potentially be overstated unless one of the more conservative indices is employed. Therefore, there is not one single approach to calculating effect sizes which is ideal in all cases; selection of the method is made on a case-by-case basis.

Interpretation of Effect Size

Cohen (1988) provided rules of thumb for characterizing what effect sizes are small (*d*=.2), medium (*d*=.5), or large (*d*=.8), based on his observations of typicality within the social sciences literature (Cohen, 1988). Detection of treatment effects is inversely related to the number of test subjects; the smaller the treatment effect is, the greater the number of subjects is required to detect it, and vice versa. Although some empirical studies suggest that Cohen's characterization is fairly accurate (Glass, 1979; Olejnik, 1984), these guidelines should only be used when no better basis for estimating the effect size index is available. The preferred manner of determining effect size is to calculate it directly from published studies or from pilot data. In the case of the existing study, we have access to both published studies as well as relevant pilot data.

As mentioned in the Introduction, there are 10 experimental studies which have investigated emotional functioning in HD. Of these, 4 were not able to be used in the power analysis for the reasons stated in Table 2.

Table 2. HD Emotion Studies Not Included in Power Analysis

Study	Reason for Omission
Gray et al., 1997	Controls were at-risk individuals found to be gene negative only after completion of the study; HD- individuals share too many environmental factors with HD+ individuals to be considered a clean comparison group.
Jacobs et al., 1995	No data provided
Sprenglemeyer et al., 1997	Case study
Sprenglemeyer et al., 2006	No SDs provided for the healthy controls

The remaining six studies contained all the necessary information required. As data were provided in distinct formats for each study, the exact calculation procedure varied. For example, when studies provided overall task performance summary data, these statistics were able to be utilized directly in the power analysis. Other studies provided summary data for performance by emotion within the experimental task; for these studies, calculation of overall task performance had to be obtained prior to this information being utilized in the power analysis. Regardless of the method utilized, overall effect and sample sizes from each publication were derived. Each set of values were equally weighted by their respective sample sizes and combined to yield the Grand Effect Size needed for power analysis of the current study. The following paragraphs detail how effect sizes were attained for each reference. See Table 3 for effect and sample size values for each of the six studies.

In the Hayes et al. (2007) study, three tasks were administered to the HD and HC group, and an effect sizes analysis was carried out for each task. Sample sizes were equivalent in the patient and control group, so this number was directly utilized in relevant calculations. An overall effect size averaged across tasks was computed.

In the Milders et al. (2003) study, only one task was administered to the HD and HC groups. However, the results of the task are presented by emotion rather than as an aggregate. As sample sizes were equivalent in the patient and control group, this number was directly utilized in relevant calculations. Effect size analyses were carried out for each individual emotion and then averaged across emotions for an overall effect size.

Montagne et al. (2006) administered one task to the HD and HC groups. The results of this task were presented by emotion rather than as a total. Sample sizes of the healthy control and patient group were unequal, so a harmonic mean was calculated and utilized in relevant calculations. Effect sizes were calculated for individual emotions and then combined into an overall effect size.

The Speedie et al. (1990) study included two sub-studies; each was carried out using the same HD group individuals, but distinctly different healthy control groups. Sample sizes of the healthy control and patient groups were unequal in both studies, so harmonic means were calculated for each study and utilized in the respective effect size calculation. These two means were further combined into one value (representing the sample size of both studies), which was used in the subsequent statistical analysis. Effect sizes were calculated independently for each study and combined into an overall effect size.

Sprenglemeyer et al. (1996) included three tasks in their study. The sample sizes of the healthy control and patient groups were unequal, but constant across tasks. Therefore, a harmonic mean for the unequal sample sizes was calculated and utilized in the weighted effect size calculations for each task. Effect sizes were calculated individually for each task and then a weighted effect size across tasks was calculated.

In the Wang et al. (2003) study, only one task was administered. Sample sizes of the healthy control and patient groups were unequal, so a harmonic mean was calculated and utilized in relevant calculations. The effect size for the one task was calculated. As there was only one task, this was the only calculation necessary.

As mentioned previously, pilot data exist that are relevant for use in this power analysis. In research previously conducted as a doctoral research project, Parkinson's disease patients were administered facial, prosodic, and lexical emotional perception tasks (Krch, 2003). Although the patient populations are distinct, the same core neuroanatomical structures are affected in both disease processes. Though it is not suspected that involvement of the same structures alone would produce similar deficits in both populations (indeed, the specific pathways involved in each disease process is distinct), the pilot research's contribution to the power analysis is more owing to the use of identical testing instruments. In Krch's study, three tasks were administered to PD and HC groups. Sample sizes were equivalent in the patient and control group, so this number was directly utilized in relevant calculations. Effect sizes were calculated individually for each task and then a weighted effect size across tasks was calculated.

Table 3. Task, Study, and Grand Effect and Sample Size Calculations

	Task		Study-wide		Grand
Study	**Effect Size**	**Sample Size**	**Effect Size**	**Sample Size**	**Effect Size**
Hayes et. al., 2007					
Vocal	1.59	14			
Lexical	0.40	14	1.19	14	
Images	1.57	14			
Krch, 2003					
Prosodic	0.16	9			
Facial	0.44	9	0.26	9	
Lexical	0.18	9			
Milders et al., 2003					
Happy	0.62	20			
Surprise	0.59	20			
Fear	2.48	20	1.48	20	
Sadness	1.34	20			
Disgust	1.67	20			
Anger	2.15	20			
Montagne et al., 2006					} 1.47
Disgust	1.31	12.63*			
Anger	0.87	12.63*			
Fear	0.67	12.63*	0.81	12.63	
Happy	0.67	12.63*			
Surprise	0.67	12.63*			
Sadness	0.67	12.63*			
Speedie et al., 1990					
Prosodic 1	1.33	8.57*	1.45	7.37	
Prosodic 2	1.62	6.46*			
Sprenglemeyer et al., 1997					
Facial hexagon	1.93	14.73*			
Facial identification	2.89	13.36*	2.16	13.79	
Prosodic	1.70	18.12*			
Wang et al., 2003					
Facial	3.20	8.73*	3.20	8.73	

*Some sample size values within this table contain decimal points, indicating that healthy control and patient group sample sizes were unequal, requiring a harmonic mean to be calculated.

Alpha Level

Alpha level is the probability of rejecting the null hypothesis when it is true (i.e., Type I error). It would seem that a reasonable approach to selecting the alpha level would be to minimize the chance of committing this error by selecting the smallest value

possible, such as .01 or .001. However, the smaller the alpha level, the larger the possibility of not rejecting the null is when the null is false (i.e., Type II error). Within the social sciences, .05 has become the conventional alpha level value, due to the fact that it provides an acceptable balance of risks of Type I and Type II errors. As such, this value will be utilized in the power analysis and for all other hypothesis testing within this study.

Power

Statistical power is the ability to detect a difference between treatment groups in the event that a true difference exists. When the statistical power of an experiment is sufficient, it is possible to detect an existing statistically significant result. If power is not sufficient, a study may not produce a statistically significant result even when that significant difference exists. (To illustrate, power in statistics may be compared to the power of a microscope. If the power of magnification is not adequate, an observer will not be able to visualize the specimen, even if it is on the slide). Statistical power is influenced by the alpha level, and as such, selection of the power should be considered in conjunction with the alpha level. Cohen recommends that experiments be designed to achieve a power of about .80 (i.e., 80 percent; Cohen, 1988). When alpha is .05, a power of .80 is associated with a 20 percent probability of a Type II error and 5 percent probability of a Type I error. This 4:1 ratio reflects a balance that appreciates the greater severity of Type I errors versus Type II errors.

Power Analysis

After determining the weighted average effect size (displayed in Table 3), alpha level, and power, the appropriate power analysis tables were selected. Tables vary based on the statistical procedure (e.g. t-test, correlation, or ANOVA) to be performed on the

data in question. In the current study, the primary type of analysis was a one-way ANOVA; accordingly, the one-way ANOVA power analysis tables were utilized (Cohen, 1988). Although the grand average effect size that resulted from these calculations was 1.47, an effect size of .80 was utilized, as this is the largest conventional effect size used in power analyses with ANOVAs (Cohen, 1988). As mentioned above, the conventionally proposed power to achieve in a study is .80. Based on an alpha level of .05, a .80 effect size, and a power of .79 (the closest value on the table to the sought after power of .80), 7 subjects per cell would be required (i.e., 7 HD patients and 7 HC subjects; Cohen, 1988). The following table (Table 4), which reflects the rate of increase in power associated with increasing sample sizes, was extracted from Cohen's (1988) power tables.

Table 4. Corresponding Change in Power with Increasing Sample Size

.8 Effect Size	
Power	**Subjects**
.79	7
.85	8
.89	9
.93	10
.97	12
.99	15
>.995	17

Substantial effort has been made to utilize conservative approaches at each stage of the power analysis process in an attempt to find the smallest effect worth detecting. In studies where several tasks were administered, rather than utilize effects from each task, the overall study-wide effect was utilized; this was done in an effort to prevent multiple-task studies from being over-represented in the overall power analysis. The effect size procedures selected were conservative as they are specialized for use with small and

disparate sample sizes. As mentioned previously, although the grand average effect size that resulted from these calculations was 1.47, a more conservative .80 effect size was utilized. Despite these methods, the resulting weighted effect size reflects a robust presence of emotional deficits in HD; it is important to note that some of these studies were performed with as few as 6 subjects, with the largest number of HD subjects being 20. The following table (Table 5) presents sample sizes for each study utilized in the power analysis.

Table 5. Sample Sizes of HD Emotion Studies Utilized in Power Analysis

Study	HD subjects	NC subjects
Hayes et al.	14	14
Krch*	9	9
Milder et al.	20	20
Montagne et al.	8	30
Speedie et al. study 1	6	15
Speedie et al. study 2	6	7
Sprenglemeyer et al. (1997) study 1	13	17
Sprenglemeyer et al. (1997) study 2	11	17
Sprenglemeyer et al. (1997) study 3	11	17
Wang et al.	6	16

*PD patients were utilized in this study

Notwithstanding, it is likely that the cautious researcher or statistician would feel uncomfortable with such a small suggested sample size. It is important to keep the following in mind: 1) Previous studies only provide estimates of effects. When the true effect is smaller than the estimate, the sacrifice is insufficient power to detect the effect. 2) Effect sizes greater than 1.0 are considered rare and are often treated as outliers unless they make up a large portion of the distribution. Because power estimates were derived from other studies and not from pilot data, except in one possible case, it was prudent to consider a larger sample size.

A sample size of 30 would put the law of large numbers into effect and significantly strengthen the probability of detecting an effect. However, the goal of a power analysis is to find an appropriate balance by taking into account the substantive goals of the study and the resources available to the researcher. As is often the case in research, a judgment call was made by the investigator; a sample size of 15 per cell, double the amount required by the power analysis, was proposed as a reasonable compromise. The total participants in the study, then, exceeded this expectation by 3 subjects per cell.

Materials

HD Patients

Prescreening

The questionnaire used for the phone screen is based on a questionnaire from the NYEB, modified for application with HD subjects and their caregivers (See Appendix B). The portion of the questionnaire applicable to HD patients assesses age, language dominance, stage of illness, neurological history secondary to HD, neurological interventions, psychiatric illness, substance abuse, and sensory system functioning.

Demographics

Interview Form. An interview was conducted in which education, occupational history, medical and psychopharmacological history, sensory system functioning, and handedness was assessed (See Appendix C). Self-report of handedness was confirmed by the 4-item behavior handedness subtest from the Coren, Porac, and Duncan lateral dominance inventory (Coren, Porac, & Duncan, 1979).

Screening

All HD subjects were administered screening tasks to assess possession of basic cognitive skills required to complete the experimental tasks. The cutoff scores to be utilized for the screening measures in this study are based on previous research with neurological populations from Dr. Borod's laboratory and are generally 2 or more SDs below the normative mean. Screening tasks were administered in a fixed order for all subjects.

Cognitive Decline. The *Dementia Rating Scale – 2* (DRS-2; Jurica, Leitten, & Mattis, 2001) is a dementia screening test that generates five subscale scores in the areas of Attention, Initiation-Perseveration, Construction, Conceptualization, and Memory. This was administered to assess the presence of dementia.

Premorbid Intellectual Functioning. The *North American Adult Reading Test* (NAART, Blair & Spreen, 1989) is a reading test of 61 irregularly spelled words of increasing difficulty (e.g., from "debt" to "syndecdoche"). From the total score, an estimate of premorbid verbal intellectual functioning can be derived. This measure was administered to obtain an estimate of level of premorbid intellectual functioning.

Language. Select subtests (Commands, Complex Ideational Material, and Reading Sentences and Paragraphs) from the *Boston Diagnostic Aphasia Examination* (BDAE, Goodglass & Kaplan, 1983) were administered to assess ability to manage the basic linguistic requirements (e.g., reading comprehension) of the experimental perception tasks. The Commands subtest requires subjects to follow commands with increasing complexity (e.g., "Point to the ceiling and then to the floor"). The Complex Ideational Material subtest requires that the subject make judgments about statements of

logic (e.g., “Will a cork sink in water?”). The Reading Sentences and Paragraphs subtest requires the subject to read text and answer basic comprehension questions.

Psychiatric Functioning. HD subjects were administered the *Personality Assessment Inventory* (PAI; Morey, 1991), a comprehensive, objective, self-administered questionnaire, to assess for significant psychiatric history, or significant alcohol or substance abuse histories. This instrument contains 344 items comprising 22 nonoverlapping full scales, including 4 validity scales, 11 clinical scales (clustered in Neurotic, Psychotic, Personality Disorders, and Behavioral Disorders), 5 treatment scales, and 2 interpersonal scales. T-scores are reviewed to determine if clinically significant elevations exist (different cutoffs exist for each scale), thereby providing a means for screening psychopathology, which may have an impact on emotion task performance in neurological populations (Jaeger, Borod, & Peselow, 1987; Liotti & Tucker, 1992; Robinson & Manes, 2000).

Medical History. The patients signed a Release of Information Consent form which gave their neurologist authorization to release information regarding diagnosis and stage of HD to the investigators of this study: diagnosis, neurological examinations, genetic test results (i.e., CAG repeat size), and family history of HD.

Experimental Emotion Tasks

In order to assess the perceptual aspects of the three emotional communication channels described above (i.e., facial, prosodic, and lexical), perceptual experimental emotional tasks were administered to subjects. The NYEB contains various experimental emotion tasks for the facial, prosodic, and lexical channels. Only one task from each of these channels was selected for inclusion in the current study, with selection based on

psychometric properties of the task, as well as logistical and administration time constraints in the present study. The Facial Identification Task (FID) was selected for the facial channel, the Prosodic Identification Task (PID) was selected for the prosodic channel, and the Emotional Sentence Identification Task (SID) was selected for the lexical channel. Each experimental task includes 8 emotions: 3 positive (Happy, Pleasant Surprise, and Interest) and 5 negative (Anger, Sadness, Unpleasant Surprise, Disgust, and Fear). Experimental tasks were administered with task order randomized across subjects.

Facial Channel. The *Facial Emotional Identification Task* requires subjects to correctly identify the emotion portrayed in a single photograph of a human face (Borod et al., 1992; 1998). The eight emotions chosen (described above) are presented. The subject is permitted to view each slide as long as desired in order to select the correct response from a list of the eight emotions, which are printed in a vertical array on an 8.5 X 11-inch card. Thirty-two trials are given, with each of the eight emotional expressions shown four times (in randomized order). Each correct item is given one point, resulting in 32 possible points on the FID.

Prosodic Channel. The *Prosodic Emotional Identification Task* involves the subject listening to emotionally intoned tape-recorded neutral-content sentences (e.g., "She went to walk the dog") and selecting which one of the eight basic emotions they believe is represented in the prosody (Borod, et al., 1992, 1998). The intoned sentences are presented twice. There are 24 trials on this task, with each of the eight emotional intonations presented three times (in randomized order). One point is given for each correct item, resulting in 24 possible points on the PID.

Lexical Channel. The *Lexical Emotional Sentence Identification Task* requires subjects to read emotionally laden sentences (e.g., "He slammed the door") and select the correct emotion represented (i.e., anger in this case). There are 24 trials on this task, with each of the eight emotions occurring three times (in randomized order). A total of 24 sentences are presented at one point per correct response, yielding a maximum score of 24.

Nonemotional Control Tasks

Control tasks were chosen based on presumed nonemotional cognitive skills underlying emotional perception. The NYEB contains various nonemotional control tasks for the facial, prosodic, and lexical channels. Only one task from each of these channels was selected for inclusion in the current study, with selection based on psychometric properties of the task, as well as logistical and administration time constraints. The Benton Facial Recognition Task (BFRT; Benton, deS Hamsher, Varney, & Spreen, 1983) was chosen for the facial channel, the Intonation Contours Perception Task (ICP; Borod et al., 1992) was chosen for the prosodic channel, and the Lexical Nonemotional Sentence Identification Task (NESID; Borod et al., 1992) was chosen for the lexical channel. Control tasks were administered for each in a fixed order for all subjects.

Facial Channel. The short form of the *Benton Facial Recognition Test* (Benton et al., 1983) was administered to assess basic visual perceptual functioning and basic face recognition. This task requires subjects to match a black-and-white photograph of a face to six possible choices. The first section of the test consists of six trials, in which a single face is matched to an array of six choices. In contrast, the second section consists of seven trials, in which a single face is matched to three out of six possible choices. Each

trial is presented for as long as a subject needs to respond. For approximately half the stimulus items, correct matches are the same face in different angles of orientation, and for the other half, correct matches are the same face with different degrees of lighting. One point is given for each correct match, resulting in 27 possible points. The total score is converted to the long-form scale (Benton et al., 1983), based on 54 possible points.

Prosodic Channel. For the *Intonations Contours Perception Task* (Borod et al., 1992), subjects listen to an audiotape which contains 3-syllable nonsense strings (e.g., ba-ta-ga; Blumstein & Cooper, 1974) and identify which of the three types of intonational stress (i.e., declarative, interrogative, or exclamatory) is represented. There are eight items for each type of intonation, yielding 24 items. Participants must identify each item from a multiple-choice response card that contains a verbal label, a punctuation mark representing the label, and a drawing which exemplifies the label (e.g., for the "exclamation", a military general is giving a private soldier a direct order). One point is given for each correct response, resulting in a maximum score of 24.

Lexical Channel. The *Lexical Nonemotional Sentence Identification Task* (Borod, et al., 1992) requires subjects to read a sentence and then determine which of the eight nonemotional category types ("characteristics of people"; body type, complexion, hair type, intelligence, personality, teeth, vision, and voice type) best represents the sentence. For example, the sentence, "He watched the concert until the end." is best represented by the category "Vision". A total of 24 sentences are presented at one point per correct response, yielding a maximum score of 24.

Awareness Functioning

The *Subjective Report Questionnaire* is a brief self-report questionnaire administered to assess HD patients' awareness of involuntary movements (Snowden, Craufurd, Griffiths, & Neary, 1998). This measure produces two values, a measure of subjective experience and a measure of subjective consequence. This questionnaire was administered to gain insight into the degree of appreciation the individual has for their deficits, and how that might be associated with caregiver burden.

Caregivers

Prescreening

The questionnaire used for the phone screen is based on a questionnaire from the NYEB, modified for application with HD subjects and their caregivers (See Appendix B). The portion of the questionnaire applicable to caregivers ascertains that participants meet age (i.e., 18 to 80), language (i.e., native language is English or acquired by age 8) and HD status (i.e., not at risk) inclusionary criteria. When these criteria were not met, caregivers and the persons with HD for whom they cared were not invited to further participate in the study.

Questionnaires

The Huntington's Disease Quality of Life Battery for Carers (HDQoL-C; Aubeeluck & Buchanan, 2004) is a self-administered questionnaire designed especially for caregivers of HD patients. It consists of 34 questions divided into four sections: 1) demographic and objective information, 2) aspects of caring, 3) satisfaction with life, and 4) feelings about life. The demographic and objective information section consists of six questions, some of which can be responded to in multiple-choice format and others in

open-ended format. Sections 2, 3, and 4 comprise 34 questions on an 11-point Likert scale (i.e., 0-10), wherein subjects are asked to circle the number they feel most accurately represents their situation. A quality of life score can be derived for each of the subsections as well as for the total measure. Higher scores represent higher quality of life.

The Zarit Burden Inventory (ZBI; Zarit, Reever, & Bach-Peterson, 1980) is a widely used self-administered questionnaire which measures a caregiver's subjective burden. It consists of 22 items on a 5-point Likert scale (0, never; 1, rarely; 2, sometimes; 3, quite frequently; 4, nearly always) that explores the physical, psychological, and social consequences of caring activities. Item ratings are summed to achieve a total score from 0 to 88, with higher scores representing greater burden.

Procedures

IRB Approval

Institutional Review Board (IRB) approval was obtained from the University of Medicine and Dentistry of New Jersey (UMDNJ; initial approval 4/18/06; protocol #0220060113) and Queens College (QC; initial approval 5/24/06; protocol #06-05-11-02) to carry out this study.

Process of Obtaining Subjects

Huntington's disease subjects and their caregivers were recruited via three methods: 1) Flyers were included as part of mailings disseminated by the Huntington's Disease Society of Americas' New Jersey Chapter, 2) Flyers were given to patients and caregivers attending Huntington's disease Clinics at UMDNJ Piscataway and Newark campuses, and 3) Announcements were made at at-risk, affected, and caregiver support

group meetings (sponsored by the HD Family Service Center) in Plainfield, Piscataway, Lakewood, and Stratford, New Jersey.

Pre-screening for HD Patients and Caregivers

HD patients and their caregivers were pre-screened for assessment of basic exclusionary criteria prior to the in-person appointment. Participants were read a narrative regarding the purpose and requirements of the study (See Appendix A) and they were given the opportunity to ask questions. Verbal permission was obtained to proceed with pre-screening questions, and a brief interview was conducted (See Appendix B), after which subjects were informed of their eligibility.

Testing

Informed consent was obtained prior to any formal testing. Though participants were offered the option of completing testing over multiple sessions, all opted to complete testing in a single session. Subjects were reimbursed for parking and were paid a $10 travel stipend to offset the cost of expenses incurred during travel. Participants were offered and provided with breaks whenever needed. All subjects that consented to participate completed the study.

HD Patients

HD subjects were administered screening, control, and experimental tasks, an interview, and two questionnaires. The patient filled out a medical release form, giving permission to obtain medical records pertaining to HD status and regarding neurological and psychiatric history. The total time for administration of consent forms, interviews, questionnaires, and experimental and control tasks was approximately 3 hours. Two of the HD subjects became quite fatigued toward the end of testing, so they were given an

abbreviated administration of the PAI. Although the use of the short form is less reliable than the full version of the same scale (Morey, 1991), and only 20 of the 22 scales are utilizable, the short form, nonetheless, retains adequate reliability and validity (Morey, 1991) and provided all crucial measures of concern in the current study.

Caregivers

Caregivers were administered two questionnaires regarding caregiver experience, the ZBI and the HDQoL-C. Total time for completion of these measures was approximately one hour.

Data Analysis

Data Screening

Summary statistics on all the variables and scatterplots of correlations among all variables with each of the caregiver burden scales were generated (See Appendix D). Descriptive statistics were inspected for out-of-range values and plausibility of means and standard deviations. Scatterplot distributions were visually inspected for outliers. Skewness, kurtosis, and the Kolmogorov-Smirnov test of normality were produced for all variables (See Appendix E). Violations of normality were addressed via the Kruskal-Wallis test in the case of ANOVAs and the Spearman rho in the case of correlations.

HD versus HC Comparisons

Broad and Individuated

A comparison was made between the HD and the HC groups. A two-tiered approach was selected in which each variable was analyzed in the context of the overall data set (i.e., “broad approach”), followed by a more specific analysis of the same variable with particular consideration to the pertinent demographics (i.e., “individuated

approach"). A broad inspection familiarizes us with of the pattern of performance between HD and HC subjects, and provides us with a foundation and context from which to interpret the finer analyses. Utilization of a "broad to specific" approach provides the opportunity to scrutinize variability within the HC sample, the appropriateness of the HC comparison group to the experimental group, and any potential power issues that may have arisen.

Broad Approach

In the broad approach, the overall HD group is simply compared to the overall HC group information.

Determination of Individuated Analyses by Demographic Matching

It is known that subject characteristics and situational factors (Borod, 2000; Ekman & Friesen, 1976; Lezak, 1995; Spreen & Strauss, 1991) can influence performance on neuropsychological tests and on emotion tasks, as well as the salience of a particular channel for emotion communication. In order to minimize the possibility that extraneous factors may affect performance on perceptual tasks of emotional processing, it was important to match the demographic features in patient and control groups as closely as possible. As such, attention was turned to the pattern of demographic effects within the healthy control group based on research examining this issue for the perception measures of the NYEB (Borod & Krch, 2008). In addition to existing tasks on the NYEB, created difference score values were also considered. These were derived by subtracting the experimental task score from the control task score for each channel. The resultant value represents emotional perception functioning after accounting for any potential general perception impairments within the respective domain. Difference score values can be

utilized in group comparisons, offering a statistical alternative to covarying for a potential confound (i.e., general perception impairments) in the absence of raw data. The following table (Table 6) summarizes the findings presented by Borod and Krch (2008):

Table 6. ANOVA for Experimental and Control Tasks, and their Difference Scores by Demographic Effect within the Healthy Control Group

	Main Effects				2-way Interactions						3-way Interactions				4-way Interaction
	A	G	R	E	A x G	A x R	A x E	G x R	G x E	R x E	A x G x R	A x G x E	A x R x E	G x R x E	A x G x R x E
Experimental Tasks															
FID	*														
PID	*														
SID	*			*		*									
Control Tasks															
BFRT															
ICP				*											
NESID	*				*						*				*
Difference Scores															
FIDDIFF	*														
PIDDIFF	*														
SIDDIFF				*		*								*	

A = Age; G = Gender; R = Ethnicity; E = Education; FID = Facial Identification Task; PID =Prosodic Identification Task; SID = Sentence Identification Task; BFRT = Benton Facial Recognition Task; ICP = Intonation Contours Perception Task; NESID = Nonemotional Sentence Identification Task; FIDDIFF = BFRT-FID; PIDDIFF = ICP-PID; and SIDDIFF = NESID-SID.

As can be seen from the above table, there was a main effect of age on the FID, PID, FIDDIFF, and PIDDIFF scores, and a main effect of education on ICP. A two-way interaction was seen on SID, a three-way interaction was seen on SIDDIFF, and a four-way interaction was seen on NESID. There were no demographic effects on the BFRT. HD versus HC analyses were carried out for each variable according to the demographics that were found to have had an effect within the HC population (See Table 7). For example, based on a significant age effect on the FID within the healthy control population, this characteristic was considered in the HD versus HC analyses on FID.

Since the majority of individuals in the overall HD group were between 40 and 59 years of age, this age group was utilized as the HC comparison cohort to which the HD sample was compared. A detailed explanation of how each analysis was carried out is provided in the Statistical Procedures section below.

Table 7. Demographics Considered for each Variable in Analyses

Task	Analysis based on:
Experimental Tasks	
FID	Age
PID	Age
SID	Age, education, and ethnicity
Control Tasks	
BFRT	Whole group
ICP	Education
NESID	Age, gender, ethnicity, education
Difference Scores	
FIDDIFF	Age
PIDDIFF	Age
SIDDIFF	Age, gender, ethnicity, education

FID = Facial Identification Task; PID =Prosodic Identification Task; SID = Sentence Identification Task; BFRT = Benton Facial Recognition Task; ICP = Intonation Contours Perception Task; NESID = Nonemotional Sentence Identification Task; FIDDIFF = BFRT-FID; PIDDIFF = ICP-PID; and SIDDIFF = NESID-SID.

The individuated approach that was performed for each variable was determined not only upon the demographics relevant to that variable, but also by the limitations of the HD sample composition and/or size. The impact of these two factors on the analyses will be discussed separately in the relevant sections. The breakdowns of sample size by demographic is found in Table 8. Education breakdown was based on the median education value in the HC sample.

Table 8. Demographics of Whole HD Group, and Nondemented and Demented Subgroups

		Age			Gender		Education	
	N	**20-39**	**40-59**	**60-81**	**M**	**F**	**≤15**	**≥16**
Overall HD Group	18	2	14	2	9	9	13	5
Nondemented Individuals	8	1	7	0	2	6	7	1
Demented Individuals	10	1	7	2	7	3	6	4

Huntington's Disease Group Partition

In addition to performing broad and individuated analyses for each variable, special consideration was made within the HD group. During testing, it became evident that a number of the individuals were dementing. It was felt that this cognitive impairment invalidated HD patients' performance on the experimental and control tasks. Based on the literature, a cutoff of 128 was utilized to identify those subjects considered to have moderate to severe dementia (Monsch et al., 1995; Montoya et al., 2006; Paulsen et al., 1995). Mean disease duration for the demented individuals was 9.7 years, and it was 8.0 years for the nondemented individuals. The demented individuals were then removed from the overall group, leaving 8 individuals in the nondemented group. All analyses involving HD subjects were carried out utilizing both the overall HD group and the subset of nondemented HD subjects.

Healthy Control Summary Data

As mentioned previously, the comparison group data were available in summary form (i.e., *M* and *SD*), in 1-way, 2-way, 3-way, and 4-way tables of age, gender, ethnicity, and education from Borod and Krch (2008). Statistical analyses are unable to be carried out utilizing statistical software with only summary data. They can, however, be accomplished through a multi-step process which involves some hand calculations.

The derivation of Sum of Squares (*SS*), degrees of freedom (*df*), and Mean Square (*MS*) values for all main effect and interaction terms are carried out using SPSS, calculation of the Error terms for between-subjects and within-subjects analyses are carried out using Excel, and calculation of the *MS* and *F*-ratio are carried out by hand. Please refer to Appendix F for a thorough elaboration of these steps and two sample problems. Comparisons for HDs versus HCs were performed in this manner.

The first sample problem utilizes only HD data, which allows the multiple-method calculations to be verified with the SPSS output that was calculated utilizing raw data. A 2 x 2 x 2 repeated-measures ANOVA was performed to assess the differences between men and women and among lesser and greater years of education on experimental and control tasks. Specifically, factors included two between-subjects factors, that is, Gender (2: male and female) and Education (2: ≤ 15 years education and ≥ 16 years of education), and one within-subjects factor, that is, Task (2: facial experimental and facial control). The dependent variable (DV) was percent correct for each task.

The second sample is provided to exemplify one of the actual analyses included in this study (i.e., the Broad Approach one-way ANOVA performed on the FIDDIFF variable, page 191), although lack of access to raw data prevented verification of the multiple-method calculations in SPSS. In this second sample, a 2 x 2 repeated-measures ANOVA was performed to assess the differences between HD and HC subjects on experimental and control tasks. Specifically, factors included one between-subjects factor, that is, Group (2: HD subjects and HC subjects), and one within-subjects factor,

that is, Task (2: facial experimental and facial control). The DV was the percent correct for each task.

Experimental Tasks

Generally speaking, group differences were assessed for each of the experimental tasks (facial, prosodic, and lexical). The DV for each task was the percent correct for that task.

Control Tasks

Analyses were conducted on the nonemotional control tasks (BFRT, ICP, and NESID) to determine whether there are any significant differences in performance between HD patients and NC subjects on control tasks. The DV was the percent correct for each task.

Difference Scores

If it were the case that HD and HC subjects performed significantly differently from each other on nonemotional control tasks, then this would be an indication that any emotional impairments found might be confounded by general perceptual impairments. To address this issue, difference scores were calculated between experimental and control tasks for each channel, and comparisons between HD and HC groups were made on difference scores. This procedure effectively parcels out any effect that the control task contributes to the dependent variable, thereby producing a residual that is a purer product of the experimental task effect on the dependent variable than the experimental measures themselves.

Statistical Procedures

Overview

There were several primary elements to the analysis and several supplementary analyses. The primary analyses included assessment of differences between HD and HC subjects on the experimental tasks, the control tasks, and the difference between these (which served as a measure of true emotional perceptual functioning, subtracting out potential general perceptual impairment); assessment of the HD subjects' ability to recognize the individual emotions; and assessment of the factors associated with increased levels of caregiver burden. In addition to primary analyses, supplementary analyses included determining the degree of interchannel correlations among NYEB experimental tasks in HD subjects; exploration of the relationship between CAG repeats and age of onset; and the exploration of the relationship between disease duration and performance on the NYEB experimental tasks. These analyses will be elaborated upon in the following sections.

Aim 1: HD versus HC Comparisons

Broad Approach

To investigate the emotional perceptual functioning of HD patients as compared to healthy controls from a broad approach, group comparisons for each of the three experimental tasks were performed. That is, three one-way analyses of variance (ANOVAs) were employed to assess differences between the two subject groups (all HDs and all HCs) for each of the three tasks (facial, prosodic, and lexical). The DV for each task was the percent correct for that task. This analysis was performed, again, with the demented individuals removed.

Individuated Approach

Facial Identification Task. Based on a significant Age effect on the FID within the healthy control population, this characteristic was considered in the HD versus HC analyses on FID. (See Appendix G for a visual summary of these analyses in tables).

This first analysis on FID involved the overall HD Group, which consists of two 20-39 y.o. individuals, fourteen 40-59 y.o. individuals, and two 60-81 y.o. individuals. There were too few HD subjects in the 20-39 and 60-81 y.o. age groups to warrant a two-way ANOVA. However, since the majority of the overall HD group is 40-59 y.o., this demographic was selected as the HC comparison group to which the HD sample was compared. Therefore, group differences between the overall HD group and 40-59 y.o. HCs were assessed through a one-way ANOVA. The DV was the percent correct on the FID. To determine if the performance of the four individuals who were outside of the majority demographic altered the results, the above analysis was repeated after having removed these individuals.

The second analysis on FID was performed with the nondemented HD subgroup, which consisted of individuals who were primarily (i.e., 7 of 8) aged 40-59. As such, this demographic was selected as the HC comparison group to which the HD sample was compared. Therefore, group differences between nondemented HD subjects and 40-59 y.o. HCs were assessed through a one-way ANOVA. The DV was the percent correct on the FID. To determine if the performance of the one individual who was outside of the majority demographic altered the results, the above analysis was repeated after having removed this individual.

Benton Facial Recognition Task. Within the healthy control sample, there were no age, gender, education, or ethnicity effects for the BFRT. As such, HD subjects and HCs were not partitioned by demographics, making the individuated analysis identical to that of the broad analysis. Therefore, no individuated analyses were carried out between HD and HC subjects for BFRT.

FIDDIFF: Difference Scores between FID and BFRT. Based on a significant Age effect on the FID-BFRT Difference Scores within the healthy control population, this characteristic was considered in the HD versus HC analyses on FIDDIFF.

The first analysis on FIDDIFF involved the overall HD group, which consists of two 20-39 y.o. individuals, fourteen 40-59 y.o. individuals, and two 60-81 y.o. individuals. There were too few HD subjects in the 20-39 and 60-81 y.o. age groups to warrant a two-way ANOVA (i.e. Age x Group). However, since the majority of the overall HD group is 40-59 y.o., this demographic was selected as the HC comparison group to which the HD sample was compared. Therefore, group differences between the overall HD group and 40-59 y.o. HCs were assessed through a one-way ANOVA. The DV was the difference score between percent correct on the FID and percent correct on the BFRT. To determine if the performance of the four individuals who were outside of the majority demographic altered the results, the above analysis was repeated after having removed these individuals.

The second analysis on FIDDIFF was performed with the nondemented HD subgroup, which consists primarily (i.e., 7 of 8) of individuals aged 40-59. As such, this demographic was selected as the HC comparison group to which the HD sample was compared. Therefore, group differences between nondemented HD subjects and 40-59

y.o. HCs were assessed through a one-way ANOVA. The DV was the difference score between percent correct on the FID and the BFRT. To determine if the performance of the one individual who was outside of the majority demographic altered the results, the above analysis was repeated after having removed this individual.

Prosodic Identification Task. Based on a significant Age effect on the PID within the healthy control population, this characteristic was considered in the HD versus HC analyses on PID.

The first analysis on PID involved the overall HD group, which consists of two 20-39 y.o. individuals, fourteen 40-59 y.o. individuals, and two 60-81 y.o. individuals. There were too few HD subjects in the 20-39 and 60-81 y.o. age groups to warrant a two-way ANOVA. However, since the majority of the overall HD group is 40-59 y.o., this demographic was selected as the HC comparison group to which the HD sample was compared. Therefore, group differences between the overall HD group and 40-59 y.o. HCs were assessed through a one-way ANOVA. The DV was the percent correct on the PID. To determine if the performance of the four individuals who were outside of the majority demographic altered the results, the above analysis was repeated after having removed these individuals.

The second analysis on PID was performed with the nondemented HD subgroup, which consists primarily (i.e., 7 of 8) of individuals aged 40-59. As such, this demographic was selected as the HC comparison group to which the HD sample was compared. Therefore, group differences between nondemented HD subjects and 40-59 y.o. HCs were assessed through a one-way ANOVA. The DV was the percent correct on the PID. To determine if the performance of the one individual who was outside of the

majority demographic altered the results, the above analysis was repeated after having removed this individual.

Intonation Contours Perception Task. Based on a significant Education effect on the ICP within the healthy control population, this characteristic was considered in the HD versus HC analyses on ICP.

The first analysis on ICP involved the overall HD group. Given that there were sufficient individuals in each cell of the education breakdown, a 2 x 2 Univariate ANOVA was performed to assess group differences. Specifically, between-subjects factors were Group (2: HD and HC) and Education (2: ≤ 15 years of education, ≥ 16 years of education). The DV was the percent correct on the ICP.

The second analysis on ICP involved the nondemented HD subgroup, which had insufficient numbers of individuals in each cell. The nondemented HD group consisted primarily (7 of 8) of individuals with ≤ 15 years of education (y.e.), so this demographic was selected as the HC group to which HD individuals were compared. Therefore, group differences between the nondemented HD subjects and HC individuals with ≤ 15 y.e. were assessed through a one-way ANOVA. The DV was the percent correct on the ICP. To determine if the performance of the one individual who was outside of the majority demographic altered the results, the above analysis was repeated after having removed this individual.

PIDDIFF: Difference Scores between PID and ICP. Based on a significant Age effect on the PIDDIFF within the healthy control population, this characteristic was considered in the HD versus HC analyses on PIDDIFF.

The first analysis on PIDDIFF involved the overall HD group, which consists of two 20-39 y.o. individuals, fourteen 40-59 y.o. individuals, and two 60-81 y.o. individuals. There were too few HD subjects in the 20-39 and 60-81 y.o. age groups to warrant a two-way ANOVA. However, since the majority of the overall HD group is 40-59 y.o., this demographic was selected as the HC comparison group to which the HD sample was compared. Therefore, group differences between the overall HD group and 40-59 y.o. HCs were assessed through a one-way ANOVA. The DV was the difference score between percent correct on the PID and the ICP. To determine if the performance of the four individuals who were outside of the majority demographic altered the results, the above analysis was repeated after having removed these individuals.

The second analysis on PIDDIFF was performed with the nondemented HD subgroup, which consists primarily (i.e., 7 of 8) of individuals aged 40-59. As such, this demographic was selected as the HC comparison group to which the HD sample was compared. Therefore, group differences between nondemented HD subjects and 40-59 y.o. HCs were assessed through a one-way ANOVA. The DV was the difference score between percent correct on the PID and the ICP. To determine if the performance of the one individual who was outside of the majority demographic altered the results, the above analysis was repeated after having removed this individual.

Sentence Identification Task. Based on significant Age, Education, and Ethnicity effects on the SID within the healthy control population, these characteristics were considered in the HD versus HC analyses on SID.

The first analysis on SID involved the overall HD group. The Age effect was addressed by taking the majority age demographic of the HD group and comparing it to

their HC counterparts. Education was addressed by considering it as a separate independent variable in the analysis. In order to address Ethnicity, no parceling was required in the HD group as it consists of all Caucasians; in the HC group, non-Caucasians were excluded. Therefore, group differences between the overall HD group and 40-59 y.o. Caucasian HCs were assessed in a 2 x 2 Univariate ANOVA. Specifically, between-subjects factors were Group (2: HD and HC) and Education (2: ≤ 15 years of education and ≥ 16 years of education). The DV was the percent correct on the SID. To determine if the performance of the four individuals, who were outside of the majority age demographic, altered the results, the above analysis was repeated after having removed these individuals.

The second analysis on SID was performed with the nondemented HD subgroup. Age and education were addressed simultaneously by comparing the majority demographics of the HD group to their HC counterparts. Seven of eight of these individuals were within the 40-59 y.o. age group and simultaneously had ≤ 15 y.e. (the remaining individual was 39 and had ≥ 16 y.e.). Ethnicity was not an issue in the HD group as they are comprised entirely of Caucasians. However, Ethnicity was addressed in the HC group by excluding all non-Caucasians from the comparison group. Given that the Kolmogorov-Smirnov test was significant for SID in the nondemented HD group, group differences between nondemented HD subjects and 40-59 y.o. Caucasian HCs with ≤ 15 y.e. were assessed in a Kruskal-Wallis ANOVA. The DV was the percent correct on the SID. To determine if the performance of the one individual who was outside of the majority age and education demographic altered the results, this individual was removed and the Kruskal-Wallis ANOVA was rerun.

Nonemotional Sentence Identification Task. Based on significant Age, Gender, Education, and Ethnicity effects on the NESID within the healthy control population, these characteristics were considered in the HD versus HC analyses on NESID.

The first analysis on NESID involved the overall HD Group. Gender served as its own control by virtue of there being equal numbers of men and women in the overall HD group. Age was addressed by selecting the majority age range of the overall HD group, 40-59 y.o., and comparing these individuals to their HC counterparts. Ethnicity, as noted above, addresses itself in the HD group and was addressed in the HC group by excluding non-Caucasians. Finally, education was addressed by considering it as a separate independent variable (IV) in the analysis. Therefore, group differences between the overall HD group and the 40-59 y.o. Caucasian HCs were assessed in a 2 x 2 Univariate ANOVA. Specifically, between-subjects factors were Group (2: HD and HC) and Education (2: $\leq$ 15 years of education and $\geq$ 16 years of education). The DV was the percent correct on the NESID. To determine if the performance of the four individuals who were outside of the majority age demographic altered the results, the above analysis was repeated after having removed these individuals.

The second analysis on NESID was performed with the nondemented HD subgroup. Age and education were able to be addressed simultaneously by comparing the majority demographics of the HD group to their HC counterparts; seven of eight of nondemented individuals were both within the 40-59 y.o. age group and had $\leq$ 15 y.e. (the remaining individual was 39 and had $\geq$ 16 y.e.). Ethnicity was addressed by excluding non-Caucasians in the HC group; the HD group already consisted of all Caucasians. Therefore, group differences between nondemented HD subjects and 40-59

y.o. Caucasian HCs with ≤ 15 y.e. were assessed in a one-way ANOVA. The DV was the percent correct on the NESID. To determine if the performance of the one individual who was outside of the majority age and education demographic altered the results, the above analysis was repeated after having removed this individual.

A third analysis was performed on NESID to address the Gender effect in the nondemented HD subgroup. As the equal gender split was not maintained in the nondemented subgroup, a separate analysis of this variable was necessitated. Six of the individuals were women and two were men. As women were in the majority, this demographic was selected as the HC comparison group to which the HD sample was compared. The Ethnicity demographic was considered again in this analysis. Therefore, group differences between the nondemented HD group and female Caucasian HCs were assessed through a one-way ANOVA. The DV was the percent correct on the NESID. To determine if the performance of the two male individuals altered the results, the above analysis was repeated after having removed these individuals.

SIDDIFF: Difference Scores between SID and NESID. Based on significant Age, Gender, Education, and Ethnicity effects on the SIDDIFF within the healthy control population, these characteristics were considered in the HD versus HC analyses on SIDDIFF.

The first analysis on SIDDIFF involved the overall HD Group. Gender served as its own control by virtue of there being equal numbers of males and females in the overall HD group. Age was addressed by selecting the majority age range of the overall HD group, (i.e., 40-59 y.o.), and comparing these individuals to their HC counterparts. Ethnicity addresses itself in the HD group by virtue of the sample consisting only of

Caucasians and is addressed in the HC group by filtering out the non-Caucasians. Finally, education was addressed by considering it as a separate IV in the analysis. Therefore, group differences between the overall HD group and 40-59 y.o. Caucasian HCs were assessed in a 2 x 2 Univariate ANOVA. Specifically, between-subjects factors were Group (2: HD and HC) and Education (2: ≤ 15 years of education, ≥ 16 years of education). The DV was the difference scores between percent correct on the SID and the NESID. To determine if the performance of the four individuals who were outside of the majority demographic altered the results, the above analysis was repeated after having removed these individuals.

The second analysis on SIDDIFF was performed with the nondemented HD subgroup. Age and education were able to be addressed simultaneously by comparing the majority demographics of the HD group to their HC counterparts; seven of the eight nondemented individuals were both within the 40-59 y.o. age group and had ≤ 15 y.e. (the remaining individual was 39 and had ≥ 16 y.e.). Ethnicity was addressed by filtering out non-Caucasians in the HC group; the HD group already consisted of all Caucasians. Therefore, group differences between nondemented HD subjects and 40-59 y.o. Caucasian HCs with ≤ 15 y.e. were assessed in a one-way ANOVA. The DV was the difference scores between percent correct on the SID and the NESID. To determine if the performance of the one individual who was outside of the majority age and education demographic altered the results, the above analysis was repeated after having removed this individual.

A third analysis was performed on SIDDIFF to address the Gender effect in the nondemented HD subgroup. As the equal gender split was not maintained in the

nondemented subgroup, a separate analysis of this variable was necessitated. Six of the individuals were female and two were male. As women were in the majority, this demographic was selected as the HC comparison group to which the HD sample was compared. The Ethnicity demographic was considered again in this analysis. Therefore, group differences between the nondemented HD group and female Caucasian HCs were assessed through a one-way ANOVA. The DV was the difference score between percent correct on the SID and the NESID. To determine if the performance of the two male individuals altered the results, the above analysis was repeated after having removed these individuals.

Aim 2: Huntington's Disease Performance by Emotions

Based on the literature's reports that HD patients are consistently worse at perceiving disgust and fear, too, to a slightly lesser degree, versus other emotions, one of the goals of the current study was to investigate whether similar patterns occurred in the current sample of HD subjects. A 3 X 8 ANOVA was performed to assess differences among tasks and emotions within the HD group. Specifically, factors included two within-subjects factors, that is, Task (3: FID, PID, and SID) and Emotion (8: Anger, Disgust, Fear, Happiness, Interest, Pleasant Surprise, Sadness, and Unpleasant Surprise). The dependent variable was the percent correct on each emotion. Significant *F*-values for any main effect were followed up with pairwise tests using the Bonferroni procedure. Subsequently, on an exploratory basis, separate analyses were performed to investigate the individual pattern of emotion recognition within each task. Three one-way ANOVAs were performed, with emotion as the within-subjects variable (8: Anger, Disgust, Fear, Happiness, Interest, Pleasant Surprise, Sadness, and Unpleasant Surprise). The dependent

variable was the percent correct on each emotion. Significant *F*-values were followed up with pairwise tests using the Bonferroni procedure.

Aim 3: Caregiver Burden Measures

The relationship between the two different caregiver burden measures was explored using the Pearson product-moment correlation procedure.

Aim 4: Emotional Perceptual Functioning and Caregiver Burden

Relationships among the three emotional perception tasks and caregiver burden were explored using the Pearson product-moment correlation procedure.

Aim 5: Psychiatric and Personality Functioning and Caregiver Burden

Relationships among psychiatric functioning, personality characteristics, and caregiver burden were explored using the Pearson product-moment correlation procedure, with one exception; one variable violated the Kolmogorov-Smirnov test of normality (See Appensix E), so a Spearman rho correlation was performed for this variable.

Aim 6: HD and Caregiver Characteristics and Caregiver Burden

Relationships among HD subject characteristics, HD subject cognitive profile, level of HD subject awareness, caregiver characteristics, and caregiver burden were explored using the Pearson product-moment and Spearman rho procedures, depending on whether the variable was normally distributed or not via the Komogorov-Smirnov test of normality. Please note that the results are displayed in Appendix E. In the case where the variable of interest was nominal or ordinal in nature, one-way ANOVAs were performed in lieu of correlations, and Kruskal-Wallis ANOVAs were performed in the case of normality violations.

Exploratory Analyses

Communication Channels

Given that previous research (Borod et al., 1998, 2000) has indicated intercorrelations exist among particular modalities, we were interested in investigating if these associations occur in the current study. We evaluated the relationships among the different communication channels using Pearson product-moment correlation.

CAG Repeat and Age of Onset

The literature states that there is a positive relationship between number of CAG repeats and age of onset of HD symptoms. To investigate this association within our sample, the relationship between CAG repeat size and age of onset was examined using Pearson product-moment correlations; only 14 subjects were utilized because the CAG repeat number was not available for four of the HD subjects.

Disease Duration and NYEB

It was thought that as the disease progresses, HD subjects would show decreasing performance on emotional perception functioning. As such, the relationships between disease duration at the time of testing and performance on the NYEB experimental tasks were explored using Pearson product-moment correlation.

RESULTS

Aim 1: HD versus HC Comparisons

Broad Approach

In the broad approach, HD subjects performed significantly worse than the HCs on all three experimental tasks: FID, F (1, 119) = 98.71, p = .000; PID, F (1, 119) = 40.34, p = .000; and SID, F (1, 119) = 39.44, p = .000. HD subjects performed worse than HCs on the nonemotional facial (BFRT, F [1, 119] = 35.35, p = .000) and prosodic (ICP, F [1, 119] = 38.85, p = .000) control tasks, but did not differ in their performance on the nonemotional lexical control task (NESID, F [1, 119] = .012, p = .730). See Table 9 for means and standard deviations for each variable for each subject group. When taking into account the contribution of nonemotional perceptual functioning, HD individuals remained impaired on the facial (FIDDIFF, F [1, 119] = 29.25, p = .000) and lexical (SIDDIFF, F [1, 119] = 34.25, p = .000) emotional tasks. However, this was not the case for the prosodic channel (PIDDIFF, F [1, 119] = 1.62, p = .205).

Table 9. Summary Statistics for Task by Group for the Overall HD Group using the Broad Approach

	Healthy Controls[a]		HD Subjects[b]	
	M	***SD***	***M***	***SD***
Experimental Tasks				
FID	73.30	11.03	43.92	14.40
PID	58.09	17.01	30.32	17.72
SID	79.45	13.13	58.10	14.33
Control Tasks				
BFRT	87.26	7.93	73.77	13.22
ICP	89.44	11.48	67.82	22.36
NESID	73.26	11.57	74.31	13.27
Difference Scores				
FIDDIFF	13.95	11.81	29.84	9.45
PIDDIFF	31.35	19.43	37.50	15.39
SIDDIFF	-6.19	15.30	16.20	12.85

FID = Facial Identification Task; PID = Prosodic Identification Task; SID = Sentence Identification Task; BFRT = Benton Facial Recognition Task; ICP = Intonation Contours Task; NESID = Nonemotional Sentence Identification Task.; FIDDIFF = BFRT-FID; PIDDIFF = ICP-PID; SIDDIFF = NESID-SID.
[a]n = 103. [b]n = 18

The reader is referred to Figures 5, 6, and 7 for graphs which display subject group performance on emotional and nonemotional tasks, separately for each channel. Note that in these graphs and all others, error bars represent SD.

Figure 5. Facial Task by Group for the Overall HD Group using the Broad Approach

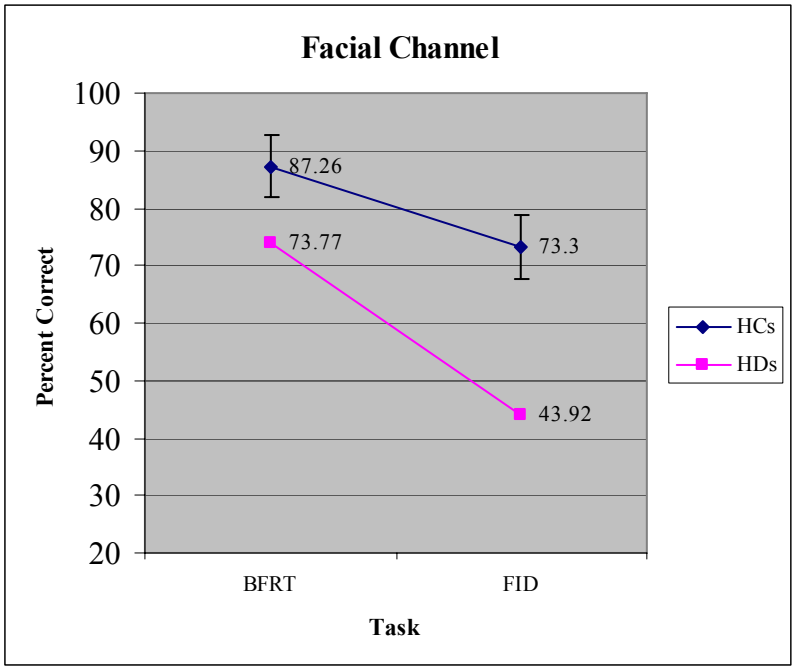


Figure 6. Prosodic Task by Group for the Overall HD Group using the Broad Approach

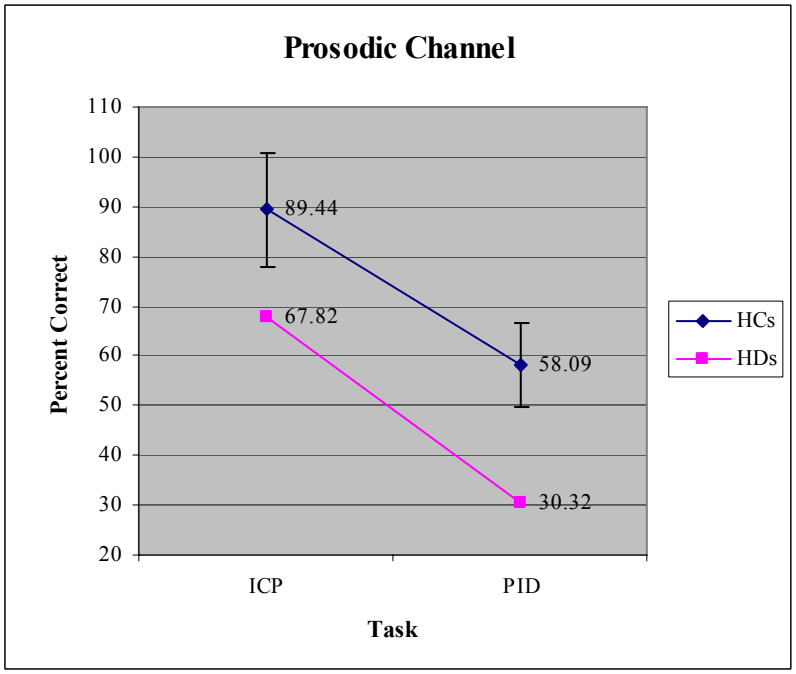
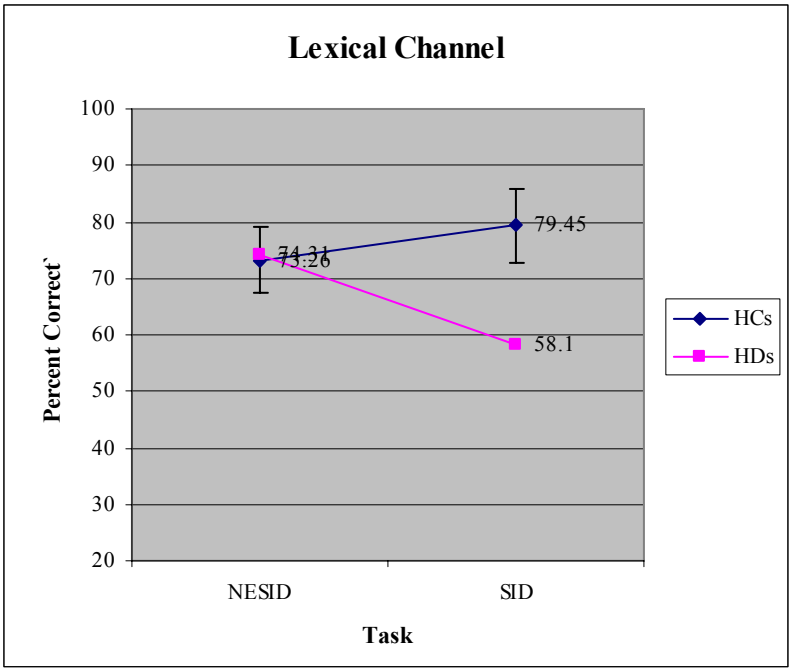


Figure 7. Lexical Task by Group for the Overall HD Group using the Broad Approach



The broad analysis was carried out again with the nondemented subgroup. The HD subjects performed significantly worse than the HCs on all three experimental tasks: FID, F (1, 109) = 20.98, p = .000; PID, F (1, 109) = 7.32, p = .008; and SID, F (1, 109) = 9.20, p = .003. HD subjects performed significantly worse than HCs on the facial perception task (BFRT, F [1, 109] = 5.08, p = .026), with a trend toward significance on the nonemotional prosodic (ICP, F [1, 109] = 3.06, p = .083) and lexical (NESID, F [1, 109] = 3.70, p = .057) tasks. See Table 10 for means and standard deviations for each variable for each subject group. When accounting for the contribution of the control task to the dependent variable, nondemented HD individuals remained impaired within the facial (FIDDIFF, F [1, 109] = 7.16, p = .009) and lexical channels (SIDDIFF, F [1, 109] = 16.34, p = .000), but not within the prosodic channel (PIDDIFF, F [1, 109] = 1.72, p = .192).

Table 10. Summary Statistics for Task by Group for the Nondemented HD Subgroup using the Broad Approach

	Healthy Controls[a]		HD Subjects[b]	
	M	***SD***	***M***	***SD***
FID	73.30	11.03	55.08	7.46
PID	58.09	17.01	41.15	17.88
SID	79.45	13.13	65.10	8.61
BFRT	87.26	7.93	80.56	10.24
ICP	89.44	11.48	81.77	17.39
NESID	73.26	11.57	81.25	6.68
FIDDIFF	13.95	11.81	25.48	10.57
PIDDIFF	31.35	19.43	40.63	16.63
SIDDIFF	-6.19	15.30	16.15	10.78

FID = Facial Identification Task; PID = Prosodic Identification Task; SID = Sentence Identification Task; BFRT = Benton Facial Recognition Task; ICP = Intonation Contours Task; NESID = Nonemotional Sentence Identification Task.; FIDDIFF = BFRT-FID; PIDDIFF = ICP-PID; SIDDIFF = NESID-SID.
[a]n = 103. [b]n = 8

The reader is referred to Figures 8, 9, and 10 for graphs which display subject group performance on emotional and nonemotional tasks, separately for each channel, using the Broad Approach.

Figure 8. Facial Task by Group for the Nondemented HD Subgroup using the Broad Approach

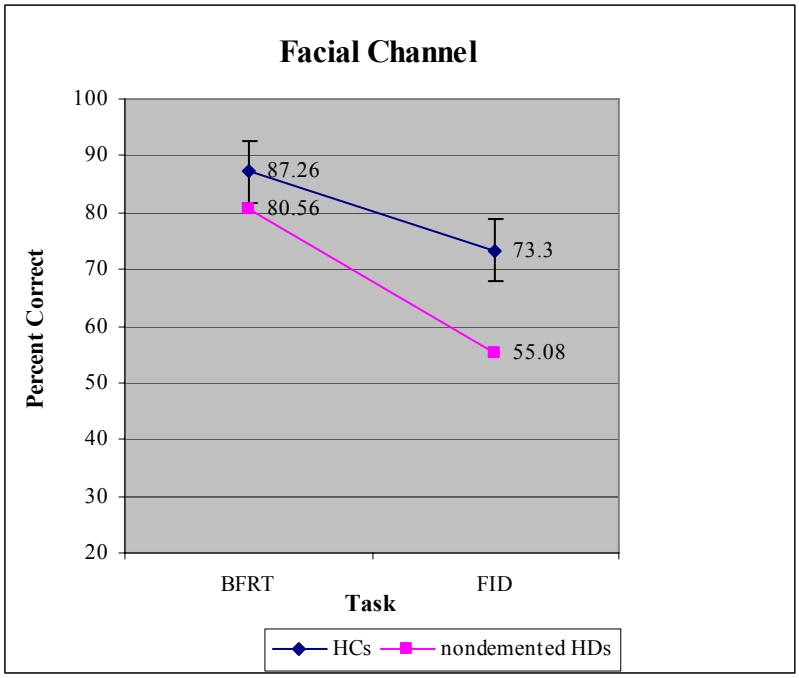


Figure 9. Prosodic Task by Group for the Nondemented HD Subgroup using the Broad Approach

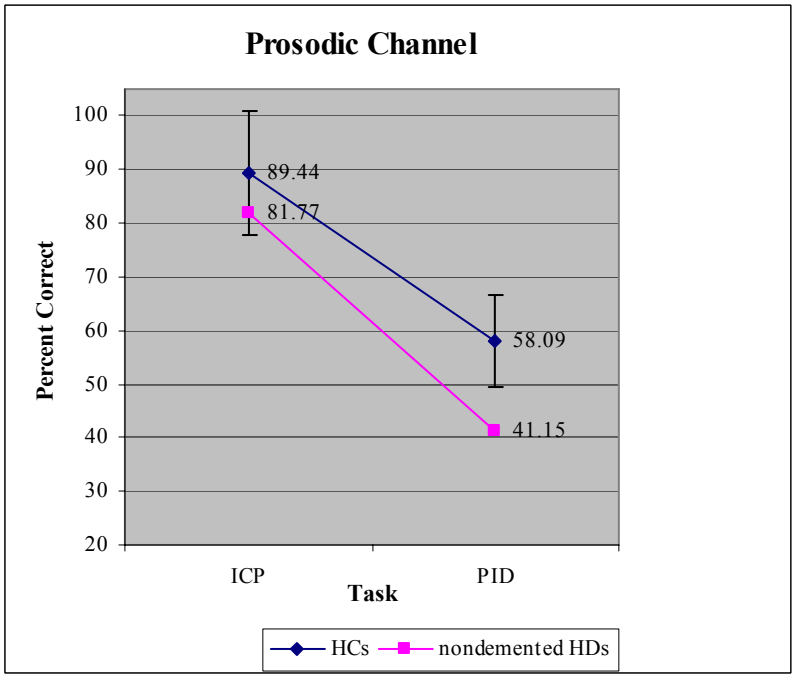
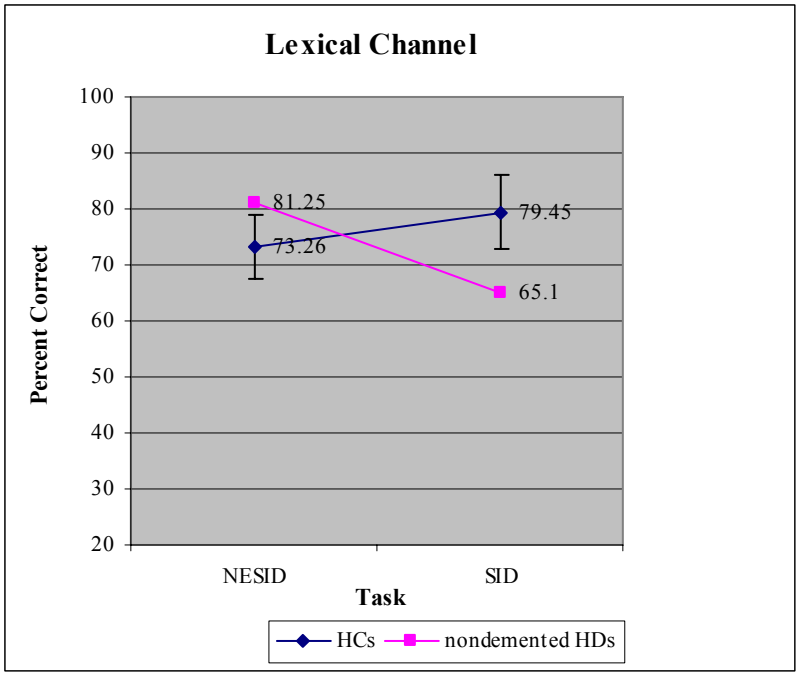


Figure 10. Lexical Task by Group for the Nondemented HD Subgroup using the Broad Approach



Individuated Approach

Facial Identification Task

Overall HD Group. HD subjects were significantly less accurate than the HCs at identifying facial emotion, F (1, 48) = 75.75, p = .000. See Table 11 for means and standard deviations for each subject group.

Table 11. FID Summary Statistics by Group for the Overall HD Group using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***N***
73.83	9.84	32	43.92	14.40	18

When this same analysis was performed after removing the four HD individuals who did not fall within the majority age demographic, the results remained significant, F (1, 44) = 71.46, p = .000.

Nondemented HD Subgroup. Nondemented HD subjects were significantly less accurate than the HCs at identifying facial emotion, F (1, 38) = 25.20, p = .000. See Table 12 for means and standard deviations for each subject group.

Table 12. FID Summary Statistics by Group for the Nondemented HD Subgroup using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
73.83	9.84	32	55.08	7.46	8

When this same analysis was performed after removing the one HD individual who did not fall within the majority age demographic, the results remained significant, F (1, 37) = 28.66, p = .000.

Benton Facial Recognition Task. Within the healthy control sample, there were no age, gender, education, or ethnicity effects; as such, HD subjects and HCs were not partitioned by demographics. The results of the overall analysis can be seen in the "Broad Approach" section above.

FIDDIFF: Difference Scores between FID and BFRT

Overall HD Group. HD subjects showed significantly greater discrepancies in performance between the experimental and control tasks than the HC subjects, F (1, 48) = 29.35, p = .000. This indicates that HD subjects remained significantly impaired on facial emotional perception, even after controlling for facial perception impairments. See Table 13 for means and standard deviations for each subject group.

Table 13. FIDDIFF Summary Statistics by Group for the Overall HD Group using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***N***
12.92	11.19	32	29.84	9.45	18

When this same analysis was performed after removing the four HD individuals who did not fall within the majority age demographic, the results remained significant, F (1, 44) = 27.74, p = .000.

Nondemented HD Subgroup. Nondemented HD subjects showed significantly greater discrepancies in performance between the experimental and control tasks than the HC subjects, F (1, 38) = 8.23, p = .007. This finding indicates that nondemented HD subjects remained significantly impaired on facial emotional perception, even after controlling for facial perception impairments. See Table 14 for means and standard deviations for each subject group.

Table 14. FIDDIFF Summary Statistics by Group for the Nondemented HD Subgroup using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
12.92	11.19	32	25.48	10.47	8

When this same analysis was performed after removing the one HD individual who did not fall within the majority age demographic, the results remained significant, F (1, 37) = 8.53, p = .006).

Prosodic Identification Task

Overall HD Group. HD subjects were significantly less accurate than HCs at identifying prosodic emotion, F (1, 48) = 39.73, p = .000. See Table 15 for means and standard deviations for each subject group.

Table 15. PID Summary Statistics by Group for the Overall HD Group using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***N***
60.03	14.96	32	30.32	17.72	18

When this same analysis was performed after removing the four HD individuals who did not fall within the majority age demographic, the results remained significant, F (1, 44) = 37.76, p = .000.

Nondemented HD Subgroup. Nondemented HD subjects were significantly less accurate than the HCs at identifying prosodic emotion, F (1, 38) = 9.45, p = .004. See Table 16 for means and standard deviations for each subject group.

Table 16. PID Summary Statistics by Group for the Nondemented HD Subgroup using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
60.03	14.96	32	41.15	17.88	8

When this same analysis was performed after removing the one HD individual who did not fall within the majority age demographic, the results remained significant, F (1, 37) = 10.18, p = .003.

Intonation Contours Perception Task

Overall HD Group. The interaction between Group and Education was not significant, F (1, 117) = 0.00, p = .960. There was no main effect of Education collapsed across experimental and control groups, F (1, 117) = 1.84, p = .178. A significant main effect of Group was found, F (1, 117) = 30.02, p = .000, wherein the HD subjects were significantly less accurate than the HC subjects at identifying nonemotional prosody. See Table 17 for means and standard deviations for each subject group by education grouping.

Table 17. ICP Summary Statistics by Education and Group for the Overall HD Group using the Individuated Approach

	Healthy Controls			HD Subjects		
	M	***SD***	***N***	***M***	***SD***	***N***
≤ 15	87.29	13.39	58	66.35	20.09	13
≥ 16	92.22	7.67	45	71.67	29.81	5
Totals	89.44	11.48	103	67.82	22.36	18

Nondemented HD Subgroup. Nondemented HD subjects were not significantly different than the HCs at identifying nonemotional prosody, F (1, 64) = 1.11, p = .296. See Table 18 for means and standard deviations for each subject group.

Table 18. ICP Summary Statistics by Group for the Nondemented HD Subgroup using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
87.29	13.39	58	81.77	17.39	8

When this same analysis was performed after removing the one HD individual who did not fall within the majority education demographic, the results remained nonsignificant, F (1, 63) = 2.17, p = .146.

PIDDIFF: Difference Scores between PID and ICP

Overall HD Group. HD subjects showed discrepancies in performance between the experimental and control tasks similar in degree to those of the HC subjects, F (1, 48) = 2.26, p = .140. This means that after controlling for facial perception impairments, HD subjects were no longer impaired on facial emotional perception. See Table 19 for means and standard deviations for each subject group.

Table 19. PIDDIFF Summary Statistics by Group for the Overall HD Group using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***N***
29.43	19.63	32	37.50	15.39	18

When this same analysis was performed after removing the four HD individuals who did not fall within the majority age demographic, the results remained nonsignificant, F (1, 44) = 1.52, p = .224.

Nondemented HD Subgroup. The degree of discrepancy that nondemented HD subjects showed between the experimental and control tasks was not significantly different than that of the HC subjects, F (1, 38) = 2.20, p = .146. This means that after controlling for facial perception impairments, HD subjects were no longer impaired on prosodic emotional perception. See Table 20 for means and standard deviations for each subject group.

Table 20. PIDDIFF Summary Statistics by Group for the Nondemented HD Subgroup using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
29.43	19.63	32	40.63	16.63	8

When this same analysis was performed after removing the one HD individual who did not fall within the majority age demographic, the results remained nonsignificant, F (1, 37) = 1.68, p = .203.

Sentence Identification Task

Overall HD Group. The interaction between Group and Education was not significant, F (1, 32) = 0.29, p = .594. There was no main effect of Education collapsed across experimental and control groups, F (1, 32) = 0.73, p = .399. A significant main effect of Group was found, F (1, 32) = 19.87, p = .000, wherein the HD subjects were significantly less accurate than the HC subjects at identifying lexical emotion. See Table 21 for means and standard deviations for each subject group by education grouping.

Table 21. SID Summary Statistics by Education and Group for the Overall HD Group using the Individuated Approach

	Healthy Controls			HD Subjects		
	M	***SD***	***N***	***M***	***SD***	***N***
≤ 15	75.93	12.46	9	57.69	14.11	13
≥ 16	82.41	10.16	9	59.17	16.51	5
Totals	79.17	11.52	18	58.10	14.33	18

When this same analysis was performed after removing the four HD individuals who did not fall within the majority age demographic, the results remained similar, with no interaction, F (1, 28) = 2.51, p = .125, no main effect of Education, F (1, 28) = 0.09, p = .769, and a significant main effect of Group, F (1, 28) = 23.52, p = .000.

Nondemented HD Subgroup. Nonparametric testing using the Kruskal-Wallis procedure revealed that nondemented HD subjects were less accurate than the HCs at identifying lexical emotion, though not at a conventionally significant level, χ^2 (1) = 3.50, p = .061. See Table 22 for medians for each subject group.

Table 22. SID Nonparametric Summary Statistics by Group for the Nondemented HD Subgroup using the Individuated Approach

Healthy Controls		HD Subjects	
Median	***n***	***Median***	***N***
11.11	9	6.63	8

When the same analysis was performed after removing the one HD individual who did not fall within the majority age and education demographic, the results remained similar for the nonparametric Kruskal-Wallis one-way ANOVA, χ^2 (1) = 2.96, p = .085.

Nonemotional Sentence Identification Task

Overall HD Group. The interaction between Group and Education was not significant, F (1, 32) = 0.28, p = .604. There was no significant main effect of Group, F (1, 32) = 0.12, p = .732, nor Education, F (1, 32) = 0.10, p = .757. See Table 23 for means and standard deviations for each subject group by education grouping.

Table 23. NESID Summary Statistics by Education and Group for the Overall HD Group using the Individuated Approach

	Healthy Controls			HD Subjects		
	M	***SD***	***n***	***M***	***SD***	***N***
≤ 15	74.54	11.50	9	75.32	11.72	13
≥ 16	75.46	10.51	9	71.67	18.02	5
Totals	75.00	10.69	18	74.31	13.27	18

When this same analysis was performed after removing the four HD individuals who did not fall within the majority age demographic, the results remained similar, with no interaction, F (1, 28) = 2.17, p = .152, no main effect of Group, F (1, 28) = 1.49, p = .233, and no main effect of Education, F (1, 28) = 1.64, p = .211.

Nondemented HD Subgroup. *Age and Education.* Nondemented HD subjects were not significantly different than the HCs in their accuracy in nonemotional lexical

perception, F (1, 15) = 2.09, p = .169. See Table 24 for means and standard deviations for each subject group.

Table 24. NESID Summary Statistics by Group for the Nondemented HD Subgroup, Age and Education Breakdown, using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
74.54	11.50	9	81.25	6.68	8

When this same analysis was performed after having removed the 39 y.o. with ≥ 16 y.e. from the group, the results remained nonsignificant, F (1, 14) = 1.41, p = .255.

Nondemented HD Subgroup. *Gender.* Nondemented HD subjects were not significantly different than the HCs in their accuracy in nonemotional lexical perception, F (1, 37) = 2.55, p = .119. See Table 25 for means and standard deviations for each subject group.

Table 25. NESID Summary Statistics by Group for the Nondemented HD Subgroup, Gender Breakdown, using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
75.00	10.49	31	81.25	6.68	8

When this same analysis was performed after having removed the two men from the HD group, the results remained nonsignificant, F (1, 35) = 1.17, p = .287.

SIDDIFF: Difference Scores between SID and NESID

Overall HD Group. The interaction between Group and Education was not significant, F (1, 32) = 0.00, p = .969. There was no main effect of Education, F (1, 32) = 0.95, p = .336. There was a significant main effect of Group, F (1, 32) = 12.35, p = .001, such that HD subjects showed significantly greater discrepancies in performance between

the experimental and control tasks than did the HC subjects. This indicates that HD subjects remained significantly impaired on lexical emotional perception, even after controlling for lexical perception functioning. See Table 26 for means and standard deviations for each subject group by education grouping.

Table 26. SIDDIFF Summary Statistics by Education and Group for the Overall HD Group using the Individuated Approach

	Healthy Controls			HD Subjects		
	M	***SD***	***N***	***M***	***SD***	***N***
≤ 15	-1.39	18.16	9	17.63	13.84	13
≥ 16	-6.94	17.05	9	12.50	10.21	5
Totals	-4.17	17.33	18	16.20	12.85	18

When this same analysis was performed after removing the four HD individuals who did not fall within the majority age demographic, the results remained similar, with no interaction, F (1, 28) = 0.02, p = .888, no main effect of Education, F (1, 28) = 0.57, p = .458, and a significant main effect of Group, F (1, 28) = 8.93, p = .006.

Nondemented HD Subgroup. *Age and Education*. Nondemented HD subjects showed significantly greater discrepancies in performance between the experimental and control tasks than did the HC subjects, F (1, 15) = 5.66, p = .031. This indicates that nondemented HD subjects remained significantly impaired on lexical emotional perception, even after controlling for lexical perception functioning. See Table 27 for means and standard deviations for each subject group.

Table 27. SIDDIFF Summary Statistics by Group for the Nondemented HD Subgroup, Age and Education Breakdown, using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
-1.39	18.16	9	16.15	10.78	8

When this same analysis was performed after having removed the 39 y.o. with ≥ 16 y.e. from the group, the results remained similar, although not significant at the conventional level, F (1, 14) = 4.34, p = .056.

Nondemented HD Subgroup. *Gender*. Nondemented HD subjects showed significantly greater discrepancies in performance between the experimental and control tasks than did the HC subjects, F (1, 37) = 12.57, p = .001. This indicates that nondemented HD subjects remained significantly impaired on lexical emotional perception, even after controlling for lexical perception functioning. See Table 28 for means and standard deviations for each subject group.

Table 28. SIDDIFF Summary Statistics by Group for the Nondemented HD Subgroup, Gender Breakdown, using the Individuated Approach

Healthy Controls			HD Subjects		
M	***SD***	***n***	***M***	***SD***	***n***
-5.38	16.18	31	16.15	10.78	8

When this same analysis was performed after having removed the two men from the HD group, the results remained significant, F (1, 35) = 7.65, p = .009.

Individuated Graphs

The reader is referred to Figures 11, 12, and 13 for graphs which display subject group performance on emotional and nonemotional tasks, separately for each channel, for the overall HD group using the Individuated Approach. Figures 14, 15, and 16 show subject group performance on emotional and nonemotional tasks, separately for each channel, for the nondemented HD subgroup using the Individuated Approach.

Figure 11. Facial Task by Group for the Overall HD Group using the Individuated Approach

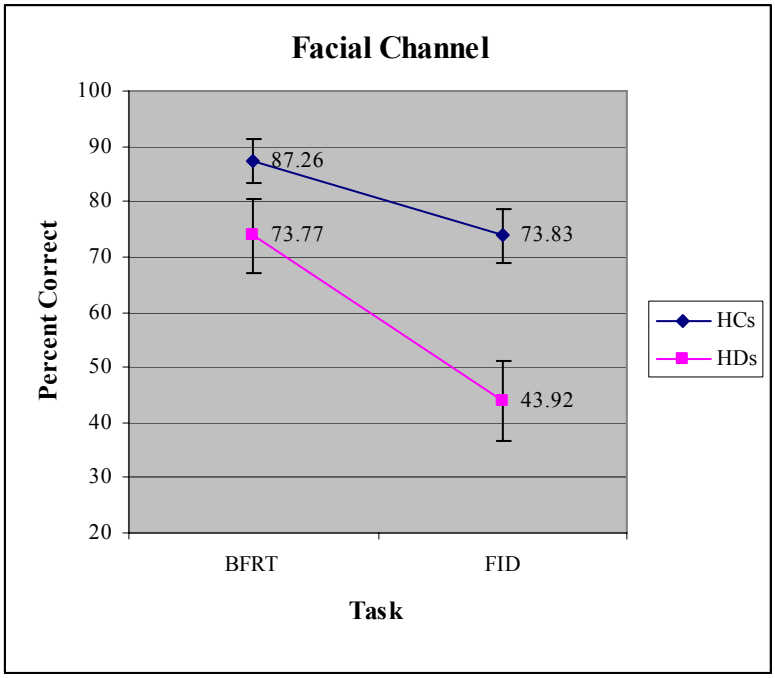


Figure 12. Prosodic Task by Group for the Overall HD Group using the Individuated Approach

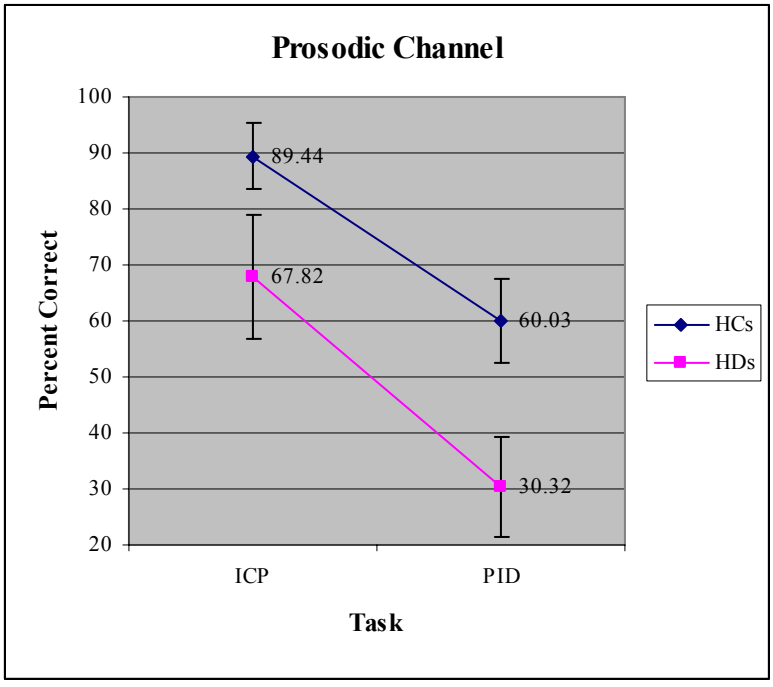


Figure 13. Lexical Task by Group for the Overall HD Group using the Individuated Approach

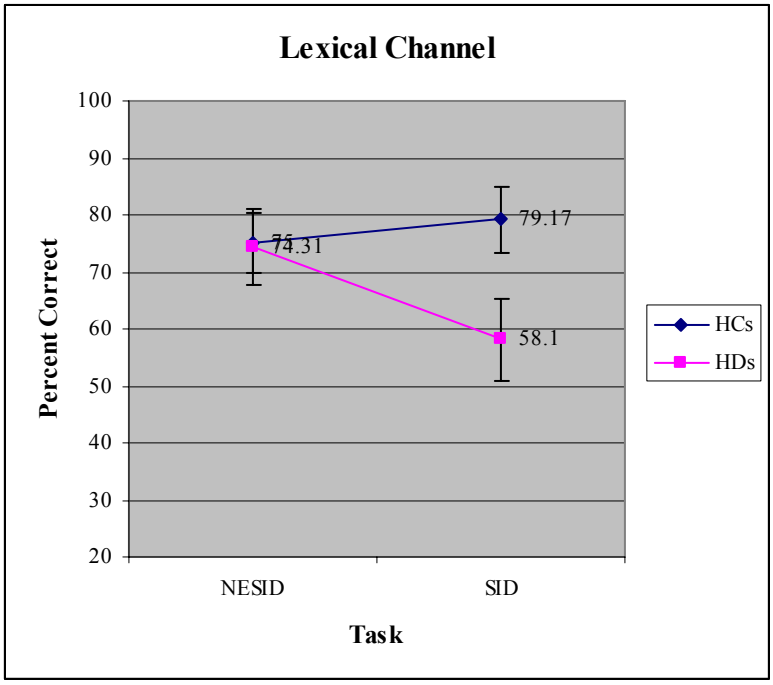


Figure 14. Facial Task by Group for the Nondemented HD Subgroup using the Individuated Approach

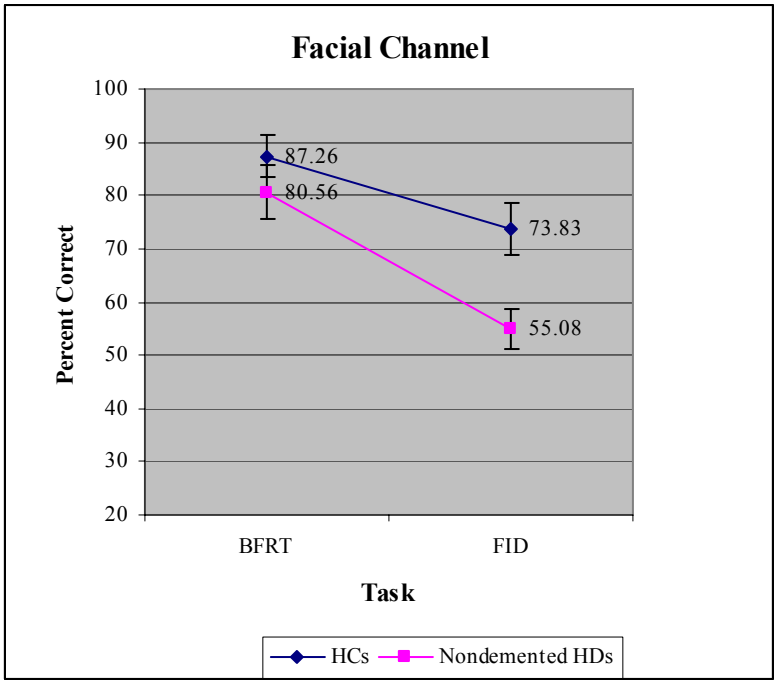


Figure 15. Prosodic Task by Group for the Nondemented HD Subgroup using the Individuated Approach

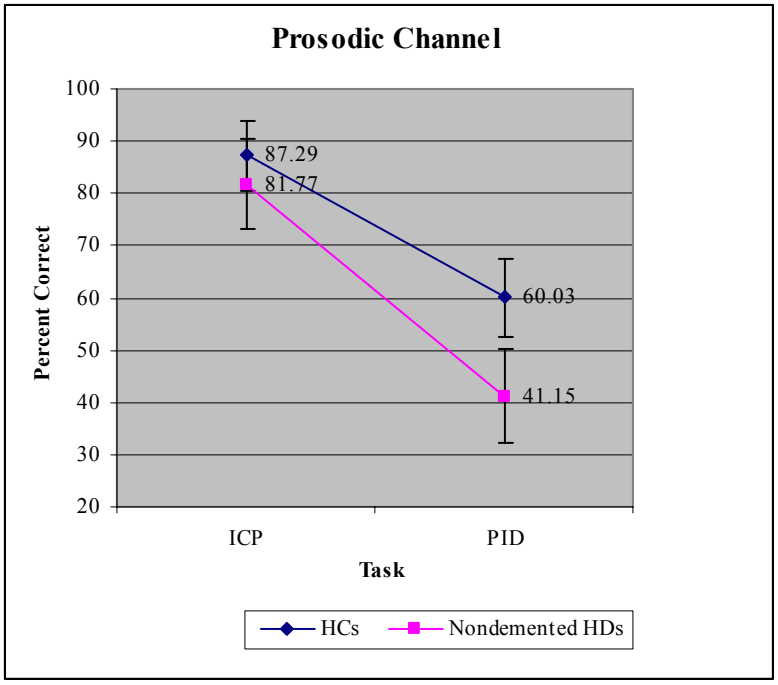
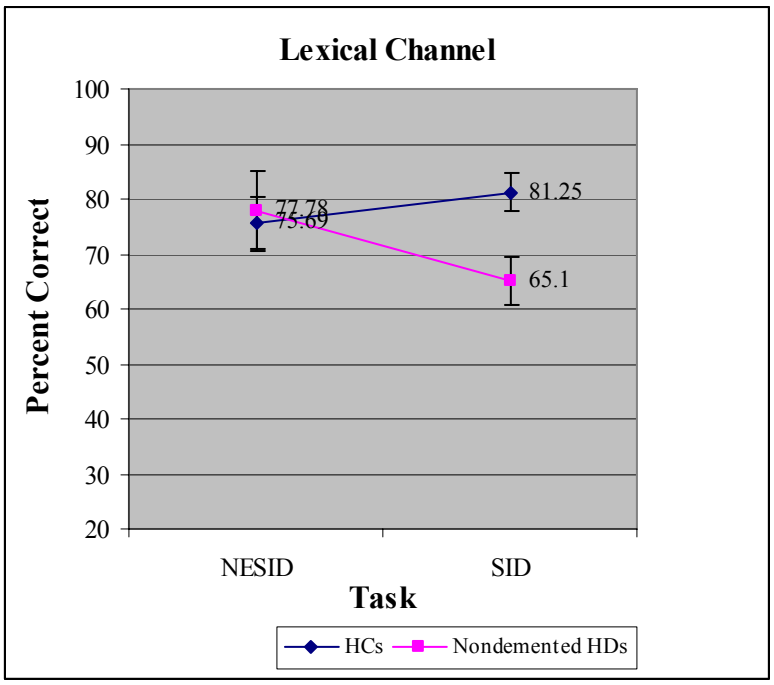


Figure 16. Lexical Task by Group for the Nondemented HD Subgroup using the Individuated Approach



Aim 2: Huntington's Disease Performance by Emotions

Emotion Performance Collapsed Across Tasks for the Overall HD Group

A significant main effect of Task, F (2, 34) = 37.31, p = .000, was found. On a post-hoc basis, overall emotion recognition performance was investigated across tasks through the use of pairwise comparisons using the Bonferroni adjustment for multiple comparisons. HD subjects performed significantly better on the lexical emotional perception task (M = 58.10, SD = 14.33) as compared to facial (M = 43.92, SD = 14.40) and prosodic (M = 30.32, SD = 17.72) tasks. They also performed significantly better on the facial task (M = 43.92, SD = 14.40) as compared to the prosodic task (M = 30.32, SD = 17.72). There was a significant main effect of Emotion, F (7, 119) = 8.30, p =.000. On a post-hoc basis, emotion recognition performance was investigated across tasks through the use of pairwise comparisons using the Bonferroni adjustment for multiple comparisons. Sadness (M = 60.80, SD = 32.36) was recognized with significantly greater accuracy than unpleasant surprise (M = 39.51, SD = 31.81), disgust (M = 37.19, SD = 32.00), fear (M = 32.10, SD = 26.72), and interest (M = 30.71, SD = 31.51). Happiness (M = 56.33, SD = 36.27) was recognized with significantly greater accuracy than fear (M = 32.10, SD = 26.72) and interest (M = 30.71, SD = 31.51). Anger (M = 52.31, SD = 33.39) was recognized with significantly greater accuracy than fear (M = 32.10, SD = 26.72) and interest (M = 30.71, SD = 31.51). There was a significant interaction between Task and Emotion, F (14, 238) = 3.88, p = .000, indicating that the pattern in HD subjects' ability to perceive emotions varied from task to task. The order of emotion recognition for FID was H > S > A > US > D > PS > F > I; the order for PID was S > A > I > PS > US > D > F and H; and the order for SID was S > H > A > PS > US > F > D > I.

These patterns will be described further in the subsequent sections. The individual data are presented in Table 29.

Table 29. Percent Correct Across Emotion Tasks for Overall HD Group

	A	D	F	H	I	PS	S	US	Overall
FID	45.8	43.1	27.8	76.4	23.6	33.3	58.3	44.4	44.10
PID	42.6	22.2	18.5	18.5	37.0	35.2	46.3	24.1	30.56
SID	68.5	46.3	50.0	74.1	31.5	63.0	77.8	51.9	57.87
Overall	52.3	37.2	32.1	56.3	30.7	43.8	60.8	39.5	

A = Anger; D = Disgust; F = Fear; H = Happy; I = Interest; PS = Pleasant Surprise; S = Sadness; US = Unpleasant Surprise; FID = Facial Identification Task; PID = Prosodic Identification Task; SID = Sentence Identification Task.

Emotion Performance within the Facial Channel

A significant main effect of emotion was found on the facial identification task, F (7, 119) = 9.04, p = .000. On a post-hoc basis, emotion recognition performance was investigated through the use of pairwise comparisons using the Bonferroni adjustment for multiple comparisons. Happiness (M = 76.39, SD = 23.44) was recognized with significantly greater accuracy than anger (M = 45.83, SD = 23.09), unpleasant surprise (M = 44.44, SD = 30.38), disgust (M = 43.06, SD = 26.85), pleasant surprise (M = 33.33, SD = 27.12), fear (M = 27.78, SD = 20.81), and interest (M = 23.61, SD = 31.47). Sadness (M = 58.33, SD = 29.70) was recognized with significantly greater accuracy than fear (M = 27.78, SD = 20.81).

Emotion Performance within the Prosodic Channel

A significant main effect of emotion was found on the prosodic identification task, F (7, 119) = 3.33, p = .003. On a post-hoc basis, emotion recognition performance was investigated through the use of pairwise comparisons using the Bonferroni adjustment for multiple comparisons. Sadness (M = 46.30, SD = 30.55) was recognized with significantly greater accuracy than unpleasant surprise (M = 22.22, SD = 25.57).

Emotion Performance within the Lexical Channel

A significant main effect of emotion was found on the prosodic identification task, F (7, 119) = 5.03, p = .000. On a post-hoc basis, emotion recognition performance was investigated through the use of pairwise comparisons using the Bonferroni adjustment for multiple comparisons. Sadness (M = 77.78, SD = 30.25) was recognized with significantly greater accuracy than interest (M = 31.48, SD = 31.25). Happiness (M = 74.07, SD = 26.95) was recognized with significantly greater accuracy than fear (M = 50.00, SD = 28.58) and interest (M = 31.48, SD = 31.25).

Analysis of Incorrect Responses on the Emotion Perception Tasks

Complementary to analyzing the accuracy of emotional perception is an exploration of HD subjects' incorrect responding. Error analysis provides insight into the subjects' perceptual mindset and provides important qualitative information about the nature of the error. Do subjects respond randomly to alternatives or do they tend to select particular alternatives? Are errors logical (e.g., confusing unpleasant surprise and fear) or are they unusual (e.g., mistaking fear for happiness)? Of particular interest is responding related to the emotion disgust, which has been found to be most often impaired. Fear and anger have also been found to be poorly recognized, but to a lesser degree. Tables 30-32 provide the pattern of emotions perceived for each emotion presented across the three channels.

Table 30. Percent of Emotions Endorsed by Emotions Presented on FID for Overall HD Group

	Subjects' responses							
Emotion presented	**A**	**D**	**F**	**H**	**I**	**PS**	**S**	**US**
Anger	**45.8**	8.3	6.9	2.8	12.5	4.2	6.9	12.5
Disgust	13.9	**43.1**	2.8	6.9	2.8	2.8	16.7	11.1
Fear	9.7	11.1	**27.8**	4.2	9.7	6.9	5.6	25.0
Happy	4.2	1.4	0	**76.4**	4.2	13.9	0	0
Interest	4.2	11.1	5.6	5.6	**23.6**	6.9	25.0	18.1
P Surprise	4.2	1.4	0	58.3	0	**33.3**	0	2.8
Sadness	19.4	6.9	6.9	1.4	0	1.4	**58.3**	5.6
U Surprise	8.3	8.3	11.1	5.6	0	13.9	8.3	**44.4**

A = Anger; D = Disgust; F = Fear; H = Happy; I = Interest; PS = Pleasant Surprise; S = Sadness; US = Unpleasant Surprise.

Within the facial task, disgust was found to be recognized fairly equivalently to other emotions. When disgust stimuli were presented and an incorrect response was produced, the alternative response most frequently endorsed was sadness, another negative valence emotion. Fear was perceived as unpleasant surprise as frequently as it was correctly identified as fear, which is not unexpected given that a fearful reaction might be one that is unanticipated (i.e., surprising) and unpleasant. Anger was very well recognized, with the most frequently endorsed alternative emotions being unpleasant surprise and interest. In general, persons tended to select interest as a catch-all category when they were unsure of which response to choose.

Table 31. Percent of Emotions Endorsed by Emotions Presented on PID for Overall HD Group

	Subjects' responses							
Emotion presented	**A**	**D**	**F**	**H**	**I**	**PS**	**S**	**US**
Anger	**42.6**	22.2	1.9	3.7	7.4	7.4	1.9	13.0
Disgust	13.0	**22.2**	7.4	11.1	11.1	14.8	9.3	11.1
Fear	5.6	7.4	**18.5**	11.1	13.0	9.3	7.4	27.8
Happy	1.9	13.0	5.6	**18.5**	22.2	33.3	3.7	1.9
Interest	3.7	1.9	1.9	9.3	**37.0**	16.7	14.8	14.8
P Surprise	5.6	3.7	3.7	22.2	5.6	**35.2**	1.9	22.2
Sadness	7.4	14.8	7.4	7.4	0	5.6	**46.3**	11.1
U Surprise	5.6	14.8	3.7	16.7	5.6	25.9	3.7	**24.1**

A = Anger; D = Disgust; F = Fear; H = Happy; I = Interest; PS = Pleasant Surprise; S = Sadness; US = Unpleasant Surprise.

The prosodic emotional identification task is inherently more difficult than the other two experimental tasks, and HD subjects' accuracy in identification across all emotions was relatively poorer. When an emotion was perceived incorrectly, subjects had no clear preference for a specific alternative emotion. This can be seen in a fairly even distribution of incorrect responses across the matrix. When presented with prosodic disgust, HD subjects were more likely to correctly recognize it than not; alternative guesses, however, appeared to be random. Interestingly, one of the most common alternative responses was pleasant surprise; a response inconsistent even in terms of valence. When presented with fear stimuli, subjects most often endorsed unpleasant surprise. Although they correctly identified fear next most frequently, other similarly frequent alternative responses were interest and happiness, both quite discrepant even in valence. Anger was (again) quite well recognized; the most frequent alternative response, however, was disgust, followed by unpleasant surprise.

Table 32. Percent of Emotions Endorsed by Emotions Presented on SID for Overall HD Group

	Subjects' responses							
Emotion presented	**A**	**D**	**F**	**H**	**I**	**PS**	**S**	**US**
Anger	**68.5**	13.0	0	0	0	0	11.1	7.4
Disgust	1.9	**46.3**	27.8	0	1.9	0	1.9	20.4
Fear	5.6	0	**50.0**	5.6	9.3	7.4	3.7	18.5
Happy	0	0	0	**74.1**	11.1	13.0	0	1.9
Interest	0	0	0	18.5	**31.5**	44.4	0	5.6
P Surprise	0	0	0	35.2	0	**63.0**	0	1.9
Sadness	3.7	3.7	3.7	0	0	0	**77.8**	11.1
U Surprise	11.1	14.8	13.0	0	0	1.9	7.4	**51.9**

A = Anger; D = Disgust; F = Fear; H = Happy; I = Interest; PS = Pleasant Surprise; S = Sadness; US = Unpleasant Surprise.

Within the lexical task, fear was recognized well relative to the accuracy with which it was recognized in the other two experimental tasks. Anger was also recognized comparatively better on this task than on facial or prosodic tasks. When anger was misperceived, it was most commonly perceived as disgust. When disgust was misperceived, it was most commonly perceived as fear, and then as unpleasant surprise. Subjects appeared to perceive the negative valence, but failed to place the stimulus into the correct narrow class. When fear was misperceived, it was most commonly perceived as unpleasant surprise.

Comparison of Emotion Recognition to Healthy Controls

A qualitative exploration of emotion recognition between HD subjects and HCs was performed. Percent correct of emotion by task was converted into a rank order table for HD subjects. Rank order of HCs emotion recognition was obtained by personal communication (J.C. Borod, March 28, 2008). Comparisons of these groups' rank order performance can be seen in Tables 33-36.

Table 33. Rank Order of Facial Emotion Recognition in Healthy Controls versus the Overall HD Group

Rank Order	Healthy Controls	HD Subjects
1	Happiness	Happiness
2	Disgust	Sadness
3	Fear	Anger
4	Pleasant surprise	Unpleasant surprise
5	Anger	Disgust
6	Sadness	Pleasant surprise
7	Unpleasant surprise	Fear
8	Interest	Interest

On the facial task, HCs and HD subjects both evidenced superior recognition of the emotion of happiness, and poor recognition of interest. Beyond this, however, their pattern of recognition for the other emotions was dissimilar from each other.

Table 34. Rank Order of Prosodic Emotion Recognition in Healthy Controls versus the Overall HD Group

Rank Order	Healthy Controls	HD Subjects
1	Anger	Sadness
2	Sadness	Anger
3	Fear	Interest
4	Interest	Pleasant surprise
5	Happiness	Unpleasant surprise
6	Pleasant surprise	Disgust
7	Unpleasant surprise	Fear and Happiness
8	Disgust	

On the prosodic task, HCs and HD subjects shared relatively better recognition of the emotions anger and sadness, and a modest recognition of interest. Both groups also did relatively poorly in recognizing disgust. The remainder of the emotions was recognized at varying levels of accuracy between groups.

Table 35. Rank Order of Lexical Emotion Recognition in Healthy Controls versus the Overall HD Group

Rank Order	Healthy Controls	HD Subjects
1	Sadness	Sadness
2	Pleasant surprise	Happiness
3	Unpleasant surprise	Anger
4	Happiness and Disgust	Pleasant surprise
5		Unpleasant surprise
6	Interest	Disgust
7	Anger	Fear
8	Fear	Interest

On the lexical task, both HCs and HD subjects recognized sadness most accurately. They shared poor recognition of fear and interest, and moderately poor recognition of disgust. There were no other similarities in the degree of recognition accuracy of various emotions between the HC and HD groups.

Table 36. Rank Order of Emotion Recognition Across Channels in Healthy Controls versus the Overall HD Group

Rank Order	Healthy Controls	HD Subjects
1	Sadness	Sadness
2	Anger	Happiness
3	Happiness	Anger
4	Fear	Pleasant surprise
5	Pleasant surprise	Unpleasant surprise
6	Disgust	Disgust
7	Unpleasant surprise	Fear
8	Interest	Interest

Averaged across tasks, HD subjects' performance more closely resembled that of their HC counterparts. Both groups showed a greater propensity for identification of sadness, anger, and happiness, and a relative difficulty identifying interest and to a lesser degree disgust. In contrast to HCs, HD subjects showed a clear-cut difficulty recognizing fear.

Aim 3: Caregiver Burden Measures

The relationship between the two caregiver burden measures, the HDQoL-C and the ZBI, was explored using Pearson product-moment correlations. The three subscale scores of the HDQoL-C (Aspects of Caring, Satisfaction with Life, and Feelings about Life) were also included in this analysis. On the HDQoL-C, higher scores indicate greater quality of life or less caregiver burden, whereas on the ZBI, higher scores indicate greater

levels of caregiver burden. See Table 37 for correlations among caregiver burden measures.

Table 37. Correlations Among Caregiver Burden Measures

	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
HDQoL Total	--	.78**	.67**	.90**	-.65**
HDQoL AC		--	.87**	.95**	-.82**
HDQoL LS			--	.91**	-.73**
HDQoL FL				--	-.79**
ZBI					--

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; AC = Aspects of Caring; SL = Satisfaction with Life; FL = Feelings about Life; ZBI = Zarit Burden Inventory.
**$p < .01$.

Each subscale of the HDQoL-C, as well as the overall score, was significantly inversely correlated with the ZBI, indicating that higher levels of burden on one scale corresponded positively with higher levels of burden on the other scale.

Aim 4: Emotional Perceptual Functioning and Caregiver Burden

The relationship between performance on the three emotional perception tasks, the three control tasks, and caregiver burden was explored using Pearson product-moment correlations. See Table 38 for correlations among caregiver burden measures and emotional and nonemotional measures of the New York Emotion Battery.

Table 38. Correlations Among Caregiver Burden Measures and Emotional and Nonemotional Measures of the New York Emotion Battery

	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
Emotional Tasks					
FID	-.09	.19	.13	.07	-.36
PID	-.17	.11	.16	.02	-.29
SID	.04	.31	.40	.26	-.49*
Control Tasks					
BFRT	-.03	.34	.25	.18	-.53*
ICP	.11	.25	.38	.26	-.51*
NESID	-.15	.03	.16	.00	-.36

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; AC = Aspects of Caring; SL = Satisfaction with Life; FL = Feelings about Life; ZBI = Zarit Burden Inventory; FID = Facial Identification Task; PID = Prosodic Identification Task; SID = Sentence Identification Task; BFRT = Benton Facial Recognition Task; ICP = Intonation Contours Task; NESID = Nonemotional Sentence Identification Task.
*$p < .05$.

There was a significant inverse correlation between the ZBI and nonemotional facial and prosodic recognition tasks and the emotional lexical perception task, meaning that greater impairments in general facial and prosodic perception, and in lexical emotional perception, were associated with higher levels of burden.

Aim 5: Psychiatric and Personality Functioning and Caregiver Burden

Psychiatric and personality functioning was assessed through administration of the PAI. Seven of the HD individuals were found to have responded invalidly to this measure and were removed from the analysis. One additional subject's data were not utilized in this analysis due to the fact that their caregiver failed to complete the study. The relationship between psychiatric and personality functioning and caregiver burden was explored through the use of a correlational analysis. Pearson product-moment correlations were utilized except where indicated. See Table 39 for correlations among caregiver burden measures and psychiatric and personality functioning measures.

Table 39. Correlations Among Caregiver Burden Measures and Psychiatric and Personality Functioning Measures

PAI Scales	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
Somatic Complaints	-.24	-.50	-.24	-.37	.74*
Anxiety	-.12	-.16	-.27	-.21	-.00
Anxiety-Related Dx	-.28	-.44	-.33	-.39	.51
Depression	-.51	-.32	-.38	-.47	.25
Mania	-.04	-.14	-.06	-.09	.48
Paranoia	-.41	.03	-.00	-.15	.02
Schizophrenia	-.06	-.28	-.17	-.19	.25
Borderline	-.65*	-.51	-.44	-.61†	.47
Antisocial	-.14	-.04	.16	-.01	.45
Alcohol Problems	-.57†	-.78**	-.54	-.71*	.54
Drug Problems	.19	-.14	-.29	-.09	.11
Aggression	-.87**	-.36	-.37	-.61†	.21
Suicidal Ideation^{S}	-.32	.11	-.42	-.42	.74*
Stress	-.46	-.24	-.23	-.35	.50
Nonsupport	-.35	-.28	-.51	-.45	.19
Treatment Rejection	.03	.18	.12	.12	-.38
Dominance	-.20	-.20	-.10	-.18	.32
Warmth	.08	-.00	-.10	-.01	.21

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; AC = Aspects of Caring; SL = Satisfaction with Life; FL = Feelings about Life; ZBI = Zarit Burden Inventory; PAI = Personality Assessment Inventory.
^{S}Spearman rho. $^{\dagger}p < .10$. $*p < .05$. $**p < .01$.

It was found that elevated somatic complaints and suicidal ideation were associated significantly with higher levels of reported burden on the Zarit Burden Inventory. Borderline features and aggression were significantly correlated with higher levels of burden on the overall HDQoL-C measure. The presence of alcohol problems was associated with lower Feelings about Life and Aspects of Caring scores. Trends toward significance were seen in the relationships between increased alcohol problems and the elevated burden on the HDQoL-C, and between borderline features and aggression, and the poorer Feelings about Life.

Aim 6: HD Subject, Caregiver Characteristics, and Caregiver Burden

HD Subject Characteristics

The relationship between HD subject characteristics and caregiver burden were explored using Pearson product-moment correlations. See Table 40 for correlations among caregiver burden measures and HD subject characteristics.

Table 40. Correlations Among Caregiver Burden Measures and HD Subject Characteristics

	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
Age	.19	.28	.42†	.32	-.29
Education	.19	.34	.14	.24	-.47†
Age of Sx Onset	.39	.46†	.50*	.49*	-.55*
Age of Diagnosis	.30	.38	.41	.39	-.43†
CAG Repeat Size	-.26	-.22	-.23	-.26	.31
Shoulson Total	.32	.43†	.35	.40	-.44†

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; AC = Aspects of Caring; SL = Satisfaction with Life; FL = Feelings about Life; ZBI = Zarit Burden Inventory. Sx = Symptom
†$p < .10$. *$p < .05$.

On the ZBI, older age of HD onset was associated significantly with decreased burden; trends toward significance were seen between greater HD subject education and decreased burden, older age of diagnosis and decreased burden, and greater total functional capacity and decreased burden. A trend toward significance was found in the association between older age of symptom onset and higher Aspects of Caring scores, and between higher total functional capacity and higher Aspects of Caring scores. A significant association was found between older age of HD symptom onset and greater Life Satisfaction, and a trend toward significance was found in the association between increasing caregiver age and Life Satisfaction. Finally, a significant relationship was found between age of symptom onset and healthier Feelings about Life.

Cognitive Profile of HD Subjects

The relationship between HD subjects' cognitive functioning and caregiver burden was explored in a correlational analysis. Pearson product-moment correlations were utilized except where indicated. See Table 41 for correlations among caregiver burden measures and HD subjects' cognitive profile.

Table 41. Correlations Among Caregiver Burden Measures and HD Subjects' Cognitive Profile

	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
BDAE Commands[S]	.08	-.05	.17	.21	-.08
BDAE Com. Id. Mat.[S]	-.26	-.23	-.16	-.31	.06
BDAE Sent & Par[S]	.24	.21	.16	.18	-.42
DRS Total	-.07	.25	.14	.10	-.17
DRS Attention[S]	.21	.15	.33	.12	-.22
DRS I/P[S]	.04	.03	.13	-.03	-.15
DRS Construction[S]	.31	.20	.41	.33	-.03
DRS Conceptualiz[S]	.01	-.22	.02	.21	-.25
DRS Memory[S]	.18	.18	.12	.08	.00
Premorbid FSIQ Est.[N]	.05	.14	.23	.15	-.29

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; AC = Aspects of Caring; SL = Satisfaction with Life; FL = Feelings about Life; ZBI = Zarit Burden Inventory; BDAE = Boston Diagnostic Aphasia Examination; DRS = Dementia Rating Scale; FSIQ = Full Scale Intelligence Quotient. [S]Spearman rho. [N]FSIQ based on the North American Adult Reading Test.

Estimated premorbid intellectual functioning, current comprehension ability, and level of current cognitive functioning were not significantly related to caregiver burden.

HD Subjects' Awareness

The relationship between HD patients' awareness functioning and caregiver burden were explored using Pearson product-moment correlations. See Table 42 for correlations among caregiver burden measures and HD subjects' awareness measures.

Table 42. Correlations Among Caregiver Burden Measures and HD Subjects' Awareness

	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
Subjective Experience	-.42	-.56*	-.44†	-.51*	.40
Subjective Consequence	.00	-.27	-.09	-.12	.10

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; AC = Aspects of Caring; SL = Satisfaction with Life; FL = Feelings about Life; ZBI = Zarit Burden Inventory.
†$p < .10$. *$p < .05$.

A significant correlation was found between higher subjective experience of motoric deficits and lower scores on the Aspects of Caring and Feelings about Life subscales. A trend toward significance was found in the association between higher subject experience of motoric deficits and lower Satisfaction with Life.

Caregiver Characteristics

The relationship between caregiver characteristics and caregiver burden was explored in a correlational analysis. Pearson product-moment correlations were utilized, except where indicated. See Table 43 for correlations among caregiver burden measures and caregiver characteristics.

Table 43. Correlations Among Caregiver Burden Measures and Caregiver Characteristics

	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
Caregiver Age	.21	.48†	.61*	.45†	-.34
Time Known HD	-.02	-.02	.10	.02	.06
Time Caring HD[S]	-.05	-.26	.11	.11	-.11
Income[S]	-.30	-.39	-.41	-.29	.03
Freq. Leisure Act.	.31	.24	.35	.33	-.10

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; AC = Aspects of Caring; SL = Satisfaction with Life; FL = Feelings about Life; ZBI = Zarit Burden Inventory.
[S]Spearman rho. †$p < .10$. *$p < .05$.

There was a significant relationship between older caregivers and higher Satisfaction with Life. A trend toward significance was found in the relationship between older caregivers and optimistic Feelings about Life, and between older caregivers and positive Aspects of Caring.

Relationships Between Dichotomous and Categorical Variables and Caregiver Burden

Some of the measures thought to be potentially related to elevated caregiver burden were either dichotomous or categorical in nature and were consequently not able to be explored using a correlational approach. Instead, one-way ANOVAs were carried out for each of these variables, separately for each caregiver burden measure (see Table 44; the dependent variables were the total score on the HDQoL-C and ZBI).

Table 44. One-way ANOVAs on Caregiver Burden

	HDQoL-C		ZBI	
	F	**p**	**F**	**p**
Patient Gender	1.74	.207	1.43	.250
Patient Occupation	1.62	.234	1.96	.164
Psychiatric Hx	2.41	.142	9.69	.007*
Neurological Hx	0.01	.935	0.00	1.00
Substance Abuse Hx	0.61	.448	0.88	.362
Antipsychotic Use	0.01	.922	0.02	.889
Antidepressant Use	1.95	.183	1.66	.217
Mood Stabilizer Use	1.62	.223	0.63	.440
Memory Medication Use	0.43	.521	0.42	.525
Shoulson Stage	1.86	.191	3.53	.057†
Demented	0.09	.773	0.10	.759
Caregiver Gender	1.46	.245	0.39	.544
Caregiver Relation	0.50	.489	0.19	.670
Cared for Other HD Family	0.02	.897	0.51	.486
Genetic Status of Children	2.36	.123	0.76	.537

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; ZBI = Zarit Burden Inventory.
†$p < .10$. *$p < .05$.

The Kruskal-Wallis ANOVA was carried out for several of the variables in which the normality assumption had been violated (see Table 45; the dependent variables were the total score on the HDQoL-C and ZBI).

Table 45. Kruskal-Wallis One-way ANOVAs on Caregiver Burden

	HDQoL-C		ZBI	
	χ^2	*p*	χ^2	*p*
Caregiver Education	3.17	.366	1.04	.792
Caregiver Disability Level	2.37	.500	0.11	.991
Hours Work/week	2.48	.480	3.01	.391
Hours Child Care/week	0.44	.801	0.08	.962
Hours Caring for HD/week	1.34	.719	0.08	.994

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; ZBI = Zarit Burden Inventory.

The only significant result found was that caregivers of HD patients with a positive psychiatric history reported significantly higher levels of caregiver burden (M = 41.67, SD = 14.66) than caregivers of HD patients without a positive psychiatric history (M = 21.13, SD = 12.23), F = 9.69 (1, 15), p = .007. A trend toward significance was seen in the relationship between Shoulson stage and caregiver burden as measured by the ZBI.

Supplementary Analyses

NYEB Interchannel Correlations

Pearson product-moment correlation coefficients were performed between the three experimental emotional tasks from the NYEB within the overall HD sample. See Table 46 for interchannel correlations among the NYEB emotion tasks for the overall HD group.

Table 46. Interchannel Correlations (and *p*-values) Among NYEB Emotional Tasks for the Overall HD Group

	FID	PID	SID
FID	--	.67	.61
		(.002)	(.007)
PID		--	.55
			(.019)
SID			--

FID = Facial Identification Task; PID = Prosodic Identification Task;
SID = Sentence Identification Task.

Within the overall HD group, significant correlations were found among all channels, with the correlation between the prosodic and facial channels being the strongest, followed by the correlation between the facial and lexical channel and the correlation between the prosodic and lexical channel. To test whether these relationships were statistically significant, Fisher's r-to-z transformations were performed. None of the resulting comparisons were significant: PID/FID versus SID/FID, $z = 0.35$, $p = .728$; PID/FID versus SID/PID, $z = 0.71$, $p = .476$; FID/SID versus SID/PID, $z = 0.37$, $p = .713$.

The following table presents the interchannel correlations on the three experimental emotional tasks from the NYEB within the nondemented HD subgroup. See Table 47 for interchannel correlations among the NYEB emotional tasks for the nondemented HD subgroup.

Table 47. Interchannel Correlations (and *p*-values) Among NYEB Emotional Tasks for the Nondemented HD Subgroup

	FID	PID	SID
FID	--	.60 (.120)	.17 (.687)
PID		--	-.28 (.502)
SID			--

FID = Facial Identification Task; PID = Prosodic Identification Task; SID = Sentence Identification Task.

Within the nondemented HD subgroup, none of the correlations were found to be significant. However, the pattern of relationships was similar to that seen in the overall HD sample, with the strongest positive relationship between the prosodic and facial channels, followed by a positive correlation between the facial and lexical channel and an inverse correlation between the prosodic and lexical channel. To test whether relationships among particular channels were statistically significant, Fisher's r-to-z

transformations were performed. The differences between the prosodic/facial and prosodic/lexical comparisons, $z = 3.08$, $p = .002$, and between the lexical/facial and lexical/prosodic comparisons, $z = 2.02$, $p = .043$, were significant. However, the comparison between the facial/prosodic and the lexical/facial channels was not significant, $z = 1.24$, $p = .214$.

CAG Repeat and Age of Onset

The relationship between number of CAG repeats and age of HD symptom onset was found to be significantly correlated, $r = -.72$, $p = .003$, indicating that a higher CAG repeat number is associated with earlier age of onset.

Disease Duration and NYEB

The relationships between disease duration and NYEB emotional perception measures and were explored using Pearson product-moment correlations. It was found that disease duration was unrelated to all three emotional channels of the NYEB. See Table 48 for interchannel correlations among the NYEB emotional tasks and disease duration.

Table 48. Correlations between Emotional Perception and Disease Duration

	Disease Duration
FID	-.44
PID	.24
SID	-.27

FID = Facial Identification Task;
PID = Prosodic Identification Task;
SID = Lexical Identification Task.

DISCUSSION

Emotional Functioning in HDs as Compared to HCs

The present study comprehensively examined emotional perception across facial, prosodic, and lexical channels in HC and HD subjects. Further, this was done using a broad and an individuated approach, and with the overall HD group and the nondemented subgroup. The broad to specific approach allowed for a clear understanding of the overarching patterns in the sample and then provided a context from which to interpret the more specific analyses. It allowed the opportunity to study the variability within the HD sample, the appropriateness of the comparison of the HC group to the experimental group, and any potential power issues that may have arisen.

The risk of performing such a large number of analyses is that every analysis will produce a different result, and that the interpretation is so complex that the beauty of the original research question is lost in the design. Fortunately, with this study, the opposite was true. In fact, the most exciting aspect of the findings was that, regardless of the approach or the HD comparison group, the pattern of the results remained consistent. For example, in the broad approach, the overall HD group was impaired on all emotional experimental tasks in comparison to the HCs; this exact pattern remained when considering only the group of nondemented HD individuals. To ensure that demographic factors did not alter the results of the broad analysis, individuated analyses customized by demographic for each variable were carried out. The overall HD group remained significantly impaired on all experimental emotional tasks. The nondemented individuals showed significant impairments on facial and prosodic emotional tasks. Nondemented

individuals were not significantly impaired on the lexical task, however, there was a trend toward impairment.

An obvious concern was the possibility that the HD subjects' emotional perceptual impairments were merely a consequence of general perception impairments. As such, the two groups were compared to each other on the nonemotional control task variables. Within the facial channel, general nonemotional facial perception impairments were found. The pattern was not as well-defined within the prosodic channel. In the broad analysis, the overall HD group was impaired, however, this effect weakened when examining the nondemented group. In the individuated approach, the overall HD group was impaired, but the effect did not occur for the nondemented group. This pattern of performance may indicate that intact nonemotional prosodic perception is dependent upon preserved intellectual functioning. As cognition declines, so does the ability to perceive linguistic prosody. Finally, within the lexical channel, HD individuals' performance was similar to that of HCs. The overall HD group did not show any significant differences in nonemotional lexical perception as compared to HCs in either the broad ($p = .730$) or individuated ($p = .732$) analyses. However, a trend toward significance, F [1, 109] = 3.70, $p = .057$, was seen in the difference between the nondemented HD individuals and the HCs in the broad analysis (HD: $M = 81.25$, $SD = 6.68$; HC: $M = 73.26$, $SD = 11.57$). Although nondemented HD subjects were not performing significantly worse than their HC counterparts in the individuated analysis, the similarity in performance was not as strong ($p = .169$) as that seen in the overall HD versus HC comparison. In summary, nondemented HD subjects had more difficulty on the nonemotional lexical control task than the overall group, which contained dementing

individuals. This pattern conflicts with reported initially preserved language function in the early stages of HD (Deus-Yela, Pujol, & Espert, 1997) and may indicate that the nondemented subgroup in the current sample is not representative of the nondemented HD population in terms of language functioning, although the deficit could be reflective of another impaired process such as executive functioning (e.g., reasoning).

Having elucidated HD subjects' performance on experimental and control tasks, the question that remained was: to what degree is the general perception impairment contributing to the pattern of deficits seen on the experimental emotional tasks? To address this question, the difference score comparisons were carried out. Within both the facial and lexical channels, relative impairments not only remained, but were robust and consistent across broad and individuated analyses in both the overall HD group and nondemented HD subgroup. In contrast, within the prosodic channel, no differences remained in broad or individuated analyses in the overall HD group or in the nondemented HD subgroup. This indicates that HD individuals are likely to have a true facial and lexical emotional impairment, but that their prosodic emotional functioning remains intact. From a neuroanatomical perspective, this pattern of functioning lends support for modality-specific processing. However, given the significant correlations among all three experimental emotional channels, at least in the overall HD group, it is likely that a general processor exists for all three communication channels, with separate channels processed secondarily and uniquely through separate modality systems (Borod et al., 1990). Proposed neural substrates of emotional processing include amygdala, orbitofrontal cortex, anterior cingulate, and posterior parietal cortex (Borod et al., 2000).

Other studies have found general perception deficits in their HD samples. None of the studies that found impairments, however, treated the general perception deficits as confounds or attempted to address them statistically through, for example, stratification or covarying. It seems that cognitive and other general perception tasks were often administered for the purpose of sample characterization rather than for the control of potential confounds in emotional processing, per se. Of note, in their discussion of limitations, many of the studies suggested that basic perceptual deficits may have confounded their findings for emotion perception functioning. With the exception of studies of presymptomatic individuals, most investigators have found their HD samples to be impaired on a wide range of screening instruments. Hayes et al.'s HD sample (2007; with a disease duration of 6.7 years), was impaired on phonemic fluency, symbol digit transcription, and a test of attention and response inhibition (i.e., Stroop test). The authors did not address these neuropsychological deficits in their study design (Hayes, Stevenson, & Coltheart, 2007). Milders et al.'s HD sample (2003; with a disease duration of 6.5 years) was impaired on vocabulary, semantic fluency, and unfamiliar face recognition (as measured by the BFRT). The mean score on the BFRT [$M = 40.2$], although lower than the one for the control sample [$M = 49.1$], was "on the border of the normal range" (which was similar to the score [$M = 39.8$] found in the current study) and as such, this factor was not addressed in the study design (Milders, Crawford, Lamb, & Simpson, 2003). Montagne et al. (2006) did not report disease duration, stating only that their subjects were early symptomatic and independent in their ADLs, with a mean MMSE score of 29.1 and a mean UHDRS score of 17.1. Their sample was found to perform well on the BFRT, and no other screening tests were administered (Montagne et

al., 2006). Sprenglemeyer et al.'s sample (1996; with a disease duration of 6.6 years) was not demented, although it is unclear how this was determined. Their sample was intact in their ability to identify young versus old, male versus female, and familiar versus unfamiliar faces. In contrast, HD subjects were impaired on tasks of gaze direction and unfamiliar faces (i.e., BFRT). They stated that HD subjects' BFRT performance was not only above chance but found to be at the low end of the low average range (i.e., 41.0) based on norms. They stated that the gaze direction impairment did not need to be addressed because performance was well above chance. Although the investigators admitted that the visual perceptual abnormalities found in their HD sample were severe, they did not attempt to control for these deficits in the study design. They further reasoned that the pattern of disproportionately affected emotions was not consistent with a generalized cognitive deterioration (Sprengelmeyer, Schroeder, Young, & Epplen, 2006). Finally, Wang et al.'s HD subjects (2003; with a disease duration of 4.3 years) were independent in ADLs, but were no longer working. Subjects showed intact performance on gender and age perception, gaze direction, and identification of familiar faces, but were impaired on semantic fluency, visual form discrimination, line orientation, and identification of unfamiliar faces (i.e., BFRT). The authors reasoned that because disgust was not the most difficult emotion to recognize by normal control group, this differential impairment in the HD group could not be attributed to a general visuospatial functioning deficit (Wang et al., 2003).

In comparison to the existing studies, the HD subjects in the current study had longer disease durations (overall group, 8.9 years; nondemented subgroup, 8 years). However, their performance on the BFRT was similar to performances reported in the

literature; overall group, 49.83; nondemented subgroup, 43.50. The overall group's performance is considered Borderline, but the nondemented subgroup's performance is considered to be in the Average range. Similar reasoning could have been used; although HD subject performance was impaired compared to the normative group on the BFRT, it was still considered to be within the normal range based on Benton's normative data. However, the fact remains that HD subjects were performing differently, and in fact, worse than the healthy controls. It was felt that the possibility that general perception functioning might impact performance on the emotional tasks could not be discounted; the only way to address that was to test for it. So in this respect, one of the current study's special strengths, as compared to the extant literature, is that it is the only one to address general cognitive and perceptual impairments in the study design.

In the HD emotion literature, screening and control tasks were administered to assess various cognitive functions and abilities. In most of the studies, tasks were administered with the goal of assessing general neuropsychological functioning. In rarer cases, the control task appears to have been selected to complement the domain of the experimental task in question (e.g., BFRT as a control for facial emotion). In our study, screening tasks were administered to assess possession of basic intellectual and language skills required to complete the experimental tasks. Additionally, perception functioning was systematically assessed for each of the three emotional (i.e., facial, prosodic, and lexical) channels.

Although HD subjects demonstrated general facial perception deficits, their emotional impairments were robust enough to endure after taking the former into account. In the lexical domain, HD subjects did not demonstrate nonemotional lexical

deficits, so their emotional lexical impairments persisted. Within the prosodic channel, a slightly different pattern emerged when considering the control task and the difference score. In this case, the overall HD group was impaired on the control task, whereas the nondemented groups' performance was intact. Regardless, both groups failed to remain impaired on emotional prosody after accounting for the control task. This may be an artifact, however, of the Prosodic Identification Task's level of difficulty. A visual inspection of the summary data for healthy controls across experimental task reveals that percent correct for the facial and lexical tasks is roughly 15 to 20 points higher than that of the prosodic task. Further, the variability of the prosodic task is 30-50% higher relative to the facial and lexical tasks, resulting in significantly greater error or "noise". Therefore, the effect was not robust enough, secondary to task difficulty, to withstand comparison to a control task. Another possibility is that the ICP task was not the ideal measure to serve as a control, and consequently factored out the wrong information. Alternative tasks, for example, might include perception of musical tones, linguistic accents, or pitch range and duration. And, of course, one additional possible explanation is that HD subjects are just not impaired on emotional prosody, per se, relative to HCs.

One could argue that it makes little sense to consider the results of analyses involving the overall HD group, because it includes some individuals whose performance was invalidated secondary to impaired cognition. On the contrary, inspection of both the overall HD group and the nondemented HD subgroup provides some clues as to the relationship between cognitive decline and emotional perceptual functioning. For example, the overall HD group was significantly impaired on the lexical emotional task relative to the HC group, whereas the nondemented HD subgroup showed a

(nonsignificant) trend toward poorer performance on lexical emotional perception, indicating that emotional impairment in the lexical domain may not occur until cognitive decline is fully underway. Early in the disease process, the neurodegeneration remains focally in the basal ganglia. As the disease progresses, degeneration involves the bilateral frontal lobes, and ultimately results in more widespread degeneration and atrophy. Relatively preserved lexical emotional functioning in HD is consistent with other language functions which are robust to cognitive decline, such as expressive vocabulary. In contrast, both the overall HD group and the nondemented subgroup were significantly impaired in comparison to HCs on the facial emotional task. This suggests that impairment in the facial domain precedes significant cognitive impairment and may begin to decline even presymptomatically. Inspection of the lexical emotional summary data reveals that mean percent correct values increase from the overall HD group to the nondemented subgroup. This example highlights the importance of considering both the significance of the test results, as well as the pattern of the data.

The nondemented HD subgroup's performance is of particular interest. The crux of the primary research question is whether there are differences in the two groups in terms of impairments. Therefore, consideration of a sample that is not dementing allows for the removal of a known confound in order to have a clearer look at the true deficits.

HD Subjects' Emotional Perception Functioning by Individual Emotions

Consistent with the HD emotion literature, the current study has unequivocally demonstrated that HD subjects are impaired in emotional perception functioning, and facial emotional perception, in particular. Beyond a channel perspective, the literature has revealed that HD individuals demonstrate differential impairments in identifying specific

emotions, the most commonly cited being disgust, and to a lesser degree, anger. The studies which have looked at presymptomatic individuals have shown exclusive disgust deficits (Gray, Young, Barker, Curtis, & Gibson, 1997; Sprengelmeyer, Schroeder, Young, & Epplen, 2006), indicating that emotional perception impairment may begin with deficits in disgust. The studies of persons with symptomatic HD show deficits ranging principally from anger and disgust (Hayes, Stevenson, & Coltheart, 2007; Montagne et al., 2006; Sprengelmeyer et al., 1997) to deficits across many emotions (Milders, Crawford, Lamb, & Simpson, 2003; Sprengelmeyer et al., 1996; Wang, Hoosain, Yang, Meng, & Wang, 2003). Consistently, though, happiness and sadness, and to a lesser degree surprise, are preserved.

To investigate the literature's reports of emotion-specific processing deficits, the present study sought to delineate the pattern of emotion performance among the HD subjects. Individual consideration was given to each of the three channels, as well to overall emotion recognition collapsed across the three channels. The objective of this emotion analysis is distinct from the channel analysis in the preceding section, in that the focus is not to detect impairments on the tasks, per se. Rather, the goal is to observe how HD affects an individual's emotion perception, irrespective of disease stage. Therefore, the demented individuals were retained in the sample and the overall HD group was utilized.

In consideration of emotion recognition across tasks, it is helpful to visualize the results for all tasks for both groups simultaneously (see Table 49).

Table 49. Rank Order of Emotion Recognition across Tasks and Groups

Huntington's Disease Subjects					Healthy Control Subjects				
Rank Order	**FID**	**PID**	**SID**	**Overall**	**Rank Order**	**FID**	**PID**	**SID**	**Overall**
1	H	S	S	S	1	H	A	S	S
2	S	A	H	H	2	D	S	PS	A
3	A	I	A	A	3	F	F	US	H
4	US	PS	PS	PS	4	PS	I	H; D	F
5	D	US	US	US	5	A	H		PS
6	PS	D	D	D	6	S	PS	I	D
7	F	F; H	F	F	7	US	US	A	US
8	I		I	I	8	I	D	F	I

A = Anger; D = Disgust; F = Fear; H = Happy; I = Interest; PS = Pleasant Surprise; S = Sadness; US = Unpleasant Surprise; FID = Facial Identification Task; PID = Prosodic Identification Task; SID = Lexical Identification Task; Overall = mean score of the 3 experimental emotion tasks.

There are several noteworthy patterns in regard to emotion recognition in the HD group when revisiting the literature reviewed in the Introduction, especially with respect to disgust and fear (highlighted in gray in Table 49 for each emotion task). Disgust is relatively poorly recognized across tasks by the HD group, though less so on the facial recognition task. HCs, in contrast, show superior facial disgust recognition, moderate lexical disgust recognition, and poor prosodic disgust recognition. Finally, fear and interest are the most poorly recognized emotions in the HD group. Fear recognition by HCs is relatively better in the facial and prosodic channels and relatively worse in the lexical channel. HD subjects are consistently better at recognizing happiness, sadness, and anger across tasks; the only exception to this is poor happiness recognition on the prosodic task. In contrast, HCs are superior at recognizing facial happiness, but are only moderately good at recognizing prosodic and lexical happiness. Finally, a striking difference between groups is that HD subjects tend to be consistent in their ability to recognize emotions across channels, whereas HCs' ability varies from channel to channel.

When comparing the patterns of emotion recognition within this current HD sample to those in the literature, there are two interesting observations. First, the finding that fear is more impaired than disgust supports Milder et al.'s (2003) findings of worse fear versus disgust recognition and contradicts Sprenglemeyer et al.'s (1996, 1997) and Gray et al.'s (1997) findings of worse disgust versus fear recognition. Additionally, fear appeared to be the only emotion impaired when studies involved presymptomatic individuals. Milders et al. calls attention to the fact that if Gray et al.'s (1997) data were presented as mean percent correct scores (instead of percentages of individuals achieving a particular score), recognition of fear actually happens to be poorer than recognition of disgust. Therefore, there seems to be an interesting debate developing regarding fear versus disgust. Differences found across tasks could be related to task type or difficulty, although the same exact facial tasks were utilized in Milders' (2003), Sprenglemeyer's (1996, 1997), and Gray's (1997) studies. Another possibility for fear versus disgust deficit differences is variability across samples in neuroanatomical degeneration. Prominent fear impairments might be reflective of a pattern of neurodegeneration distinct to that found when disgust impairments are more prominent. Similar to the way that symptom presentation can vary widely between individuals with HD, so too may the pattern of neurodegeneration. Both the basal ganglia and the amygdala are structures known to degenerate in HD. It is possible, then, that patients who have greater degeneration in the amygdala region relative to those with relatively greater degeneration in the basal ganglia would have relatively more severe fear than disgust deficits (Adolphs et al., 1994; Sprenglemeyer et al., 1997). The disgust deficits may be more prevalent as the basal ganglia are member structures of all five of the frontal-subcortical circuits

proposed to be affected in HD, whereas the amygdala is a member structure of only the anterior cingulate and the lateral orbitofrontal circuits. Further inspection of the relationship between neurodegeneration and clinical symptoms may reveal that individuals who have relatively more deficits in fear recognition also have relatively more apathy, aggression, personality changes, and emotional dysfunction versus those with more prominent disgust deficits may have more motor and cognitive impairments.

Milders et al. notes that their normal control group performed similarly (poorly) on fear recognition, which they reasoned was related to the nature of fear being a more difficult emotion to recognize, in general. However, relatively good fear recognition in the facial and prosodic tasks in this study's healthy controls sample argues against this.

A second notable comparison between this study's sample and the literature is that the unique pattern of performance on the prosodic task in the sample is virtually identical to Sprenglemeyer et al.'s (2006) reports. In that study, no significant differences were found between the HD group and the normal control group on sadness and anger, with significantly poorer performance by the HD group for happiness, surprise, fear, and disgust. The pattern in this study's sample is identical, except that normal controls also did poorly at recognizing disgust.

After having considered the pattern of emotion recognition within the HD sample, the pattern within the HD group was compared to that of the HC group. In the HC group, the only consistency in emotion recognition across tasks was inconsistency; HCs exhibit a different pattern of emotion recognition within each channel. This is in contrast to the HD group's emotion recognition performance, which is relatively stable across channels. If, however, the overall pattern of emotion recognition in the HCs is compared to that of

the HD subjects, sadness, anger, and happiness share the top three rankings for both groups, disgust shares ranking 6, and interest shares the last rank. However, making comparisons to the overall standings within the HC group is deceiving given that it is a composite of disparate patterns in each of the channels. The order of emotion recognition by normal controls in Sprengelmeyer et al's. (1996) study is happy, sad, disgust, surprise, anger, and fear for the facial emotion hexagon; and happy, sad, surprise, anger, disgust, and fear on the facial emotional identification task. Ekman and Friesen's normal controls are reported to recognize happy best, then disgust, surprise, sad, fear, and anger. This study's healthy control population did not appear to perform similarly to the data reported in the literature.

One final comment should be made regarding the rank order comparisons: although access to the rank order of emotion recognition of healthy controls provides a context for interpretation of the normality or lack thereof in the pattern of emotion recognition in HDs, caution should be made in any interpretation as rank order information can be misleading. To make this case, the mock data in Table 50 are considered.

Table 50. Mock Rank Order and Percent Correct Data

Group 1		Group 2	
Rank	**Percent**	**Rank**	**Percent**
Anger	98	Anger	95
Disgust	97	Sad	32
Sad	40	Disgust	31

Based on only rank order information, it would appear that that sadness recognition is poorer in group 1 than sadness recognition in group 2. However, attention to the actual percent correct data indicates otherwise.

Factors Associated with Elevated Caregiver Burden in HD Caregivers

Higher levels of caregiver burden on the overall HDQoL-C and each of the subscales were found to be strongly associated with higher levels of caregiver burden on the ZBI. Given that the ZBI is one of the most widely used caregiver burden instruments, the HDQoL-C's relationship to it implies good construct validity. In the results section, correlations that were found to be either significant or a trend toward significance were mentioned. However, given that the nature of the investigation of factors relating to caregiver burden is exploratory, a more liberal viewpoint was warranted. Therefore, in addition to the significant findings summarized in the results section, correlations considered at least moderate (i.e., .40 and above; Cohen, 1988) were reassessed within the overall pattern of correlations. To emphasize this point, the tables containing the correlational analyses are reproduced below, with any correlation above .40 highlighted n gray. Note that only those variables where significant correlations were found, as well as where moderate but nonsignificant correlations are newly being considered are included below; all other variables were removed from the tables for the sake of simplicity.

The HDQoL-C's subscales (i.e., Satisfaction with Life, Feelings about Life, and Aspects of Care) were included in the correlational analyses to provide a more comprehensive understanding regarding the caregiving issues specific to HD. The Satisfaction with Life subscale provides insight into a caregiver's overall quality of life issues and personal issues. The Feelings about Life subscale elicits information about a caregiver's positive and negative feelings about living with HD. The Aspects of Care subscale provides information about the practical aspects of caring for an individual with

HD, such as levels of support, access to professionals, long-term and genetic issues, and daily hassles.

A moderate correlation was found between lexical emotional perception and the Satisfaction with Life. On the ZBI, greater burden was associated with nonemotional facial and prosodic and emotional lexical impairments. Similarly, communication difficulties in stroke survivors were found to be associated with lower caregiver quality of life (Watson et al., 1998; White et al., 2003). See Truncated Table 38 for trends toward significance and significant relationships among caregiver burden measures and emotional and nonemotional perception measures.

Truncated Table 38. Trends and Significant Relationships Among Caregiver Burden Measures and Measures of the New York Emotion Battery

	HDQoL SL	ZBI
SID	.40	-.49*
BFRT	.25	-.53*
ICP	.38	-.51*

SID = Lexical Identification Task; BFRT = Benton Facial Recognition Task; ICP = Intonation Contours Perception Task.
*$p < .05$.

Inspection of the PAI scales after including moderate correlations provides a more exhaustive look at the personality and psychiatric variables that may contribute to increased caregiver burden. It becomes evident, for example, that alcohol problems and borderline characteristics are consistently related to caregiver burden across all caregiver burden measures. Additionally, it was discovered that the increased burden as measured by the ZBI is related to six additional variables: increased HD subject stress, increased alcohol problems, antisocial characteristics, borderline characteristics, mania, and anxiety-related disorders. It appeared initially that only more "positive psychiatric symptoms" were associated with increased caregiver burden, which is indeed confirmed

by the significant relationship between a positive psychiatric history and elevated caregiver burden. However, consideration of moderate relationships reveals that depression in the HD individual can also negatively impact caregivers.

Personality and psychiatric functioning directly impacts a person's behavior and in turn, their ability to function successfully in their relationships. The general caregiver burden literature clearly states that the presence of behavioral disturbances in the patient can lead to increased caregiver burden (Coen et al., 2007) and declining mental and physical health of the caregiver (Hooker et al., 2002). Specifically, anger and aggression were found to be predictors of caregiver role overload (Bedard et al., 1997). More specificially, the HD literature replicates the findings that impaired mood and behavior are the most distressing to caregivers (Hayden et al., 1980; LoGiudice & Hassett, 2005; Semple, 1995; Tyler et al., 1983). See Truncated Tables 39 and 44 for trends toward significance and significant relationships among caregiver burden measures and psychiatric and personality functioning measures.

Truncated Table 39. Trends and Significant Relationships Among Caregiver Burden Measures and Psychiatric and Personality Functioning Measures

PAI Scales	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
Somatic Complaints	-.24	-.50	-.24	-.37	.74*
Anxiety-Related Dx	-.28	-.44	-.33	-.39	.51
Depression	-.51	-.32	-.38	-.47	.25
Paranoia	-.41	.03	-.00	-.15	.02
Borderline	-.65*	-.51	-.44	-.61†	.47
Antisocial	-.14	-.04	.16	-.01	.45
Alcohol Problems	-.57†	-.78**	-.54	-.71*	.54
Aggression	-.87**	-.36	-.37	-.61†	.21
Suicidal Ideation^{S}	-.32	.11	-.42	-.42	.74*
Stress	-.46	-.24	-.23	-.35	.50
Nonsupport	-.35	-.28	-.51	-.45	.19

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; AC = Aspects of Caring; SL = Satisfaction with Life; FL = Feelings about Life; ZBI = Zarit Burden Inventory; Dx = Diagnosis.
^{S}Spearman rho. $^{\dagger}p < .10$. $*p < .05$. $**p < .01$.

Truncated Table 44. Psychiatric Functioning and Caregiver Burden

	ZBI	
	F	**p**
Psychiatric Hx	9.69	.007*

ZBI = Zarit Burden Inventory.
*$p < .05$

Several characteristics of the HD patients have been found to be related to increased caregiver burden. Given the relatively exploratory nature of the application of the HDQoL-C in general, and more specifically to the relationship between the two burden measures, additional consideration of trends and moderate correlations was found to be helpful in looking at patterns of significant relationships. Reevaluation of HD subject characteristics reveals that age of diagnosis is related not just to the ZBI, but also to the HDQoL-C subscales. A similar pattern is seen between the Shoulson total and caregiver burden. Evidence to support these findings comes from a study which reported a significant association between lower self-care abilities and caregiver burden (Watson et al., 1998). See Truncated Table 40 for trends toward significance and significant relationships among caregiver burden measures and HD subject characteristics.

Truncated Table 40. Trends and Significant Relationships Among Caregiver Burden Measures and HD Subject Characteristics

	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
Age	.28	.42†	.32	-.29
Education	.34	.14	.24	-.47†
Age of Sx Onset	.46†	.50*	.49*	-.55*
Age of Diagnosis	.38	.41	.39	-.43†
Shoulson Total	.43†	.35	.40	-.44†

Sx = Symptom.
†$p < .10$. *$p < .05$.

Moderate relationships are seen between language comprehension ability of the HD individual and caregiver burden; poorer comprehension is associated with increased

caregiver burden. This finding supports the general theme of impaired communication (in this case, comprehension) in contributing to increased caregiver burden (Watson et al., 1998; White et al., 2003). See Truncated Table 41 for trends toward significance and significant relationships among caregiver burden measures and HD subjects' cognitive profile.

Truncated Table 41. Trends and Significant Relationships Among Caregiver Burden Measures and HD Subjects' Cognitive Profile

	HDQoL SL	ZBI
BDAE Sent & Par[S]	.16	-.42
DRS Construction[S]	.41	-.03

[S]Spearman rho.

Including moderate correlations clarifies the notion that intact subjective experience is globally related to caregiver burden. It was expected that a decreased awareness of subjective awareness of deficits would lead to increased grief in the caregiver. However, the opposite was found to be true. Rather, the more aware the HD subject is of their deficit, the more distressing this is for the caregiver. There are not any known studies that exist to corroborate or dispute this finding. See Truncated Table 42 for trends toward significance and significant relationships among caregiver burden measures and HD subjects' awareness.

Truncated Table 42: Trends and Significant Relationships Among Caregiver Burden Measures and HD Subjects' Awareness

	HDQoL Tot.	HDQoL AC	HDQoL SL	HDQoL FL	ZBI
Subjective Experience	-.42	-.56*	-.44†	-.51*	.40

†$p < .10$. *$p < .05$.

In the current study, a significant relationship was found between caregiver age and increased burden, specifically burden related to practical Aspects of Caring,

Satisfaction with Life, and Feelings about living with HD. This finding replicates findings in disabled elderly, stroke, and hip and knee replacement literature, which found significant inverse relationships between caregiver age and caregiver burden (Watson et al., 1998; Llacer et al., 2002). Lower income is moderately correlated with lower Life Satisfaction, and to a lesser degree, Aspects of Caring. Income was also reported in the literature have a negative impact on caregivers (Hans & Gilmore, 1968; McGarva, 2001). See Truncated Table 43 for trends toward significance and significant relationships among caregiver burden measures and caregiver characteristics.

Truncated Table 43. Trends and Significant Relationships Among Caregiver Burden Measures and Caregiver Characteristics

	HDQoL AC	HDQoL SL	HDQoL FL
Caregiver Age	.48†	.61*	.45†
Income[S]	-.39	-.41	-.29

[S]Spearman rho. †$p < .10$. *$p < .05$.

Other factors that were found in the HD caregiving literature to be associated with increased burden that were not found in the current study were caring for more than one HD family member and worry of transmission of HD to children. The current sample was not variable enough to provide accurate answers to these questions.

Relationships Among Communication Channels and Disease Characteristics

NYEB Interchannel Correlations

Regarding communication channels, previous research has demonstrated (Borod et al., 1998, 2000) that particular channels are more related to each other than to others. In particular, the relationship between facial and prosodic nonverbal channels has been reported as positive and significant, whereas the relationship between nonverbal and verbal channels is typically not significant (e.g., Borod, Koff, Lorch, & Nicholas, 1985;

Borod et al., 1990, 2000). This provides evidence of a general processor for the identification of emotional stimuli across the three communication channels, with facial, prosodic, and lexical information secondarily processed through separate sensory modality systems (Borod et al., 2000). Proposed neural substrates involved in the processing of emotional stimuli include amygdala, orbitofrontal cortex, anterior cingulate, and posterior parietal cortex (Borod et al., 2000).

In the HD subjects in this study, this same pattern was seen. Within the overall HD group, the relationship between the facial and prosodic nonverbal channels was the strongest, and indeed positive and significant. The relationship between the nonverbal and verbal channels, although significant, was weaker than the relationship between the facial and prosodic nonverbal channels. The nondemented HD subgroup showed the same pattern, albeit at a level which did not reach the conventional level of significance. Within the nondemented HD subgroup, the relationship between the facial and prosodic nonverbal channels was positive, but not significant. It was the strongest of the three relationships, however, in terms of magnitude. The relationships between the nonverbal and verbal channels were weaker. In particular, the relationship between the facial and lexical channels was positive and the relationship between the prosodic and the lexical channels was actually negative, albeit low in magnitude. The relationship between facial and prosodic perception was significantly stronger than the relationship between lexical and prosodic perception, but not significantly stronger than the relationship between lexical and facial perception. The relationship between lexical and facial perception was significantly stronger than the relationship between lexical and prosodic perception. This trend of a positive association between face and voice and relatively poorer associations

between the lexical and nonverbal channels is consistent with studies of individuals with brain damage (Borod et al., 1985).

The Relationship Between CAG Repeat Size and Age of Onset

The current study's finding that a greater number of CAG repeats was associated with an earlier age of onset of Huntington's disease symptoms is consistent with the literature (The U.S.-Venezuela Collaborative Research Project & Wexler, 2004). That this significant effect was seen despite the limited sample size (i.e., n = 14) and the fairly limited age range and CAG repeat lengths of the subjects, speaks to the strength of this effect. The following table (Table 51) illustrates the genetic data:

Table 51. The Relationship Between Age of Onset and CAG Repeat Size

Age of Onset	Repeat size
29	51
30	42
31	47
32	48
32	49
35	47
38	43
39	45
41	47
45	43
47	42
47	45
50	44
65	39

The Relationship Between Disease Duration and Emotional Perception

Ostensibly, it would seem logical that the further along in the HD disease process, the more impaired emotional perception becomes. However, disease duration was unrelated to all three emotional channels. Upon further consideration, with minor exception, when emotional perception impairments were present in the overall HD group,

the same pattern was seen in the nondemented subgroup. This consistency across level of cognitive decline evidences that the onset of emotional deficits occurs well before a more general decline in cognition, and quite possibly predates other signs and symptoms of the disease by years. As such, it is not surprising that a significant correlation between disease duration and emotional perception functioning was not established in the current study. Perhaps a relationship would become apparent if presymptomatic individuals were included in the sample range.

Demographic Considerations

The individuals with HD who participated in this study were similar to the majority of subject samples reported in the literature in terms of age. However, the disease duration of this study's HD group was approximately two to two and half years further into the disease process (overall group versus nondemented subgroup) than those symptomatic subjects reported in the literature. In a disease, whose progression from onset until death spans only 15 to 20 years, two years is a rather large portion of time, during which a significant amount of decline in function can occur. The HD subjects in this study had a relatively higher education level (i.e., overall group, 14.2; nondemented group, 14.5) than those subjects from other studies: Hayes et al., 11.8; Milders et al., 11.6; Sprenglemeyer et al. (1996), 9.0; Sprenglemeyer et al. (2006), 11.3; and Wang et al., 8.5. The HD subjects in this study were comparable (i.e., overall group, 98.6; nondemented group, 105.0) in reported estimated IQs of those subjects from the literature: Milders, et al., 105.8; Sprenglemeyer et al. (1996), 105.6; and Sprenglemeyer et al. (2006), 113.2. The current HD sample differs from most of the studies in terms of ethnicity and in some cases, dominant language: Sprenglemeyer et al.'s subjects are

German; Speedie et al.'s and Jacobs et al.'s subjects are American; Wang et al.'s subjects are Chinese; Milder et al.'s and Gray et al.'s subjects are English; Hayes et al.'s subjects are Australian; and Montagne et al.'s subjects are Dutch.

Although the HD individuals were similar demographically to HD individuals in existing studies in terms of age and intellectual functioning, they differed somewhat in disease duration, education level, ethnicity, and language. Despite existing differences, it was nice to see an overall emotional perception impairment, with consistency across individual emotional recognition performance. Additionally, that this pattern is consistent within a limited number of studies is encouraging to the validity of the current and existing findings and the potential for replication.

In terms of demographics comparison to the HC sample, the HD subjects were quite similar; the mean age of the HD group was similar to that of the HCs. The HD group had equal number of males and females, and their education split was also fairly equivalent. The only factor they didn't vary on was ethnicity. Given the demographic effects found in the HC population utilized in this study, a close demographics matching of groups was crucial to lend credence to any potential significant results. It could be argued that basing the individuated analyses between HD subjects and HCs on demographics that were found to be significant only within the HC population would obviate detection of a distinct pattern occurring in the HD population or in an interaction between the two, and consequently potentially significant results in analyses between the two groups. For this reason, the more closely matched the groups are on demographics, the less likely this scenario could impact the results. In the current study, five of the nine variables (i.e., emotional task scores, nonemotional task scores, and

nonemotional/emotional difference scores) in the primary analyses were examined with respect to at least one demographic characteristic, three variables were examined with respect to at least three demographic dimensions; and only one variable was not examined with respect to any demographic characteristic.

Conclusions and Recommendations for Future Research

The unique aspect of this study is the use of an emotion battery that simultaneously examines facial, prosodic, and lexical perception in HD subjects and healthy controls. Whereas other studies tested selective channels, this study examines all three. This is particularly important in light of the relative dearth of studies investigating prosodic emotional perception (only four such studies exist) in HD and the virtual lack of research on lexical emotional perception (only one such study exists) in HD. In addition, the emotion tasks have analogous psychometric properties (e.g., structure, item difficulty, and administration procedures). Nonemotional control tasks were selected a priori for each channel and task based on the belief that these factors that may impact a subject's performance on the emotional tasks. The presence of these three tasks makes this study unique among the existing research. Although the sample size was small, deficits in emotion perception have proved to be robust and can be established even in small sample sizes. Further the use of a carefully constructed emotion battery, including multiple modalities and control tasks, the power of the statistical design is increased (Borod et al., 1998).

In this study, HD subjects exhibited emotional perception deficits across all three emotional channels. After accounting for nonemotional perceptual functioning, HD subjects remained impaired on the facial and lexical channels, irrespective of dementia

status of HD subjects or of demographic class of the HC. In some cases, the effect was dampened, but not eliminated. Better understanding of nonverbal communication is hoped to allow development of therapies aimed at improving emotional health in persons with Huntington's disease and to improve communication patterns between Huntington's disease patients and their caregivers.

Among their own cohort, HD subjects performed relatively better on some emotions than on others. HD subjects tended to recognize happiness the best, and to be differentially impaired in disgust and fear recognition. The results are largely consistent across channels, with the exception of the prosodic channel, wherein happiness is perceived poorly. Better understanding of nonverbal communication is hoped to allow development of therapies aimed at improving emotional health in persons with Huntington's disease and improve communication patterns between Huntington's disease patients and their caregivers. Specifically, therapeutic goals may include increasing awareness and identification of a partner's emotions and increasing the ability to express thoughts and feelings related to the disease-related stressors.

In terms of caregiving, this study has served to provide valuable information about the significant positive association between a widely used existing, but nonspecific, caregiver burden measure and the newly devised HD-specific caregiver burden measure. Through utilization of these two measures, various characteristics were found to be associated with increased burden. These were aggression, anger, behavioral disturbances, impaired cognition, impaired mood, lower self care abilities, younger caregiver age, financial strain, and limited social support. The knowledge gained will be of benefit to others in the future. Better understanding of the experience of caregivers of HD patients

is hoped to allow us to provide psychoeducation regarding impact of caring on mental and physical health as well as provide appropriate services for these caregivers. Further, more widespread application of the HDQoL-C may be indicated clinically as a way to obtain a baseline of caregiver burden, and to assess success of treatments and interventions.

There are several means by which future studies could be improved over the current study. One suggestion is to decrease the number of emotions to be included in the study in an effort to decrease the level of difficulty of the tasks. It appeared that when HD subjects were unclear about which emotion to select, they chose interest, and to a lesser degree pleasant and unpleasant surprise, as a default.

The PID task tends to be differentially more difficult a task. This is evident not only in the performance of the HD subjects, but also in that of the healthy controls; both groups performed most poorly on this channel as compared to the lexical and facial channels. In addition to emotional prosodic perception impairments, HD subjects were also significantly more impaired than HCs in nonemotional prosodic perception. When accounting for the linguistic prosody impairments, however, the HD subjects did not show a greater discrepancy in performance between the experimental and control tasks than the HCs. This indicates that HD subjects did not have a selective deficit in emotional prosodic processing, rather, they had deficits in both emotional and nonemotional prosodic perception relative to HCs. However, it is possible that the failure of the emotional prosody deficits in HD subjects to remain significant after accounting for the control task is due to PID being a disproportionately difficult task. Perhaps the items that

comprise this measure could be adjusted to better match the level of difficulty of the lexical and facial tasks.

Reconsideration might be given to the nonemotional control tasks selected, in particular within the prosody channel. In the current study, the task selected to represent nonemotional prosodic functioning assessed intonational stress (i.e., declarative, interrogative, or exclamatory). One possibility is that assessing intonational prosody is simply not sufficient in attempting to control for nonemotional perceptual factors; alternative factors include pitch, rhythm, and/or timbre. To some extent, there is no perfect nonemotional control task, rather selection is a matter of what appears most appropriate. However, perhaps reconsideration of the options available would be appropriate for future studies.

While the time involved was not prohibitive, it appeared that some potential subjects decided not to participate due to the time commitment involved. The measure that contributed greatest to lengthening the overall time of administration was the PAI. In this particular sample, this measure was invalidated due to the cognitive impairment of a large number of the participants, rendering its administration useless. Should future studies utilize subjects as late as Shoulson Stage 3, the elimination of the PAI is strongly suggested. A preferred approach would be to recruit only subjects who are in Shoulson Stage 1 and the early part of Stage 2 (i.e., subjects with a Shoulson TFC Scale score of greater than or equal to 9). Subjects in late Stage 2 are beginning to decline cognitively, and subjects in Stage 3 are invariably demented. Earlier staged individuals would be more cognitively intact, and consequently more tolerant of the length of the testing and

more resilient to the demands of the PAI (which has the potential to provide extremely useful information).

Obtaining imaging data would be extremely interesting so as to be able to correlate emotional perception impairments with neurodegeneration. Further, the use of the multichannel approach (i.e., facial, prosodic, and lexical perception) in conjunction with imaging would provide a uniquely comprehensive look at these emotion-brain relationships. Application of imaging would also help delineate the issue of a double dissociation between disgust and anger.

In general, a larger sample size is recommended. Generally speaking, the true problem of having a small sample size is the concern about whether or not the subjects sampled are representative of the larger population. It is less related to the sample size itself or to the statistical significance that can be reached. Certainly a smaller sample results in decreased power and increased likelihood of failing to find significance. However, in the current study, robust significance was found in the face of these obstacles, testifying to the strength of the emotional impairment effect. However, the small sample size was more problematic in respect to the caregiver analysis. There were moderate to high value correlations that did not reach statistical significance, implying that this piece of the experiment was underpowered. The use of one-tailed tests, when particular predictions were made, would have bolstered the frequency of significant correlations.

The current study included a number of individuals who had significant psychiatric, alcohol, substance abuse, and neurological histories. A rational recommendation would be that future studies exclude persons with such complicated

histories. However, the nature of HD is such that these problems are often inherent in the disease and exclusions of these individuals would too severely limit the number of individuals participating. Additionally, inclusion of only individuals without complicated histories would really not be a representative sample. It is unclear if the decision to include these individuals in the current study is consistent with what other researchers have done. One study reported mild depression in two subjects and another study reported that eight out of twelve individuals were taking psychotropics, however, the remainder failed to mention this issue. Given the nature of the disease, it is suspected that these studies did not exclude HD subjects on these bases. In this vein, doing so in future studies may not necessarily be realistic.

A final recommendation for future studies is that healthy control raw data be utilized in lieu of healthy control summary data. This would provide more a greater degree of flexibility in the statistical approaches utilized. Further, it would allow for a decrease in the number of comparisons made, thereby increasing the strength of the study.

Overall, this study's similarities and discrepancies with previous findings provide ample questions for the future study of emotional perception in HD and a caregiver's experience.

Appendix A: Phone Screen Narrative

This is Denise Krch. I am one of the investigators of the HD emotion study at UMDNJ… I'm calling to talk to you about participation in the study. I will probably need about 15 minutes of your time. Is now convenient?

This study requires that both you and your caregiver (you and the HD person you care for) must participate as a pair. One may not participate without the other. You probably know that this study consists of simple paper and pencil tasks. I'd like to explain the study in more detail to you, answer any questions you have, and then, with your permission, collect some screening information from you over the phone. You should know up-front, that if you do not meet the required criteria **at any point during the study** – during our phone screen or during the in-person testing, the information collected from you will be destroyed.

First, some information about the study…
[If speaking to HD Patient]: At the in-person appointment, you will be asked to give your consent to participate. After consenting, the first few tasks given are screening tasks that cannot be performed over the phone because they require you to be present. If you qualify, you will participate in an interview, complete two questionnaires and some paper-and-pencil tasks. I will be asking you to read some words, draw simple shapes… You will look at pictures, read sentences, or listen to tape recordings and make simple judgments about what you see or hear. Finally, I will ask you to sign a release form giving us permission to obtain medical information from your neurologist. The total time to finish all tasks will be about 3 hours. You are welcome to take breaks during testing and if you need to stop testing, a second session can be scheduled to finish the tests. Regardless of whether you complete just the screening tasks or all of the tasks, you will be reimbursed for travel and parking. Do you have any questions about any part of the study?
[If speaking to caregiver]: At the in-person appointment, you will be asked to give your consent to participate. After consenting, you will be asked to fill out two questionnaires regarding your experience as a caregiver. These questionnaires take about 1 hour to complete. Do you have any questions about any part of the study?

[After all questions answered to participants' satisfaction:]
Ok, let me explain why I will be asking you questions before we meet personally. The pre-screening is used to see if you meet preliminary criteria to participate in the study. If not, this will save time and effort for both of us. It's okay to decline answering any screening questions you don't feel comfortable answering. After the phone interview, I will let you know if you qualify for the study, and, if you are interested in participating, we can set up an appointment for you to come in. **Is it okay to go ahead and ask you the questions?** [If so, administer phone screening]

If you think of any questions after we hang up, please feel free to call me back. My phone number is 732.235.5992.

Appendix B: Phone Screen

Subject Name:_____

Phone: _____

Contacted:_____

Scheduled:_____

Between ages of 18 and 80?_____

LANGUAGE:
Are you a native English speaker? YES NO

In not, spoke English by what age?_____

Other Languages Spoken: _____ Fluent? _____

NEUROLOGICAL HISTORY:
Do you have a history of a learning disability? YES NO
Was it ever diagnosed by a professional? YES NO
Have you ever had a serious problem with reading, writing, spelling, or arithmetic? YES NO

Describe...._____

Have you ever hit your head? YES NO When? _____

Describe:_____

Lost consciousness? YES NO When? _____

Describe: (how long was LOC?)_____

Been in a car accident? YES NO When? _____

Describe:_____

Have you ever been hospitalized for any reason? YES NO

Why?_____

Do you have a history of any of the following neurological problems?

Congenital Abhealthy controlities	YES NO	Alzheimer's Disease	YES NO
Head Injury	YES NO	Pick's Disease	YES NO
Dementia	YES NO	Parkinson's disease	YES NO
Korsakoff's Syndrome	YES NO	Creutzfeld-Jacob Disease	YES NO
Epilepsy	YES NO	Healthy control Pressure Hydrocephalus	YES NO
Pseudobulbar Palsy	YES NO	Multiple Sclerosis	YES NO
Subcortical Motor Disease	YES NO	Arteriosclerosis	YES NO
Schilder's Disease	YES NO	CVA	YES NO
Wilson's Disease	YES NO	Other:	YES NO
Seizure disorder	YES NO		

Had you ever been to see a neurologist before your diagnosis of HD? YES NO

This phone screen was adapted from the New York Emotion Battery (Borod et al., 1992) phone screen for use with HD subjects and their caregivers.

DRUG AND ALCOHOL USE:

Have you ever used recreational drugs (e.g. marijuana, speed, cocaine, crack, angel dust, heroine)? YES NO

If yes, what and for how long?_____

Has drug use ever interfered with your daily functioning? YES NO

How often do you drink alcohol?_____

How much per day/week?_____

What kind? _____

Has alcohol ever interfered with your daily activities? YES NO

Have you ever had a blackout while drinking? YES NO

Have you ever had a hangover after drinking? YES NO

SENSORY SYSTEM FUNCTIONING:

How is your hearing?_____

What is your visual capacity? _____

Do you wear glasses or contacts? YES NO

If yes, for how long have you had them? _____

Near-sighted? Far sighted?_____

HUNTINGTON'S SPECIFIC QUESTIONS:

At what age did you first notice onset of symptoms?

What symptoms were noticed at onset?

At what age did you seek medical consultation? (i.e. How long after onset of symptoms)

At what age were you diagnosed with HD?

How were you diagnosed (e.g. neurological exam, gene testing, established family history)?

Briefly describe some of your current symptoms (e.g. motor, cognitive, psychiatric…)

Have you experienced any difficulties with speech since your diagnosis? YES NO

This phone screen was adapted from the New York Emotion Battery (Borod et al., 1992) phone screen for use with HD subjects and their caregivers.

Are you aware of what stage of the illness you are in (i.e. Shoulson 1-5)?
Shoulson Functional Capacity Scale (Shoulson & Fahn, 1979)

Occupation:	Finances	ADL*:
0-Unable	0-Unable	0-Total Care
1-Marginal work only	1-Major assistance	1-Gross tasks only
2-Reduced capacity for usual job	2-Slight assistance	2-Minimal impairment
3-Healthy control	3-Healthy control	3-Healthy control

Domestic Chores:	Care Level:	Functional Information obtained from:
0-Unable	0-Full time skilled nurse	1-Patient only
1-Impaired	1-Home or chronic care	2-Patient and family/companion
2-Healthy control	2-Home	

Total Functional Capacity: _____
11-13 – **Stage 1** 7-10 – **Stage 2** 3-6 – **Stage 3** 1-2 – **Stage 4** 0 – **Stage 5**
* self-care (feeding, bathing, dressing, grooming), work, homemaking, and leisure.

PREMORBID PSYCHOLOGICAL FUNCTIONING:

Have you ever experienced any type of visual or auditory hallucination prior to or since your diagnosis (including medication-related)? YES NO

Have you ever been to see any kind of counselor, social worker, psychologist, psychiatrist? YES NO
Please describe (When, duration, reason)_____

_____

Would you consider yourself currently depressed? YES NO
Does your mood ever interfere with your daily functioning?_____

_____

FAMILY HISTORY:

Which of your family members have HD? Have they been formally diagnosed?

_____

_____

Caregiver Screening

Caregiver identified (e.g. spouse, sibling, child):_____

Phone: _____

Contacted:_____

Scheduled:_____

Between ages of 18 and 80?_____
Are you diagnosed with HD? YES NO
Are you at-risk for HD? YES NO
Native English speaker? YES NO
If not, spoke English by what age?_____
Other Languages Spoken: _____ Fluent? _____

This phone screen was adapted from the New York Emotion Battery (Borod et al., 1992) phone screen for use with HD subjects and their caregivers.

Appendix C: HD In-Person Interview

ID #: _____

Ethnicity:_____

Age:_____

Gender: _____

EDUCATION:

What is the highest level of education that you obtained?

Grade School: 1-6	College: 13-16	Major: _____
Junior High: 7-9	Masters: 17-18	Major: _____
High School: 10-12	M.D./Ph.D.: up to 20	Area: _____

OCCUPATIONAL HISTORY:

What is/was your occupational title? _____

What are/were your job duties? _____

_____

What other jobs/careers have you held?

CAREER	# Years Employed	Primary?
1. _____	_____	_____
2. _____	_____	_____
3. _____	_____	_____

Hollingshead Rating: _____

MEDICAL AND PSYCHOPHARMACOLOGICAL HISTORY:

Are you aware of any medical problems that you were born with? YES NO

Do you have allergies? YES NO

What prescription medications are you taking?

Name	**Reason**	**Duration**
_____	_____	_____
_____	_____	_____
_____	_____	_____
_____	_____	_____

Have you ever taken medications like tranquilizers, antidepressants, sleeping pills, or stimulants? (valium, halcion, prozac, elavil, xanax, ativan, dexedrine, librium, haldol) YES NO

Which:_____

This interview was adapted from the New York Emotion Battery (Borod et al., 1992) interview for use with HD subjects and their caregivers.

Are you under the care of a doctor/neurologist? YES NO

Name:_____

Phone #: _____

SENSORY SYSTEM FUNCTIONING:

Do you have any problems with your sense of smell? YES NO

If yes, please explain_____

HANDEDNESS:

Do you consider yourself:

1	2	3
Left Handed	Ambidextrous	Right Handed

In childhood or as an adult, were you ever forced to switch your handedness? YES NO

In which hand would you:	**Left**	**Amb.**	**Right**
…Throw a ball to hit a target?	1	2	3
…Draw?	1	2	3
…Use an eraser?	1	2	3
…Remove the top card when dealing?	1	2	3

FOOT AND EYE USE:

(FOOT)	**Left**	**Amb.**	**Right**
With which foot do you kick a ball?	1	2	3
…Pick up a pebble with your toes?	1	2	3
…Step up onto a chair with first?	1	2	3
…Stamp out a cigarette?	1	2	3
(EYE)	**Left**	**Amb.**	**Right**
Which eye would you use to peep through a keyhole?	1	2	3
…Look into a dark bottle to see how full it was?	1	2	3
…To sight down a rifle?	1	2	3
…Look through a telescope?	1	2	3

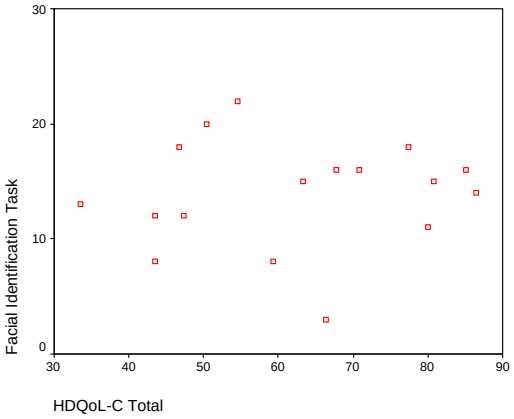
Are (or were) either of your parents or any of your full siblings Left handed or Amb.? YES NO

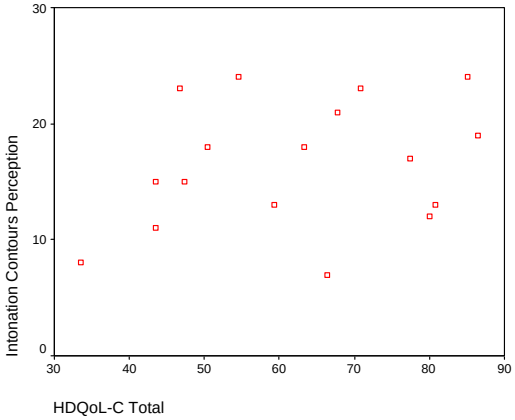
If so who? _____ How many full siblings are LEFT or AMB.?
_____ How many full siblings do you have?
_____

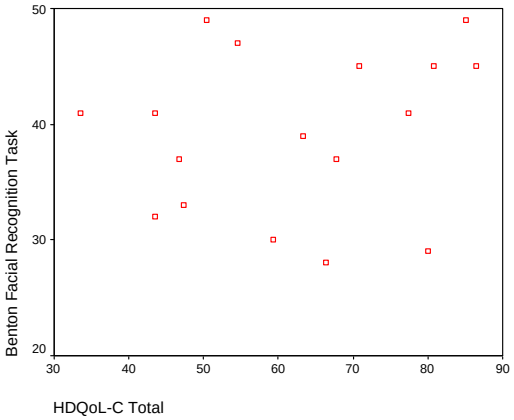
This interview was adapted from the New York Emotion Battery (Borod et al., 1992) interview for use with HD subjects and their caregivers.

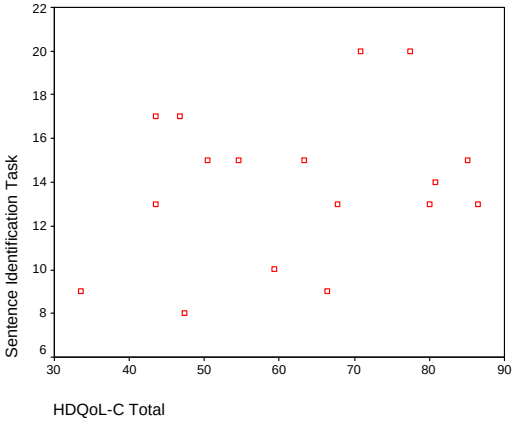
Appendix D: Scatterplots
Huntington's Disease Quality of Life for Carers

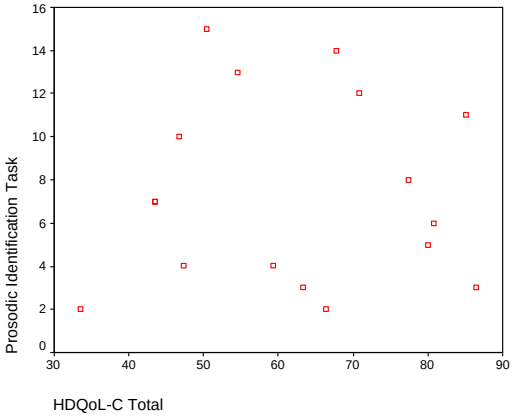
New York Emotion Battery

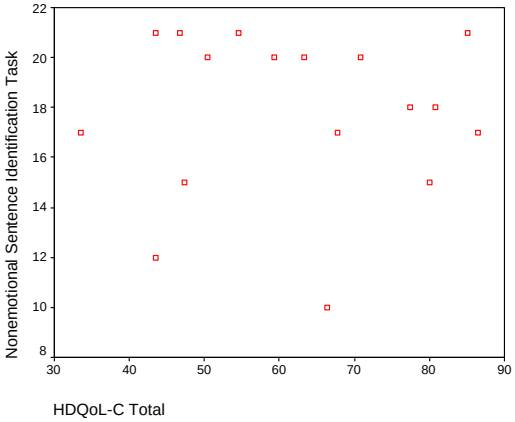




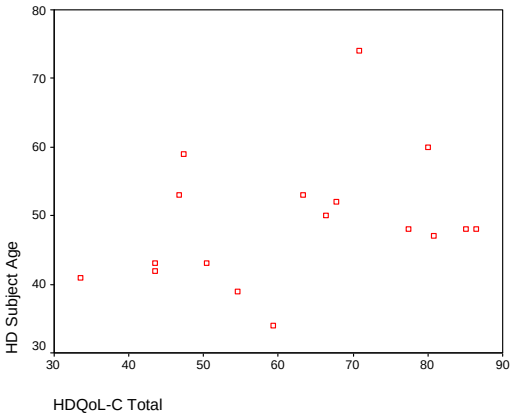


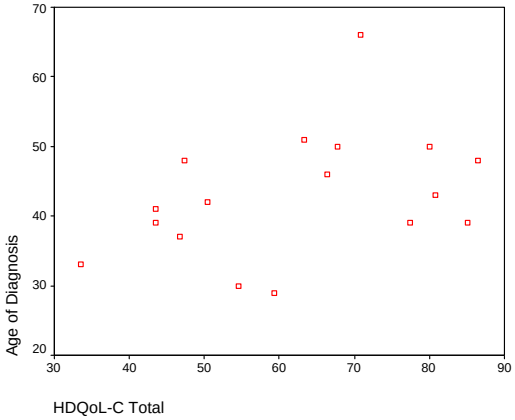


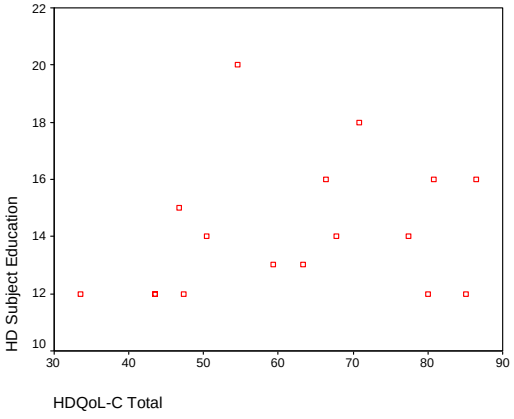


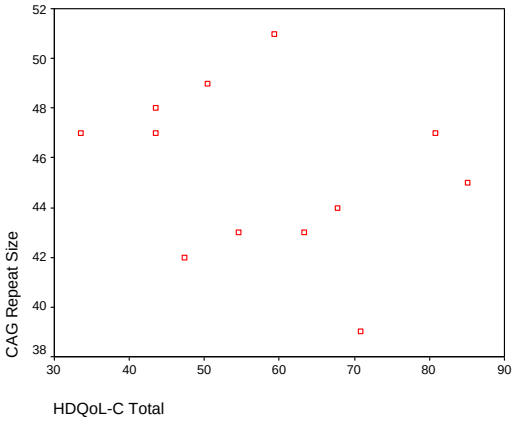


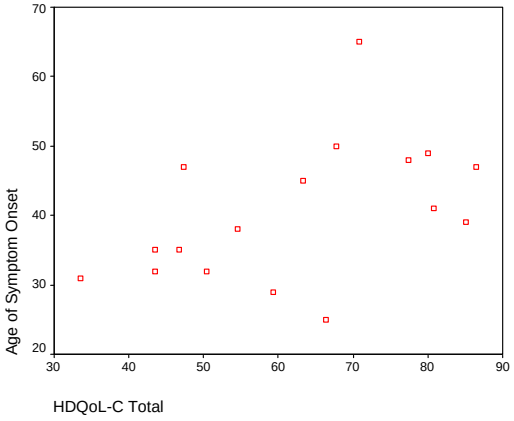
HD Subject Characteristics

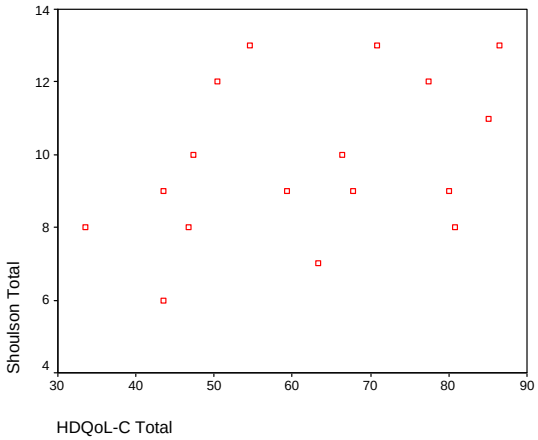




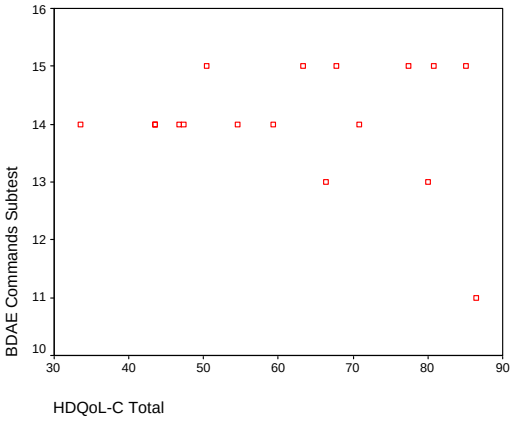


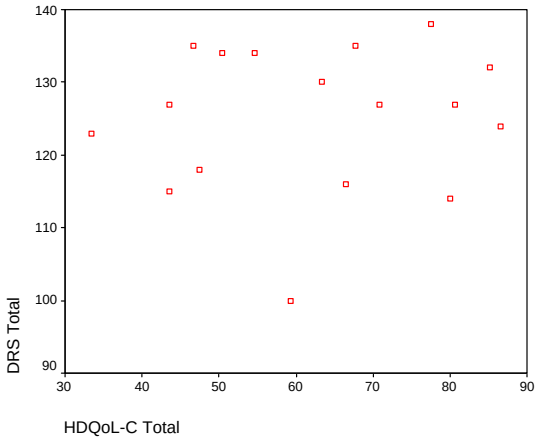


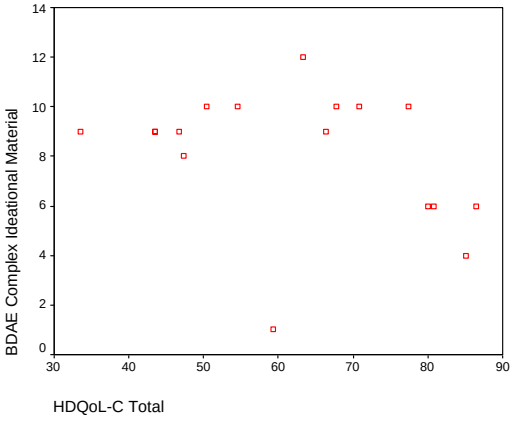


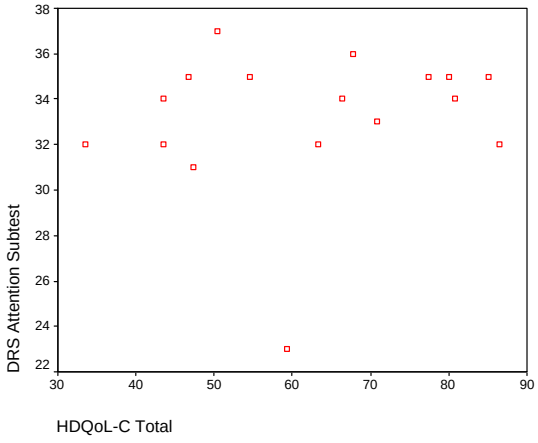


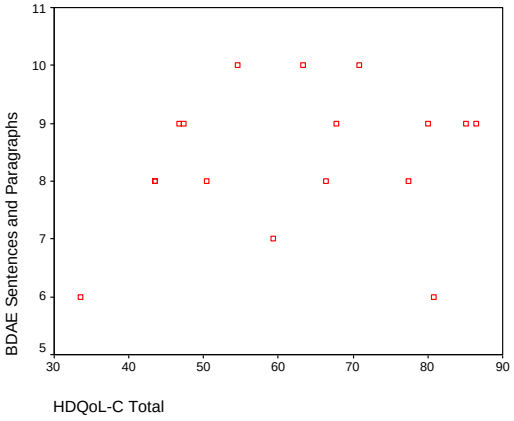
Cognitive Profile of HD subjects

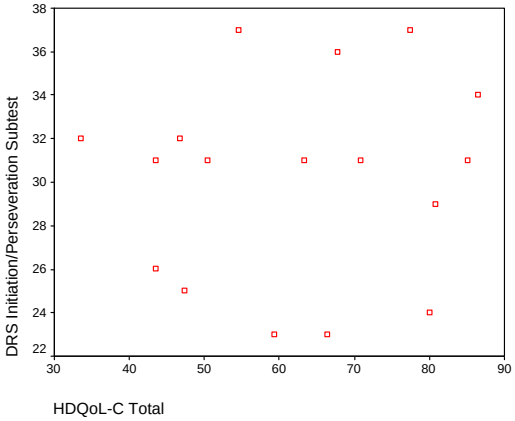


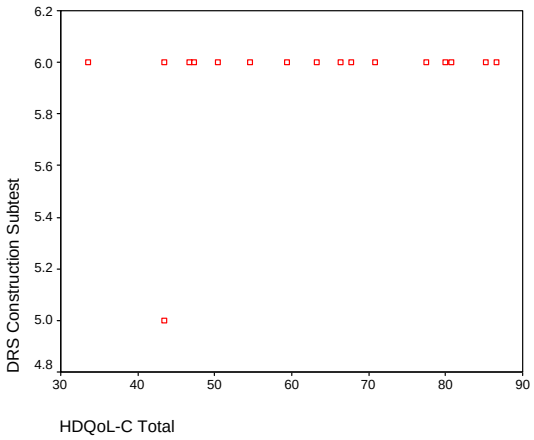


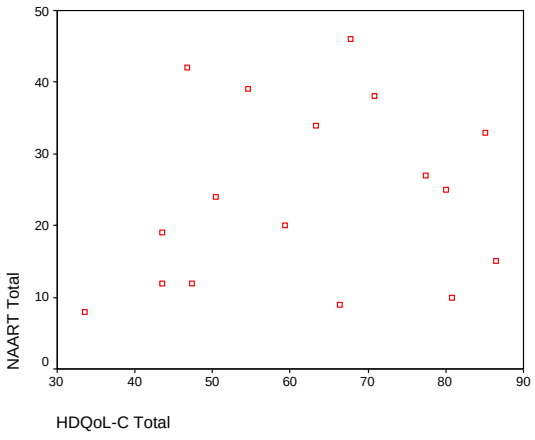


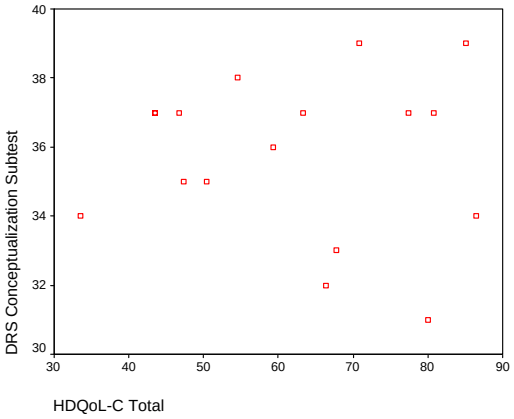


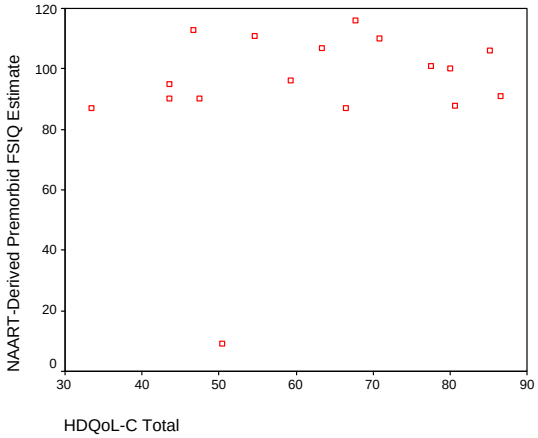


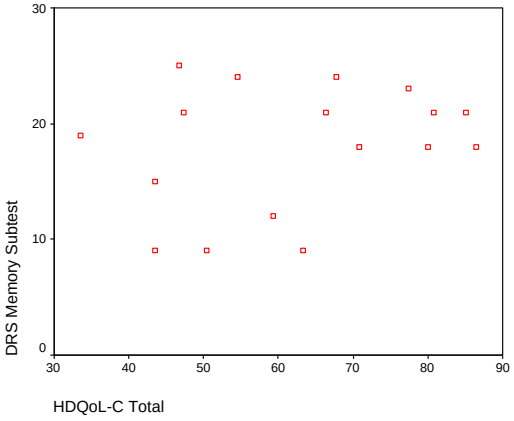



DRS Construction Subtest
6.2
6.0
5.8
5.6
5.4
5.2
5.0
4.8
30
40
50
60
70
80
90
HDQoL-C Total

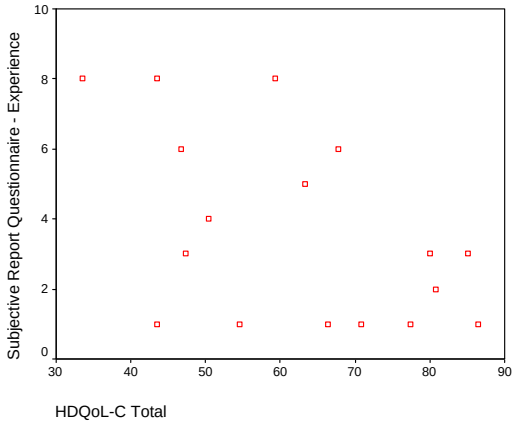

NAART Total
50
40
30
20
10
0
30
40
50
60
70
80
90
HDQoL-C Total

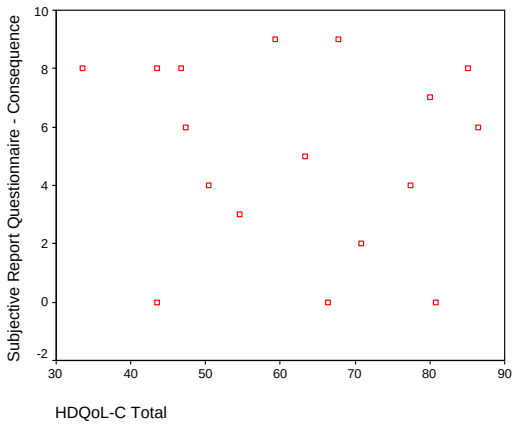

DRS Conceptualization Subtest
40
38
36
34
32
30
30
40
50
60
70
80
90
HDQoL-C Total


NAART-Derived Premorbid FSIQ Estimate
120
100
80
60
40
20
0
30
40
50
60
70
80
90
HDQoL-C Total

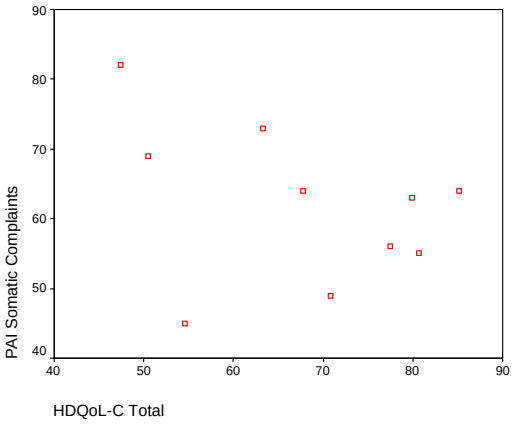

DRS Memory Subtest
30
20
10
0
30
40
50
60
70
80
90
HDQoL-C Total

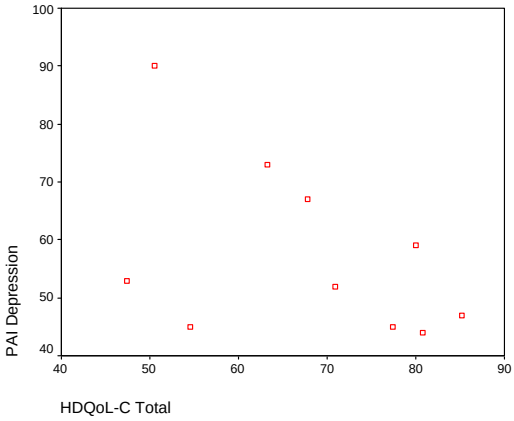
HD Subjects' Awareness

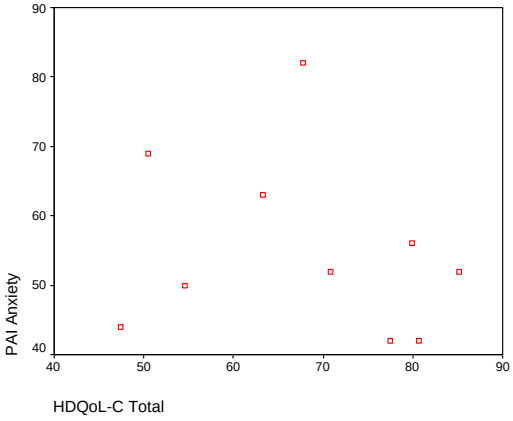


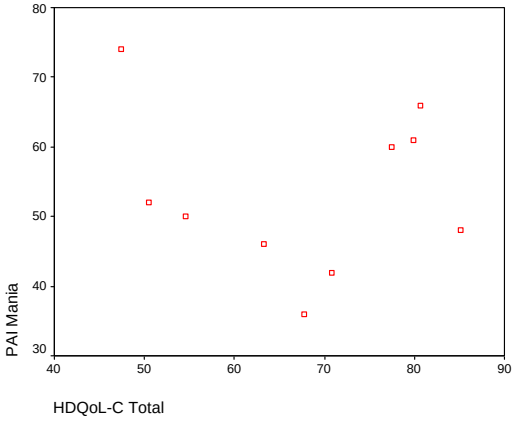


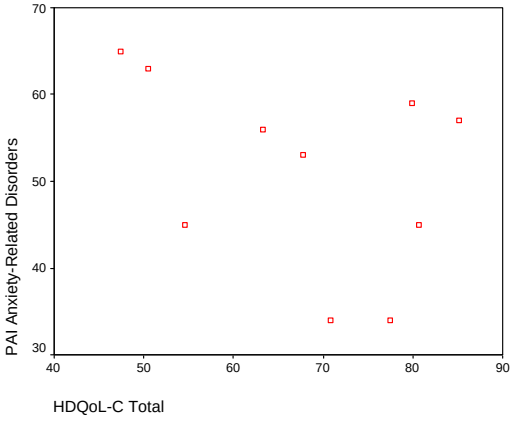
Psychiatric and Personality Functioning

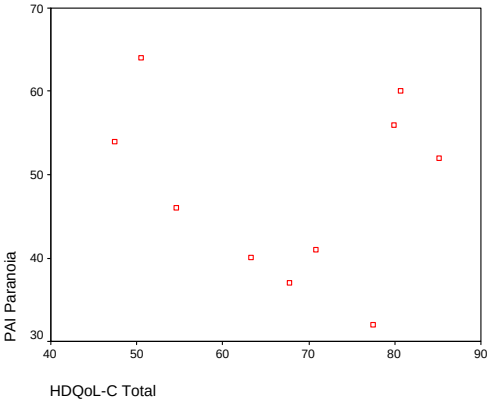


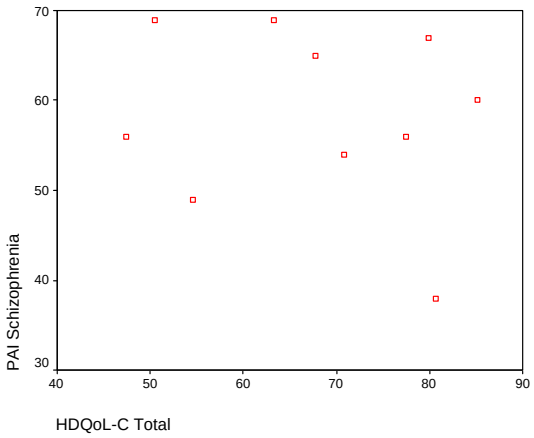


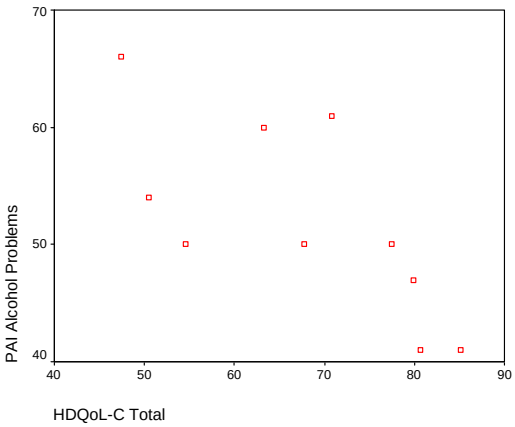


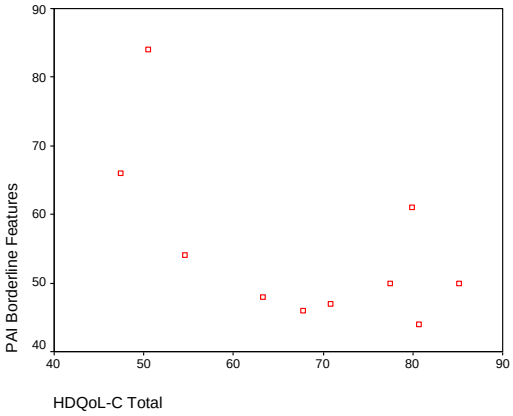


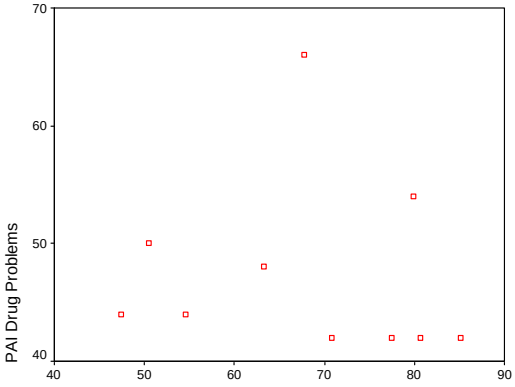


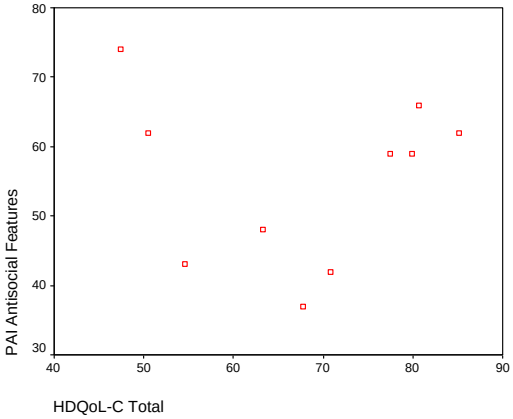


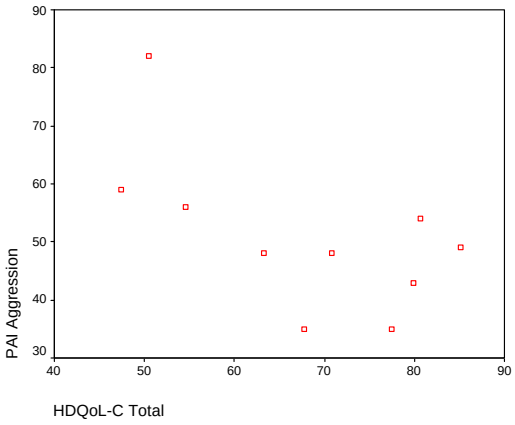

PAI Schizophrenia
70
60
50
40
30
40
50
60
70
80
90
HDQoL-C Total

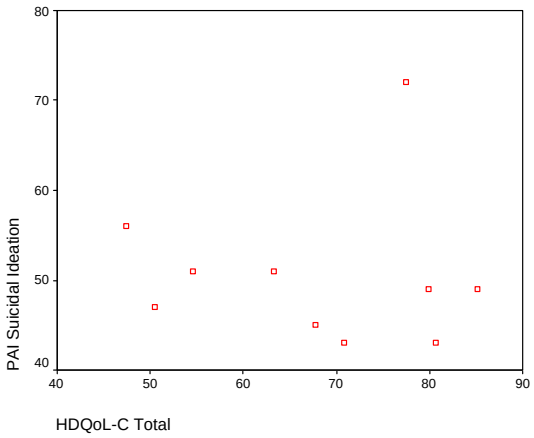

PAI Alcohol Problems
70
60
50
40
40
50
60
70
80
90
HDQoL-C Total

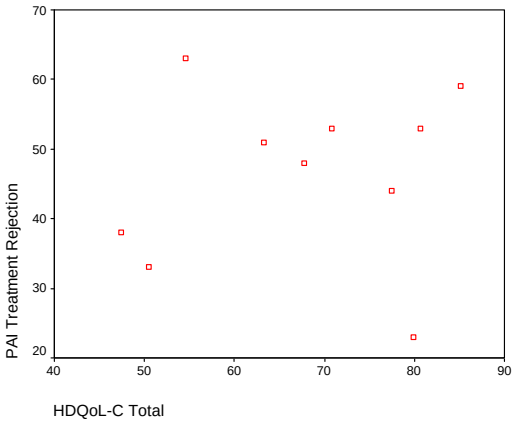

PAI Borderline Features
90
80
70
60
50
40
40
50
60
70
80
90
HDQoL-C Total

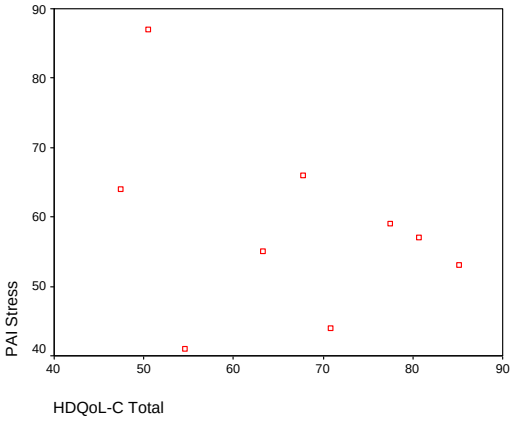

PAI Drug Problems
70
60
50
40
40
50
60
70
80
90
HDQoL-C Total

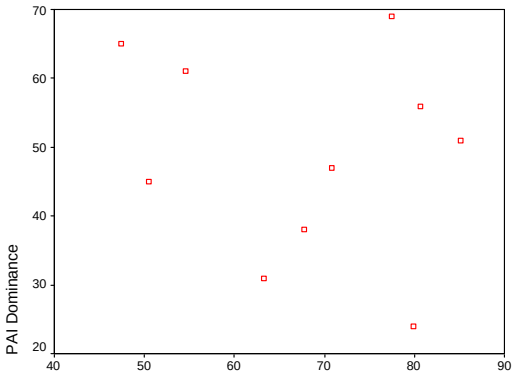

PAI Antisocial Features
80
70
60
50
40
30
40
50
60
70
80
90
HDQoL-C Total

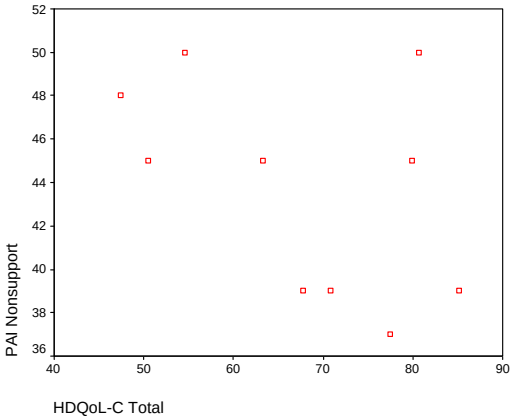

PAI Aggression
90
80
70
60
50
40
30
40
50
60
70
80
90
HDQoL-C Total

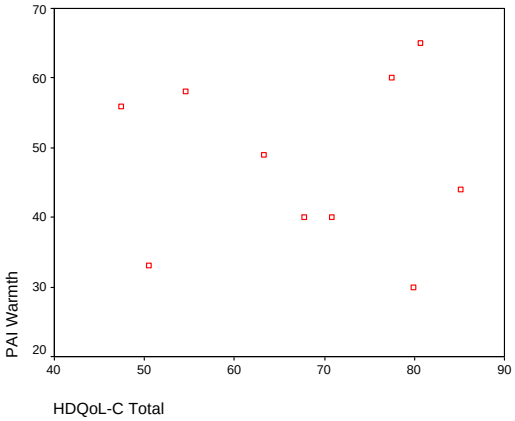

PAI Suicidal Ideation
80
70
60
50
40
40
50
60
70
80
90
HDQoL-C Total


PAI Treatment Rejection
70
60
50
40
30
20
40
50
60
70
80
90
HDQoL-C Total

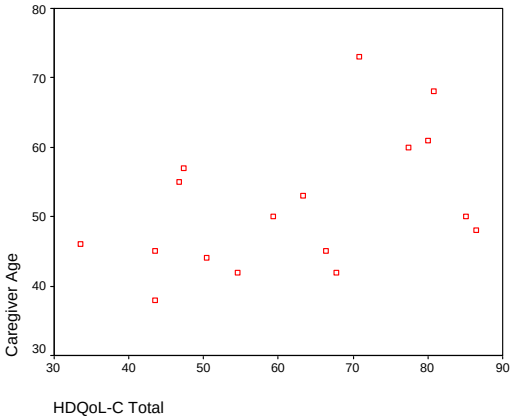

PAI Stress
90
80
70
60
50
40
40
50
60
70
80
90
HDQoL-C Total

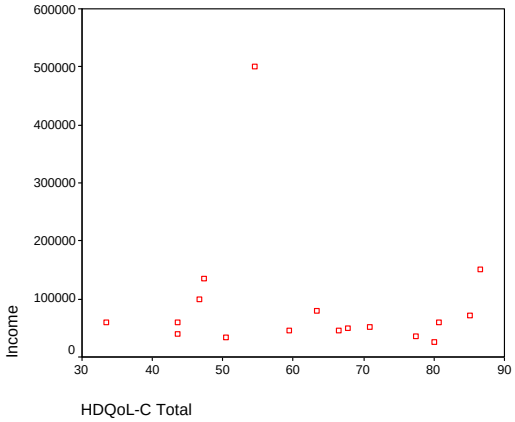

PAI Dominance
70
60
50
40
30
20
40
50
60
70
80
90
HDQoL-C Total

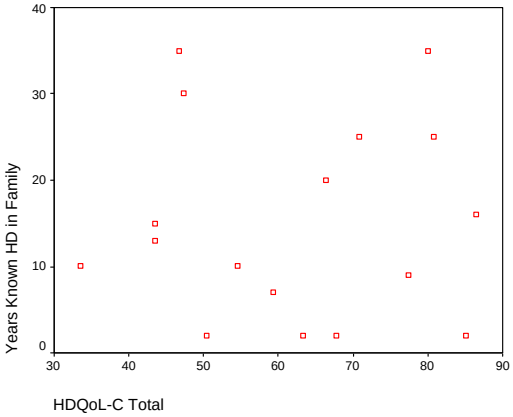

PAI Nonsupport
52
50
48
46
44
42
40
38
36
40
50
60
70
80
90
HDQoL-C Total

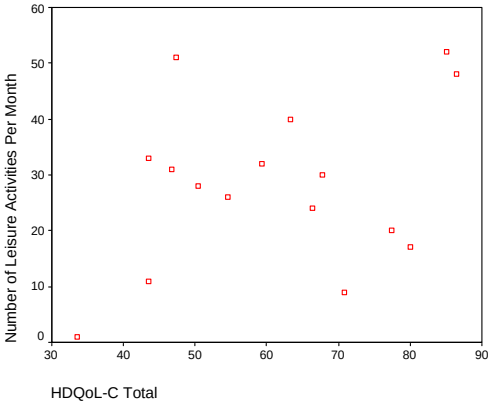

PAI Warmth
70
60
50
40
30
20
40
50
60
70
80
90
HDQoL-C Total

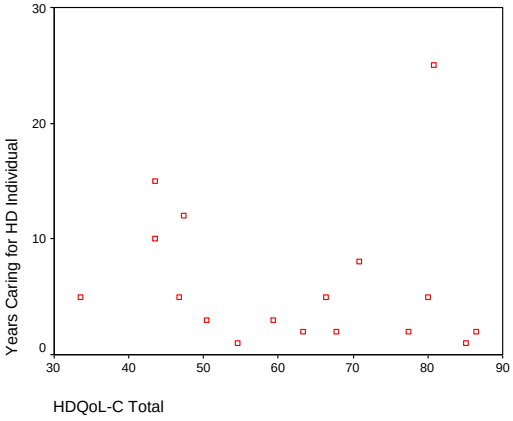
Caregiver Characteristics





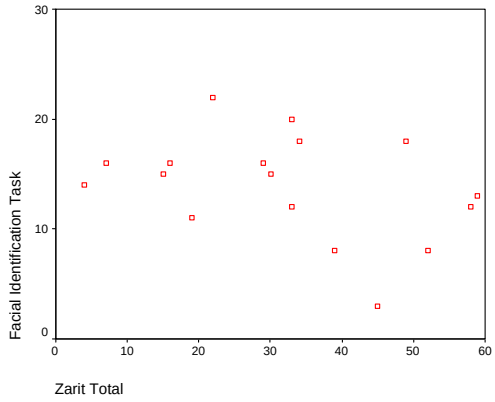


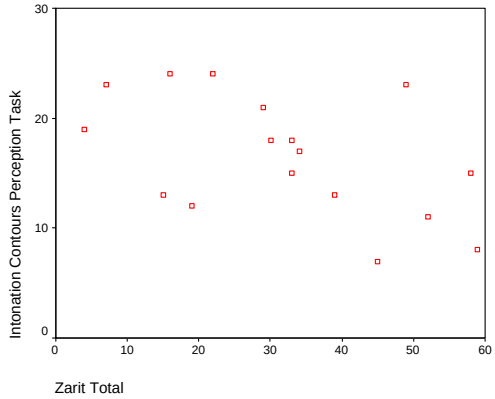


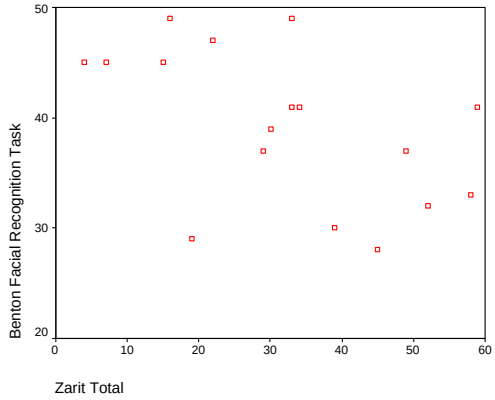


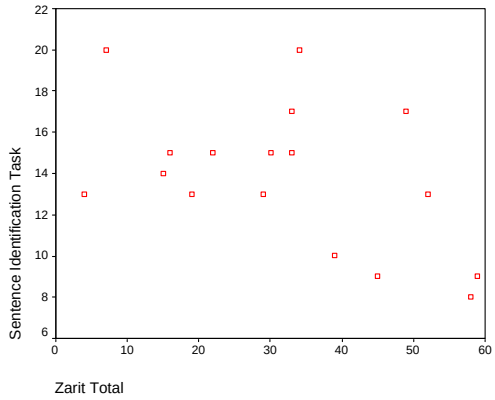
Zarit Burden Inventory

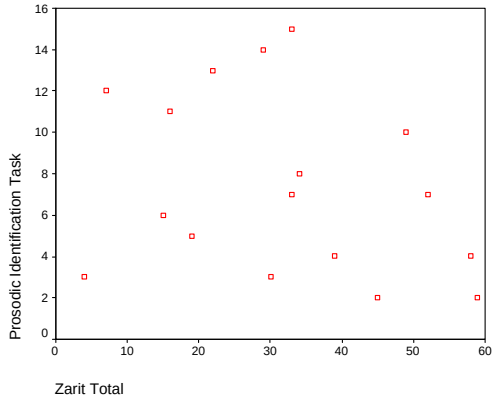
New York Emotion Battery

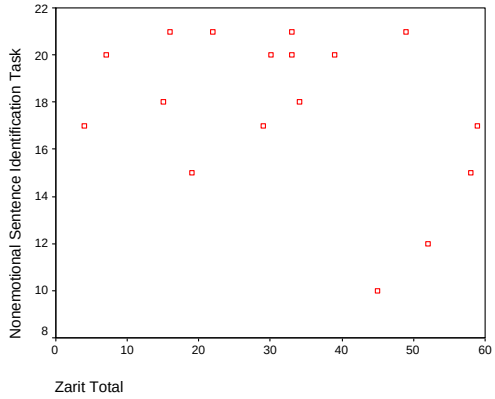




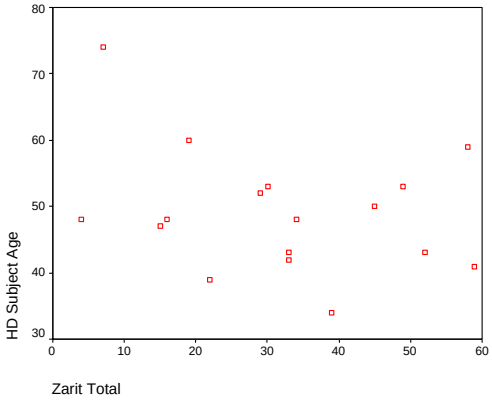


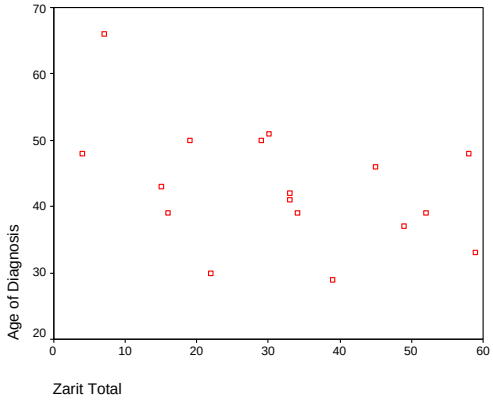


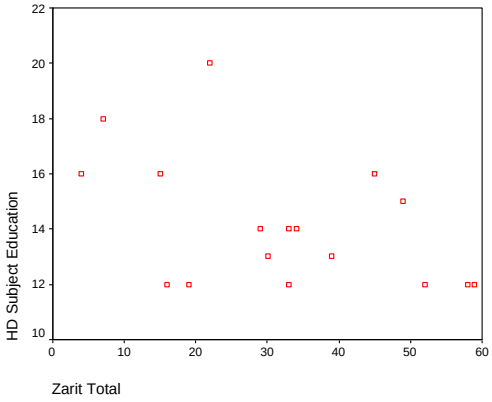


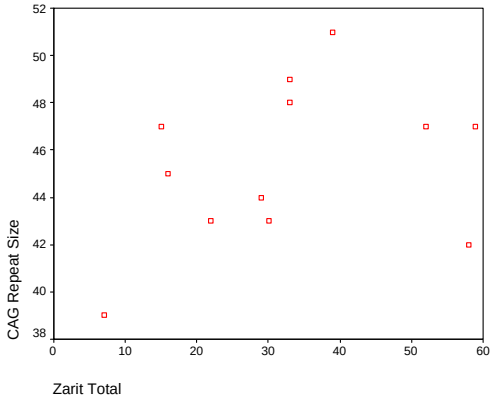


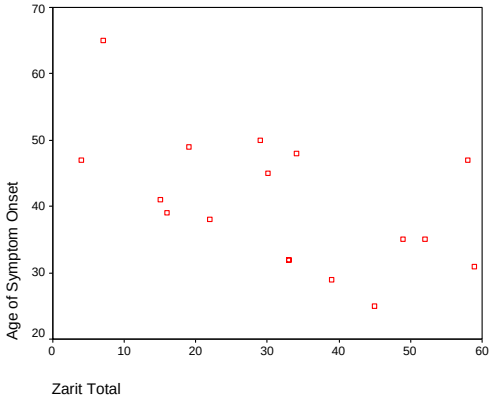
HD Subject Characteristics

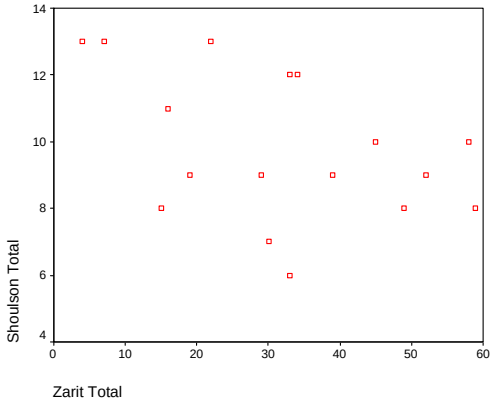




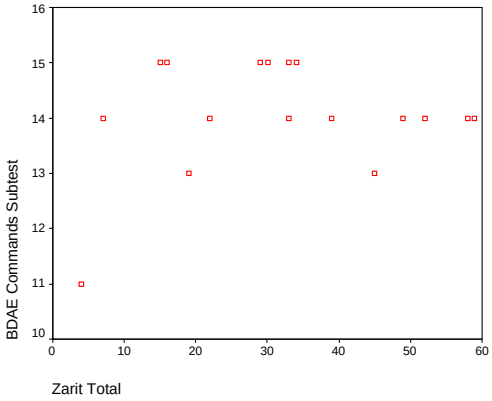


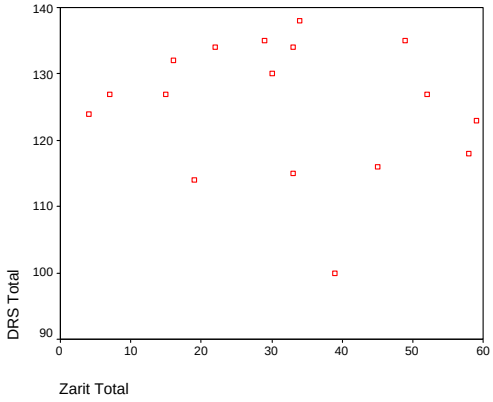


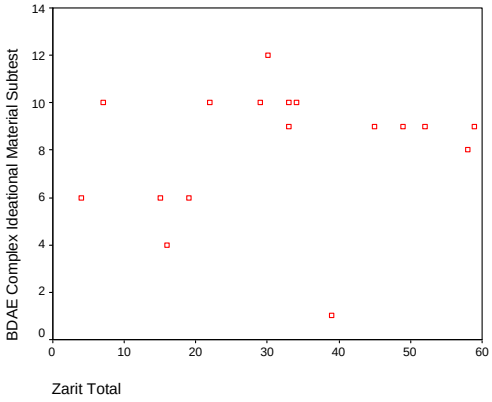


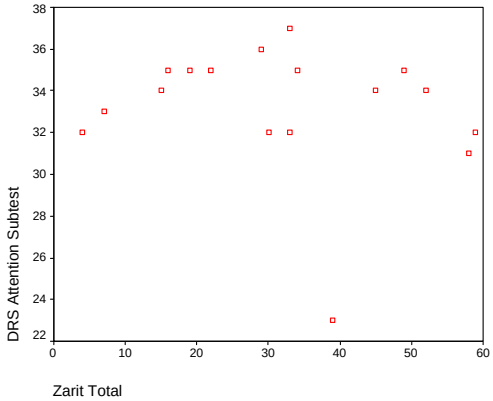


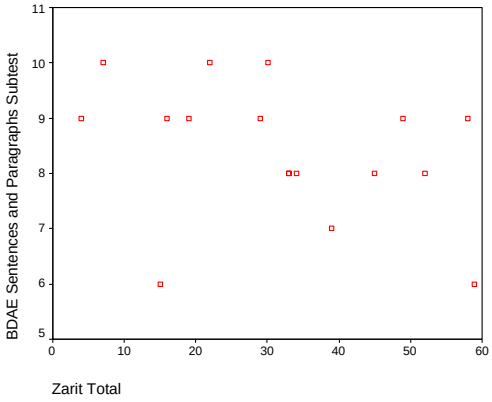
Cognitive Profile of HD subjects

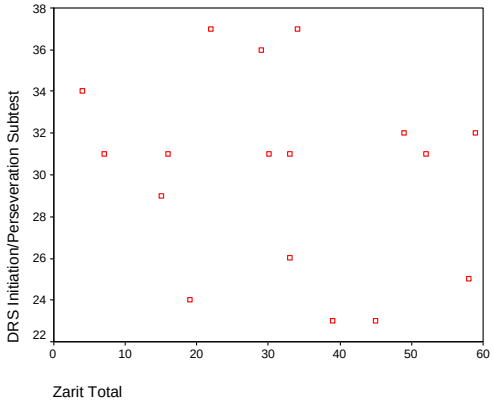


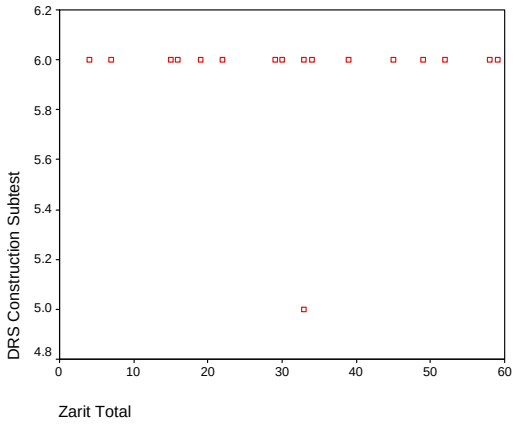




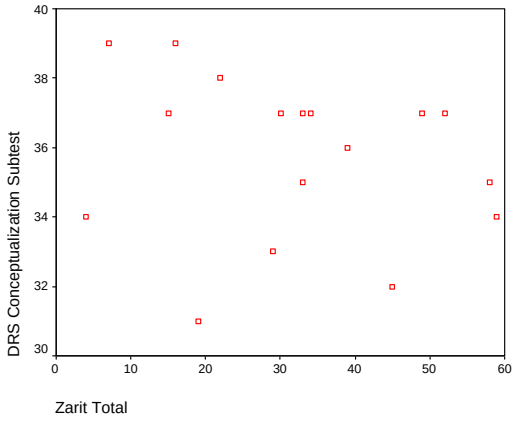


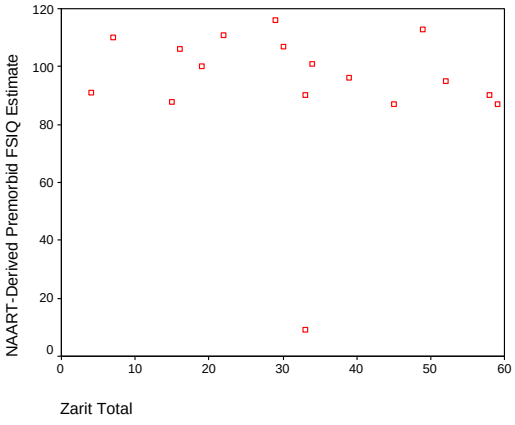


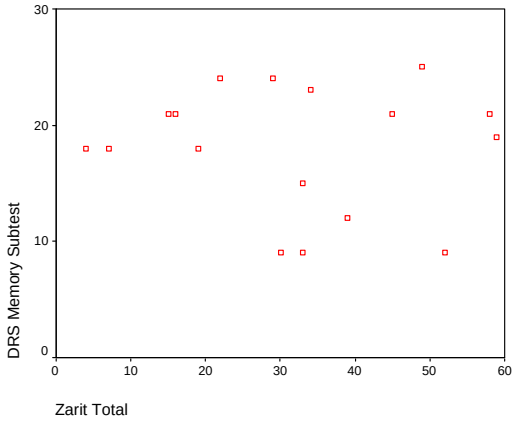



DRS Construction Subtest
6.2
6.0
5.8
5.6
5.4
5.2
5.0
4.8
0
10
20
30
40
50
60
Zarit Total

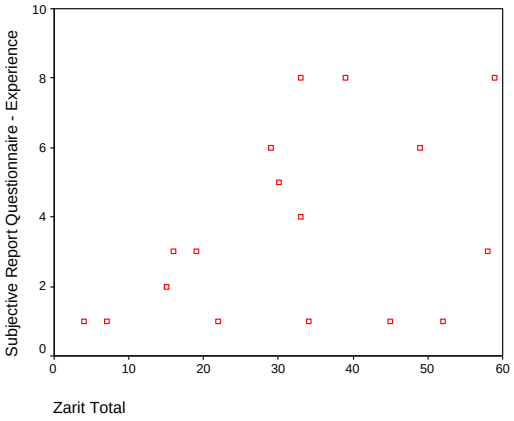

NAART Total
50
40
30
20
10
0
0
10
20
30
40
50
60
Zarit Total

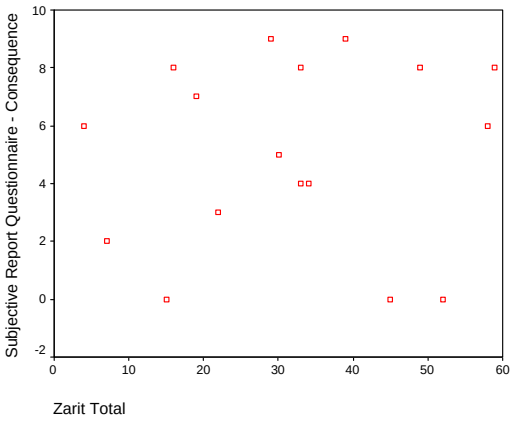

DRS Conceptualization Subtest
40
38
36
34
32
30
0
10
20
30
40
50
60
Zarit Total


NAART-Derived Premorbid FSIQ Estimate
120
100
80
60
40
20
0
0
10
20
30
40
50
60
Zarit Total

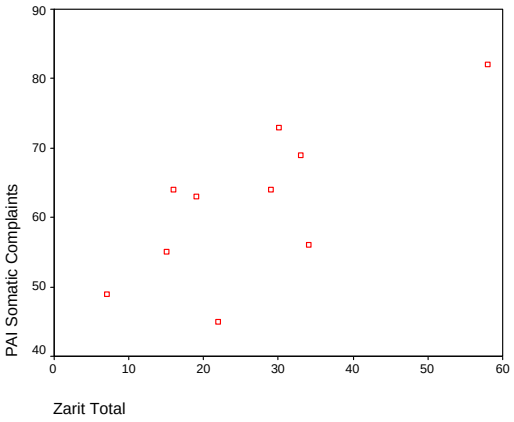

DRS Memory Subtest
30
20
10
0
0
10
20
30
40
50
60
Zarit Total

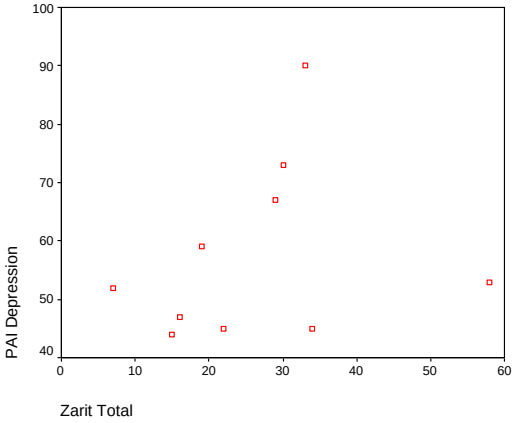
HD Subjects' Awareness

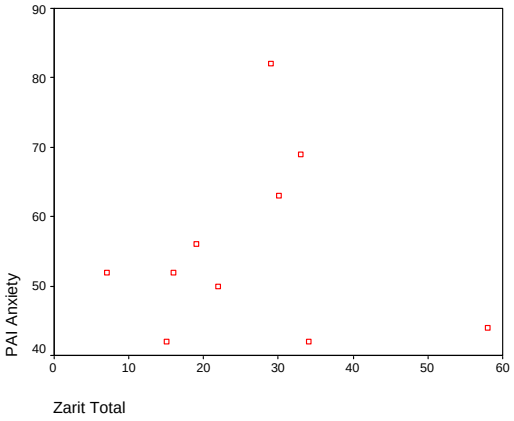


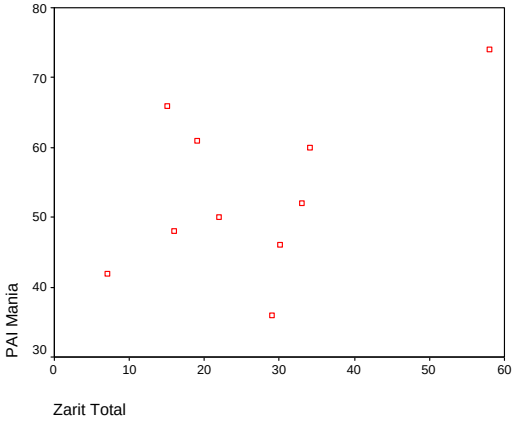


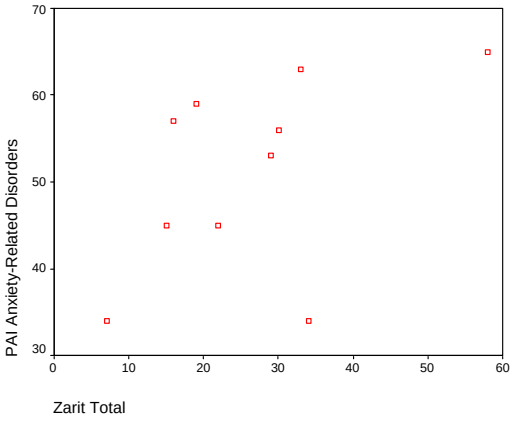
Psychiatric and Personality Functioning

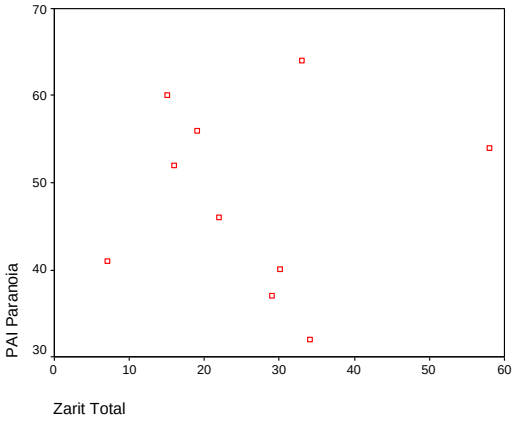


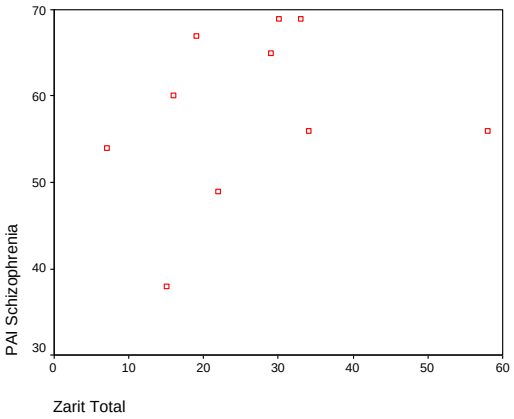


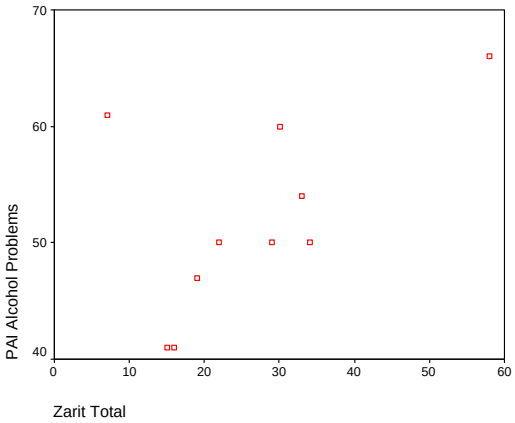


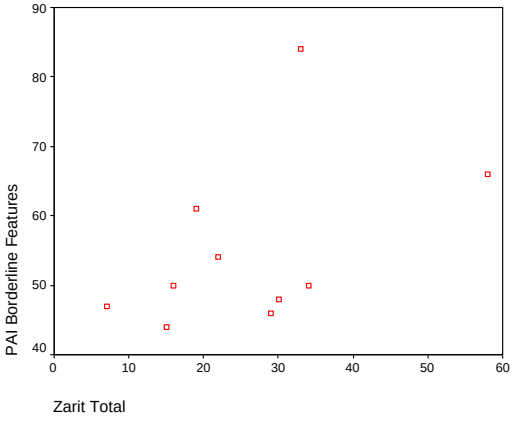


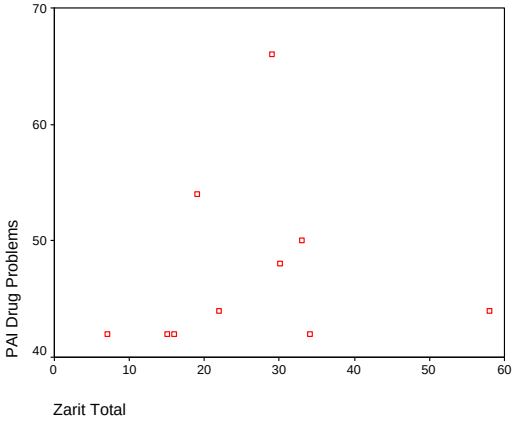


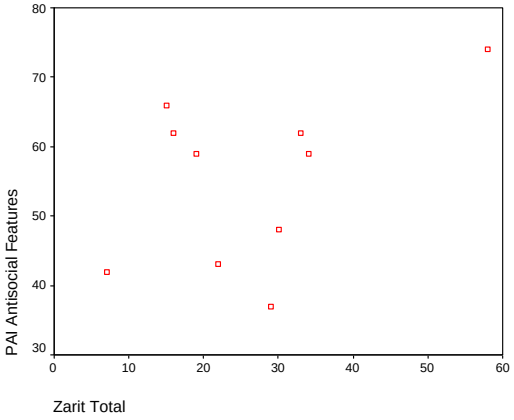


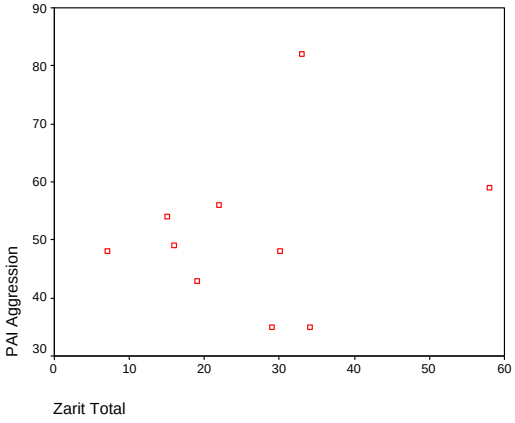

PAI Schizophrenia
70
60
50
40
30
0
10
20
30
40
50
60
Zarit Total

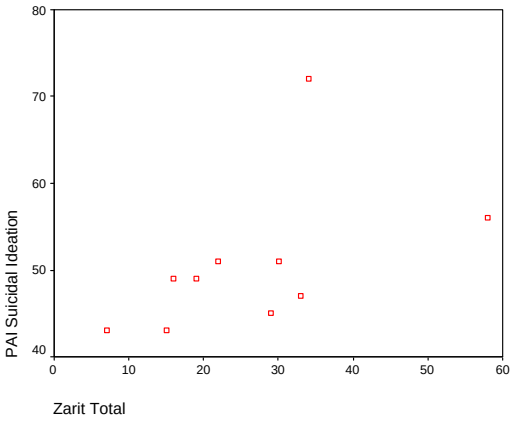

PAI Alcohol Problems
70
60
50
40
0
10
20
30
40
50
60
Zarit Total

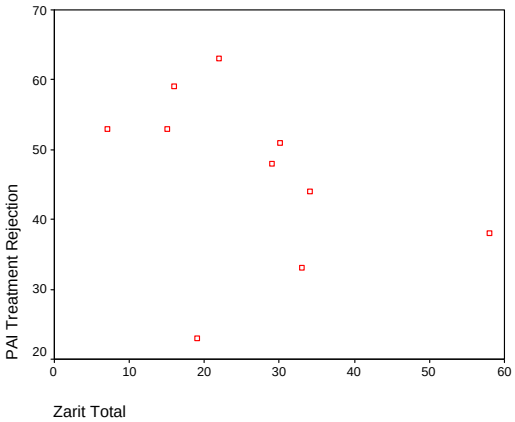

PAI Borderline Features
90
80
70
60
50
40
0
10
20
30
40
50
60
Zarit Total

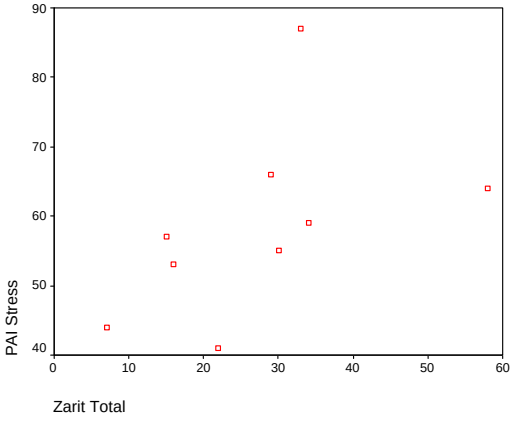

PAI Drug Problems
70
60
50
40
0
10
20
30
40
50
60
Zarit Total

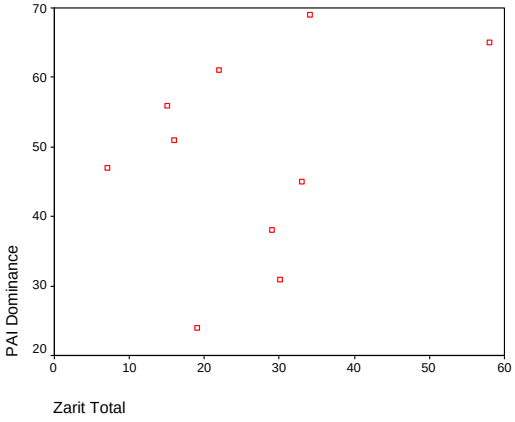

PAI Antisocial Features
80
70
60
50
40
30
0
10
20
30
40
50
60
Zarit Total

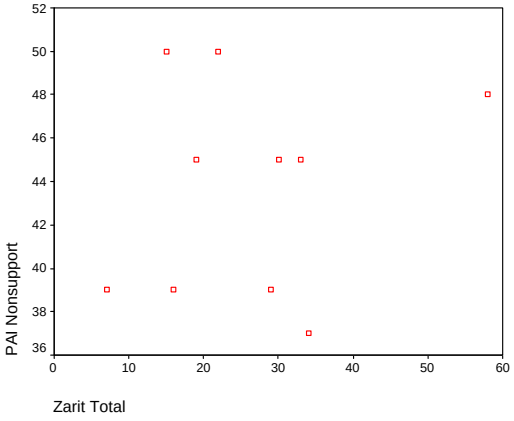

PAI Aggression
90
80
70
60
50
40
30
0
10
20
30
40
50
60
Zarit Total

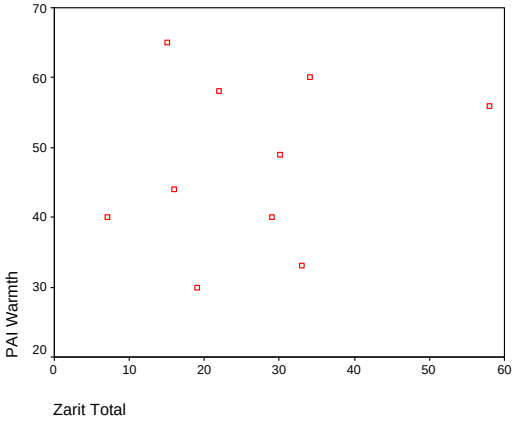

PAI Suicidal Ideation
80
70
60
50
40
0
10
20
30
40
50
60
Zarit Total


PAI Treatment Rejection
70
60
50
40
30
20
0
10
20
30
40
50
60
Zarit Total

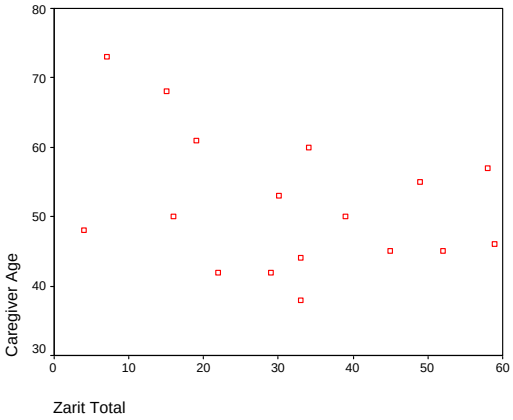

PAI Stress
90
80
70
60
50
40
0
10
20
30
40
50
60
Zarit Total

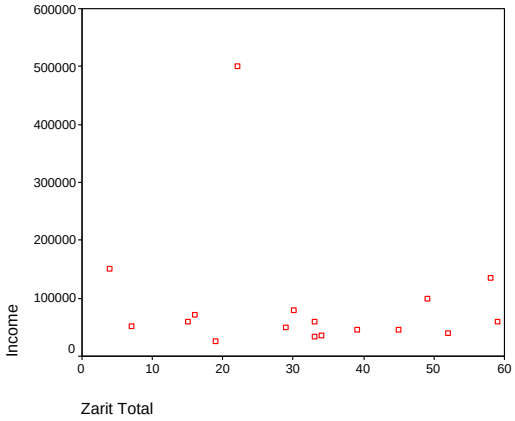

PAI Dominance
70
60
50
40
30
20
0
10
20
30
40
50
60
Zarit Total

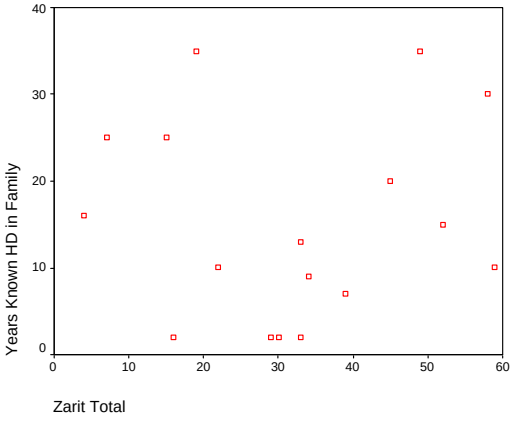

PAI Nonsupport
52
50
48
46
44
42
40
38
36
0
10
20
30
40
50
60
Zarit Total

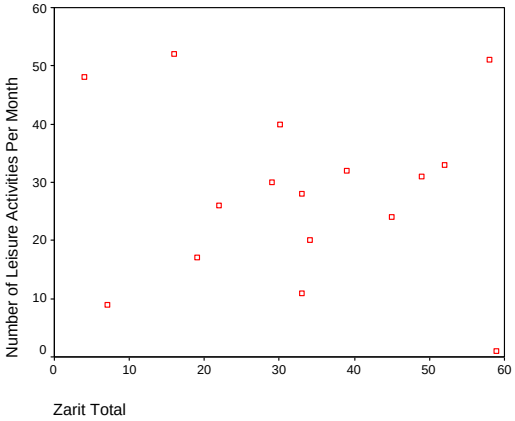

PAI Warmth
70
60
50
40
30
20
0
10
20
30
40
50
60
Zarit Total

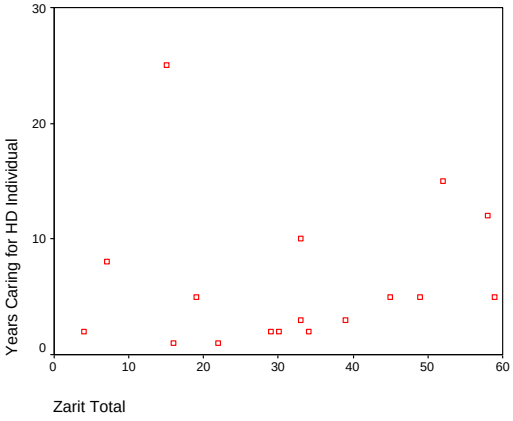
Caregiver Characteristics











Appendix E: Skewness, Kurtosis, and Normality of Variables

Skewness, Kurtosis and Normality Values for Caregiver Burden Measures

	Skewness[S]	Kurtosis[K]	Kolmogorov-Smirnov p
HDQoL-C Aspects of Caring	-0.06	-0.95	.200
HDQoL-C Satisfaction w Life	0.04	-1.15	.200
HDQoL-C Feelings about Life	0.11	-0.31	.200
HDQoL-C Total Score	-0.04	-1.21	.200
Zarit Total Score	0.04	-0.90	.200

HDQoL-C = Huntington's Disease Quality of Life for Carers Questionnaire; ZBI = Zarit Burden Inventory. [S]*SE* = .550. [K]*SE* = 1.063.

Skewness, Kurtosis and Normality Values for Measures of the New York Emotion Battery – Overall HD Group

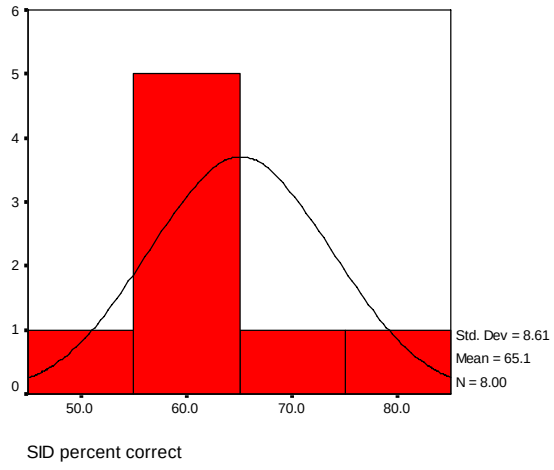
	Skewness[S]	Kurtosis[K]	Kolmogorov-Smirnov p
FID	-0.66	0.76	.200
PID	0.50	-1.07	.200
SID	-0.02	-0.28	.184
BFRT	-0.30	-1.20	.200
ICP	-0.03	-1.03	.200
NESID	-1.14	0.90	.065
FIDDIFF	-0.33	-1.24	.200
PIDDIFF	0.32	-0.65	.200
SIDDIFF	-0.14	0.00	.123

FID = Facial Identification Task; PID = Prosodic Identification Task; SID = Sentence Identification Task; BFRT = Benton Facial Recognition Task; ICP = Intonation Contours Task; NESID = Nonemotional Sentence Identification Task.; FIDDIFF = BFRT - FID; PIDDIFF = ICP - PID; SIDDIFF = NESID - SID. [S]*SE* = .536. [K]*SE* = 1.038.

Skewness, Kurtosis and Normality Values for Measures of the New York Emotion Battery – Nondemented HD Subgroup

	Skewness[S]	Kurtosis[K]	Kolmogorov-Smirnov p
FID	0.93	0.03	.144
PID	-0.51	-0.97	.200
SID	1.45	2.97	**.002** [N]
BFRT	-0.13	-2.36	.200
ICP	-0.69	-0.03	.200
NESID	-0.55	-1.56	.162
FIDDIFF	0.63	-0.91	.200
PIDDIFF	-0.40	-0.61	.200
SIDDIFF	-1.99	4.55	.091

FID = Facial Identification Task; PID = Prosodic Identification Task; SID = Sentence Identification Task; BFRT = Benton Facial Recognition Task; ICP = Intonation Contours Task; NESID = Nonemotional Sentence Identification Task.; FIDDIFF = BFRT - FID; PIDDIFF = ICP - PID; SIDDIFF = NESID - SID. [N] Normality violation. [S]*SE* = .752. [K]*SE* = 1.481.



Skewness, Kurtosis and Normality Values for HD Subject Characteristics

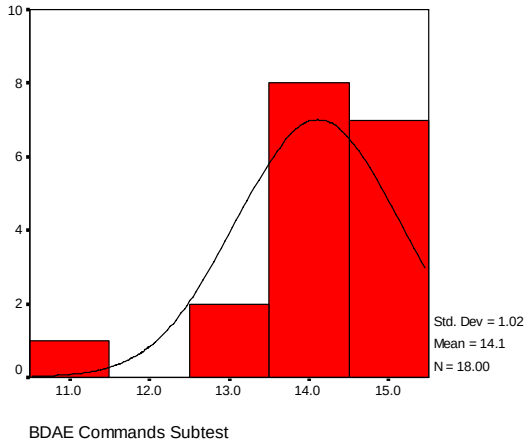
	Skewness	Kurtosis	Kolmogorov-Smirnov p
Age	1.13 [S1]	2.32 [K1]	.200
Education	1.13 [S1]	0.96 [K1]	.067
Age of Symptom Onset	0.76 [S1]	0.67 [K1]	.200
Age Diagnosed	0.70 [S1]	1.62 [K1]	.200
CAG Repeat Size	-0.04 [S2]	-0.29 [K2]	.200
Shoulson Total	0.00 [S1]	-1.10 [K1]	.197

[S1]*SE* = .536. [S2]*SE* = .597. [K1]*SE* = 1.038. [K2]*SE* = 1.154.

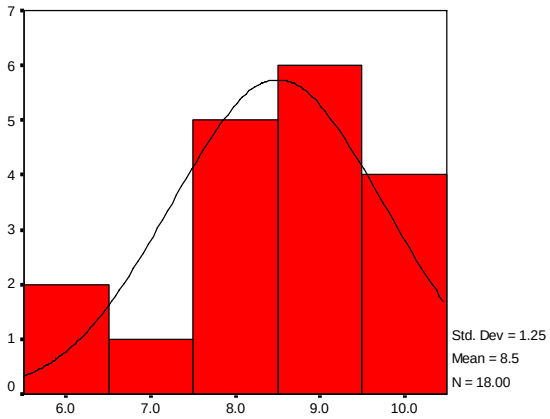
Skewness, Kurtosis and Normality Values for HD Subjects' Cognitive Profile

	Skewness[S]	Kurtosis[K]	Kolmogorov-Smirnov p
BDAE Commands	-1.73	4.14	**.000** [N]
BDAE Complex Ideational Material	-1.20	1.58	**.002** [N]
BDAE Sentences and Paragraphs.	-0.71	-0.02	**.033** [N]
DRS Total	-0.99	0.87	.200
DRS Attention	-2.21	6.93	**.032** [N]
DRS Initiation/Perseveration	-0.29	-0.97	**.036** [N]
DRS Construction	-2.71	5.98	**.000** [N]
DRS Conceptualization	-0.60	-0.44	**.015** [N]
DRS Memory	-0.76	-0.61	**.043** [N]
NAART Premorbid IQ Estimate	-2.99	11.08	.200

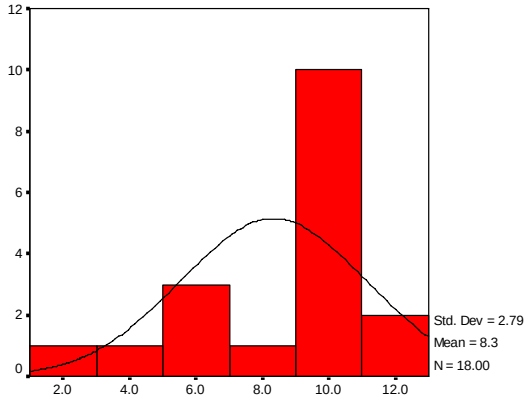
BDAE = Boston Diagnostic Aphasia Examination; DRS = Dementia Rating Scale; NAART = North American Adult Reading Test.
[N] Normality violation. [S]*SE* = .536. [K]*SE* = 1.038.



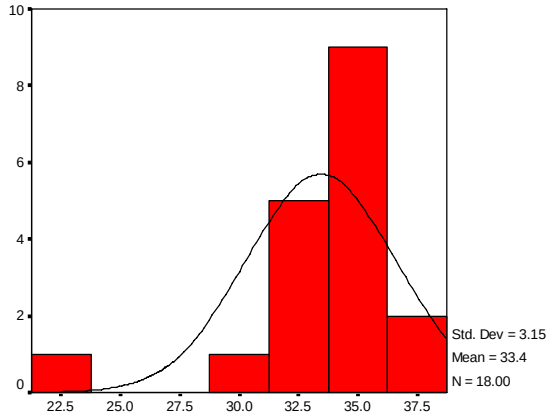
BDAE Commands Subtest



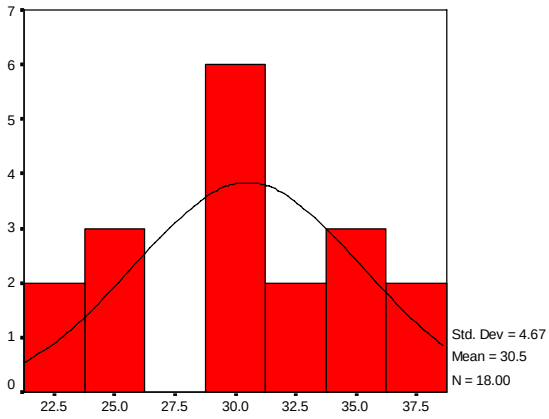
BDAE Sentences and Paragraphs



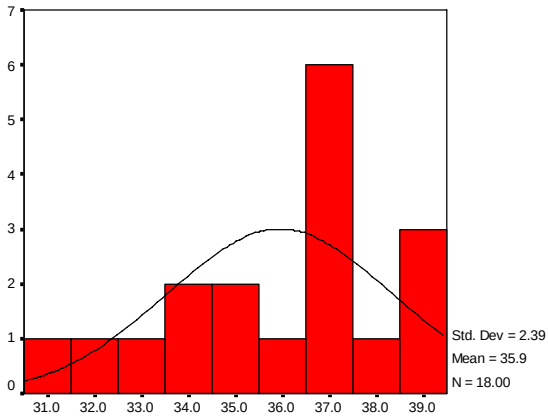
BDAE Complex Ideational Material



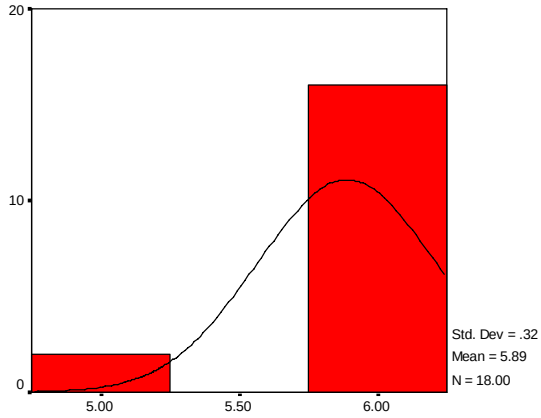
DRS Attention Subtest



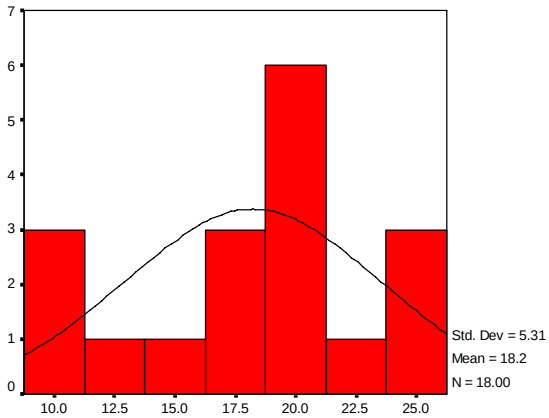
DRS Initiation/Perseveration Subtest



DRS Conceptualization Subtest



DRS Construction Subtest



DRS Memory Subtest

Skewness, Kurtosis and Normality Values for HD Subjects' Awareness

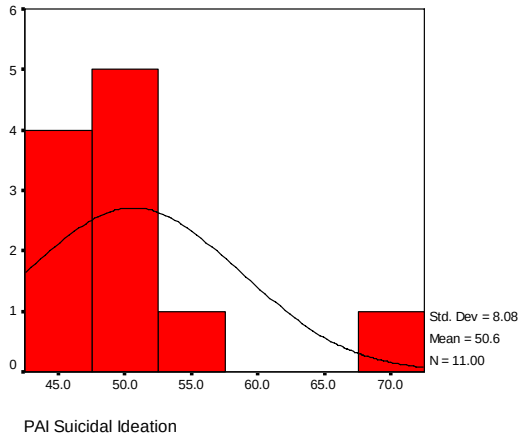
	Skewness[S]	Kurtosis[K]	Kolmogorov-Smirnov p
Experience	0.43	-1.43	.119
Consequence	-0.55	-1.06	.110

[N] Normality violation. [S]*SE* = .536. [K]*SE* = 1.038.

Skewness, Kurtosis and Normality Values for Psychiatric and Personality Functioning Measures

	Skewness	Kurtosis	Kolmogorov-Smirnov p
PAI Somatic Complaints	0.95 [S1]	1.15 [K1]	.200
PAI Anxiety	0.94 [S1]	0.70 [K1]	.200
PAI Anxiety Disorders	0.05 [S1]	-0.18 [K1]	.200
PAI Depression	1.10 [S1]	0.84 [K1]	.200
PAI Mania	0.56 [S1]	-0.29 [K1]	.200
PAI Paranoia	0.12 [S1]	-1.08 [K1]	.200
PAI Schizophrenia	-0.51 [S1]	0.31 [K1]	.200
PAI Borderline	1.45 [S1]	2.12 [K1]	.108
PAI Antisocial	-0.06 [S1]	-0.88 [K1]	.200
PAI Alcohol Problems	0.40 [S1]	-0.85 [K1]	.200
PAI Drug Problems	1.85 [S1]	3.72 [K1]	.114
PAI Aggression	1.21 [S1]	2.85 [K1]	.200
PAI Suicidal Ideation	2.04 [S1]	5.18 [K1]	**.006** [N]
PAI Stress	1.17 [S2]	2.12 [K2]	.200
PAI Nonsupport	0.13 [S1]	-1.69 [K1]	.062
PAI Treatment Rejection	-0.38 [S1]	-0.80 [K1]	.200
PAI Dominance	-0.49 [S1]	-0.79 [K1]	.200
PAI Warmth	-0.02 [S1]	-1.30 [K1]	.200

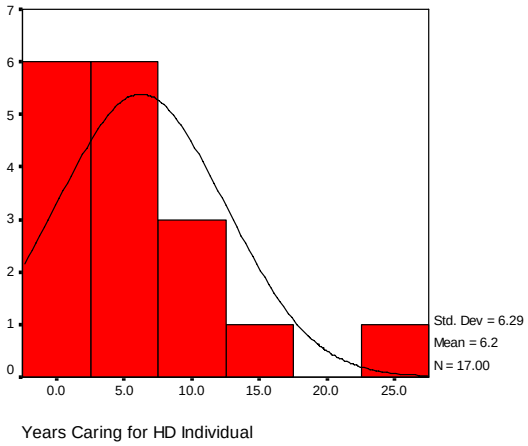
PAI = Personality Assessment Inventory.
[N] Normality violation. [S1]*SE* = .661. [S2]*SE* = .687. [K1]*SE* = 1.279. [K2]*SE* = 1.334.

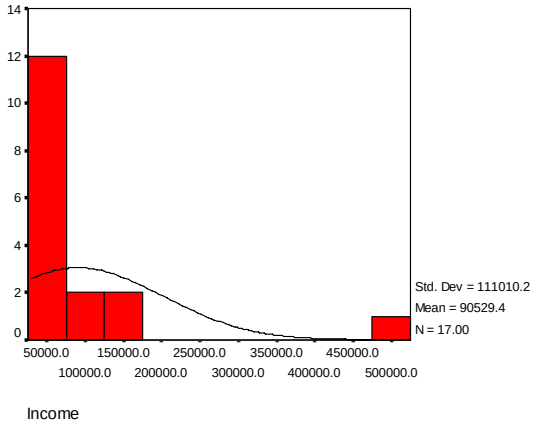


Skewness, Kurtosis and Normality Values for Caregiver Characteristics

	Skewness[S]	Kurtosis[K]	Kolmogorov-Smirnov p
Caregiver Age	0.82	0.70	.200
Time Known HD Family	0.51	-0.94	.200
Time Caring for HD Individual	1.95	4.15	**.001** [N]
Income	3.51	13.25	**.001** [N]
Leisure Activities	-0.01	-0.48	.200

[N] Normality violation. [S]*SE* = .550. [K]*SE* = 1.063.

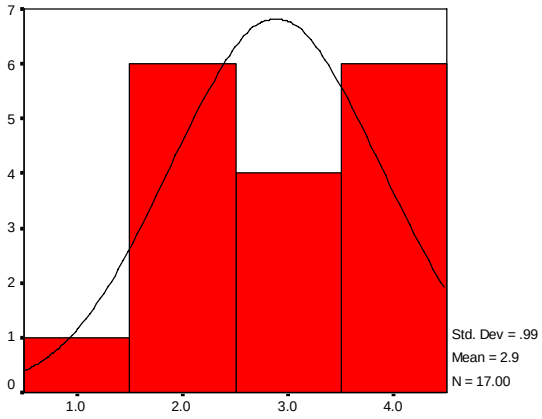




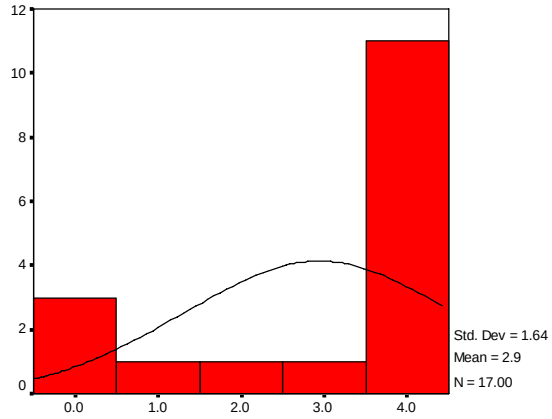
Skewness, Kurtosis and Normality Values for Ordinal and Categorical Variables

	Skewness	Kurtosis	Kolmogorov-Smirnov p
Patient Occupation	-0.32 [S1]	-0.14 [K1]	.200
Caregiver Education	-0.17 [S2]	-1.26 [K2]	**.022** [N]
Caregiver Disability	1.63 [S2]	2.56 [K2]	**.000** [N]
Hours Paid Work/Week	-1.15 [S2]	-0.45 [K2]	**.000** [N]
Hours Caring Children	0.76 [S3]	-1.13 [K3]	**.013** [N]
Hours Caring HD Indiv.	-0.37 [S4]	-1.63 [K4]	**.001** [N]

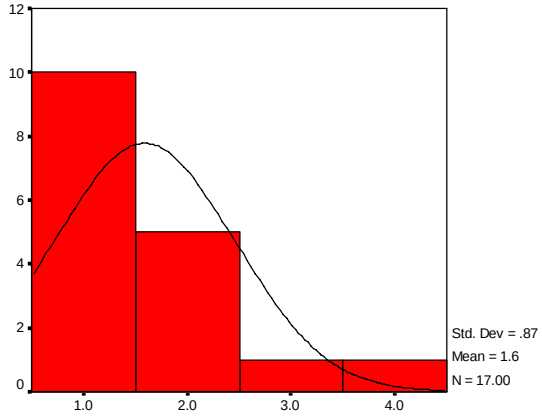
[N] Normality violation. [S1]*SE* = .536. [S2]*SE* = .550. [S3]*SE* = .564. [S4]*SE* = .580. [K1]*SE* = 1.038. [K2]*SE* = 1.063. [K3]*SE* = 1.091. [K4]*SE* = 1.121.



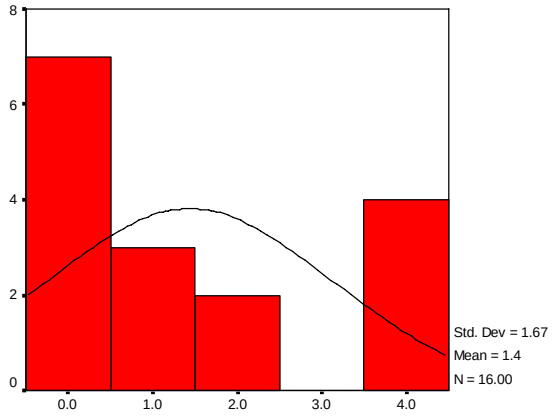
Caregiver education



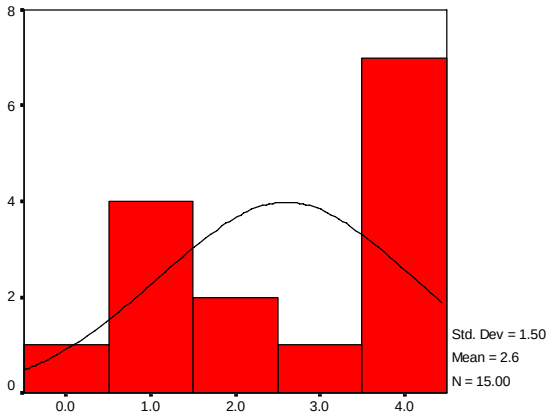
Hours paid work per week



Level of Disability



Hours Caring for Children Per Week



Hours Caring for HD Individual Per Week

Appendix F: Multiple-Method Calculation Explanation

Overview

The HD versus HC comparisons were carried out in three steps: 1) derivation of Sum of Squares (*SS*), degrees of freedom (*df*), and Mean Square (*MS*) values for all main effect and interaction terms using SPSS (version 11.5.2.1; SPSS Inc; Chicago, IL), 2) calculation of the Error terms for between and within analyses using Microsoft Excel (version 2003; Microsoft Corp; Redmond, WA), and 3) calculation of the *MS* and *F*-ratio by hand.

SPSS Analysis

The means (*M*) for the experimental and control tasks, weighted by their sample sizes (*n*), were input into SPSS. A repeated measures ANOVA was performed on this summary data. The output included *SS*, *df*, and *MS* values for all main effect and interaction terms. However, with only summary data, SPSS is unable to calculate the Error *SS* (and consequently, the *F* and *p* values for all main effects and interactions cannot be calculated). The Error *SS* can be calculated manually using Excel.

Excel Analysis

The Error *SS* analysis can be automated using an Excel spreadsheet. Existing *M*, standard deviation (*SD*), and sample size values for experimental and control tasks, and the derived difference scores, broken down by demographic categories (e.g., males with less than or equal to 15 years of education), were input. The Error SS (*ESS*) for the within-subjects analysis can be obtained by squaring the standard deviation of the difference scores (SD_{diff}) and summing across categories. To compute the *ESS* for the between-subjects analysis, the correlation between repeated measures is needed.

Correlations between tasks (e.g., FID and BFRT), based on the standard deviations of the difference scores, were calculated for each difference score. This calculation is based on the usual formula for the variance of a difference in paired data (Keppel, 1991). These correlations were then used to compute the pooled standard deviation (SD_p) between subjects for the between-subjects main effects and interactions using the usual formula for the variance of a sum of observations for paired data (Keppel, 1991). These calculations were carried out utilizing appropriate formulae within Excel. With both the summary and the derived data, the between and within *ESS* were computed for each demographic category. The *ESS* between and within were summed across demographic categories for an overall between and within *ESS*.

Hand Analysis

The two *ESS*s (between and within) obtained from the Excel analysis, along with the *df* from the SPSS output, were plugged into formulae for Error Mean Square (*EMS*) and calculated by hand. The resulting *EMS* was utilized in the (denominator of the) *F*-ratio calculations for main effects and interactions. Level of significance for each *F* statistic was calculated using a free online *p*-value calculator for the *F*-test (Soper, 2008).

Sample of SPSS, Excel, and Hand Analysis

Below is a sample problem utilizing only HD data, which allows the multiple-method calculations to be verified with the SPSS output that was calculated utilizing raw data. Descriptive statistics for FID, BFRT, and the Difference Score of these two tasks are found below in a 2 (Gender) x 2 (Education) x 3 (Task) table.

		FID Education			**BFRT Education**			**Diff Scores Education***	
		≤15	**≥16**		**≤15**	**≥16**		**≤15**	**≥16**
Gender	M	*M*=39.38	37.50	M	71.11	75.46	M	31.74	37.96
		SD=9.78	18.92		13.71	15.74		7.65	3.95
		N=5	4		5	4		5	4
	F	46.88	68.75	F	72.92	87.04	F	26.04	18.29
		12.61	0		13.35	0		10.04	0
		8	1		8	1		8	1

*Information added for sake of comprehensiveness; data not utilized in analyses.

SPSS Analysis

Using codes for gender (1 = male and 2 = female) and education (1 = ≤15 yrs education and 2 = ≥16 yrs education), the following data were entered into SPSS:

Education code	**Gender Code**	**FID *M***	**BFRT *M***	***n* by cell**
1	1	39.38	71.11	5
2	1	37.50	75.46	4
1	2	46.88	72.92	8
2	2	68.75	87.04	1

A 2 x 2 x 2 repeated-measures ANOVA was performed in SPSS utilizing experimental and control tasks means, weighted by sample size, to assess the differences between men and women and among lesser and greater years of education on experimental and control tasks. Specifically, factors included two between-subjects factors, that is, Gender (2: male and female) and Education (2: ≤15 years education and

≥16 years of education), and one within-subjects factor, this is, Task (2: facial experimental and facial control). The DV was percent correct for each task.

The following output was produced from SPSS:

Tests of Within-Subjects Contrasts

Source	*SS*	*df*	*MS*	*F*	*p*
Task	4127.74	1	4127.74	-	-
Task x Gender	204.33	1	204.33	-	-
Task x Education	0.74	1	0.74	-	-
Task x Gender x Education	62.06	1	62.06	-	-
Error	0.00	14	0.00	-	-

Tests of Between-Subjects Effects

Source	*SS*	*df*	*MS*	*F*	*p*
Gender	862.71	1	862.71	-	-
Education	469.87	1	469.87	-	-
Gender x Education	356.65	1	356.65	-	-
Error	0.00	14	0.00	-	-

From the above tables, it is observed that the *ESS*s are missing, consequently precluding *F* and *p* from being calculated.

Excel Analysis

In order to obtain these the *ESSs*, the following information was entered into Excel:

		FID		BFRT		Difference scores	
Gender	**Education**	***M***	***SD***	***M***	***SD***	***M***	***SD***
Male	≤15	39.38	9.78	71.11	13.71	31.74	7.65
Male	≥16	37.50	18.92	75.46	15.74	34.51	3.95
female	≤15	48.88	12.61	72.92	13.35	26.04	10.04
female	≥16	68.75	0.00	87.04	0.00	18.29	0.00

Utilizing the above data, the correlation (*r*) and the SD_p were calculated in Excel by utilizing the following formulae. The formula for *r* comes from solving for *r* in the usual formula for the standard deviation of a paired difference (Keppel, 1991). The

formula for the SD_p comes from the usual formula for the standard deviation of a paired sum (Keppel, 1991).

$$r = \frac{var_1 + var_2 - var_{diff}}{2*SD_1*SD_2} \qquad SD_p = \sqrt{(var_1 + var_2 + (2*SD_1*SD_2*r)}$$

The following data resulted:

Gender	Education	*r*	SD_p
Male	≤15	0.84	22.56
Male	≥16	0.99	34.59
Female	≤15	0.70	23.95
Female	≥16	-	-

Utilizing sample size, SD_p, and SD_{diff}, *ESS* within and between were calculated in Excel using the following formulae:

$$ESS_{within} = \frac{(n\text{-}1)*SD_{diff}}{2} \qquad ESS_{between} = \frac{(n\text{-}1)*SD_p}{2}$$

The following data resulted:

Gender	Education	SS_{within}	$SS_{between}$
male	≤15	117.03	1017.50
male	≥16	23.35	1794.18
female	≤15	353.02	2008.11
female	≥16	0	0
		∑ = 493.40	∑ = 4819.79

Hand Analysis

Utilizing the overall *ESS* for within and between, along with the *df*, *EMS* within and between were calculated by hand:

$$EMS_{within} = \frac{ESS_{within}}{df_{within}} = \frac{493.40}{14} = 35.24$$

$$EMS_{between} = \frac{ESS_{between}}{df_{between}} = \frac{4819.79}{14} = 344.27$$

*EMS*s within and between were utilized to calculate *F*-ratios for respective main effects and interactions.

$$F_{\text{source}} = \frac{MS_{\text{source}}}{EMS_{\text{error}}}$$

The following *F* statistics resulted (level of significance for each *F* statistic was calculated using a free online *p*-value calculator for the *F*-test (Soper, 2008):

Tests of Within-Subjects Contrasts

Source	***F***	***p****
Task	117.12	.000
Task x Gender	5.80	.030
Task x Education	0.02	.887
Task x Gender x Education	1.76	.206

**p* values were calculated using Soper, D.S. (2008) "The Free Statistics Calculators Website", *Online Software*, http://www.danielsoper.com/statcalc/

Tests of Between-Subjects Effects

Source	***F***	***p****
Gender	2.51	.136
Education	1.37	.262
Gender x Education	1.04	.326

**p* values were calculated using Soper, D.S. (2008) "The Free Statistics Calculators Website", *Online Software*, http://www.danielsoper.com/statcalc/

Data Checking

Once the multiple-method analysis was completed, the same analysis was performed utilizing the raw data in SPSS, thereby verifying that multiple-method calculations were accurate. The SPSS output can be seen in the following tables:

Tests of Within-Subjects Contrasts

Source	***SS***	***df***	***MS***	***F***	***p***
Task	4127.74	1	4127.74	117.12	.000
Task x Gender	204.33	1	204.33	5.80	.030
Task x Education	0.74	1	0.74	0.02	.887
Task x Gender x Education	62.06	1	62.06	1.76	.206
Error	493.40	14	35.24	-	-

Tests of Between-Subjects Effects

Source	*SS*	*df*	*MS*	*F*	*p*
Gender	862.71	1	862.71	2.51	.136
Education	469.87	1	469.87	1.37	.262
Gender x Education	356.65	1	356.65	1.04	.326
Error	4819.79	14	344.27	-	-

Completing Multiple Analyses

Once the formulae were programmed into the Excel spreadsheet, calculations of r, SD_p, SD_{within}, and $SD_{between}$ were quickly and easily performed for subsequent analyses by simply entering the respective M, SD, and n for those analyses into the appropriate cells.

Sample HD versus HC Analysis

In the above example, it was shown that the results of the multiple-method calculations (i.e., SPSS, Excel, and hand analyses) performed utilizing summary statistics were virtually identical to the results of the SPSS calculations performed on the raw data. Below, an actual HD versus HC analysis is illustrated. Note that the result from the Task x Group interaction is equivalent to the result of the Broad Approach one-way ANOVA performed on the FIDDIFF variable found in the Results Section on page 84. Inaccessibility to raw data prevents a proof from being performed. Descriptive statistics for FID, BFRT and the Difference Score of these two tasks are found below in a 2 (Gender) x 3 (Task) table.

		FID		BFRT		Difference Scores*
Group		43.92		73.77		29.84
	HD	14.40	HD	13.22	HD	9.45
		18		18		18
		73.30		87.26		13.95
	HC	11.03	HC	7.93	HC	11.81
		103		103		103

*Information added for sake of comprehensiveness; data not utilized in analyses.

SPSS Analysis

Using codes for group (1 = HD and 2 = HC), the following data were entered into SPSS:

Group Code	FID *M*	BFRT *M*	*n* by cell
1	43.92	73.77	18
2	73.30	87.26	103

A 2 x 2 repeated-measures ANOVA was performed in SPSS utilizing experimental and control tasks means, weighted by sample size, to assess the differences among HD and HC subjects on experimental and control tasks. Specifically, factors included one between-subjects factors, that is, Group (2: HD subjects and HC subjects), and one within subjects factor, this is, Task (2: facial experimental and facial control). The DV was percent correct for each task. The following output was produced from SPSS:

Tests of Within-Subjects Contrasts

Source	*SS*	*df*	*MS*	*F*	*p*
Task	14694.58	1	14694.58	-	-
Task x Group	1933.87	1	1933.87	-	-
Error	0.00	119	0.00	-	-

Tests of Between-Subjects Effects

Source	*SS*	*df*	*MS*	*F*	*p*
Group	14077.92	1	14077.92	-	-
Error	0.00	119	0.00	-	-

From the above tables, it is observed that the *ESS*s are missing, consequently precluding *F* and *p* from being calculated.

Excel Analysis

In order to obtain these the *ESS*s, the following information was entered into Excel:

	FID		BFRT		Difference scores	
Group	***M***	***SD***	***M***	***SD***	***M***	***SD***
HD	43.92	14.40	73.77	13.22	29.84	9.45
HC	73.30	11.03	87.26	7.93	13.95	11.81

Utilizing the above data, the r and the SD_p were calculated in Excel by utilizing the following formulae:

$$r = \frac{\text{var}_1 + \text{var}_2 - \text{var}_{diff}}{2*SD_1*SD_2} \qquad SD_p = \sqrt{(\text{var}_1 + \text{var}_2 + (2*SD_1*SD_2*r)}$$

The following data resulted:

Group	***r***	SD_p
HD	0.77	25.98
HC	0.26	15.15

Utilizing sample size, SD_p, and SD_{diff}, *ESS* within and between were calculated in Excel using the following formulae:

$$ESS_{within} = \frac{(n\text{-}1)*SD_{diff}}{2} \qquad ESS_{between} = \frac{(n\text{-}1)*SD_{pooled}}{2}$$

The following data resulted:

Group	SS_{within}	$SS_{between}$
HD	759.07	5737.11
HC	7113.28	11710.39
	$\sum$ = 7872.35	$\sum$ = 17447.50

Hand Analysis

Utilizing the overall *ESS* for within and between, along with the *df*, *EMS* within and between were calculated by hand:

$$EMS_{within} = \frac{ESS_{within}}{df_{within}} = \frac{7872.35}{119} = 66.15$$

$$EMS_{between} = \frac{ESS_{between}}{df_{between}} = \frac{17447.5}{119} = 146.62$$

*EMS*s within and between were utilized to calculate *F*-ratios for respective main effects and interactions.

$$F_{source} = \frac{MS_{source}}{EMS_{error}}$$

The following *F* statistics resulted (level of significance for each *F* statistic was calculated using a free online *p*-value calculator for the *F*-test (Soper, 2008):

Tests of Within-Subjects Contrasts

Source	***F***	***p****
Task	222.13	.000
Task x Group	29.25	.000

**p* values were calculated using Soper, D.S. (2008) "The Free Statistics Calculators Website", *Online Software*, http://www.danielsoper.com/statcalc/

Note that the result from the Task x Group interaction in the table above is equivalent to the result of the Broad Approach one-way ANOVA performed on the FIDDIFF variable found in the Results Section.

Tests of Between-Subjects Effects

Source	***F***	***p****
Group	96.02	.000

**p* values were calculated using Soper, D.S. (2008) "The Free Statistics Calculators Website", *Online Software*, http://www.danielsoper.com/statcalc/

Appendix G: Individuated Analyses

FID (by age)

HD Group	n	HD Subject Breakdown:	N	HC Subject Breakdown:	Analysis:
Whole	18	All	32	40-59 y.o.	1-way ANOVA
	14	40-59 y.o.	32	40-59 y.o.	1-way ANOVA
Nondemented Subgroup	8	All	32	40-59 y.o.	1-way ANOVA
	7	40-59 y.o.	32	40-59 y.o.	1-way ANOVA

PID (by age)

HD Group	n	HD Subject Breakdown:	n	HC Subject Breakdown:	Analysis:
Whole	18	All	32	40-59 y.o.	1-way ANOVA
	14	40-59 y.o.	32	40-59 y.o.	1-way ANOVA
Nondemented Subgroup	8	All	32	40-59 y.o.	1-way ANOVA
	7	40-59 y.o.	32	40-59 y.o.	1-way ANOVA

SID (by age, education, and ethnicity)

HD Group	n	HD Subject Breakdown:	n	HC Subject Breakdown:	Analysis:
Whole	18	All (13 ≤15 y.e. vs. 5 ≥ 16 y.e.)	18	40-59 year-old Caucasians (9 ≤15 y.e vs. 9 ≥16 y.e.)	Group x Educ. 2-way ANOVA
	14	40-59 y.o. (11 ≤15 y.e vs. 3 ≥16 y.e.)	18	40-59 year-old Caucasians (9 ≤15 y.e vs. 9 ≥16 y.e.)	Group x Educ. 2-way ANOVA
Nondemented Subgroup	8	All	9	40-59 y.o. Caucasians w ≤15 y.e.	1-way ANOVA
	7	40-59 y.o. w ≤15 y.e.	9	40-59 y.o. Caucasians w ≤15 y.e.	1-way ANOVA

BFRT

HD Group	n	HD Subject Breakdown:	n	HC Subject Breakdown:	Analysis:
Whole	18	All	103	All	1-way ANOVA
Nondemented Subgroup	8	All	103	All	1-way ANOVA

ICP (by education)

HD Group	n	HD Subject Breakdown:	n	HC Subject Breakdown:	Analysis:
Whole	18	All (13 ≤15 y.e vs. 5 ≥16 y.e.)	103	All (58 ≤15 y.e vs. 45 ≥16 y.e.)	Group x Educ. 2-way ANOVA
Nondemented Subgroup	8	All	58	w ≤15 y.e.	1-way ANOVA
	7	w ≤15 y.e.	58	w ≤15 y.e.	1-way ANOVA

NESID (by age, gender, ethnicity, and education)

HD Group	n	HD Subject Breakdown:	n	HC Subject Breakdown:	Analysis:
Whole*	18	All (13 ≤15 y.e vs. 5 ≥16 y.e.)	18	40-59 y.o. Caucasians (9 ≤15 y.e vs. 9 ≥16 y.e.)	Group x Educ. 2-way ANOVA
	14	40-59 y.o. (11 ≤15 y.e vs. 3 ≥16 y.e.)	18	40-59 y.o. Caucasians (9 ≤15 y.e vs. 9 ≥16 y.e.)	Group x Educ. 2-way ANOVA
Nondemented Subgroup	8	All	31	Caucasian women	1-way ANOVA
	6	Females	31	Caucasian women	1-way ANOVA
	8	All	9	40-59 y.o. Caucasians w ≤15 y.e.	1-way ANOVA
	7	40-59 y.o. w ≤15 y.e.	9	40-59 y.o. Caucasians w ≤15 y.e.	1-way ANOVA

* Gender served as its own control by virtue of there being equal numbers of men and women in the overall HD group.

FIDDIFF (by age)

HD Group	n	HD Subject Breakdown:	n	HC Subject Breakdown:	Analysis:
Whole	18	All	32	40-59 y.o.	1-way ANOVA
	14	40-59 y.o.	32	40-59 y.o.	1-way ANOVA
Nondemented Subgroup	8	All	32	40-59 y.o.	1-way ANOVA
	7	40-59 y.o.	32	40-59 y.o.	1-way ANOVA

PIDDIFF (by age)

HD Group	n	HD Subject Breakdown:	n	HC Subject Breakdown:	Analysis:
Whole	18	All	32	40-59 y.o.	1-way ANOVA
	14	40-59 y.o.	32	40-59 y.o.	1-way ANOVA
Nondemented Subgroup	8	All	32	40-59 y.o.	1-way ANOVA
	7	40-59 y.o.	32	40-59 y.o.	1-way ANOVA

SIDDIFF (by age, gender, ethnicity, and education)

HD Group	n	HD Subject Breakdown:	n	HC Subject Breakdown:	Analysis:
Whole*	18	All (13 ≤15 y.e vs. 5 ≥16 y.e.)	18	40-59 y.o. Caucasians (9 ≤15 y.e vs. 9 ≥16 y.e.)	Group x Educ. 2-way ANOVA
	14	40-59 y.o. (11 ≤15 y.e vs. 3 ≥16 y.e.)	18	40-59 y.o. Caucasians (9 ≤15 y.e vs. 9 ≥16 y.e.)	Group x Educ. 2-way ANOVA
Nondemented Subgroup	8	All	31	Caucasian women	1-way ANOVA
	6	Females	31	Caucasian women	1-way ANOVA
	8	All	9	40-59 y.o. Caucasians w ≤15 y.e.	1-way ANOVA
	7	40-59 y.o. w ≤15 y.e.	9	40-59 y.o. Caucasians w ≤15 y.e.	1-way ANOVA

* Gender served as its own control by virtue of there being equal numbers of men and women in the overall HD group.

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