DOES THE NONVERBAL LEARNING DISABILITIES (NLD) SCALE DISTINGUISH BETWEEN SUBTYPES OF PERVASIVE DEVELOPMENTAL DISORDER?

by
Chand Taneja
B.Sc. University of Victoria, 1996

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ABSTRACT

The NLD Scale, developed by Rourke in 1993, is an instrument designed to assist in the diagnosis of nonverbal learning disabilities (NLD). The purpose of the present investigation, which uses the NLD Scale, was twofold. The first goal was to evaluate the validity and internal consistency of the NLD Scale, which is a relatively new tool. The second goal was to determine if the NLD Scale could distinguish between individuals from 3 different subtypes of pervasive developmental disorder (PDD): (1) high-functioning autism (HFA), (2) Asperger syndrome (AS) and (3) pervasive developmental disorder-not otherwise specified (PDD-NOS). This was done in order to investigate recent findings that the NLD neuropsychological profile is characteristic of persons with AS, but not those with HFA (e.g., Klin, Volkmar, Sparrow, Cicchetti & Rourke, 1995). A preliminary test of the validity of the NLD Scale appears to be adequate, indicating that the NLD Scale appears to be a useful tool to identify symptoms characteristic of the NLD syndrome. However, due to the broad range of behavioural characteristics analyzed by the NLD Scale, it has somewhat low internal consistency. Next, the PDD groups were compared using one-way between groups ANOVAs and Jonckheere's test of trend on each of the following NLD Scale measures: (1) neuropsychological functioning, (2) academic achievement, (3) social-emotional and adaptive functioning, (4) total scores, and (5) individual questions. Group differences were significant on the section of the NLD Scale measuring neuropsychological functioning, verifying recent findings that individuals with AS, but not HFA, share characteristics with the NLD syndrome. However, subtypes of PDD were not differentiated by means of academic achievement, social-emotional and adaptive functioning or total NLD Scale scores. Also, the PDD-NOS group did not differ significantly from either of the two other groups on any of the NLD Scale sections. In addition, few individual questions significantly differentiated the three groups. Future research and clinical use of the NLD Scale are encouraged.

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CHAPTER I

INTRODUCTION

High-Functioning Autism, Asperger Syndrome and Pervasive Developmental Disorder Not Otherwise Specified

Background

In 1943, Leo Kanner, a psychiatrist in Baltimore, was the first to clearly describe children with "autistic disturbances of affective contact," later referred to as autism. Kanner provided detailed descriptions of children who displayed difficulties with communication and socialization, resisted change in the environment, demonstrated stereotypic behaviours and often had some isolated interest or ability.

Unaware of Kanner's work, the Viennese pediatrician, Hans Asperger, was concurrently studying a similar disorder that he called "autistic psychopathy" (Asperger, 1944/1991). Asperger's description was different from Kanner's; for example, speech was less commonly delayed, motor impairments were more common, cognitive deficits were not apparent, only males were affected, and onset was later than found in autism. Yet, both Asperger and Kanner used the term "autistic" to signify extreme egocentrism and withdrawal from other people. This sharing of terminology may have ignited a controversy concerning the relationship between these disorders.

Since Kanner's introduction to autism, there has been an abundance of research focused on this disorder. However, there was little direct mention of Asperger's original report until Lorna Wing's 1981 clinical account of what she called "Asperger's syndrome" (Wing, 1981). Wing modified Asperger's description somewhat; for example, she included observations of its existence in females as well as males, and the occasional appearance of language problems and mental retardation. Subsequently, research in this area has grown dramatically, and the term "Asperger syndrome" (AS) has become part of

the diagnostic nomenclature (American Psychiatric Association, 1994; World Health Organization, 1993).

By definition, high-functioning autism (HFA), a subtype of autism where there are no signs of mental retardation, differs from AS in terms of language functioning. Nevertheless, because it appears that both these syndromes share several features, they have traditionally been placed on a continuum of severity, with AS considered to be less severe than HFA. At present, the distinction between AS and HFA is not clearly agreed upon (Ozonoff, South, & Miller, 2000). However, recent research is revealing qualitative differences between AS and HFA. These conditions appear to have unique patterns of strengths and weaknesses, leading to the notion that a continuum of severity alone may not adequately describe the relationship between AS and HFA. For example, it has been found that they differ on measures including, language development and competence, adaptive behaviours, (e.g., Szatmari, Archer, Fisman, Streiner, & Wilson, 1995), visual-spatial functioning, visual memory (e.g., Klin, Volkmar, Sparrow, Cicchetti, & Rourke, 1995), psychomotor skills (e.g., Iwanaga, Kawasaki, & Tsuchida, 2000; Klin et al., 1995; Ozonoff, Rogers, & Pennington, 1991; Szatmari, Tuff, Finlayson, & Bartolucci, 1990), "theory of mind" deficits, "executive" dysfunction (e.g., Ozonoff et al., 1991) and prognosis (e.g., Szatmari, Bartolucci, & Bremner, 1989). Nevertheless, some researchers including Szatmari and colleagues still conclude that there is a lack of differentiation in neurocognitive functioning, and so AS and HFA should be categorized together rather than as separate syndromes.

Justifying the division of diagnostic terms, Klin et al. (1995) report several areas of differences between the HFA and AS. For example, a relative strength in verbal functioning, but a relative weakness in visual-spatial performance was displayed by the AS group, and essentially opposite findings were demonstrated by the HFA group. In

addition, there were differences between the groups on fine and gross motor skills, nonverbal concept formation, visual and verbal memory, articulation, vocabulary and verbal output. Similarly, Sparrow, Klin, Volkmar, Cicchetti and Rourke (unpublished) found that when analyzing preschool evaluations and information acquired from the Autism Diagnostic Interview-Revised (Lord, Rutter, & LeCouteur, 1994), distinctions were found between the AS and HFA groups that corresponded to findings by Klin et al. (1995). For example, the AS group demonstrated social awkwardness, verbal precociousness, visual and/or visual-motor deficits, and all absorbing circumscribed interests. The HFA group was characterized by social withdrawal, verbal delays, auditory-verbal deficits and pronounced motor stereotypies.

One reason for the discrepancy in conclusions made by researchers may be the use of different diagnostic criteria employed by various researchers in the field autism. Klin et al. (1995) adopted a more stringent diagnostic procedure, whereas Szatmari and colleagues appear to embrace a broader definition of AS.

A third disorder, which is quite similar to HFA and AS is called pervasive developmental disorder-not otherwise specified (PDD-NOS). At present this condition is poorly defined, but in general, individuals with this condition have some features of autism, such as unusual sensitivities and difficulties with social interaction, but they do not meet all the criteria for either a diagnosis of autism or AS.

Past research has shown several similarities between PDD-NOS and AS. For example, Kurita (1997) found that individuals with comparable psychometric intelligence diagnosed with AS or PDD-NOS were similar on various measures including obstetrical risk factors, early developmental milestones (e.g., motor landmarks), and EEG abnormalities. Although, Kurita's finding that the AS group was "less autistic" than the PDD-NOS group according to scores on the Tokyo version of the Childhood Autism

Rating Scale (CARS), caution should be taken when interpreting results, because additional studies are still required to replicate these findings.

Thus far, most neurobiological information on AS and PDD-NOS has been extrapolated from work done on autism. This was necessitated by the fact that research specifically addressing AS and PDD-NOS is minimally represented in the literature. Because neurobiological studies generally cannot clearly differentiate between these conditions at this point in time, one is forced to rely on neuropsychological data to understand the qualitative distinctions between these conditions.

One area of current research in AS is understanding its association with the nonverbal learning disabilities (NLD) syndrome. The term "nonverbal learning disabilities", originally coined by Myklebust in 1975, has become a subject of much research (e.g., Rourke, 1989, 1995). The NLD syndrome is characterized by a profile of neuropsychological assets and deficits including, intact verbal processing in the presence of deficits in visual-spatial functioning and significant difficulties in social interactions. Unlike autism, which is speculated to be associated with left hemisphere dysfunction (e.g., Dawson, Finley, Phillips, & Galpert, 1986; Rumsey, 1992), AS and the NLD syndrome are thought to be related to underlying right hemisphere dysfunction (Gunter, Ghaziuddin, & Ellis, unpublished; McKelvey, Lambert, Mottron, & Shevell, 1995; Rourke, 1989, 1995). Therefore, associating AS with NLD appears to provide a comprehensible means of distinguishing AS from HFA.

In order to describe the areas of phenomenological overlap and divergence between HFA, AS, and PDD-NOS, this paper provides a summary of background information including current neurobiological and neuropsychological findings on these three disorders. There will be a focus on the controversial relationship between AS and HFA. and an attempt to examine their relationship to PDD-NOS, an infrequently studied

condition. Also, the close association between AS and the syndrome of NLD will be reviewed. Properties including, validity and internal consistency of the NLD Scale (Rourke, 1993), a fairly new instrument designed to identify individuals with NLD, will be investigated. And finally, the discriminatory power of the NLD Scale in respect to HFA, AS, and PDD-NOS, will be evaluated.

Definitions and Associated Features of PDDs

Pervasive Developmental Disorder. Pervasive developmental disorder (PDD) is not itself a diagnosis, but instead a category that encompasses a number of syndromes. Autism, AS, Rett's Disorder, Childhood Disintegrative Disorder and PDD-NOS are often collectively referred to as PDDs or autistic spectrum disorders (American Psychiatric Association, 1994, 2000). PDDs may be found in up to one percent of the school age population (Kadesjo, Gillberg, Hagberg, 1999). The main characteristics of PDDs include impairments in empathy and social understanding, difficulties with communication, and a restricted range of behaviours and interests (American Psychiatric Association, 1994, 2000; World Health Organization, 1993). Symptoms may overlap with other disorders including: semantic-pragmatic language disorder, obsessive-compulsive personality disorder (OCPD), social phobia, schizotypal personality disorders, paranoid personality disorders and possibly childhood-onset schizophrenia (Gillberg, 1998; Rapin & Katzman, 1998).

The use of explicit operational definitions is important when attempting to distinguish between subtypes of PDD. Thus far, there has been much ambiguity in defining AS and PDD-NOS, and also understanding the extent of overlap between autism, AS and PDD-NOS (Gillberg, 1989, 1998; Wing, 1981). To add to the confusion, researchers including Wing (1981) and Gillberg (1998) assert that a diagnosis may change over time. For example, a child may be considered to have symptoms of autism at

one point in time and symptoms of AS at another date. Presently, this viewpoint is considered controversial, and is an issue that requires further research.

Autistic Disorder and High-Functioning Autism. Autistic disorder (as labeled in the DSM-IV) or Childhood Autism (as labeled in the ICD-10) is more commonly known as autism, and is the most severe of the PDDs. It is characterized by three main areas of disturbance: (1) impairments in social interaction, (2) impairments in communication skills, and (3) restricted, repetitive and stereotyped patterns of behaviour (see Table 1). Within each of these three broad areas more specific criteria are also found (American Psychiatric Association, 1994, 2000).

Impairments in social interaction may be demonstrated by difficulties with gestures, facial expressions, obstacles forming usual peer relations, difficulties with spontaneous sharing of interests with others, and a lack of social-emotional reciprocity. Impairments in communication may be marked by a delay or complete lack of the development of speech, which is not accompanied by gestures or other attempts to communicate. In individuals with autism who are able to speak, there is often difficulty maintaining reciprocal conversations and their language may be characterized by stereotyped and repetitive use of words, or unusual pitch, intonation, rate or rhythm. Behavioural abnormalities may include fixation with restricted interests, rigid adherence to routines, stereotyped and repetitive movements (e.g., hand-flapping), and unusual preoccupation with parts of objects. In addition, features of autism are seen in children by 3 years of age. Also, symptoms are not better described by any other disorder.

The estimated prevalence for children with autism is 1 case per 2000 individuals (American Psychiatric Association, 2000); however, rates as high as 1 case per 1000 individuals have been reported (Gillberg & Wing, 1999). Differences in rates of prevalence are possibly due to varying criteria used (e.g., DSM-IV, ICD-10 or other

Table 1

Diagnostic Criteria for Autistic Disorder

A. A total of six (or more) items from (1), (2), and (3), with at least two from (1), and one each from (2) and (3):

(1) qualitative impairment in social interaction, as manifested by at least two of the following:

(a) marked impairments in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body posture, and gestures to regulate social interaction

(b) failure to develop peer relationships appropriate to developmental level

(c) a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people, (e.g., by a lack of showing, bringing, or pointing out objects of interest to other people)

(d) lack of social or emotional reciprocity

(2) qualitative impairments in communication as manifested by at least one of the following:

(a) delay in, or total lack of, the development of spoken language (not accompanied by an attempt to compensate through alternative modes of communication such as gesture or mime)

(b) in individuals with adequate speech, marked impairment in the ability to initiate or sustain a conversation with others

(c) stereotyped and repetitive use of language or idiosyncratic language

(d) lack of varied, spontaneous make-believe play or social imitative play appropriate to developmental level

(3) restricted repetitive and stereotyped patterns of behavior, interests and activities, as manifested by at least two of the following:

(a) encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus

(b) apparently inflexible adherence to specific, nonfunctional routines or rituals

(c) stereotyped and repetitive motor mannerisms (e.g., hand or finger flapping or twisting, or complex whole-body movements)

(d) persistent preoccupation with parts of objects

B. Delays or abnormal functioning in at least one of the following areas, with onset prior to age 3 years:

(1) social interaction

(2) language as used in social communication

(3) symbolic or imaginative play

C. The disturbance is not better accounted for by Rett's Disorder or Childhood Disintegrative Disorder.

Note. From "Diagnostic and statistical manual of mental disorders, (4th ed.), Text Revision (DSM-IV-TR)". By American Psychiatric Association (APA), 2000, p. 75.

modified criteria). Those with autism have a male to female ratio of approximately 4:1 (Gillberg, 1995), although, the male to female ratio is lower in subpopulations of autism with severe to profound mental retardation.

High-functioning autism (HFA) is a category of autism without signs of mental retardation, determined by a psychometric intelligence score (IQ) generally above 70. Cases of HFA constitute less than 25% of autism cases (American Psychiatric Association, 1994).

Asperger Syndrome. Diagnostic criteria for AS are generally adopted from the ICD-10 (World Health Organization, 1993) or the DSM-IV (American Psychiatric Association, 1994, 2000). Previous to the publication of the DSM-IV diagnostic criteria for AS (known as Asperger's Disorder in the DSM-IV), it was very difficult to define this condition. Whereas DSM-IV criteria may not be perfect, they provide clear, standardized inclusion and exclusion criteria for AS. The DSM-IV clinical criteria for AS include impairments in social interaction and restricted repetitive and stereotyped patterns of behavior as described in autism (see Table 2). These disturbances are stated to cause impairments in functioning (e.g., occupational and social functioning). Unlike autism, there is no clinically significant delay in the development of language, cognitive skills or self-help skills. Other than difficulties with reciprocal social exchanges, adaptive behavior and curiosity about the environment are not affected. In addition, criteria are not met for any another disorder. According to the ICD-10, motor clumsiness and isolated special skills, often related to abnormal preoccupations, are also common but not necessary for diagnosis.

AS clearly differs from the majority of autism cases where signs of mental retardation are present, and language skills and prognosis are poorer. However, it is more difficult to distinguish AS from HFA due to somewhat similar cognitive abilities. One

Table 2

Diagnostic Criteria for Asperger's Disorder

A. Qualitative impairment in social interaction, as manifested by at least two of the following:

(1) marked impairment in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction

(2) failure to develop peer relationships appropriate to developmental level

(3) a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g., by a lack of showing, bringing, or pointing out objects of interest to other people)

(4) lack of social or emotional reciprocity

B. Restricted repetitive and stereotyped patterns of behavior, interests, and activities, as manifested by at least one of the following:

(1) encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus

(2) apparently inflexible adherence to specific, nonfunctional routines or rituals

(3) stereotyped and repetitive motor mannerisms (e.g., hand or finger flapping or twisting, or complex whole-body movements)

(4) persistent preoccupation with parts of objects

C. The disturbance causes clinically significant impairment in social, occupational, or other important areas of functioning.

D. There is no clinically significant general delay in language (e.g., single words used by age 2 years, communicative phrases used by age 3 years).

E. There is no clinically significant delay in cognitive development or in the development of age-appropriate self-help skills, adaptive behavior (other than in social interaction), and curiosity about the environment in childhood.

F. Criteria are not met for another specific Pervasive Developmental Disorder or Schizophrenia.

Note. From "Diagnostic and statistical manual of mental disorders, (4th ed.), Text Revision (DSM-IV-TR)". By American Psychiatric Association (APA), 2000, p. 84.

method of differentiating these conditions is based on profiles of verbal and visual-spatial functioning. AS is characterized by significantly better verbal functioning compared to visual-spatial functioning, in contrast to HFA, in which nonverbal skills are higher or not significantly different from verbal skills (Ehlers et al., 1997; Klin et al., 1995; Volkmar et al., 1994). It should be noted that although language is relatively good in AS, speech might still be odd. For example, inflection and intonation may not be as rigid and monotonic as in autism, but speech may be marked by poor prosody or may even be incoherent. In contrast to HFA, AS is also usually characterized by marked verbosity. Also motor mannerisms are more common in HFA, whereas preoccupations with specific topics are more commonly observed in AS (American Psychiatric Association, 2000; Volkmar et al., 1994). Finally, social interactions are more frequently attempted by persons with AS compared to those with HFA (American Psychiatric Association, 2000).

Due to past problems defining AS, determining its prevalence has been complex. In general, AS has been found to be more common than autism. In 1993, Ehlers and Gillberg found a prevalence of 36 to 71 per 10 000 for suspected cases of AS, and a male to female ratio estimated of approximately 4:1. Kopp and Gillberg (1992) suggest that girls experience less obvious impairments, because their social skills are usually better when compared to boys.

Symptoms of AS may overlap with features from several different psychiatric disorders outside of the autistic spectrum (Ghaziuddin, Weidmer-Mikhail & Ghaziuddin, 1998). Young children with AS are likely to exhibit externalizing problems, particularly attention-deficit hyperactivity disorder, whereas adolescents and adults often experience internalizing disorders, most commonly depression (Gillberg & Gillberg, 1989). In addition, Ehlers and Gillberg (1993) found that tics and occasionally Tourette's syndrome were seen in children with AS. There is also an overlap in symptoms of the NLD

syndrome and AS, but not between NLD and HFA (Brumback, Harper, & Weinberg, 1996; Klin et al., 1995; Rourke & Tsatsanis, 2000; Sparrow et al., unpublished). This will be discussed in detail in a following section of this paper. There may also be an overlap between NLD and PDD-NOS because many features of AS and PDD-NOS are shared (e.g., Kurita, 1997).

Pervasive Developmental Disorder Not Otherwise Specified. PDD-NOS is classified along with AS in the DSM-IV, rather than having its own operationalized definition (American Psychiatric Association, 1994, 2000). PDD-NOS, known as atypical autism in ICD-10, is a poorly defined diagnosis that involves some symptoms of autistic disorder without meeting its full criteria, or for that matter, criteria for any other disorder (Mahoney et al., 1998). There are many similarities between PDD-NOS and other forms of PDD including impairment in the development of social interaction and communication skills, and stereotyped behaviors, interests and activities may be present. According to the DSM-IV, PDD-NOS can be differentiated from AS and autism because those with PDD-NOS experience a later age of onset, atypical or subthreshold symptomatology, or all of the above (American Psychiatric Association, 1994, 2000). Most often those with PDD-NOS are less severely socially dysfunctional than those with AS or autism. It is reportedly found in approximately 2 to 4 in 10 000 children (Gillberg, 1995), but due to poorly defined diagnostic criteria its prevalence cannot be certain. Similarly, interpreting research studies is also complicated by the lack of specific guidelines for this condition.

The Syndrome of Nonverbal Learning Disabilities

It is commonly stated that the left cerebral hemisphere usually underlies the use and understanding of verbal material, whereas the right cerebral hemisphere appears to be responsible for visual-spatial functioning, including nonverbal communication. Features

of AS, but not HFA, are remarkably similar to right-hemisphere dysfunctions. The "right-hemispheric dysfunction syndrome" (Semrud-Clikeman & Hynd, 1990) better known as "the syndrome of Nonverbal Learning Disabilities" (NLD) follows a complex pattern and involves several assets and deficits in functioning (Rourke, 1989,1995). The NLD neurocognitive profile is found in individuals suffering from a variety of neurological and neuroendocrine disorders. These include, some forms of hydrocephalus, callosal agenesis, congenital hypothyroidism, William syndrome, AS, and various forms of white matter and/or right hemisphere tissue damage (detailed information about the clinical manifestations of these disorders can be found in Rourke, 1995). This section reviews NLD and discusses its relationship with AS. Research evidence will be provided to illustrate the correspondence between neuropsychological strengths and weaknesses of these syndromes.

Characteristics of the NLD Syndrome

The subtype of learning disability known as the NLD syndrome is comprised of a number of neuropsychological assets and deficits (Rourke, 1989, 1995; Rourke & Tsatsanis, 2000). According to Rourke (1989, 1995), a pattern of neuropsychological assets and deficits can lead to a particular pattern of academic strengths and weaknesses in addition to specific psychosocial difficulties. Further information on the dynamics of NLD can be found in Rourke's NLD model, which integrates cognitive, brain maturational and experiential factors (see Rourke, 1989, pp.80-110; Rourke, 1995, p.6-18). The principal assets and deficits as described by Rourke are as follows:

Assets. Neuropsychological assets include psycholinguistic skills such as rote verbal learning and memory skills, regular phoneme-grapheme matching, a high amount of verbal output (e.g., verbosity of a repetitive, straightforward, rote nature), and verbal classification. In addition to advanced levels of verbal memory and attention, auditory

perception may also be an asset. Also, even though complex psychomotor tasks may be difficult for these individuals, simple motor activities may in contrast seem easy for individuals with NLD. Graphomotor skills may become an asset in later years, and with regard to academics, word recognition, spelling, and verbatim memory may be advanced compared to other measures of scholastic achievement.

Deficits. Deficits found in NLD include difficulties with visual-spatial organization. This is especially true of material that requires working within a novel framework and information that cannot be verbalized.

Tactile-perceptual deficits may be common in persons with NLD, especially on the left side of the body. Simple tactile perception may improve with age; although, complex tactile perceptual tasks continue to be problematic. Also, bilateral psychomotor coordination deficiencies, again more marked on the left side of the body may be apparent in individuals with NLD. Simple motor skills (e.g., finger tapping and static steadiness) may improve with age, whereas relatively more difficult psychomotor tasks (e.g., those requiring novelty) tend to worsen with age compared to same-aged peers. Graphomotor skills may also be a deficit in early years for these children. Poor psychomotor skills, in addition to general clumsiness may contribute to their social awkwardness.

Individuals with NLD have trouble dealing with novel and complex situations, in part due to their over-reliance on prosaic, rote behaviours. Difficulty with novel experiences is likely to continue, if not worsen, with age. Nonverbal problem-solving, concept formation skills, and understanding informational feedback in novel or otherwise complex situations, may also be difficult for persons with NLD. Deficits in understanding cause-and-effect relationships and appreciating incongruities may result in a difficulty for them to understand humor. Again, these difficulties may persist or even worsen with age.

Language content may be unusual and pragmatics may be poor in individuals with NLD. Also, their speech may be characterized by having minimal prosody. Due to a strength in language in comparison to nonverbal skills, children and adults with NLD may overly rely on verbal communication in social interactions instead of incorporating gestures and other forms of nonverbal communication.

Individuals may have a distorted sense of time, whereby they may not be able to estimate the duration of time of a common activity, or they may make poor estimates of the time of day.

As a result of neuropsychological deficits, individuals with NLD may have difficulty with academic achievement in the areas of mechanical arithmetic, mathematics, science, and reading comprehension. Although rote memorization is one of their strengths, higher-level mathematics requires spatial visualization and representation, which may be difficult for persons with NLD.

Individuals with NLD exhibit deficits in social interaction skills, marked by difficulties appreciating nonverbal forms of communication and adapting to novel situations. These social-emotional problems may arise, in part, due to the interaction of the neuropsychological difficulties outlined above. For example, problems with visual-perceptual organizational skills may result in difficulty interpreting facial expressions and gestures, thus contributing to their problems interacting socially. Likewise, poor reasoning and concept formation skills may contribute to adaptive difficulties such as misinterpreting emotional state from facial expressions and failing to understand humor. These individuals may exhibit activity levels that deviate from the norm; for example, they are often hyperactive during early childhood and become increasingly hypoactive with increasing age. Furthermore, social withdrawal, social isolation and risk for

"internalized" forms of psychopathology (e.g., depression and suicide attempts) may develop during late childhood and adolescence, in part due to social pressure and ridicule.

White Matter Model

According to Rourke (1989, 1995), difficulties experienced by individuals with NLD result from a neurodevelopmental disorder explained in the "White Matter Model". Essentially, white matter destruction or dysfunction is presumed to cause the symptoms of NLD, with increasing damage resulting in higher risks of NLD. The clinical presentation of the NLD syndrome results from the lesion type and the stage of development when the lesion occurs.

The right hemisphere is particularly affected because it has a higher ratio of white matter to gray matter, and utilizes intermodal integration across long communication pathways more than the left hemisphere does. Because the right and left hemispheres have distinct roles, individuals with NLD may have difficulty with tasks that the right hemisphere is thought to be responsible for (Rourke & Tsatsanis, 2000). For example, the right hemisphere has been said to assemble descriptive systems (e.g., strategies to decode language or music), whereas the left hemisphere integrates and applies these well-formed descriptive systems (Goldberg & Costa, 1981). Furthermore, right hemisphere functions may involve processing novel events and learning via experimentation or inductive reasoning, whereas the left hemisphere deals with predictable or routine events, and learns by deductive construction of preexisting knowledge (Goldberg & Costa, 1981). Thus, damage to the right hemisphere, as found in NLD, results in difficulty with right hemisphere functions, such as dealing with novelty or comprehending abstractions and generalization, and using inductive reasoning (Rourke & Tsatsanis, 2000).

Relationship between NLD and PDDs

The syndrome of NLD and AS appear to be closely related disorders. Therefore, the NLD model appears to be useful in explaining symptoms found in AS. Developmental factors which cause disturbances in the neurodevelopment of white matter may subsequently result in symptoms characteristic of AS (Ellis & Gunter, 1999; Rickerby et al., 1991). The White Matter Model proposed by Rourke (1989, 1995) accounts well for AS, although the etiology of AS still remains unclear. Neuroimaging studies often support the white matter model by revealing abnormalities in the right hemisphere and corpus callosum of patients with AS (Berthier, Bayes, & Tolosa, 1993; David, Wacharasindhu, & Lishman, 1993; McKelvey et al., 1995). The following section reviews the assets and deficits found in both AS and NLD (Rourke & Tsatsanis, 2000).

Assets of children with AS that also fit in the NLD syndrome include higher VIQ than PIQ, marked verbosity, and strengths in rote verbal-memory skills, grammar, single word reading and spelling. Although simple motor skills are usually considered a strength in children with NLD, this is not usually true of children with AS.

Numerous characteristic deficits in AS are also consistent with core deficits exhibited in the syndrome of NLD. They include: (1) impaired reciprocal social interactions (pedantic one-sided conversations, limited understanding of humor, and difficulty forming friendships), (2) atypical nonverbal communication (abnormalities in prosody and emotionality in speech and gestures, and impaired face recognition and expression), (3) stereotyped behaviours, (4) motor impairments (e.g., motor incoordination and/or clumsiness), (5) difficulties with problem solving, (6) poor reading comprehension, and (7) difficulty adapting to novel situations. Also common to both AS and NLD are emotional disturbances in particular, externalizing problems such as attention-deficit hyperactivity disorder in children, and internalizing problems including

depression in adolescence and adulthood (Ghaziuddin, Weidmer-Mikhail, & Ghaziuddin 1998). Impairments in arithmetic and bilateral tactile-perception are characteristic of NLD, but remain relatively unexplored in AS (Ellis & Gunter, 1999).

Characteristic strengths and weakness found in neuropsychological assessments show that children with AS probably have right hemisphere dysfunction and clearly fall under the classification of NLD (Ellis, Ellis, Fraser, & Deb, 1994; Klin et al., 1995; Rourke, 1989, 1995; Rourke & Tsatsanis, 2000). For example, Ellis et al., 1994 noted that individuals with AS exhibited symptoms characteristic of the syndrome of NLD, including difficulty judging social situations and strengths in verbal functioning.

Klin et al. (1995) were able to distinguish AS from HFA using NLD as a neuropsychological model of AS. They examined 19 individuals with HFA and 21 individuals with AS. All subjects had Full Scale IQ's (FSIQ's) over 70. Two clinical neuropsychologists independently rated the individuals' performance on 22 items thought to be strengths and weaknesses in Rourke's NLD model. They found higher VIQ and lower PIQ for the AS group, but not any significant verbal-performance discrepancy in scores for the HFA group. Also, they detected a high correspondence in the strengths and weaknesses characteristic of the NLD profile in all individuals in the AS group. In contrast, only one of the individuals with HFA exhibited the NLD profile. For example, researchers found that individuals with AS had several areas of deficit characteristic of NLD including deficits in fine and gross motor skills, visual-motor integration, visual-spatial perception, nonverbal concept formation, and visual memory. Also similar to NLD, AS was not associated with deficits in articulation, verbal output, auditory perception, vocabulary, and verbal memory. Overall, this study did show an overlap in neuropsychological strengths and weakness between AS and NLD but not between HFA and NLD.

Although, children fitting into a diagnosis of AS generally also meet the criteria for NLD, this is not true of individuals with HFA. For example, children with HFA do not have a substantially stronger verbal than visual-spatial functioning. In fact, either the opposite is true or else the difference is not clinically significant. Thus, the NLD phenotype may help differentiate between AS and HFA (Klin et al., 1995; Rourke & Tsatsanis, 2000). Neuropsychological strengths and weakness in PDD-NOS and AS may be similar, but research is necessary to determine if indeed a relationship between NLD and PDD-NOS exists.

Research studies on white matter development and inter-hemisphere communication are warranted for persons with PDDs. For example, postmortem studies of persons with PDDs, may also determine if brain structures or specific tracts are especially vulnerable. In general, discovering histological evidence and corresponding deficits and capacities should be continued.

The NLD Scale

It appears that the subtypes of PDD are heterogeneous, but overlapping conditions. Various methodologies should be employed to help better understand their similarities and differences. Because of the resemblance between behaviours characteristic of NLD and AS, but not HFA, one means of differentiating AS from HFA is by using criteria for NLD. Similarly, those with PDD-NOS appear to share many characteristic of AS (e.g., Kurita, 1997), and may also be found to display behaviours commonly noted in individuals with NLD.

This study will utilize the NLD Scale, which is composed of items that determine if an individual has behaviours characteristic of NLD, in order to help differentiate and describe subtypes of PDD.

Administration

The NLD Scale was developed by Rourke in 1993 in order to evaluate individuals, as young as seven years of age, with potential nonverbal learning disabilities (see Appendix for a copy of the NLD Scale and scoring criteria). The NLD Scale, constructed to identify the behaviours characteristic of individuals with NLD, is given to a parent or guardian to fill out. The parent answers questions on a three-point scale, stating if a behaviour has been evident or not. For example, they may check off (i) no, not at all; never, (ii) somewhat; every once in a while, or (iii) yes, very much; frequently. It is an advantageous tool because questions are straightforward and administration can be completed within a short period of time. Although the NLD Scale itself provides useful information, and may be an effective instrument to screen for NLD, it is especially useful when included as part of a complete neuropsychological assessment.

Measures

The NLD Scale has a total of 40 questions, which may be separated into three sections:

1. Neuropsychological functioning (questions 1-23). The first section has 23 questions tapping into various aspects of the child or adolescent's behaviour, such as verbal and visual-spatial functioning, memory and attention, sensitivity to sounds, motor skills, exploration of the environment, problem-solving abilities, understanding of time, and dealing with novelty and social skills.

2. Academic achievement (questions 24-30). The second section is comprised of seven questions related to academic achievement. This section is used to record observations of achievement in: neatness of handwriting, single word reading, spelling, rote memory, reading comprehension, arithmetic and concept formation.

3. Social-emotional and adaptive functioning (questions 31-40). The third section consists of ten questions on social, emotional and adaptive behaviours. For example, the reporter notes how the child interacts with same-age peers, adults and family members. Also, physical activity level and emotion in social situations is recorded.

Reliability of the NLD Scale

Although the NLD Scale appears to have potential benefit in clinical practice, its use in research studies is in its early stages. Van der Vlugt and his colleagues in the Netherlands have initiated investigations on the utility of this tool. Three studies on test-retest reliability have recently resulted in acceptable reliability coefficients (e.g., Study 1, n = 400, reliability coefficient = 0.82; Study 2, n = 379, reliability coefficient = 0.87; Study 3, n = 290, reliability coefficient = 0.92). In these studies, parents of children with NLD responded to NLD Scale questions on two occasions three weeks apart. Study 1 also consisted of a test of interrater reliability. When the NLD Scale was given to both a parent and a teacher an interrater reliability coefficient of 0.67 was obtained (van der Vlugt, unpublished). Hopefully the present study will support ongoing studies in order to help establish the usefulness of the NLD Scale.

Research on the Neurobiology of Pervasive Developmental Disorders

Although the etiologies of HFA, AS, and PDD-NOS are presently unknown, they are considered to be neurodevelopmental disorders. The following section provides a brief review of the nature of the involvement of brain dysfunction in the origins of PDD. Results from neuroimaging and laboratory studies are examined, and the contribution of genetic factors is also discussed. Present neurobiological studies have not been successful in distinguishing HFA, AS, and PDD-NOS in part because research on AS and PDD-NOS is relatively infrequent (Schultz, Romanski, & Tsatsanis, 2000). A reliance on

information from studies of autistic disorder is of some benefit, although this creates difficulty when attempting to distinguish the conditions qualitatively.

Brain Dysfunction and Medical Conditions

There are differences in obstetrical factors between cases of PDD (Ghaziuddin, Shakal, & Tsai, 1995). Males with AS have lower Apgar scores at one minute after birth, but are less irritable and have better muscle tone than neonates with HFA. In addition, mothers of infants with AS are more often older (above 30 years of age) than the mothers of the infants with HFA. According to Kurita (1997) obstetrical risk factors and early developmental milestones are not substantially different between infants with AS or PDD-NOS.

Factors associated with PDDs are often complex and highly variable. Often cases of AS have indications of major prenatal, perinatal, or postnatal distress (e.g., anoxia, hypoglycemia or hemorrhage related to trauma), some of which include brain dysfunction (Wing, 1981; Gillberg, 1989; Rickerby, Carruthers & Mitchell, 1991). Patients with HFA, and to a lesser extent AS and PDD-NOS, have also been reported to have co-morbid medical disorders including epilepsy, neurocutaneous disorders (e.g., tuberous sclerosis), Marfan-like syndromes, Kleine-Levin syndrome, hypothyroidism, seizures, fragile X syndrome and other chromosomal abnormalities (Bolton & Griffiths, 1997; Bonnet & Gao, 1996; Gillberg, 1998).

EEG and Neuroimaging Findings

Electroencephalogram (EEG) and neuroimaging technology is continuously adding to research on PDDs. Nevertheless, it should be noted that no single abnormality characterizes all subjects with PDD, and in some individuals no irregularities are detected. An overview of some studies follows:

Brain Size, Cerebral Asymmetries, and Ventricular Size. Various abnormalities of brain structure in individuals with autism have been described in past research although findings are not consistent. A study by Hier, LeMay and Rosenberger (1979) revealed that 57% of individuals with autism displayed a pattern of cerebral asymmetry, where their right parietooccipital region was wider than the left. However, subsequent studies of cerebral asymmetry did not replicate these results (Rumsey et al., 1988; Tsai, Jacoby, & Stewart, 1983; Tsai, Jacoby, Stewart, & Beisler, 1982).

Findings suggest that compared to normal controls, male patients with autism have enlarged brains due to both greater brain tissue volume and greater lateral ventricle volume (Piven et al., 1995). This may result from abnormal neuronal proliferation and/or cell death. Jacobson, LeCouteur, Howlin, and Rutter (1988) found brain enlargement was due to larger third ventricles but not lateral ventricles in males with autism. However, another study revealed that there was no significant difference between the volumes of the third ventricle or lateral ventricles in cases of autism compared to controls (Creasey et al., 1986). Interpreting these results is unfortunately hindered due to a lack of replication and inconsistent methodology.

Research on brain enlargement, cerebral asymmetries, and ventricular size is sparse in the AS and PDD-NOS literature, so no conclusions about similarities and distinctions between individuals with different subtypes of PDD can be drawn.

Temporal Lobe and Limbic System. When looking at subcortical structures, Creasey et al. (1986) noted no notable difference between the volumes of cerebrospinal fluid, white matter, gray matter, the caudate nuclei, lenticular nuclei, or thalami, in males with autism compared to controls. However, subtle cellular changes have been consistently found in areas of the limbic circuits including the hippocampus and amygdala (Rapin & Katzman, 1998). These changes may be linked to the difficulties in

social interaction, attention, expressing emotion, learning and motivation in those diagnosed with autism (Delong, 1992).

Anomalies in temporal lobe neurons are also intriguing, especially because they are conceivably related to deficits in the recognition of face and emotion by persons with PDD (Schultz et al., 2000). The amygdala, an area that plays a role in emotional arousal, and its connections to the temporal and prefrontal cortex, are frequently suspected to be abnormal in those with PDD. Lesions to the amygdala in infant monkeys have been found to produce behaviours similar to those depicted in persons with social disabilities (Bachevalier, 1994). As the monkeys matured, they displayed minimal eye contact, withdrew from social interaction and displayed expressionless faces. Even more fascinating, they engaged in motor stereotypies when exposed to stressful social situations.

Baron-Cohen et al. (1999) found evidence of amygdala dysfunction in individuals with AS and HFA during a functional magnetic resonance imaging (fMRI) investigation of social intelligence (e.g., the ability to interpret others' mental states, to empathize with others, and to interact socially). Increased activity was found in the superior temporal gyrus (STG), the prefrontal cortex and the amygdala of controls without a PDD. However, those with AS and HFA activated fronto-temporal regions but not the amygdala when making cognitive inferences from pictures of eyes. These researchers proposed that the left amygdala may be critically involved in identifying mental states and emotional information from complex visual stimulation. Perhaps to compensate, persons with AS and HFA rely on temporal structures more for aiding in verbally labeling complex visual stimuli when they have difficulty judging the emotional qualities of the stimuli. The performance of the individuals with AS and HFA were not distinguished from one another, thus future studies are necessary to understand how these

groups may differ from each other, and also from PDD-NOS, in brain structure activation during social situations.

Cerebellar Dysfunction. A study by McKelvey et al. (1995) revealed dysfunction in the right hemisphere of the cerebellum in patients with AS. Similarly, research in the autism literature has inconsistently reported irregularities in the cerebellum. For example, hypoplasia of the cerebellum in patients with HFA has been reported by some investigators (e.g., Courchesne, Yeung-Courchesne, Press, Hesselink, & Jernigan, 1988; Hashimoto et al., 1995) whereas others found an increase in cerebellar volume (e.g., Piven, Saliba, Bailey, & Arndt, 1997). A recent study comparing individuals with autism, AS, and PDD-NOS found no significant difference between-groups in the volume of the cerebellum, midbrain, pons, and medulla (Schultz et al., 1999).

White Matter Abnormalities. A consistent finding in the literature is that the corpus callosum, particularly in the posterior subregions, is smaller in cases of autism compared to normal controls (Egaas, Courchesne, & Saitoh, 1995). Similarly, callosal anomalies have been seen in cases of AS, but further studies may be necessary to replicate these finding (David et al., 1993; Lincoln, Courchesne, Allen, Hanson & Ene, 1998).

The corpus callosum is a region consisting of mostly myelinated neurons. Another area highly associated with myelinated fibers (white matter) is the right cerebral hemisphere. Neuroimaging has shown that right hemisphere dysfunction is common in cases of PDD. This may be linked to the role of the right hemisphere in visual-spatial and social-emotional functioning (Mandal, Mohanty, Pandey, & Mohanty, 1996).

A single photon emission computed topography (SPECT) study revealed hypoperfusion in various regions of the right hemisphere in three patients with AS (McKelvey et al., 1995). Similarly, a magnetic resonance imaging (MRI) study showed

that right hemisphere abnormalities were more common in children with a combined diagnosis of AS and Tourette's syndrome (71%), than in cases of Tourette's syndrome without AS (11%) (Berthier et al., 1993). Researchers of these studies attributed abnormalities to decreased neuronal migration, decreased fiber tract elaboration or neuronal shedding, which corresponds to other investigations which have linked AS to right hemisphere dysfunction and more specifically to the syndrome of NLD (Rourke, 1989).

A recent case study of an 11 year old boy with AS revealed a white matter lesion of the right middle temporal gyrus, an area important in the perception of facial expressions and eye gaze (Volkmar, Klin, Schultz, Rubin, & Bronen, 2000). This abnormality, visualized on repeated MRI evaluations, corresponded to his neuropsychological profile, which indicated a substantial strength in verbal (WISC-III Verbal IQ = 150) compared to nonverbal skills (WISC-III Performance IQ = 116).

Right hemisphere lesions have also been visualized in other cases of PDD. Taylor, Neville and Cross (1999) found that in epilepsy surgery candidates, 6 out of 8 patients with AS, and 10 out of 11 with other autistic spectrum disorders (autistic disorder n = 3, PDD-NOS n = 7, and childhood disintegrative disorder n = 1) displayed right hemisphere abnormality. Unfortunately the sample size was small, and the authors did not mention the specific diagnosis (e.g., autistic disorder, PDD-NOS or childhood disintegrative disorder) of the subject without the right hemisphere lesion, and thus a lesion on the left side. Further MRI research will be useful for understanding the relationship between subtypes of PDD and right hemisphere dysfunction.

Left Hemisphere Dysfunction. The observation that language impairments constitute a major symptom of autism has led some researchers to speculate that this may be a result of brain damage to the left hemisphere (Fein, Humes, Kaplan, Lucci, &

Waterhouse, 1984). A positron emission tomography (PET) study suggested language dominance is atypical in some cases of HFA (Muller et al., 1999). For example, it was noted that compared to normal controls, patients with HFA showed a reversed hemispheric dominance in response to a verbal auditory stimulus, and a reduction in activity of the auditory cortex and cerebellum during acoustic stimulation. Left hemisphere dysfunction is infrequently reported in PDD literature, but it may hypothetically be correlated with verbal communication deficits in HFA, AS, and PDD-NOS. More research would unquestionably be worthwhile.

Frontal Lobe. Frontal cortex abnormalities have been visualized in persons with autism when compared to normal control groups (Carper & Courchesne, 2000). Frontal lobe anomalies visualized on MRI have been thought to result in social-emotional problems, along with motor and executive dysfunction in persons with PDDs (Bonnet & Gao, 1996; Schultz et al., 2000). Irregularities in frontal lobe regions may help explain impairments seen in individuals with PDD such as, "theory of mind" deficits. In PET studies related to theory of mind tasks, young men with AS had increased blood flow in areas of the frontal lobe that were discrepant from areas of activation in normal controls (Happé, Ehlers, & Frackowiak, 1996). Frontal lobe neuroimaging comparing HFA, AS, and PDD-NOS are necessary; however, some studies on theory of mind tasks and their possible relation to frontal lobe functioning are addressed later in this paper.

Biochemical Pathology

Laboratory studies have illustrated abnormalities in various proteins found in the cerebrospinal fluid (CSF) of children with autism and AS. CSF levels of glial fibrillary acidic protein (GFA-p) and ganglioside GM-1, were higher in children with AS compared to controls, but lower than levels found in children with autism (Ahlsen et al., 1993).

Laboratory findings that show cerebrospinal fluid protein levels that distinguish AS from HFA, are intriguing and should be studied further.

There may be synaptic abnormalities in individuals with PDDs. Studies of neurotransmitters and receptors show some atypical distribution of serotonin in brain pathways (Chugani et al., 1996). Dopamine systems have also been implicated in the pathophysiology of autism (McDougle et al., 1998). In addition, atypical melatonin levels, related to difficulties in sleeping, have been found in individuals with a PDD (Nir et al., 1995; Patzold, Richdale, & Tonge, 1998). Whereas such abnormalities in biochemical variables may underlie the pathophysiology of PDD, further research is required to understand if their role differs in individuals with HFA, AS, and PDD-NOS.

Genetic Factors

It has been hypothesized that genetic factors are at least partially responsible for causing PDDs, but the mechanism of genetic transmission has not yet been determined. It is suspected that multiple genetic and environmental factors interact to produce HFA, AS, and PDD-NOS.

Autism has been related to various chromosomal abnormalities including fragile X (Payton, Steele, Wenger, & Minshew, 1989), tuberous sclerosis (Hunt & Dennis, 1987), 15q duplications, deletion of a portion of the 2q37 chromosome (Ghaziuddin & Burmeister, 1999), and susceptibility on chromosomes 13 and 7 (Barrett et al., 1999). Family studies report a higher prevalence of autistic-like symptoms in relatives of an individual with autism compared to the overall population (Bolton & Rutter, 1990). For example, the rate of autism in siblings of affected individuals is approximately 2 to 3%, which is 20 to 50 times greater than the incidence in the general population (Smalley, Asarnow, & Spence, 1988). This is consistent with twin studies, which also support the role of genetics in the pathogenesis of autism (Bolton & Rutter, 1990). Twin studies show

an increased concordance in monozygotic twins compared to dizygotic twins. Discordant twins also often show significant social problems even though they may not meet the criteria for autism. However, not all monozygotic twins are concordant, supporting the influence of non-genetic factors, such as brain dysfunction.

Individuals with AS are relatively less prone to show signs of brain dysfunction than those with autism, and it is speculated that genetics may play a larger role in AS than HFA (DeLong & Dwyer, 1988; Volkmar, Klin, & Pauls, 1998). Asperger (1944/1991) noted that mild forms of social problems were readily apparent in family members, particularly the fathers of children with AS. For example, he described some parents as eccentric and high strung, but despite his early suggestions, research on the genetic aspects of AS has been limited. Several case studies support Asperger's original impression, but there is a lack of controlled studies. Wing (1981) found that 5 of 16 fathers and 2 of 24 mothers had traits common to their children with AS. In addition, Volkmar, Klin and Pauls (1998) described a case where a father and son both exhibited symptoms of AS. This investigation indicated that both the father and son had significantly higher verbal cognitive abilities than performance or nonverbal abilities, and both had similar structural anomalies visualized by MRI. Volkmar, Klin and Pauls (1998) also reported social and developmental difficulties in approximately 33% of fathers and 14% of mothers. Fewer parents (6% of fathers and 2% of mothers) are reported to have difficulties with language.

According to DeLong and Dwyer (1988), the incidence of depression in children with AS is significantly higher that those with autism, supporting the notion that AS is more strongly influenced by familial factors than autism. Interestingly, Cleaver & Whitman (1988) found that depression might be linked to the right hemisphere because it is seen in 66.3% of NLD cases compared to 55.6% of reading disabled. Furthermore,

relatives of individuals with AS generally express phenotypes similar to those with the disorder to a greater extent than relatives of individuals with autism (Gillberg, 1989).

Unfortunately there does not appear to be research published on the genetics of PDD-NOS. It is conceivable that HFA, AS, and PDD-NOS may share the same underlying genetic liability with differences in the severity of the phenotype. It has been reported that there may be evidence for an etiological relationship between autism and AS (Volkmar et al., 1998). Autism in siblings of those with AS is reported in 3.5% of cases, and rates are also relatively high in first cousins. Generally, family members express less severe forms of deficits than those afflicted with AS; however, the frequency of first-degree relatives with AS or similar conditions is high (46%) and provides convincing evidence for the involvement of genetic factors in AS (Volkmar et al., 1998).

The contribution of genetics or for that matter environmental factors, in the etiology of AS, as well as, HFA and PDD-NOS are not clear. Extensive research in this area is likely to continue to provide important information about the causes of PDD. Until then, it will not be known whether or not HFA, AS, and PDD-NOS are genetically alike.

Summary of Neurobiological Findings

HFA, AS, and PDD-NOS are developmental disorders with an organic basis of non-specific origin. It is speculated that brain dysfunction, genetics, and other neurobiological factors contribute significantly to the majority of PDD cases. It should be noted that whereas various abnormalities of brain structure and function have been proposed, no specific brain structures, neural systems, or particular genes are consistently found to identify individuals with HFA, AS or PDD-NOS. Persons with autism often display increased brain volume and abnormalities of language dominance in the left hemisphere. Individuals with AS may show more anomalies in the right hemisphere, but overall display less neurological damage than found in autism. Many patients with a PDD

appear to show dysfunction of the cerebellum, frontal lobes and corpus callosum. Unfortunately, interpretation of neuroimaging data appears to be hindered by methodological difficulties. Furthermore, it is important to remember that the various brain regions work in cooperation with each other, thus complicating our understanding of brain-behaviour relationships.

Research on genetics, implying that heredity plays a larger role in AS than HFA is interesting. While there is overwhelming evidence for genetic involvement in these disorders, the exact modes of inheritance have not yet been elucidated. Until additional research is completed, understanding of the neurophysiological characteristics of HFA, AS, and PDD-NOS remain insufficient to explain the etiologies of these disorders or to devise specific pharmacological interventions. In closer reference to the present investigation, because neurobiological studies have not resulted in a clear method of qualitatively distinguishing subtypes of PDD, neuropsychological studies are very important. Recent neuropsychological research has resulted in revealing different strengths and weaknesses in persons with either HFA or AS, and may be of great benefit when attempting to distinguish between these conditions.

Research on the Neuropsychology of Pervasive Developmental Disorders

Neuropsychology enhances understanding of how the brain mediates behaviour. Accounts of PDD are usually limited to key symptoms; however, this section reviews research in all the major domains of neuropsychological functioning. Neuropsychological studies provide understanding about the variability in the expression of HFA, AS, and PDD-NOS, and thus appear to support the idea that these syndromes are unique from one another. Whereas the following characteristics relate to most cases, the clinical heterogeneity among individuals with these conditions should not be ignored.

Psychometric Intelligence

Cases of AS can frequently be distinguished from HFA by means of psychometric intelligence even though both groups are generally considered to be free of major cognitive deficits, possessing Full Scale IQ's (FSIQ's) usually greater than 70. Researchers suggest that Verbal IQ (VIQ) is higher, but Performance IQ (PIQ) is lower in individuals with AS compared to those with HFA (Ehlers et al., 1997; Gillberg, 1989; Iwanaga et al., 2000; Klin et al., 1995; Ozonoff & Farham, 1994; Ramberg, Ehlers, & Nyden, 1996; Szatmari et al., 1989; Volkmar et al., 1994). Individuals with HFA either have a higher PIQ than VIQ, or no significant discrepancy between performance and verbal functioning (Ehlers et al., 1997; Klin et al., 1995).

When comparing performance on WISC subtests, Ehlers et al. (1997) found that those with AS displayed strengths on verbal tests including, Comprehension, Similarities, Information and Vocabulary. In contrast, they had more difficulty on tests of visual-spatial functioning, particularly the Object Assembly subtest. In addition, attention was considered to be a weakness, represented by the subject's relatively poorer performance on the Arithmetic and Coding. In comparison, the HFA group showed a strength in the visual-spatial domain, characterized by significantly better scores on the Block Design subtest.

Similar to patients with AS, those with PDD-NOS are generally thought to demonstrate a relative strength in verbal skills in comparison to visual-spatial skills, although high-quality research studies are necessary to confirm this.

Language

By definition, language skills are used to distinguish individuals with AS from those with HFA. Most children with autism display delays and impairments in language abilities and are often mute. If language develops, it may be odd due to echolalia, peculiar

choices of words, or unusual intonation, or speech may be fluent but not meaningful (Fine, Bartolucci, & Ginsberg, 1991). Individuals with HFA often achieve learning through rote memory, but skills of verbal memory are not as proficient as in normal subjects.

Generally, individuals with AS do not demonstrate clinically significant delays in language, and in fact there may be a fascination with letters and numbers, and a precociousness in learning to talk. For example, Gunter et al. (unpublished) reported that individuals with AS had no difficulties with literal language in the 'Unusual Metaphors' test, or verbal memory assessed by the Warrington Recognition Memory Test for words.

Overall, language skills are relatively better in AS compared to HFA. Klin et al. (1995) found that consistent with their higher VIQ scores, skills including verbal memory, articulation, vocabulary and verbal output are generally better in those with AS than those with HFA. Similarly, Ozonoff, Rogers and Pennington (1991) noted that VIQ and verbal memory scores reflected that participants with AS had superior verbal abilities compared to the HFA group.

In spite of the fact that researchers have found individuals with AS and PDD-NOS to have better verbal skills than individuals with HFA, higher VIQ does not necessarily mean that those with AS and PDD-NOS are free from speech and language impairments. Although both the ICD-10 and DSM-IV criteria for AS suggest that there should be no early signs of language impairments, there is no mention of the possibility of demonstrating speech or language problems later in development (American Psychiatric Association, 1994, 2000; World Health Organization, 1993). In fact, most children who meet the criteria for AS and PDD-NOS express some oddness in their speech and language (Gillberg, 1991; 1998; Klin, 1994; Tsai, 1998).

Peculiarities of language are especially noticeable in a conversation with a child or adult afflicted with a PDD. For example, difficulties using cohesive links to create a reciprocal discussion may contribute to the breakdown of conversation and create frustration in social interactions (Fine et al., 1991). People with a PDD have difficulties understanding humor because they only grasp the literal meanings of jokes, and fail to comprehend figurative speech such as metaphors and proverbs (Gunter et al., unpublished; Luiselli, Taras, & Lennon, 1998; Tsai, 1998). They also exhibit poor prosody of language (e.g., speech volume, intonation and inflection) and may sound monotonic and precociously formal during childhood (Gillberg & Gillberg, 1989; Klin, 1994; Tsai, 1998).

Wing (1981) proposed that pedantic speech is a common characteristic of AS. Pedantic speech refers to discussion of narrow and intense interests, the use of obscure words, and sometimes an overall deficient vocabulary. Ghaziuddin and Gerstein (1996) state that pedantic speech may be useful in distinguishing AS from HFA because it was found in 76% of AS patients, but only 31% with HFA. Although not necessarily related to psychometric intelligence scores, pedantic speech is common in children with AS who spend much of their time gathering factual information on idiosyncratic topics that are often of little interest to others. In addition, individuals with AS may be markedly verbose, especially when talking about a favorite subject (Klin, 1994)

Overall, verbal functioning is generally superior in those with AS and PDD-NOS in comparison to individuals with HFA, and can be useful to distinguish these conditions. Nevertheless, patients with AS and PDD-NOS are not entirely free of abnormalities in speech and language, and regardless of whether or not there is a language delay, most individuals with PDDs have communication problems serious enough to affect their social-emotional development.

Sensory-Perceptual Functioning

Minshew, Goldstein and Siegel (1997), who have studied neuropsychological functioning in autism, reported that visual-spatial functioning is intact in non-mentally retarded individuals with autism. In fact, they report that visual-spatial functioning is clearly an area of strength in autism. Visual-spatial difficulties may be more representative of children with AS and PDD-NOS than those with HFA, which corresponds to verbal-performance discrepancies often revealed on tests of psychometric intelligence

Klin et al. (1995) found that 16 of 21 individuals with AS exhibited deficits in visual-spatial perception, compared to only 5 of 19 with HFA. Nearly all of those in the AS group had difficulty with visual memory, whereas less than half of those with HFA had a similar problem. Correspondingly, Ehlers et al. (1997) noted that those with HFA evidenced superior visual-spatial skills, particularly on the Block Design and Object Assembly subtest of the WISC-R compared to those with AS.

Holistic organization of visual stimuli may be difficult for those with PDDs. For example, on the Rey Osterreith Complex Figure Test, individuals with AS have been shown to lack hierarchical organization in their drawings (Gunter et al., unpublished). Because they do not integrate parts into whole images, children with PDDs also have problems with facial perception, (Davies, Bishop, Manstead, & Tantam, 1994; Gillberg, 1998; Kracke, 1994). A lack of expected gaze, especially during speech, is common and may be related to decreased attention and response to others' affect and facial expressions (Gillberg, 1998; Tantam, Holmes, & Cordess 1993). Consequently, problems with visual perception (e.g., misinterpretation of facial expressions) may contribute to serious deficits in social interaction.

Davies et al. (1994) found that children with PDDs performed worse than normal controls on tests involving facial and non-facial stimuli. Interestingly, those with PDDs in the high ability group performed worse than controls on all tests of visual perception, whereas those in the low ability group did not function significantly different from control subjects. Davies et al. (1994) did not indicate if systematic differences in performance were correlated to diagnostic labels (e.g., AS compared to HFA).

A recent investigation showed that children with autism had significant problems with face recognition; but unexpectedly, children with PDD-NOS did not have the same difficulty (Klin et al., 1999). To better understand the differences in visual-spatial performance by HFA, AS, and PDD-NOS groups further studies must be conducted.

In addition to problems with visual perception, persons with a PDD have difficulties with other forms of sensory-perceptual functioning. Hypersensitivity to environmental noises is common; but some individuals may also be hyposensitive to sound. For example, they may not acknowledge the voices of people talking to them (Asperger, 1944/1991) and therefore may be mistakenly assumed to be deaf. A deficit in auditory perception is not predictive of AS, although it appears to be more common in HFA (Klin et al., 1995; Sparrow et al., unpublished).

In addition to variable sensitivity to sounds, those with PDDs are often said to be hypersensitive to touch (Asperger, 1944/1991; Grandin, 1992; Tsai, 1998). They may like or dislike some sorts of tactile stimulation (e.g., rough materials or the feel of water), and may be hyposensitive to pain, cold, or heat.

Their sense of taste is usually thought to be odd, because they often have pronounced preferences for certain foods, and dislike of many other foods. Ornitz and Ritvo (1968) suggest that inconsistent and disordered sensory perceptions may be due to inadequate modulation of sensory input in children with autism. However, variability in

stimulus selectivity may also be conceivably related to problems with maintaining attention in individuals with PDDs.

Motor Functioning

One of the controversial issues in the diagnosis of AS versus HFA is whether or not motor skills are a distinguishing feature. The following section describes research on motor skills of children with PDDs, and the potential of using motor clumsiness as a diagnostic criterion for AS.

Populations with PDD exhibit a high incidence of movement difficulties. Most children with AS demonstrated impaired motor skills, and perform below average for their level of psychometric intelligence on standardized tests of motor functioning in the absence of any known neurological disease (Ghaziuddin, Tsai, & Ghaziuddin, 1992). In 1944 Asperger described patients as being "motorically clumsy" and exhibiting poor handwriting as a potential result of clumsiness. Wing (1981) also indicated that 90% of those with AS performed poorly at games requiring motor skill.

In recent times, motor performance in individuals with PDDs has been investigated by several researchers (e.g., Gillberg, 1989; Iwanaga et al., 2000; Klin et al., 1995; Ozonoff et al., 1991; Szatmari, Tuff, Finlayson, & Bartolucci, 1990). Ozonoff et al. (1991) found motor speed and coordination to be better in individuals with HFA than those with AS. Iwanaga et al., (2000) also found that on the Japanese version of the Miller Assessment for preschoolers, more children with AS than HFA had a sensory-motor dysfunction. Similarly, Klin et al. (1995) reported significant deficits in gross and fine motor in children with AS compared to children with HFA.

Other researchers were unable to find a significant difference in motor functioning between AS and HFA groups (Ghaziuddin & Butler, 1998; Ghaziuddin, Butler, Tsai, & Ghaziuddin, 1994; Ghaziuddin et al., 1992; Manjiviona & Prior, 1995). The investigation

by Manjiviona and Prior (1995) measured motor impairment in children using the Test of Motor Impairment – Henderson Revision (TOMI-H), and found that 50% of children with AS and 67% of children with HFA had definite fine and gross motor problems. Similarly, Ghaziuddin and Butler (1998) assessed children with the Bruininks Oseretsky test, a standardized test of motor coordination, and found that children with AS were less impaired than those with autistic disorder. Children with AS were also less impaired than a group of children with PDDNOS, however the differences did not reach statistical significance.

The diagnostic criteria for AS outlined in the ICD-10 mentions that motor clumsiness may be a common feature of AS, but it is not essential for diagnosis. Although a difficult concept to define, "clumsiness" does not necessarily appear to be unique to AS (e.g., Ghaziuddin et al., 1994; Ghaziuddin & Butler, 1998). Also, findings do not rule out the possibility that motor deficits in individuals with different subtypes of PDD could display different patterns (Ghaziuddin & Butler, 1998). Currently it is safe to assert that most patients with a PDD are likely to show problems with motor coordination, but discretion must be taken before including clumsiness as a specific diagnostic feature of AS. The controversy over whether or not motor skills should be regarded as a differentiating feature of AS remains to be settled.

Frontal Lobe Functioning

Executive Function. "Executive function" (EF) is usually described as including abilities such as planning, sustaining attention, goal-directed behavior, self-monitoring, working memory, cognitive flexibility, and inhibitory control. However, EF is not a clearly defined construct, and understanding the relationships between the different functions is still a relatively new area of research. Executive dysfunction is common among individuals with PDDs and in most cases the frontal cortex has been implicated

(Nyden, Gillberg, Hjelmquist, & Heiman, 1999; Ozonoff et al., 1991; Prior, Dahlstrom, & Squires, 1990; Szatmari et al., 1990). Thus far, neuroimaging studies have not consistently shown disturbances in the frontal cortex of individuals with a PDD, but future studies may help account for difficulties in EF.

Disturbances in one particular type of EF, namely attention, are commonly reported in cases of PDD. Asperger (1944/1991) described children as "not tuned in" because their concentration and attention were poor. Individuals with PDDs may have difficulty shifting attention or appear distractible in school during conversations with others, and these difficulties may lead to further problems in academics and social situations over time. In contrast to their problems maintaining attention, at times these individuals can focus for long periods of time on restricted activities of their choice.

One of the most frequently used neuropsychological measures of EF is the Wisconsin Card Sorting Test (WCST). Individuals with PDDs demonstrate poorer performance compared to controls on the WCST (Ozonoff et al., 1991; Szatmari et al., 1990), which taps into skills including categorization, selective attention, shifting set and responding to feedback (Anderson, Damasio, Jones, & Tranel, 1991). Performance by those with a PDD can be described as inflexible, rigid and perseverative. In general, these individuals often have trouble holding back responses and also applying knowledge meaningfully. They also perform poorly on other tests of EF such as the Tower of Hanoi and the Tower of London (Ozonoff et al., 2000).

Interestingly, individuals with PDD are often intolerant to change or inconsistencies in their daily schedules, and transition from one activity to another may be very difficult for them to make (Tsai, 1998). Their style of thinking may appear very inflexible, demonstrated by their repetitive verbal statements and questions. Other

examples of intolerance to change in daily situations include throwing temper tantrums when an object of attachment is taken away or when new foods are presented.

Craig and Baron-Cohen (1999) assert that impoverished creativity in children with autism and AS is additional evidence for dysfunction of EF. They have found that these children exhibit a lack of imagination on several tasks. For example, these children generated few novel changes to an object, and fewer suggestions on an imaginative fluency measure, when compared to controls.

Overall, neuropsychological tests reveal that deficits of EF are common to most cases of PDD. Thus, EF cannot clearly be used as a means of differentiating HFA from AS and PDD-NOS.

Theory of Mind. "Theory of mind" or "mentalizing" deficits are common in children with PDD (Happé, 1994). As a result of PET studies showing abnormal blood flow in the frontal lobe of AS individuals compared to a control group, it is presently believed that mentalizing is at least partially a function of the frontal lobe. Generally, mentalizing is the empathic ability to appreciate what another person is thinking. This may include other people's intentions, desires and beliefs, which may be different from our own. Experiments have repeatedly concluded that children with PDDs often fail theory of mind tasks. Because of "mind blindness" children with PDDs may have difficulty predicting or anticipating another person's behaviour and intentions or motives, and may even have difficulty trying to understand their own behaviour. In addition, they may have trouble understanding emotions, deception, and differentiating fact from fiction. This possibly makes anticipating turn-taking and topic maintenance difficult. Interestingly, Asperger (1944/1991) had suggested that some of his cases might have acted maliciously at times because they did not have any understanding that they were hurting other people's feelings.

Baron-Cohen and Jolliffe (1997) found that adults with AS and HFA may continue to do poorly on advanced tests of theory of mind. Researchers have found that adults with AS tend to pass basic theory of mind tasks (e.g., Bowler, 1992; Happé et al., 1996; Ozonoff, Rogers, & Pennington, 1991; Prior et al., 1990). According to Ozonoff et al. (1991) the ability of individuals with AS to pass theory of mind tasks is a distinguishing feature between AS and HFA. Specifically, the AS group performed significantly better than the HFA group on both basic and complex theory of mind composites. Ozonoff et al. (1991) also suggest that the mentalizing abilities demonstrated by the individuals with AS is related to their relative strength in the verbal domain. Although tests are passed in formal testing, generalization of skills to real-life settings is minimal (Bowler, 1992).

Recently, Sicotte and Stemberger (1999) administered theory of mind tasks to children with PDD-NOS. They found that these individuals were deficient on this task, although not to the same extent as individuals with autism who were tested in previous studies. Therefore, individuals with AS and PDD-NOS have difficulty with theory of mind tasks, but perhaps not to the same degree as those with HFA.

More studies are necessary to understand why individuals with AS and PDD-NOS are often able to pass these tests, but still remain socially impaired. Perhaps a failure of their application of skills, rather than the absence of ability, contributes to their difficulties in social situations.

Central Coherence Deficit. Research indicates that children with PDDs have a central coherence deficit, meaning that they perform poorly when attempting to bring diverse pieces of information together to form a higher-level meaning in context (Ellis & Gunter, 1999; Jolliffe & Baron-Cohen, 1999). More simply stated, they focus on detail and tend not to see the whole picture. An example of this difficulty is apparent when they

attempt to process information from facial expressions holistically in order to judge emotions.

On a test of visual-spatial skills and attention to detail, requiring the detection of a geometric figure within a complex diagram (Embedded Figure Test), children with AS perform faster than those without a PDD. In contrast, when recalling the Rey-Osterreith Complex Figure, these children struggle due to poor structural coherence (Ellis & Gunter, 1999; Ellis & Leafhead, 1996). In addition, they may have difficulty in choosing, prioritizing, organizing and comprehending connections and generalizing skills and knowledge. Individuals with autism have even greater difficulty that those with AS when attempting to achieve coherence (Jolliffe & Baron-Cohen, 1999).

Similar to children with HFA and AS, those with PDD-NOS have clear difficulties in the area of abstract thought and comprehension (Tsai, 1998). However, investigations are required to see if central coherence deficits are also a common feature of PDD-NOS, and how this impacts daily functioning in comparison to levels of dysfunction seen in HFA and AS.

Social-Emotional Functioning

Many researchers believe that the fundamental disorder in children with PDDs is their difficulty in interacting socially (Asperger, 1944/1991; Gillberg & Gillberg, 1989; Scott, 1985; Wing, 1981). Reported problems with social-emotional functioning can include difficulty understanding and interpreting details relating to the social environment, for example, other people's feelings, thoughts, and motives, or information about social rules, or relations between people. Reasons for their deficient social skills likely result from numerous other problems including difficulties with verbal and nonverbal communication, visual-spatial problems (e.g., interpreting emotion from facial expressions), and challenges with academic material.

According to some researchers (e.g., Klin, 2000; Ozonoff et al., 1991) individuals with HFA and AS do not differ on measures of social-emotional functioning, however other researchers have found that those with HFA have somewhat more difficulty (e.g., Bartolucci & Bremner, 1989; Sparrow et al., unpublished; Szatmari et al. 2000).

Children and adults with AS have much difficulty in social relations with others and those with HFA are usually not even interested in social interactions (Sparrow et al., unpublished). Szatmari, Bartolucci and Bremner (1989) compared children matched on IQ and found that those with AS had social impairments similar to those with HFA. Nevertheless, they did note that the subjects with AS did not show a significant lack of social responsiveness toward their primary caregivers, as did subjects with HFA. Szatmari et al. (2000) found that children, aged four to six years, with either HFA or AS had significant difficulties with social skills, but interestingly, after a two year follow-up, those with AS showed a greater improvement in social skills than those with HFA.

Individuals with AS and PDD-NOS tend to have somewhat better social skills when compared to those with autism; nevertheless, interacting with others is still a fundamental problem for them (Njardvik, Matson & Cherry, 1999). Serra, Minderaa, van Geert, and Jackson, (1995) examined 24 children with PDD-NOS and according to their symptom check-list, nearly all of these subjects had trouble inferring other people's emotions, thoughts and intentions, and displayed difficulty understanding social information and social rules. However, Serra et al. (1995) reported that these individuals tend to seek interaction with others. Also, children with PDD-NOS may be considered to be overly persistent in their ways of thinking. In addition, they may act chaotically when under pressure or when faced with novel situations. According to parental ratings on the Child Behaviour Checklist (CBCL), PDD-NOS children show significantly greater problems with behaviour than normal age-matched peers (Serra et al., 1995).

Specifically, Serra et al. (1995) examined "person perception" a social-cognitive skill that involves how a child conceptualizes other people, their thoughts, intentions, attitudes, personality traits and emotions ("central" or psychological aspects), and also their physical characteristics, for example, age, name or gender, and also overt behaviour ("peripheral" or external aspects). When compared to normal control subjects, Serra et al. (1995) found that individuals with PDD-NOS had more difficulty describing another person without a considerable level of instruction. The PDD-NOS children did not differ significantly from normal children with respect to the use of peripheral statements to describe another person, nevertheless they did tend to use fewer central statements. It should be noted that in both the experimental and control groups, older children (10 to 12 years of age) used more central statements than younger children (7 to 9 years of age). Also, the fewer central statements made by the PDD-NOS group were speculated to result from a difference in WISC-R IQ level (i.e., mean of 95 for the PDD-NOS group, and 107 for the normal controls). A re-examination of this task using 27 children with PDD-NOS and 31 normal controls matched on age, gender and IQ resulted in the same findings (Serra, Minderaa, van Geert, & Jackson, 1999). Overall Serra et al. found that children with PDD-NOS may have the skills to infer other people's mental states, but are unable to spontaneously use these skills. Serra et al. (1995, 1999) concluded that similar to performance on "theory of mind" tasks that are passed by individuals with AS, but not with HFA (see Bowler 1992), some subgroups of PDD may perform well on social-cognitive tasks, despite their failure in everyday social situations.

According to psychometric tests individuals with AS and PDD-NOS generally have relatively adequate language abilities, but their nonverbal communication style is quite unusual (Tsai, 1998). For example, eye contact is not appropriately maintained, which is especially apparent during conversations with others. Their understanding and

use of gestures and facial expression is poor, and they often display a "flat affect". Although most facial expressions appear flattened, they may at times strongly demonstrate feelings of anger and frustration. Individuals with HFA generally show similar problems with gaze, gestures and affect, as those with AS and PDD-NOS, in addition to their problems with language (Hobson, 1986).

In general, for most individuals with PDDs, extreme impairment in socialization begins in infancy and often continues throughout adulthood. For example, smiling, babbling and gestures may be utilized less frequently when compared to normally developing infants (Tsai, 1998). Play is restricted and repetitive in children with PDDs. Deficient social skills and restricted interests become more apparent during preschool and elementary years and efforts to overcome social deficits are frequently ineffective (Nordin & Gillberg, 1998; Wing, 1981). These individuals often misunderstand rules of social interactions, jokes and metaphors (Luiselli et al., 1998; Tsai, 1998). Misinterpretations may become upsetting in many circumstances. For example, a child with a PDD may be disturbed when watching magic shows where a woman is sawn in half because he or she does not understand that it is not real. In addition, these children have difficulty forming peer relationships and they often socialize relatively better with adults. Children with PDD may appear egocentric, insensitive and lacking in tact. They experience frequent problems with family and friends, and often remain socially isolated (De Long & Dwyer, 1988).

For individuals with AS, late adolescence is sometimes associated with a variety of problems including severe cases of catatonia (Nordin & Gillberg, 1998; Wing, 1981). Adolescents and adults with AS frequently face problems resulting from their attempts to "fit in'. They realize that they are different, and as a result they may become withdrawn, anxious and depressed (De Long & Dwyer, 1988). Generalized anxiety and depression

may be related to misunderstanding of language and social cues, limited comprehension of behavioural expectations, and social stigma in addition to biological and genetic factors (Wing, 1981; De Long & Dwyer, 1988; Ghaziuddin et al.1998). Sadly, incidence of suicide may also be high (Nordin & Gillberg, 1998; Wing, 1981).

Restricted Range of Interests, Activities or Behaviours

Another feature of PDD, perhaps related to minimal social interaction, is a restricted range of interests. For example, a child may avoid novel activities and instead become preoccupied with a certain toy, television program or odd items (e.g., keys or maps). Individuals with autistic spectrum disorders may engage in stereotyped behaviours such as repetitive motor movements including, rhythmic rocking, monotonous play with a particular toy, hand flapping or spinning objects (Asperger, 1944/1991; Tsai, 1998; Wing, 1981). Preoccupations with objects and motor stereotypies are more common in those with HFA than those with AS; however, individuals with AS are more likely to be fixated on all absorbing circumscribed interests (Sparrow et al., unpublished).

Persons with PDDs are likely to be poor in maintaining attention in topics outside their main interests. Topics that children with AS are interested in are often more intellectually advanced than those in which children with HFA take an interest. In addition, isolated special skills, often related to abnormal preoccupations, seem more common in AS than HFA (Volkmar et al., 1994). Also, in comparison to HFA, individuals with AS are known to spend more time collecting an extraordinary number of facts on a narrow topic that is of interest to them.

Academic Achievement

During the school years, significant strengths and weaknesses in academics are found in children with PDD. Children with AS often perform well in vocabulary, spelling, grammar, mechanical reading and arithmetic, but have difficulties with

comprehension, writing and other areas of scholastic performance (Wing, 1981). Similar patterns of performance have been found in persons with HFA; however, they may exhibit more problems with verbal tasks such as vocabulary (Klin et al., 1995). Research in this area is scarce and more studies of academic achievement in individuals with PDD are essential. Although it appears that most individuals with a PDD struggle with certain skills, such as comprehension (Tsai, 1998), at this point, differences in the academic profiles of persons with AS, HFA, and PDD have not been well documented.

Prognosis

PDD continues throughout the lifespan. In spite of the fact that children with autism are generally diagnosed around three years of age, those with AS and PDD-NOS may not be recognized until sometime later in childhood. Communication problems are experienced as early as infancy and deficient social skills, academic difficulties and restricted interests become apparent during preschool and elementary years (Tsai, 1998; Wing, 1981). Late adolescence and adulthood may be marked by behavioural impairments, socialization and communication problems, and psychiatric disorders (e.g., depression) (De Long & Dwyer, 1988; Wing, 1981). Adults with PDD may be labeled "eccentric" because they have narrow interests; however, they are often able to work and function relatively well in day-to-day life. This is especially true if their occupations are tailored around their specific areas of interest. In general, individuals with AS and PDD-NOS are reported to have better outcomes in terms of self-care and academic growth than those with autism. This may be because outcome significantly improves with the presence of language and higher psychometric IQ for most persons with any subtype of PDD (Happé & Frith, 1996; Gillberg 1991, 1998).

It appears that outcome is generally better in those with AS in contrast to autism (Szatmari, Bartolucci, & Bremner, 1989). For example, those with autism spend more

years in special education and are less able to live independently than individuals with AS. Nevertheless, those with AS may be more vulnerable than those with autism to developing a variety of psychiatric disorders. Due to the reduced severity of PDD-NOS in comparison to other PDDs, it is expected that persons with this condition have relatively better outcomes than individuals with HFA (Tsai, 1998). Nevertheless, research is required to confirm this.

There is no cure for AS, HFA or PDD-NOS. Accurate diagnosis and understanding of outcome may provide information to formulate appropriate intervention plans. Also, treatment may be tailored to the individual's strengths and weaknesses. Additional studies of outcome for individuals with PDDs are crucial to better comprehend differences in developmental progression of these syndromes, and therefore provide further justification for differentiating between HFA, AS, and PDD-NOS.

Summary of Neuropsychological Findings

Neuropsychological literature shows that the expression of a PDD is highly complex and diverse. Because of neuropsychological differences found between subtypes of PDD, conditions such as HFA, AS, and PDD-NOS should be viewed as separate conditions with overlapping symptoms. For example, persons with AS and PDD-NOS usually have better verbal functioning but poorer visual-spatial functioning compared to individuals with HFA that are matched on IQ. All groups usually have deficits on theory of mind tasks, although patients with AS and PDD-NOS are somewhat more successful on these tasks compared to individuals with HFA, presumably because of relatively superior language skills. In general, PDDs are characterized by difficulties in communication; speech tends to be odd, and nonverbal communication using facial expressions, gaze and gestures is poorly developed. Pedantic speech and special interests to the point of obsession are found more commonly in cases of AS compared to HFA.

Motor mannerisms appear to be more common in HFA than AS or PDD-NOS. In addition, motor skills may be more deficient in AS relative to HFA, however not all studies confirm this. Social-emotional functioning is problematic for all groups, although individuals with AS and PDD-NOS seem less aloof and make a clear effort to interact in social situations, whereas most of those with HFA avoid social intercourse. Finally, prognosis is somewhat better in cases of AS and PDD-NOS compared to HFA; this is often attributed to their relatively better language skills.

Statement of Purpose

Rationale

Research has been presented to describe the relationship between subtypes of PDD, and the similarities between AS and NLD. There is an ongoing debate concerning the matter of AS and HFA being distinct syndromes or merely different due to varying levels of severity. For example, AS has often been a term applied to a milder version of the disorder. Despite evidence demonstrating qualitative differences between AS and HFA (e.g., Ehlers et al., 1997; Klin et al., 1995; Sparrow et al., unpublished), few studies have distinguished these conditions beyond looking at them along a continuum.

The goal of the present study is to employ the NLD Scale in order to see if it can provide further evidence that AS, in contrast to HFA, shares a characteristic phenotype with the NLD syndrome (e.g., Gunter et al., unpublished; Rourke, 1989, 1995), therefore illustrating that AS and HFA are qualitatively distinct. The present research is unique from previous studies scrutinizing the differences between AS and HFA because it uses a new instrument, the NLD Scale. Also, this study includes a group of individuals neglected in most studies, specifically those with PDD-NOS, in order to provide a better understanding of this condition.

Use of the NLD Scale in research studies is still in its preliminary stages. Thus, the reliability and validity of the NLD Scale are in the process of being established. The present research provides an attempt to begin to test the usefulness of this tool by investigating its validity and internal consistency on a sample of individuals with PDD. Ongoing studies by van der Vlugt and his colleagues have established that test-retest and interrater reliability of the NLD Scale are adequate. With respect to validity, it appears that the NLD Scale has face validity, in that it appears to measure characteristics observed in children and adolescents with NLD; nevertheless, a study of the objective validity of the NLD Scale would be beneficial.

Because no previous research has been conducted using the NLD Scale to distinguish AS and PDD-NOS from HFA, the present study will provide new and useful information. For example, rather than clinicians, raters for the NLD Scale are parents who constantly observe their children's behaviours. Use of the NLD Scale in this study is appropriate because past investigations have demonstrated a striking parallel between the behavioural characteristics of the NLD syndrome and AS, but not between the NLD syndrome and HFA. It is expected that this project will further support the correspondence between NLD and AS, and perhaps to some extent between NLD and PDD-NOS.

Limitations

Although the NLD Scale appears to be useful as a tool to distinguish those with and without NLD, it is not without some shortcomings. The NLD Scale has not been widely used in research and optimally more comparisons between groups positive for NLD and groups that have other learning disabilities or no disabilities are necessary. Such comparisons are not yet published; nevertheless, there is evidence that the NLD scale is a beneficial tool to identify individuals with NLD (e.g., van der Vlugt, unpublished).

At this point in time the NLD Scale has no cut-off scores to distinguish persons with or without NLD. Although this may make it more difficult to interpret results, it is still possible to calculate if groups score significantly differently from each other by means of statistical analysis.

There is a definite need to investigate the characteristics of PDD-NOS; however, a major limitation of this study is that there is no universally agreed upon definition for PDD-NOS. Since the publication of the DSM-IV, there has been a standardized definition of AS which has allowed researchers to more easily study differences between AS and HFA. Studies conducted before this must obviously be interpreted with caution. Likewise, research that involves subjects with PDD-NOS must be carefully examined. Nevertheless, researchers at the Yale University Child Study Center have been involved in numerous controlled studies using patients with PDD-NOS. They include experts in the field of PDDs who, in fact, contributed to the definitions found in the DSM-IV. Hopefully, research studies such as the present one will contribute to a better understanding of PDD-NOS, and perhaps foster the development of a clearer definition of PDD-NOS.

Hypotheses

There is a long-standing debate concerning whether or not subtypes of PDD exist on a continuum of severity. The term "autistic spectrum disorders" might lead some to believe this, but there is evidence that shows that these syndromes are characterized by varying cognitive strengths and weaknesses. The present study aims to help clarify if these syndromes have differences significant enough to warrant their unique names.

This study utilizes the NLD Scale in order to test its discriminating ability with respect to HFA, AS, and PDD-NOS. Comparing scores obtained by individuals with AS and HFA on the NLD Scale is expected to provide further evidence that the NLD profile

is also characteristic of AS, but not of HFA. This would support recent neuropsychological studies suggesting that AS and HFA are qualitatively different rather than existing on a continuum (e.g., Klin et al., 1995; Sparrow et al., unpublished). In addition, it is expected that subjects with PDD-NOS will score similarly to those with AS, displaying characteristics of NLD to a greater extent than individuals with HFA. This is hypothesized because past studies (e.g., Kurita, 1997) have shown AS to be similar to PDD-NOS, although by definition (American Psychiatric Association, 2000), deficits in PDD-NOS are less numerous and/or not as severe as those seen in AS. Based on the previously outlined research the following predictions were made:

Study 1: Performance of the PDD subgroups on the NLD Scale. Individuals with AS are expected to receive the highest mean total score on the NLD Scale, followed by those with PDD-NOS, and individuals with HFA are expected to receive the lowest scores. In other words, those with AS are expected to share the most characteristics, and those with HFA are expected to share the least characteristics with the syndrome of NLD. In addition to total scores on the NLD Scale, subsections of the scale and the individual questions they are composed of will be analyzed to identify more clearly how the subtypes of PDD are similar or different:

(a) Similar to total scores, it is hypothesized that the AS group will score highest on the first section of the NLD Scale, meaning that they will display neuropsychological strengths and weakness characteristic of the syndrome of NLD. Meanwhile, the PDD-NOS group will score lower than the AS group, and the HFA group is expected to score the lowest on this section, suggesting that their neuropsychological profile is less characteristic of NLD.

(b) On the academic achievement section of the NLD Scale, individuals with AS and PDD-NOS are hypothesized to have higher mean scores than the HFA

group. The AS and PDD-NOS groups are not expected to have scores that are significantly different from one another. This is hypothesized because, unlike HFA, the AS and PDD-NOS groups are speculated to share academic strengths (e.g., verbal functioning) and weaknesses (e.g., visual-spatial functioning) seen in individuals with NLD.

(c) All groups are hypothesized to have scores that do not differ significantly on the third section, comprised mostly of questions related to social-emotional and adaptive functioning, because all subtypes of PDD are thought to share a social disability.

Study 2: Validity of the NLD Scale. In order to test the validity of the NLD Scale, total scores will be compared to scores obtained using an algorithm that classifies individuals as having a low, questionable or probable chance of displaying NLD. This algorithm is translated from a previously established algorithm developed by Pelletier, Ahmad and Rourke (unpublished) according to neuropsychological strengths and weaknesses reported (e.g., using test scores on nonverbal memory, verbal and nonverbal intelligence, fine motor skills, and achievement). Individuals with a probable chance of having NLD according to neuropsychological profiles are expected to score high on the NLD Scale. Individuals who show fewer characteristics of NLD according to their neuropsychological profiles, and thus have a questionable chance of having NLD, are expected to score significantly lower on the NLD Scale. Finally, individuals who do not show characteristics of NLD according to their neuropsychological strengths and weakness, and thus have a low chance of having NLD, are expected to score the lowest on the NLD Scale. Differences between these three groups are expected to be statistically significant. The most pronounced effects are expected on the neuropsychological functioning section of the scale, because questions in this section are most closely related

to the domains of functioning measured by the neuropsychological tests used in the algorithm.

Study 3: Reliability of the NLD Scale. The internal consistency of this scale and its subsections will be tested with the expectation that the alpha levels will be acceptable. For example, Cronbach's alpha coefficients near 0.7 are expected. However, internal consistency coefficients are not expected to be significantly higher than this because the items found on the NLD Scale examine a variety of behaviours rather than a single characteristic deficit.

CHAPTER II

METHOD

Background

Archival data for this study was obtained from the Yale University Child Study Center, which does extensive research on PDDs. This research program consists of a multidisciplinary clinical approach, assessing individuals by various methods, including neuropsychological evaluations, neuroimaging and genetic screening. However, the present study was limited to data obtained using the NLD Scale, and various subtests from the Yale Social Learning Disabilities Project Neuropsychological Testing Battery.

Subjects

The participants in this study included 16 males who met criteria for HFA, 21 who met criteria for AS, and 12 who met criteria for PDD-NOS. Criteria for inclusion in the HFA, AS or PDD-NOS groups were stringent. Only cases meeting full criteria following meticulous medical record review and diagnostic evaluation were included. All subjects were examined and diagnosed by (a minimum of) an experienced child psychiatrist and an experienced clinical neuropsychologist. Diagnosis was confirmed by means of specific procedures, including independent clinical diagnosis based on DSM-IV criteria.

To assist in diagnosis, information was provided by clinical observation of current social and communicative behaviour using the Autism Diagnostic Observation Schedule (ADOS), a standardized instrument for diagnosis (Lord, Rutter, DiLavore, & Risi, 1999). This instrument consists of a series of structured and semi-structured stimuli for social interaction. Specific target behaviours are coded and the quality of social and interpersonal behaviours is rated.

In addition, parent reports of their child's social interaction, communication, and repetitive or stereotyped behaviour impairments were provided by means of a standardized interview using the Autism Diagnostic Interview-Revised (ADI-R; Lord et al., 1994). The ADI-R consists of a series of questions on education, treatment history, family constellation, social skills, communication, play, and questions on behavioural difficulties, including restricted and repetitive behaviours. Each question on the ADI-R is scored on a scale of zero to three, where zero is normal and three is severely disabled.

Both the ADOS and ADI-R have criteria for overall diagnosis as well as for each of the three DSM-IV domains of autism. In addition, both instruments have demonstrated good reliability and validity when used by trained examiners (Lord et al., 1994; Lord et al., 1989).

Subjects were matched as closely as possible in terms of age and FSIQ. The sample was restricted to individuals with a level of psychometric intelligence above the range of mental retardation (e.g., FSIQ>70). Using only high-functioning individuals was necessary to maintain a relatively homogeneous sample. Also, the sample was limited to males because the number of subjects was too small to permit analysis of gender differences. Finally, to be included in the study, children were at least seven years of age. This was required to be included in the Yale Social Disabilities Project, and also it is also the minimum age for testing according to present NLD Scale instructions. Adolescents and adults were also included in this study.

Measures

In addition to the NLD Scale, data from the following tests were utilized as dependent variables in this research: Wechsler Adult Intelligence Scale – third edition (WAIS-III; Wechsler, 1997), or Wechsler Intelligence Scale for Children – third edition (WISC-III; Wechsler, 1991), the Purdue Pegboard (Tiffin, 1968), the Finger Windows

subtest of the Wide Range Assessment of Memory and Learning (WRAML; 1990), and various achievement tests. Each will be briefly described here in turn, and mention of its utility made.

Wechsler Adult Intelligence Scale – third edition. The Wechsler Adult Intelligence Scale – third edition (WAIS-III; Wechsler, 1997) is a test of psychometric intelligence for individuals over the age of 16. It is composed of 14 subtests that sample a wide range of cognitive abilities. It is categorized into 3 scales, Verbal (VIQ), Performance (PIQ) and Full Scale (FSIQ). The Verbal Scale is composed of seven subtests, including Vocabulary, Comprehension, Similarities, Arithmetic, Digit Span, Information, and Letter-Number Sequencing. The Performance Scale is composed of seven subtests, including Picture Completion, Digit Symbol, Block Design, Picture Arrangement, Matrix Reasoning, Object Assembly, and Symbol Search. The Full Scale is a composite of all subtests except Object Assembly.

Wechsler Intelligence Scale for Children – third edition. The Wechsler Intelligence Scale for Children – third edition (WISC-III; Wechsler, 1991) is similar to the WAIS-III, and it is developed to assess psychometric intelligence in children ranging in age from 6 to 16 years. This test is comprised of six verbal subtests (i.e., Vocabulary, Comprehension, Similarities, Arithmetic, Digit Span, and Information), and seven performance subtests (i.e., Picture Completion, Block Design, Picture Arrangement, Object Assembly, Symbol Search, Coding and Mazes). Subtest scores, excluding Digit Span and Mazes, are combined to calculate VIQ, PIQ, and FSIQ.

Wechsler tests of intelligence generally have very good to excellent split-half and test-retest reliability for Verbal, Performance and Full Scale IQ's as well as other indices, including factor scores (Spreen & Strauss, 1998). In terms of validity, there is also high correlation between Wechsler IQ's and other tests of psychometric intelligence, as well

as, tests of academic achievement.

Purdue Pegboard. The Purdue Pegboard (Tiffin, 1968) measures dexterity of the fingers and hands. Pins are picked up and placed into a pegboard with the dominant hand for 30 seconds. This is repeated with the non-dominant hand, and again with both hands. This test has been shown to have moderately high test-retest reliability ($r = 0.82$) (Kelland, Lewis, & Gurevitch, 1992).

Wide Range Assessment of Memory and Learning. The Wide Range Assessment of Memory and Learning (WRAML; Sheslow & Adams, 1990) measures an individual's verbal and visual memory. It is usually administered to children 5 to 17 years of age. The present study utilized only one subtest from the WRAML, the Finger Windows subtest. Finger Windows subtest is a measure of nonverbal memory that requires subjects to put their fingers through a series of holes immediately following the test administrator's demonstration. Sheslow and Adams report moderately good test-retest reliability coefficients (.61 - .84), and coefficient alpha measures ranging from .78 to .96.

Achievement tests. Measures of academic achievement were extracted from a variety of tests, depending on what could be found in the patients' clinical files. Only subtests of basic reading (decoding) and arithmetic were obtained for this study.

Reading and Arithmetic subtests from the Wide Range Achievement Test, WRAT-R (Jastak & Wilkinson, 1984) or WRAT-3, (Wilkinson, 1993) were collected from patient files. These tests measure basic educational skills in persons 5 to 75 years of age. Reliability estimates of WRAT-3 subtests are high. For example, test-retest correlations range from .91 to .98 when individuals 6 – 16 years were re-tested after a month (Wilkinson, 1993).

If a file did not contain WRAT-R or WRAT-3 standard scores, scores from the Wechsler Individual Achievement Test, (WIAT; The Psychological Corporation, 1992)

were used. The WIAT is used to individuals between the ages of 5 and 19 on a variety of achievement measures. For this study only scores from the Basic Reading and Numerical Operations subtests were collected. This standardized achievement battery has moderately high test-retest and split-half reliability (.69 - .98). Also, it allows for meaningful comparisons with measures of general ability particularly the WISC III. In addition, the WIAT correlates substantially, generally above .70, with other tests of achievement including, WRAT-3, Woodcock-Johnson Psycho-Educational Battery-Revised, and Kaufman Test of Educational Achievement.

If test scores from neither the WRAT-3 nor the WIAT were available, standard scores from reading (Letter-Word Identification) and arithmetic (Applied Problems) scores were obtained from the Woodcock-Johnson Psycho-Educational Battery-Revised (WJ-R; Woodcock & Johnson, 1989) or the Kaufman Test of Educational Achievement, (K-TEA; Kaufman & Kaufman, 1985).

Procedure

Data collection.

Archival data from subjects assessed at the Yale Child Study Center were obtained. Data not already available on computer were obtained from case files and inputted into a Microsoft Excel file. Standard scoring procedures for the NLD Scale were employed to score individual questions and to arrive at a total score. Finally, various statistical tests, as described below, were conducted using SPSS-10 software (SPSS, 2000).

Descriptive statistics.

Descriptive statistics including means and standard deviations for age, FSIQ, VIQ and PIQ, and scores on the NLD Scale (i.e., total scores and subsection scores) were calculated for the entire PDD sample, and also for the three groups of interest (HFA, AS,

and PDD-NOS). Following this, one-way between-groups ANOVAs were used to determine if the groups were matched on age and FSIQ.

Study 1: Performance of the PDD subgroups on the NLD Scale.

To test the first hypothesis, stating that the AS group will have the highest total scores on the NLD Scale followed by the PDD-NOS and HFA groups, a one-way between-groups ANOVA and Jonckheere's test of trend were run. The independent variable was the mutually exclusive group, either HFA, AS or PDD-NOS, in which the participant was placed. The dependent variable was the total score obtained on the NLD Scale.

A similar procedure was used when analyzing the difference between scores obtained by the HFA, AS, and PDD-NOS groups on subsections of the NLD Scale: (1) neuropsychological functioning, (2) academic achievement, and (3) social-emotional and adaptive functioning.

To determine if there was a significant difference between responses on individual questions, the three PDD groups were again compared using one-way between-groups ANOVAs and Jonckheere's test of trend. Thus, selective NLD behavioural characteristics prevalent in each PDD group could be detected. For all analyses, a significance level of $p < .05$ was used.

Study 2: Validity of the NLD Scale.

The same subjects were used in a validity analysis. Neuropsychological profiles were analyzed to determine which subjects were positive for NLD. Classification rules for NLD (Pelletier, Ahmad, & Rourke, unpublished) were modified using available tests thought to measure characteristics of NLD (see Table 3). In particular, scores from the WISC-III or WAIS-III, the Purdue Pegboard, the Finder Windows subtest of the WRAML, and academic achievement scores from various tests (e.g., K-TEA, WIAT,

Table 3

Rules of NLD Classification & Scoring Algorithm for testing the Validity of the NLD Scale

Provisional rules of NLD classification (Pelletier, Ahmad, & Rourke, unpublished). The numbers in parentheses represent the percentage of children classified as having NLD who exhibit this feature.	Rules of NLD classification to be used in the present study (modified by Tsatsanis & Taneja). Criteria were translated according to availability of tests in the Yale Child Study Center neuropsychological battery.
1. Target Test at least 1 standard deviation below the mean (90)	Finger Windows subtest from the WRAML at least 1 standard deviation below the mean
2. No or very minimal simple tactile imperception and suppression versus very poor finger agnosia, finger dysgraphesthesia, and astereognosis composite (90)	No tests of tactile perception or finger agnosia tests included in the present study
3. Two of WISC Vocabulary, Similarities, and Information are the highest of the Verbal scales (76)	Two of WISC-III or WAIS-III Vocabulary, Similarities, and Information were the highest of the Verbal Scales
4. Two of WISC Block Design, Object Assembly, and Coding subtests are the lowest of the Performance scales (76)	Two of WISC-III or WAIS-III Block Design, Object Assembly, and Coding (or Digit Symbol) subtests were the lowest of the Performance Scales
5. WRAT Standard Score for Reading is at least 8 points greater than Arithmetic (72)	Standard scores for reading (decoding) at least 8 points greater than arithmetic subtests on a test of achievement (e.g., K-TEA, WRAT- R, WRAT-3, WIAT, or WJ-R Tests of Achievement)
6. Tactual Performance Test right, left, and both hand times become progressively worse vis-a-vis norms (66)	Tactual Performance Test not included in the present study
7. Normal to superior grip strength versus mildly to moderately impaired Grooved Pegboard (59)	Purdue Pegboard performance at least 1 standard deviation below the mean
8. WISC VIQ greater than PIQ at least 10 points (41)	Criterion of VIQ greater than PIQ by at least 10 points omitted

WRAT-R, WRAT-3, WJ-R Tests of Achievement) were developed into an algorithm. Because of the variety of achievement tests used, this measure was included only for interest's sake, and statistical analyses were conducted with and without taking into account the difference between reading and arithmetic scores.

Changes in NLD criteria from Pelletier, Ahmad and Rourke's original rules included use of the Finger Windows test instead of the Target Test as a measure of nonverbal memory, and use of the Purdue Pegboard instead of the Grooved Pegboard as a measure of fine motor skill. In addition, measures of tactile perception were not available for the present investigation. Finally, unlike criteria formulated by Pelletier et al., the present study did not include the NLD criterion stating that WISC VIQ should be greater than PIQ at least 10 points. This rule was excluded for two reasons. First, it was omitted because there is an overlap between this rule and two of the other rules (e.g., (1) Two of WISC Vocabulary, Similarities, and Information are the highest of the Verbal scales, and (2) Two of WISC Block Design, Object Assembly, and Coding subtests are the lowest of the Performance scales). Second, a VIQ greater than PIQ by 10 points is the least effective criterion in identifying cases of NLD. Although it might be incorrectly assumed that VIQ must be higher than PIQ in all cases of NLD, in fact Pelletier et al., noted that only 41% of their NLD cases met this criterion. However, it will be noted if individuals with a Verbal IQ at least ten points greater than their Performance IQ will have higher scores on the NLD Scale than do persons without such a discrepancy.

For this study, a subject received a point for meeting each of the following NLD criteria:

1. Finger Windows at least one standard deviation below the mean.
2. Two of WISC-III or WAIS-III Vocabulary, Similarities, and Information were the highest of the Verbal Scales.

3. Two of WISC-III or WAIS-III Block Design, Object Assembly, and Coding (or Digit Symbol) subtests were the lowest of the Performance Scales.
4. Standard scores for reading (decoding) were at least 8 points greater than arithmetic subtests on a test of achievement (e.g., the K-TEA, WRAT- R, WRAT-3, WIAT, or WJ-R Tests of Achievement).
5. Purdue Pegboard performance was at least one standard deviation below the mean.

Pelletier et al. are investigating the use of the criteria found in table 3 to diagnosis NLD: subjects meeting the either the first five features or seven of the eight features are classified as having definite NLD, those with five or six of these eight features are thought to have probable NLD, those with three or four of the eight features are thought to have a questionable chance of displaying NLD and those with one or two features are thought to have a low probability of exhibiting the NLD syndrome. In the present study, subjects were classified as having either a low (1 point out of 5), questionable (2 or 3 points out of 5), or probable chance of displaying the syndrome of NLD (4 or 5 points out of 5). The responses on the NLD Scale, both total scores and subsection scores, were then compared to the placement along this NLD continuum (i.e., low, questionable, or probable) using one-way between-groups ANOVAs.

Study 3: Reliability of the NLD Scale.

Finally, adequacy of the NLD Scale's internal consistency was measured. Despite optimal testing conditions, no test is perfectly reliable. In the present study, internal consistency was measured using the Cronbach's alpha test of reliability. Cronbach's alpha was also run for each of the three subsections to assess the internal consistency of each domain of the scale.

CHAPTER III

RESULTS

Descriptive Statistics

The sample in this study consisted of 21 males diagnosed with AS, 16 with HFA and 12 with PDD-NOS. Of these subjects, 1 person with AS, 3 with HFA and 1 with PDD-NOS were left-handed and the rest were right-handed. Descriptive statistics for age, FSIQ, VIQ and PIQ were calculated (see Table 4).

Subjects ranged in age from 8.1 to 37.9 years (M = 16.0, SD = 7.2). A one-way between-groups ANOVA revealed that the HFA group was older than the AS and PDD-NOS groups [F (2,46) = 3.25; p= .05]. The HFA group ranged in age from 10.6 to 37.9 years (M = 19.5, SD = 8.4), the AS group were 8.1 to 36.3 years (M = 14.7, SD = 6.7), and the PDD-NOS group ranged from 8.9 to 26.3 years (M = 13.4, SD = 4.6).

Ranges of IQ scores for the entire sample were as follows: FSIQ, 70 to 144 (M = 100.9, SD = 21.2), VIQ, 72 to 153 (M = 106.9, SD = 22.6), and PIQ, 67 to 136 (M = 93.9, SD = 18.7). One-way between-groups ANOVAs showed that the groups were not different on FSIQ [F (2,46) = .84; p = .44], VIQ [F (2,46) = 1.86; p = .17], or PIQ [F (2,46) = .04; p = .96]. Ranges of IQ scores for the groups under investigation are found in Table 4.

Study 1: Performance of the PDD subgroups on the NLD Scale

The number of subjects in each of the PDD groups is small (N=49 total; n=16 with HFA; n=21 with AS; n=12 with PDD-NOS), thus the statistical power of each of the following analyses is low. It may be complicated to interpret the results of these analyses, because of the small number of subjects in each group, significant differences may not be exposed. Nevertheless, the findings that are significant are expected to be robust.

To test the main hypothesis, means of the total scores on the NLD Scale were

Table 4

Mean, Standard Deviation and Range of Age and IQ Scores for Individuals with AS, HFA and PDD-NOS

	HFA (n=16)			AS (n=21)			PDD-NOS (n=12)		
	M	SD	Range	M	SD	Range	M	SD	Range
Age	19.5	8.4	10.6-37.9	14.7	6.6	8.1-36.2	13.4	4.6	8.9-26.3
FSIQ	95.4	21.4	70-141	102.8	18.6	72-132	104.9	25.4	75-144
VIQ	98.3	24.0	72-150	109.9	17.8	75-140	113.2	26.6	81-153
PIQ	92.8	18.8	67-135	94.2	18.5	68-136	94.8	20.6	69-127

calculated and compared for each of the three groups (see Table 5). Each of the 40 questions can be allotted a score of 0, 1 or 2; therefore, the highest score that can be achieved on the NLD Scale is 80 points. For this study, two of the questions were omitted and consequently only a maximum score of 76 could be attained. The AS group had the highest mean total score (M = 47.1, SD = 6.9) followed by the PDD-NOS group (M = 44.3, SD = 6.6), and the HFA group had the lowest mean total score (M = 42.8, SD = 6.9). However, an ANOVA revealed that differences between the groups on total NLD Scale scores were not significant [F (2,46) = 1.94; p = .16].

Subjects were also compared on subsections of the NLD Scale (see Table 5). When evaluated on the first section of the NLD Scale, composed of 23 questions on neuropsychological functioning, a significant group difference was found [F (2,46) = 3.13; p = .05]. Those with AS had the highest mean score on these items (M = 26.2, SD = 4.1), followed by individuals with PDD-NOS (M = 24.2, SD = 3.5), and finally, individuals with HFA had the lowest mean score on this section (M = 23.1, SD = 3.5). Post hoc tests confirmed that the difference between the AS and HFA groups was, indeed, significant.

In contrast, when evaluated on the second section of the NLD Scale, composed of 6 questions on academic achievement, no significant group differences were found [F (2,46) = .77; p = .47]. Individuals with AS had the highest mean score, reflecting a greater proportion of NLD characteristics (M = 8.9, SD = 2.2), followed by those with HFA (M = 8.5, SD = 2.1) and PDD-NOS (M = 7.9, SD = 2.0).

The third section of the NLD Scale is composed of 10 questions on psychosocial and adaptive functioning. However, a version of the NLD Scale that omitted questions number 33 and 34 was administered to 31 of the 49 individuals. Because of this, analysis of group differences was conducted on the 8 remaining questions. No significant group

Table 5

Analysis of the Performance of Individual's with Subtypes of Pervasive Developmental Disorder on the NLD Scale

Section of NLD Scale	PDD Group	M	SD	F (2,46)	p
Total Score on NLD Scale	HFA (n=16)	42.8	6.9		
	AS (n=21)	47.1	6.9	1.94	.16
	PDD-NOS (n=12)	44.3	6.6		
Neuropsychological Functioning (questions 1 to 23)	HFA (n=16)	23.1	3.5		
	AS (n=21)	26.2	4.1	3.13	.05*
	PDD-NOS (n=12)	24.2	3.5		
Academic Achievement (questions 24 to 30)	HFA (n=16)	8.5	2.1		
	AS (n=21)	8.9	2.2	.77	.47
	PDD-NOS (n=12)	7.9	2.0		
Social-Emotional and Adaptive Functioning (questions 31 to 40)	HFA (n=16)	11.1	3.6		
	AS (n=21)	12.0	2.5	.61	.55
	PDD-NOS (n=12)	12.2	2.6		

Note. * $p \leq 0.05$

differences were found [F (2,46) = .61; p = .55]. Individuals with PDD-NOS had a mean score of (M = 12.2, SD = 2.6), followed by the AS group (M = 12.0, SD = 2.5) and finally by the HFA group (M = 11.1, SD = 3.6).

In addition, performance of the three PDD groups on total and subsections of the NLD Scale were compared using Jonckheere's test of trend. No statistically significant linear trends were detected when comparing individuals who scored above the median with those who scored equal or below the median on these measures.

Responses on individual questions of the NLD Scale were compared among the three PDD groups using one-way between-groups ANOVAs (see Table 6). Significant findings were only revealed on questions 20 and 21. For Question 20, "Speaks more frequently and at greater length than others of his/her age," parents of children with AS responded "yes, very much so" significantly more often than did either parents of individuals with HFA or PDD-NOS [F (2,46) = 12.83; p < .01]. For Question 21, "Prefers to spend time talking or reading rather than engaging in physical activities," parents of children with AS responded "yes, very much so" significantly more often than the HFA group [F (2,46) = 3.35; p = .04].

When examining individual questions the trend wherein the AS group had the highest mean score, the PDD-NOS group scored in the middle, and the HFA group had the lowest mean score, and was only detected on only 12 of 38 questions. Although, when the scores obtained by the AS and PDD-NOS groups were averaged, they were found to be higher than mean scores achieved by the HFA group on 27 of the 38 questions. Similarly, when scores reported by the PDD-NOS and HFA groups were averaged, they were found to be lower than the scores attained by the AS group on 27 of the 38 questions.

Table 6

Analysis of PDD Group Differences on Individual Questions from the NLD Scale.

	HFA	M AS	PDD-NOS	F (2, 46)	p
Neuropsychological Functioning					
(1) is appropriately responsive to noises or sounds [1234]	1.38	1.43	1.42	0.03	0.97
(2) follows verbal commands	1.56	1.29	1.33	1.33	0.28
(3) is attentive to auditory-verbal input [234]	1.19	1.38	1.08	1.21	0.31
(4) easily remembers verbal material [1234]	1.13	1.52	1.25	1.67	0.20
(5) engages in simple, repetitive motoric activities	1.38	1.00	0.75	2.49	0.09
(6) explores objects by touch	1.06	0.67	0.83	1.12	0.34
(7) engages in visual exploration of environments [234]	0.50	0.57	0.58	0.09	0.92
(8) is attentive to visual stimuli [1234]	0.44	0.62	0.58	0.46	0.63
(9) remembers what he/she sees [234]	0.13	0.24	0.25	0.44	0.65
(10) engages in age-appropriate psychomotor activities [1234]	0.94	1.29	1.17	1.35	0.27
(11) relishes new environments; explores them actively [2]	1.06	1.00	1.42	1.26	0.30
(12) is eager to engage in new activities [234]	1.06	1.38	1.50	2.07	0.14
(13) seeks out and enjoys problem-solving activities [3]	1.44	1.43	1.42	<0.01	>0.99
(14) has age-appropriate understanding of concepts	1.13	0.95	1.25	0.62	0.55
(15) echoes (i.e., repeats verbatim) verbal messages [3]	1.00	0.86	0.42	1.93	0.16
(16) remembers what is said to him/her [234]	1.38	1.48	1.33	0.27	0.77
(17) exhibits age-appropriate pronunciation of words [1234]	0.19	0.38	0.25	0.53	0.59
(18) speaks out of turn; interrupts when others are trying to speak [34]	1.44	1.52	1.25	0.56	0.57
(19) has age-appropriate orientation to day of the week, time of day, and other dimensions of time [234]	0.19	0.38	0.42	0.66	0.52
(20) Speak more frequently and at greater length than others of his/her age? [1234]	0.50	1.71	0.92	12.83	<0.01*
(21) Prefer to spend time talking or reading rather than engaging in physical (including sporting) activities? [1234]	1.19	1.76	1.42	3.35	0.04**
(22) Prefer to interact with younger persons or adults rather than same-age children/adolescents? [1234]	1.38	1.81	1.58	2.22	0.12

Table 6 (continued)

Analysis of PDD Group Differences on Individual Questions from the NLD Scale

	M			F	p
	HFA	AS	PDD-NOS	(2, 46)	
(23) Shy away from new, novel, or complicated social events? [24]	1.50	1.52	1.75	0.67	0.52
Academic Achievement					
(24) exhibits neat (appropriate for age) handwriting [234]	1.13	1.48	1.17	0.58	0.57
(25) reads single words at or above age-expectation [24]	1.69	1.71	1.83	0.24	0.79
(26) spells words at or above age-expectation [1234]	1.38	1.52	1.42	0.14	0.87
(27) recalls (recites) academic material easily [234]	1.38	1.62	1.67	0.90	0.42
(28) comprehends reading material at or above age level [3]	1.19	0.81	0.42	3.08	0.06
(29) does age-appropriate arithmetic calculations [1234]	0.69	0.90	0.75	0.32	0.73
Social-Emotional and Adaptive Functioning					
(30) deals well with scientific concepts and reasoning [3]	0.88	0.81	0.67	0.27	0.76
(31) seeks out new friends and new experiences	1.69	1.57	1.75	0.42	0.66
(32) behaves appropriately with same-age children [234]	1.44	1.57	1.58	0.30	0.74
(33) prefers company of family members to that of others	-	-	-	-	-
(34) behaves appropriately with adults	-	-	-	-	-
(35) reacts with appropriate emotion in social situations [24]	1.38	1.38	1.58	0.56	0.58
(36) exhibits an age-appropriate level of general physical activity [1234]	1.19	1.48	1.25	0.98	0.38
(37) exhibits age-appropriate levels of skill in psychomotor (including sport) activities [1234]	1.31	1.62	1.50	1.13	0.33
(38) tends to play with younger children	1.44	1.14	0.92	0.69	0.51
(39) displays an age-appropriate range of emotional responsivity [2]	1.56	1.48	1.67	0.41	0.67
(40) is socially "popular" with age-mates [234]	1.69	1.81	1.92	0.88	0.42

Note. * $p < 0.01$; ** $p < 0.05$

[1] AS>PDD>HFA

[2] AS+PDD>HFA

[3] AS>PDD+HFA

[4] AS>IIFA

According to results of Jonckheere's test of trend, a trend such that the AS group scored the highest followed by the PDD-NOS group and finally the HFA group, was only statistically significant on three questions. These include questions 20, and 21, as detected by the one-way between-groups ANOVA's, and also question 22 ("Prefer to interact with younger persons or adults rather than same-age children/adolescents").

Study 2: Validity of the NLD Scale

Tests were conducted to see if subjects rated as having a high probability of being positive for NLD, according to neuropsychological profiles, would score higher on the NLD Scale than would those rated as having a low probability of displaying NLD (see Table 7). Individuals who have a probable chance of displaying the NLD syndrome scored significantly higher on the NLD Scale than did those who had a low or questionable chance of exhibiting NLD [F (2,46) = 9.24; $p < .01$]. Similar findings were found for scores on the neuropsychological functioning section of the NLD Scale [F (2,46) = 11.25; $p < .01$], but not for scores obtained on the academic achievement [F (2,46) = 2.56; $p = .12$] and social-emotional functioning [F (2,46) = 2.41; $p = .10$] sections.

When analyzing discrepancies between Verbal and Performance IQ scores, it was found that both the AS (M = 15.7, SD = 12.7) and the PDD-NOS (M = 18.3, SD = 12.2) groups had much greater discrepancies than the HFA group (M = 5.5, SD = 18.7). The difference in these IQ score discrepancies across groups was statistically significant [F (2,46) = 3.17; $p = .05$]. The majority of those with AS and PDD-NOS had higher verbal scores than performance scores. For example, it was found that 76.0% of individuals with AS and 75.0% of individuals with PDD-NOS had a VIQ greater than PIQ by at least 10 points, yet only 37.5% of individuals with HFA had a VIQ greater than PIQ by at least 10 points.

Table 7

Data Analysis Using One-Way ANOVAS to Investigate the Validity of the NLD Scale

Section of NLD Scale	Probability of Displaying NLD	M	SD	F (2,46)	p
Total Score on NLD Scale	Low (n=9)	37.4	5.5		
	Questionable (n= 28)	46.0	5.4	9.24	<.01*
	Probable (n= 12)	48.2	7.4		
Neuropsychological Functioning (questions 1 to 23)	Low (n=9)	20.2	3.2		
	Questionable (n= 28)	25.2	2.8	11.25	<.01*
	Probable (n= 12)	26.9	4.3		
Academic Achievement (questions 24 to 30)	Low (n=9)	7.2	.8		
	Questionable (n= 28)	8.9	2.3	2.26	.12
	Probable (n= 12)	8.7	1.8		
Social-Emotional and Adaptive Functioning (questions 31 to 40)	Low (n=9)	10.0	3.4		
	Questionable (n= 28)	12.0	2.6	2.41	.10
	Probable (n= 12)	12.6	2.8		

Note. * $p < 0.01$

Interestingly, an ANOVA revealed that subjects with a VIQ of at least ten points greater than their PIQ did not display higher scores on the NLD Scale [F (2,46) = .02; p = .88]. Similarly, there was no significant relationship between the discrepancies between VIQ and PIQ on any of the NLD Scale subsections (e.g., neuropsychological functioning [F (2,46) = .88; p = .35], academic achievement [F (2,46) = .16; p = .69], the social-emotional and adaptive behaviours [F (2,46) = .38; p = .54]). The NLD criterion, originally used by Pelletier et al. (2000), which states that VIQ should be greater that PIQ by ten points, did not predict a high score on the NLD Scale or its subtests. Therefore, the choice of excluding this criterion from the modified classification rules for NLD appears justified.

Adding the criterion of standard scores for reading (decoding) being at least 8 points greater than arithmetic on tests of achievement was problematic. Subjects were administered various tests of achievement (e.g., the K-TEA, WRAT- R, WRAT-3, WIAT, or WJ-R Tests of Achievement) at different dates, up to five years prior to their other neuropsychological tests. Also, 11 of the 49 subjects did not have scores from achievement tests recorded in their files. Therefore, this criterion may not add valuable information to the present validity determinations. When including achievement scores in NLD classification rules, subjects rated as having a high probability of being positive for NLD, still appeared to score higher on the NLD Scale than those rated as having a low or questionable chance of NLD. However, results did not reach a commonly accepted level of statistical significance for total NLD Scale scores [F (2,46) = 2.83; p = .07], scores on the academic achievement section [F (2,46) = 1.74; p = .19], or scores on the social-emotional functioning section [F (2,46) = .52; p = .60]. A statistically significant difference was found only on the first section of the scale, which consists of 23 questions based on neuropsychological functioning [F (2,46) = 3.73; p = .03].

Study 3: Reliability of the NLD Scale

Finally, adequacy of the NLD Scale's internal consistency was measured using the Cronbach's alpha test of reliability. The coefficient alpha for the NLD Scale was found to be 0.66.

Cronbach's alpha was also determined for each of the three subsections to assess the internal consistency of each domain of the scale. For the first section of the scale, consisting of 23 questions on neuropsychological functioning, a coefficient alpha of 0.36 was obtained. Item analysis of the academic achievement section resulted in a coefficient alpha of 0.06. Finally, when calculating the internal consistency for the third section (i.e., the 8 items on social-emotional and adaptive functioning that were answered by responders), there was a coefficient alpha of 0.74.

CHAPTER IV

DISCUSSION

Summary and Interpretation of Research Findings

The aim of the present study was twofold. The first goal was to evaluate the validity and internal consistency of the NLD Scale, because it is a relatively new instrument. Second, the NLD Scale was employed to examine its discriminating ability in respect to subtypes of PDD. Thus, the main question of interest was if there were behavioural differences between individuals with HFA, AS, and PDD-NOS that could be identified using the NLD Scale. It was hypothesized that those with AS would have the highest number of characteristics typically identifying those with NLD, followed by those with PDD-NOS, whereas those with HFA would have the least in common with the NLD syndrome. The main hypotheses of this investigation were tested using one-way between-groups ANOVAs and Jonckheere's test of trend.

Study 1: Performance of the PDD Subgroups on the NLD Scale

Although a carefully selected, relatively homogeneous group of individuals with PDD were chosen for this study, the use of specific and narrow diagnostic criteria resulted in a limited sample size. In reflection of the small sample size used, the power of each of the following analyses was generally low. The consequence of low statistical power is that results are prone to type II error. Nevertheless, several interesting findings emerged.

Comparison of Total Scores on the NLD Scale. The decision to include AS as a separate diagnostic category in the DSM-IV has been met with tremendous controversy due to its questionable validity outside of the autistic spectrum. Therefore, it is important to continue to look for differences between PDDs. In the first analysis of this study, the total NLD Scale scores achieved by each group were examined. Although a trend was

seen, such that the AS group scored the highest on mean total scores, followed by the PDD-NOS group and finally the HFA group, a significant difference was not revealed. Because the analysis of total NLD Scale scores did not differentiate the three PDD groups, further analyses were conducted with hopes of understanding which particular sections of the NLD Scale do or do not allow for a distinction between HFA, AS, and PDD-NOS.

Comparison of Scores from the Neuropsychological Functioning Section of the NLD Scale. It was hypothesized that the AS group would score highest on the first section of the NLD Scale, whereas the PDD-NOS group would score somewhat lower than the AS group, and finally the HFA group was expected to score the lowest. Results clearly indicate that individuals with HFA score lower on the NLD Scale than individuals with AS when compared on neuropsychological characteristics. This corresponds with past research that indicates that AS closely resembles Rourke's description of the NLD syndrome, whereas autism reflects a different neuropsychological profile (Gunter et al., unpublished; Klin et al., 1995; Rourke & Tsatsanis, 2000). Results confirm that use of strict diagnostic criteria, as applied by Klin and colleagues, is necessary to uncover differences between AS and HFA.

Despite this, no differences were found on neuropsychological characteristics between PDD-NOS and HFA, or PDD-NOS and AS. A lack of a robust overlap between characteristics of PDD-NOS and the NLD syndrome suggests that unlike AS, PDD-NOS does not appear to share a common phenotype with NLD. Further investigations are essential before it can be concluded that PDD-NOS and NLD are clinically dissimilar.

Comparison of Scores from the Academic Achievement Section of the NLD Scale.

Individuals with AS and PDD-NOS are thought to share academic strengths, for example,

linguistic function, and weaknesses, for example, visual-spatial functioning, therefore they were hypothesized to score similarly on the academic achievement section of the NLD Scale. HFA is not expected to have the same academic profile as individuals with NLD, and thus the HFA group was expected to score lower than persons with AS and PDD-NOS on this section of the scale. Reasons for formulating this hypothesis included knowledge of the differing profiles exhibited by groups of individuals with AS and HFA on intellectual scales, such as the Wechsler scales (Ehlers et al., 1997), and neuropsychological tests of language (Klin et al., 1995).

Despite the hypothesis formed, the NLD Scale was unable to discriminate between the PDD subtypes based on academic achievement. Due to the limited research on the academic functioning of persons with PDDs, it is difficult to draw any conclusions based on this finding. Nevertheless, there is a possibility the individuals with different types of PDD do not differ significantly in terms of scholastic functioning, because they all have difficulties on certain aspects of achievement such as comprehension. In fact, one study was found where subjects with HFA were found to perform in similar manner as those with NLD (Minshew, Goldstein, Taylor, & Siegal (1994). Specifically, they noted both HFA and NLD group were poor at comprehension but performed adequately on tests of reading, spelling and computation.

The NLD Scale was not able to distinguish PDDs based on academic functioning. However, the observation that the academic achievement section of the NLD Scale is only composed of six questions must not be ignored. This brief report of academic performance may not be sufficient to discriminate subtypes of PDD. Additional measures of scholastic achievement should be studied to decide if PDDs could be differentiated based on academics.

Comparison of Scores from the Social-Emotional and Adaptive Functioning Section of the NLD Scale. Social-emotional and adaptive functioning are clearly weaknesses found in all cases of PDD. Although some researchers have found that psychosocial functioning is somewhat better in individuals with AS and PDD-NOS than those with HFA (Bartolucci & Bremner, 1989; Njardvik et al., 1999; Sparrow et al., unpublished; Szatmari et al., 2000), others report little difference (e.g., Klin, 2000; Ozonoff et al., 1991). In general, it makes sense that there was no significant difference in scores on the NLD Scale subsection composed of mainly questions targeting social impairment. This is because what ties the PDDs together is the assumption that they all suffer from a serious social disability.

Comparison of Scores from Individual Questions of the NLD Scale. Due to small sample sizes the analyses of individual questions were generally inconclusive. Also, potential responses on these questions (e.g., "no"; "somewhat", and "yes, very much") lack a wide range of choice, and more differences were likely to have been detected if the NLD Scale required more specific responses (e.g., a scale of composed of 5 possible responses).

Findings did suggest that the AS group is reported to be more talkative than the HFA and PDD-NOS groups. Overall, poorer linguistic functioning demonstrated by the HFA group in comparison to the AS and PDD-NOS groups, corresponds with the DSM-IV definition of autism (American Psychiatric Association, 1994, 2000).

Even though the AS group was expected to have the highest mean score, followed by the PDD-NOS and then the HFA group on individual questions, this trend was only seen on 12 of 38 questions (and in fact a statistically significant trend was only found on questions 20, 21, and 22). A higher rate was found when the scores obtained by the AS and PDD-NOS group were combined and compared to the responses by the HFA group.

In this case the HFA group scored the lowest on 27 of the 38 questions. Correspondingly, when scores of the PDD-NOS and HFA groups were averaged, they were lower than the scores by the AS group on 27 of the 38 questions. Although of interest, these findings should be replicated using a larger sample size and greater range of response possibilities on questions, so that potential statistical significance can be determined.

Study 2: Validity of the NLD Scale

Results showed that there was a significant correlation between subjects who were NLD positive according to modified classification rules for NLD based on neuropsychological tests and total scores on the NLD Scale. This confirms that the NLD Scale is an effective tool for identifying characteristics of NLD. Validity tests of the first subsection of the NLD Scale, based on neuropsychological functioning, also produced a statistically significant group difference. Even though the sections on academic achievement and social-emotional functioning showed a similar trend, they did not reach a level of statistical significance. Therefore, in comparison to the academic and social sections, the neuropsychological functioning section of the scale best identifies characteristics of NLD. This makes conceptual sense, because the tests employed (e.g., WISC-III, WAIS-III, Purdue Pegboard, Finger Windows of the WRAML, and various achievement tests) evaluate domains of functioning most similar to functions questioned on the neuropsychological functioning section of the NLD Scale. Caution must be taken when interpreting scores on the academic achievement and social-emotional functioning subsections, as they are less indicative of a diagnosis of NLD.

It is frequently assumed that persons with a significantly greater Verbal IQ than Performance IQ are positive for NLD, but this is an overly simplistic view. A comparison between persons with and without significantly higher Verbal compared to Performance IQ scores revealed no significant differences on NLD Scale total scores. Therefore, this

supported the choice of excluding this criterion from NLD classification rules used during validity testing. Results of this analysis reinforces Rourke's (1989, 1995, 2000) notion that there are many criteria that must be taken into account before a diagnosis of NLD can be made.

The criterion stating that standard scores for reading (decoding) should be at least 8 points greater than arithmetic on tests of achievement was also not included in this study. This is because no one test of achievement was used throughout the sample. Also, 11 of the 49 subjects were not even assessed on an academic achievement test. When this criterion was supplemented, only the neuropsychological functioning section was found to identify characteristics of NLD. However, extreme caution must be taken before interpreting these results. Despite the lack of academic testing conducted for many of the subjects in this study, future studies investigating the validity of the NLD Scale should definitely include the criteria of reading being significantly better than arithmetic.

Inclusion of tests of tactile perception in the NLD classification rules used for this study would have been beneficial. According to Pelletier et al. (unpublished), no or very minimal simple tactile imperception and suppression compared to very poor finger agnosia, finger dysgraphesthesia, and astereognosis is characteristic of 90% of their sample of NLD subjects. Similarly, 66% of their sample had difficulties with the Tactual Performance Test (right, left, and both hand times become progressively worse in comparison to norms).

The Finger Windows subtest of the WRAML appears to be a good substitution for the Target Test when assessing non-verbal memory functions. However, the substitution of the Purdue Pegboard for the Grooved Pegboard must be discussed. Although Pelletier et al. (unpublished) utilized the Grooved Pegboard in their NLD classification criteria, due to the lack of availability of this test in the Yale Social Learning Disabilities Project

Neuropsychological Testing Battery, the present investigation instead relied on scores achieved on the Purdue Pegboard. The Grooved Pegboard test requires more complex visual-motor coordination than the Purdue Pegboard, because each peg must be turned into the proper position before it can fit into the board. Therefore, the Purdue Pegboard may not be as complex as the grooved pegboard task. Because individuals with NLD usually have a deficit in complex motor skills, rather than simple ones, substituting the Grooved Pegboard with the Purdue Pegboard may have resulted in the identification of fewer cases of NLD.

Study 3: Reliability of the NLD Scale

Cronbach's coefficient alpha was calculated to measure the internal consistency of the NLD Scale and also its subsections. A perfectly reliable measure would have a coefficient alpha of 1, and if there were no relation between items a coefficient of 0 would result. The NLD Scale in its entirety had an adequate internal consistency, represented by a coefficient alpha of 0.66.

The section on social-emotional and adaptive behaviours had good internal consistency (coefficient alpha = 0.74), because it is composed of many items measuring social interaction skills. Nevertheless, it also contains items that appear to extend beyond the domain of social interaction. For example, questions number 36 and 37 query the area of motor functioning; although it can be argued that levels of physical activity and clumsiness have a direct impact on social interactions and more specifically popularity among school children.

In contrast, the sections testing neuropsychological functioning (coefficient alpha = 0.36) and academic achievement, (coefficient alpha = 0.06) clearly contained items that were not measuring the same domain of functioning. For example, the section on neuropsychological functioning consists of item tapping into various aspects of

neuropsychological functioning including verbal and visual-spatial functioning, memory and attention, sensory perception, motor skills, problem-solving abilities, understanding of time, dealing with novelty and even social skills. Similarly, the section on academic achievement contains questions on a variety of scholastic behaviours for example, handwriting, spelling, single word reading, reading comprehension memory and arithmetic. In addition, the academic section was relatively small, as it consisted of merely six questions. This may have lowered the coefficient alpha even further.

Of importance is the notion that test-retest and interrater reliability are better measures of a test's value than internal consistency. It can be argued that Cronbach's alpha simply measures consistency amongst test items rather than allowing for a true appraisal of an instruments' reliability. Van der Vlugt's (unpublished) studies have demonstrated that the NLD Scale is undoubtedly a reliable and practical tool. Continued research and clinical use of this scale is supported.

Implications of Findings

There is presently no consistent evidence that neurobiological traits can clearly distinguish between subtypes of PDD. Thus, neuropsychological characteristics appear to be a more useful means of differentiating between these conditions. This body of research presents evidence that the neuropsychological properties characteristic of the syndrome of NLD is a means of distinguishing HFA from AS. Overall, the idea that HFA and AS are distinct syndromes with common overlapping phenotype (as described by Klin et al., 1995), appears to be the best way of making sense of these disorders.

It was hypothesized that individuals with PDD-NOS would display a number of characteristics seen in the NLD syndrome, although to a lesser extent than AS, because of the relatively milder severity of PDD-NOS compared to AS. Nevertheless it was suspected that PDD-NOS cases would display more behaviours characteristic of NLD

than those in the HFA group, due to conceivably differing neuropsychological strengths and weaknesses. Although a trend in this direction was noted when comparing NLD total scores, results were not statistically significant. Overall, the present investigation did not find that PDD-NOS could be distinguished from either AS or HFA using the NLD Scale. Lack of significance is theorized to result from the small sample size used. Nevertheless, a closer look must be taken at the DSM-IV and ICD-10 definitions of PDD-NOS (or atypical autism).

The researchers who diagnosed individuals into the three PDD groups used in this study were, to say the least, extremely competent, and based their decisions on strict criteria. Nevertheless, whereas DSM-IV and ICD-10 may adequately be used to diagnose lower functioning patients with autistic disorder, this task is more difficult when differentiating between HFA, AS, and PDD-NOS. One reason for this difficulty is that PDD-NOS is not even operationally defined in the DSM-IV. This renders it especially difficult to discriminate PDD-NOS groups from other disorders when relying on either the ICD-10 or DSM-IV (Buitelaar, Van der Gaag, Klin, & Volkmar, 1999; Mahoney et al., 1998). Therefore, it is highly recommended that the vague descriptions of PDD-NOS in the ICD-10 and DSM-IV be refined to make classification of PDDs less challenging.

At present there is little research using the NLD Scale. It appears to be a highly useful tool, and studies such as the present investigation, may allow for widespread use of this tool in the future. At the moment, the NLD Scale is an effective screener of the NLD syndrome, although some modifications may allow for improvement in the detection of NLD.

Limitations and Future Directions

The present study analyzed the results obtained from the NLD Scale; however, the major limitation of this study is the small sample size. In the future, larger samples

should be used to differentiate groups more clearly.

Past studies comparing groups of PDD which were written before 1994 are at a clear disadvantage, because AS did not become a separate category in the DSM until the publication of the fourth edition. The present study used a well-characterized sample of high-functioning individuals of autism, AS and PDD-NOS. Specific and reliable diagnostic criteria were employed, and individuals were well-screened for medical and psychiatric conditions. The groups were also well-matched in terms of gender and IQ, although individuals with HFA had a significantly higher mean age. Because sample size was already relatively small, an attempt to match the groups more closely in age was not made. This was not thought to have a significant effect on the response style exhibited by this group on the NLD Scale. Nevertheless, future studies should attempt to match groups on age as there is a possibility that children with HFA may possibly vary in their behaviour, language and social skills over time. A study by Church and Coplan (1995) showed that children with HFA have a milder symptom profile as they approach teen years.

PDDs are highly heterogeneous, and may be associated with differing etiological mechanisms and comorbid symptoms. The present study was limited to only high-functioning males. Future research should continue its efforts in distinguishing between HFA, AS, and PDD-NOS, although it is important to consider using a larger and wider ranging sample in order to elucidate importance differences that may exist between these groups. For example, to permit further generalization it would be worthwhile to repeat the present investigation using females, older adults, low-functioning individuals with PDDs, various populations of individuals with a learning disability and of course normal control subjects. Use of such populations would help clarify where the normal spectrum ends and where pathology begins.

In addition to sample size and subject selection, there are limitations associated with the use of the NLD Scale. It is acknowledged that the NLD Scale is a new instrument that may require some updates before it is established as a widely administered instrument. Cut-off scores should be included so that the scale could be more useful in determining if a person is either positive or negative for displaying the syndrome of NLD. Also, this brief parental report of functioning, in particular the academic achievement section, which consists of merely six questions, may not be sufficient to characterize PDDs. Even though this instrument was not specifically designed to differentiate PDDs, supplementing the scale with more questions may also enhance its ability to distinguish between individuals with and without the syndrome of NLD. To make the NLD Scale a more effective tool, the range of possible responses may be increased from three to perhaps four or five responses (e.g., "no, never", "rarely", "yes, regularly", and "yes, always"). In addition, some modifications can be made to the language used on this scale so that parents more easily understand it. For example, terminology such as, "motoric activities" or "auditory-verbal input" could be avoided.

It is known that many individuals with AS share a phenotype with NLD, but not all individuals with NLD exhibit the signs of AS. This study is only considered to represent an initial step towards understanding how NLD overlaps with HFA, AS, and PDD-NOS. Clearly, research supports Rourke's (1989, 1995) theoretical framework suggesting that the NLD model can account for the apparently unconnected deficits characteristic of AS. For example, neuropsychological functions, which may include psychomotor functioning, visual-spatial organization, and verbal skills, appear more similar in AS and NLD compared to HFA and NLD. Furthermore, Rourke's white matter model may provide a comprehensive mechanism for understanding AS, because of the possibility that white matter tracts can be differentially affected and thus result in a wide

range of deficits, as exhibited by AS. A more detailed understanding of the relationship between NLD and PDDs cannot be provided until further neuropsychological research is conducted.

It may be troublesome to establish the distinction between subtypes of PDD without confirmation from accurate knowledge of etiology and pathogenesis of these disorders. Despite the fact that the present investigation supports the view that individuals with AS display virtually all of the assets and deficits of NLD (e.g., Klin et al., 1995; Rourke, 1995; Rourke & Tsatsanis, 2000), at the present time the association between AS and the white matter model of brain damage proposed by Rourke (1995) requires further analysis. Similarly, evidence of right hemisphere dysfunction in AS, as opposed to left hemisphere dysfunction in HFA is relatively sparse. This is even truer of PDD-NOS, where neurobiological literature is practically non-existent. Therefore, further neurobiological research on the similarities and differences between NLD and subtypes of PDD is strongly encouraged. Similarly, studies on the developmental progression of these disorders may also contribute to a better understanding of how these syndromes may overlap, and how they differ. Research in this area would be especially useful if conducted with infants and preschoolers, to understand if observed differences between those with HFA, AS, and PDD-NOS are due to varying levels of ability in language and IQ, or if discrepancies result from distinct developmental pathways.

A review of the literature clearly alerts us to the fact that PDD-NOS research is sparse. The lack of a clear definition suggests that researchers employ the concept of PDD-NOS inconsistently. Well-validated rules used to identify those with PDD-NOS need to be made universal. Following this, it will be crucial to research the neuropsychological strengths of individuals with PDD-NOS, and to compare their profiles with those found for HFA, AS and control subjects.

Distinguishing subtypes of PDD allows for individuals with different strengths and weaknesses to take advantage of opportunities based on their specific needs. Therefore, such knowledge may permit the enhancement of current educational and treatment programs.

Conclusions

Since Wing's (1981) publication of 19 case reports of children with AS, there has been a growing body of research dedicated to either placing AS along a spectrum of autistic disorders, or distinguishing these syndromes. With the definition of AS, now mutually exclusive from autism in both the DSM-IV and ICD-10, the task of understanding these disorders as separate conditions has been somewhat less complicated. Despite this, a definition of PDD-NOS is still in the early stages of development, and so understanding its similarities and differences from the other PDDs is still a complex undertaking.

Collectively, PDDs are accepted to be neurodevelopmental disorders, although an exhaustive account of the neurobiology of these conditions is lacking. The central issue at hand is whether HFA, AS, and PDD-NOS are distinctive in ways that are independent of the definitions used to assign group membership. The present study relied on neuropsychological characteristics to help distinguish these conditions. More specifically, this paper described the use of the NLD Scale, an instrument used to aid diagnosis of individuals with NLD, and then its potential utility to differentiate different subtypes of PDD.

The psychometric properties of the NLD Scale appear adequate although additional investigation is required. Also, modifications to this tool, such as the inclusion of supplementary questions and cut-off scores, may improve its clinical utility.

Results show that AS can be differentiated from HFA by means of this scale,

although only based on neuropsychological characteristics. Measures of academic functioning and social-emotional functioning are less apt to differentiate these groups. Individuals with PDD-NOS do not appear to be easily differentiated from either HFA or AS. Perhaps the development of a valid operational definition of PDD-NOS would help clarify the distinction between these groups.

Although ongoing study should include larger and more widespread populations, the positive results of this study suggest a need to examine the use of the NLD Scale more closely. Furthermore, continued neuropsychological research aimed at examining the similarities and differences of subtypes of PDD is encouraged in order to improve the understanding of these disorders.

APPENDIX

NLD Scale (Byron P. Rourke, University of Windsor & Yale University)

Instructions: Please answer each question in terms of your experience with your child. Of interest is whether and to what extent a behaviour occurs regularly or typically, NOT whether and to what extent the child is capable of exhibiting the behaviour. In other words, this section deals with what the child DOES regularly or habitually, rather than what the child is capable of doing but does not typically do in everyday life.

Three point-scale: All of the questions are to be answered on a three-point scale, as follows: (I) No, not at all, never,
(II) Somewhat; every once in a while,
(III) Yes, very much, frequently.

Your Child	No	Somewhat	Yes, Very Much
Neuropsychological Functioning			
(1) is appropriately responsive to noises or sounds	-	-	-
(2) follows verbal commands	-	-	-
(3) is attentive to auditory-verbal input	-	-	-
(4) easily remembers verbal material	-	-	-
(5) engages in simple, repetitive motoric activities	-	-	-
(6) explores objects by touch	-	-	-
(7) engages in visual exploration of environments	-	-	-
(8) is attentive to visual stimuli	-	-	-
(9) remembers what he/she sees	-	-	-
(10) engages in age-appropriate psychomotor activities	-	-	-
(11) relishes new environments; explores them actively	-	-	-
(12) is eager to engage in new activities	-	-	-
(13) seeks out and enjoys problem-solving activities	-	-	-

(14) has age-appropriate understanding of concepts - - -

(15) echoes (i.e., repeats verbatim) verbal messages - - -

(16) remembers what is said to him/her - - -

(17) exhibits age-appropriate pronunciation of words - - -

(18) speaks out of turn; interrupts when others are trying to speak - - -

(19) has age-appropriate orientation to day of the week, time of day, and other dimensions of time - - -

(20) Speak more frequently and at greater length than others of his/her age? - - -

(21) Prefer to spend time talking or reading rather than engaging in physical (including sporting) activities? - - -

(22) Prefer to interact with younger persons or adults rather than same-age children/adolescents? - - -

(23) Shy away from new, novel, or complicated social events? - - -

Academic Achievement

(24) exhibits neat (appropriate for age) handwriting - - -

(25) reads single words at or above age-expectation - - -

(26) spells words at or above age-expectation - - -

(27) recalls (recites) academic material easily - - -

(28) comprehends reading material at or above age level - - -

(29) does age-appropriate arithmetic calculations - - -

Social-Emotional and Adaptive Functioning

(30) deals well with scientific concepts and reasoning - - -

(31) seeks out new friends and new experiences - - -

(32) behaves appropriately with same-age children	-	-	-
(33) prefers company of family members to that of others	-	-	-
(34) behaves appropriately with adults	-	-	-
(35) reacts with appropriate emotion in social situations	-	-	-
(36) exhibits an age-appropriate level of general physical activity	-	-	-
(37) exhibits age-appropriate levels of skill in psychomotor (including sport) activities	-	-	-
(38) tends to play with younger children	-	-	-
(39) displays an age-appropriate range of emotional responsivity	-	-	-
(40) is socially "popular" with age-mates	-	-	-

NLD Scale - Scoring

In order to proceed with the assessment of the reliability and validity of the scale, it is necessary to have a clear scoring system available. The one that we are using now is as follows:

(a) Each item is scored on a scale of 0, 1, and 2, corresponding to the three response categories.

(b) This yields a total possible score of 0 to 80; the higher the score, the more likely that the child/adolescent exhibits NLD.

(c) The items have been arranged in such a manner that the direction of the scoring needs to be adjusted in terms of the criterion just above (b).

The following (18) items are those where a response of yes, very much yields a score of 2; yes, a little, 1; and, no, not at all, 0:

1, 2, 3, 4, 5, 15, 16, 18, 20, 21, 22, 23, 25, 26, 27, 33, 34, 38.

The remainder (22) of the items are those where a response of no, not at all yields a score of 2; yes, a little, 1; and, yes, very much, 0.

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VITA AUCTORIS

Chand Taneja was born on December 17, 1974 in Victoria, British Columbia. Her interest in Pervasive Developmental Disorders was stimulated when she did volunteer work at Queen Alexandria Centre for Children's Health (1995-1998; Summer 1999). Her work with children with autism and Asperger's syndrome continued when employed by the Laurel Group (1997-1998), and the B.C. Ministry of Child and Families (Summer 1999). She graduated with distinction in 1996, receiving her Bachelor of Science degree in Biology and Psychology from the University of Victoria. Following this she enrolled in the graduate program in Clinical Neuropsychology at the University of Windsor, in Windsor, Ontario. She continues to pursue her doctoral degree in Clinical Neuropsychology at the University of Windsor.